(up to 10 years from the start of the experiment or demonstration project).

[48 FR 7575, Feb. 23, 1983, as amended at 52 FR 37605, Oct. 8, 1987; 55 FR 51687, Dec. 17, 1990; 62 FR 38451, July 18, 1997]

APPENDIX 1 TO SUBPART P OF PART 404—LISTING OF IMPAIRMENTS

The body system listings in parts A and B of the Listing of Impairments will no longer be effective on the following dates unless extended by the Commissioner or revised and promulgated again.

1. Low Birth Weight and Failure to Thrive (100.00): June 12, 2020.
2. Musculoskeletal System (1.00 and 101.00): January 27, 2017.
3. Special Senses and Speech (2.00 and 102.00): April 29, 2018.
4. Respiratory System (3.00 and 103.00): January 27, 2017.
5. Cardiovascular System (4.00 and 104.00): January 27, 2017.
6. Digestive System (5.00 and 105.00): January 27, 2017.
7. Genitourinary Disorders (6.00 and 106.00): December 9, 2019.
8. Hematological Disorders (7.00 and 107.00): May 18, 2020.
9. Skin Disorders (8.00 and 108.00): January 27, 2017.
10. Endocrine Disorders (9.00 and 109.00): June 7, 2016.
11. Congenital Disorders That Affect Multiple Body Systems (10.00 and 110.00): April 5, 2018.
12. Neurological (11.00 and 111.00): January 27, 2017.
13. Mental Disorders (12.00 and 112.00): January 27, 2017.
14. Cancer (Malignant Neoplastic Diseases) (13.00 and 113.00): July 20, 2020.
15. Immune System Disorders (14.00 and 114.00): June 16, 2016.

Part A

Criteria applicable to individuals age 18 and over and to children under age 18 where criteria are appropriate.

Sec.

1.00 MUSCULOSKELETAL SYSTEM

A. *Disorders of the musculoskeletal system* may result from hereditary, congenital, or acquired pathologic processes. Impairments may result from infectious, inflammatory, or degenerative processes, traumatic or developmental events, or neoplastic, vascular, or toxic/metabolic diseases.

B. *Loss of function.*

1. *General.* Under this section, loss of function may be due to bone or joint deformity or destruction from any cause; miscellaneous disorders of the spine with or without radiculopathy or other neurological deficits; amputation; or fractures or soft tissue injuries, including burns, requiring prolonged periods of immobility or convalescence. The provisions of 1.02 and 1.03 notwithstanding, inflammatory arthritis is evaluated under 14.09 (see 14.00D6). Impairments with neurological causes are to be evaluated under 11.00ff.

2. How We Define Loss of Function in These Listings

a. *General.* Regardless of the cause(s) of a musculoskeletal impairment, functional loss for purposes of these listings is defined as the inability to ambulate effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment, or the inability to perform fine and gross movements effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment. The inability to ambulate effectively or the inability to perform fine and gross movements effectively must have lasted, or be expected to last, for at least 12 months. For the purposes of these criteria, consideration of the ability to perform these activities must be from a physical standpoint alone. When there is an inability to perform these activities due to a mental impairment, the criteria in 12.00ff are to be used. We will determine whether an individual can ambulate effectively or can perform fine and gross movements effectively based on the medical and other evidence in the case record, generally without developing additional evidence about the individual's ability to perform the specific activities listed as examples in 1.00B2b(2) and 1.00B2c.

b. What We Mean by Inability To Ambulate Effectively

(1) *Definition.* Inability to ambulate effectively means an extreme limitation of the ability to walk; *i.e.*, an impairment(s) that

interferes very seriously with the individual's ability to independently initiate, sustain, or complete activities. Ineffective ambulation is defined generally as having insufficient lower extremity functioning (see 1.00J) to permit independent ambulation without the use of a hand-held assistive device(s) that limits the functioning of both upper extremities. (Listing 1.05C is an exception to this general definition because the individual has the use of only one upper extremity due to amputation of a hand.)

(2) *To ambulate effectively*, individuals must be capable of sustaining a reasonable walking pace over a sufficient distance to be able to carry out activities of daily living. They must have the ability to travel without companion assistance to and from a place of employment or school. Therefore, examples of ineffective ambulation include, but are not limited to, the inability to walk without the use of a walker, two crutches or two canes, the inability to walk a block at a reasonable pace on rough or uneven surfaces, the inability to use standard public transportation, the inability to carry out routine ambulatory activities, such as shopping and banking, and the inability to climb a few steps at a reasonable pace with the use of a single hand rail. The ability to walk independently about one's home without the use of assistive devices does not, in and of itself, constitute effective ambulation.

c. *What we mean by inability to perform fine and gross movements effectively.* Inability to perform fine and gross movements effectively means an extreme loss of function of both upper extremities; *i.e.*, an impairment(s) that interferes very seriously with the individual's ability to independently initiate, sustain, or complete activities. To use their upper extremities effectively, individuals must be capable of sustaining such functions as reaching, pushing, pulling, grasping, and fingering to be able to carry out activities of daily living. Therefore, examples of inability to perform fine and gross movements effectively include, but are not limited to, the inability to prepare a simple meal and feed oneself, the inability to take care of personal hygiene, the inability to sort and handle papers or files, and the inability to place files in a file cabinet at or above waist level.

d. *Pain or other symptoms.* Pain or other symptoms may be an important factor contributing to functional loss. In order for pain or other symptoms to be found to affect an individual's ability to perform basic work activities, medical signs or laboratory findings must show the existence of a medically determinable impairment(s) that could reasonably be expected to produce the pain or other symptoms. The musculoskeletal listings that include pain or other symptoms among their criteria also include criteria for limitations in functioning as a result of the listed impairment, including limitations caused by pain. It is, therefore, important to evaluate the intensity and persistence of such pain or other symptoms carefully in order to determine their impact on the individual's functioning under these listings. See also §§ 404.1525(f) and 404.1529 of this part, and §§ 416.925(f) and 416.929 of part 416 of this chapter.

C. Diagnosis and Evaluation

1. *General.* Diagnosis and evaluation of musculoskeletal impairments should be supported, as applicable, by detailed descriptions of the joints, including ranges of motion, condition of the musculature (e.g., weakness, atrophy), sensory or reflex changes, circulatory deficits, and laboratory findings, including findings on x-ray or other appropriate medically acceptable imaging. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Purchase of certain medically acceptable imaging.* While any appropriate medically acceptable imaging is useful in establishing the diagnosis of musculoskeletal impairments, some tests, such as CAT scans and MRIs, are quite expensive, and we will not routinely purchase them. Some, such as myelograms, are invasive and may involve significant risk. We will not order such tests. However, when the results of any of these tests are part of the existing evidence in the case record we will consider them together with the other relevant evidence.

3. *Consideration of electrodiagnostic procedures.* Electrodiagnostic procedures may be useful in establishing the clinical diagnosis, but do not constitute alternative criteria to the requirements of 1.04.

D. *The physical examination* must include a detailed description of the rheumatological, orthopedic, neurological, and other findings appropriate to the specific impairment being evaluated. These physical findings must be determined on the basis of objective observation during the examination and not simply a report of the individual's allegation; e.g., ''He says his leg is weak, numb.'' Alternative testing methods should be used to verify the abnormal findings; e.g., a seated straight-leg raising test in addition to a supine straight-leg raising test. Because abnormal physical findings may be intermittent, their presence over a period of time must be established by a record of ongoing management and evaluation. Care must be taken to ascertain that the reported examination findings are consistent with the individual's daily activities.

E. Examination of the Spine

1. *General.* Examination of the spine should include a detailed description of gait, range of motion of the spine given quantitatively in degrees from the vertical position (zero degrees) or, for straight-leg raising from the sitting and supine position (zero degrees), any other appropriate tension signs, motor and sensory abnormalities, muscle spasm, when present, and deep tendon reflexes. Observations of the individual during the examination should be reported; e.g., how he or she gets on and off the examination table. Inability to walk on the heels or toes, to squat, or to arise from a squatting position, when appropriate, may be considered evidence of significant motor loss. However, a report of atrophy is not acceptable as evidence of significant motor loss without circumferential measurements of both thighs and lower legs, or both upper and lower arms, as appropriate, at a stated point above and below the knee or elbow given in inches or centimeters. Additionally, a report of atrophy should be accompanied by measurement of the strength of the muscle(s) in question generally based on a grading system of 0 to 5, with 0 being complete loss of strength and 5 being maximum strength. A specific description of atrophy of hand muscles is acceptable without measurements of atrophy but should include measurements of grip and pinch strength.

2. *When neurological abnormalities persist.* Neurological abnormalities may not completely subside after treatment or with the passage of time. Therefore, residual neurological abnormalities that persist after it has been determined clinically or by direct surgical or other observation that the ongoing or progressive condition is no longer present will not satisfy the required findings in 1.04. More serious neurological deficits (paraparesis, paraplegia) are to be evaluated under the criteria in 11.00ff.

F. *Major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

G. *Measurements of joint motion* are based on the techniques described in the chapter on the extremities, spine, and pelvis in the current edition of the ''Guides to the Evaluation of Permanent Impairment'' published by the American Medical Association.

H. Documentation

1. *General.* Musculoskeletal impairments frequently improve with time or respond to treatment. Therefore, a longitudinal clinical record is generally important for the assessment of severity and expected duration of an impairment unless the claim can be decided favorably on the basis of the current evidence.

2. *Documentation of medically prescribed treatment and response.* Many individuals, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever evidence of such treatment is available it must be considered.

3. *When there is no record of ongoing treatment.* Some individuals will not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In such cases, evaluation will be made on the basis of the current objective medical evidence and other available evidence, taking into consideration the individual's medical history, symptoms, and medical source opinions. Even though an individual who does not receive treatment may not be able to show an impairment that meets the criteria of one of the musculoskeletal listings, the individual may have an impairment(s) equivalent in severity to one of the listed impairments or be disabled based on consideration of his or her residual functional capacity (RFC) and age, education and work experience.

4. *Evaluation when the criteria of a musculoskeletal listing are not met.* These listings are only examples of common musculoskeletal disorders that are severe enough to prevent a person from engaging in gainful activity. Therefore, in any case in which an individual has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will consider medical equivalence. (See §§ 404.1526 and 416.926.) Individuals who have an impairment(s) with a level of severity that does not meet or equal the criteria of the musculoskeletal listings may or may not have the RFC that would enable them to engage in substantial gainful activity. Evaluation of the impairment(s) of these individuals should proceed through the final steps of the sequential evaluation process in §§ 404.1520 and 416.920 (or, as appropriate, the steps in the medical improvement review standard in §§ 404.1594 and 416.994).

I. Effects of Treatment

1. *General.* Treatments for musculoskeletal disorders may have beneficial effects or adverse side effects. Therefore, medical treatment (including surgical treatment) must be

considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the disorder, and in terms of any side effects that may further limit the individual.

2. *Response to treatment.* Response to treatment and adverse consequences of treatment may vary widely. For example, a pain medication may relieve an individual's pain completely, partially, or not at all. It may also result in adverse effects, e.g., drowsiness, dizziness, or disorientation, that compromise the individual's ability to function. Therefore, each case must be considered on an individual basis, and include consideration of the effects of treatment on the individual's ability to function.

3. *Documentation.* A specific description of the drugs or treatment given (including surgery), dosage, frequency of administration, and a description of the complications or response to treatment should be obtained. The effects of treatment may be temporary or long-term. As such, the finding regarding the impact of treatment must be based on a sufficient period of treatment to permit proper consideration or judgment about future functioning.

J. Orthotic, Prosthetic, or Assistive Devices

1. *General.* Consistent with clinical practice, individuals with musculoskeletal impairments may be examined with and without the use of any orthotic, prosthetic, or assistive devices as explained in this section.

2. *Orthotic devices.* Examination should be with the orthotic device in place and should include an evaluation of the individual's maximum ability to function effectively with the orthosis. It is unnecessary to routinely evaluate the individual's ability to function without the orthosis in place. If the individual has difficulty with, or is unable to use, the orthotic device, the medical basis for the difficulty should be documented. In such cases, if the impairment involves a lower extremity or extremities, the examination should include information on the individual's ability to ambulate effectively without the device in place unless contraindicated by the medical judgment of a physician who has treated or examined the individual.

3. *Prosthetic devices.* Examination should be with the prosthetic device in place. In amputations involving a lower extremity or extremities, it is unnecessary to evaluate the individual's ability to walk without the prosthesis in place. However, the individual's medical ability to use a prosthesis to ambulate effectively, as defined in 1.00B2b, should be evaluated. The condition of the stump should be evaluated without the prosthesis in place.

4. *Hand-held assistive devices.* When an individual with an impairment involving a lower extremity or extremities uses a hand-held assistive device, such as a cane, crutch or walker, examination should be with and without the use of the assistive device unless contraindicated by the medical judgment of a physician who has treated or examined the individual. The individual's ability to ambulate with and without the device provides information as to whether, or the extent to which, the individual is able to ambulate without assistance. The medical basis for the use of any assistive device (e.g., instability, weakness) should be documented. The requirement to use a hand-held assistive device may also impact on the individual's functional capacity by virtue of the fact that one or both upper extremities are not available for such activities as lifting, carrying, pushing, and pulling.

K. *Disorders of the spine*, listed in 1.04, result in limitations because of distortion of the bony and ligamentous architecture of the spine and associated impingement on nerve roots (including the cauda equina) or spinal cord. Such impingement on nerve tissue may result from a herniated nucleus pulposus, spinal stenosis, arachnoiditis, or other miscellaneous conditions. Neurological abnormalities resulting from these disorders are to be evaluated by referral to the neurological listings in 11.00ff, as appropriate. (See also 1.00B and E.)

1. *Herniated nucleus pulposus* is a disorder frequently associated with the impingement of a nerve root. Nerve root compression results in a specific neuro-anatomic distribution of symptoms and signs depending upon the nerve root(s) compromised.

2. Spinal Arachnoiditis

a. *General.* Spinal arachnoiditis is a condition characterized by adhesive thickening of the arachnoid which may cause intermittent ill-defined burning pain and sensory dysesthesia, and may cause neurogenic bladder or bowel incontinence when the cauda equina is involved.

b. *Documentation.* Although the cause of spinal arachnoiditis is not always clear, it may be associated with chronic compression or irritation of nerve roots (including the cauda equina) or the spinal cord. For example, there may be evidence of spinal stenosis, or a history of spinal trauma or meningitis. Diagnosis must be confirmed at the time of surgery by gross description, microscopic examination of biopsied tissue, or by findings on appropriate medically acceptable imaging. Arachnoiditis is sometimes used as a diagnosis when such a diagnosis is unsupported by clinical or laboratory findings. Therefore, care must be taken to ensure that the diagnosis is documented as described in 1.04B. Individuals with arachnoiditis, particularly when it involves the lumbosacral spine, are generally unable to sustain any given position or posture for more than a short period of time due to pain.

3. *Lumbar spinal stenosis* is a condition that may occur in association with degenerative processes, or as a result of a congenital anomaly or trauma, or in association with Paget's disease of the bone. *Pseudoclaudication,* which may result from lumbar spinal stenosis, is manifested as pain and weakness, and may impair ambulation. Symptoms are usually bilateral, in the low back, buttocks, or thighs, although some individuals may experience only leg pain and, in a few cases, the leg pain may be unilateral. The pain generally does not follow a particular neuro-anatomical distribution, *i.e.*, it is distinctly different from the radicular type of pain seen with a herniated intervertebral disc, is often of a dull, aching quality, which may be described as ''discomfort'' or an ''unpleasant sensation,'' or may be of even greater severity, usually in the low back and radiating into the buttocks region bilaterally. The pain is provoked by extension of the spine, as in walking or merely standing, but is reduced by leaning forward. The distance the individual has to walk before the pain comes on may vary. Pseudoclaudication differs from peripheral vascular claudication in several ways. Pedal pulses and Doppler examinations are unaffected by pseudoclaudication. Leg pain resulting from peripheral vascular claudication involves the calves, and the leg pain in vascular claudication is ordinarily more severe than any back pain that may also be present. An individual with vascular claudication will experience pain after walking the same distance time after time, and the pain will be relieved quickly when walking stops.

4. *Other miscellaneous conditions* that may cause weakness of the lower extremities, sensory changes, areflexia, trophic ulceration, bladder or bowel incontinence, and that should be evaluated under 1.04 include, but are not limited to, osteoarthritis, degenerative disc disease, facet arthritis, and vertebral fracture. Disorders such as spinal dysrhaphism (e.g., spina bifida), diastematomyelia, and tethered cord syndrome may also cause such abnormalities. In these cases, there may be gait difficulty and deformity of the lower extremities based on neurological abnormalities, and the neurological effects are to be evaluated under the criteria in 11.00ff.

L. *Abnormal curvatures of the spine.* Abnormal curvatures of the spine (specifically, scoliosis, kyphosis and kyphoscoliosis) can result in impaired ambulation, but may also adversely affect functioning in body systems other than the musculoskeletal system. For example, an individual's ability to breathe may be affected; there may be cardiac difficulties (e.g., impaired myocardial function); or there may be disfigurement resulting in withdrawal or isolation. When there is impaired ambulation, evaluation of equivalence may be made by reference to 14.09A. When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 14.09C. When there is respiratory or cardiac involvement or an associated mental disorder, evaluation may be made under 3.00ff, 4.00ff, or 12.00ff, as appropriate. Other consequences should be evaluated according to the listing for the affected body system.

M. *Under continuing surgical management*, as used in 1.07 and 1.08, refers to surgical procedures and any other associated treatments related to the efforts directed toward the salvage or restoration of functional use of the affected part. It may include such factors as post-surgical procedures, surgical complications, infections, or other medical complications, related illnesses, or related treatments that delay the individual's attainment of maximum benefit from therapy. When burns are not under continuing surgical management, see 8.00F.

N. *After maximum benefit from therapy has been achieved* in situations involving fractures of an upper extremity (1.07), or soft tissue injuries (1.08), *i.e.*, there have been no significant changes in physical findings or on appropriate medically acceptable imaging for any 6-month period after the last definitive surgical procedure or other medical intervention, evaluation must be made on the basis of the demonstrable residuals, if any. A finding that 1.07 or 1.08 is met must be based on a consideration of the symptoms, signs, and laboratory findings associated with recent or anticipated surgical procedures and the resulting recuperative periods, including any related medical complications, such as infections, illnesses, and therapies which impede or delay the efforts toward restoration of function. Generally, when there has been no surgical or medical intervention for 6 months after the last definitive surgical procedure, it can be concluded that maximum therapeutic benefit has been reached. Evaluation at this point must be made on the basis of the demonstrable residual limitations, if any, considering the individual's impairment-related symptoms, signs, and laboratory findings, any residual symptoms, signs, and laboratory findings associated with such surgeries, complications, and recuperative periods, and other relevant evidence.

O. *Major function of the face and head,* for purposes of listing 1.08, relates to impact on any or all of the activities involving vision, hearing, speech, mastication, and the initiation of the digestive process.

P. *When surgical procedures have been performed,* documentation should include a copy of the operative notes and available pathology reports.

Q. *Effects of obesity.* Obesity is a medically determinable impairment that is often associated with disturbance of the musculoskeletal system, and disturbance of this system can be a major cause of disability in individuals with obesity. The combined effects of obesity with musculoskeletal impairments can be greater than the effects of each of the impairments considered separately. Therefore, when determining whether an individual with obesity has a listing-level impairment or combination of impairments, and when assessing a claim at other steps of the sequential evaluation process, including when assessing an individual's residual functional capacity, adjudicators must consider any additional and cumulative effects of obesity.

1.01 Category of Impairments, Musculoskeletal

1.02 *Major dysfunction of a joint(s) (due to any cause):* Characterized by gross anatomical deformity (e.g., subluxation, contracture, bony or fibrous ankylosis, instability) and chronic joint pain and stiffness with signs of limitation of motion or other abnormal motion of the affected joint(s), and findings on appropriate medically acceptable imaging of joint space narrowing, bony destruction, or ankylosis of the affected joint(s). With:

A. Involvement of one major peripheral weight-bearing joint (*i.e.*, hip, knee, or ankle), resulting in inability to ambulate effectively, as defined in 1.00B2b;

or

B. Involvement of one major peripheral joint in each upper extremity (*i.e.*, shoulder, elbow, or wrist-hand), resulting in inability to perform fine and gross movements effectively, as defined in 1.00B2c.

1.03 *Reconstructive surgery or surgical arthrodesis of a major weight-bearing joint,* with inability to ambulate effectively, as defined in 1.00B2b, and return to effective ambulation did not occur, or is not expected to occur, within 12 months of onset.

1.04 *Disorders of the spine* (e.g., herniated nucleus pulposus, spinal arachnoiditis, spinal stenosis, osteoarthritis, degenerative disc disease, facet arthritis, vertebral fracture), resulting in compromise of a nerve root (including the cauda equina) or the spinal cord. With:

A. Evidence of nerve root compression characterized by neuro-anatomic distribution of pain, limitation of motion of the spine, motor loss (atrophy with associated muscle weakness or muscle weakness) accompanied by sensory or reflex loss and, if there is involvement of the lower back, positive straight-leg raising test (sitting and supine);

or

B. Spinal arachnoiditis, confirmed by an operative note or pathology report of tissue biopsy, or by appropriate medically acceptable imaging, manifested by severe burning or painful dysesthesia, resulting in the need for changes in position or posture more than once every 2 hours;

or

C. Lumbar spinal stenosis resulting in pseudoclaudication, established by findings on appropriate medically acceptable imaging, manifested by chronic nonradicular pain and weakness, and resulting in inability to ambulate effectively, as defined in 1.00B2b.

1.05 *Amputation (due to any cause).*

A. Both hands; or

or

B. One or both lower extremities at or above the tarsal region, with stump complications resulting in medical inability to use a prosthetic device to ambulate effectively, as defined in 1.00B2b, which have lasted or are expected to last for at least 12 months;

or

C. One hand and one lower extremity at or above the tarsal region, with inability to ambulate effectively, as defined in 1.00B2b; OR

D. Hemipelvectomy or hip disarticulation.

1.06 *Fracture of the femur, tibia, pelvis, or one or more of the tarsal bones.* With:

A. Solid union not evident on appropriate medically acceptable imaging and not clinically solid;

and

B. Inability to ambulate effectively, as defined in 1.00B2b, and return to effective ambulation did not occur or is not expected to occur within 12 months of onset.

1.07 *Fracture of an upper extremity* with nonunion of a fracture of the shaft of the humerus, radius, or ulna, under continuing surgical management, as defined in 1.00M, directed toward restoration of functional use of the extremity, and such function was not restored or expected to be restored within 12 months of onset.

1.08 *Soft tissue injury (e.g., burns)* of an upper or lower extremity, trunk, or face and head, under continuing surgical management, as defined in 1.00M, directed toward the salvage or restoration of major function, and such major function was not restored or expected to be restored within 12 months of onset. Major function of the face and head is described in 1.00O.

2.00 Special Senses and Speech

A. *How do we evaluate visual disorders?*

1. *What are visual disorders?* Visual disorders are abnormalities of the eye, the optic nerve, the optic tracts, or the brain that may cause a loss of visual acuity or visual fields. A loss of visual acuity limits your ability to

distinguish detail, read, or do fine work. A loss of visual fields limits your ability to perceive visual stimuli in the peripheral extent of vision.

2. *How do we define statutory blindness?* Statutory blindness is blindness as defined in sections 216(i)(1) and 1614(a)(2) of the Social Security Act (Act).

a. The Act defines blindness as central visual acuity of 20/200 or less in the better eye with the use of a correcting lens. We use your best-corrected central visual acuity for distance in the better eye when we determine if this definition is met. (For visual acuity testing requirements, see 2.00A5.)

b. The Act also provides that an eye that has a visual field limitation such that the widest diameter of the visual field subtends an angle no greater than 20 degrees is considered as having a central visual acuity of 20/200 or less. (For visual field testing requirements, see 2.00A6.)

c. You have statutory blindness only if your visual disorder meets the criteria of 2.02 or 2.03A. You do not have statutory blindness if your visual disorder medically equals the criteria of 2.02 or 2.03A or meets or medically equals the criteria of 2.03B, 2.03C, 2.04A, or 2.04B because your disability is based on criteria other than those in the statutory definition of blindness.

3. *What evidence do we need to establish statutory blindness under title XVI?* To establish that you have statutory blindness under title XVI, we need evidence showing only that your central visual acuity in your better eye or your visual field in your better eye meets the criteria in 2.00A2, provided that those measurements are consistent with the other evidence in your case record. We do not need documentation of the cause of your blindness. Also, there is no duration requirement for statutory blindness under title XVI (see §§ 416.981 and 416.983 of this chapter).

4. *What evidence do we need to evaluate visual disorders, including those that result in statutory blindness under title II?* To evaluate your visual disorder, we usually need a report of an eye examination that includes measurements of your best-corrected central visual acuity (see 2.00A5) or the extent of your visual fields (see 2.00A6), as appropriate. If you have visual acuity or visual field loss, we need documentation of the cause of the loss. A standard eye examination will usually indicate the cause of any visual acuity loss. A standard eye examination can also indicate the cause of some types of visual field deficits. Some disorders, such as cortical visual disorders, may result in abnormalities that do not appear on a standard eye examination. If the standard eye examination does not indicate the cause of your vision loss, we will request the information used to establish the presence of your visual disorder. If your visual disorder does not satisfy the criteria in 2.02, 2.03, or 2.04, we will request a description of how your visual disorder affects your ability to function.

5. *How do we measure your best-corrected central visual acuity?*

a. *Visual acuity testing.* When we need to measure your best-corrected central visual acuity (your optimal visual acuity attainable with the use of a corrective lens), we use visual acuity testing for distance that was carried out using Snellen methodology or any other testing methodology that is comparable to Snellen methodology.

(i) Your best-corrected central visual acuity for distance is usually measured by determining what you can see from 20 feet. If your visual acuity is measured for a distance other than 20 feet, we will convert it to a 20-foot measurement. For example, if your visual acuity is measured at 10 feet and is reported as 10/40, we will convert this measurement to 20/80.

(ii) A visual acuity recorded as CF (counts fingers), HM (hand motion only), LP or LPO (light perception or light perception only), or NLP (no light perception) indicates that no optical correction will improve your visual acuity. If your central visual acuity in an eye is recorded as CF, HM, LP or LPO, or NLP, we will determine that your best-corrected central visual acuity is 20/200 or less in that eye.

(iii) We will not use the results of pinhole testing or automated refraction acuity to determine your best-corrected central visual acuity. These tests provide an estimate of potential visual acuity but not an actual measurement of your best-corrected central visual acuity.

b. *Other test charts.* Most test charts that use Snellen methodology do not have lines that measure visual acuity between 20/100 and 20/200. Some test charts, such as the Bailey-Lovie or the Early Treatment Diabetic Retinopathy Study (ETDRS), used mostly in research settings, have such lines. If your visual acuity is measured with one of these charts, and you cannot read any of the letters on the 20/100 line, we will determine that you have statutory blindness based on a visual acuity of 20/200 or less. For example, if your best-corrected central visual acuity for distance in the better eye is 20/160 using an ETDRS chart, we will find that you have statutory blindness. Regardless of the type of test chart used, you do not have statutory blindness if you can read at least one letter on the 20/100 line. For example, if your best-corrected central visual acuity for distance in the better eye is 20/125 + 1 using an ETDRS chart, we will find that you do not have statutory blindness because you are able to read one letter on the 20/100 line.

c. *Testing using a specialized lens.* In some instances, you may have visual acuity testing performed using specialized lens, such as a contact lens. We will use the visual acuity measurements obtained with a specialized

lens only if you have demonstrated the ability to use the specialized lens on a sustained basis. We will not use visual acuity measurements obtained with telescopic lenses.

d. *Cycloplegic refraction* is an examination of the eye performed after administering cycloplegic eye drops capable of relaxing the ability of the pupil to become smaller and temporarily paralyzing the focusing muscles. If your case record contains the results of cycloplegic refraction, we may use the results to determine your best-corrected central visual acuity. We will not purchase cycloplegic refraction.

e. *Visual evoked response (VER) testing* measures your response to visual events and can often detect dysfunction that is undetectable through other types of examinations. If you have an absent response to VER testing in your better eye, we will determine that your best-corrected central visual acuity is 20/200 or less in that eye and that your visual acuity loss satisfies the criterion in 2.02 when these test results are consistent with the other evidence in your case record. If you have a positive response to VER testing in an eye, we will not use that result to determine your best-corrected central visual acuity in that eye.

6. *How do we measure your visual fields?*

a. *General.* We generally need visual field testing when you have a visual disorder that could result in visual field loss, such as glaucoma, retinitis pigmentosa, or optic neuropathy, or when you display behaviors that suggest a visual field loss. When we need to measure the extent of your visual field loss, we use visual field testing (also referred to as perimetry) carried out using automated static threshold perimetry performed on an acceptable perimeter. (For perimeter requirements, see 2.00A9.)

b. *Automated static threshold perimetry requirements.*

(i) The test must use a white size III Goldmann stimulus and a 31.5 apostilb (asb) white background (or a 10 candela per square meter (cd/m^2) white background). The stimuli test locations must be no more than 6 degrees apart horizontally or vertically. Measurements must be reported on standard charts and include a description of the size and intensity of the test stimulus.

(ii) We measure the extent of your visual field loss by determining the portion of the visual field in which you can see a white III4e stimulus. The ''III'' refers to the standard Goldmann test stimulus size III ($4\ mm^2$), and the ''4e'' refers to the standard Goldmann intensity filter (0 decibel (dB) attenuation, which allows presentation of the maximum luminance) used to determine the intensity of the stimulus.

(iii) In automated static threshold perimetry, the intensity of the stimulus varies. The intensity of the stimulus is expressed in decibels (dB). A perimeter's maximum stimulus luminance is usually assigned the value 0 dB. We need to determine the dB level that corresponds to a 4e intensity for the particular perimeter being used. We will then use the dB printout to determine which points you see at a 4e intensity level (a ''seeing point''). For example:

A. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 10,000 asb, a 10 dB stimulus is equivalent to a 4e stimulus. Any point you see at 10 dB or greater is a seeing point.

B. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 4,000 asb, a 6 dB stimulus is equivalent to a 4e stimulus. Any point you see at 6 dB or greater is a seeing point.

C. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 1,000 asb, a 0 dB stimulus is equivalent to a 4e stimulus. Any point you see at 0 dB or greater is a seeing point.

c. *Evaluation under 2.03A.* To determine statutory blindness based on visual field loss in your better eye (2.03A), we need the results of a visual field test that measures the central 24 to 30 degrees of your visual field; that is, the area measuring 24 to 30 degrees from the point of fixation. Acceptable tests include the Humphrey Field Analyzer (HFA) 30–2, HFA 24–2, and Octopus 32.

d. *Evaluation under 2.03B.* To determine whether your visual field loss meets listing 2.03B, we use the mean deviation or defect (MD) from acceptable automated static threshold perimetry that measures the central 30 degrees of the visual field. MD is the average sensitivity deviation from normal values for all measured visual field locations. When using results from HFA tests, which report the MD as a negative number, we use the absolute value of the MD to determine whether your visual field loss meets listing 2.03B. We cannot use tests that do not measure the central 30 degrees of the visual field, such as the HFA 24–2, to determine if your impairment meets or medically equals 2.03B.

e. *Other types of perimetry.* If the evidence in your case contains visual field measurements obtained using manual or automated kinetic perimetry, such as Goldmann perimetry or the HFA ''SSA Test Kinetic,'' we can generally use these results if the kinetic test was performed using a white III4e stimulus projected on a white 31.5 asb ($10\ cd/m^2$) background. Automated kinetic perimetry, such as the HFA ''SSA Test Kinetic,'' does not detect limitations in the central visual field because testing along a meridian stops when you see the stimulus. If your visual disorder has progressed to the point at which it is likely to result in a significant limitation in the central visual field, such as a scotoma (see 2.00A6h), we will not use *automated* kinetic perimetry to determine the extent of

your visual field loss. Instead, we will determine the extent of your visual field loss using automated static threshold perimetry or manual kinetic perimetry.

f. *Screening tests.* We will not use the results of visual field screening tests, such as confrontation tests, tangent screen tests, or automated static screening tests, to determine that your impairment meets or medically equals a listing or to evaluate your residual functional capacity. We can consider normal results from visual field screening tests to determine whether your visual disorder is severe when these test results are consistent with the other evidence in your case record. (See §§404.1520(c), 404.1521, 416.920(c), and 416.921 of this chapter.) We will not consider normal test results to be consistent with the other evidence if the clinical findings indicate that your visual disorder has progressed to the point that it is likely to cause visual field loss, or you have a history of an operative procedure for retinal detachment.

g. *Use of corrective lenses.* You must not wear eyeglasses during visual field testing because they limit your field of vision. You may wear contact lenses to correct your visual acuity during the visual field test to obtain the most accurate visual field measurements. For this single purpose, you do not need to demonstrate that you have the ability to use the contact lenses on a sustained basis.

h. *Scotoma.* A scotoma is a field defect or non-seeing area (also referred to as a ''blind spot'') in the visual field surrounded by a normal field or seeing area. When we measure your visual field, we subtract the length of any scotoma, other than the normal blind spot, from the overall length of any diameter on which it falls.

7. *How do we determine your visual acuity efficiency, visual field efficiency, and visual efficiency?*

a. *General. Visual efficiency,* a calculated value of your remaining visual function, is the combination of your *visual acuity efficiency* and your *visual field efficiency* expressed as a percentage.

b. *Visual acuity efficiency.* Visual acuity efficiency is a percentage that corresponds to the best-corrected central visual acuity for distance in your better eye. See Table 1.

TABLE 1—VISUAL ACUITY EFFICIENCY

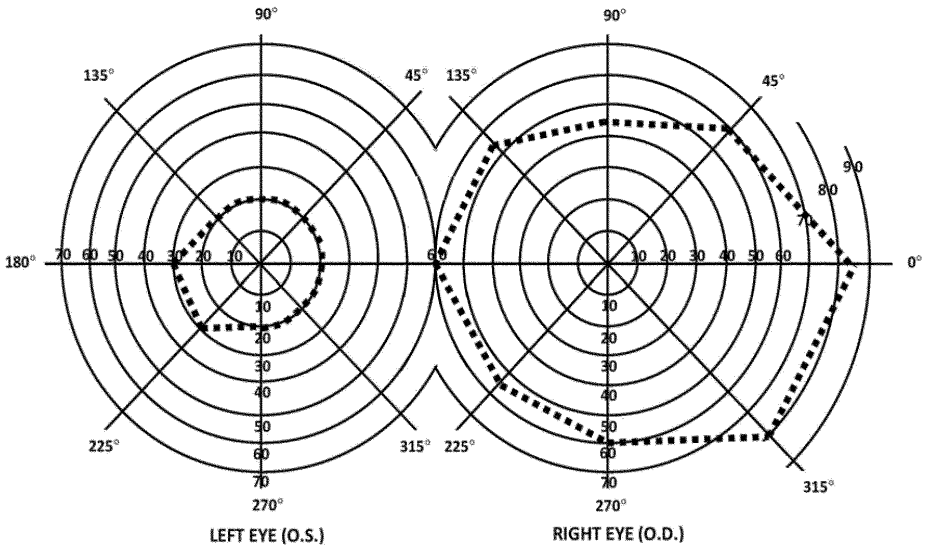
Snellen best-corrected central visual acuity for distance		Visual acuity efficiency (%) (2.04A)
English	Metric	
20/16	6/5	100
20/20	6/6	100
20/25	6/7.5	95
20/30	6/9	90
20/40	6/12	85
20/50	6/15	75
20/60	6/18	70
20/70	6/21	65
20/80	6/24	60
20/100	6/30	50

c. *Visual field efficiency.* Visual field efficiency is a percentage that corresponds to the visual field in your better eye. Under 2.03C, we require kinetic perimetry to determine your visual field efficiency percentage. We calculate the visual field efficiency percentage by adding the number of degrees you see along the eight principal meridians found on a visual field chart (0, 45, 90, 135, 180, 225, 270, and 315) in your better eye and dividing by 5. For example, in Figure 1:

A. The diagram of the left eye illustrates a visual field, as measured with a III4e stimulus, contracted to 30 degrees in two meridians (180 and 225) and to 20 degrees in the remaining six meridians. The visual efficiency percentage of this field is: $((2 \times 30) + (6 \times 20)) \div 5 = 36$ percent.

B. The diagram of the right eye illustrates the extent of a normal visual field as measured with a III4e stimulus. The sum of the eight principal meridians of this field is 500 degrees. The visual efficiency percentage of this field is $500 \div 5 = 100$ percent.

Figure 1:



d. *Visual efficiency.* Under 2.04A, we calculate the visual efficiency percentage by multiplying your visual acuity efficiency percentage (see 2.00A7b) by your visual field efficiency percentage (see 2.00A7c) and dividing by 100. For example, if your visual acuity efficiency percentage is 75 and your visual field efficiency percentage is 36, your visual efficiency percentage is: (75 × 36) ÷ 100 = 27 percent.

8. *How do we determine your visual acuity impairment value, visual field impairment value, and visual impairment value?*

a. *General. Visual impairment value,* a calculated value of your loss of visual function, is the combination of your *visual acuity impairment value* and your *visual field impairment value.*

b. *Visual acuity impairment value.* Your visual acuity impairment value corresponds to the best-corrected central visual acuity for distance in your better eye. See Table 2.

TABLE 2—VISUAL ACUITY IMPAIRMENT VALUE

Snellen best-corrected central visual acuity for distance		Visual acuity impairment value (2.04B)
English	Metric	
20/16	6/5	0.00
20/20	6/6	0.00
20/25	6/7.5	0.10
20/30	6/9	0.18
20/40	6/12	0.30
20/50	6/15	0.40
20/60	6/18	0.48
20/70	6/21	0.54
20/80	6/24	0.60
20/100	6/30	0.70

c. *Visual field impairment value.* Your visual field impairment value corresponds to the visual field in your better eye. Using the MD from acceptable automated static threshold perimetry, we calculate the visual field impairment value by dividing the absolute value of the MD by 22. For example, if your MD on an HFA 30–2 is −16, your visual field impairment value is: $-16\surd \div 22 = 0.73$.

d. *Visual impairment value.* Under 2.04B, we calculate the visual impairment value by adding your visual acuity impairment value (see 2.00A8b) and your visual field impairment value (see 2.00A8c). For example, if your visual acuity impairment value is 0.48 and your visual field impairment value is 0.73, your visual impairment value is: 0.48 + 0.73 = 1.21.

9. *What are our requirements for an acceptable perimeter?* We will use results from automated static threshold perimetry performed on a perimeter that:

a. Uses optical projection to generate the test stimuli.

b. Has an internal normative database for automatically comparing your performance with that of the general population.

c. Has a statistical analysis package that is able to calculate visual field indices, particularly MD.

d. Demonstrates the ability to correctly detect visual field loss and correctly identify normal visual fields.

e. Demonstrates good test-retest reliability.

f. Has undergone clinical validation studies by three or more independent laboratories with results published in peer-reviewed ophthalmic journals.

B. *How do we evaluate hearing loss?*

1. *What evidence do we need?*

a. We need evidence showing that you have a medically determinable impairment that causes your hearing loss and audiometric measurements of the severity of your hearing loss. We generally require both a complete otologic examination and audiometric testing to establish that you have a medically determinable impairment that causes your hearing loss. You should have this audiometric testing within 2 months of the complete otologic examination. Once we have evidence that you have a medically determinable impairment, we can use the results of later audiometric testing to assess the severity of your hearing loss without another complete otologic examination. We will consider your test scores together with any other relevant information we have about your hearing, including information from outside of the test setting.

b. The complete otologic examination must be performed by a licensed physician (medical or osteopathic doctor). It must include your medical history, your description of how your hearing loss affects you, and the physician's description of the appearance of the external ears (pinnae and external ear canals), evaluation of the tympanic membranes, and assessment of any middle ear abnormalities.

c. Audiometric testing must be performed by, or under the direct supervision of, an otolaryngologist or by an audiologist qualified to perform such tests. We consider an audiologist to be qualified if he or she is currently and fully licensed or registered as a clinical audiologist by the State or U.S. territory in which he or she practices. If no licensure or registration is available, the audiologist must be currently certified by the American Board of Audiology or have a Certificate of Clinical Competence (CCC–A) from the American Speech-Language-Hearing Association (ASHA).

2. *What audiometric testing do we need when you do not have a cochlear implant?*

a. We generally need pure tone air conduction and bone conduction testing, speech reception threshold (SRT) testing (also referred to as ''spondee threshold'' or ''ST'' testing), and word recognition testing (also referred to as ''word discrimination'' or ''speech discrimination'' testing). This testing must be conducted in a sound-treated booth or room and must be in accordance with the most recently published standards of the American National Standards Institute (ANSI). Each ear must be tested separately.

b. You must not wear hearing aids during the testing. Additionally, a person described in 2.00B1c must perform an otoscopic examination immediately before the audiometric testing. (An *otoscopic examination* provides a description of the appearance of your external ear canals and an evaluation of the tympanic membranes. In these rules, we use the term to include otoscopic examinations performed by physicians and otoscopic inspections performed by audiologists and others.) The otoscopic examination must show that there are no conditions that would prevent valid audiometric testing, such as fluid in the ear, ear infection, or obstruction in an ear canal. The person performing the test should also report on any other factors, such as your cooperation with the test, that can affect the interpretation of the test results.

c. To determine whether your hearing loss meets the air and bone conduction criteria in 2.10A, we will average your air and bone conduction hearing thresholds at 500, 1000, and 2000 Hertz (Hz). If you do not have a response at a particular frequency, we will use a threshold of 5 decibels (dB) over the limit of the audiometer.

d. The SRT is the minimum dB level required for you to recognize 50 percent of the words on a standard list of spondee words. (Spondee words are two-syllable words that have equal stress on each syllable.) The SRT is usually within 10 dB of the average pure tone air conduction hearing thresholds at 500, 1000, and 2000 Hz. If the SRT is not within 10 dB of the average pure tone air conduction threshold, the reason for the discrepancy must be documented. If we cannot determine that there is a medical basis for the discrepancy, we will not use the results of the testing to determine whether your hearing loss meets a listing.

e. Word recognition testing determines your ability to recognize a standardized list of phonetically balanced monosyllabic words in the absence of any visual cues. This testing must be performed in quiet. The list may be recorded or presented live, but in either case the words should be presented at a level of amplification that will measure your maximum ability to discriminate words, usually 35 to 40 dB above your SRT. However, the

amplification level used in the testing must be medically appropriate, and you must be able to tolerate it. If you cannot be tested at 35 to 40 dB above your SRT, the person who performs the test should report your word recognition testing score at your highest comfortable level of amplification.

3. *What audiometric testing do we need when you have a cochlear implant?*

a. If you have a cochlear implant, we will consider you to be disabled until 1 year after initial implantation.

b. After that period, we need word recognition testing performed with any version of the Hearing in Noise Test (HINT) to determine whether your impairment meets 2.11B. This testing must be conducted in quiet in a sound field. Your implant must be functioning properly and adjusted to your normal settings. The sentences should be presented at 60 dB HL (Hearing Level) and without any visual cues.

4. *How do we evaluate your word recognition ability if you are not fluent in English?*

If you are not fluent in English, you should have word recognition testing using an appropriate word list for the language in which you are most fluent. The person conducting the test should be fluent in the language used for the test. If there is no appropriate word list or no person who is fluent in the language and qualified to perform the test, it may not be possible to measure your word recognition ability. If your word recognition ability cannot be measured, your hearing loss cannot meet 2.10B or 2.11B. Instead, we will consider the facts of your case to determine whether you have difficulty understanding words in the language in which you are most fluent, and if so, whether that degree of difficulty medically equals 2.10B or 2.11B. For example, we will consider how you interact with family members, interpreters, and other persons who speak the language in which you are most fluent.

C. *How do we evaluate vertigo associated with disturbances of labyrinthine-vestibular function, including Meniere's disease?*

1. These disturbances of balance are characterized by an hallucination of motion or loss of position sense and a sensation of dizziness which may be constant or may occur in paroxysmal attacks. Nausea, vomiting, ataxia, and incapacitation are frequently observed, particularly during the acute attack. It is important to differentiate the report of rotary vertigo from that of ''dizziness'' which is described as lightheadedness, unsteadiness, confusion, or syncope.

2. Meniere's disease is characterized by paroxysmal attacks of vertigo, tinnitus, and fluctuating hearing loss. Remissions are unpredictable and irregular, but may be longlasting; hence, the severity of impairment is best determined after prolonged observation and serial reexaminations.

3. The diagnosis of a vestibular disorder requires a comprehensive neuro-otolaryngologic examination with a detailed description of the vertiginous episodes, including notation of frequency, severity, and duration of the attacks. Pure tone and speech audiometry with the appropriate special examinations, such as Bekesy audiometry, are necessary. Vestibular functions is assessed by positional and caloric testing, preferably by electronystagmography. When polytomograms, contrast radiography, or other special tests have been performed, copies of the reports of these tests should be obtained in addition to appropriate medically acceptable imaging reports of the skull and temporal bone. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

D. *Loss of speech.* In evaluating the loss of speech, the ability to produce speech by any means includes the use of mechanical or electronic devices that improve voice or articulation. Impairments of speech may also be evaluated under the body system for the underlying disorder, such as neurological disorders, 11.00ff.

E. *How do we evaluate impairments that do not meet one of the special senses and speech listings?*

1. These listings are only examples of common special senses and speech disorders that we consider severe enough to prevent an individual from doing any gainful activity. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. If you have a medically determinable impairment(s) that does not meet a listing, we will determine whether the impairment(s) medically equals a listing. (See §§ 404.1526 and 416.926.) If you have an impairment(s) that does not meet or medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. Therefore, we proceed to the fourth, and if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920. When we decide whether you continue to be disabled, we use the rules in §§ 404.1594, 416.994, or 416.994a, as appropriate.

2.01 Category of Impairments, Special Senses and Speech

2.02 *Loss of central visual acuity.* Remaining vision in the better eye after best correction is 20/200 or less.

2.03 *Contraction of the visual field in the better eye,* with:

A. The widest diameter subtending an angle around the point of fixation no greater than 20 degrees.

OR

B. An MD of 22 decibels or greater, determined by automated static threshold perimetry that measures the central 30 degrees of the visual field (see 2.00A6d).

OR

C. A visual field efficiency of 20 percent or less, determined by kinetic perimetry (see 2.00A7c).

2.04 *Loss of visual efficiency, or visual impairment, in the better eye:*

A. A visual efficiency percentage of 20 or less after best correction (see 2.00A7d).

OR

B. A visual impairment value of 1.00 or greater after best correction (see 2.00A8d).

2.07 *Disturbance of labyrinthine-vestibular function (including Meniere's disease),* characterized by a history of frequent attacks of balance disturbance, tinnitus, and progressive loss of hearing. With both A and B:

A. Disturbed function of vestibular labyrinth demonstrated by caloric or other vestibular tests; and

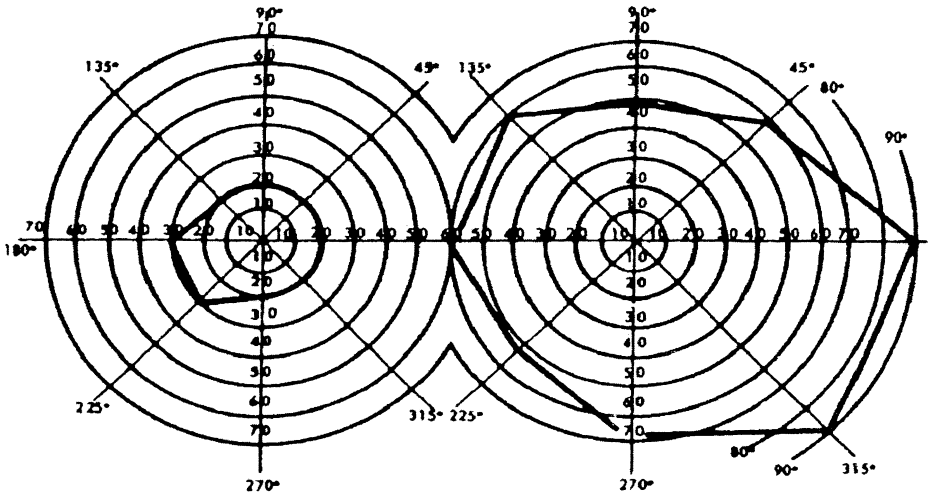
B. Hearing loss established by audiometry.

2.09 *Loss of speech* due to any cause, with inability to produce by any means speech that can be heard, understood, or sustained.

TABLE 1—PERCENTAGE OF VISUAL ACUITY EFFICIENCY CORRESPONDING TO THE BEST-CORRECTED VISUAL ACUITY MEASUREMENT FOR DISTANCE IN THE BETTER EYE

Snellen		Percent visual acuity efficiency
English	Metric	
20/16	6/5	100
20/20	6/6	100
20/25	6/7.5	95
20/30	6/9	90
20/40	6/12	85
20/50	6/15	75
20/60	6/18	70
20/70	6/21	65
20/80	6/24	60
20/100	6/30	50

TABLE 2—CHART OF VISUAL FIELDS



1. The diagram of the right eye illustrates the extent of a normal visual field as measured with a III4e stimulus. The sum of the eight principal meridians of this field is 500 degrees.

2. The diagram of the left eye illustrates a visual field contracted to 30 degrees in two meridians and to 20 degrees in the remaining six meridians. The percent of visual field efficiency of this field is: $(2 \times 30) + (6 \times 20) = 180 \div 500 = 0.36$ or 36 percent visual field efficiency.

2.10 *Hearing loss not treated with cochlear implantation.*

A. An average air conduction hearing threshold of 90 decibels or greater in the better ear and an average bone conduction hearing threshold of 60 decibels or greater in the better ear (*see* 2.00B2c).

OR

B. A word recognition score of 40 percent or less in the better ear determined using a standardized list of phonetically balanced monosyllabic words (see 2.00B2e).

2.11 *Hearing loss treated with cochlear implantation.*

A. Consider under a disability for 1 year after initial implantation.

OR

B. If more than 1 year after initial implantation, a word recognition score of 60 percent or less determined using the HINT (*see* 2.00B3b).

3.00 Respiratory System

A. *Introduction.* The listings in this section describe impairments resulting from respiratory disorders based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of treatment prescribed by a treating source. Respiratory disorders along with any associated impairment(s) must be established by medical evidence. Evidence must be provided in sufficient detail to permit an independent reviewer to evaluate the severity of the impairment.

Many individuals, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is evidence of such treatment, the longitudinal clinical record must include a description of the treatment prescribed by the treating source and response in addition to information about the nature and severity of the impairment. It is important to document any prescribed treatment and response, because this medical management may have improved the individual's functional status. The longitudinal record should provide information regarding functional recovery, if any.

Some individuals will not have received ongoing treatment or have an ongoing relationship with the medical community, despite the existence of a severe impairment(s). An individual who does not receive treatment may or may not be able to show the existence of an impairment that meets the criteria of these listings. Even if an individual does not show that his or her impairment meets the criteria of these listings, the individual may have an impairment(s) equivalent in severity to one of the listed impairments or be disabled because of a limited residual functional capacity. Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the individual's functioning, and the frequency, severity, and duration of symptoms. Also, the asthma listing specifically includes a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.

Impairments caused by chronic disorders of the respiratory system generally produce irreversible loss of pulmonary function due to ventilatory impairments, gas exchange abnormalities, or a combination of both. The most common symptoms attributable to these disorders are dyspnea on exertion, cough, wheezing, sputum production, hemoptysis, and chest pain. Because these symptoms are common to many other diseases, a thorough medical history, physical examination, and chest x-ray or other appropriate imaging technique are required to establish chronic pulmonary disease. Pulmonary function testing is required to assess the severity of the respiratory impairment once a disease process is established by appropriate clinical and laboratory findings.

Alterations of pulmonary function can be due to obstructive airway disease (e.g., emphysema, chronic bronchitis, asthma), restrictive pulmonary disorders with primary loss of lung volume (e.g., pulmonary resection, thoracoplasty, chest cage deformity as in kyphoscoliosis or obesity), or infiltrative interstitial disorders (e.g., diffuse pulmonary fibrosis). Gas exchange abnormalities without significant airway obstruction can be produced by interstitial disorders. Disorders involving the pulmonary circulation (e.g., primary pulmonary hypertension, recurrent thromboembolic disease, primary or secondary pulmonary vasculitis) can produce pulmonary vascular hypertension and, eventually, pulmonary heart disease (cor pulmonale) and right heart failure. Persistent hypoxemia produced by any chronic pulmonary disorder also can result in chronic pulmonary hypertension and right heart failure. Chronic infection, caused most frequently by mycobacterial or mycotic organisms, can produce extensive and progressive lung destruction resulting in marked loss of pulmonary function. Some disorders, such as bronchiectasis, cystic fibrosis, and asthma, can be associated with intermittent exacerbations of such frequency and intensity that they produce a disabling impairment, even when pulmonary function during periods of relative clinical stability is relatively well-maintained.

Respiratory impairments usually can be evaluated under these listings on the basis of a complete medical history, physical examination, a chest x-ray or other appropriate imaging techniques, and spirometric pulmonary function tests. In some situations, most typically with a diagnosis of diffuse interstitial fibrosis or clinical findings suggesting cor pulmonale, such as cyanosis or

secondary polycythemia, an impairment may be underestimated on the basis of spirometry alone. More sophisticated pulmonary function testing may then be necessary to determine if gas exchange abnormalities contribute to the severity of a respiratory impairment. Additional testing might include measurement of diffusing capacity of the lungs for carbon monoxide or resting arterial blood gases. Measurement of arterial blood gases during exercise is required infrequently. In disorders of the pulmonary circulation, right heart catheterization with angiography and/or direct measurement of pulmonary artery pressure may have been done to establish a diagnosis and evaluate severity. When performed, the results of the procedure should be obtained. Cardiac catheterization will not be purchased.

These listings are examples of common respiratory disorders that are severe enough to prevent a person from engaging in any gainful activity. When an individual has a medically determinable impairment that is not listed, an impairment which does not meet a listing, or a combination of impairments no one of which meets a listing, we will consider whether the individual's impairment or combination of impairments is medically equivalent in severity to a listed impairment. Individuals who have an impairment(s) with a level of severity which does not meet or equal the criteria of the listings may or may not have the residual functional capacity (RFC) which would enable them to engage in substantial gainful activity. Evaluation of the impairment(s) of these individuals will proceed through the final steps of the sequential evaluation process.

B. *Mycobacterial, mycotic, and other chronic persistent infections of the lung.* These disorders are evaluated on the basis of the resulting limitations in pulmonary function. Evidence of chronic infections, such as active mycobacterial diseases or mycoses with positive cultures, drug resistance, enlarging parenchymal lesions, or cavitation, is not, by itself, a basis for determining that an individual has a disabling impairment expected to last 12 months. In those unusual cases of pulmonary infection that persist for a period approaching 12 consecutive months, the clinical findings, complications, therapeutic considerations, and prognosis must be carefully assessed to determine whether, despite relatively well-maintained pulmonary function, the individual nevertheless has an impairment that is expected to last for at least 12 consecutive months and prevent gainful activity.

C. *Episodic respiratory disease.* When a respiratory impairment is episodic in nature, as can occur with exacerbations of asthma, cystic fibrosis, bronchiectasis, or chronic asthmatic bronchitis, the frequency and intensity of episodes that occur despite prescribed treatment are often the major criteria for determining the level of impairment. Documentation for these exacerbations should include available hospital, emergency facility and/or physician records indicating the dates of treatment; clinical and laboratory findings on presentation, such as the results of spirometry and arterial blood gas studies (ABGS); the treatment administered; the time period required for treatment; and the clinical response. Attacks of asthma, episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum), or respiratory failure as referred to in paragraph B of 3.03, 3.04, and 3.07, are defined as prolonged symptomatic episodes lasting one or more days and requiring intensive treatment, such as intravenous bronchodilator or antibiotic administration or prolonged inhalational bronchodilator therapy in a hospital, emergency room or equivalent setting. Hospital admissions are defined as inpatient hospitalizations for longer than 24 hours. The medical evidence must also include information documenting adherence to a prescribed regimen of treatment as well as a description of physical signs. For asthma, the medical evidence should include spirometric results obtained between attacks that document the presence of baseline airflow obstruction.

D. *Cystic fibrosis* is a disorder that affects either the respiratory or digestive body systems or both and is responsible for a wide and variable spectrum of clinical manifestations and complications. Confirmation of the diagnosis is based upon an elevated sweat sodium concentration or chloride concentration accompanied by one or more of the following: the presence of chronic obstructive pulmonary disease, insufficiency of exocrine pancreatic function, meconium ileus, or a positive family history. The quantitative pilocarpine iontophoresis procedure for collection of sweat content must be utilized. Two methods are acceptable: the ''Procedure for the Quantitative Iontophoretic Sweat Test for Cystic Fibrosis'' published by the Cystic Fibrosis Foundation and contained in, ''A Test for Concentration of Electrolytes in Sweat in Cystic Fibrosis of the Pancreas Utilizing Pilocarpine Iontophoresis,'' Gibson, I.E., and Cooke, R.E., *Pediatrics,* Vol. 23: 545, 1959; or the ''Wescor Macroduct System.'' To establish the diagnosis of cystic fibrosis, the sweat sodium or chloride content must be analyzed quantitatively using an acceptable laboratory technique. Another diagnostic test is the ''CF gene mutation analysis'' for homozygosity of the cystic fibrosis gene. The pulmonary manifestations of this disorder should be evaluated under 3.04. The nonpulmonary aspects of cystic fibrosis should be evaluated under the digestive body system (5.00). Because cystic fibrosis may involve the respiratory and digestive body systems, the combined effects of the involvement of

these body systems must be considered in case adjudication.

E. *Documentation of pulmonary function testing.* The results of spirometry that are used for adjudication under paragraphs A and B of 3.02 and paragraph A of 3.04 should be expressed in liters (L), body temperature and pressure saturated with water vapor (BTPS). The reported one-second forced expiratory volume (FEV_1) and forced vital capacity (FVC) should represent the largest of at least three satisfactory forced expiratory maneuvers. Two of the satisfactory spirograms should be reproducible for both pre-bronchodilator tests and, if indicated, post-bronchodilator tests. A value is considered reproducible if it does not differ from the largest value by more than 5 percent or 0.1 L, whichever is greater. The highest values of the FEV_1 and FVC, whether from the same or different tracings, should be used to assess the severity of the respiratory impairment. Peak flow should be achieved early in expiration, and the spirogram should have a smooth contour with gradually decreasing flow throughout expiration. The zero time for measurement of the FEV_1 and FVC, if not distinct, should be derived by linear back-extrapolation of peak flow to zero volume. A spirogram is satisfactory for measurement of the FEV_1 if the expiratory volume at the back-extrapolated zero time is less than 5 percent of the FVC or 0.1 L, whichever is greater. The spirogram is satisfactory for measurement of the FVC if maximal expiratory effort continues for at least 6 seconds, or if there is a plateau in the volume-time curve with no detectable change in expired volume (VE) during the last 2 seconds of maximal expiratory effort.

Spirometry should be repeated after administration of an aerosolized bronchodilator under supervision of the testing personnel if the pre-bronchodilator FEV_1 value is less than 70 percent of the predicted normal value. Pulmonary function studies should not be performed unless the clinical status is stable (e.g., the individual is not having an asthmatic attack or suffering from an acute respiratory infection or other chronic illness). Wheezing is common in asthma, chronic bronchitis, or chronic obstructive pulmonary disease and does not preclude testing. The effect of the administered bronchodilator in relieving bronchospasm and improving ventilatory function is assessed by spirometry. If a bronchodilator is not administered, the reason should be clearly stated in the report. Pulmonary function studies performed to assess airflow obstruction without testing after bronchodilators cannot be used to assess levels of impairment in the range that prevents any gainful work activity, unless the use of bronchodilators is contraindicated. Post-bronchodilator testing should be performed 10 minutes after bronchodilator administration. The dose and name of the bronchodilator administered should be specified. The values in paragraphs A and B of 3.02 must only be used as criteria for the level of ventilatory impairment that exists during the individual's most stable state of health (*i.e.*, any period in time except during or shortly after an exacerbation).

The appropriately labeled spirometric tracing, showing the claimant's name, date of testing, distance per second on the abscissa and distance per liter (L) on the ordinate, must be incorporated into the file. The manufacturer and model number of the device used to measure and record the spirogram should be stated. The testing device must accurately measure both time and volume, the latter to within 1 percent of a 3 L calibrating volume. If the spirogram was generated by any means other than direct pen linkage to a mechanical displacement-type spirometer, the testing device must have had a recorded calibration performed previously on the day of the spirometric measurement.

If the spirometer directly measures flow, and volume is derived by electronic integration, the linearity of the device must be documented by recording volume calibrations at three different flow rates of approximately 30 L/min (3 L/6 sec), 60 L/min (3 L/3 sec), and 180 L/min (3 L/sec). The volume calibrations should agree to within 1 percent of a 3 L calibrating volume. The proximity of the flow sensor to the individual should be noted, and it should be stated whether or not a BTPS correction factor was used for the calibration recordings and for the individual's actual spirograms.

The spirogram must be recorded at a speed of at least 20 mm/sec, and the recording device must provide a volume excursion of at least 10 mm/L. If reproductions of the original spirometric tracings are submitted, they must be legible and have a time scale of at least 20 mm/sec and a volume scale of at least 10 mm/L to permit independent measurements. Calculation of FEV_1 from a flow-volume tracing is not acceptable, *i.e.*, the spirogram and calibrations must be presented in a volume-time format at a speed of at least 20 mm/sec and a volume excursion of at least 10 mm/L to permit independent evaluation.

A statement should be made in the pulmonary function test report of the individual's ability to understand directions as well as his or her effort and cooperation in performing the pulmonary function tests.

The pulmonary function tables in 3.02 and 3.04 are based on measurement of standing height without shoes. If an individual has marked spinal deformities (e.g., kyphoscoliosis), the measured span between the fingertips with the upper extremities abducted 90 degrees should be substituted for

height when this measurement is greater than the standing height without shoes.

F. *Documentation of chronic impairment of gas exchange.*

1. *Diffusing capacity of the lungs for carbon monoxide (DLCO).* A diffusing capacity of the lungs for carbon monoxide study should be purchased in cases in which there is documentation of chronic pulmonary disease, but the existing evidence, including properly performed spirometry, is not adequate to establish the level of functional impairment. Before purchasing DLCO measurements, the medical history, physical examination, reports of chest x-ray or other appropriate imaging techniques, and spirometric test results must be obtained and reviewed because favorable decisions can often be made based on available evidence without the need for DLCO studies. Purchase of a DLCO study may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

The DLCO should be measured by the single breath technique with the individual relaxed and seated. At sea level, the inspired gas mixture should contain approximately 0.3 percent carbon monoxide (CO), 10 percent helium (He), 21 percent oxygen (O_2), and the balance nitrogen. At altitudes above sea level, the inspired O_2 concentration may be raised to provide an inspired O_2 tension of approximately 150 mm Hg. Alternatively, the sea level mixture may be employed at altitude and the measured DLCO corrected for ambient barometric pressure. Helium may be replaced by another inert gas at an appropriate concentration. The inspired volume (VI) during the DLCO maneuver should be at least 90 percent of the previously determined vital capacity (VC). The inspiratory time for the VI should be less than 2 seconds, and the breath-hold time should be between 9 and 11 seconds. The washout volume should be between 0.75 and 1.00 L, unless the VC is less than 2 L. In this case, the washout volume may be reduced to 0.50 L; any such change should be noted in the report. The alveolar sample volume should be between 0.5 and 1.0 L and be collected in less than 3 seconds. At least 4 minutes should be allowed for gas washout between repeat studies.

A DLCO should be reported in units of ml CO, standard temperature, pressure, dry (STPD)/min/mm Hg uncorrected for hemoglobin concentration and be based on a single-breath alveolar volume determination. Abnormal hemoglobin or hematocrit values, and/or carboxyhemoglobin levels should be reported along with diffusing capacity.

The DLCO value used for adjudication should represent the mean of at least two acceptable measurements, as defined above. In addition, two acceptable tests should be within 10 percent of each other or 3 ml CO(STPD)/min/mm Hg, whichever is larger. The percent difference should be calculated as $100 \times (\text{test } 1 - \text{test } 2)/\text{average DLCO}$.

The ability of the individual to follow directions and perform the test properly should be described in the written report. The report should include tracings of the VI, breath-hold maneuver, and VE appropriately labeled with the name of the individual and the date of the test. The time axis should be at least 20 mm/sec and the volume axis at least 10 mm/L. The percentage concentrations of inspired O_2 and inspired and expired CO and He for each of the maneuvers should be provided. Sufficient data must be provided, including documentation of the source of the predicted equation, to permit verification that the test was performed adequately, and that, if necessary, corrections for anemia or carboxyhemoglobin were made appropriately.

2. *Arterial blood gas studies (ABGS).* An ABGS performed at rest (while breathing room air, awake and sitting or standing) or during exercise should be analyzed in a laboratory certified by a State or Federal agency. If the laboratory is not certified, it must submit evidence of participation in a national proficiency testing program as well as acceptable quality control at the time of testing. The report should include the altitude of the facility and the barometric pressure on the date of analysis.

Purchase of resting ABGS may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided. If the results of a DLCO study are greater than 40 percent of predicted normal but less than 60 percent of predicted normal, purchase of resting ABGS should be considered. Before purchasing resting ABGS, a program physician, preferably one experienced in the care of patients with pulmonary disease, must review all clinical and laboratory data short of this procedure, including spirometry, to determine whether obtaining the test would present a significant risk to the individual.

3. *Exercise testing.* Exercise testing with measurement of arterial blood gases during exercise may be appropriate in cases in which there is documentation of chronic pulmonary disease, but full development, short of exercise testing, is not adequate to establish if the impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided. In this context, ''full development'' means that results from spirometry and measurement of DLCO and resting ABGS have been obtained from treating sources or through purchase. Exercise arterial blood gas measurements will be required infrequently and should be purchased only after careful review of the medical history, physical examination, chest x-ray or other appropriate imaging techniques, spirometry, DLCO, electrocardiogram (ECG),

hematocrit or hemoglobin, and resting blood gas results by a program physician, preferably one experienced in the care of patients with pulmonary disease, to determine whether obtaining the test would presents a significant risk to the individual. Oximetry and capillary blood gas analysis are not acceptable substitutes for the measurement of arterial blood gases. Arterial blood gas samples obtained after the completion of exercise are not acceptable for establishing an individual's functional capacity.

Generally, individuals with a DLCO greater than 60 percent of predicted normal would not be considered for exercise testing with measurement of blood gas studies. The exercise test facility must be provided with the claimant's clinical records, reports of chest x-ray or other appropriate imaging techniques, and any spirometry, DLCO, and resting blood gas results obtained as evidence of record. The testing facility must determine whether exercise testing present a significant risk to the individual; if it does, the reason for not performing the test must be reported in writing.

4. *Methodology.* Individuals considered for exercise testing first should have resting arterial blood partial pressure of oxygen (PO_2), resting arterial blood partial pressure of carbon dioxide (PCO_2) and negative log of hydrogen ion concentration (pH) determinations by the testing facility. The sample should be obtained in either the sitting or standing position. The individual should then perform exercise under steady state conditions, preferably on a treadmill, breathing room air, for a period of 4 to 6 minutes at a speed and grade providing an oxygen consumption of approximately 17.5 ml/kg/min (5 METs). If a bicycle ergometer is used, an exercise equivalent of 5 METs (e.g., 450 kpm/min, or 75 watts, for a 176 pound (80 kilogram) person) should be used. If the individual is able to complete this level of exercise without achieving listing-level hypoxemia, then he or she should be exercised at higher workloads to determine exercise capacity. A warm-up period of treadmill walking or cycling may be performed to acquaint the individual with the exercise procedure. If during the warm-up period the individual cannot achieve an exercise level of 5 METs, a lower workload may be selected in keeping with the estimate of exercise capacity. The individual should be monitored by ECG throughout the exercise and in the immediate post-exercise period. Blood pressure and an ECG should be recorded during each minute of exercise. During the final 2 minutes of a specific level of steady state exercise, an arterial blood sample should be drawn and analyzed for oxygen pressure (or tension) (PO_2), carbon dioxide pressure (or tension) (PCO_2), and pH. At the discretion of the testing facility, the sample may be obtained either from an indwelling arterial catheter or by direct arterial puncture. If possible, in order to evaluate exercise capacity more accurately, a test site should be selected that has the capability to measure minute ventilation, O_2 consumption, and carbon dioxide (CO_2) production. If the claimant fails to complete 4 to 6 minutes of steady state exercise, the testing laboratory should comment on the reason and report the actual duration and levels of exercise performed. This comment is necessary to determine if the individual's test performance was limited by lack of effort or other impairment (e.g., cardiac, peripheral vascular, musculoskeletal, neurological).

The exercise test report should contain representative ECG strips taken before, during and after exercise; resting and exercise arterial blood gas values; treadmill speed and grade settings, or, if a bicycle ergometer was used, exercise levels expressed in watts or kpm/min; and the duration of exercise. Body weight also should be recorded. If measured, O_2 consumption (STPD), minute ventilation (BTPS), and CO_2 production (STPD) also should be reported. The altitude of the test site, its normal range of blood gas values, and the barometric pressure on the test date must be noted.

G. *Chronic cor pulmonale and pulmonary vascular disease.* The establishment of an impairment attributable to irreversible cor pulmonale secondary to chronic pulmonary hypertension requires documentation by signs and laboratory findings of right ventricular overload or failure (e.g., an early diastolic right-sided gallop on auscultation, neck vein distension, hepatomegaly, peripheral edema, right ventricular outflow tract enlargement on x-ray or other appropriate imaging techniques, right ventricular hypertrophy on ECG, and increased pulmonary artery pressure measured by right heart catheterization available from treating sources). Cardiac catheterization will not be purchased. Because hypoxemia may accompany heart failure and is also a cause of pulmonary hypertension, and may be associated with hypoventilation and respiratory acidosis, arterial blood gases may demonstrate hypoxemia (decreased PO_2), CO_2 retention (increased PCO_2), and acidosis (decreased pH). Polycythemia with an elevated red blood cell count and hematocrit may be found in the presence of chronic hypoxemia.

P-pulmonale on the ECG does not establish chronic pulmonary hypertension or chronic cor pulmonale. Evidence of florid right heart failure need not be present at the time of adjudication for a listing (e.g., 3.09) to be satisfied, but the medical evidence of record should establish that cor pulmonale is chronic and irreversible.

H. *Sleep-related breathing disorders.* Sleep-related breathing disorders (sleep apneas) are caused by periodic cessation of respiration associated with hypoxemia and frequent

arousals from sleep. Although many individuals with one of these disorders will respond to prescribed treatment, in some, the disturbed sleep pattern and associated chronic nocturnal hypoxemia cause daytime sleepiness with chronic pulmonary hypertension and/or disturbances in cognitive function. Because daytime sleepiness can affect memory, orientation, and personality, a longitudinal treatment record may be needed to evaluate mental functioning. Not all individuals with sleep apnea develop a functional impairment that affects work activity. When any gainful work is precluded, the physiologic basis for the impairment may be chronic cor pulmonale. Chronic hypoxemia due to episodic apnea may cause pulmonary hypertension (see 3.00G and 3.09). Daytime somnolence may be associated with disturbance in cognitive vigilance. Impairment of cognitive function may be evaluated under organic mental disorders (12.02).

I. *Effects of obesity.* Obesity is a medically determinable impairment that is often associated with disturbance of the respiratory system, and disturbance of this system can be a major cause of disability in individuals with obesity. The combined effects of obesity with respiratory impairments can be greater than the effects of each of the impairments considered separately. Therefore, when determining whether an individual with obesity has a listing-level impairment or combination of impairments, and when assessing a claim at other steps of the sequential evaluation process, including when assessing an individual's residual functional capacity, adjudicators must consider any additional and cumulative effects of obesity.

3.01 Category of Impairments, Respiratory System.

3.02 *Chronic pulmonary insufficiency.*

A. Chronic obstructive pulmonary disease, due to any cause, with the FEV_1 equal to or less than the values specified in table I corresponding to the person's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

TABLE I

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
154 or less	60 or less	1.05
155–160	61–63	1.15
161–165	64–65	1.25
166–170	66–67	1.35
171–175	68–69	1.45
176–180	70–71	1.55
181 or more	72 or more	1.65

Or

B. Chronic restrictive ventilatory disease, due to any cause, with the FVC equal to or less than the values specified in table II corresponding to the person's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

TABLE II

Height without shoes (centimeters)	Height without shoes (inches)	FVC equal to or less than (L, BTPS)
154 or less	60 or less	1.25
155–160	61–63	1.35
161–165	64–65	1.45
166–170	66–67	1.55
171–175	68–69	1.65
176–180	70–71	1.75
181 or more	72 or more	1.85

Or

C. Chronic impairment of gas exchange due to clinically documented pulmonary disease. With:

1. Single breath DLCO (see 3.00F1) less than 10.5 ml/min/mm Hg or less than 40 percent of the predicted normal value. (Predicted values must either be based on data obtained at the test site or published values from a laboratory using the same technique as the test site. The source of the predicted values should be reported. If they are not published, they should be submitted in the form of a table or nomogram); or

2. Arterial blood gas values of PO_2 and simultaneously determined PCO_2 measured while at rest (breathing room air, awake and sitting or standing) in a clinically stable condition on at least two occasions, three or more weeks apart within a 6-month period, equal to or less than the values specified in the applicable table III-A or III-B or III-C:

TABLE III—A

[Applicable at test sites less than 3,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 equal to or less than (mm. Hg)
30 or below	65
31	64
32	63
33	62
34	61
35	60
36	59
37	58
38	57
39	56
40 or above	55

TABLE III—B

[Applicable at test sites 3,000 through 6,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 equal to or less than (mm. Hg)
30 or below	60
31	59
32	58
33	57
34	56
35	55
36	54
37	53
38	52
39	51
40 or above	50

TABLE III—C

[Applicable at test sites over 6,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 or equal to or less than (mm. Hg)
30 or below	55
31	54
32	53
33	52
34	51
35	50
36	49
37	48
38	47
39	46
40 or above	45

Or

3. Arterial blood gas values of PO_2 and simultaneously determined PCO_2 during steady state exercise breathing room air (level of exercise equivalent to or less than 17.5 ml O_2 consumption/kg/min or 5 METs) equal to or less than the values specified in the applicable table III-A or III-B or III-C in 3.02C2.

3.03 *Asthma*. With:

A. Chronic asthmatic bronchitis. Evaluate under the criteria for chronic obstructive pulmonary disease in 3.02A;

Or

B. Attacks (as defined in 3.00C), in spite of prescribed treatment and requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each in-patient hospitalization for longer than 24 hours for control of asthma counts as two attacks, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of attacks.

3.04 *Cystic fibrosis*. With:

A. An FEV_1 equal to or less than the appropriate value specified in table IV corresponding to the individual's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

Or

B. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;

Or

C. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased bacterial infection occurring at least once every 6 months and requiring intravenous or nebulization antimicrobial therapy.

TABLE IV

[Applicable only for evaluation under 3.04A—cystic fibrosis]

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
154 or less	60 or less	1.45
155–159	61–62	1.55
160–164	63–64	1.65
165–169	65–66	1.75
170–174	67–68	1.85
175–179	69–70	1.95
180 or more	71 or more	2.05

3.05 [Reserved]

3.06 *Pneumoconiosis* (demonstrated by appropriate imaging techniques). Evaluate under the appropriate criteria in 3.02.

3.07 *Bronchiectasis* (demonstrated by appropriate imaging techniques). With:

A. Impairment of pulmonary function due to extensive disease. Evaluate under the appropriate criteria in 3.02;

Or

B. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation of at least 12 consecutive months must be used to determine the frequency of episodes.

3.08 *Mycobacterial, mycotic, and other chronic persistent infections of the lung* (see 3.00B). Evaluate under the appropriate criteria in 3.02.

3.09 *Cor pulmonale secondary to chronic pulmonary vascular hypertension*. Clinical evidence of cor pulmonale (documented according to 3.00G) with:

A. Mean pulmonary artery pressure greater than 40 mm Hg;

Or

B. Arterial hypoxemia. Evaluate under the criteria in 3.02C2.

3.10 *Sleep-related breathing disorders.* Evaluate under 3.09 (chronic cor pulmonale) or 12.02 (organic mental disorders).

3.11 *Lung transplant.* Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment.

4.00 CARDIOVASCULAR SYSTEM

A. General

1. *What do we mean by a cardiovascular impairment?*

a. We mean any disorder that affects the proper functioning of the heart or the circulatory system (that is, arteries, veins, capillaries, and the lymphatic drainage). The disorder can be congenital or acquired.

b. Cardiovascular impairment results from one or more of four consequences of heart disease:

(i) Chronic heart failure or ventricular dysfunction.

(ii) Discomfort or pain due to myocardial ischemia, with or without necrosis of heart muscle.

(iii) Syncope, or near syncope, due to inadequate cerebral perfusion from any cardiac cause, such as obstruction of flow or disturbance in rhythm or conduction resulting in inadequate cardiac output.

(iv) Central cyanosis due to right-to-left shunt, reduced oxygen concentration in the arterial blood, or pulmonary vascular disease.

c. Disorders of the veins or arteries (for example, obstruction, rupture, or aneurysm) may cause impairments of the lower extremities (peripheral vascular disease), the central nervous system, the eyes, the kidneys, and other organs. We will evaluate peripheral vascular disease under 4.11 or 4.12 and impairments of another body system(s) under the listings for that body system(s).

2. *What do we consider in evaluating cardiovascular impairments?* The listings in this section describe cardiovascular impairments based on symptoms, signs, laboratory findings, response to a regimen of prescribed treatment, and functional limitations.

3. *What do the following terms or phrases mean in these listings?*

a. *Medical consultant* is an individual defined in §§404.1616(a) and 416.1016(a). This term does not include medical sources who provide consultative examinations for us. We use the abbreviation ''MC'' throughout this section to designate a medical consultant.

b. *Persistent* means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been present, or is expected to be present, for a continuous period of at least 12 months, such that a pattern of continuing severity is established.

c. *Recurrent* means that the longitudinal clinical record shows that, within a consecutive 12-month period, the finding(s) occurs at least three times, with intervening periods of improvement of sufficient duration that it is clear that separate events are involved.

d. *Appropriate medically acceptable imaging* means that the technique used is the proper one to evaluate and diagnose the impairment and is commonly recognized as accurate for assessing the cited finding.

e. *A consecutive 12-month period* means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with an application or continuing disability review.

f. *Uncontrolled* means the impairment does not adequately respond to standard prescribed medical treatment.

B. Documenting Cardiovascular Impairment

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular impairment. A longitudinal clinical record covering a period of not less than 3 months of observations and treatment is usually necessary, unless we can make a determination or decision based on the current evidence.

2. *Why is a longitudinal clinical record important?* We will usually need a longitudinal clinical record to assess the severity and expected duration of your impairment(s). If you have a listing-level impairment, you probably will have received medically prescribed treatment. Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your treating or other medical source. It should also include your response to this medical management, as well as information about the nature and severity of your impairment. The record will provide us with information on your functional status over an extended period of time and show whether your ability to function is improving, worsening, or unchanging.

3. *What if you have not received ongoing medical treatment?*

a. You may not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In this situation, we will base our evaluation on the current objective medical evidence and the other evidence we have. If you do not receive treatment, you cannot show an impairment that meets the criteria of most of these listings. However, we may find you disabled because you have another impairment(s) that

in combination with your cardiovascular impairment medically equals the severity of a listed impairment or based on consideration of your residual functional capacity and age, education, and work experience.

b. Unless we can decide your claim favorably on the basis of the current evidence, a longitudinal record is still important. In rare instances where there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the severity and duration of your impairment.

4. *When will we wait before we ask for more evidence?*

a. We will wait when we have information showing that your impairment is not yet stable and the expected change in your impairment might affect our determination or decision. In these situations, we need to wait to properly evaluate the severity and duration of your impairment during a stable period. Examples of when we might wait are:

(i) If you have had a recent acute event; for example, a myocardial infarction (heart attack).

(ii) If you have recently had a corrective cardiac procedure; for example, coronary artery bypass grafting.

(iii) If you have started new drug therapy and your response to this treatment has not yet been established; for example, beta-blocker therapy for dilated congestive cardiomyopathy.

b. In these situations, we will obtain more evidence 3 months following the event before we evaluate your impairment. However, we will not wait if we have enough information to make a determination or decision based on all of the relevant evidence in your case.

5. *Will we purchase any studies?* In appropriate situations, we will purchase studies necessary to substantiate the diagnosis or to document the severity of your impairment, generally after we have evaluated the medical and other evidence we already have. We will not purchase studies involving exercise testing if there is significant risk involved or if there is another medical reason not to perform the test. We will follow sections 4.00C6, 4.00C7, and 4.00C8 when we decide whether to purchase exercise testing.

6. *What studies will we not purchase?* We will not purchase any studies involving cardiac catheterization, such as coronary angiography, arteriograms, or electrophysiological studies. However, if the results of catheterization are part of the existing evidence we have, we will consider them together with the other relevant evidence. See 4.00C15a.

C. Using Cardiovascular Test Results

1. *What is an ECG?*

a. *ECG* stands for *electrocardiograph* or *electrocardiogram.* An electrocardiograph is a machine that records electrical impulses of your heart on a strip of paper called an electrocardiogram or a *tracing.* To record the ECG, a technician positions a number of small contacts (or *leads*) on your arms, legs, and across your chest to connect them to the ECG machine. An ECG may be done while you are resting or exercising.

b. The ECG tracing may indicate that you have a heart abnormality. It may indicate that your heart muscle is not getting as much oxygen as it needs (ischemia), that your heart rhythm is abnormal (arrhythmia), or that there are other abnormalities of your heart, such as left ventricular enlargement.

2. *How do we evaluate ECG evidence?* We consider a number of factors when we evaluate ECG evidence:

a. An original or legible copy of the 12-lead ECG obtained at rest must be appropriately dated and labeled, with the standardization inscribed on the tracing. Alteration in standardization of specific leads (such as to accommodate large QRS amplitudes) must be identified on those leads.

(i) Detailed descriptions or computer-averaged signals without original or legible copies of the ECG as described in listing 4.00C2a are not acceptable.

(ii) The effects of drugs or electrolyte abnormalities must be considered as possible noncardiac causes of ECG abnormalities of ventricular repolarization; that is, those involving the ST segment and T wave. If available, the predrug (especially digitalis glycosides) ECG should be submitted.

b. ECGs obtained in conjunction with treadmill, bicycle, or arm exercise tests should meet the following specifications:

(i) ECG reports must include the original calibrated ECG tracings or a legible copy.

(ii) A 12-lead baseline ECG must be recorded in the upright position before exercise.

(iii) A 12-lead ECG should be recorded at the end of each minute of exercise.

(iv) If ECG documentation of the effects of hyperventilation is obtained, the exercise test should be deferred for at least 10 minutes because metabolic changes of hyperventilation may alter the physiologic and ECG-recorded response to exercise.

(v) Post-exercise ECGs should be recorded using a generally accepted protocol consistent with the prevailing state of medical knowledge and clinical practice.

(vi) All resting, exercise, and recovery ECG strips must have the standardization inscribed on the tracing. The ECG strips should be labeled to indicate the date, the times recorded and the relationship to the stage of the exercise protocol. The speed and grade (treadmill test) or work rate (bicycle or arm ergometric test) should be recorded. The highest level of exercise achieved, heart rate and blood pressure levels during testing, and the reason(s) for terminating the test

(including limiting signs or symptoms) must be recorded.

3. *What are exercise tests and what are they used for?*

a. Exercise tests have you perform physical activity and record how your cardiovascular system responds. Exercise tests usually involve walking on a treadmill, but other forms of exercise, such as an exercise bicycle or an arm exercise machine, may be used. Exercise testing may be done for various reasons; such as to evaluate the severity of your coronary artery disease or peripheral vascular disease or to evaluate your progress after a cardiac procedure or an acute event, like a myocardial infarction (heart attack). Exercise testing is the most widely used testing for identifying the presence of myocardial ischemia and for estimating maximal aerobic capacity (usually expressed in METs—metabolic equivalents) if you have heart disease.

b. We include exercise tolerance test (ETT) criteria in 4.02B3 (chronic heart failure) and 4.04A (ischemic heart disease). To meet the ETT criteria in these listings, the ETT must be a sign-or symptom-limited test in which you exercise while connected to an ECG until you develop a sign or symptom that indicates that you have exercised as much as is considered safe for you.

c. In 4.12B, we also refer to exercise testing for peripheral vascular disease. In this test, you walk on a treadmill, usually for a specified period of time, and the individual who administers the test measures the effect of exercise on the flow of blood in your legs, usually by using ultrasound. The test is also called an exercise Doppler test. Even though this test is intended to evaluate peripheral vascular disease, it will be stopped for your safety if you develop abnormal signs or symptoms because of heart disease.

d. Each type of test is done in a certain way following specific criteria, called a *protocol*. For our program, we also specify certain aspects of how any exercise test we purchase is to be done. See 4.00C10 and 4.00C17.

4. *Do ETTs have limitations?* An ETT provides an estimate of aerobic capacity for walking on a grade, bicycling, or moving one's arms in an environmentally controlled setting. Therefore, ETT results do not correlate with the ability to perform other types of exertional activities, such as lifting and carrying heavy loads, and do not provide an estimate of the ability to perform activities required for work in all possible work environments or throughout a workday. Also, certain medications (such as beta blockers) and conduction disorders (such as left or right bundle branch blocks) can cause false-negative or false-positive results. Therefore, we must consider the results of an ETT together with all the other relevant evidence in your case record.

5. *How does an ETT with measurement of maximal or peak oxygen uptake VO_2) differ from other ETTs?* Occasionally, medical evidence will include the results of an ETT with VO_2 While ETTs without measurement of VO_2 provide only an estimate of aerobic capacity, measured maximal or peak oxygen uptake provides an accurate measurement of aerobic capacity, which is often expressed in METs (metabolic equivalents). The MET level may not be indicated in the report of attained maximal or peak VO_2 testing, but can be calculated as follows: 1 MET = 3.5 milliliters (ml) of oxygen uptake per kilogram (kg) of body weight per minute. For example, a 70 kg (154 lb.) individual who achieves a maximal or peak VO_2 of 1225 ml in 1 minute has attained 5 METs (1225 ml/70 kg/1 min = 17.5 ml/kg/min. 17.5/3.5 = 5 METs).

6. *When will we consider whether to purchase an exercise test?*

a. We will consider whether to purchase an exercise test when:

(i) There is a question whether your cardiovascular impairment meets or medically equals the severity of one of the listings, or there is no timely test in the evidence we have (see 4.00C9), and we cannot find you disabled on some other basis; or

(ii) We need to assess your residual functional capacity and there is insufficient evidence in the record to make a determination or decision.

b. We will not purchase an exercise test when we can make our determination or decision based on the evidence we already have.

7. *What must we do before purchasing an exercise test?*

a. Before we purchase an exercise test, an MC, preferably one with experience in the care of patients with cardiovascular disease, must review the pertinent history, physical examinations, and laboratory tests that we have to determine whether the test would present a significant risk to you or if there is some other medical reason not to purchase the test (see 4.00C8).

b. If you are under the care of a treating source (see §§404.1502 and 416.902) for a cardiovascular impairment, this source has not performed an exercise test, and there are no reported significant risks to testing, we will request a statement from that source explaining why it was not done or should not be done before we decide whether we will purchase the test.

c. The MC, in accordance with the regulations and other instructions on consultative examinations, will generally give great weight to the treating source's opinion about the risk of exercise testing to you and will generally not override it. In the rare situation in which the MC does override the treating source's opinion, the MC must prepare a written rationale documenting the reasons for overriding the opinion.

d. If you do not have a treating source or we cannot obtain a statement from your treating source, the MC is responsible for assessing the risk to exercise testing based on a review of the records we have before purchasing an exercise test for you.

e. We must also provide your records to the medical source who performs the exercise test for review prior to conducting the test if the source does not already have them. The medical source who performs the exercise test has the ultimate responsibility for deciding whether you would be at risk.

8. *When will we not purchase an exercise test or wait before we purchase an exercise test?*

a. We will not purchase an exercise test when an MC finds that you have one of the following significant risk factors:

(i) Unstable angina not previously stabilized by medical treatment.

(ii) Uncontrolled cardiac arrhythmias causing symptoms or hemodynamic compromise.

(iii) An implanted cardiac defibrillator.

(iv) Symptomatic severe aortic stenosis.

(v) Uncontrolled symptomatic heart failure.

(vi) Aortic dissection.

(vii) Severe pulmonary hypertension (pulmonary artery systolic pressure greater than 60 mm Hg).

(viii) Left main coronary stenosis of 50 percent or greater that has not been bypassed.

(ix) Moderate stenotic valvular disease with a systolic gradient across the aortic valve of 50 mm Hg or greater.

(x) Severe arterial hypertension (systolic greater than 200 mm Hg or diastolic greater than 110 mm Hg).

(xi) Hypertrophic cardiomyopathy with a systolic gradient of 50 mm Hg or greater.

b. We also will not purchase an exercise test when you are prevented from performing exercise testing due to another impairment affecting your ability to use your arms and legs.

c. We will not purchase an ETT to document the presence of a cardiac arrhythmia.

d. We will wait to purchase an exercise test until 3 months after you have had one of the following events. This will allow for maximal, attainable restoration of functional capacity.

(i) Acute myocardial infarction.

(ii) Surgical myocardial revascularization (bypass surgery).

(iii) Other open-heart surgical procedures.

(iv) Percutaneous transluminal coronary angioplasty with or without stenting.

e. If you are deconditioned after an extended period of bedrest or inactivity and could improve with activity, or if you are in acute heart failure and are expected to improve with treatment, we will wait an appropriate period of time for you to recuperate before we purchase an exercise test.

9. *What do we mean by a ''timely'' test?*

a. We consider exercise test results to be timely for 12 months after the date they are performed, provided there has been no change in your clinical status that may alter the severity of your cardiovascular impairment.

b. However, an exercise test that is older than 12 months, especially an abnormal one, can still provide information important to our adjudication. For example, a test that is more than 12 months old can provide evidence of ischemic heart disease or peripheral vascular disease, information on decreased aerobic capacity, or information about the duration or onset of your impairment. Such tests can be an important component of the longitudinal record.

c. When we evaluate a test that is more than 12 months old, we must consider the results in the context of all the relevant evidence, including why the test was performed and whether there has been an intervening event or improvement or worsening of your impairment.

d. We will purchase a new exercise test only if we cannot make a determination or decision based on the evidence we have.

10. *How must ETTs we purchase be performed?*

a. The ETT must be a sign- or symptom-limited test characterized by a progressive multistage regimen. It must be performed using a generally accepted protocol consistent with the prevailing state of medical knowledge and clinical practice. A description of the protocol that was followed must be provided, and the test must meet the requirements of 4.00C2b and this section. A radionuclide perfusion scan may be useful for detecting or confirming ischemia when resting ECG abnormalities, medications, or other factors may decrease the accuracy of ECG interpretation of ischemia. (The perfusion imaging is done at the termination of exercise, which may be at a higher MET level than that at which ischemia first occurs. If the imaging confirms the presence of reversible ischemia, the exercise ECG may be useful for detecting the MET level at which ischemia initially appeared.) Exercise tests may also be performed using echocardiography to detect stress-induced ischemia and left ventricular dysfunction (see 4.00C12 and 4.00C13).

b. The exercise test must be paced to your capabilities and be performed following the generally accepted standards for adult exercise test laboratories. With a treadmill test, the speed, grade (incline), and duration of exercise must be recorded for each exercise test stage performed. Other exercise test protocols or techniques should use similar workloads. The exercise protocol may need to be modified in individual cases to allow for a lower initial workload with more slowly graded increments than the standard Bruce protocol.

c. Levels of exercise must be described in terms of workload and duration of each stage; for example, treadmill speed and grade, or bicycle ergometer work rate in kpm/min or watts.

d. The exercise laboratory's physical environment, staffing, and equipment must meet the generally accepted standards for adult exercise test laboratories.

11. *How do we evaluate ETT results?* We evaluate ETT results on the basis of the work level at which the test becomes abnormal, as documented by onset of signs or symptoms and any ECG or imaging abnormalities. The absence of an ischemic response on an ETT alone does not exclude the diagnosis of ischemic heart disease. We must consider the results of an ETT in the context of all of the other evidence in your case record.

12. *When are ETTs done with imaging?* When resting ECG abnormalities preclude interpretation of ETT tracings relative to ischemia, a radionuclide (for example, thallium-201 or technetium-99m) perfusion scan or echocardiography in conjunction with an ETT provides better results. You may have resting ECG abnormalities when you have a conduction defect—for example, Wolff-Parkinson-White syndrome, left bundle branch block, left ventricular hypertrophy—or when you are taking digitalis or other antiarrhythmic drugs, or when resting ST changes are present. Also, these techniques can provide a reliable estimate of ejection fraction.

13. *Will we purchase ETTs with imaging?* We may purchase an ETT with imaging in your case after an MC, preferably one with experience in the care of patients with cardiovascular disease, has reviewed your medical history and physical examination, any report(s) of appropriate medically acceptable imaging, ECGs, and other appropriate tests. We will consider purchasing an ETT with imaging when other information we have is not adequate for us to assess whether you have severe ventricular dysfunction or myocardial ischemia, there is no significant risk involved (see 4.00C8a), and we cannot make our determination or decision based on the evidence we already have.

14. *What are drug-induced stress tests?* These tests are designed primarily to provide evidence about myocardial ischemia or prior myocardial infarction, but do not require you to exercise. These tests are used when you cannot exercise or cannot exercise enough to achieve the desired cardiac stress. Drug-induced stress tests can also provide evidence about heart chamber dimensions and function; however, these tests do not provide information about your aerobic capacity and cannot be used to help us assess your ability to function. Some of these tests use agents, such as Persantine or adenosine, that dilate the coronary arteries and are used in combination with nuclear agents, such as thallium or technetium (for example, Cardiolyte or Myoview), and a myocardial scan. Other tests use agents, such as dobutamine, that stimulate the heart to contract more forcefully and faster to simulate exercise and are used in combination with a 2-dimensional echocardiogram. We may, when appropriate, purchase a drug-induced stress test to confirm the presence of myocardial ischemia after a review of the evidence in your file by an MC, preferably one with experience in the care of patients with cardiovascular disease.

15. *How do we evaluate cardiac catheterization evidence?*

a. We will not purchase cardiac catheterization; however, if you have had catheterization, we will make every reasonable effort to obtain the report and any ancillary studies. We will consider the quality and type of data provided and its relevance to the evaluation of your impairment. For adults, we generally see two types of catheterization reports: Coronary arteriography and left ventriculography.

b. For coronary arteriography, the report should provide information citing the method of assessing coronary arterial lumen diameter and the nature and location of obstructive lesions. Drug treatment at baseline and during the procedure should be reported. Some individuals with significant coronary atherosclerotic obstruction have collateral vessels that supply the myocardium distal to the arterial obstruction so that there is no evidence of myocardial damage or ischemia, even with exercise. When the results of quantitative computer measurements and analyses are included in your case record, we will consider them in interpreting the severity of stenotic lesions.

c. For left ventriculography, the report should describe the wall motion of the myocardium with regard to any areas of hypokinesis (abnormally decreased motion), akinesis (lack of motion), or dyskinesis (distortion of motion), and the overall contraction of the ventricle as measured by the ejection fraction. Measurement of chamber volumes and pressures may be useful. Quantitative computer analysis provides precise measurement of segmental left ventricular wall thickness and motion. There is often a poor correlation between left ventricular function at rest and functional capacity for physical activity.

16. *What details should exercise Doppler test reports contain?* The reports of exercise Doppler tests must describe the level of exercise; for example, the speed and grade of the treadmill settings, the duration of exercise, symptoms during exercise, and the reasons for stopping exercise if the expected level of exercise was not attained. They must also include the blood pressures at the ankle and other pertinent sites measured after exercise

and the time required for the systolic blood pressure to return toward or to the pre-exercise level. The graphic tracings, if available, should also be included with the report. All tracings must be annotated with the standardization used by the testing facility.

17. *How must exercise Doppler tests we purchase be performed?* When we purchase an exercise Doppler test, you must exercise on a treadmill at 2 mph on a 12 percent grade for up to 5 minutes. The reports must include the information specified in 4.00C16. Because this is an exercise test, we must evaluate whether such testing would put you at significant risk, in accordance with the guidance found in 4.00C6, 4.00C7, and 4.00C8.

D. Evaluating Chronic Heart Failure

1. *What is chronic heart failure (CHF)?*

a. *CHF* is the inability of the heart to pump enough oxygenated blood to body tissues. This syndrome is characterized by symptoms and signs of pulmonary or systemic congestion (fluid retention) or limited cardiac output. Certain laboratory findings of cardiac functional and structural abnormality support the diagnosis of CHF. There are two main types of CHF:

(i) *Predominant systolic dysfunction* (the inability of the heart to contract normally and expel sufficient blood), which is characterized by a dilated, poorly contracting left ventricle and reduced ejection fraction (abbreviated EF, it represents the percentage of the blood in the ventricle actually pumped out with each contraction), and

(ii) *Predominant diastolic dysfunction* (the inability of the heart to relax and fill normally), which is characterized by a thickened ventricular muscle, poor ability of the left ventricle to distend, increased ventricular filling pressure, and a normal or increased EF.

b. CHF is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, or rheumatic, congenital, or other heart disease. However, if the CHF is the result of primary pulmonary hypertension secondary to disease of the lung (cor pulmonale), we will evaluate your impairment using 3.09, in the respiratory system listings.

2. *What evidence of CHF do we need?*

a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization.

(i) Abnormal cardiac imaging showing increased left ventricular end diastolic diameter (LVEDD), decreased EF, increased left atrial chamber size, increased ventricular filling pressures measured at cardiac catheterization, or increased left ventricular wall or septum thickness, provides objective measures of both left ventricular function and structural abnormality in heart failure.

(ii) An LVEDD greater than 6.0 cm or an EF of 30 percent or less measured during a period of stability (that is, not during an episode of acute heart failure) may be associated clinically with systolic failure.

(iii) Left ventricular posterior wall thickness added to septal thickness totaling 2.5 cm or greater with left atrium enlarged to 4.5 cm or greater may be associated clinically with diastolic failure.

(iv) However, these measurements alone do not reflect your functional capacity, which we evaluate by considering all of the relevant evidence. In some situations, we may need to purchase an ETT to help us assess your functional capacity.

(v) Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema. These findings need not be present on each report, since CHF may be controlled by prescribed treatment.

b. To establish that you have *chronic* heart failure, your medical history and physical examination should describe characteristic symptoms and signs of pulmonary or systemic congestion or of limited cardiac output associated with the abnormal findings on appropriate medically acceptable imaging. When an acute episode of heart failure is triggered by a remediable factor, such as an arrhythmia, dietary sodium overload, or high altitude, cardiac function may be restored and a chronic impairment may not be present.

(i) Symptoms of congestion or of limited cardiac output include easy fatigue, weakness, shortness of breath (dyspnea), cough, or chest discomfort at rest or with activity. Individuals with CHF may also experience shortness of breath on lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). They may also experience cardiac arrhythmias resulting in palpitations, lightheadedness, or fainting.

(ii) Signs of congestion may include hepatomegaly, ascites, increased jugular venous distention or pressure, rales, peripheral edema, or rapid weight gain. However, these signs need not be found on all examinations because fluid retention may be controlled by prescribed treatment.

3. *Is it safe for you to have an ETT, if you have CHF?* The presence of CHF is not necessarily a contraindication to an ETT, unless you are having an acute episode of heart failure. Measures of cardiac performance are valuable in helping us evaluate your ability to do work-related activities. Exercise testing has been safely used in individuals with CHF; therefore, we may purchase an ETT for evaluation under 4.02B3 if an MC, preferably one experienced in the care of patients with

cardiovascular disease, determines that there is no significant risk to you. (See 4.00C6 for when we will consider the purchase of an ETT. See 4.00C7–4.00C8 for what we must do before we purchase an ETT and when we will not purchase one.) ST segment changes from digitalis use in the treatment of CHF do not preclude the purchase of an ETT.

4. *How do we evaluate CHF using 4.02?*

a. We must have objective evidence, as described in 4.00D2, that you have chronic heart failure.

b. To meet the required level of severity for this listing, your impairment must satisfy the requirements of one of the criteria in A and one of the criteria in B.

c. In 4.02B2, the phrase *periods of stabilization* means that, for at least 2 weeks between episodes of acute heart failure, there must be objective evidence of clearing of the pulmonary edema or pleural effusions and evidence that you returned to, or you were medically considered able to return to, your prior level of activity.

d. Listing 4.02B3c requires a decrease in systolic blood pressure below the baseline level (taken in the standing position immediately prior to exercise) or below any systolic pressure reading recorded during exercise. This is because, normally, systolic blood pressure and heart rate increase gradually with exercise. Decreases in systolic blood pressure below the baseline level that occur during exercise are often associated with ischemia-induced left ventricular dysfunction resulting in decreased cardiac output. However, a blunted response (that is, failure of the systolic blood pressure to rise 10 mm Hg or more), particularly in the first 3 minutes of exercise, may be drug-related and is not necessarily associated with left ventricular dysfunction. Also, some individuals with increased sympathetic responses because of deconditioning or apprehension may increase their systolic blood pressure and heart rate above their baseline level just before and early into exercise. This can be associated with a drop in systolic pressure in early exercise that is not due to left ventricular dysfunction. Therefore, an early decrease in systolic blood pressure must be interpreted within the total context of the test; that is, the presence or absence of symptoms such as lightheadedness, ischemic changes, or arrhythmias on the ECG.

E. Evaluating Ischemic Heart Disease

1. *What is ischemic heart disease (IHD)? IHD* results when one or more of your coronary arteries is narrowed or obstructed or, in rare situations, constricted due to vasospasm, interfering with the normal flow of blood to your heart muscle (ischemia). The obstruction may be the result of an embolus, a thrombus, or plaque. When heart muscle tissue dies as a result of the reduced blood supply, it is called a myocardial infarction (heart attack).

2. *What causes chest discomfort of myocardial origin?*

a. Chest discomfort of myocardial ischemic origin, commonly known as angina pectoris, is usually caused by coronary artery disease (often abbreviated CAD). However, ischemic discomfort may be caused by a noncoronary artery impairment, such as aortic stenosis, hypertrophic cardiomyopathy, pulmonary hypertension, or anemia.

b. Instead of typical angina pectoris, some individuals with IHD experience atypical angina, anginal equivalent, variant angina, or silent ischemia, all of which we may evaluate using 4.04. We discuss the various manifestations of ischemia in 4.00E3–4.00E7.

3. *What are the characteristics of typical angina pectoris?* Discomfort of myocardial ischemic origin (angina pectoris) is discomfort that is precipitated by effort or emotion and promptly relieved by rest, sublingual nitroglycerin (that is, nitroglycerin tablets that are placed under the tongue), or other rapidly acting nitrates. Typically, the discomfort is located in the chest (usually substernal) and described as pressing, crushing, squeezing, burning, aching, or oppressive. Sharp, sticking, or cramping discomfort is less common. Discomfort occurring with activity or emotion should be described specifically as to timing and usual inciting factors (type and intensity), character, location, radiation, duration, and response to nitrate treatment or rest.

4. *What is atypical angina? Atypical angina* describes discomfort or pain from myocardial ischemia that is felt in places other than the chest. The common sites of cardiac pain are the inner aspect of the left arm, neck, jaw(s), upper abdomen, and back, but the discomfort or pain can be elsewhere. When pain of cardiac ischemic origin presents in an atypical site in the absence of chest discomfort, the source of the pain may be difficult to diagnose. To represent atypical angina, your discomfort or pain should have precipitating and relieving factors similar to those of typical chest discomfort, and we must have objective medical evidence of myocardial ischemia; for example, ECG or ETT evidence or appropriate medically acceptable imaging.

5. *What is anginal equivalent?* Often, individuals with IHD will complain of shortness of breath (dyspnea) on exertion without chest pain or discomfort. In a minority of such situations, the shortness of breath is due to myocardial ischemia; this is called *anginal equivalent*. To represent anginal equivalent, your shortness of breath should have precipitating and relieving factors similar to those of typical chest discomfort, and we must have objective medical evidence of myocardial ischemia; for example, ECG or ETT evidence or appropriate medically acceptable

imaging. In these situations, it is essential to establish objective evidence of myocardial ischemia to ensure that you do not have effort dyspnea due to non-ischemic or non-cardiac causes.

6. *What is variant angina?*

a. *Variant angina* (Prinzmetal's angina, vasospastic angina) refers to the occurrence of anginal episodes at rest, especially at night, accompanied by transitory ST segment elevation (or, at times, ST depression) on an ECG. It is due to severe spasm of a coronary artery, causing ischemia of the heart wall, and is often accompanied by major ventricular arrhythmias, such as ventricular tachycardia. We will consider variant angina under 4.04 only if you have spasm of a coronary artery in relation to an obstructive lesion of the vessel. If you have an arrhythmia as a result of variant angina, we may consider your impairment under 4.05.

b. Variant angina may also occur in the absence of obstructive coronary disease. In this situation, an ETT will not demonstrate ischemia. The diagnosis will be established by showing the typical transitory ST segment changes during attacks of pain, and the absence of obstructive lesions shown by catheterization. Treatment in cases where there is no obstructive coronary disease is limited to medications that reduce coronary vasospasm, such as calcium channel blockers and nitrates. In such situations, we will consider the frequency of anginal episodes despite prescribed treatment when evaluating your residual functional capacity.

c. Vasospasm that is catheter-induced during coronary angiography is not variant angina.

7. *What is silent ischemia?*

a. Myocardial ischemia, and even myocardial infarction, can occur without perception of pain or any other symptoms; when this happens, we call it *silent ischemia*. Pain sensitivity may be altered by a variety of diseases, most notably diabetes mellitus and other neuropathic disorders. Individuals also vary in their threshold for pain.

b. Silent ischemia occurs most often in:

(i) Individuals with documented past myocardial infarction or established angina without prior infarction who do not have chest pain on ETT, but have a positive test with ischemic abnormality on ECG, perfusion scan, or other appropriate medically acceptable imaging.

(ii) Individuals with documented past myocardial infarction or angina who have ST segment changes on ambulatory monitoring (Holter monitoring) that are similar to those that occur during episodes of angina. ST depression shown on the ambulatory recording should not be interpreted as positive for ischemia unless similar depression is also seen during chest pain episodes annotated in the diary that the individual keeps while wearing the Holter monitor.

c. ST depression can result from a variety of factors, such as postural changes and variations in cardiac sympathetic tone. In addition, there are differences in how different Holter monitors record the electrical responses. Therefore, we do not consider the Holter monitor reliable for the diagnosis of silent ischemia except in the situation described in 4.00E7b(ii).

8. *What other sources of chest discomfort are there?* Chest discomfort of nonischemic origin may result from other cardiac impairments, such as pericarditis. Noncardiac impairments may also produce symptoms mimicking that of myocardial ischemia. These impairments include acute anxiety or panic attacks, gastrointestinal tract disorders, such as esophageal spasm, esophagitis, hiatal hernia, biliary tract disease, gastritis, peptic ulcer, and pancreatitis, and musculoskeletal syndromes, such as chest wall muscle spasm, chest wall syndrome (especially after coronary bypass surgery), costochondritis, and cervical or dorsal spine arthritis. Hyperventilation may also mimic ischemic discomfort. Thus, in the absence of documented myocardial ischemia, such disorders should be considered as possible causes of chest discomfort.

9. *How do we evaluate IHD using 4.04?*

a. We must have objective evidence, as described under 4.00C, that your symptoms are due to myocardial ischemia.

b. Listing-level changes on the ECG in 4.04A1 are the classically accepted changes of horizontal or downsloping ST depression occurring both during exercise and recovery. Although we recognize that ischemic changes may at times occur only during exercise or recovery, and may at times be upsloping with only junctional ST depression, such changes can be false positive; that is, occur in the absence of ischemia. Diagnosis of ischemia in this situation requires radionuclide or echocardiogram confirmation. See 4.00C12 and 4.00C13.

c. Also in 4.04A1, we require that the depression of the ST segment last for at least 1 minute of recovery because ST depression that occurs during exercise but that rapidly normalizes in recovery is a common false-positive response.

d. In 4.04A2, we specify that the ST elevation must be in non-infarct leads during both exercise and recovery. This is because, in the absence of ECG signs of prior infarction, ST elevation during exercise denotes ischemia, usually severe, requiring immediate termination of exercise. However, if there is baseline ST elevation in association with a prior infarction or ventricular aneurysm, further ST elevation during exercise does not necessarily denote ischemia and could be a false-positive ECG response. Diagnosis of ischemia in this situation requires radionuclide or echocardiogram confirmation. See 4.00C12 and 4.00C13.

e. Listing 4.04A3 requires a decrease in systolic blood pressure below the baseline level (taken in the standing position immediately prior to exercise) or below any systolic pressure reading recorded during exercise. This is the same finding required in 4.02B3c. See 4.00D4d for full details.

f. In 4.04B, each of the three ischemic episodes must require revascularization or be not amenable to treatment. *Revascularization* means angioplasty (with or without stent placement) or bypass surgery. However, reocclusion that occurs after a revascularization procedure but during the same hospitalization and that requires a second procedure during the same hospitalization will not be counted as another ischemic episode. Not amenable means that the revascularization procedure could not be done because of another medical impairment or because the vessel was not suitable for revascularization.

g. We will use 4.04C only when you have symptoms due to myocardial ischemia as described in 4.00E3–4.00E7 while on a regimen of prescribed treatment, you are at risk for exercise testing (see 4.00C8), and we do not have a timely ETT or a timely normal drug-induced stress test for you. See 4.00C9 for what we mean by a timely test.

h. In 4.04C1 the term *nonbypassed* means that the blockage is in a vessel that is potentially bypassable; that is, large enough to be bypassed and considered to be a cause of your ischemia. These vessels are usually major arteries or one of a major artery's major branches. A vessel that has become obstructed again after angioplasty or stent placement and has remained obstructed or is not amenable to another revascularization is considered a nonbypassed vessel for purposes of this listing. When you have had revascularization, we will not use the pre-operative findings to assess the current severity of your coronary artery disease under 4.04C, although we will consider the severity and duration of your impairment prior to your surgery in making our determination or decision.

F. Evaluating Arrhythmias

1. *What is an arrhythmia?* An *arrhythmia* is a change in the regular beat of the heart. Your heart may seem to skip a beat or beat irregularly, very quickly (tachycardia), or very slowly (bradycardia).

2. *What are the different types of arrhythmias?*

a. There are many types of arrhythmias. Arrhythmias are identified by where they occur in the heart (atria or ventricles) and by what happens to the heart's rhythm when they occur.

b. Arrhythmias arising in the cardiac atria (upper chambers of the heart) are called atrial or supraventricular arrhythmias. Ventricular arrhythmias begin in the ventricles (lower chambers). In general, ventricular arrhythmias caused by heart disease are the most serious.

3. *How do we evaluate arrhythmias using 4.05?*

a. We will use 4.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator and you have uncontrolled recurrent episodes of syncope or near syncope. If your arrhythmias are controlled, we will evaluate your underlying heart disease using the appropriate listing. For other considerations when we evaluate arrhythmias in the presence of an implanted cardiac defibrillator, see 4.00F4.

b. We consider *near syncope* to be a period of altered consciousness, since syncope is a loss of consciousness or a faint. It is not merely a feeling of light-headedness, momentary weakness, or dizziness.

c. For purposes of 4.05, there must be a documented association between the syncope or near syncope and the recurrent arrhythmia. The recurrent arrhythmia, not some other cardiac or non-cardiac disorder, must be established as the cause of the associated symptom. This documentation of the association between the symptoms and the arrhythmia may come from the usual diagnostic methods, including Holter monitoring (also called ambulatory electrocardiography) and tilt-table testing with a concurrent ECG. Although an arrhythmia may be a coincidental finding on an ETT, we will not purchase an ETT to document the presence of a cardiac arrhythmia.

4. *What will we consider when you have an implanted cardiac defibrillator and you do not have arrhythmias that meet the requirements of 4.05?*

a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in individuals who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group at risk for sudden cardiac death consists of individuals with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in individuals with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator is a unique form of treatment; it rescues an individual from what may have been cardiac arrest. However, as a consequence of the shock(s), individuals may experience psychological distress, which we may evaluate under the mental disorders listings in 12.00ff.

b. Most implantable cardiac defibrillators have rhythm-correcting and pacemaker capabilities. In some individuals, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. (The shock is like being kicked in the chest.) Implanted cardiac

defibrillators may deliver inappropriate shocks, often repeatedly, in response to benign arrhythmias or electrical malfunction. Also, exposure to strong electrical or magnetic fields, such as from MRI (magnetic resonance imaging), can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. We must consider the frequency of, and the reason(s) for, the shocks when evaluating the severity and duration of your impairment.

c. In general, the exercise limitations imposed on individuals with an implanted cardiac defibrillator are those dictated by the underlying heart impairment. However, the exercise limitations may be greater when the implanted cardiac defibrillator delivers an inappropriate shock in response to the increase in heart rate with exercise, or when there is exercise-induced ventricular arrhythmia.

G. Evaluating Peripheral Vascular Disease

1. *What is peripheral vascular disease (PVD)?* Generally, *PVD* is any impairment that affects either the arteries (peripheral arterial disease) or the veins (venous insufficiency) in the extremities, particularly the lower extremities. The usual effect is blockage of the flow of blood either from the heart (arterial) or back to the heart (venous). If you have peripheral arterial disease, you may have pain in your calf after walking a distance that goes away when you rest (intermittent claudication); at more advanced stages, you may have pain in your calf at rest or you may develop ulceration or gangrene. If you have venous insufficiency, you may have swelling, varicose veins, skin pigmentation changes, or skin ulceration.

2. *How do we assess limitations resulting from PVD?* We will assess your limitations based on your symptoms together with physical findings, Doppler studies, other appropriate non-invasive studies, or angiographic findings. However, if the PVD has resulted in amputation, we will evaluate any limitations related to the amputation under the musculoskeletal listings, 1.00ff.

3. *What is brawny edema? Brawny edema* (4.11A) is swelling that is usually dense and feels firm due to the presence of increased connective tissue; it is also associated with characteristic skin pigmentation changes. It is not the same thing as pitting edema. Brawny edema generally does not pit (indent on pressure), and the terms are not interchangeable. Pitting edema does not satisfy the requirements of 4.11A.

4. *What is lymphedema and how will we evaluate it?*

a. *Lymphedema* is edema of the extremities due to a disorder of the lymphatic circulation; at its worst, it is called elephantiasis. Primary lymphedema is caused by abnormal development of lymph vessels and may be present at birth (congenital lymphedema), but more often develops during the teens (lymphedema praecox). It may also appear later, usually after age 35 (lymphedema tarda). Secondary lymphedema is due to obstruction or destruction of normal lymphatic channels due to tumor, surgery, repeated infections, or parasitic infection such as filariasis. Lymphedema most commonly affects one extremity.

b. Lymphedema does not meet the requirements of 4.11, although it may medically equal the severity of that listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular listing, such as 4.11, or a musculoskeletal listing, such as 1.02A or 1.03. If no listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we assess your residual functional capacity.

5. *When will we purchase exercise Doppler studies for evaluating peripheral arterial disease (PAD)?* If we need additional evidence of your PAD, we will generally purchase exercise Doppler studies (see 4.00C16 and 4.00C17) when your resting ankle/brachial systolic blood pressure ratio is at least 0.50 but less than 0.80, and only rarely when it is 0.80 or above. We will not purchase exercise Doppler testing if you have a disease that results in abnormal arterial calcification or small vessel disease, but will use your resting toe systolic blood pressure or resting toe/brachial systolic blood pressure ratio. (See 4.00G7c and 4.00G8.) There are no current medical standards for evaluating exercise toe pressures. Because any exercise test stresses your entire cardiovascular system, we will purchase exercise Doppler studies only after an MC, preferably one with experience in the care of patients with cardiovascular disease, has determined that the test would not present a significant risk to you and that there is no other medical reason not to purchase the test (see 4.00C6, 4.00C7, and 4.00C8).

6. *Are there any other studies that are helpful in evaluating PAD?* Doppler studies done using a recording ultrasonic Doppler unit and strain-gauge plethysmography are other useful tools for evaluating PAD. A recording Doppler, which prints a tracing of the arterial pulse wave in the femoral, popliteal, dorsalis pedis, and posterior tibial arteries, is an excellent evaluation tool to compare wave forms in normal and compromised peripheral blood flow. Qualitative analysis of the pulse wave is very helpful in the overall assessment of the severity of the occlusive disease. Tracings are especially helpful in assessing severity if you have small vessel disease related to diabetes mellitus or other diseases with similar vascular changes, or diseases causing medial calcifications when ankle pressure is either normal or falsely high.

7. *How do we evaluate PAD under 4.12?*

a. The ankle blood pressure referred to in 4.12A and B is the higher of the pressures recorded from the posterior tibial and dorsalis pedis arteries in the affected leg. The higher pressure recorded from the two sites is the more significant measurement in assessing the extent of arterial insufficiency. Techniques for obtaining ankle systolic blood pressures include Doppler (See 4.00C16 and 4.00C17), plethysmographic studies, or other techniques. We will request any available tracings generated by these studies so that we can review them.

b. In 4.12A, the ankle/brachial systolic blood pressure ratio is the ratio of the systolic blood pressure at the ankle to the systolic blood pressure at the brachial artery; both taken at the same time while you are lying on your back. We do not require that the ankle and brachial pressures be taken on the same side of your body. This is because, as with the ankle pressure, we will use the higher brachial systolic pressure measured. Listing 4.12A is met when your resting ankle/brachial systolic blood pressure ratio is less than 0.50. If your resting ankle/brachial systolic blood pressure ratio is 0.50 or above, we will use 4.12B to evaluate the severity of your PAD, unless you also have a disease causing abnormal arterial calcification or small vessel disease, such as diabetes mellitus. See 4.00G7c and 4.00G8.

c. We will use resting toe systolic blood pressures or resting toe/brachial systolic blood pressure ratios (determined the same way as ankle/brachial ratios, see 4.00G7b) when you have intermittent claudication and a disease that results in abnormal arterial calcification (for example, Monckeberg's sclerosis or diabetes mellitus) or small vessel disease (for example, diabetes mellitus). These diseases may result in misleadingly high blood pressure readings at the ankle. However, high blood pressures due to vascular changes related to these diseases seldom occur at the toe level. While the criteria in 4.12C and 4.12D are intended primarily for individuals who have a disease causing abnormal arterial calcification or small vessel disease, we may also use them for evaluating anyone with PAD.

8. *How are toe pressures measured?* Toe pressures are measured routinely in most vascular laboratories through one of three methods: most frequently, photoplethysmography; less frequently, plethysmography using strain gauge cuffs; and Doppler ultrasound. Toe pressure can also be measured by using any blood pressure cuff that fits snugly around the big toe and is neither too tight nor too loose. A neonatal cuff or a cuff designed for use on fingers or toes can be used in the measurement of toe pressure.

9. *How do we use listing 4.12 if you have had a peripheral graft?* Peripheral grafting serves the same purpose as coronary grafting; that is, to bypass a narrow or obstructed arterial segment. If intermittent claudication recurs or persists after peripheral grafting, we may purchase Doppler studies to assess the flow of blood through the bypassed vessel and to establish the current severity of the peripheral arterial impairment. However, if you have had peripheral grafting done for your PAD, we will not use the findings from before the surgery to assess the current severity of your impairment, although we will consider the severity and duration of your impairment prior to your surgery in making our determination or decision.

H. Evaluating Other Cardiovascular Impairments

1. *How will we evaluate hypertension?* Because *hypertension* (high blood pressure) generally causes disability through its effects on other body systems, we will evaluate it by reference to the specific body system(s) affected (heart, brain, kidneys, or eyes) when we consider its effects under the listings. We will also consider any limitations imposed by your hypertension when we assess your residual functional capacity.

2. *How will we evaluate symptomatic congenital heart disease? Congenital heart disease* is any abnormality of the heart or the major blood vessels that is present at birth. Because of improved treatment methods, more children with congenital heart disease are living to adulthood. Although some types of congenital heart disease may be corrected by surgery, many individuals with treated congenital heart disease continue to have problems throughout their lives (symptomatic congenital heart disease). If you have congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias, we will evaluate your impairment under 4.02 or 4.05. Otherwise, we will evaluate your impairment under 4.06.

3. *What is cardiomyopathy and how will we evaluate it? Cardiomyopathy* is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy). There are various types of cardiomyopathy, which fall into two major categories: *Ischemic* and *nonischemic* cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: Dilated, hypertrophic, and restrictive. We will evaluate cardiomyopathy under 4.02, 4.04, 4.05, or 11.04, depending on its effects on you.

4. *How will we evaluate valvular heart disease?* We will evaluate valvular heart disease under the listing appropriate for its effect on

you. Thus, we may use 4.02, 4.04, 4.05, 4.06, or an appropriate neurological listing in 11.00ff.

5. *What do we consider when we evaluate heart transplant recipients?*

a. After your heart transplant, we will consider you disabled for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year.

b. However, heart transplant patients generally meet our definition of disability before they undergo transplantation. We will determine the onset of your disability based on the facts in your case.

c. We will not assume that you became disabled when your name was placed on a transplant waiting list. This is because you may be placed on a waiting list soon after diagnosis of the cardiac disorder that may eventually require a transplant. Physicians recognize that candidates for transplantation often have to wait months or even years before a suitable donor heart is found, so they place their patients on the list as soon as permitted.

d. When we do a continuing disability review to determine whether you are still disabled, we will evaluate your residual impairment(s), as shown by symptoms, signs, and laboratory findings, including any side effects of medication. We will consider any remaining symptoms, signs, and laboratory findings indicative of cardiac dysfunction in deciding whether medical improvement (as defined in §§404.1594 and 416.994) has occurred.

6. *When does an aneurysm have ''dissection not controlled by prescribed treatment,'' as required under 4.10?* An aneurysm (or bulge in the aorta or one of its major branches) is *dissecting* when the inner lining of the artery begins to separate from the arterial wall. We consider the dissection not controlled when you have persistence of chest pain due to progression of the dissection, an increase in the size of the aneurysm, or compression of one or more branches of the aorta supplying the heart, kidneys, brain, or other organs. An aneurysm with dissection can cause heart failure, renal (kidney) failure, or neurological complications. If you have an aneurysm that does not meet the requirements of 4.10 and you have one or more of these associated conditions, we will evaluate the condition(s) using the appropriate listing.

7. *What is hyperlipidemia and how will we evaluate it? Hyperlipidemia* is the general term for an elevation of any or all of the lipids (fats or cholesterol) in the blood; for example, hypertriglyceridemia, hypercholesterolemia, and hyperlipoproteinemia. These disorders of lipoprotein metabolism and transport can cause defects throughout the body. The effects most likely to interfere with function are those produced by atherosclerosis (narrowing of the arteries) and coronary artery disease. We will evaluate your lipoprotein disorder by considering its effects on you.

8. *What is Marfan syndrome and how will we evaluate it?*

a. Marfan syndrome is a genetic connective tissue disorder that affects multiple body systems, including the skeleton, eyes, heart, blood vessels, nervous system, skin, and lungs. There is no specific laboratory test to diagnose Marfan syndrome. The diagnosis is generally made by medical history, including family history, physical examination, including an evaluation of the ratio of arm/leg size to trunk size, a slit lamp eye examination, and a heart test(s), such as an echocardiogram. In some cases, a genetic analysis may be useful, but such analyses may not provide any additional helpful information.

b. The effects of Marfan syndrome can range from mild to severe. In most cases, the disorder progresses as you age. Most individuals with Marfan syndrome have abnormalities associated with the heart and blood vessels. Your heart's mitral valve may leak, causing a heart murmur. Small leaks may not cause symptoms, but larger ones may cause shortness of breath, fatigue, and palpitations. Another effect is that the wall of the aorta may be weakened and abnormally stretch (aortic dilation). This aortic dilation may tear, dissect, or rupture, causing serious heart problems or sometimes sudden death. We will evaluate the manifestations of your Marfan syndrome under the appropriate body system criteria, such as 4.10, or if necessary, consider the functional limitations imposed by your impairment.

I. Other Evaluation Issues

1. *What effect does obesity have on the cardiovascular system and how will we evaluate it?* Obesity is a medically determinable impairment that is often associated with disorders of the cardiovascular system. Disturbance of this system can be a major cause of disability if you have obesity. Obesity may affect the cardiovascular system because of the increased workload the additional body mass places on the heart. Obesity may make it harder for the chest and lungs to expand. This can mean that the respiratory system must work harder to provide needed oxygen. This in turn would make the heart work harder to pump blood to carry oxygen to the body. Because the body would be working harder at rest, its ability to perform additional work would be less than would otherwise be expected. Thus, the combined effects of obesity with cardiovascular impairments can be greater than the effects of each of the impairments considered separately. We must consider any additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular impairment or a listing-level cardiovascular impairment (or a combination of impairments

that medically equals the severity of a listed impairment), and when we assess your residual functional capacity.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, we may need to defer evaluation of the impairment for a period of up to 3 months from the date treatment began to permit consideration of treatment effects, unless we can make a determination or decision using the evidence we have. See 4.00B4.

3. *How do we evaluate impairments that do not meet one of the cardiovascular listings?*

a. These listings are only examples of common cardiovascular impairments that we consider severe enough to prevent you from doing any gainful activity. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

b. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairments(s) medically equals a listing. (See §§404.1526 and 416.926.) If you have a severe impairment(s) that does not meet or medically equal the criteria of a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. Therefore, we proceed to the fourth and, if necessary, the fifth steps of the sequential evaluation process in §§404.1520 and 416.920. If you are an adult, we use the rules in §§404.1594 or 416.994, as appropriate, when we decide whether you continue to be disabled.

4.01 Category of Impairments, Cardiovascular System

4.02 *Chronic heart failure* while on a regimen of prescribed treatment, with symptoms and signs described in 4.00D2. The required level of severity for this impairment is met when the requirements in *both A and B* are satisfied.

A. Medically documented presence of one of the following:

1. Systolic failure (see 4.00D1a(i)), with left ventricular end diastolic dimensions greater than 6.0 cm or ejection fraction of 30 percent or less during a period of stability (not during an episode of acute heart failure); or

2. Diastolic failure (see 4.00D1a(ii)), with left ventricular posterior wall plus septal thickness totaling 2.5 cm or greater on imaging, with an enlarged left atrium greater than or equal to 4.5 cm, with normal or elevated ejection fraction during a period of stability (not during an episode of acute heart failure);

AND

B. Resulting in one of the following:

1. Persistent symptoms of heart failure which very seriously limit the ability to independently initiate, sustain, or complete activities of daily living in an individual for whom an MC, preferably one experienced in the care of patients with cardiovascular disease, has concluded that the performance of an exercise test would present a significant risk to the individual; or

2. Three or more separate episodes of acute congestive heart failure within a consecutive 12-month period (see 4.00A3e), with evidence of fluid retention (see 4.00D2b(ii)) from clinical and imaging assessments at the time of the episodes, requiring acute extended physician intervention such as hospitalization or emergency room treatment for 12 hours or more, separated by periods of stabilization (see 4.00D4c); or

3. Inability to perform on an exercise tolerance test at a workload equivalent to 5 METs or less due to:

a. Dyspnea, fatigue, palpitations, or chest discomfort; or

b. Three or more consecutive premature ventricular contractions (ventricular tachycardia), or increasing frequency of ventricular ectopy with at least 6 premature ventricular contractions per minute; or

c. Decrease of 10 mm Hg or more in systolic pressure below the baseline systolic blood pressure or the preceding systolic pressure measured during exercise (see 4.00D4d) due to left ventricular dysfunction, despite an increase in workload; or

d. Signs attributable to inadequate cerebral perfusion, such as ataxic gait or mental confusion.

4.04 *Ischemic heart disease*, with symptoms due to myocardial ischemia, as described in 4.00E3–4.00E7, while on a regimen of prescribed treatment (see 4.00B3 if there is no regimen of prescribed treatment), with one of the following:

A. Sign-or symptom-limited exercise tolerance test demonstrating at least one of the following manifestations at a workload equivalent to 5 METs or less:

1. Horizontal or downsloping depression, in the absence of digitalis glycoside treatment or hypokalemia, of the ST segment of at least −0.10 millivolts (−1.0 mm) in at least 3 consecutive complexes that are on a level baseline in any lead other than aVR, and depression of at least −0.10 millivolts lasting for at least 1 minute of recovery; or

2. At least 0.1 millivolt (1 mm) ST elevation above resting baseline in non-infarct

leads during both exercise and 1 or more minutes of recovery; or

3. Decrease of 10 mm Hg or more in systolic pressure below the baseline blood pressure or the preceding systolic pressure measured during exercise (see 4.00E9e) due to left ventricular dysfunction, despite an increase in workload; or

4. Documented ischemia at an exercise level equivalent to 5 METs or less on appropriate medically acceptable imaging, such as radionuclide perfusion scans or stress echocardiography.

OR

B. Three separate ischemic episodes, each requiring revascularization or not amenable to revascularization (see 4.00E9f), within a consecutive 12-month period (see 4.00A3e).

OR

C. Coronary artery disease, demonstrated by angiography (obtained independent of Social Security disability evaluation) or other appropriate medically acceptable imaging, and in the absence of a timely exercise tolerance test or a timely normal drug-induced stress test, an MC, preferably one experienced in the care of patients with cardiovascular disease, has concluded that performance of exercise tolerance testing would present a significant risk to the individual, with both 1 and 2:

1. Angiographic evidence showing:

a. 50 percent or more narrowing of a nonbypassed left main coronary artery; or

b. 70 percent or more narrowing of another nonbypassed coronary artery; or

c. 50 percent or more narrowing involving a long (greater than 1 cm) segment of a nonbypassed coronary artery; or

d. 50 percent or more narrowing of at least two nonbypassed coronary arteries; or

e. 70 percent or more narrowing of a bypass graft vessel; and

2. Resulting in very serious limitations in the ability to independently initiate, sustain, or complete activities of daily living.

4.05 *Recurrent arrhythmias*, not related to reversible causes, such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled (see 4.00A3f), recurrent (see 4.00A3c) episodes of cardiac syncope or near syncope (see 4.00F3b), despite prescribed treatment (see 4.00B3 if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography, or by other appropriate medically acceptable testing, coincident with the occurrence of syncope or near syncope (see 4.00F3c).

4.06 *Symptomatic congenital heart disease* (cyanotic or acyanotic), documented by appropriate medically acceptable imaging (see 4.00A3d) or cardiac catheterization, with one of the following:

A. Cyanosis at rest, and:

1. Hematocrit of 55 percent or greater; or

2. Arterial O_2 saturation of less than 90 percent in room air, or resting arterial PO_2 of 60 Torr or less.

OR

B. Intermittent right-to-left shunting resulting in cyanosis on exertion (e.g., Eisenmenger's physiology) and with arterial PO_2 of 60 Torr or less at a workload equivalent to 5 METs or less.

OR

C. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure.

4.09 *Heart transplant*. Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under the appropriate listing.

4.10 *Aneurysm of aorta or major branches*, due to any cause (e.g., atherosclerosis, cystic medial necrosis, Marfan syndrome, trauma), demonstrated by appropriate medically acceptable imaging, with dissection not controlled by prescribed treatment (see 4.00H6).

4.11 *Chronic venous insufficiency* of a lower extremity with incompetency or obstruction of the deep venous system and one of the following:

A. Extensive brawny edema (see 4.00G3) involving at least two-thirds of the leg between the ankle and knee or the distal one-third of the lower extremity between the ankle and hip.

OR

B. Superficial varicosities, stasis dermatitis, and either recurrent ulceration or persistent ulceration that has not healed following at least 3 months of prescribed treatment.

4.12 *Peripheral arterial disease*, as determined by appropriate medically acceptable imaging (see 4.00A3d, 4.00G2, 4.00G5, and 4.00G6), causing intermittent claudication (see 4.00G1) and one of the following:

A. Resting ankle/brachial systolic blood pressure ratio of less than 0.50.

OR

B. Decrease in systolic blood pressure at the ankle on exercise (see 4.00G7a and 4.00C16–4.00C17) of 50 percent or more of pre-exercise level and requiring 10 minutes or more to return to pre-exercise level.

OR

C. Resting toe systolic pressure of less than 30 mm Hg (see 4.00G7c and 4.00G8).

OR

D. Resting toe/brachial systolic blood pressure ratio of less than 0.40 (see 4.00G7c).

5.00 DIGESTIVE SYSTEM

A. *What kinds of disorders do we consider in the digestive system?* Disorders of the digestive system include gastrointestinal hemorrhage,

hepatic (liver) dysfunction, inflammatory bowel disease, short bowel syndrome, and malnutrition. They may also lead to complications, such as obstruction, or be accompanied by manifestations in other body systems.

B. *What documentation do we need?* We need a record of your medical evidence, including clinical and laboratory findings. The documentation should include appropriate medically acceptable imaging studies and reports of endoscopy, operations, and pathology, as appropriate to each listing, to document the severity and duration of your digestive disorder. Medically acceptable imaging includes, but is not limited to, x-ray imaging, sonography, computerized axial tomography (CAT scan), magnetic resonance imaging (MRI), and radionuclide scans. *Appropriate* means that the technique used is the proper one to support the evaluation and diagnosis of the disorder. The findings required by these listings must occur within the period we are considering in connection with your application or continuing disability review.

C. *How do we consider the effects of treatment?*

1. Digestive disorders frequently respond to medical or surgical treatment; therefore, we generally consider the severity and duration of these disorders within the context of prescribed treatment.

2. We assess the effects of treatment, including medication, therapy, surgery, or any other form of treatment you receive, by determining if there are improvements in the symptoms, signs, and laboratory findings of your digestive disorder. We also assess any side effects of your treatment that may further limit your functioning.

3. To assess the effects of your treatment, we may need information about:

a. The treatment you have been prescribed (for example, the type of medication or therapy, or your use of parenteral (intravenous) nutrition or supplemental enteral nutrition via a gastrostomy);

b. The dosage, method, and frequency of administration;

c. Your response to the treatment;

d. Any adverse effects of such treatment; and

e. The expected duration of the treatment.

4. Because the effects of treatment may be temporary or long-term, in most cases we need information about the impact of your treatment, including its expected duration and side effects, over a sufficient period of time to help us assess its outcome. When adverse effects of treatment contribute to the severity of your impairment(s), we will consider the duration or expected duration of the treatment when we assess the duration of your impairment(s).

5. If you need parenteral (intravenous) nutrition or supplemental enteral nutrition via a gastrostomy to avoid debilitating complications of a digestive disorder, this treatment will not, in itself, indicate that you are unable to do any gainful activity, except under 5.07, short bowel syndrome (see 5.00F).

6. If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the severity and duration of your digestive impairment on the basis of the current medical and other evidence in your case record. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the digestive system listings, but your digestive impairment may medically equal a listing or be disabling based on consideration of your residual functional capacity, age, education, and work experience.

D. *How do we evaluate chronic liver disease?*

1. *General. Chronic liver disease* is characterized by liver cell necrosis, inflammation, or scarring (fibrosis or cirrhosis), due to any cause, that persists for more than 6 months. Chronic liver disease may result in portal hypertension, cholestasis (suppression of bile flow), extrahepatic manifestations, or liver cancer. (We evaluate liver cancer under 13.19.) Significant loss of liver function may be manifested by hemorrhage from varices or portal hypertensive gastropathy, ascites (accumulation of fluid in the abdominal cavity), hydrothorax (ascitic fluid in the chest cavity), or encephalopathy. There can also be progressive deterioration of laboratory findings that are indicative of liver dysfunction. Liver transplantation is the only definitive cure for end stage liver disease (ESLD).

2. *Examples of chronic liver disease* include, but are not limited to, chronic hepatitis, alcoholic liver disease, non-alcoholic steatohepatitis (NASH), primary biliary cirrhosis (PBC), primary sclerosing cholangitis (PSC), autoimmune hepatitis, hemochromatosis, drug-induced liver disease, Wilson's disease, and serum alpha-1 antitrypsin deficiency. Acute hepatic injury is frequently reversible, as in viral, drug-induced, toxin-induced, alcoholic, and ischemic hepatitis. In the absence of evidence of a chronic impairment, episodes of acute liver disease do not meet 5.05.

3. *Manifestations of chronic liver disease.*

a. *Symptoms* may include, but are not limited to, pruritis (itching), fatigue, nausea, loss of appetite, or sleep disturbances. Symptoms of chronic liver disease may have a poor correlation with the severity of liver disease and functional ability.

b. *Signs* may include, but are not limited to, jaundice, enlargement of the liver and spleen, ascites, peripheral edema, and altered mental status.

c. *Laboratory findings* may include, but are not limited to, increased liver enzymes, increased serum total bilirubin, increased ammonia levels, decreased serum albumin, and

abnormal coagulation studies, such as increased International Normalized Ratio (INR) or decreased platelet counts. Abnormally low serum albumin or elevated INR levels indicate loss of synthetic liver function, with increased likelihood of cirrhosis and associated complications. However, other abnormal lab tests, such as liver enzymes, serum total bilirubin, or ammonia levels, may have a poor correlation with the severity of liver disease and functional ability. A liver biopsy may demonstrate the degree of liver cell necrosis, inflammation, fibrosis, and cirrhosis. If you have had a liver biopsy, we will make every reasonable effort to obtain the results; however, we will not purchase a liver biopsy. Imaging studies (CAT scan, ultrasound, MRI) may show the size and consistency (fatty liver, scarring) of the liver and document ascites (see 5.00D6).

4. *Chronic viral hepatitis infections.*

a. *General.*

(i) *Chronic viral hepatitis* infections are commonly caused by hepatitis C virus (HCV), and to a lesser extent, hepatitis B virus (HBV). Usually, these are slowly progressive disorders that persist over many years during which the symptoms and signs are typically nonspecific, intermittent, and mild (for example, fatigue, difficulty with concentration, or right upper quadrant pain). Laboratory findings (liver enzymes, imaging studies, liver biopsy pathology) and complications are generally similar in HCV and HBV. The spectrum of these chronic viral hepatitis infections ranges widely and includes an asymptomatic state; insidious disease with mild to moderate symptoms associated with fluctuating liver tests; extrahepatic manifestations; cirrhosis, both compensated and decompensated; ESLD with the need for liver transplantation; and liver cancer. Treatment for chronic viral hepatitis infections varies considerably based on medication tolerance, treatment response, adverse effects of treatment, and duration of the treatment. Comorbid disorders, such as HIV infection, may affect the clinical course of viral hepatitis infection(s) or may alter the response to medical treatment.

(ii) We evaluate all types of chronic viral hepatitis infections under 5.05 or any listing in an affected body system(s). If your impairment(s) does not meet or medically equal a listing, we will consider the effects of your hepatitis when we assess your residual functional capacity.

b. *Chronic hepatitis B virus (HBV) infection.*

(i) *Chronic HBV* infection is diagnosed by the detection of hepatitis B surface antigen (HBsAg) in the blood for at least 6 months. In addition, detection of the hepatitis B envelope antigen (HBeAg) suggests an increased likelihood of progression to cirrhosis and ESLD.

(ii) The therapeutic goal of treatment is to suppress HBV replication and thereby prevent progression to cirrhosis and ESLD. Treatment usually includes a combination of interferon injections and oral antiviral agents. Common adverse effects of treatment are the same as noted in 5.00D4c(ii) for HCV, and generally end within a few days after treatment is discontinued.

c. *Chronic hepatitis C virus (HCV) infection.*

(i) *Chronic HCV infection* is diagnosed by the detection of hepatitis C viral RNA in the blood for at least 6 months. Documentation of the therapeutic response to treatment is also monitored by the quantitative assay of serum HCV RNA (''HCV viral load''). Treatment usually includes a combination of interferon injections and oral ribavirin; whether a therapeutic response has occurred is usually assessed after 12 weeks of treatment by checking the HCV viral load. If there has been a substantial reduction in HCV viral load (also known as early viral response, or EVR), this reduction is predictive of a sustained viral response with completion of treatment. Combined therapy is commonly discontinued after 12 weeks when there is no early viral response, since in that circumstance there is little chance of obtaining a sustained viral response (SVR). Otherwise, treatment is usually continued for a total of 48 weeks.

(ii) Combined interferon and ribavirin treatment may have significant adverse effects that may require dosing reduction, planned interruption of treatment, or discontinuation of treatment. Adverse effects may include: Anemia (ribavirin-induced hemolysis), neutropenia, thrombocytopenia, fever, cough, fatigue, myalgia, arthralgia, nausea, loss of appetite, pruritis, and insomnia. Behavioral side effects may also occur. Influenza-like symptoms are generally worse in the first 4 to 6 hours after each interferon injection and during the first weeks of treatment. Adverse effects generally end within a few days after treatment is discontinued.

d. *Extrahepatic manifestations of HBV and HCV.* In addition to their hepatic manifestations, both HBV and HCV may have significant extrahepatic manifestations in a variety of body systems. These include, but are not limited to: Keratoconjunctivitis (sicca syndrome), glomerulonephritis, skin disorders (for example, lichen planus, porphyria cutanea tarda), neuropathy, and immune dysfunction (for example, cryoglobulinemia, Sjögren's syndrome, and vasculitis). The extrahepatic manifestations of HBV and HCV may not correlate with the severity of your hepatic impairment. If your impairment(s) does not meet or medically equal a listing in an affected body system(s), we will consider the effects of your extrahepatic manifestations when we assess your residual functional capacity.

5. *Gastrointestinal hemorrhage* (5.02 and 5.05A). Gastrointestinal hemorrhaging can result in hematemesis (vomiting of blood),

melena (tarry stools), or hematochezia (bloody stools). Under 5.02, the required transfusions of at least 2 units of blood must be at least 30 days apart and occur at least three times during a consecutive 6-month period. Under 5.05A, *hemodynamic instability* is diagnosed with signs such as pallor (pale skin), diaphoresis (profuse perspiration), rapid pulse, low blood pressure, postural hypotension (pronounced fall in blood pressure when arising to an upright position from lying down) or syncope (fainting). Hemorrhaging that results in hemodynamic instability is potentially life-threatening and therefore requires hospitalization for transfusion and supportive care. Under 5.05A, we require only one hospitalization for transfusion of at least 2 units of blood.

6. *Ascites or hydrothorax* (5.05B) indicates significant loss of liver function due to chronic liver disease. We evaluate ascites or hydrothorax that is not attributable to other causes under 5.05B. The required findings must be present on at least two evaluations at least 60 days apart within a consecutive 6-month period and despite continuing treatment as prescribed.

7. *Spontaneous bacterial peritonitis* (5.05C) is an infectious complication of chronic liver disease. It is diagnosed by ascitic peritoneal fluid that is documented to contain an absolute neutrophil count of at least 250 cells/mm^3. The required finding in 5.05C is satisfied with one evaluation documenting peritoneal fluid infection. We do not evaluate other causes of peritonitis that are unrelated to chronic liver disease, such as tuberculosis, malignancy, and perforated bowel, under this listing. We evaluate these other causes of peritonitis under the appropriate body system listings.

8. *Hepatorenal syndrome* (5.05D) is defined as functional renal failure associated with chronic liver disease in the absence of underlying kidney pathology. Hepatorenal syndrome is documented by elevation of serum creatinine, marked sodium retention, and oliguria (reduced urine output). The requirements of 5.05D are satisfied with documentation of any one of the three laboratory findings on one evaluation. We do not evaluate known causes of renal dysfunction, such as glomerulonephritis, tubular necrosis, drug-induced renal disease, and renal infections, under this listing. We evaluate these other renal impairments under 6.00ff.

9. *Hepatopulmonary syndrome* (5.05E) is defined as arterial deoxygenation (hypoxemia) that is associated with chronic liver disease due to intrapulmonary arteriovenous shunting and vasodilatation in the absence of other causes of arterial deoxygenation. Clinical manifestations usually include dyspnea, orthodeoxia (increasing hypoxemia with erect position), platypnea (improvement of dyspnea with flat position), cyanosis, and clubbing. The requirements of 5.05E are satisfied with documentation of any one of the findings on one evaluation. In 5.05E1, we require documentation of the altitude of the testing facility because altitude affects the measurement of arterial oxygenation. We will not purchase the specialized studies described in 5.05E2; however, if you have had these studies at a time relevant to your claim, we will make every reasonable effort to obtain the reports for the purpose of establishing whether your impairment meets 5.05E2.

10. *Hepatic encephalopathy* (5.05F).

a. *General*. Hepatic encephalopathy usually indicates severe loss of hepatocellular function. We define hepatic encephalopathy under 5.05F as a recurrent or chronic neuropsychiatric disorder, characterized by abnormal behavior, cognitive dysfunction, altered state of consciousness, and ultimately coma and death. The diagnosis is established by changes in mental status associated with fleeting neurological signs, including ''flapping tremor'' (asterixis), characteristic electroencephalographic (EEG) abnormalities, or abnormal laboratory values that indicate loss of synthetic liver function. We will not purchase the EEG testing described in 5.05F3b; however, if you have had this test at a time relevant to your claim, we will make every reasonable effort to obtain the report for the purpose of establishing whether your impairment meets 5.05F.

b. *Acute encephalopathy*. We will not evaluate your acute encephalopathy under 5.05F if it results from conditions other than chronic liver disease, such as vascular events and neoplastic diseases. We will evaluate these other causes of acute encephalopathy under the appropriate body system listings.

11. *End stage liver disease (ESLD) documented by scores from the SSA Chronic Liver Disease (SSA CLD) calculation* (5.05G).

a. We will use the SSA CLD score to evaluate your ESLD under 5.05G. We explain how we calculate the SSA CLD score in b. through g. of this section.

b. To calculate the SSA CLD score, we use a formula that includes three laboratory values: Serum total bilirubin (mg/dL), serum creatinine (mg/dL), and International Normalized Ratio (INR). The formula for the SSA CLD score calculation is:

$$9.57 \times [\text{Log}_e(\text{serum creatinine mg/dL})] + 3.78 \times [\text{Log}_e(\text{serum total bilirubin mg/dL})] + 11.2 \times [\text{Log}_e(\text{INR})] + 6.43$$

c. When we indicate ''Log_e'' in the formula for the SSA CLD score calculation, we mean the ''base *e* logarithm'' or ''natural logarithm'' (ln) of a numerical laboratory value, not the ''base 10 logarithm'' or ''common logarithm'' (log) of the laboratory value, and not the actual laboratory value. For example, if an individual has laboratory values of

serum creatinine 1.2 mg/dL, serum total bilirubin 2.2 mg/dL, and INR 1.0, we would compute the SSA CLD score as follows:

9.57 × [Log_e(serum creatinine 1.2 mg/dL) = 0.182]
+ 3.78 × [Log_e(serum total bilirubin 2.2 mg/dL) = 0.788]
+ 11.2 × [Log_e(INR 1.0) = 0]
+ *6.43*

= 1.74 + 2.98 + 0 + 6.43
= 11.15, which is then rounded to an SSA CLD score of 11.

d. For any SSA CLD score calculation, all of the required laboratory values must have been obtained within 30 days of each other. If there are multiple laboratory values within the 30-day interval for any given laboratory test (serum total bilirubin, serum creatinine, or INR), we will use the highest value for the SSA CLD score calculation. We will round all laboratory values less than 1.0 up to 1.0.

e. Listing 5.05G requires two SSA CLD scores. The laboratory values for the second SSA CLD score calculation must have been obtained at least 60 days after the latest laboratory value for the first SSA CLD score and within the required 6-month period. We will consider the date of each SSA CLD score to be the date of the first laboratory value used for its calculation.

f. If you are in renal failure or on dialysis within a week of any serum creatinine test in the period used for the SSA CLD calculation, we will use a serum creatinine of 4, which is the maximum serum creatinine level allowed in the calculation, to calculate your SSA CLD score.

g. If you have the two SSA CLD scores required by 5.05G, we will find that your impairment meets the criteria of the listing from at least the date of the first SSA CLD score.

12. *Liver transplantation* (5.09) may be performed for metabolic liver disease, progressive liver failure, life-threatening complications of liver disease, hepatic malignancy, and acute fulminant hepatitis (viral, drug-induced, or toxin-induced). We will consider you to be disabled for 1 year from the date of the transplantation. Thereafter, we will evaluate your residual impairment(s) by considering the adequacy of post-transplant liver function, the requirement for post-transplant antiviral therapy, the frequency and severity of rejection episodes, comorbid complications, and all adverse treatment effects.

E. *How do we evaluate inflammatory bowel disease (IBD)?*

1. *Inflammatory bowel disease* (5.06) includes, but is not limited to, Crohn's disease and ulcerative colitis. These disorders, while distinct entities, share many clinical, laboratory, and imaging findings, as well as similar treatment regimens. Remissions and exacerbations of variable duration are the hallmark of IBD. Crohn's disease may involve the entire alimentary tract from the mouth to the anus in a segmental, asymmetric fashion. Obstruction, stenosis, fistulization, perineal involvement, and extraintestinal manifestations are common. Crohn's disease is rarely curable and recurrence may be a lifelong problem, even after surgical resection. In contrast, ulcerative colitis only affects the colon. The inflammatory process may be limited to the rectum, extend proximally to include any contiguous segment, or involve the entire colon. Ulcerative colitis may be cured by total colectomy.

2. Symptoms and signs of IBD include diarrhea, fecal incontinence, rectal bleeding, abdominal pain, fatigue, fever, nausea, vomiting, arthralgia, abdominal tenderness, palpable abdominal mass (usually inflamed loops of bowel) and perineal disease. You may also have signs or laboratory findings indicating malnutrition, such as weight loss, edema, anemia, hypoalbuminemia, hypokalemia, hypocalcemia, or hypomagnesemia.

3. IBD may be associated with significant extraintestinal manifestations in a variety of body systems. These include, but are not limited to, involvement of the eye (for example, uveitis, episcleritis, iritis); hepatobiliary disease (for example, gallstones, primary sclerosing cholangitis); urologic disease (for example, kidney stones, obstructive hydronephrosis); skin involvement (for example, erythema nodosum, pyoderma gangrenosum); or non-destructive inflammatory arthritis. You may also have associated thromboembolic disorders or vascular disease. These manifestations may not correlate with the severity of your IBD. If your impairment does not meet any of the criteria of 5.06, we will consider the effects of your extraintestinal manifestations in determining whether you have an impairment(s) that meets or medically equals another listing, and we will also consider the effects of your extraintestinal manifestations when we assess your residual functional capacity.

4. Surgical diversion of the intestinal tract, including ileostomy and colostomy, does not preclude any gainful activity if you are able to maintain adequate nutrition and function of the stoma. However, if you are not able to maintain adequate nutrition, we will evaluate your impairment under 5.08.

F. *How do we evaluate short bowel syndrome (SBS)?*

1. *Short bowel syndrome* (5.07) is a disorder that occurs when ischemic vascular insults (for example, volvulus), trauma, or IBD complications require surgical resection of more than one-half of the small intestine, resulting in the loss of intestinal absorptive surface and a state of chronic malnutrition. The management of SBS requires long-term parenteral nutrition via an indwelling central venous catheter (central line); the process is

often referred to as *hyperalimentation* or *total parenteral nutrition* (TPN). Individuals with SBS can also feed orally, with variable amounts of nutrients being absorbed through their remaining intestine. Over time, some of these individuals can develop additional intestinal absorptive surface, and may ultimately be able to be weaned off their parenteral nutrition.

2. Your impairment will continue to meet 5.07 as long as you remain dependent on daily parenteral nutrition via a central venous catheter for most of your nutritional requirements. Long-term complications of SBS and parenteral nutrition include central line infections (with or without septicemia), thrombosis, hepatotoxicity, gallstones, and loss of venous access sites. Intestinal transplantation is the only definitive treatment for individuals with SBS who remain chronically dependent on parenteral nutrition.

3. To document SBS, we need a copy of the operative report of intestinal resection, the summary of the hospitalization(s) including: Details of the surgical findings, medically appropriate postoperative imaging studies that reflect the amount of your residual small intestine, or if we cannot get one of these reports, other medical reports that include details of the surgical findings. We also need medical documentation that you are dependent on daily parenteral nutrition to provide most of your nutritional requirements.

G. *How do we evaluate weight loss due to any digestive disorder?*

1. In addition to the impairments specifically mentioned in these listings, other digestive disorders, such as esophageal stricture, pancreatic insufficiency, and malabsorption, may result in significant weight loss. We evaluate weight loss due to any digestive disorder under 5.08 by using the Body Mass Index (BMI). We also provide a criterion in 5.06B for lesser weight loss resulting from IBD.

2. BMI is the ratio of your weight to the square of your height. Calculation and interpretation of the BMI are independent of gender in adults.

a. We calculate BMI using inches and pounds, meters and kilograms, or centimeters and kilograms. We must have measurements of your weight and height without shoes for these calculations.

b. We calculate BMI using one of the following formulas:

English Formula

$$\text{BMI} = \left(\frac{\text{Weight in Pounds}}{(\text{Height in Inches}) \times (\text{Height in Inches})} \right) \times 703$$

Metric Formula

$$\text{BMI} = \frac{\text{Weight in Kilograms}}{(\text{Height in Meters}) \times (\text{Height in Meters})}$$

Or

$$\text{BMI} = \left(\frac{\text{Weight in Kilograms}}{(\text{Height in Centimeters}) \times (\text{Height in Centimeters})} \right) \times 10{,}000$$

H. *What do we mean by the phrase ''consider under a disability for 1 year''?* We use the phrase ''consider under a disability for 1 year'' following a specific event in 5.02, 5.05A, and 5.09 to explain how long your impairment can meet the requirements of those particular listings. This phrase does not refer to the date on which your disability began, only to the date on which we must reevaluate whether your impairment continues to meet a listing or is otherwise disabling. For example, if you have received a liver transplant, you may have become disabled before the transplant because of chronic liver disease. Therefore, we do not restrict our determination of the onset of disability to the date of the specified event. We will establish an onset date earlier than the date of

the specified event if the evidence in your case record supports such a finding.

I. *How do we evaluate impairments that do not meet one of the digestive disorder listings?*

1. These listings are only examples of common digestive disorders that we consider severe enough to prevent you from doing any gainful activity. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system. For example, if you have hepatitis B or C and you are depressed, we will evaluate your impairment under 12.04.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§ 404.1526 and 416.926.) If your impairment(s) does not meet or medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. In this situation, we will proceed to the fourth, and if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920. When we decide whether you continue to be disabled, we use the rules in §§ 404.1594, 416.994, and 416.994a as appropriate.

5.01 *Category of Impairments, Digestive System*

5.02 *Gastrointestinal hemorrhaging from any cause, requiring blood transfusion* (with or without hospitalization) of at least 2 units of blood per transfusion, and occurring at least three times during a consecutive 6-month period. The transfusions must be at least 30 days apart within the 6-month period. Consider under a disability for 1 year following the last documented transfusion; thereafter, evaluate the residual impairment(s).

5.03–5.04 [Reserved]

5.05 *Chronic liver disease,* with:

A. Hemorrhaging from esophageal, gastric, or ectopic varices or from portal hypertensive gastropathy, demonstrated by endoscopy, x-ray, or other appropriate medically acceptable imaging, resulting in hemodynamic instability as defined in 5.00D5, and requiring hospitalization for transfusion of at least 2 units of blood. Consider under a disability for 1 year following the last documented transfusion; thereafter, evaluate the residual impairment(s).

OR

B. Ascites or hydrothorax not attributable to other causes, despite continuing treatment as prescribed, present on at least two evaluations at least 60 days apart within a consecutive 6-month period. Each evaluation must be documented by:

1. Paracentesis or thoracentesis; or

2. Appropriate medically acceptable imaging or physical examination and one of the following:

a. Serum albumin of 3.0 g/dL or less; or

b. International Normalized Ratio (INR) of at least 1.5.

OR

C. Spontaneous bacterial peritonitis with peritoneal fluid containing an absolute neutrophil count of at least 250 cells/mm^3.

OR

D. Hepatorenal syndrome as described in 5.00D8, with one of the following:

1. Serum creatinine elevation of at least 2 mg/dL; or

2. Oliguria with 24-hour urine output less than 500 mL; or

3. Sodium retention with urine sodium less than 10 mEq per liter.

OR

E. Hepatopulmonary syndrome as described in 5.00D9, with:

1. Arterial oxygenation (P_aO_2) on room air of:

a. 60 mm Hg or less, at test sites less than 3000 feet above sea level, or

b. 55 mm Hg or less, at test sites from 3000 to 6000 feet, or

c. 50 mm Hg or less, at test sites above 6000 feet; or

2. Documentation of intrapulmonary arteriovenous shunting by contrast-enhanced echocardiography or macroaggregated albumin lung perfusion scan.

OR

F. Hepatic encephalopathy as described in 5.00D10, with 1 and either 2 or 3:

1. Documentation of abnormal behavior, cognitive dysfunction, changes in mental status, or altered state of consciousness (for example, confusion, delirium, stupor, or coma), present on at least two evaluations at least 60 days apart within a consecutive 6-month period; and

2. History of transjugular intrahepatic portosystemic shunt (TIPS) or any surgical portosystemic shunt: or

3. One of the following occurring on at least two evaluations at least 60 days apart within the same consecutive 6-month period as in F1:

a. Asterixis or other fluctuating physical neurological abnormalities; or

b. Electroencephalogram (EEG) demonstrating triphasic slow wave activity; or

c. Serum albumin of 3.0 g/dL or less; or

d. International Normalized Ratio (INR) of 1.5 or greater.

OR

G. End stage liver disease with SSA CLD scores of 22 or greater calculated as described in 5.00D11. Consider under a disability from at least the date of the first score.

5.06 *Inflammatory bowel disease (IBD)* documented by endoscopy, biopsy, appropriate

medically acceptable imaging, or operative findings with:

A. Obstruction of stenotic areas (not adhesions) in the small intestine or colon with proximal dilatation, confirmed by appropriate medically acceptable imaging or in surgery, requiring hospitalization for intestinal decompression or for surgery, and occurring on at least two occasions at least 60 days apart within a consecutive 6-month period;

OR

B. Two of the following despite continuing treatment as prescribed and occurring within the same consecutive 6-month period:

1. Anemia with hemoglobin of less than 10.0 g/dL, present on at least two evaluations at least 60 days apart; or

2. Serum albumin of 3.0 g/dL or less, present on at least two evaluations at least 60 days apart; or

3. Clinically documented tender abdominal mass palpable on physical examination with abdominal pain or cramping that is not completely controlled by prescribed narcotic medication, present on at least two evaluations at least 60 days apart; or

4. Perineal disease with a draining abscess or fistula, with pain that is not completely controlled by prescribed narcotic medication, present on at least two evaluations at least 60 days apart; or

5. Involuntary weight loss of at least 10 percent from baseline, as computed in pounds, kilograms, or BMI, present on at least two evaluations at least 60 days apart; or

6. Need for supplemental daily enteral nutrition via a gastrostomy or daily parenteral nutrition via a central venous catheter.

5.07 *Short bowel syndrome (SBS),* due to surgical resection of more than one-half of the small intestine, with dependence on daily parenteral nutrition via a central venous catheter (see 5.00F).

5.08 *Weight loss due to any digestive disorder* despite continuing treatment as prescribed, with BMI of less than 17.50 calculated on at least two evaluations at least 60 days apart within a consecutive 6-month period.

5.09 *Liver transplantation.* Consider under a disability for 1 year following the date of transplantation; thereafter, evaluate the residual impairment(s) (see 5.00D12 and 5.00H).

6.00 GENITOURINARY DISORDERS

A. Which disorders do we evaluate under these listings?

We evaluate genitourinary disorders resulting in chronic kidney disease (CKD). Examples of such disorders include chronic glomerulonephritis, hypertensive nephropathy, diabetic nephropathy, chronic obstructive uropathy, and hereditary nephropathies. We also evaluate nephrotic syndrome due to glomerular dysfunction under these listings.

B. What evidence do we need?

1. We need evidence that documents the signs, symptoms, and laboratory findings of your CKD. This evidence should include reports of clinical examinations, treatment records, and documentation of your response to treatment. Laboratory findings, such as serum creatinine or serum albumin levels, may document your kidney function. We generally need evidence covering a period of at least 90 days unless we can make a fully favorable determination or decision without it.

2. *Estimated glomerular filtration rate (eGFR).* The eGFR is an estimate of the filtering capacity of the kidneys that takes into account serum creatinine concentration and other variables, such as your age, gender, and body size. If your medical evidence includes eGFR findings, we will consider them when we evaluate your CKD under 6.05.

3. *Kidney or bone biopsy.* If you have had a kidney or bone biopsy, we need a copy of the pathology report. When we cannot get a copy of the pathology report, we will accept a statement from an acceptable medical source verifying that a biopsy was performed and describing the results.

C. What other factors do we consider when we evaluate your genitourinary disorder?

1. *Chronic hemodialysis or peritoneal dialysis.*

a. Dialysis is a treatment for CKD that uses artificial means to remove toxic metabolic byproducts from the blood. Hemodialysis uses an artificial kidney machine to clean waste products from the blood; peritoneal dialysis uses a dialyzing solution that is introduced into and removed from the abdomen (peritoneal cavity) either continuously or intermittently. Under 6.03, your ongoing dialysis must have lasted or be expected to last for a continuous period of at least 12 months. To satisfy the requirements in 6.03, we will accept a report from an acceptable medical source that describes your CKD and your current dialysis, and indicates that your dialysis will be ongoing.

b. If you are undergoing chronic hemodialysis or peritoneal dialysis, your CKD may meet our definition of disability before you started dialysis. We will determine the onset of your disability based on the facts in your case record.

2. *Kidney transplant.*

a. If you receive a kidney transplant, we will consider you to be disabled under 6.04 for 1 year from the date of transplant. After that, we will evaluate your residual impairment(s) by considering your post-transplant function, any rejection episodes you have had, complications in other body systems,

and any adverse effects related to ongoing treatment.

b. If you received a kidney transplant, your CKD may meet our definition of disability before you received the transplant. We will determine the onset of your disability based on the facts in your case record.

3. *Renal osteodystrophy*. This condition is the bone degeneration resulting from chronic kidney disease-mineral and bone disorder (CKD–MBD). CKD–MBD occurs when the kidneys are unable to maintain the necessary levels of minerals, hormones, and vitamins required for bone structure and function. Under 6.05B1, ''severe bone pain'' means frequent or intractable (resistant to treatment) bone pain that interferes with physical activity or mental functioning.

4. *Peripheral neuropathy*. This disorder results when the kidneys do not adequately filter toxic substances from the blood. These toxins can adversely affect nerve tissue. The resulting neuropathy may affect peripheral motor or sensory nerves, or both, causing pain, numbness, tingling, and muscle weakness in various parts of the body. Under 6.05B2, the peripheral neuropathy must be a severe impairment. (See §§ 404.1520(c), 404.1521, 416.920(c), and 416.921 of this chapter.) It must also have lasted or be expected to last for a continuous period of at least 12 months.

5. *Fluid overload syndrome*. This condition occurs when excess sodium and water retention in the body due to CKD results in vascular congestion. Under 6.05B3, we need a description of a physical examination that documents signs and symptoms of vascular congestion, such as congestive heart failure, pleural effusion (excess fluid in the chest), ascites (excess fluid in the abdomen), hypertension, fatigue, shortness of breath, or peripheral edema.

6. *Anasarca* (generalized massive edema or swelling). Under 6.05B3 and 6.06B, we need a description of the extent of edema, including pretibial (in front of the tibia), periorbital (around the eyes), or presacral (in front of the sacrum) edema. We also need a description of any ascites, pleural effusion, or pericardial effusion.

7. *Anorexia (diminished appetite) with weight loss*. Anorexia is a frequent sign of CKD and can result in weight loss. We will use body mass index (BMI) to determine the severity of your weight loss under 6.05B4. (BMI is the ratio of your measured weight to the square of your measured height.) The formula for calculating BMI is in section 5.00G.

8. *Complications of CKD*. The hospitalizations in 6.09 may be for different complications of CKD. Examples of complications from CKD that may result in hospitalization include stroke, congestive heart failure, hypertensive crisis, or acute kidney failure requiring a short course of hemodialysis. If the CKD complication occurs during a hospitalization that was initially for a co-occurring condition, we will evaluate it under our rules for determining medical equivalence. (See §§ 404.1526 and 416.926 of this chapter.) We will evaluate co-occurring conditions, including those that result in hospitalizations, under the listings for the affected body system or under our rules for medical equivalence.

D. How do we evaluate disorders that do not meet one of the genitourinary listings?

1. The listed disorders are only examples of common genitourinary disorders that we consider severe enough to prevent you from doing any gainful activity. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§ 404.1526 and 416.926 of this chapter.) Genitourinary disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not meet or medically equal the criteria of a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. We proceed to the fourth and, if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920 of this chapter. We use the rules in §§ 404.1594 and 416.994 of this chapter, as appropriate, when we decide whether you continue to be disabled.

6.01 Category of Impairments, Genitourinary Disorders

6.03 *Chronic kidney disease*, with chronic hemodialysis or peritoneal dialysis (see 6.00C1).

6.04 *Chronic kidney disease*, with kidney transplant. Consider under a disability for 1 year following the transplant; thereafter, evaluate the residual impairment (see 6.00C2).

6.05 *Chronic kidney disease*, with impairment of kidney function, with A and B:

A. Reduced glomerular filtration evidenced by one of the following laboratory findings documented on at least two occasions at least 90 days apart during a consecutive 12-month period:

1. Serum creatinine of 4 mg/dL or greater; or

2. Creatinine clearance of 20 ml/min. or less; or

3. Estimated glomerular filtration rate (eGFR) of 20 ml/min/1.73m^2 or less.

AND

B. One of the following:

1. Renal osteodystrophy (see 6.00C3) with severe bone pain and imaging studies documenting bone abnormalities, such as osteitis fibrosa, osteomalacia, or pathologic fractures; or

2. Peripheral neuropathy (see 6.00C4); or

3. Fluid overload syndrome (see 6.00C5) documented by one of the following:

a. Diastolic hypertension greater than or equal to diastolic blood pressure of 110 mm Hg despite at least 90 consecutive days of prescribed therapy, documented by at least two measurements of diastolic blood pressure at least 90 days apart during a consecutive 12-month period; or

b. Signs of vascular congestion or anasarca (see 6.00C6) despite at least 90 consecutive days of prescribed therapy, documented on at least two occasions at least 90 days apart during a consecutive 12-month period; or

4. Anorexia with weight loss (see 6.00C7) determined by body mass index (BMI) of 18.0 or less, calculated on at least two occasions at least 90 days apart during a consecutive 12-month period.

6.06 *Nephrotic syndrome,* with A and B:

A. Laboratory findings as described in 1 or 2, documented on at least two occasions at least 90 days apart during a consecutive 12-month period:

1. Proteinuria of 10.0 g or greater per 24 hours; or

2. Serum albumin of 3.0 g/dL or less, and

a. Proteinuria of 3.5 g or greater per 24 hours; or

b. Urine total-protein-to-creatinine ratio of 3.5 or greater.

AND

B. Anasarca (see 6.00C6) persisting for at least 90 days despite prescribed treatment.

6.09 *Complications of chronic kidney disease* (see 6.00C8) requiring at least three hospitalizations within a consecutive 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

7.00 Hematological Disorders

A. What hematological disorders do we evaluate under these listings?

1. We evaluate non-malignant (non-cancerous) hematological disorders, such as hemolytic anemias (7.05), disorders of thrombosis and hemostasis (7.08), and disorders of bone marrow failure (7.10). These disorders disrupt the normal development and function of white blood cells, red blood cells, platelets, and clotting-factor proteins (factors).

2. We evaluate malignant (cancerous) hematological disorders, such as lymphoma, leukemia, and multiple myeloma, under the appropriate listings in 13.00, except for lymphoma associated with human immunodeficiency virus (HIV) infection, which we evaluate under 14.08E.

B. What evidence do we need to document that you have a hematological disorder?

We need the following evidence to document that you have a hematological disorder:

1. A laboratory report of a definitive test that establishes a hematological disorder, signed by a physician; or

2. A laboratory report of a definitive test that establishes a hematological disorder that is not signed by a physician and a report from a physician that states you have the disorder; or

3. When we do not have a laboratory report of a definitive test, a persuasive report from a physician that a diagnosis of your hematological disorder was confirmed by appropriate laboratory analysis or other diagnostic method(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test or tests for diagnosing your disorder and provide the results, or explain how your diagnosis was established by other diagnostic method(s) consistent with the prevailing state of medical knowledge and clinical practice.

4. We will make every reasonable effort to obtain the results of appropriate laboratory testing you have had. We will not purchase complex, costly, or invasive tests, such as tests of clotting-factor proteins, and bone marrow aspirations.

C. What are hemolytic anemias, and how do we evaluate them under 7.05?

1. *Hemolytic anemias, both congenital and acquired,* are disorders that result in premature destruction of red blood cells (RBCs). Hemolytic disorders include abnormalities of hemoglobin structure (hemoglobinopathies), abnormal RBC enzyme content and function, and RBC membrane (envelope) defects that are congenital or acquired. The diagnosis of hemolytic anemia is based on hemoglobin electrophoresis or analysis of the contents of the RBC (enzymes) and membrane. Examples of congenital hemolytic anemias include sickle cell disease, thalassemia and their variants, and hereditary spherocytosis. Acquired hemolytic anemias may result from autoimmune disease (for example, systemic lupus erythematosus) or mechanical devices (for example, heart valves, intravascular patches).

2. The hospitalizations in 7.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include osteomyelitis, painful (vaso-occlusive)

crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver) failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and stroke. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

3. For 7.05C, we do not require hemoglobin to be measured during a period in which you are free of pain or other symptoms of your disorder. We will accept hemoglobin measurements made while you are experiencing complications of your hemolytic anemia.

4. 7.05D refers to the most serious type of beta thalassemia major in which the bone marrow cannot produce sufficient numbers of normal RBCs to maintain life. The only available treatments for beta thalassemia major are life-long RBC transfusions (sometimes called hypertransfusion) or bone marrow transplantation. For purposes of 7.05D, we do not consider prophylactic RBC transfusions to prevent strokes or other complications in sickle cell disease and its variants to be of equal significance to life-saving RBC transfusions for beta thalassemia major. However, we will consider the functional limitations associated with prophylactic RBC transfusions and any associated side effects (for example, iron overload) under 7.18 and any affected body system(s). We will also evaluate strokes and resulting complications under 11.00 and 12.00.

D. What are disorders of thrombosis and hemostasis, and how do we evaluate them under 7.08?

1. *Disorders of thrombosis and hemostasis* include both clotting and bleeding disorders, and may be congenital or acquired. These disorders are characterized by abnormalities in blood clotting that result in hypercoagulation (excessive blood clotting) or hypocoagulation (inadequate blood clotting). The diagnosis of a thrombosis or hemostasis disorder is based on evaluation of plasma clotting-factor proteins (factors) and platelets. Protein C or protein S deficiency and Factor V Leiden are examples of hypercoagulation disorders. Hemophilia, von Willebrand disease, and thrombocytopenia are examples of hypocoagulation disorders. Acquired excessive blood clotting may result from blood protein defects and acquired inadequate blood clotting (for example, acquired hemophilia A) may be associated with inhibitor autoantibodies.

2. The hospitalizations in 7.08 do not all have to be for the same complication of a disorder of thrombosis and hemostasis. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include anemias, thromboses, embolisms, and uncontrolled bleeding requiring multiple factor concentrate infusions or platelet transfusions. We will also consider any surgery that you have, even if it is not related to your hematological disorder, to be a complication of your disorder of thrombosis and hemostasis if you require treatment with clotting-factor proteins (for example, factor VIII or factor IX) or anticoagulant medication to control bleeding or coagulation in connection with your surgery. We will count the hours you receive emergency treatment in a comprehensive hemophilia treatment center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

E. What are disorders of bone marrow failure, and how do we evaluate them under 7.10?

1. *Disorders of bone marrow failure* may be congenital or acquired, characterized by bone marrow that does not make enough healthy RBCs, platelets, or granulocytes (specialized types of white blood cells); there may also be a combined failure of these bone marrow-produced cells. The diagnosis is based on peripheral blood smears and bone marrow aspiration or bone marrow biopsy, but not peripheral blood smears alone. Examples of these disorders are myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis. Acquired disorders of bone marrow failure may result from viral infections, chemical exposure, or immunologic disorders.

2. The hospitalizations in 7.10A do not all have to be for the same complication of bone marrow failure. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include uncontrolled bleeding, anemia, and systemic bacterial, viral, or fungal infections.

3. For 7.10B, the requirement of life-long RBC transfusions to maintain life in myelodysplastic syndromes or aplastic anemias has the same meaning as it does for beta thalassemia major. (See 7.00C4.)

F. How do we evaluate bone marrow or stem cell transplantation under 7.17?

We will consider you to be disabled for 12 months from the date of bone marrow or stem cell transplantation, or we may consider you to be disabled for a longer period if you are experiencing any serious post-transplantation complications, such as graft-versus-host (GVH) disease, frequent infections after immunosuppressive therapy, or significant deterioration of organ systems. We do not restrict our determination of the onset of disability to the date of the transplantation in 7.17. We may establish an earlier onset date of disability due to your

transplantation if evidence in your case record supports such a finding.

G. How do we use the functional criteria in 7.18?

1. When we use the functional criteria in 7.18, we consider all relevant information in your case record to determine the impact of your hematological disorder on your ability to function independently, appropriately, effectively, and on a sustained basis in a work setting. Factors we will consider when we evaluate your functioning under 7.18 include, but are not limited to: Your symptoms, the frequency and duration of complications of your hematological disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

2. *Repeated complications* means that the complications occur on an average of three times a year, or once every 4 months, each lasting 2 weeks or more; or the complications do not last for 2 weeks but occur substantially more frequently than three times in a year or once every 4 months; or they occur less frequently than an average of three times a year or once every 4 months but last substantially longer than 2 weeks. Your impairment will satisfy this criterion regardless of whether you have the same kind of complication repeatedly, all different complications, or any other combination of complications; for example, two of the same kind of complication and a different one. You must have the required number of complications with the frequency and duration required in this section. Additionally, the complications must occur within the period we are considering in connection with your application or continuing disability review.

3. To satisfy the functional criteria in 7.18, your hematological disorder must result in a ''marked'' level of limitation in one of three general areas of functioning: Activities of daily living, social functioning, or difficulties in completing tasks due to deficiencies in concentration, persistence, or pace. Functional limitations may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This limitation could result from persistent or intermittent symptoms, such as pain, severe fatigue, or malaise, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. (*Severe fatigue* means a frequent sense of exhaustion that results in significant reduced physical activity or mental function. *Malaise* means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.) You may also have limitations because of your treatment and its side effects.

4. *Marked* limitation means that the symptoms and signs of your hematological disorder interfere *seriously* with your ability to function. Although we do not require the use of such a scale, ''marked'' would be the fourth point on a five-point scale consisting of no limitation, mild limitation, moderate limitation, marked limitation, and extreme limitation. We do not define ''marked'' by a specific number of different activities of daily living or different behaviors in which your social functioning is impaired, or a specific number of tasks that you are able to complete, but by the nature and overall degree of interference with your functioning. You may have a marked limitation when several activities or functions are impaired, or even when only one is impaired. Additionally, you need not be totally precluded from performing an activity to have a marked limitation, as long as the degree of limitation interferes seriously with your ability to function independently, appropriately, and effectively. The term ''marked'' does not imply that you must be confined to bed, hospitalized, or in a nursing home.

5. *Activities of daily living* include, but are not limited to, such activities as doing household chores, grooming and hygiene, using a post office, taking public transportation, or paying bills. We will find that you have a ''marked'' limitation in activities of daily living if you have a serious limitation in your ability to maintain a household or take public transportation because of symptoms such as pain, severe fatigue, anxiety, or difficulty concentrating, caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to perform some self-care activities.

6. *Social functioning* includes the capacity to interact with others independently, appropriately, effectively, and on a sustained basis. It includes the ability to communicate effectively with others. We will find that you have a ''marked'' limitation in maintaining social functioning if you have a serious limitation in social interaction on a sustained basis because of symptoms such as pain, severe fatigue, anxiety, or difficulty concentrating, or a pattern of exacerbation and remission, caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to communicate with close friends or relatives.

7. *Completing tasks in a timely manner* involves the ability to sustain concentration, persistence, or pace to permit timely completion of tasks commonly found in work settings. We will find that you have a ''marked'' limitation in completing tasks if you have a serious limitation in your ability to sustain concentration or pace adequate to complete work-related tasks because of

symptoms, such as pain, severe fatigue, anxiety, or difficulty concentrating caused by your hematological disorder (including complications of the disorder) or its treatment, even if you are able to do some routine activities of daily living.

H. How do we consider your symptoms, including your pain, severe fatigue, and malaise?

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your hematological disorder(s) meets or medically equals a listing, or in our determination whether you are otherwise able to work. We cannot consider your symptoms unless you have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 7.00 and in our other regulations. (See sections 404.1528, 404.1529, 416.928, and 416.929 of this chapter.) Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

I. How do we evaluate episodic events in hematological disorders?

Some of the listings in this body system require a specific number of events within a consecutive 12-month period. (See 7.05, 7.08, and 7.10A.) When we use such criteria, a consecutive 12-month period means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with your application or continuing disability review. These events must occur at least 30 days apart to ensure that we are evaluating separate events.

J. How do we evaluate hematological disorders that do not meet one of these listings?

1. These listings are only common examples of hematological disorders that we consider severe enough to prevent a person from doing any gainful activity. If your disorder does not meet the criteria of any of these listings, we must consider whether you have a disorder that satisfies the criteria of a listing in another body system. For example, we will evaluate hemophilic joint deformity or bone or joint pain from myelofibrosis under 1.00; polycythemia vera under 3.00, 4.00, or 11.00; chronic iron overload resulting from repeated RBC transfusion (transfusion hemosiderosis) under 3.00, 4.00, or 5.00; and the effects of intracranial bleeding or stroke under 11.00 or 12.00.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See sections 404.1526 and 416.926 of this chapter.) Hematological disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. We proceed to the fourth, and, if necessary, the fifth steps of the sequential evaluation process in sections 404.1520 and 416.920. We use the rules in sections 404.1594, 416.994, and 416.994a of this chapter, as appropriate, when we decide whether you continue to be disabled.

7.01 Category of Impairments, Hematological Disorders

7.05 *Hemolytic anemias*, including sickle cell disease, thalassemia, and their variants (see 7.00C), with:

A. Documented painful (vaso-occlusive) crises requiring parenteral (intravenous or intramuscular) narcotic medication, occurring at least six times within a 12-month period with at least 30 days between crises.

OR

B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 7.00C2).

OR

C. Hemoglobin measurements of 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12-month period with at least 30 days between measurements.

OR

D. Beta thalassemia major requiring lifelong RBC transfusions at least once every 6 weeks to maintain life (see 7.00C4).

7.08 *Disorders of thrombosis and hemostasis*, including hemophilia and thrombocytopenia (see 7.00D), with complications requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive hemophilia treatment center immediately before the hospitalization (see 7.00D2).

7.10 *Disorders of bone marrow failure*, including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis (see 7.00E), with:

A. Complications of bone marrow failure requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department immediately before the hospitalization (see 7.00E2).

OR

B. Myelodysplastic syndromes or aplastic anemias requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 7.00E3).

7.17 *Hematological disorders treated by bone marrow or stem cell transplantation* (see 7.00F). Consider under a disability for at least 12 consecutive months from the date of transplantation. After that, evaluate any residual impairment(s) under the criteria for the affected body system.

7.18 *Repeated complications of hematological disorders* (see 7.00G2), including those complications listed in 7.05, 7.08, and 7.10 but without the requisite findings for those listings, or other complications (for example, anemia, osteonecrosis, retinopathy, skin ulcers, silent central nervous system infarction, cognitive or other mental limitation, or limitation of joint movement), resulting in significant, documented symptoms or signs (for example, pain, severe fatigue, malaise, fever, night sweats, headaches, joint or muscle swelling, or shortness of breath), and one of the following at the marked level (see 7.00G4):

A. Limitation of activities of daily living (see 7.00G5).

B. Limitation in maintaining social functioning (see 7.00G6).

C. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace (see 7.00G7).

8.00 Skin Disorders

A. *What skin disorders do we evaluate with these listings?* We use these listings to evaluate skin disorders that may result from hereditary, congenital, or acquired pathological processes. The kinds of impairments covered by these listings are: Ichthyosis, bullous diseases, chronic infections of the skin or mucous membranes, dermatitis, hidradenitis suppurativa, genetic photosensitivity disorders, and burns.

B. *What documentation do we need?* When we evaluate the existence and severity of your skin disorder, we generally need information about the onset, duration, frequency of flareups, and prognosis of your skin disorder; the location, size, and appearance of lesions; and, when applicable, history of exposure to toxins, allergens, or irritants, familial incidence, seasonal variation, stress factors, and your ability to function outside of a highly protective environment. To confirm the diagnosis, we may need laboratory findings (for example, results of a biopsy obtained independently of Social Security disability evaluation or blood tests) or evidence from other medically acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

C. *How do we assess the severity of your skin disorder(s)?* We generally base our assessment of severity on the extent of your skin lesions, the frequency of flareups of your skin lesions, how your symptoms (including pain) limit you, the extent of your treatment, and how your treatment affects you.

1. *Extensive skin lesions.* Extensive skin lesions are those that involve multiple body sites or critical body areas, and result in a very serious limitation. Examples of extensive skin lesions that result in a very serious limitation include but are not limited to:

a. Skin lesions that interfere with the motion of your joints and that very seriously limit your use of more than one extremity; that is, two upper extremities, two lower extremities, or one upper and one lower extremity.

b. Skin lesions on the palms of both hands that very seriously limit your ability to do fine and gross motor movements.

c. Skin lesions on the soles of both feet, the perineum, or both inguinal areas that very seriously limit your ability to ambulate.

2. *Frequency of flareups.* If you have skin lesions, but they do not meet the requirements of any of the listings in this body system, you may still have an impairment that prevents you from doing any gainful activity when we consider your condition over time, especially if your flareups result in extensive skin lesions, as defined in C1 of this section. Therefore, if you have frequent flareups, we may find that your impairment(s) is medically equal to one of these listings even though you have some periods during which your condition is in remission. We will consider how frequent and serious your flareups are, how quickly they resolve, and how you function between flareups to determine whether you have been unable to do any gainful activity for a continuous period of at least 12 months or can be expected to be unable to do any gainful activity for a continuous period of at least 12 months. We will also consider the frequency of your flareups when we determine whether you have a severe impairment and when we need to assess your residual functional capacity.

3. *Symptoms (including pain).* Symptoms (including pain) may be important factors contributing to the severity of your skin disorder(s). We assess the impact of symptoms as explained in §§ 404.1528, 404.1529, 416.928, and 416.929 of this chapter.

4. *Treatment.* We assess the effects of medication, therapy, surgery, and any other form of treatment you receive when we determine the severity and duration of your impairment(s). Skin disorders frequently respond

to treatment; however, response to treatment can vary widely, with some impairments becoming resistant to treatment. Some treatments can have side effects that can in themselves result in limitations.

a. We assess the effects of continuing treatment as prescribed by determining if there is improvement in the symptoms, signs, and laboratory findings of your disorder, and if you experience side effects that result in functional limitations. To assess the effects of your treatment, we may need information about:

i. The treatment you have been prescribed (for example, the type, dosage, method, and frequency of administration of medication or therapy);

ii. Your response to the treatment;

iii. Any adverse effects of the treatment; and

iv. The expected duration of the treatment.

b. Because treatment itself or the effects of treatment may be temporary, in most cases sufficient time must elapse to allow us to evaluate the impact and expected duration of treatment and its side effects. Except under 8.07 and 8.08, you must follow continuing treatment as prescribed for at least 3 months before your impairment can be determined to meet the requirements of a skin disorder listing. (See 8.00H if you are not undergoing treatment or did not have treatment for 3 months.) We consider your specific response to treatment when we evaluate the overall severity of your impairment.

D. *How do we assess impairments that may affect the skin and other body systems?* When your impairment affects your skin and has effects in other body systems, we first evaluate the predominant feature of your impairment under the appropriate body system. Examples include, but are not limited to the following.

1. *Tuberous sclerosis* primarily affects the brain. The predominant features are seizures, which we evaluate under the neurological listings in 11.00, and developmental delays or other mental disorders, which we evaluate under the mental disorders listings in 12.00.

2. *Malignant tumors of the skin* (for example, malignant melanomas) are cancers, or neoplastic diseases, which we evaluate under the listings in 13.00.

3. *Autoimmune disorders and other immune system disorders* (for example, systemic lupus erythematosus (SLE), scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than one body system. We first evaluate these disorders under the immune system disorders listings in 14.00. We evaluate SLE under 14.02, scleroderma under 14.04, HIV infection under 14.08, and Sjögren's syndrome under 14.10.

4. *Disfigurement or deformity* resulting from skin lesions may result in loss of sight, hearing, speech, and the ability to chew (mastication). We evaluate these impairments and their effects under the special senses and speech listings in 2.00 and the digestive system listings in 5.00. Facial disfigurement or other physical deformities may also have effects we evaluate under the mental disorders listings in 12.00, such as when they affect mood or social functioning.

E. *How do we evaluate genetic photosensitivity disorders?*

1. *Xeroderma pigmentosum (XP).* When you have XP, your impairment meets the requirements of 8.07A if you have clinical and laboratory findings showing that you have the disorder. (See 8.00E3.) People who have XP have a lifelong hypersensitivity to all forms of ultraviolet light and generally lead extremely restricted lives in highly protective environments in order to prevent skin cancers from developing. Some people with XP also experience problems with their eyes, neurological problems, mental disorders, and problems in other body systems.

2. *Other genetic photosensitivity disorders.* Other genetic photosensitivity disorders may vary in their effects on different people, and may not result in an inability to engage in any gainful activity for a continuous period of at least 12 months. Therefore, if you have a genetic photosensitivity disorder other than XP (established by clinical and laboratory findings as described in 8.00E3), you must show that you have either extensive skin lesions or an inability to function outside of a highly protective environment to meet the requirements of 8.07B. You must also show that your impairment meets the duration requirement. By *inability to function outside of a highly protective environment* we mean that you must avoid exposure to ultraviolet light (including sunlight passing through windows and light from unshielded fluorescent bulbs), wear protective clothing and eyeglasses, and use opaque broad-spectrum sunscreens in order to avoid skin cancer or other serious effects. Some genetic photosensitivity disorders can have very serious effects in other body systems, especially special senses and speech (2.00), neurological (11.00), mental (12.00), and neoplastic (13.00). We will evaluate the predominant feature of your impairment under the appropriate body system, as explained in 8.00D.

3. *Clinical and laboratory findings.*

a. *General.* We need documentation from an acceptable medical source, as defined in §§ 404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment. In general, we must have evidence of appropriate laboratory testing showing that you have XP or another genetic photosensitivity disorder. We will find that you have XP or another genetic photosensitivity disorder based on a report

from an acceptable medical source indicating that you have the impairment, supported by definitive genetic laboratory studies documenting appropriate chromosomal changes, including abnormal DNA repair or another DNA or genetic abnormality specific to your type of photosensitivity disorder.

b. *What we will accept as medical evidence instead of the actual laboratory report.* When we do not have the actual laboratory report, we need evidence from an acceptable medical source that includes appropriate clinical findings for your impairment and that is persuasive that a positive diagnosis has been confirmed by appropriate laboratory testing at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive genetic laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

F. *How do we evaluate burns?* Electrical, chemical, or thermal burns frequently affect other body systems; for example, musculoskeletal, special senses and speech, respiratory, cardiovascular, renal, neurological, or mental. Consequently, we evaluate burns the way we evaluate other disorders that can affect the skin and other body systems, using the listing for the predominant feature of your impairment. For example, if your soft tissue injuries are under continuing surgical management (as defined in 1.00M), we will evaluate your impairment under 1.08. However, if your burns do not meet the requirements of 1.08 and you have extensive skin lesions that result in a very serious limitation (as defined in 8.00C1) that has lasted or can be expected to last for a continuous period of at least 12 months, we will evaluate them under 8.08.

G. *How do we determine if your skin disorder(s) will continue at a disabling level of severity in order to meet the duration requirement?* For all of these skin disorder listings except 8.07 and 8.08, we will find that your impairment meets the duration requirement if your skin disorder results in extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. By *persist,* we mean that the longitudinal clinical record shows that, with few exceptions, your lesions have been at the level of severity specified in the listing. For 8.07A, we will presume that you meet the duration requirement. For 8.07B and 8.08, we will consider all of the relevant medical and other information in your case record to determine whether your skin disorder meets the duration requirement.

H. *How do we assess your skin disorder(s) if your impairment does not meet the requirements of one of these listings?*

1. These listings are only examples of common skin disorders that we consider severe enough to prevent you from engaging in any gainful activity. For most of these listings, if you do not have continuing treatment as prescribed, if your treatment has not lasted for at least 3 months, or if you do not have extensive skin lesions that have persisted for at least 3 months, your impairment cannot meet the requirements of these skin disorder listings. (This provision does not apply to 8.07 and 8.08.) However, we may still find that you are disabled because your impairment(s) meets the requirements of a listing in another body system or medically equals the severity of a listing. (See §§404.1526 and 416.926 of this chapter.) We may also find you disabled at the last step of the sequential evaluation process.

2. If you have not received ongoing treatment or do not have an ongoing relationship with the medical community despite the existence of a severe impairment(s), or if your skin lesions have not persisted for at least 3 months but you are undergoing continuing treatment as prescribed, you may still have an impairment(s) that meets a listing in another body system or that medically equals a listing. If you do not have an impairment(s) that meets or medically equals a listing, we will assess your residual functional capacity and proceed to the fourth and, if necessary, the fifth step of the sequential evaluation process in §§404.1520 and 416.920 of this chapter. When we decide whether you continue to be disabled, we use the rules in §§404.1594 and 416.994 of this chapter.

8.01 Category of Impairments, Skin Disorders

8.02 *Ichthyosis,* with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.03 *Bullous disease* (for example, pemphigus, erythema multiforme bullosum, epidermolysis bullosa, bullous pemphigoid, dermatitis herpetiformis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.04 *Chronic infections of the skin or mucous membranes,* with extensive fungating or extensive ulcerating skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.05 *Dermatitis* (for example, psoriasis, dyshidrosis, atopic dermatitis, exfoliative dermatitis, allergic contact dermatitis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.06 *Hidradenitis suppurativa,* with extensive skin lesions involving both axillae, both inguinal areas or the perineum that persist for at least 3 months despite continuing treatment as prescribed.

8.07 *Genetic photosensitivity disorders*, established as described in 8.00E.

A. Xeroderma pigmentosum. Consider the individual disabled from birth.

B. Other genetic photosensitivity disorders, with:

1. Extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months, or

2. Inability to function outside of a highly protective environment for a continuous period of at least 12 months (see 8.00E2).

8.08 *Burns,* with extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months (see 8.00F).

9.00 ENDOCRINE DISORDERS

A. *What is an endocrine disorder?*

An endocrine disorder is a medical condition that causes a hormonal imbalance. When an endocrine gland functions abnormally, producing either too much of a specific hormone (hyperfunction) or too little (hypofunction), the hormonal imbalance can cause various complications in the body. The major glands of the endocrine system are the pituitary, thyroid, parathyroid, adrenal, and pancreas.

B. *How do we evaluate the effects of endocrine disorders?* We evaluate impairments that result from endocrine disorders under the listings for other body systems. For example:

1. *Pituitary gland disorders* can disrupt hormone production and normal functioning in other endocrine glands and in many body systems. The effects of pituitary gland disorders vary depending on which hormones are involved. For example, when pituitary hypofunction affects water and electrolyte balance in the kidney and leads to diabetes insipidus, we evaluate the effects of recurrent dehydration under 6.00.

2. *Thyroid gland disorders* affect the sympathetic nervous system and normal metabolism. We evaluate thyroid-related changes in blood pressure and heart rate that cause arrhythmias or other cardiac dysfunction under 4.00; thyroid-related weight loss under 5.00; hypertensive cerebrovascular accidents (strokes) under 11.00; and cognitive limitations, mood disorders, and anxiety under 12.00.

3. *Parathyroid gland disorders* affect calcium levels in bone, blood, nerves, muscle, and other body tissues. We evaluate parathyroid-related osteoporosis and fractures under 1.00; abnormally elevated calcium levels in the blood (hypercalcemia) that lead to cataracts under 2.00; kidney failure under 6.00; and recurrent abnormally low blood calcium levels (hypocalcemia) that lead to increased excitability of nerves and muscles, such as tetany and muscle spasms, under 11.00.

4. *Adrenal gland disorders* affect bone calcium levels, blood pressure, metabolism, and mental status. We evaluate adrenal-related osteoporosis with fractures that compromises the ability to walk or to use the upper extremities under 1.00; adrenal-related hypertension that worsens heart failure or causes recurrent arrhythmias under 4.00; adrenal-related weight loss under 5.00; and mood disorders under 12.00.

5. *Diabetes mellitus and other pancreatic gland disorders* disrupt the production of several hormones, including insulin, that regulate metabolism and digestion. Insulin is essential to the absorption of glucose from the bloodstream into body cells for conversion into cellular energy. The most common pancreatic gland disorder is *diabetes mellitus* (DM). There are two major types of DM: type 1 and type 2. Both type 1 and type 2 DM are chronic disorders that can have serious disabling complications that meet the duration requirement. Type 1 DM—previously known as ''juvenile diabetes'' or ''insulin-dependent diabetes mellitus'' (IDDM)—is an absolute deficiency of insulin production that commonly begins in childhood and continues throughout adulthood. Treatment of type 1 DM always requires lifelong daily insulin. With type 2 DM—previously known as ''adult-onset diabetes mellitus'' or ''non-insulin-dependent diabetes mellitus'' (NIDDM)—the body's cells resist the effects of insulin, impairing glucose absorption and metabolism. Treatment of type 2 DM generally requires lifestyle changes, such as increased exercise and dietary modification, and sometimes insulin in addition to other medications. While both type 1 and type 2 DM are usually controlled, some persons do not achieve good control for a variety of reasons including, but not limited to, hypoglycemia unawareness, other disorders that can affect blood glucose levels, inability to manage DM due to a mental disorder, or inadequate treatment.

a. *Hyperglycemia.* Both types of DM cause hyperglycemia, which is an abnormally high level of blood glucose that may produce acute and long-term complications. Acute complications of hyperglycemia include diabetic ketoacidosis. Long-term complications of chronic hyperglycemia include many conditions affecting various body systems.

(i) *Diabetic ketoacidosis (DKA).* DKA is an acute, potentially life-threatening complication of DM in which the chemical balance of the body becomes dangerously hyperglycemic and acidic. It results from a severe insulin deficiency, which can occur due to missed or inadequate daily insulin therapy or in association with an acute illness. It usually requires hospital treatment to correct the acute complications of dehydration, electrolyte imbalance, and insulin deficiency. You may have serious complications resulting from your treatment, which we evaluate under the affected body system. For example, we evaluate cardiac arrhythmias under 4.00, intestinal necrosis under 5.00, and cerebral edema and seizures under

11.00. Recurrent episodes of DKA may result from mood or eating disorders, which we evaluate under 12.00.

(ii) *Chronic hyperglycemia.* Chronic hyperglycemia, which is longstanding abnormally high levels of blood glucose, leads to long-term diabetic complications by disrupting nerve and blood vessel functioning. This disruption can have many different effects in other body systems. For example, we evaluate diabetic peripheral neurovascular disease that leads to gangrene and subsequent amputation of an extremity under 1.00; diabetic retinopathy under 2.00; coronary artery disease and peripheral vascular disease under 4.00; diabetic gastroparesis that results in abnormal gastrointestinal motility under 5.00; diabetic nephropathy under 6.00; poorly healing bacterial and fungal skin infections under 8.00; diabetic peripheral and sensory neuropathies under 11.00; and cognitive impairments, depression, and anxiety under 12.00.

b. *Hypoglycemia.* Persons with DM may experience episodes of hypoglycemia, which is an abnormally low level of blood glucose. Most adults recognize the symptoms of hypoglycemia and reverse them by consuming substances containing glucose; however, some do not take this step because of hypoglycemia unawareness. Severe hypoglycemia can lead to complications, including seizures or loss of consciousness, which we evaluate under 11.00, or altered mental status and cognitive deficits, which we evaluate under 12.00.

C. *How do we evaluate endocrine disorders that do not have effects that meet or medically equal the criteria of any listing in other body systems?* If your impairment(s) does not meet or medically equal a listing in another body system, you may or may not have the residual functional capacity to engage in substantial gainful activity. In this situation, we proceed to the fourth and, if necessary, the fifth steps of the sequential evaluation process in §§404.1520 and 416.920. When we decide whether you continue to be disabled, we use the rules in §§404.1594, 416.994, and 416.994a.

10.00 Congenital Disorders That Affect Multiple Body Systems

A. *Which disorder do we evaluate under this body system?* Although Down syndrome exists in non-mosaic and mosaic forms, we evaluate only non-mosaic Down syndrome under this body system.

B. *What is non-mosaic Down syndrome?* Non-mosaic Down syndrome is a genetic disorder. Most people with non-mosaic Down syndrome have three copies of chromosome 21 in all of their cells (chromosome 21 trisomy); some have an extra copy of chromosome 21 attached to a different chromosome in all of their cells (chromosome 21 translocation). Virtually all people with non-mosaic Down syndrome have characteristic facial or other physical features, delayed physical development, and intellectual disability. People with non-mosaic Down syndrome may also have congenital heart disease, impaired vision, hearing problems, and other disorders. We evaluate non-mosaic Down syndrome under 10.06. If you have non-mosaic Down syndrome documented as described in 10.00C, we consider you disabled from birth.

C. What evidence do we need to document non-mosaic Down syndrome under 10.06?

1. Under 10.06A, we will find you disabled based on laboratory findings.

a. To find that your disorder meets 10.06A, we need a copy of the laboratory report of karyotype analysis, which is the definitive test to establish non-mosaic Down syndrome. We will not purchase karyotype analysis. We will not accept a fluorescence in situ hybridization (FISH) test because it does not distinguish between the mosaic and non-mosaic forms of Down syndrome.

b. If a physician (see §§404.1513(a)(1) and 416.913(a)(1) of this chapter) has not signed the laboratory report of karyotype analysis, the evidence must also include a physician's statement that you have Down syndrome.

c. For purposes of 10.06A, we do not require additional evidence stating that you have the distinctive facial or other physical features of Down syndrome.

2. If we do not have a laboratory report of karyotype analysis showing that you have non-mosaic Down syndrome, we may find you disabled under 10.06B or 10.06C.

a. Under 10.06B, we need a physician's report stating: (i) your karyotype diagnosis or evidence that documents your type of Down syndrome is consistent with prior karyotype analysis (for example, reference to a diagnosis of ''trisomy 21''), and (ii) that you have the distinctive facial or other physical features of Down syndrome. We do not require a detailed description of the facial or other physical features of the disorder. However, we will not find that your disorder meets 10.06B if we have evidence—such as evidence of functioning inconsistent with the diagnosis—that indicates that you do not have non-mosaic Down syndrome.

b. If we do not have evidence of prior karyotype analysis (you did not have testing, or you had testing but we do not have information from a physician about the test results), we will find that your disorder meets 10.06C if we have: (i) a physician's report stating that you have the distinctive facial or other physical features of Down syndrome, and (ii) evidence that your functioning is consistent with a diagnosis of non-mosaic Down syndrome. This evidence may include medical or nonmedical information about your physical and mental abilities, including information about your education, work history, or the results of psychological testing. However, we will not find that your

disorder meets 10.06C if we have evidence—such as evidence of functioning inconsistent with the diagnosis—that indicates that you do not have non-mosaic Down syndrome.

D. How do we evaluate mosaic Down syndrome and other congenital disorders that affect multiple body systems?

1. *Mosaic Down syndrome.* Approximately 2 percent of people with Down syndrome have the mosaic form. In mosaic Down syndrome, there are some cells with an extra copy of chromosome 21 and other cells with the normal two copies of chromosome 21. Mosaic Down syndrome can be so slight as to be undetected clinically, but it can also be profound and disabling, affecting various body systems.

2. *Other congenital disorders that affect multiple body systems.* Other congenital disorders, such as congenital anomalies, chromosomal disorders, dysmorphic syndromes, inborn metabolic syndromes, and perinatal infectious diseases, can cause deviation from, or interruption of, the normal function of the body or can interfere with development. Examples of these disorders include both the juvenile and late-onset forms of Tay-Sachs disease, trisomy X syndrome (XXX syndrome), fragile X syndrome, phenylketonuria (PKU), caudal regression syndrome, and fetal alcohol syndrome. For these disorders and other disorders like them, the degree of deviation, interruption, or interference, as well as the resulting functional limitations and their progression, may vary widely from person to person and may affect different body systems.

3. *Evaluating the effects of mosaic Down syndrome or another congenital disorder under the listings.* When the effects of mosaic Down syndrome or another congenital disorder that affects multiple body systems are sufficiently severe we evaluate the disorder under the appropriate affected body system(s), such as musculoskeletal, special senses and speech, neurological, or mental disorders. Otherwise, we evaluate the specific functional limitations that result from the disorder under our other rules described in 10.00E.

E. What if your disorder does not meet a listing?

If you have a severe medically determinable impairment(s) that does not meet a listing, we will consider whether your impairment(s) medically equals a listing. See §§ 404.1526 and 416.926 of this chapter. If your impairment(s) does not meet or medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. We proceed to the fourth, and if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920 of this chapter. We use the rules in §§ 404.1594 and 416.994 of this chapter, as appropriate, when we decide whether you continue to be disabled.

10.01 CATEGORY OF IMPAIRMENTS, CONGENITAL DISORDERS THAT AFFECT MULTIPLE BODY SYSTEMS

10.06 *Non-mosaic Down syndrome* (chromosome 21 trisomy or chromosome 21 translocation), documented by:

A. A laboratory report of karyotype analysis signed by a physician, or both a laboratory report of karyotype analysis not signed by a physician *and* a statement by a physician that you have Down syndrome (see 10.00C1), or

B. A physician's report stating that you have chromosome 21 trisomy or chromosome 21 translocation consistent with prior karyotype analysis with the distinctive facial or other physical features of Down syndrome (see 10.00C2a), or

C. A physician's report stating that you have Down syndrome with the distinctive facial or other physical features *and* evidence demonstrating that you function at a level consistent with non-mosaic Down syndrome (see 10.00C2b).

11.00 NEUROLOGICAL

A. *Epilepsy.* In epilepsy, regardless of etiology, degree of impairment will be determined according to type, frequency, duration, and sequelae of seizures. At least one detailed description of a typical seizure is required. Such description includes the presence or absence of aura, tongue bites, sphincter control, injuries associated with the attack, and postictal phenomena. The reporting physician should indicate the extent to which description of seizures reflects his own observations and the source of ancillary information. Testimony of persons other than the claimant is essential for description of type and frequency of seizures if professional observation is not available.

Under 11.02 and 11.03, the criteria can be applied only if the impairment persists despite the fact that the individual is following prescribed antiepileptic treatment. Adherence to prescribed antiepileptic therapy can ordinarily be determined from objective clinical findings in the report of the physician currently providing treatment for epilepsy. Determination of blood levels of phenytoin sodium or other antiepileptic drugs may serve to indicate whether the prescribed medication is being taken. When seizures are occurring at the frequency stated in 11.02 or 11.03, evalution of the severity of the impairment must include consideration of the serum drug levels. Should serum drug levels appear therapeutically inadequate, consideration should be given as to whether this is caused by individual idiosyncrasy in absorption of metabolism of the drug. Blood drug levels should be evaluated in conjunction

with all the other evidence to determine the extent of compliance. When the reported blood drug levels are low, therefore, the information obtained from the treating source should include the physician's statement as to why the levels are low and the results of any relevant diagnostic studies concerning the blood levels. Where adequate seizure control is obtained only with unusually large doses, the possibility of impairment resulting from the side effects of this medication must be also assessed. Where documentation shows that use of alcohol or drugs affects adherence to prescribed therapy or may play a part in the precipitation of seizures, this must also be considered in the overall assessment of impairment level.

B. *Brain tumors.* We evaluate malignant brain tumors under the criteria in 13.13. For benign brain tumors, we determine the severity and duration of the impairment on the basis of symptoms, signs, and laboratory findings (11.05).

C. *Persistent disorganization of motor function* in the form of paresis or paralysis, tremor or other involuntary movements, ataxia and sensory disturbances (any or all of which may be due to cerebral, cerebellar, brain stem, spinal cord, or peripheral nerve dysfunction) which occur singly or in various combinations, frequently provides the sole or partial basis for decision in cases of neurological impairment. The assessment of impairment depends on the degree of interference with locomotion and/or interference with the use of fingers, hands, and arms.

D. *In conditions which are episodic in character,* such as multiple sclerosis or myasthenia gravis, consideration should be given to frequency and duration of exacerbations, length of remissions, and permanent residuals.

E. *Multiple sclerosis.* The major criteria for evaluating impairment caused by multiple sclerosis are discussed in listing 11.09. Paragraph A provides criteria for evaluating disorganization of motor function and gives reference to 11.04B (11.04B then refers to 11.00C). Paragraph B provides references to other listings for evaluating visual or mental impairments caused by multiple sclerosis. Paragraph C provides criteria for evaluating the impairment of individuals who do not have muscle weakness or other significant disorganization of motor function at rest, but who do develop muscle weakness on activity as a result of fatigue.

Use of the criteria in 11.09C is dependent upon (1) documenting a diagnosis of multiple sclerosis, (2) obtaining a description of fatigue considered to be characteristic of multiple sclerosis, and (3) obtaining evidence that the system has actually become fatigued. The evaluation of the magnitude of the impairment must consider the degree of exercise and the severity of the resulting muscle weakness.

The criteria in 11.09C deals with motor abnormalities which occur on activity. If the disorganization of motor function is present at rest, paragraph A must be used, taking into account any further increase in muscle weakness resulting from activity.

Sensory abnormalities may occur, particularly involving central visual acuity. The decrease in visual acuity may occur after brief attempts at activity involving near vision, such as reading. This decrease in visual acuity may not persist when the specific activity is terminated, as with rest, but is predictably reproduced with resumption of the activity. The impairment of central visual acuity in these cases should be evaluated under the criteria in listing 2.02, taking into account the fact that the decrease in visual acuity will wax and wane.

Clarification of the evidence regarding central nervous system dysfunction responsible for the symptoms may require supporting technical evidence of functional impairment such as evoked response tests during exercise.

F. *Traumatic brain injury (TBI).* The guidelines for evaluating impairments caused by cerebral trauma are contained in 11.18. Listing 11.18 states that cerebral trauma is to be evaluated under 11.02, 11.03, 11.04, and 12.02, as applicable.

TBI may result in neurological and mental impairments with a wide variety of posttraumatic symptoms and signs. The rate and extent of recovery can be highly variable and the long-term outcome may be difficult to predict in the first few months post-injury. Generally, the neurological impairment(s) will stabilize more rapidly than any mental impairment(s). Sometimes a mental impairment may appear to improve immediately following TBI and then worsen, or, conversely, it may appear much worse initially but improve after a few months. Therefore, the mental findings immediately following TBI may not reflect the actual severity of your mental impairment(s). The actual severity of a mental impairment may not become apparent until 6 months post-injury.

In some cases, evidence of a profound neurological impairment is sufficient to permit a finding of disability within 3 months post-injury. If a finding of disability within 3 months post-injury is not possible based on any neurological impairment(s), we will defer adjudication of the claim until we obtain evidence of your neurological or mental impairments at least 3 months post-injury. If a finding of disability still is not possible at that time, we will again defer adjudication of the claim until we obtain evidence at least 6 months post-injury. At that time, we will fully evaluate any neurological and mental impairments and adjudicate the claim.

G. *Amyotrophic Lateral Sclerosis (ALS).* 1. Amyotrophic lateral sclerosis (ALS), sometimes called Lou Gehrig's disease, is a progressive, invariably fatal neurological disease that attacks the nerve cells (motor neurons) responsible for controlling voluntary muscles. Eventually, all muscles under voluntary control are affected, and individuals with ALS ultimately lose their ability to move their arms and legs, and their capacity to swallow, speak, and breath. Most people with ALS die from respiratory failure. There is currently no cure for ALS, and most treatments are designed only to relieve symptoms and improve the quality of life.

2. Diagnosis of ALS is based on history, neurological findings consistent with the diagnosis of ALS, and electrophysiological and neuroimaging testing to rule out other impairments that may cause similar signs and symptoms. The diagnosis may also be supported by electrophysiological studies (electromyography or nerve conduction studies), but these tests may be negative or only suggestive of the diagnosis. There is no single test that establishes the existence of ALS.

3. For purposes of 11.10, documentation of the diagnosis must be by generally accepted methods consistent with the prevailing state of medical knowledge and clinical practice. The evidence should include documentation of a clinically appropriate medical history, neurological findings consistent with the diagnosis of ALS, and the results of any electrophysiological and neuroimaging testing.

11.01 Category of Impairments, Neurological

11.02 *Epilepsy—convulsive epilepsy, (grand mal or psychomotor), documented by detailed description of a typical seizure pattern, including all associated phenomena; occurring more frequently than once a month in spite of at least 3 months of prescribed treatment.* With:

A. Daytime episodes (loss of consciousness and convulsive seizures) or

B. Nocturnal episodes manifesting residuals which interfere significantly with activity during the day.

11.03 *Epilepsy—nonconvulsive epilepsy (petit mal, psychomotor, or focal), documented by detailed description of a typical seizure pattern, including all associated phenomena; occurring more frequently than once weekly in spite of at least 3 months of prescribed treatment.* With alteration of awareness or loss of consciousness and transient postictal manifestations of unconventional behavior or significant interference with activity during the day.

11.04 *Central nervous system vascular accident.* With one of the following more than 3 months post-vascular accident:

A. Sensory or motor aphasia resulting in ineffective speech or communication; or

B. Significant and persistent disorganization of motor function in two extremities, resulting in sustained disturbance of gross and dexterous movements, or gait and station (see 11.00C).

11.05 *Benign brain tumors.* Evaluate under 11.02, 11.03, 11.04, or the criteria of the affected body system.

11.06 *Parkinsonian syndrome* with the following signs: Significant rigidity, brady kinesia, or tremor in two extremities, which, singly or in combination, result in sustained disturbance of gross and dexterous movements, or gait and station.

11.07 *Cerebral palsy.* With:

A. IQ of 70 or less; or

B. Abnormal behavior patterns, such as destructiveness or emotional instability: or

C. Significant interference in communication due to speech, hearing, or visual defect; or

D. Disorganization of motor function as described in 11.04B.

11.08 *Spinal cord or nerve root lesions, due to any cause* with disorganization of motor function as described in 11.04B.

11.09 *Multiple sclerosis.* With:

A. Disorganization of motor function as described in 11.04B; or

B. Visual or mental impairment as described under the criteria in 2.02, 2.03, 2.04, or 12.02; or

C. Significant, reproducible fatigue of motor function with substantial muscle weakness on repetitive activity, demonstrated on physical examination, resulting from neurological dysfunction in areas of the central nervous system known to be pathologically involved by the multiple sclerosis process.

11.10 *Amyotrophic lateral sclerosis* established by clinical and laboratory findings, as described in 11.00G.

11.11 *Anterior poliomyelitis.* With:

A. Persistent difficulty with swallowing or breathing; or

B. Unintelligible speech; or

C. Disorganization of motor function as described in 11.04B.

11.12 *Myasthenia gravis.* With:

A. Significant difficulty with speaking, swallowing, or breathing while on prescribed therapy; or

B. Significant motor weakness of muscles of extremities on repetitive activity against resistance while on prescribed therapy.

11.13 *Muscular dystrophy* with disorganization of motor function as described in 11.04B.

11.14 *Peripheral neuropathies.*

With disorganization of motor function as described in 11.04B, in spite of prescribed treatment.

11.15 [Reserved]

11.16 *Subacute combined cord degeneration (pernicious anemia) with disorganization of motor function as decribed in 11.04B or 11.15B, not significantly improved by prescribed treatment.*

11.17 *Degenerative disease not listed elsewhere, such as Huntington's chorea, Friedreich's ataxia, and spino-cerebellar degeneration*. With:

A. Disorganization of motor function as described in 11.04B; or

B. Chronic brain syndrome. Evaluate under 12.02.

11.18 *Cerebral trauma:*

Evaluate under the provisions of 11.02, 11.03, 11.04 and 12.02, as applicable.

11.19 *Syringomyelia*.

With:

A. Significant bulbar signs; or

B. Disorganization of motor function as described in 11.04B.

12.00 MENTAL DISORDERS

A. *Introduction*. The evaluation of disability on the basis of mental disorders requires documentation of a medically determinable impairment(s), consideration of the degree of limitation such impairment(s) may impose on your ability to work, and consideration of whether these limitations have lasted or are expected to last for a continuous period of at least 12 months. The listings for mental disorders are arranged in nine diagnostic categories: Organic mental disorders (12.02); schizophrenic, paranoid and other psychotic disorders (12.03); affective disorders (12.04); intellectual disability (12.05); anxiety-related disorders (12.06); somatoform disorders (12.07); personality disorders (12.08); substance addiction disorders (12.09); and autistic disorder and other pervasive developmental disorders (12.10). Each listing, except 12.05 and 12.09, consists of a statement describing the disorder(s) addressed by the listing, paragraph A criteria (a set of medical findings), and paragraph B criteria (a set of impairment-related functional limitations). There are additional functional criteria (paragraph C criteria) in 12.02, 12.03, 12.04, and 12.06, discussed herein. We will assess the paragraph B criteria before we apply the paragraph C criteria. We will assess the paragraph C criteria only if we find that the paragraph B criteria are not satisfied. We will find that you have a listed impairment if the diagnostic description in the introductory paragraph and the criteria of both paragraphs A and B (or A and C, when appropriate) of the listed impairment are satisfied.

The criteria in paragraph A substantiate medically the presence of a particular mental disorder. Specific symptoms, signs, and laboratory findings in the paragraph A criteria of any of the listings in this section cannot be considered in isolation from the description of the mental disorder contained at the beginning of each listing category. Impairments should be analyzed or reviewed under the mental category(ies) indicated by the medical findings. However, we may also consider mental impairments under physical body system listings, using the concept of medical equivalence, when the mental disorder results in physical dysfunction. (See, for instance, 12.00D12 regarding the evaluation of anorexia nervosa and other eating disorders.)

The criteria in paragraphs B and C describe impairment-related functional limitations that are incompatible with the ability to do any gainful activity. The functional limitations in paragraphs B and C must be the result of the mental disorder described in the diagnostic description, that is manifested by the medical findings in paragraph A.

The structure of the listing for intellectual disability (12.05) is different from that of the other mental disorders listings. Listing 12.05 contains an introductory paragraph with the diagnostic description for intellectual disability. It also contains four sets of criteria (paragraphs A through D). If your impairment satisfies the diagnostic description in the introductory paragraph and any one of the four sets of criteria, we will find that your impairment meets the listing. Paragraphs A and B contain criteria that describe disorders we consider severe enough to prevent your doing any gainful activity without any additional assessment of functional limitations. For paragraph C, we will assess the degree of functional limitation the additional impairment(s) imposes to determine if it significantly limits your physical or mental ability to do basic work activities, *i.e.*, is a ''severe'' impairment(s), as defined in §§ 404.1520(c) and 416.920(c). If the additional impairment(s) does not cause limitations that are ''severe'' as defined in §§ 404.1520(c) and 416.920(c), we will not find that the additional impairment(s) imposes ''an additional and significant work-related limitation of function,'' even if you are unable to do your past work because of the unique features of that work. Paragraph D contains the same functional criteria that are required under paragraph B of the other mental disorders listings.

The structure of the listing for substance addiction disorders, 12.09, is also different from that for the other mental disorder listings. Listing 12.09 is structured as a reference listing; that is, it will only serve to indicate which of the other listed mental or physical impairments must be used to evaluate the behavioral or physical changes resulting from regular use of addictive substances.

The listings are so constructed that an individual with an impairment(s) that meets or is equivalent in severity to the criteria of a listing could not reasonably be expected to do any gainful activity. These listings are only examples of common mental disorders that are considered severe enough to prevent an individual from doing any gainful activity. When you have a medically determinable severe mental impairment that does

not satisfy the diagnostic description or the requirements of the paragraph A criteria of the relevant listing, the assessment of the paragraph B and C criteria is critical to a determination of equivalence.

If your impairment(s) does not meet or is not equivalent in severity to the criteria of any listing, you may or may not have the residual functional capacity (RFC) to do substantial gainful activity (SGA). The determination of mental RFC is crucial to the evaluation of your capacity to do SGA when your impairment(s) does not meet or equal the criteria of the listings, but is nevertheless severe.

RFC is a multidimensional description of the work-related abilities you retain in spite of your medical impairments. An assessment of your RFC complements the functional evaluation necessary for paragraphs B and C of the listings by requiring consideration of an expanded list of work-related capacities that may be affected by mental disorders when your impairment(s) is severe but neither meets nor is equivalent in severity to a listed mental disorder.

B. *Need for medical evidence.* We must establish the existence of a medically determinable impairment(s) of the required duration by medical evidence consisting of symptoms, signs, and laboratory findings (including psychological test findings). Symptoms are your own description of your physical or mental impairment(s). Psychiatric signs are medically demonstrable phenomena that indicate specific psychological abnormalities, e.g., abnormalities of behavior, mood, thought, memory, orientation, development, or perception, as described by an appropriate medical source. Symptoms and signs generally cluster together to constitute recognizable mental disorders described in the listings. The symptoms and signs may be intermittent or continuous depending on the nature of the disorder.

C. *Assessment of severity.* We measure severity according to the functional limitations imposed by your medically determinable mental impairment(s). We assess functional limitations using the four criteria in paragraph B of the listings: Activities of daily living; social functioning; concentration, persistence, or pace; and episodes of decompensation. Where we use ''marked'' as a standard for measuring the degree of limitation, it means more than moderate but less than extreme. A marked limitation may arise when several activities or functions are impaired, or even when only one is impaired, as long as the degree of limitation is such as to interfere seriously with your ability to function independently, appropriately, effectively, and on a sustained basis. See §§ 404.1520a and 416.920a.

1. *Activities of daily living* include adaptive activities such as cleaning, shopping, cooking, taking public transportation, paying bills, maintaining a residence, caring appropriately for your grooming and hygiene, using telephones and directories, and using a post office. In the context of your overall situation, we assess the quality of these activities by their independence, appropriateness, effectiveness, and sustainability. We will determine the extent to which you are capable of initiating and participating in activities independent of supervision or direction.

We do not define ''marked'' by a specific number of different activities of daily living in which functioning is impaired, but by the nature and overall degree of interference with function. For example, if you do a wide range of activities of daily living, we may still find that you have a marked limitation in your daily activities if you have serious difficulty performing them without direct supervision, or in a suitable manner, or on a consistent, useful, routine basis, or without undue interruptions or distractions.

2. *Social functioning* refers to your capacity to interact independently, appropriately, effectively, and on a sustained basis with other individuals. Social functioning includes the ability to get along with others, such as family members, friends, neighbors, grocery clerks, landlords, or bus drivers. You may demonstrate impaired social functioning by, for example, a history of altercations, evictions, firings, fear of strangers, avoidance of interpersonal relationships, or social isolation. You may exhibit strength in social functioning by such things as your ability to initiate social contacts with others, communicate clearly with others, or interact and actively participate in group activities. We also need to consider cooperative behaviors, consideration for others, awareness of others' feelings, and social maturity. Social functioning in work situations may involve interactions with the public, responding appropriately to persons in authority (e.g., supervisors), or cooperative behaviors involving coworkers.

We do not define ''marked'' by a specific number of different behaviors in which social functioning is impaired, but by the nature and overall degree of interference with function. For example, if you are highly antagonistic, uncooperative, or hostile but are tolerated by local storekeepers, we may nevertheless find that you have a marked limitation in social functioning because that behavior is not acceptable in other social contexts.

3. *Concentration, persistence, or pace* refers to the ability to sustain focused attention and concentration sufficiently long to permit the timely and appropriate completion of tasks commonly found in work settings. Limitations in concentration, persistence, or pace are best observed in work settings, but may also be reflected by limitations in other settings. In addition, major limitations in

this area can often be assessed through clinical examination or psychological testing. Wherever possible, however, a mental status examination or psychological test data should be supplemented by other available evidence.

On mental status examinations, concentration is assessed by tasks such as having you subtract serial sevens or serial threes from 100. In psychological tests of intelligence or memory, concentration is assessed through tasks requiring short-term memory or through tasks that must be completed within established time limits.

In work evaluations, concentration, persistence, or pace is assessed by testing your ability to sustain work using appropriate production standards, in either real or simulated work tasks (e.g., filing index cards, locating telephone numbers, or disassembling and reassembling objects). Strengths and weaknesses in areas of concentration and attention can be discussed in terms of your ability to work at a consistent pace for acceptable periods of time and until a task is completed, and your ability to repeat sequences of action to achieve a goal or an objective.

We must exercise great care in reaching conclusions about your ability or inability to complete tasks under the stresses of employment during a normal workday or work week based on a time-limited mental status examination or psychological testing by a clinician, or based on your ability to complete tasks in other settings that are less demanding, highly structured, or more supportive. We must assess your ability to complete tasks by evaluating all the evidence, with an emphasis on how independently, appropriately, and effectively you are able to complete tasks on a sustained basis.

We do not define ''marked'' by a specific number of tasks that you are unable to complete, but by the nature and overall degree of interference with function. You may be able to sustain attention and persist at simple tasks but may still have difficulty with complicated tasks. Deficiencies that are apparent only in performing complex procedures or tasks would not satisfy the intent of this paragraph B criterion. However, if you can complete many simple tasks, we may nevertheless find that you have a marked limitation in concentration, persistence, or pace if you cannot complete these tasks without extra supervision or assistance, or in accordance with quality and accuracy standards, or at a consistent pace without an unreasonable number and length of rest periods, or without undue interruptions or distractions.

4. *Episodes of decompensation* are exacerbations or temporary increases in symptoms or signs accompanied by a loss of adaptive functioning, as manifested by difficulties in performing activities of daily living, maintaining social relationships, or maintaining concentration, persistence, or pace. Episodes of decompensation may be demonstrated by an exacerbation in symptoms or signs that would ordinarily require increased treatment or a less stressful situation (or a combination of the two). Episodes of decompensation may be inferred from medical records showing significant alteration in medication; or documentation of the need for a more structured psychological support system (e.g., hospitalizations, placement in a halfway house, or a highly structured and directing household); or other relevant information in the record about the existence, severity, and duration of the episode.

The term *repeated episodes of decompensation, each of extended duration* in these listings means three episodes within 1 year, or an average of once every 4 months, each lasting for at least 2 weeks. If you have experienced more frequent episodes of shorter duration or less frequent episodes of longer duration, we must use judgment to determine if the duration and functional effects of the episodes are of equal severity and may be used to substitute for the listed finding in a determination of equivalence.

D. *Documentation.* The evaluation of disability on the basis of a mental disorder requires sufficient evidence to (1) establish the presence of a medically determinable mental impairment(s), (2) assess the degree of functional limitation the impairment(s) imposes, and (3) project the probable duration of the impairment(s). See §§ 404.1512 and 416.912 for a discussion of what we mean by ''evidence'' and how we will assist you in developing your claim. Medical evidence must be sufficiently complete and detailed as to symptoms, signs, and laboratory findings to permit an independent determination. In addition, we will consider information you provide from other sources when we determine how the established impairment(s) affects your ability to function. We will consider all relevant evidence in your case record.

1. *Sources of evidence.*

a. *Medical evidence.* There must be evidence from an acceptable medical source showing that you have a medically determinable mental impairment. See §§ 404.1508, 404.1513, 416.908, and 416.913. We will make every reasonable effort to obtain all relevant and available medical evidence about your mental impairment(s), including its history, and any records of mental status examinations, psychological testing, and hospitalizations and treatment. Whenever possible, and appropriate, medical source evidence should reflect the medical source's considerations of information from you and other concerned persons who are aware of your activities of daily living; social functioning; concentration, persistence, or pace; or episodes of decompensation. Also, in accordance with standard clinical practice, any medical

source assessment of your mental functioning should take into account any sensory, motor, or communication abnormalities, as well as your cultural and ethnic background.

b. *Information from the individual.* Individuals with mental impairments can often provide accurate descriptions of their limitations. The presence of a mental impairment does not automatically rule you out as a reliable source of information about your own functional limitations. When you have a mental impairment and are willing and able to describe your limitations, we will try to obtain such information from you. However, you may not be willing or able to fully or accurately describe the limitations resulting from your impairment(s). Thus, we will carefully examine the statements you provide to determine if they are consistent with the information about, or general pattern of, the impairment as described by the medical and other evidence, and to determine whether additional information about your functioning is needed from you or other sources.

c. *Other information.* Other professional health care providers (e.g., psychiatric nurse, psychiatric social worker) can normally provide valuable functional information, which should be obtained when available and needed. If necessary, information should also be obtained from nonmedical sources, such as family members and others who know you, to supplement the record of your functioning in order to establish the consistency of the medical evidence and longitudinality of impairment severity, as discussed in 12.00D2. Other sources of information about functioning include, but are not limited to, records from work evaluations and rehabilitation progress notes.

2. *Need for longitudinal evidence.* Your level of functioning may vary considerably over time. The level of your functioning at a specific time may seem relatively adequate or, conversely, rather poor. Proper evaluation of your impairment(s) must take into account any variations in the level of your functioning in arriving at a determination of severity over time. Thus, it is vital to obtain evidence from relevant sources over a sufficiently long period prior to the date of adjudication to establish your impairment severity.

3. *Work attempts.* You may have attempted to work or may actually have worked during the period of time pertinent to the determination of disability. This may have been an independent attempt at work or it may have been in conjunction with a community mental health or sheltered program, and it may have been of either short or long duration. Information concerning your behavior during any attempt to work and the circumstances surrounding termination of your work effort are particularly useful in determining your ability or inability to function in a work setting. In addition, we should also examine the degree to which you require special supports (such as those provided through supported employment or transitional employment programs) in order to work.

4. *Mental status examination.* The mental status examination is performed in the course of a clinical interview and is often partly assessed while the history is being obtained. A comprehensive mental status examination generally includes a narrative description of your appearance, behavior, and speech; thought process (e.g., loosening of associations); thought content (e.g., delusions); perceptual abnormalities (e.g., hallucinations); mood and affect (e.g., depression, mania); sensorium and cognition (e.g., orientation, recall, memory, concentration, fund of information, and intelligence); and judgment and insight. The individual case facts determine the specific areas of mental status that need to be emphasized during the examination.

5. *Psychological testing.*

a. Reference to a ''standardized psychological test'' indicates the use of a psychological test measure that has appropriate validity, reliability, and norms, and is individually administered by a qualified specialist. By ''qualified,'' we mean the specialist must be currently licensed or certified in the State to administer, score, and interpret psychological tests and have the training and experience to perform the test.

b. Psychological tests are best considered as standardized sets of tasks or questions designed to elicit a range of responses. Psychological testing can also provide other useful data, such as the specialist's observations regarding your ability to sustain attention and concentration, relate appropriately to the specialist, and perform tasks independently (without prompts or reminders). Therefore, a report of test results should include both the objective data and any clinical observations.

c. The salient characteristics of a good test are: (1) Validity, *i.e.,* the test measures what it is supposed to measure; (2) reliability, *i.e.,* the consistency of results obtained over time with the same test and the same individual; (3) appropriate normative data, *i.e.,* individual test scores can be compared to test data from other individuals or groups of a similar nature, representative of that population; and (4) wide scope of measurement, *i.e.,* the test should measure a broad range of facets/aspects of the domain being assessed. In considering the validity of a test result, we should note and resolve any discrepancies between formal test results and the individual's customary behavior and daily activities.

6. *Intelligence tests.*

a. The results of standardized intelligence tests may provide data that help verify the presence of intellectual disability or organic mental disorder, as well as the extent of any

compromise in cognitive functioning. However, since the results of intelligence tests are only part of the overall assessment, the narrative report that accompanies the test results should comment on whether the IQ scores are considered valid and consistent with the developmental history and the degree of functional limitation.

b. Standardized intelligence test results are essential to the adjudication of all cases of intellectual disability that are not covered under the provisions of 12.05A. Listing 12.05A may be the basis for adjudicating cases where the results of standardized intelligence tests are unavailable, e.g., where your condition precludes formal standardized testing.

c. Due to such factors as differing means and standard deviations, identical IQ scores obtained from different tests do not always reflect a similar degree of intellectual functioning. The IQ scores in 12.05 reflect values from tests of general intelligence that have a mean of 100 and a standard deviation of 15; e.g., the Wechsler series. IQs obtained from standardized tests that deviate from a mean of 100 and a standard deviation of 15 require conversion to a percentile rank so that we can determine the actual degree of limitation reflected by the IQ scores. In cases where more than one IQ is customarily derived from the test administered, e.g., where verbal, performance, and full scale IQs are provided in the Wechsler series, we use the lowest of these in conjunction with 12.05.

d. Generally, it is preferable to use IQ measures that are wide in scope and include items that test both verbal and performance abilities. However, in special circumstances, such as the assessment of individuals with sensory, motor, or communication abnormalities, or those whose culture and background are not principally English-speaking, measures such as the Test of Nonverbal Intelligence, Third Edition (TONI–3), Leiter International Performance Scale-Revised (Leiter-R), or Peabody Picture Vocabulary Test—Third Edition (PPVT-III) may be used.

e. We may consider exceptions to formal standardized psychological testing when an individual qualified by training and experience to perform such an evaluation is not available, or in cases where appropriate standardized measures for your social, linguistic, and cultural background are not available. In these cases, the best indicator of severity is often the level of adaptive functioning and how you perform activities of daily living and social functioning.

7. *Personality measures and projective testing techniques.* Results from standardized personality measures, such as the Minnesota Multiphasic Personality Inventory-Revised (MMPI-II), or from projective types of techniques, such as the Rorschach and the Thematic Apperception Test (TAT), may provide useful data for evaluating several types of mental disorders. Such test results may be useful for disability evaluation when corroborated by other evidence, including results from other psychological tests and information obtained in the course of the clinical evaluation, from treating and other medical sources, other professional health care providers, and nonmedical sources. Any inconsistency between test results and clinical history and observation should be explained in the narrative description.

8. *Neuropsychological assessments.* Comprehensive neuropsychological examinations may be used to establish the existence and extent of compromise of brain function, particularly in cases involving organic mental disorders. Normally, these examinations include assessment of cerebral dominance, basic sensation and perception, motor speed and coordination, attention and concentration, visual-motor function, memory across verbal and visual modalities, receptive and expressive speech, higher-order linguistic operations, problem-solving, abstraction ability, and general intelligence. In addition, there should be a clinical interview geared toward evaluating pathological features known to occur frequently in neurological disease and trauma, e.g., emotional lability, abnormality of mood, impaired impulse control, passivity and apathy, or inappropriate social behavior. The specialist performing the examination may administer one of the commercially available comprehensive neuropsychological batteries, such as the Luria-Nebraska or the Halstead-Reitan, or a battery of tests selected as relevant to the suspected brain dysfunction. The specialist performing the examination must be properly trained in this area of neuroscience.

9. *Screening tests.* In conjunction with clinical examinations, sources may report the results of screening tests; *i.e.,* tests used for gross determination of level of functioning. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

10. *Traumatic brain injury (TBI).* In cases involving TBI, follow the documentation and evaluation guidelines in 11.00F.

11. *Anxiety disorders.* In cases involving agoraphobia and other phobic disorders, panic disorders, and posttraumatic stress disorders, documentation of the anxiety reaction is essential. At least one detailed description of your typical reaction is required. The description should include the nature, frequency, and duration of any panic attacks or other reactions, the precipitating and exacerbating factors, and the functional effects. If the description is provided by a medical source, the reporting physician or psychologist should indicate the extent to which

the description reflects his or her own observations and the source of any ancillary information. Statements of other persons who have observed you may be used for this description if professional observation is not available.

12. *Eating disorders.* In cases involving anorexia nervosa and other eating disorders, the primary manifestations may be mental or physical, depending upon the nature and extent of the disorder. When the primary functional limitation is physical, e.g., when severe weight loss and associated clinical findings are the chief cause of inability to work, we may evaluate the impairment under the appropriate physical body system listing. Of course, we must also consider any mental aspects of the impairment, unless we can make a fully favorable determination or decision based on the physical impairment(s) alone.

E. *Chronic mental impairments.* Particular problems are often involved in evaluating mental impairments in individuals who have long histories of repeated hospitalizations or prolonged outpatient care with supportive therapy and medication. For instance, if you have chronic organic, psychotic, and affective disorders, you may commonly have your life structured in such a way as to minimize your stress and reduce your symptoms and signs. In such a case, you may be much more impaired for work than your symptoms and signs would indicate. The results of a single examination may not adequately describe your sustained ability to function. It is, therefore, vital that we review all pertinent information relative to your condition, especially at times of increased stress. We will attempt to obtain adequate descriptive information from all sources that have treated you in the time period relevant to the determination or decision.

F. *Effects of structured settings.* Particularly in cases involving chronic mental disorders, overt symptomatology may be controlled or attenuated by psychosocial factors such as placement in a hospital, halfway house, board and care facility, or other environment that provides similar structure. Highly structured and supportive settings may also be found in your home. Such settings may greatly reduce the mental demands placed on you. With lowered mental demands, overt symptoms and signs of the underlying mental disorder may be minimized. At the same time, however, your ability to function outside of such a structured or supportive setting may not have changed. If your symptomatology is controlled or attenuated by psychosocial factors, we must consider your ability to function outside of such highly structured settings. For these reasons, identical paragraph C criteria are included in 12.02, 12.03, and 12.04. The paragraph C criterion of 12.06 reflects the uniqueness of agoraphobia, an anxiety disorder manifested by an overwhelming fear of leaving the home.

G. *Effects of medication.* We must give attention to the effects of medication on your symptoms, signs, and ability to function. While drugs used to modify psychological functions and mental states may control certain primary manifestations of a mental disorder, e.g., hallucinations, impaired attention, restlessness, or hyperactivity, such treatment may not affect all functional limitations imposed by the mental disorder. In cases where overt symptomatology is attenuated by the use of such drugs, particular attention must be focused on the functional limitations that may persist. We will consider these functional limitations in assessing the severity of your impairment. See the paragraph C criteria in 12.02, 12.03, 12.04, and 12.06.

Drugs used in the treatment of some mental illnesses may cause drowsiness, blunted effect, or other side effects involving other body systems. We will consider such side effects when we evaluate the overall severity of your impairment. Where adverse effects of medications contribute to the impairment severity and the impairment(s) neither meets nor is equivalent in severity to any listing but is nonetheless severe, we will consider such adverse effects in the RFC assessment.

H. *Effects of treatment.* With adequate treatment some individuals with chronic mental disorders not only have their symptoms and signs ameliorated, but they also return to a level of function close to the level of function they had before they developed symptoms or signs of their mental disorders. Treatment may or may not assist in the achievement of a level of adaptation adequate to perform sustained SGA. See the paragraph C criteria in 12.02, 12.03, 12.04, and 12.06.

I. *Technique for reviewing evidence in mental disorders claims to determine the level of impairment severity.* We have developed a special technique to ensure that we obtain, consider, and properly evaluate all the evidence we need to evaluate impairment severity in claims involving mental impairment(s). We explain this technique in §§ 404.1520a and 416.920a.

12.01 Category of Impairments—Mental

12.02 *Organic Mental Disorders:* Psychological or behaviorial abnormalities associated with a dysfunction of the brain. History and physical examination or laboratory tests demonstrate the presence of a specific organic factor judged to be etiologically related to the abnormal mental state and loss of previously acquired functional abilities.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in C are satisfied.

A. Demonstration of a loss of specific cognitive abilities or affective changes and the

medically documented persistence of at least one of the following:

1. Disorientation to time and place; or

2. Memory impairment, either short-term (inability to learn new information), intermediate, or long-term (inability to remember information that was known sometime in the past); or

3. Perceptual or thinking disturbances (e.g., hallucinations, delusions); or

4. Change in personality; or

5. Disturbance in mood; or

6. Emotional lability (e.g., explosive temper outbursts, sudden crying, etc.) and impairment in impulse control; or

7. Loss of measured intellectual ability of at least 15 I.Q. points from premorbid levels or overall impairment index clearly within the severely impaired range on neuropsychological testing, e.g., the Luria-Nebraska, Halstead-Reitan, etc.;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic organic mental disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:

1. Repeated episodes of decompensation, each of extended duration; or

2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or

3. Current history of 1 or more years' inability to function outside a highly supportive living arrangement, with an indication of continued need for such an arrangement.

12.03 *Schizophrenic, Paranoid and Other Psychotic Disorders:* Characterized by the onset of psychotic features with deterioration from a previous level of functioning.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in C are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one or more of the following:

1. Delusions or hallucinations; or

2. Catatonic or other grossly disorganized behavior; or

3. Incoherence, loosening of associations, illogical thinking, or poverty of content of speech if associated with one of the following:

a. Blunt affect; or

b. Flat affect; or

c. Inappropriate affect;

or

4. Emotional withdrawal and/or isolation;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic schizophrenic, paranoid, or other psychotic disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:

1. Repeated episodes of decompensation, each of extended duration; or

2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or

3. Current history of 1 or more years' inability to function outside a highly supportive living arrangement, with an indication of continued need for such an arrangement.

12.04 *Affective Disorders:* Characterized by a disturbance of mood, accompanied by a full or partial manic or depressive syndrome. Mood refers to a prolonged emotion that colors the whole psychic life; it generally involves either depression or elation.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in C are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one of the following:

1. Depressive syndrome characterized by at least four of the following:

a. Anhedonia or pervasive loss of interest in almost all activites; or

b. Appetite disturbance with change in weight; or

c. Sleep disturbance; or

d. Psychomotor agitation or retardation; or

e. Decreased energy; or
f. Feelings of guilt or worthlessness; or
g. Difficulty concentrating or thinking; or
h. Thoughts of suicide; or
i. Hallucinations, delusions, or paranoid thinking; or

2. Manic syndrome characterized by at least three of the following:
a. Hyperactivity; or
b. Pressure of speech; or
c. Flight of ideas; or
d. Inflated self-esteem; or
e. Decreased need for sleep; or
f. Easy distractability; or
g. Involvement in activities that have a high probability of painful consequences which are not recognized; or
h. Hallucinations, delusions or paranoid thinking;

or

3. Bipolar syndrome with a history of episodic periods manifested by the full symptomatic picture of both manic and depressive syndromes (and currently characterized by either or both syndromes);

AND

B. Resulting in at least two of the following:
1. Marked restriction of activities of daily living; or
2. Marked difficulties in maintaining social functioning; or
3. Marked difficulties in maintaining concentration, persistence, or pace; or
4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic affective disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:
1. Repeated episodes of decompensation, each of extended duration; or
2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or
3. Current history of 1 or more years' inability to function outside a highly supportive living arrangement, with an indication of continued need for such an arrangement.

12.05 *Intellectual disability:* Intellectual disability refers to significantly subaverage general intellectual functioning with deficits in adaptive functioning initially manifested during the developmental period; *i.e.*, the evidence demonstrates or supports onset of the impairment before age 22.

The required level of severity for this disorder is met when the requirements in A, B, C, or D are satisfied.

A. Mental incapacity evidenced by dependence upon others for personal needs (e.g., toileting, eating, dressing, or bathing) and inability to follow directions, such that the use of standardized measures of intellectual functioning is precluded;

OR

B. A valid verbal, performance, or full scale IQ of 59 or less;

OR

C. A valid verbal, performance, or full scale IQ of 60 through 70 and a physical or other mental impairment imposing an additional and significant work-related limitation of function;

OR

D. A valid verbal, performance, or full scale IQ of 60 through 70, resulting in at least two of the following:
1. Marked restriction of activities of daily living; or
2. Marked difficulties in maintaining social functioning; or
3. Marked difficulties in maintaining concentration, persistence, or pace; or
4. Repeated episodes of decompensation, each of extended duration.

12.06 *Anxiety Related Disorders:* In these disorders anxiety is either the predominant disturbance or it is experienced if the individual attempts to master symptoms; for example, confronting the dreaded object or situation in a phobic disorder or resisting the obsessions or compulsions in obsessive compulsive disorders.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in both A and C are satisfied.

A. Medically documented findings of at least one of the following:
1. Generalized persistent anxiety accompanied by three out of four of the following signs or symptoms:
a. Motor tension; or
b. Autonomic hyperactivity; or
c. Apprehensive expectation; or
d. Vigilance and scanning;

or

2. A persistent irrational fear of a specific object, activity, or situation which results in a compelling desire to avoid the dreaded object, activity, or situation; or
3. Recurrent severe panic attacks manifested by a sudden unpredictable onset of intense apprehension, fear, terror and sense of impending doom occurring on the average of at least once a week; or
4. Recurrent obsessions or compulsions which are a source of marked distress; or

5. Recurrent and intrusive recollections of a traumatic experience, which are a source of marked distress;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

OR

C. Resulting in complete inability to function independently outside the area of one's home.

12.07 *Somatoform Disorders:* Physical symptoms for which there are no demonstrable organic findings or known physiological mechanisms.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented by evidence of one of the following:

1. A history of multiple physical symptoms of several years duration, beginning before age 30, that have caused the individual to take medicine frequently, see a physician often and alter life patterns significantly; or

2. Persistent nonorganic disturbance of one of the following:

a. Vision; or

b. Speech; or

c. Hearing; or

d. Use of a limb; or

e. Movement and its control (e.g., coordination disturbance, psychogenic seizures, akinesia, dyskinesia; or

f. Sensation (e.g., diminished or heightened).

3. Unrealistic interpretation of physical signs or sensations associated with the preoccupation or belief that one has a serious disease or injury;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

12.08 *Personality Disorders:* A personality disorder exists when personality traits are inflexible and maladaptive and cause either significant impairment in social or occupational functioning or subjective distress. Characteristic features are typical of the individual's long-term functioning and are not limited to discrete episodes of illness.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Deeply ingrained, maladaptive patterns of behavior associated with one of the following:

1. Seclusiveness or autistic thinking; or

2. Pathologically inappropriate suspiciousness or hostility; or

3. Oddities of thought, perception, speech and behavior; or

4. Persistent disturbances of mood or affect; or

5. Pathological dependence, passivity, or aggressivity; or

6. Intense and unstable interpersonal relationships and impulsive and damaging behavior;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

12.09 *Substance Addiction Disorders:* Behavioral changes or physical changes associated with the regular use of substances that affect the central nervous system.

The required level of severity for these disorders is met when the requirements in any of the following (A through I) are satisfied.

A. Organic mental disorders. Evaluate under 12.02.

B. Depressive syndrome. Evaluate under 12.04.

C. Anxiety disorders. Evaluate under 12.06.

D. Personality disorders. Evaluate under 12.08.

E. Peripheral neuropathies. Evaluate under 11.14.

F. Liver damage. Evaluate under 5.05.

G. Gastritis. Evaluate under 5.00.

H. Pancreatitis. Evaluate under 5.08.

I. Seizures. Evaluate under 11.02 or 11.03.

12.10 *Autistic disorder and other pervasive developmental disorders:* Characterized by qualitative deficits in the development of reciprocal social interaction, in the development of verbal and nonverbal communication skills, and in imaginative activity. Often, there is a markedly restricted repertoire of activities and interests, which frequently are stereotyped and repetitive.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of the following:

1. For autistic disorder, all of the following:

a. Qualitative deficits in reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity; and

c. Markedly restricted repertoire of activities and interests;

OR

2. For other pervasive developmental disorders, both of the following:

a. Qualitative deficits in reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

13.00 Cancer (Malignant Neoplastic Diseases)

A. *What impairments do these listings cover?* We use these listings to evaluate all cancers (malignant neoplastic diseases), except certain cancers associated with human immunodeficiency virus (HIV) infection. If you have HIV infection, we use the criteria in 14.08E to evaluate carcinoma of the cervix, Kaposi sarcoma, lymphoma, and squamous cell carcinoma of the anal canal and anal margin.

B. *What do we consider when we evaluate cancer under these listings?* We will consider factors including:

1. Origin of the cancer.

2. Extent of involvement.

3. Duration, frequency, and response to anticancer therapy.

4. Effects of any post-therapeutic residuals.

C. *How do we apply these listings?* We apply the criteria in a specific listing to a cancer originating from that specific site.

D. *What evidence do we need?*

1. We need medical evidence that specifies the type, extent, and site of the primary, recurrent, or metastatic lesion. When the primary site cannot be identified, we will use evidence documenting the site(s) of metastasis to evaluate the impairment under 13.27.

2. For operative procedures, including a biopsy or a needle aspiration, we generally need a copy of both the:

a. Operative note, and

b. Pathology report.

3. When we cannot get these documents, we will accept the summary of hospitalization(s) or other medical reports. This evidence should include details of the findings at surgery and, whenever appropriate, the pathological findings.

4. In some situations, we may also need evidence about recurrence, persistence, or progression of the cancer, the response to therapy, and any significant residuals. (See 13.00G.)

E. *When do we need longitudinal evidence?*

1. *Cancer with distant metastases.* We generally do not need longitudinal evidence for cancer that has metastasized beyond the regional lymph nodes because this cancer usually meets the requirements of a listing. Exceptions are for cancer with distant metastases that we expect to respond to anticancer therapy. For these exceptions, we usually need a longitudinal record of 3 months after therapy starts to determine whether the therapy achieved its intended effect, and whether this effect is likely to persist.

2. *Other cancers.* When there are no distant metastases, many of the listings require that we consider your response to initial anticancer therapy; that is, the initial planned treatment regimen. This therapy may consist of a single modality or a combination of modalities; that is, multimodal therapy. (See 13.00I4.)

3. *Types of treatment.*

a. Whenever the initial planned therapy is a single modality, enough time must pass to allow a determination about whether the therapy will achieve its intended effect. If the treatment fails, the failure often happens within 6 months after treatment starts, and there will often be a change in the treatment regimen.

b. Whenever the initial planned therapy is multimodal, we usually cannot make a determination about the effectiveness of the therapy until we can determine the effects of all the planned modalities. In some cases, we may need to defer adjudication until we can assess the effectiveness of therapy. However, we do not need to defer adjudication to determine whether the therapy will achieve its intended effect if we can make a fully favorable determination or decision based on the length and effects of therapy, or the residuals of the cancer or therapy (see 13.00G).

c. We need evidence under 13.02E, 13.11D, and 13.14C to establish that your treating source initiated multimodal anticancer therapy. We do not need to make a determination about the length or effectiveness of your therapy. Multimodal therapy has been initiated, and satisfies the requirements in 13.02E, 13.11D, and 13.14C, when your treating source starts the first modality. We may defer adjudication if your treating source plans multimodal therapy and has not yet initiated it.

F. *How do we evaluate impairments that do not meet one of the cancer listings?*

1. These listings are only examples of cancer that we consider severe enough to prevent you from doing any gainful activity. If your severe impairment(s) does not meet the

criteria of any of these listings, we must also consider whether you have an impairment(s) that meets the criteria of a listing in another body system.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§ 404.1526 and 416.926 of this chapter.) If your impairment(s) does not meet or medically equal a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. In that situation, we proceed to the fourth, and, if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920 of this chapter. We use the rules in §§ 404.1594 and 416.994 of this chapter, as appropriate, when we decide whether you continue to be disabled.

G. *How do we consider the effects of anticancer therapy?*

1. *How we consider the effects of anticancer therapy under the listings.* In many cases, cancers meet listing criteria only if the therapy is not effective and the cancer persists, progresses, or recurs. However, as explained in the following paragraphs, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in the case record.

2. *Effects can vary widely.*

a. We consider each case on an individual basis because the therapy and its toxicity may vary widely. We will request a specific description of the therapy, including these items:

i. Drugs given.

ii. Dosage.

iii. Frequency of drug administration.

iv. Plans for continued drug administration.

v. Extent of surgery.

vi. Schedule and fields of radiation therapy.

b. We will also request a description of the complications or adverse effects of therapy, such as the following:

i. Continuing gastrointestinal symptoms.

ii. Persistent weakness.

iii. Neurological complications.

iv. Cardiovascular complications.

v. Reactive mental disorders.

3. *Effects of therapy may change.* The severity of the adverse effects of anticancer therapy may change during treatment; therefore, enough time must pass to allow us to evaluate the therapy's effect. The residual effects of treatment are temporary in most instances; however, on occasion, the effects may be disabling for a consecutive period of at least 12 months. In some situations, very serious adverse effects may interrupt and prolong multimodal anticancer therapy for a continuous period of almost 12 months. In these situations, we may determine there is an expectation that your impairment will preclude you from engaging in any gainful activity for at least 12 months.

4. *When the initial anticancer therapy is effective.* We evaluate any post-therapeutic residual impairment(s) not included in these listings under the criteria for the affected body system. We must consider any complications of therapy. When the residual impairment(s) does not meet or medically equal a listing, we must consider its effect on your ability to do substantial gainful activity.

H. *How long do we consider your impairment to be disabling?*

1. In some listings, we specify that we will consider your impairment to be disabling until a particular point in time (for example, until at least 12 months from the date of transplantation). We may consider your impairment to be disabling beyond this point when the medical and other evidence justifies it.

2. When a listing does not contain such a specification, we will consider an impairment(s) that meets or medically equals a listing in this body system to be disabling until at least 3 years after onset of complete remission. When the impairment(s) has been in complete remission for at least 3 years, that is, the original tumor or a recurrence (or relapse) and any metastases have not been evident for at least 3 years, the impairment(s) will no longer meet or medically equal the criteria of a listing in this body system.

3. Following the appropriate period, we will consider any residuals, including residuals of the cancer or therapy (see 13.00G), in determining whether you are disabled. If you have a recurrence or relapse of your cancer, your impairment may meet or medically equal one of the listings in this body system again.

I. *What do we mean by the following terms?*

1. *Anticancer therapy* means surgery, radiation, chemotherapy, hormones, immunotherapy, or bone marrow or stem cell transplantation. When we refer to surgery as an anticancer treatment, we mean surgical excision for treatment, not for diagnostic purposes.

2. *Inoperable* means surgery is thought to be of no therapeutic value or the surgery cannot be performed; for example, when you cannot tolerate anesthesia or surgery because of another impairment(s), or you have a cancer that is too large or that has invaded crucial structures. This term does not include situations in which your cancer could have been surgically removed but another method of treatment was chosen; for example, an attempt at organ preservation. Your physician may determine whether the cancer is inoperable before or after you receive neoadjuvant therapy. *Neoadjuvant therapy* is anticancer therapy, such as chemotherapy or

radiation, given before surgery in order to reduce the size of the cancer.

3. *Metastases* means the spread of cancer cells by blood, lymph, or other body fluid. This term does not include the spread of cancer cells by direct extension of the cancer to other tissues or organs.

4. *Multimodal therapy* means anticancer therapy that is a combination of at least two types of treatment given in close proximity as a unified whole and usually planned before any treatment has begun. There are three types of treatment modalities: surgery, radiation, and systemic drug therapy (chemotherapy, hormone therapy, and immunotherapy or biological modifier therapy). Examples of multimodal therapy include:

a. Surgery followed by chemotherapy or radiation.

b. Chemotherapy followed by surgery.

c. Chemotherapy and concurrent radiation.

5. *Persistent* means the planned initial anticancer therapy failed to achieve a complete remission of your cancer; that is, your cancer is evident, even if smaller, after the therapy has ended.

6. *Progressive* means the cancer becomes more extensive after treatment; that is, there is evidence that your cancer is growing after you have completed at least half of your planned initial anticancer therapy.

7. *Recurrent or relapse* means the cancer that was in complete remission or entirely removed by surgery has returned.

8. *Unresectable* means surgery or surgeries did not completely remove the cancer. This term includes situations in which your cancer is incompletely resected or the surgical margins are positive. It does not include situations in which there is a finding of a positive margin(s) if additional surgery obtains a margin(s) that is clear. It also does not include situations in which the cancer is completely resected but you are receiving adjuvant therapy. *Adjuvant therapy* is anticancer therapy, such as chemotherapy or radiation, given after surgery in order to eliminate any remaining cancer cells or lessen the chance of recurrence.

J. *Can we establish the existence of a disabling impairment prior to the date of the evidence that shows the cancer satisfies the criteria of a listing?* Yes. We will consider factors such as:

1. The type of cancer and its location.

2. The extent of involvement when the cancer was first demonstrated.

3. Your symptoms.

K. *How do we evaluate specific cancers?*

1. *Lymphoma.*

a. Many indolent (non-aggressive) lymphomas are controlled by well-tolerated treatment modalities, although the lymphomas may produce intermittent symptoms and signs. We may defer adjudicating these cases for an appropriate period after therapy is initiated to determine whether the therapy will achieve its intended effect, which is usually to stabilize the disease process. (See 13.00E3.) Once your disease stabilizes, we will assess severity based on the extent of involvement of other organ systems and residuals from therapy.

b. A change in therapy for indolent lymphomas is usually an indicator that the therapy is not achieving its intended effect. However, your impairment will not meet the requirements of 13.05A2 if your therapy is changed solely because you or your physician chooses to change it and not because of a failure to achieve stability.

c. We consider Hodgkin lymphoma that recurs more than 12 months after completing initial anticancer therapy to be a new disease rather than a recurrence.

2. *Leukemia.*

a. *Acute leukemia.* The initial diagnosis of acute leukemia, including the accelerated or blast phase of chronic myelogenous (granulocytic) leukemia, is based on definitive bone marrow examination. Additional diagnostic information is based on chromosomal analysis, cytochemical and surface marker studies on the abnormal cells, or other methods consistent with the prevailing state of medical knowledge and clinical practice. Recurrent disease must be documented by peripheral blood, bone marrow, or cerebrospinal fluid examination, or by testicular biopsy. The initial and follow-up pathology reports should be included.

b. *Chronic myelogenous leukemia (CML).* We need a diagnosis of CML based on documented granulocytosis, including immature forms such as differentiated or undifferentiated myelocytes and myeloblasts, and a chromosomal analysis that demonstrates the Philadelphia chromosome. In the absence of a chromosomal analysis, or if the Philadelphia chromosome is not present, the diagnosis may be made by other methods consistent with the prevailing state of medical knowledge and clinical practice. The requirement for CML in the accelerated or blast phase is met in 13.06B if laboratory findings show the proportion of blast (immature) cells in the peripheral blood or bone marrow is 10 percent or greater.

c. *Chronic lymphocytic leukemia.*

i. We require the diagnosis of chronic lymphocytic leukemia (CLL) to be documented by evidence of a chronic lymphocytosis of at least 10,000 cells/mm^3 for 3 months or longer, or other acceptable diagnostic techniques consistent with the prevailing state of medical knowledge and clinical practice.

ii. We evaluate the complications and residual impairment(s) from CLL under the appropriate listings, such as 13.05A2 or the hematological listings (7.00).

d. *Elevated white cell count.* In cases of chronic leukemia (either myelogenous or

lymphocytic), an elevated white cell count, in itself, is not a factor in determining the severity of the impairment.

3. *Macroglobulinemia or heavy chain disease.* We require the diagnosis of these diseases to be confirmed by protein electrophoresis or immunoelectrophoresis. We evaluate the resulting impairment(s) under the appropriate listings, such as 13.05A2 or the hematological listings (7.00).

4. *Primary breast cancer.*

a. We evaluate bilateral primary breast cancer (synchronous or metachronous) under 13.10A, which covers local primary disease, and not as a primary disease that has metastasized.

b. We evaluate secondary lymphedema that results from anticancer therapy for breast cancer under 13.10E if the lymphedema is treated by surgery to salvage or restore the functioning of an upper extremity. Secondary lymphedema is edema that results from obstruction or destruction of normal lymphatic channels. We may not restrict our determination of the onset of disability to the date of the surgery; we may establish an earlier onset date of disability if the evidence in your case record supports such a finding.

5. *Carcinoma-in-situ.* Carcinoma-in-situ, or preinvasive carcinoma, usually responds to treatment. When we use the term ''carcinoma'' in these listings, it does not include carcinoma-in-situ.

6. *Primary central nervous system (CNS) cancers.* We use the criteria in 13.13 to evaluate cancers that originate within the CNS (that is, brain and spinal cord cancers).

a. The CNS cancers listed in 13.13A1 are highly malignant and respond poorly to treatment, and therefore we do not require additional criteria to evaluate them. We do not list pituitary gland cancer (for example, pituitary gland carcinoma) in 13.13A1, although this CNS cancer is highly malignant and responds poorly to treatment. We evaluate pituitary gland cancer under 13.13A1 and do not require additional criteria to evaluate it.

b. We consider a CNS tumor to be malignant if it is classified as Grade II, Grade III, or Grade IV under the World Health Organization (WHO) classification of tumors of the CNS (*WHO Classification of Tumours of the Central Nervous System,* 2007).

c. We evaluate benign (for example, WHO Grade I) CNS tumors under 11.05. We evaluate metastasized CNS cancers from non-CNS sites under the primary cancers (see 13.00C). We evaluate any complications of CNS cancers, such as resultant neurological or psychological impairments, under the criteria for the affected body system.

7. *Primary peritoneal carcinoma.* We use the criteria in 13.23E to evaluate primary peritoneal carcinoma in women because this cancer is often indistinguishable from ovarian cancer and is generally treated the same way as ovarian cancer. We use the criteria in 13.15A to evaluate primary peritoneal carcinoma in men because many of these cases are similar to malignant mesothelioma.

8. *Prostate cancer.* We exclude ''biochemical recurrence'' in 13.24A, which is defined as an increase in the serum prostate-specific antigen (PSA) level following the completion of the hormonal intervention therapy. We need corroborating evidence to document recurrence, such as radiological studies or findings on physical examination.

9. *Melanoma.* We evaluate malignant melanoma that affects the skin (cutaneous melanoma), eye (ocular melanoma), or mucosal membranes (mucosal melanoma) under 13.29. We evaluate melanoma that is not malignant that affects the skin (benign melanocytic tumor) under the listings in 8.00 or other affected body systems.

L. *How do we evaluate cancer treated by bone marrow or stem cell transplantation, including transplantation using stem cells from umbilical cord blood?* Bone marrow or stem cell transplantation is performed for a variety of cancers. We require the transplantation to occur before we evaluate it under these listings. We do not need to restrict our determination of the onset of disability to the date of the transplantation (13.05, 13.06, or 13.07) or the date of first treatment under the treatment plan that includes transplantation (13.28). We may be able to establish an earlier onset date of disability due to your transplantation if the evidence in your case record supports such a finding.

1. *Acute leukemia (including T-cell lymphoblastic lymphoma) or accelerated or blast phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of transplantation, whichever is later.

2. *Lymphoma, multiple myeloma, or chronic phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 12 months from the date of transplantation.

3. *Other cancers.* We will evaluate any other cancer treated with bone marrow or stem cell transplantation under 13.28, regardless of whether there is another listing that addresses that impairment. The length of time we will consider you to be disabled depends on whether you undergo allogeneic or autologous transplantation.

a. *Allogeneic bone marrow or stem cell transplantation.* If you undergo allogeneic transplantation (transplantation from an unrelated donor or a related donor other than an identical twin), we will consider you to be disabled until at least 12 months from the date of transplantation.

b. *Autologous bone marrow or stem cell transplantation.* If you undergo autologous transplantation (transplantation of your own cells or cells from your identical twin (syngeneic transplantation)), we will consider you to be disabled until at least 12 months from the date of the first treatment under the treatment plan that includes transplantation. The first treatment usually refers to the initial therapy given to prepare you for transplantation.

4. *Evaluating disability after the appropriate time period has elapsed.* We consider any residual impairment(s), such as complications arising from:

a. Graft-versus-host (GVH) disease.

b. Immunosuppressant therapy, such as frequent infections.

c. Significant deterioration of other organ systems.

13.01 Category of Impairments, Malignant Neoplastic Diseases

13.02 *Soft tissue cancers of the head and neck (except salivary glands—13.08—and thyroid gland—13.09).*

A. Inoperable or unresectable.

OR

B. Persistent or recurrent disease following initial anticancer therapy, except persistence or recurrence in the true vocal cord.

OR

OR

C. With metastases beyond the regional lymph nodes.

OR

D. Small-cell (oat cell) carcinoma.

OR

E. Soft tissue cancers originating in the head and neck treated with multimodal anticancer therapy (see 13.00E3c). Consider under a disability until at least 18 months from the date of diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.03 *Skin.*

A. Sarcoma or carcinoma with metastases to or beyond the regional lymph nodes.

OR

B. Carcinoma invading deep extradermal structures (for example, skeletal muscle, cartilage, or bone).

1. Recurrent after wide excision (except an additional primary melanoma at a different site, which is not considered to be recurrent disease).

2. With metastases as described in a, b, or c:

a. Metastases to one or more clinically apparent nodes; that is, nodes that are detected by imaging studies (excluding lymphoscintigraphy) or by clinical examination.

b. If the nodes are not clinically apparent, with metastases to four or more nodes.

c. Metastases to adjacent skin (satellite lesions) or distant sites.

13.04 *Soft tissue sarcoma.*

A. With regional or distant metastases.

OR

B. Persistent or recurrent following initial anticancer therapy.

13.05 *Lymphoma (including mycosis fungoides, but excluding T-cell lymphoblastic lymphoma—13.06).* (*See* 13.00K1 and 13.00K2c.)

A. Non-Hodgkin's lymphoma, as described in 1 or 2:

1. Aggressive lymphoma (including diffuse large B-cell lymphoma) persistent or recurrent following initial anticancer therapy.

2. Indolent lymphoma (including mycosis fungoides and follicular small cleaved cell) requiring initiation of more than one (single mode or multimodal) anticancer treatment regimen within a period of 12 consecutive months. Consider under a disability from at least the date of initiation of the treatment regimen that failed within 12 months.

OR

B. Hodgkin lymphoma with failure to achieve clinically complete remission, or recurrent lymphoma within 12 months of completing initial anticancer therapy.

OR

C. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

D. Mantle cell lymphoma.

13.06 *Leukemia.* (*See* 13.00K2.)

A. Acute leukemia (including T-cell lymphoblastic lymphoma). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Chronic myelogenous leukemia, as described in 1 or 2:

1. Accelerated or blast phase (see 13.00K2b). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

2. Chronic phase, as described in a or b:

a. Consider under a disability until at least 12 months from the date of bone marrow or

stem cell transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

b. Progressive disease following initial anticancer therapy.

13.07 *Multiple myeloma (confirmed by appropriate serum or urine protein electrophoresis and bone marrow findings).*

A. Failure to respond or progressive disease following initial anticancer therapy.

OR

B. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.08 *Salivary glands*—carcinoma or sarcoma with metastases beyond the regional lymph nodes.

13.09 *Thyroid gland.*

A. Anaplastic (undifferentiated) carcinoma.

OR

B. Carcinoma with metastases beyond the regional lymph nodes progressive despite radioactive iodine therapy.

OR

C. Medullary carcinoma with metastases beyond the regional lymph nodes.

13.10 *Breast (except sarcoma—13.04).* (*See* 13.00K4.)

A. Locally advanced cancer (inflammatory carcinoma, cancer of any size with direct extension to the chest wall or skin, or cancer of any size with metastases to the ipsilateral internal mammary nodes).

OR

B. Carcinoma with metastases to the supraclavicular or infraclavicular nodes, to 10 or more axillary nodes, or with distant metastases.

OR

C. Recurrent carcinoma, except local recurrence that remits with anticancer therapy.

OR

D. Small-cell (oat cell) carcinoma.

OR

E. With secondary lymphedema that is caused by anticancer therapy and treated by surgery to salvage or restore the functioning of an upper extremity. (See 13.00K4b.) Consider under a disability until at least 12 months from the date of the surgery that treated the secondary lymphedema. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.11 *Skeletal system*—sarcoma.

A. Inoperable or unresectable.

OR

B. Recurrent cancer (except local recurrence) after initial anticancer therapy.

OR

C. With distant metastases.

OR

D. All other cancers originating in bone with multimodal anticancer therapy (see 13.00E3c). Consider under a disability for 12 months from the date of diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.12 *Maxilla, orbit, or temporal fossa.*

A. Sarcoma or carcinoma of any type with regional or distant metastases.

OR

B. Carcinoma of the antrum with extension into the orbit or ethmoid or sphenoid sinus.

OR

C. Cancer with extension to the orbit, meninges, sinuses, or base of the skull.

13.13 *Nervous system.* (See 13.00K6.)

A. Primary central nervous system (CNS; that is, brain and spinal cord) cancers, as described in 1, 2, or 3:

1. Glioblastoma multiforme, ependymoblastoma, and diffuse intrinsic brain stem gliomas (see 13.00K6a).

2. Any Grade III or Grade IV CNS cancer (see 13.00K6b), including astrocytomas, sarcomas, and medulloblastoma and other primitive neuroectodermal tumors (PNETs).

3. Any primary CNS cancer, as described in a or b:

a. Metastatic.

b. Progressive or recurrent following initial anticancer therapy.

OR

B. Primary peripheral nerve or spinal root cancers, as described in 1 or 2:

1. Metastatic.

2. Progressive or recurrent following initial anticancer therapy.

13.14 *Lungs.*

A. Non-small-cell carcinoma—inoperable, unresectable, recurrent, or metastatic disease to or beyond the hilar nodes.

OR

B. Small-cell (oat cell) carcinoma.

OR

C. Carcinoma of the superior sulcus (including Pancoast tumors) with multimodal anticancer therapy (see 13.00E3c). Consider under a disability until at least 18 months from the date of diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.15 *Pleura or mediastinum.*

A. Malignant mesothelioma of pleura.

OR

B. Tumors of the mediastinum, as described in 1 or 2:

1. With metastases to or beyond the regional lymph nodes.

2. Persistent or recurrent following initial anticancer therapy.

OR

C. Small-cell (oat cell) carcinoma.

13.16 *Esophagus or stomach.*

A. Carcinoma or sarcoma of the esophagus.

OR

B. Carcinoma or sarcoma of the stomach, as described in 1 or 2:

1. Inoperable, unresectable, extending to surrounding structures, or recurrent.

2. With metastases to or beyond the regional lymph nodes.

OR

C. Small-cell (oat cell) carcinoma.

13.17 *Small intestine*—carcinoma, sarcoma, or carcinoid.

A. Inoperable, unresectable, or recurrent.

OR

B. With metastases beyond the regional lymph nodes.

OR

C. Small-cell (oat cell) carcinoma.

13.18 *Large intestine (from ileocecal valve to and including anal canal).*

A. Adenocarcinoma that is inoperable, unresectable, or recurrent.

OR

B. Squamous cell carcinoma of the anus, recurrent after surgery.

OR

C. With metastases beyond the regional lymph nodes.

OR

D. Small-cell (oat cell) carcinoma.

13.19 *Liver or gallbladder*—cancer of the liver, gallbladder, or bile ducts.

13.20 *Pancreas.*

A. Carcinoma (except islet cell carcinoma).

OR

B. Islet cell carcinoma that is physiologically active and is either inoperable or unresectable.

13.21 *Kidneys, adrenal glands, or ureters*—carcinoma.

A. Inoperable, unresectable, or recurrent.

OR

B. With metastases to or beyond the regional lymph nodes.

13.22 *Urinary bladder*—carcinoma.

A. With infiltration beyond the bladder wall.

OR

B. Recurrent after total cystectomy.

OR

C. Inoperable or unresectable.

OR

D. With metastases to or beyond the regional lymph nodes.

OR

E. Small-cell (oat cell) carcinoma.

13.23 *Cancers of the female genital tract*—carcinoma or sarcoma (including primary peritoneal carcinoma).

A. Uterus (corpus), as described in 1, 2, or 3:

1. Invading adjoining organs.

2. With metastases to or beyond the regional lymph nodes.

3. Persistent or recurrent following initial anticancer therapy.

OR

B. Uterine cervix, as described in 1, 2, or 3:

1. Extending to the pelvic wall, lower portion of the vagina, or adjacent or distant organs.

2. Persistent or recurrent following initial anticancer therapy.

3. With metastases to distant (for example, para-aortic or supraclavicular) lymph nodes.

OR

C. Vulva or vagina, as described in 1, 2, or 3:

1. Invading adjoining organs.

2. With metastases to or beyond the regional lymph nodes.

3. Persistent or recurrent following initial anticancer therapy.

OR

D. Fallopian tubes, as described in 1 or 2:

1. Extending to the serosa or beyond.

2. Persistent or recurrent following initial anticancer therapy.

E. Ovaries, as described in 1 or 2:

1. All cancers except germ-cell cancers, with at least one of the following:

a. Extension beyond the pelvis; for example, implants on, or direct extension to, peritoneal, omental, or bowel surfaces.

b. Metastases to or beyond the regional lymph nodes.

c. Recurrent following initial anticancer therapy.

2. Germ-cell cancers—progressive or recurrent following initial anticancer therapy.

OR

F. Small-cell (oat cell) carcinoma.

13.24 *Prostate gland*—carcinoma.

A. Progressive or recurrent (not including biochemical recurrence) despite initial hormonal intervention. (See 13.00K8.)

OR

B. With visceral metastases (metastases to internal organs).

OR

C. Small cell (oat cell) carcinoma.

13.25 *Testicles*—cancer with metastatic disease progressive or recurrent following initial chemotherapy.

13.26 *Penis*—carcinoma with metastases to or beyond the regional lymph nodes.

13.27 *Primary site unknown after appropriate search for primary*—metastatic carcinoma or sarcoma, except for squamous cell carcinoma confined to the neck nodes.

13.28 *Cancer treated by bone marrow or stem cell transplantation.* (See 13.00L.)

A. Allogeneic transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Autologous transplantation. Consider under a disability until at least 12 months from the date of the first treatment under the treatment plan that includes transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.29 *Malignant melanoma* (including skin, ocular, or mucosal melanomas), as described in either A, B, or C:

A. Recurrent (except an additional primary melanoma at a different site, which is not considered to be recurrent disease) following either 1 or 2:

1. Wide excision (skin melanoma).

2. Enucleation of the eye (ocular melanoma).

OR

B. With metastases as described in 1, 2, or 3:

1. Metastases to one or more clinically apparent nodes; that is, nodes that are detected by imaging studies (excluding lymphoscintigraphy) or by clinical evaluation (palpable).

2. If the nodes are not clinically apparent, with metastases to four or more nodes.

3. Metastases to adjacent skin (satellite lesions) or distant sites (for example, liver, lung, or brain).

OR

C. Mucosal melanoma.

14.00 Immune System Disorders

A. *What disorders do we evaluate under the immune system disorders listings?*

1. *We evaluate immune system disorders that cause dysfunction in one or more components of your immune system.*

a. The dysfunction may be due to problems in antibody production, impaired cell-mediated immunity, a combined type of antibody/cellular deficiency, impaired phagocytosis, or complement deficiency.

b. Immune system disorders may result in recurrent and unusual infections, or inflammation and dysfunction of the body's own tissues. Immune system disorders can cause a deficit in a single organ or body system that results in extreme (that is, very serious) loss of function. They can also cause lesser degrees of limitations in two or more organs or body systems, and when associated with symptoms or signs, such as severe fatigue, fever, malaise, diffuse musculoskeletal pain, or involuntary weight loss, can also result in extreme limitation.

c. We organize the discussions of immune system disorders in three categories: Autoimmune disorders; Immune deficiency disorders, excluding human immunodeficiency virus (HIV) infection; and HIV infection.

2. *Autoimmune disorders (14.00D).* Autoimmune disorders are caused by dysfunctional immune responses directed against the body's own tissues, resulting in chronic, multisystem impairments that differ in clinical manifestations, course, and outcome. They are sometimes referred to as rheumatic diseases, connective tissue disorders, or collagen vascular disorders. Some of the features of autoimmune disorders in adults differ from the features of the same disorders in children.

3. *Immune deficiency disorders, excluding HIV infection (14.00E).* Immune deficiency disorders are characterized by recurrent or unusual infections that respond poorly to treatment, and are often associated with complications affecting other parts of the body. Immune deficiency disorders are classified as either *primary* (congenital) or *acquired*. Individuals with immune deficiency disorders also have an increased risk of malignancies and of having autoimmune disorders.

4. *Human immunodeficiency virus (HIV) infection (14.00F).* HIV infection may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions, as described in 14.08.

B. *What information do we need to show that you have an immune system disorder?* Generally, we need your medical history, a report(s) of a physical examination, a report(s) of laboratory findings, and in some instances, appropriate medically acceptable imaging or tissue biopsy reports to show that you have an immune system disorder. Therefore, we will make every reasonable effort to obtain your medical history, medical findings, and results of laboratory tests. We explain the information we need in more detail in the sections below.

C. Definitions

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Constitutional symptoms or signs*, as used in these listings, means severe fatigue, fever, malaise, or involuntary weight loss. *Severe fatigue* means a frequent sense of exhaustion that results in significantly reduced physical activity or mental function. *Malaise* means

frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.

3. *Disseminated* means that a condition is spread over a considerable area. The type and extent of the spread will depend on your specific disease.

4. *Dysfunction* means that one or more of the body regulatory mechanisms are impaired, causing either an excess or deficiency of immunocompetent cells or their products.

5. *Extra-articular* means ''other than the joints''; for example, an organ(s) such as the heart, lungs, kidneys, or skin.

6. *Inability to ambulate effectively* has the same meaning as in 1.00B2b.

7. *Inability to perform fine and gross movements effectively* has the same meaning as in 1.00B2c.

8. *Major peripheral joints* has the same meaning as in 1.00F.

9. *Persistent* means that a sign(s) or symptom(s) has continued over time. The precise meaning will depend on the specific immune system disorder, the usual course of the disorder, and the other circumstances of your clinical course.

10. *Recurrent* means that a condition that previously responded adequately to an appropriate course of treatment returns after a period of remission or regression. The precise meaning, such as the extent of response or remission and the time periods involved, will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

11. *Resistant to treatment* means that a condition did not respond adequately to an appropriate course of treatment. Whether a response is adequate or a course of treatment is appropriate will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

12. *Severe* means medical severity as used by the medical community. The term does not have the same meaning as it does when we use it in connection with a finding at the second step of the sequential evaluation processes in §§ 404.1520, 416.920, and 416.924.

D. How do we document and evaluate the listed autoimmune disorders?

1. *Systemic lupus erythematosus (14.02).*

a. *General.* Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that can affect any organ or body system. It is frequently, but not always, accompanied by constitutional symptoms or signs (severe fatigue, fever, malaise, involuntary weight loss). Major organ or body system involvement can include: Respiratory (pleuritis, pneumonitis), cardiovascular (endocarditis, myocarditis, pericarditis, vasculitis), renal (glomerulonephritis), hematologic (anemia, leukopenia, thrombocytopenia), skin (photosensitivity), neurologic (seizures), mental (anxiety, fluctuating cognition (''lupus fog''), mood disorders, organic brain syndrome, psychosis), or immune system disorders (inflammatory arthritis). Immunologically, there is an array of circulating serum auto-antibodies and pro- and anti-coagulant proteins that may occur in a highly variable pattern.

b. *Documentation of SLE.* Generally, but not always, the medical evidence will show that your SLE satisfies the criteria in the current ''Criteria for the Classification of Systemic Lupus Erythematosus'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

2. *Systemic vasculitis (14.03).*

a. *General.*

(i) Vasculitis is an inflammation of blood vessels. It may occur acutely in association with adverse drug reactions, certain chronic infections, and occasionally, malignancies. More often, it is chronic and the cause is unknown. Symptoms vary depending on which blood vessels are involved. Systemic vasculitis may also be associated with other autoimmune disorders; for example, SLE or dermatomyositis.

(ii) There are several clinical patterns, including but not limited to polyarteritis nodosa, Takayasu's arteritis (aortic arch arteritis), giant cell arteritis (temporal arteritis), and Wegener's granulomatosis.

b. *Documentation of systemic vasculitis.* Angiography or tissue biopsy confirms a diagnosis of systemic vasculitis when the disease is suspected clinically. When you have had angiography or tissue biopsy for systemic vasculitis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase angiography or tissue biopsy.

3. *Systemic sclerosis (scleroderma) (14.04).*

a. *General.* Systemic sclerosis (scleroderma) constitutes a spectrum of disease in which thickening of the skin is the clinical hallmark. Raynaud's phenomenon, often medically severe and progressive, is present frequently and may be the peripheral manifestation of a vasospastic abnormality in the heart, lungs, and kidneys. The CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) is a variant that may slowly progress over years to the generalized process, systemic sclerosis.

b. *Diffuse cutaneous systemic sclerosis.* In diffuse cutaneous systemic sclerosis (also known as diffuse scleroderma), major organ or systemic involvement can include the gastrointestinal tract, lungs, heart, kidneys,

and muscle in addition to skin or blood vessels. Although arthritis can occur, joint dysfunction results primarily from soft tissue/cutaneous thickening, fibrosis, and contractures.

c. *Localized scleroderma (linear scleroderma and morphea).*

(i) Localized scleroderma (linear scleroderma and morphea) is more common in children than in adults. However, this type of scleroderma can persist into adulthood. To assess the severity of the impairment, we need a description of the extent of involvement of linear scleroderma and the location of the lesions. For example, linear scleroderma involving the arm but not crossing any joints is not as functionally limiting as sclerodactyly (scleroderma localized to the fingers). Linear scleroderma of a lower extremity involving skin thickening and atrophy of underlying muscle or bone can result in contractures and leg length discrepancy. In such cases, we may evaluate your impairment under the musculoskeletal listings (1.00).

(ii) When there is isolated morphea of the face causing facial disfigurement from unilateral hypoplasia of the mandible, maxilla, zygoma, or orbit, adjudication may be more appropriate under the criteria in the affected body system, such as special senses and speech (2.00) or mental disorders (12.00).

(iii) Chronic variants of these syndromes include disseminated morphea, Shulman's disease (diffuse fasciitis with eosinophilia), and eosinophilia-myalgia syndrome (often associated with toxins such as toxic oil or contaminated tryptophan), all of which can impose medically severe musculoskeletal dysfunction and may also lead to restrictive pulmonary disease. We evaluate these variants of the disease under the criteria in the musculoskeletal listings (1.00) or respiratory system listings (3.00).

d. *Documentation of systemic sclerosis (scleroderma).* Documentation involves differentiating the clinical features of systemic sclerosis (scleroderma) from other autoimmune disorders. However, there may be an overlap.

4. *Polymyositis and dermatomyositis (14.05).*

a. *General.* Polymyositis and dermatomyositis are related disorders that are characterized by an inflammatory process in striated muscle, occurring alone or in association with other autoimmune disorders or malignancy. The most common manifestations are symmetric weakness, and less frequently, pain and tenderness of the proximal limb-girdle (shoulder or pelvic) musculature. There may also be involvement of the cervical, cricopharyngeal, esophageal, intercostal, and diaphragmatic muscles.

b. *Documentation of polymyositis and dermatomyositis.* Generally, but not always, polymyositis is associated with elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, and aldolase), and characteristic abnormalities on electromyography and muscle biopsy. In dermatomyositis there are characteristic skin findings in addition to the findings of polymyositis. When you have had electromyography or muscle biopsy for polymyositis or dermatomyositis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase electromyography or muscle biopsy.

c. *Additional information about how we evaluate polymyositis and dermatomyositis under the listings.*

(i) Weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to ambulate effectively. Weakness of your shoulder girdle muscles may result in your inability to perform lifting, carrying, and reaching overhead, and also may seriously affect your ability to perform activities requiring fine movements. We evaluate these limitations under 14.05A.

(ii) We use the malignant neoplastic diseases listings (13.00) to evaluate malignancies associated with polymyositis or dermatomyositis. We evaluate the involvement of other organs/body systems under the criteria for the listings in the affected body system.

5. *Undifferentiated and mixed connective tissue disease (14.06).*

a. *General.* This listing includes syndromes with clinical and immunologic features of several autoimmune disorders, but which do not satisfy the criteria for any of the specific disorders described. For example, you may have clinical features of SLE and systemic vasculitis, and the serologic (blood test) findings of rheumatoid arthritis.

b. *Documentation of undifferentiated and mixed connective tissue disease.* Undifferentiated connective tissue disease is diagnosed when clinical features and serologic (blood test) findings, such as rheumatoid factor or antinuclear antibody (consistent with an autoimmune disorder) are present but do not satisfy the criteria for a specific disease. Mixed connective tissue disease (MCTD) is diagnosed when clinical features and serologic findings of two or more autoimmune diseases overlap.

6. *Inflammatory arthritis (14.09).*

a. *General.* The spectrum of inflammatory arthritis includes a vast array of disorders that differ in cause, course, and outcome. Clinically, inflammation of major peripheral joints may be the dominant manifestation causing difficulties with ambulation or fine and gross movements; there may be joint pain, swelling, and tenderness. The arthritis may affect other joints, or cause less limitation in ambulation or the performance of

fine and gross movements. However, in combination with extra-articular features, including constitutional symptoms or signs (severe fatigue, fever, malaise, involuntary weight loss), inflammatory arthritis may result in an extreme limitation.

b. *Inflammatory arthritis involving the axial spine (spondyloarthropathy).* In adults, inflammatory arthritis involving the axial spine may be associated with disorders such as:

(i) Reiter's syndrome;

(ii) Ankylosing spondylitis;

(iii) Psoriatic arthritis;

(iv) Whipple's disease;

(v) Behçet's disease; and

(vi) Inflammatory bowel disease.

c. *Inflammatory arthritis involving the peripheral joints.* In adults, inflammatory arthritis involving peripheral joints may be associated with disorders such as:

(i) Rheumatoid arthritis;

(ii) Sjögren's syndrome;

(iii) Psoriatic arthritis;

(iv) Crystal deposition disorders (gout and pseudogout);

(v) Lyme disease; and

(vi) Inflammatory bowel disease.

d. *Documentation of inflammatory arthritis.* Generally, but not always, the diagnosis of inflammatory arthritis is based on the clinical features and serologic findings described in the most recent edition of the Primer on the Rheumatic Diseases published by the Arthritis Foundation.

e. *How we evaluate inflammatory arthritis under the listings.*

(i) Listing-level severity in 14.09A and 14.09C1 is shown by an impairment that results in an ''extreme'' (very serious) limitation. In 14.09A, the criterion is satisfied with persistent inflammation or deformity in one major peripheral weight-bearing joint resulting in the inability to ambulate effectively (as defined in 14.00C6) or one major peripheral joint in each upper extremity resulting in the inability to perform fine and gross movements effectively (as defined in 14.00C7). In 14.09C1, if you have the required ankylosis (fixation) of your cervical or dorsolumbar spine, we will find that you have an extreme limitation in your ability to see in front of you, above you, and to the side. Therefore, inability to ambulate effectively is implicit in 14.09C1, even though you might not require bilateral upper limb assistance.

(ii) Listing-level severity is shown in 14.09B, 14.09C2, and 14.09D by inflammatory arthritis that involves various combinations of complications of one or more major peripheral joints or other joints, such as inflammation or deformity, extra-articular features, repeated manifestations, and constitutional symptoms or signs. Extra-articular impairments may also meet listings in other body systems.

(iii) Extra-articular features of inflammatory arthritis may involve any body system; for example: Musculoskeletal (heel enthesopathy), ophthalmologic (iridocyclitis, keratoconjunctivitis sicca, uveitis), pulmonary (pleuritis, pulmonary fibrosis or nodules, restrictive lung disease), cardiovascular (aortic valve insufficiency, arrhythmias, coronary arteritis, myocarditis, pericarditis, Raynaud's phenomenon, systemic vasculitis), renal (amyloidosis of the kidney), hematologic (chronic anemia, thrombocytopenia), neurologic (peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss), mental (cognitive dysfunction, poor memory), and immune system (Felty's syndrome (hypersplenism with compromised immune competence)).

(iv) If both inflammation and chronic deformities are present, we evaluate your impairment under the criteria of any appropriate listing.

7. *Sjögren's syndrome (14.10).*

a. *General.*

(i) Sjögren's syndrome is an immune-mediated disorder of the exocrine glands. Involvement of the lacrimal and salivary glands is the hallmark feature, resulting in symptoms of dry eyes and dry mouth, and possible complications, such as corneal damage, blepharitis (eyelid inflammation), dysphagia (difficulty in swallowing), dental caries, and the inability to speak for extended periods of time. Involvement of the exocrine glands of the upper airways may result in persistent dry cough.

(ii) Many other organ systems may be involved, including musculoskeletal (arthritis, myositis), respiratory (interstitial fibrosis), gastrointestinal (dysmotility, dysphagia, involuntary weight loss), genitourinary (interstitial cystitis, renal tubular acidosis), skin (purpura, vasculitis), neurologic (central nervous system disorders, cranial and peripheral neuropathies), mental (cognitive dysfunction, poor memory), and neoplastic (lymphoma). Severe fatigue and malaise are frequently reported. Sjögren's syndrome may be associated with other autoimmune disorders (for example, rheumatoid arthritis or SLE); usually the clinical features of the associated disorder predominate.

b. *Documentation of Sjögren's syndrome.* If you have Sjögren's syndrome, the medical evidence will generally, but not always, show that your disease satisfies the criteria in the current ''Criteria for the Classification of Sjögren's Syndrome'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

E. How do we document and evaluate immune deficiency disorders, excluding HIV infection?

1. *General.*

a. Immune deficiency disorders can be classified as:

(i) *Primary* (congenital); for example, X-linked agammaglobulinemia, thymic hypoplasia (DiGeorge syndrome), severe combined immunodeficiency (SCID), chronic granulomatous disease (CGD), C1 esterase inhibitor deficiency.

(ii) *Acquired;* for example, medication-related.

b. Primary immune deficiency disorders are seen mainly in children. However, recent advances in the treatment of these disorders have allowed many affected children to survive well into adulthood. Occasionally, these disorders are first diagnosed in adolescence or adulthood.

2. *Documentation of immune deficiency disorders.* The medical evidence must include documentation of the specific type of immune deficiency. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

3. *Immune deficiency disorders treated by stem cell transplantation.*

a. *Evaluation in the first 12 months.* If you undergo stem cell transplantation for your immune deficiency disorder, we will consider you disabled until at least 12 months from the date of the transplant.

b. *Evaluation after the 12-month period has elapsed.* After the 12-month period has elapsed, we will consider any residuals of your immune deficiency disorder as well as any residual impairment(s) resulting from the treatment, such as complications arising from:

(i) Graft-versus-host (GVH) disease.

(ii) Immunosuppressant therapy, such as frequent infections.

(iii) Significant deterioration of other organ systems.

4. *Medication-induced immune suppression.* Medication effects can result in varying degrees of immune suppression, but most resolve when the medication is ceased. However, if you are prescribed medication for long-term immune suppression, such as after an organ transplant, we will evaluate:

a. The frequency and severity of infections.

b. Residuals from the organ transplant itself, after the 12-month period has elapsed.

c. Significant deterioration of other organ systems.

F. How do we document and evaluate human immunodeficiency virus (HIV) infection?

Any individual with HIV infection, including one with a diagnosis of acquired immune deficiency syndrome (AIDS), may be found disabled under 14.08 if his or her impairment meets the criteria in that listing or is medically equivalent to the criteria in that listing.

1. *Documentation of HIV infection.* The medical evidence must include documentation of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for HIV infection, we will make every reasonable effort to obtain reports of the results of that testing. However, we will not purchase laboratory testing to establish whether you have HIV infection.

a. *Definitive documentation of HIV infection.* A definitive diagnosis of HIV infection is documented by one or more of the following laboratory tests:

(i) HIV antibody tests. HIV antibodies are usually first detected by an ELISA screening test performed on serum. Because the ELISA can yield false positive results, confirmation is required using a more definitive test, such as a Western blot or an immunofluorescence assay.

(ii) Positive ''viral load'' (VL) tests. These tests are normally used to quantitate the amount of the virus present but also document HIV infection. Such tests include the quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT-PCR).

(iii) HIV DNA detection by polymerase chain reaction (PCR).

(iv) A specimen that contains HIV antigen (for example, serum specimen, lymphocyte culture, or cerebrospinal fluid).

(v) A positive viral culture for HIV from peripheral blood mononuclear cells (PBMC).

(vi) Other tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge.

b. *Other acceptable documentation of HIV infection.* We may also document HIV infection without the definitive laboratory evidence described in 14.00F1a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. If no definitive laboratory evidence is available, we may document HIV infection by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence. For example, we will accept a diagnosis of HIV infection without definitive laboratory evidence of the HIV infection if you have an opportunistic disease that is predictive of a defect in cell-mediated immunity (for example, toxoplasmosis of the brain, *Pneumocystis* pneumonia (PCP)), and there is no other known cause of diminished resistance to that disease (for example, long-term steroid treatment, lymphoma). In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

2. *CD4 tests.* Individuals who have HIV infection or other disorders of the immune system may have tests showing a reduction of either the absolute count or the percentage of their T-helper lymphocytes (CD4 cells). The extent of immune suppression correlates with the level or rate of decline of the CD4 count. Generally, when the CD4 count is below 200/mm^3 (or below 14 percent of the total lymphocyte count) the susceptibility to opportunistic infection is greatly increased. Although a reduced CD4 count alone does not establish a definitive diagnosis of HIV infection, a CD4 count below 200 does offer supportive evidence when there are clinical findings, but not a definitive diagnosis of an opportunistic infection(s). However, a reduced CD4 count alone does not document the severity or functional consequences of HIV infection.

3. *Documentation of the manifestations of HIV infection.* The medical evidence must also include documentation of the manifestations of HIV infection. Documentation may be by laboratory evidence or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

a. *Definitive documentation of the manifestations of HIV infection.* The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serologic test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). We will make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histologic or other test has been performed, the evidence should include a copy of the appropriate report. If we cannot obtain the report, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including appropriate medically acceptable imaging studies) or microscopic examination of the appropriate tissues or body fluids.

b. *Other acceptable documentation of the manifestations of HIV infection.* We may also document manifestations of HIV infection without the definitive laboratory evidence described in 14.00F3a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. For example, many conditions are now commonly diagnosed based on some or all of the following: Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing. The following are examples of how we may document manifestations of HIV infection with other appropriate evidence.

(i) Although a definitive diagnosis of PCP requires identifying the organism in bronchial washings, induced sputum, or lung biopsy, these tests are frequently bypassed if PCP can be diagnosed presumptively. Supportive evidence may include: Fever, dyspnea, hypoxia, CD4 count below 200, and no evidence of bacterial pneumonia. Also supportive are bilateral lung interstitial infiltrates on x-ray, a typical pattern on CAT scan, or a gallium scan positive for pulmonary uptake. Response to anti-PCP therapy usually requires 5–7 days, and such a response can be supportive of the diagnosis.

(ii) Documentation of *Cytomegalovirus* (CMV) disease (14.08D) may present special problems because definitive diagnosis (except for chorioretinitis, which may be diagnosed by an ophthalmologist or optometrist on funduscopic examination) requires identification of viral inclusion bodies or a positive culture from the affected organ and the absence of any other infectious agent likely to be causing the disease. A positive serology test does not establish a definitive diagnosis of CMV disease, but does offer supportive evidence of a presumptive diagnosis of CMV disease. Other clinical findings that support a presumptive diagnosis of CMV may include: Fever, urinary culture positive for CMV, and CD4 count below 200. A clear response to anti-CMV therapy also supports a diagnosis.

(iii) A definitive diagnosis of toxoplasmosis of the brain is based on brain biopsy, but this procedure carries significant risk and is not commonly performed. This condition is usually diagnosed presumptively based on symptoms or signs of fever, headache, focal neurologic deficits, seizures, typical lesions on brain imaging, and a positive serology test.

(iv) Candidiasis of the esophagus (also known as *Candida* esophagitis) may be presumptively diagnosed based on symptoms of retrosternal pain on swallowing (odynophagia) and either oropharyngeal thrush (white patches or plaques) diagnosed on physical examination or by microscopic documentation of *Candida* fungal elements from a noncultured specimen scraped from the oral mucosa. Treatment with oral (systemic) antifungal agents usually produces improvement after 5 or more days of therapy, and such a response can be supportive of the diagnosis.

4. *HIV infection manifestations specific to women.*

a. *General.* Most women with severe immunosuppression secondary to HIV infection exhibit the typical opportunistic infections and other conditions, such as PCP, *Candida* esophagitis, wasting syndrome, cryptococcosis, and toxoplasmosis. However,

HIV infection may have different manifestations in women than in men. Adjudicators must carefully scrutinize the medical evidence and be alert to the variety of medical conditions specific to, or common in, women with HIV infection that may affect their ability to function in the workplace.

b. *Additional considerations for evaluating HIV infection in women.* Many of these manifestations (for example, vulvovaginal candidiasis, pelvic inflammatory disease) occur in women with or without HIV infection, but can be more severe or resistant to treatment, or occur more frequently in a woman whose immune system is suppressed. Therefore, when evaluating the claim of a woman with HIV infection, it is important to consider gynecologic and other problems specific to women, including any associated symptoms (for example, pelvic pain), in assessing the severity of the impairment and resulting functional limitations. We may evaluate manifestations of HIV infection in women under the specific criteria (for example, cervical cancer under 14.08E), under an applicable general category (for example, pelvic inflammatory disease under 14.08A4) or, in appropriate cases, under 14.08K.

5. *Involuntary weight loss.* For purposes of 14.08H, an involuntary weight loss of at least 10 percent of baseline is always considered ''significant.'' Loss of less than 10 percent may or may not be significant, depending on the individual's baseline weight and body habitus. For example, a 7-pound weight loss in a 100-pound woman who is 63 inches tall might be considered significant; but a 14-pound weight loss in a 200-pound woman who is the same height might not be significant. HIV infection that affects the digestive system and results in malnutrition can also be evaluated under 5.08.

G. How do we consider the effects of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

1. *General.* If your impairment does not otherwise meet the requirements of a listing, we will consider your medical treatment in terms of its effectiveness in improving the signs, symptoms, and laboratory abnormalities of your specific immune system disorder or its manifestations, and in terms of any side effects that limit your functioning. We will make every reasonable effort to obtain a specific description of the treatment you receive (including surgery) for your immune system disorder. We consider:

a. The effects of medications you take.

b. Adverse side effects (acute and chronic).

c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).

d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).

e. Variability of your response to treatment (see 14.00G2).

f. The interactive and cumulative effects of your treatments. For example, many individuals with immune system disorders receive treatment both for their immune system disorders and for the manifestations of the disorders or co-occurring impairments, such as treatment for HIV infection and hepatitis C. The interactive and cumulative effects of these treatments may be greater than the effects of each treatment considered separately.

g. The duration of your treatment.

h. Any other aspects of treatment that may interfere with your ability to function.

2. *Variability of your response to treatment.* Your response to treatment and the adverse or beneficial consequences of your treatment may vary widely. The effects of your treatment may be temporary or long term. For example, some individuals may show an initial positive response to a drug or combination of drugs followed by a decrease in effectiveness. When we evaluate your response to treatment and how your treatment may affect you, we consider such factors as disease activity before treatment, requirements for changes in therapeutic regimens, the time required for therapeutic effectiveness of a particular drug or drugs, the limited number of drug combinations that may be available for your impairment(s), and the time-limited efficacy of some drugs. For example, an individual with HIV infection or another immune deficiency disorder who develops pneumonia or tuberculosis may not respond to the same antibiotic regimen used in treating individuals without HIV infection or another immune deficiency disorder, or may not respond to an antibiotic that he or she responded to before. Therefore, we must consider the effects of your treatment on an individual basis, including the effects of your treatment on your ability to function.

3. *How we evaluate the effects of treatment for autoimmune disorders on your ability to function.* Some medications may have acute or long-term side effects. When we consider the effects of corticosteroids or other treatments for autoimmune disorders on your ability to function, we consider the factors in 14.00G1 and 14.00G2. Long-term corticosteroid treatment can cause ischemic necrosis of bone, posterior subcapsular cataract, weight gain, glucose intolerance, increased susceptibility to infection, and osteoporosis that may result in a loss of function. In addition, medications used in the treatment of autoimmune disorders may also have effects on mental functioning, including cognition (for example, memory), concentration, and mood.

4. *How we evaluate the effects of treatment for immune deficiency disorders, excluding HIV infection, on your ability to function.* When we consider the effects of your treatment for

your immune deficiency disorder on your ability to function, we consider the factors in 14.00G1 and 14.00G2. A frequent need for treatment such as intravenous immunoglobulin and gamma interferon therapy can be intrusive and interfere with your ability to work. We will also consider whether you have chronic side effects from these or other medications, including severe fatigue, fever, headaches, high blood pressure, joint swelling, muscle aches, nausea, shortness of breath, or limitations in mental function including cognition (for example, memory), concentration, and mood.

5. *How we evaluate the effects of treatment for HIV infection on your ability to function.*

a. *General.* When we consider the effects of antiretroviral drugs (including the effects of highly active antiretroviral therapy (HAART)) and the effects of treatments for the manifestations of HIV infection on your ability to function, we consider the factors in 14.00G1 and 14.00G2. Side effects of antiretroviral drugs include, but are not limited to: Bone marrow suppression, pancreatitis, gastrointestinal intolerance (nausea, vomiting, diarrhea), neuropathy, rash, hepatotoxicity, lipodystrophy (fat redistribution, such as ''buffalo hump''), glucose intolerance, and lactic acidosis. In addition, medications used in the treatment of HIV infection may also have effects on mental functioning, including cognition (for example, memory), concentration, and mood, and may result in malaise, severe fatigue, joint and muscle pain, and insomnia. The symptoms of HIV infection and the side effects of medication may be indistinguishable from each other. We will consider all of your functional limitations, whether they result from your symptoms or signs of HIV infection or the side effects of your treatment.

b. *Structured treatment interruptions.* A structured treatment interruption (STI, also called a ''drug holiday'') is a treatment practice during which your treating source advises you to stop taking your medications temporarily. An STI in itself does not imply that your medical condition has improved; nor does it imply that you are noncompliant with your treatment because you are following your treating source's advice. Therefore, if you have stopped taking medication because your treating source prescribed or recommended an STI, we will not find that you are failing to follow treatment or draw inferences about the severity of your impairment on this fact alone. We will consider why your treating source has prescribed or recommended an STI and all the other information in your case record when we determine the severity of your impairment.

6. *When there is no record of ongoing treatment.* If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the medical severity and duration of your immune system disorder on the basis of the current objective medical evidence and other evidence in your case record, taking into consideration your medical history, symptoms, clinical and laboratory findings, and medical source opinions. If you have just begun treatment and we cannot determine whether you are disabled based on the evidence we have, we may need to wait to determine the effect of the treatment on your ability to function. The amount of time we need to wait will depend on the facts of your case. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the immune system disorders listings, but your immune system disorder may medically equal a listing or be disabling based on a consideration of your residual functional capacity, age, education, and work experience.

H. How do we consider your symptoms, including your pain, severe fatigue, and malaise?

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your immune system disorder(s) meets or medically equals a listing or in our determination whether you are otherwise able to work. In order for us to consider your symptoms, you must have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 14.00 and in our other regulations. See §§ 404.1528, 404.1529, 416.928, and 416.929. Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

I. How do we use the functional criteria in these listings?

1. The following listings in this body system include standards for evaluating the functional limitations resulting from immune system disorders: 14.02B, for systemic lupus erythematosus; 14.03B, for systemic vasculitis; 14.04D, for systemic sclerosis (scleroderma); 14.05E, for polymyositis and dermatomyositis; 14.06B, for undifferentiated and mixed connective tissue disease; 14.07C, for immune deficiency disorders, excluding HIV infection; 14.08K, for HIV infection; 14.09D, for inflammatory arthritis; and 14.10B, for Sjögren's syndrome.

2. When we use one of the listings cited in 14.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder on your ability to function on a sustained basis. Important factors we will consider when we evaluate your functioning under these listings include, but are not limited to: Your symptoms, the frequency and duration of manifestations of your immune system disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

3. As used in these listings, ''repeated'' means that the manifestations occur on an average of three times a year, or once every 4 months, each lasting 2 weeks or more; or the manifestations do not last for 2 weeks but occur substantially more frequently than three times in a year or once every 4 months; or they occur less frequently than an average of three times a year or once every 4 months but last substantially longer than 2 weeks. Your impairment will satisfy this criterion regardless of whether you have the same kind of manifestation repeatedly, all different manifestations, or any other combination of manifestations; for example, two of the same kind of manifestation and a different one. You must have the required number of manifestations with the frequency and duration required in this section. Also, the manifestations must occur within the period covered by your claim.

4. To satisfy the functional criterion in a listing, your immune system disorder must result in a ''marked'' level of limitation in one of three general areas of functioning: Activities of daily living, social functioning, or difficulties in completing tasks due to deficiencies in concentration, persistence, or pace. Functional limitation may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This could result from persistent or intermittent symptoms, such as depression, severe fatigue, or pain, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. You may also have limitations because of your treatment and its side effects (see 14.00G).

5. When ''marked'' is used as a standard for measuring the degree of functional limitation, it means more than moderate but less than extreme. We do not define ''marked'' by a specific number of different activities of daily living in which your functioning is impaired, different behaviors in which your social functioning is impaired, or tasks that you are able to complete, but by the nature and overall degree of interference with your functioning. You may have a marked limitation when several activities or functions are impaired, or even when only one is impaired. Also, you need not be totally precluded from performing an activity to have a marked limitation, as long as the degree of limitation seriously interferes with your ability to function independently, appropriately, and effectively. The term ''marked'' does not imply that you must be confined to bed, hospitalized, or in a nursing home.

6. *Activities of daily living* include, but are not limited to, such activities as doing household chores, grooming and hygiene, using a post office, taking public transportation, or paying bills. We will find that you have a ''marked'' limitation of activities of daily living if you have a serious limitation in your ability to maintain a household or take public transportation because of symptoms, such as pain, severe fatigue, anxiety, or difficulty concentrating, caused by your immune system disorder (including manifestations of the disorder) or its treatment, even if you are able to perform some self-care activities.

7. *Social functioning* includes the capacity to interact independently, appropriately, effectively, and on a sustained basis with others. It includes the ability to communicate effectively with others. We will find that you have a ''marked'' limitation in maintaining social functioning if you have a serious limitation in social interaction on a sustained basis because of symptoms, such as pain, severe fatigue, anxiety, or difficulty concentrating, or a pattern of exacerbation and remission, caused by your immune system disorder (including manifestations of the disorder) or its treatment, even if you are able to communicate with close friends or relatives.

8. *Completing tasks in a timely manner* involves the ability to sustain concentration, persistence, or pace to permit timely completion of tasks commonly found in work settings. We will find that you have a ''marked'' limitation in completing tasks if you have a serious limitation in your ability to sustain concentration or pace adequate to complete work-related tasks because of symptoms, such as pain, severe fatigue, anxiety, or difficulty concentrating, caused by your immune system disorder (including manifestations of the disorder) or its treatment, even if you are able to do some routine activities of daily living.

J. How do we evaluate your immune system disorder when it does not meet one of these listings?

1. These listings are only examples of immune system disorders that we consider severe enough to prevent you from doing any gainful activity. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. Individuals with immune system disorders, including HIV infection, may manifest signs or symptoms of a mental impairment or of another physical impairment. We may evaluate these impairments under any affected body system. For example, we will evaluate:

a. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 1.00.

b. Ocular involvement, such as dry eye, under 2.00.

c. Respiratory impairments, such as pleuritis, under 3.00.

d. Cardiovascular impairments, such as cardiomyopathy, under 4.00.

e. Digestive impairments, such as hepatitis (including hepatitis C) or weight loss as a result of HIV infection that affects the digestive system, under 5.00.

f. Genitourinary impairments, such as nephropathy, under 6.00.

g. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 7.00.

h. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 8.00.

i. Neurologic impairments, such as neuropathy or seizures, under 11.00.

j. Mental disorders, such as depression, anxiety, or cognitive deficits, under 12.00.

k. Allergic disorders, such as asthma or atopic dermatitis, under 3.00 or 8.00 or under the criteria in another affected body system.

l. Syphilis or neurosyphilis under the criteria for the affected body system; for example, 2.00 Special senses and speech, 4.00 Cardiovascular system, or 11.00 Neurological.

3. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§ 404.1526 and 416.926.) If it does not, you may or may not have the residual functional capacity to engage in substantial gainful activity. Therefore, we proceed to the fourth, and if necessary, the fifth steps of the sequential evaluation process in §§ 404.1520 and 416.920. We use the rules in §§ 404.1594, 416.994, and 416.994a as appropriate, when we decide whether you continue to be disabled.

14.01 *Category of Impairments, Immune System Disorders.*

14.02 *Systemic lupus erythematosus.* As described in 14.00D1. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Repeated manifestations of SLE, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.03 *Systemic vasculitis.* As described in 14.00D2. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Repeated manifestations of systemic vasculitis, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.04 *Systemic sclerosis (scleroderma).* As described in 14.00D3. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. With one of the following:

1. Toe contractures or fixed deformity of one or both feet, resulting in the inability to ambulate effectively as defined in 14.00C6; or

2. Finger contractures or fixed deformity in both hands, resulting in the inability to perform fine and gross movements effectively as defined in 14.00C7; or

3. Atrophy with irreversible damage in one or both lower extremities, resulting in the inability to ambulate effectively as defined in 14.00C6; or

4. Atrophy with irreversible damage in both upper extremities, resulting in the inability to perform fine and gross movements effectively as defined in 14.00C7.

or

C. Raynaud's phenomenon, characterized by:

1. Gangrene involving at least two extremities; or

2. Ischemia with ulcerations of toes or fingers, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively as defined in 14.00C6 and 14.00C7;

or

D. Repeated manifestations of systemic sclerosis (scleroderma), with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.05 *Polymyositis and dermatomyositis.* As described in 14.00D4. With:

A. Proximal limb-girdle (pelvic or shoulder) muscle weakness, resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively as defined in 14.00C6 and 14.00C7.

or

B. Impaired swallowing (dysphagia) with aspiration due to muscle weakness.

or

C. Impaired respiration due to intercostal and diaphragmatic muscle weakness.

or

D. Diffuse calcinosis with limitation of joint mobility or intestinal motility.

or

E. Repeated manifestations of polymyositis or dermatomyositis, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.06 *Undifferentiated and mixed connective tissue disease.* As described in 14.00D5. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Repeated manifestations of undifferentiated or mixed connective tissue disease, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.07 *Immune deficiency disorders, excluding HIV infection.* As described in 14.00E. With:

A. One or more of the following infections. The infection(s) must either be resistant to treatment or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

or

B. Stem cell transplantation as described under 14.00E3. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

or

C. Repeated manifestations of an immune deficiency disorder, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social function.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.08 *Human immunodeficiency virus (HIV) infection.* With documentation as described in 14.00F and one of the following:

A. Bacterial infections:

1. Mycobacterial infection (for example, caused by *M. avium-intracellulare, M. kansasii,* or *M. tuberculosis*) at a site other than the lungs, skin, or cervical or hilar lymph nodes, or pulmonary tuberculosis resistant to treatment; or

2. Nocardiosis; or

3. *Salmonella* bacteremia, recurrent non-typhoid; or

4. Multiple or recurrent bacterial infections, including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment three or more times in a 12-month period. or

B. Fungal infections:

1. Aspergillosis; or

2. Candidiasis involving the esophagus, trachea, bronchi, or lungs, or at a site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes; or

3. Coccidioidomycosis, at a site other than the lungs or lymph nodes; or

4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis); or

5. Histoplasmosis, at a site other than the lungs or lymph nodes; or

6. Mucormycosis; or
7. *Pneumocystis* pneumonia or extrapulmonary *Pneumocystis* infection. or

C. Protozoan or helminthic infections:
1. Cryptosporidiosis, isosporiasis, or microsporidiosis, with diarrhea lasting for 1 month or longer; or
2. Strongyloidiasis, extra-intestinal; or
3. Toxoplasmosis of an organ other than the liver, spleen, or lymph nodes. or

D. Viral infections:
1. *Cytomegalovirus* disease (documented as described in 14.00F3b(ii)) at a site other than the liver, spleen or lymph nodes; or
2. Herpes simplex virus causing:
a. Mucocutaneous infection (for example, oral, genital, perianal) lasting for 1 month or longer; or
b. Infection at a site other than the skin or mucous membranes (for example, bronchitis, pneumonitis, esophagitis, or encephalitis); or
c. Disseminated infection; or
3. Herpes zoster:
a. Disseminated; or
b. With multidermatomal eruptions that are resistant to treatment; or
4. Progressive multifocal leukoencephalopathy.

or

E. Malignant neoplasms:
1. Carcinoma of the cervix, invasive, FIGO stage II and beyond; or
2. Kaposi's sarcoma with:
a. Extensive oral lesions; or
b. Involvement of the gastrointestinal tract, lungs, or other visceral organs; or
3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease); or
4. Squamous cell carcinoma of the anal canal or anal margin.

or

F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above), with extensive fungating or ulcerating lesions not responding to treatment (for example, dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal *Candida,* condyloma caused by human *Papillomavirus,* genital ulcerative disease).

or

G. HIV encephalopathy, characterized by cognitive or motor dysfunction that limits function and progresses.

or

H. HIV wasting syndrome, characterized by involuntary weight loss of 10 percent or more of baseline (computed based on pounds, kilograms, or body mass index (BMI)) or other significant involuntary weight loss as described in 14.00F5, and in the absence of a concurrent illness that could explain the findings. With either:

1. Chronic diarrhea with two or more loose stools daily lasting for 1 month or longer; or
2. Chronic weakness and documented fever greater than 38 °C (100.4 °F) for the majority of 1 month or longer.

or

I. Diarrhea, lasting for 1 month or longer, resistant to treatment, and requiring intravenous hydration, intravenous alimentation, or tube feeding.

or

J. One or more of the following infections (other than described in A-I, above). The infection(s) must either be resistant to treatment or require hospitalization or intravenous treatment three or more times in a 12-month period.
1. Sepsis; or
2. Meningitis; or
3. Pneumonia; or
4. Septic arthritis; or
5. Endocarditis; or
6. Sinusitis documented by appropriate medically acceptable imaging.

or

K. Repeated (as defined in 14.00I3) manifestations of HIV infection, including those listed in 14.08A–J, but without the requisite findings for those listings (for example, carcinoma of the cervix not meeting the criteria in 14.08E, diarrhea not meeting the criteria in 14.08I), or other manifestations (for example, oral hairy leukoplakia, myositis, pancreatitis, hepatitis, peripheral neuropathy, glucose intolerance, muscle weakness, cognitive or other mental limitation) resulting in significant, documented symptoms or signs (for example, severe fatigue, fever, malaise, involuntary weight loss, pain, night sweats, nausea, vomiting, headaches, or insomnia) and one of the following at the marked level:
1. Limitation of activities of daily living.
2. Limitation in maintaining social functioning.
3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.09 *Inflammatory arthritis.* As described in 14.00D6. With:

A. Persistent inflammation or persistent deformity of:
1. One or more major peripheral weight-bearing joints resulting in the inability to ambulate effectively (as defined in 14.00C6); or
2. One or more major peripheral joints in each upper extremity resulting in the inability to perform fine and gross movements effectively (as defined in 14.00C7).

or

B. Inflammation or deformity in one or more major peripheral joints with:
1. Involvement of two or more organs/body systems with one of the organs/body systems

involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

C. Ankylosing spondylitis or other spondyloarthropathies, with:

1. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 45° or more of flexion from the vertical position (zero degrees); or

2. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 30° or more of flexion (but less than 45°) measured from the vertical position (zero degrees), and involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity.

or

D. Repeated manifestations of inflammatory arthritis, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.10 *Sjögren's syndrome.* As described in 14.00D7. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Repeated manifestations of Sjögren's syndrome, with at least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss) and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

Part B

Medical criteria for the evaluation of impairments of children under age 18 (where criteria in part A do not give appropriate consideration to the particular disease process in childhood).

Sec.

100.00 Low Birth Weight and Failure to Thrive

A. *What conditions do we evaluate under these listings?* We evaluate low birth weight (LBW) in infants from birth to attainment of age 1 and failure to thrive (FTT) in infants and toddlers from birth to attainment of age 3.

B. *How do we evaluate disability based on LBW under 100.04?* In 100.04A and 100.04B, we use an infant's birth weight as documented by an original or certified copy of the infant's birth certificate or by a medical record signed by a physician. *Birth weight* means the first weight recorded after birth. In 100.04B, *gestational age* is the infant's age based on the date of conception as recorded in the medical record. If the infant's impairment meets the requirements for listing 100.04A or 100.04B, we will follow the rule in §416.990(b)(11) of this chapter.

C. *How do we evaluate disability based on FTT under 100.05?*

1. *General.* We establish FTT with or without a known cause when we have documentation of an infant's or a toddler's growth failure and developmental delay from an acceptable medical source(s) as defined in §416.913(a) of this chapter. We require documentation of growth measurements in 100.05A and developmental delay described in 100.05B or 100.05C within the same consecutive 12-month period. The dates of developmental testing and reports may be different from the dates of growth measurements. After the attainment of age 3, we evaluate growth failure under the affected body system(s).

2. *Growth failure.* Under 100.05A, we use the appropriate table(s) under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile. The child does not need to have a digestive disorder for purposes of 100.05.

a. For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

b. For children age 2 to attainment of age 3, we use the body mass index (BMI)-for-age table corresponding to the child's gender (Table III or Table IV).

c. BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

d. *Growth measurements.* The weight-for-length measurements for children from birth to the attainment of age 2 and BMI-for-age measurements for children age 2 to attainment of age 3 that are required for this listing must be obtained within a 12-month period and at least 60 days apart. If a child attains age 2 during the evaluation period, additional measurements are not needed. Any measurements taken before the child attains age 2 can be used to evaluate the impairment under the appropriate listing for the child's age. If the child attains age 3 during the evaluation period, the measurements can be used to evaluate the impairment in the affected body system.

3. *Developmental delay.*

a. Under 100.05B and C, we use reports from acceptable medical sources to establish delay in a child's development.

b. Under 100.05B, we document the severity of developmental delay with results from a standardized developmental assessment, which compares a child's level of development to the level typically expected for his or her chronological age. If the child was born prematurely, we may use the corrected chronological age (CCA) for comparison. (See §416.924b(b) of this chapter.) CCA is the chronological age adjusted by a period of gestational prematurity. CCA = (chronological age)—(number of weeks premature). Acceptable medical sources or early intervention specialists, physical or occupational therapists, and other sources may conduct standardized developmental assessments and developmental screenings. The results of these tests and screenings must be accompanied by a statement or records from an acceptable medical source who established the child has a developmental delay.

c. Under 100.05C, when there are no results from a standardized developmental assessment in the case record, we need narrative developmental reports from the child's medical sources in sufficient detail to assess the severity of his or her developmental delay. A narrative developmental report is based on clinical observations, progress notes, and well-baby check-ups. To meet the requirements for 100.05C, the report must include: The child's developmental history; examination findings (with abnormal findings noted on repeated examinations); and an overall assessment of the child's development (that is, more than one or two isolated skills) by the medical source. Some narrative developmental reports may include results from developmental screening tests, which can identify a child who is not developing or achieving skills within expected timeframes. Although medical sources may refer to screening test results as supporting evidence in the narrative developmental report, screening test results alone cannot establish a diagnosis or the severity of developmental delay.

D. *How do we evaluate disorders that do not meet one of these listings?*

1. We may find infants disabled due to other disorders when their birth weights are greater than 1200 grams but less than 2000 grams and their weight and gestational age do not meet listing 100.04. The most common disorders of prematurity and LBW include retinopathy of prematurity (ROP), chronic lung disease of infancy (CLD, previously known as bronchopulmonary dysplasia, or BPD), intraventricular hemorrhage (IVH), necrotizing enterocolitis (NEC), and periventricular leukomalacia (PVL). Other disorders include poor nutrition and growth failure, hearing disorders, seizure disorders, cerebral palsy, and developmental disorders. We evaluate these disorders under the affected body systems.

2. We may evaluate infants and toddlers with growth failure that is associated with a known medical disorder under the body system of that medical disorder, for example, the respiratory or digestive body systems.

3. If an infant or toddler has a severe medically determinable impairment(s) that does not meet the criteria of any listing, we must also consider whether the child has an impairment(s) that medically equals a listing (see §416.926 of this chapter). If the child's impairment(s) does not meet or medically equal a listing, we will determine whether the child's impairment(s) functionally equals the listings (see §416.926a of this chapter) considering the factors in §416.924a of this chapter. We use the rule in §416.994a of this chapter when we decide whether a child continues to be disabled.

100.01 Category of Impairments, Low Birth Weight and Failure to Thrive

100.04 *Low birth weight in infants from birth to attainment of age 1.*

A. Birth weight (see 100.00B) of less than 1200 grams.

OR

B. The following gestational age and birth weight:

Gestational age (in weeks)	Birth weight
37–40	2000 grams or less.
36	1875 grams or less.
35	1700 grams or less.
34	1500 grams or less.
33	1325 grams or less.
32	1250 grams or less.

100.05 *Failure to thrive in children from birth to attainment of age 3* (see 100.00C), documented by A and B, or A and C.

A. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2,* three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table in listing 105.08B1; or

2. *For children age 2 to attainment of age 3,* three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table in listing 105.08B2.

AND

B. Developmental delay (see 100.00C1 and C3), established by an acceptable medical source and documented by findings from one current report of a standardized developmental assessment (see 100.00C3b) that:

1. Shows development not more than two-thirds of the level typically expected for the child's age; or

2. Results in a valid score that is at least two standard deviations below the mean.

OR

C. Developmental delay (see 100.00C3), established by an acceptable medical source and documented by findings from two narrative developmental reports (see 100.00C3c) that:

1. Are dated at least 120 days apart (see 100.00C1); and

2. Indicate current development not more than two-thirds of the level typically expected for the child's age.

101.00 Musculoskeletal System

A. *Disorders of the musculoskeletal system* may result from hereditary, congenital, or acquired pathologic processes. Impairments may result from infectious, inflammatory, or degenerative processes, traumatic or developmental events, or neoplastic, vascular, or toxic/metabolic diseases.

B. Loss of Function

1. *General.* Under this section, loss of function may be due to bone or joint deformity or destruction from any cause; miscellaneous disorders of the spine with or without radiculopathy or other neurological deficits; amputation; or fractures or soft tissue injuries, including burns, requiring prolonged periods of immobility or convalescence. The provisions of 101.02 and 101.03 notwithstanding, inflammatory arthritis is evaluated under 114.09 (see 114.00D6). Impairments with neurological causes are to be evaluated under 111.00ff.

2. How We Define Loss of Function in These Listings

a. *General.* Regardless of the cause(s) of a musculoskeletal impairment, functional loss for purposes of these listings is defined as the inability to ambulate effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment, or the inability to perform fine and gross movements effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment. The inability to ambulate effectively or the inability to perform fine and gross movements effectively must have lasted, or be expected to last, for at least 12 months. For the purposes of these criteria, consideration of the ability to perform these activities must be from a physical standpoint alone. When there is an inability to perform these activities due to a mental impairment, the criteria in 112.00ff are to be used. We will determine whether a child can ambulate effectively or can perform fine and gross movements effectively based on the medical and other evidence in the case record, generally without developing additional evidence about the child's ability to perform the specific activities listed as examples in 101.00B2b(2) and (3) and 101.00B2c(2) and (3).

b. What We Mean by Inability To Ambulate Effectively

(1) *Definition.* Inability to ambulate effectively means an extreme limitation of the ability to walk; *i.e.*, an impairment that interferes very seriously with the child's ability to independently initiate, sustain, or complete activities. Ineffective ambulation is defined generally as having insufficient lower extremity functioning (see 101.00J) to permit independent ambulation without the use of a hand-held assistive device(s) that limits the functioning of both upper extremities. (Listing 101.05C is an exception to this general definition because the child has the use of only one upper extremity due to amputation of a hand.)

(2) *How we assess inability to ambulate effectively for children too young to be expected to walk independently.* For children who are too young to be expected to walk independently, consideration of function must be based on assessment of limitations in the ability to perform comparable age-appropriate activities with the lower extremities, given normal developmental expectations. For such children, an extreme level of limitation means skills or performance at no greater than one-half of age-appropriate expectations based on an overall developmental assessment rather than on one or two isolated skills.

(3) *How we assess inability to ambulate effectively for older children.* Older children, who

would be expected to be able to walk when compared to other children the same age who do not have impairments, must be capable of sustaining a reasonable walking pace over a sufficient distance to be able to carry out age-appropriate activities. They must have the ability to travel age-appropriately without extraordinary assistance to and from school or a place of employment. Therefore, examples of ineffective ambulation for older children include, but are not limited to, the inability to walk without the use of a walker, two crutches or two canes, the inability to walk a block at a reasonable pace on rough or uneven surfaces, the inability to use standard public transportation, the inability to carry out age-appropriate school activities independently, and the inability to climb a few steps at a reasonable pace with the use of a single hand rail. The ability to walk independently about the child's home or a short distance at school without the use of assistive devices does not, in and of itself, constitute effective ambulation.

c. What We Mean by Inability To Perform Fine and Gross Movements Effectively

(1) *Definition.* Inability to perform fine and gross movements effectively means an extreme loss of function of both upper extremities; *i.e.*, an impairment that interferes very seriously with the child's ability to independently initiate, sustain, or complete activities. To use their upper extremities effectively, a child must be capable of sustaining such functions as reaching, pushing, pulling, grasping, and fingering in an age-appropriate manner to be able to carry out age-appropriate activities.

(2) *How we assess inability to perform fine and gross movements in very young children.* For very young children, we consider limitations in the ability to perform comparable age-appropriate activities involving the upper extremities compared to the ability of children the same age who do not have impairments. For such children, an extreme level of limitation means skills or performance at no greater than one-half of age-appropriate expectations based on an overall developmental assessment.

(3) *How we assess inability to perform fine and gross movements in older children.* For older children, examples of inability to perform fine and gross movements effectively include, but are not limited to, the inability to prepare a simple meal and feed oneself, the inability to take care of personal hygiene, or the inability to sort and handle papers or files, depending upon which activities are age-appropriate.

d. *Pain or other symptoms.* Pain or other symptoms may be an important factor contributing to functional loss. In order for pain or other symptoms to be found to affect a child's ability to function in an age-appropriate manner or to perform basic work activities, medical signs or laboratory findings must show the existence of a medically determinable impairment(s) that could reasonably be expected to produce the pain or other symptoms. The musculoskeletal listings that include pain or other symptoms among their criteria also include criteria for limitations in functioning as a result of the listed impairment, including limitations caused by pain. It is, therefore, important to evaluate the intensity and persistence of such pain or other symptoms carefully in order to determine their impact on the child's functioning under these listings. See also §§ 404.1525(f) and 404.1529 of this part, and §§ 416.925(f) and 416.929 of part 416 of this chapter.

C. Diagnosis and Evaluation

1. *General.* Diagnosis and evaluation of musculoskeletal impairments should be supported, as applicable, by detailed descriptions of the joints, including ranges of motion, condition of the musculature (e.g., weakness, atrophy), sensory or reflex changes, circulatory deficits, and laboratory findings, including findings on x-ray or other appropriate medically acceptable imaging. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Purchase of certain medically acceptable imaging.* While any appropriate medically acceptable imaging is useful in establishing the diagnosis of musculoskeletal impairments, some tests, such as CAT scans and MRIs, are quite expensive, and we will not routinely purchase them. Some, such as myelograms, are invasive and may involve significant risk. We will not order such tests. However, when the results of any of these tests are part of the existing evidence in the case record we will consider them together with the other relevant evidence.

3. *Consideration of electrodiagnostic procedures.* Electrodiagnostic procedures may be useful in establishing the clinical diagnosis, but do not constitute alternative criteria to the requirements of 101.04.

D. *The physical examination* must include a detailed description of the rheumatological, orthopedic, neurological, and other findings appropriate to the specific impairment being evaluated. These physical findings must be determined on the basis of objective observation during the examination and not simply a report of the child's allegation; e.g., ''He says his leg is weak, numb.'' Alternative testing methods should be used to verify the abnormal findings; e.g., a seated straight-leg raising test in addition to a supine straight-

leg raising test. Because abnormal physical findings may be intermittent, their presence over a period of time must be established by a record of ongoing management and evaluation. Care must be taken to ascertain that the reported examination findings are consistent with the child's age and activities.

E. Examination of the Spine

1. *General.* Examination of the spine should include a detailed description of gait, range of motion of the spine given quantitatively in degrees from the vertical position (zero degrees) or, for straight-leg raising from the sitting and supine position (zero degrees), any other appropriate tension signs, motor and sensory abnormalities, muscle spasm, when present, and deep tendon reflexes. Observations of the child during the examination should be reported; e.g., how he or she gets on and off the examination table. Inability to walk on the heels or toes, to squat, or to arise from a squatting position, when appropriate, may be considered evidence of significant motor loss. However, a report of atrophy is not acceptable as evidence of significant motor loss without circumferential measurements of both thighs and lower legs, or both upper and lower arms, as appropriate, at a stated point above and below the knee or elbow given in inches or centimeters. Additionally, a report of atrophy should be accompanied by measurement of the strength of the muscle(s) in question generally based on a grading system of 0 to 5, with 0 being complete loss of strength and 5 being maximum strength. A specific description of atrophy of hand muscles is acceptable without measurements of atrophy but should include measurements of grip and pinch strength. However, because of the unreliability of such measurement in younger children, these data are not applicable to children under 5 years of age.

2. *When neurological abnormalities persist.* Neurological abnormalities may not completely subside after treatment or with the passage of time. Therefore, residual neurological abnormalities that persist after it has been determined clinically or by direct surgical or other observation that the ongoing or progressive condition is no longer present will not satisfy the required findings in 101.04. More serious neurological deficits (paraparesis, paraplegia) are to be evaluated under the criteria in 111.00ff.

F. *Major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

G. *Measurements of joint motion* are based on the techniques described in the chapter on the extremities, spine, and pelvis in the current edition of the ''Guides to the Evaluation of Permanent Impairment'' published by the American Medical Association.

H. Documentation.

1. *General.* Musculoskeletal impairments frequently improve with time or respond to treatment. Therefore, a longitudinal clinical record is generally important for the assessment of severity and expected duration of an impairment unless the child is a newborn or the claim can be decided favorably on the basis of the current evidence.

2. *Documentation of medically prescribed treatment and response.* Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever evidence of such treatment is available it must be considered.

3. *When there is no record of ongoing treatment.* Some children will not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In such cases, evaluation will be made on the basis of the current objective medical evidence and other available evidence, taking into consideration the child's medical history, symptoms, and medical source opinions. Even though a child who does not receive treatment may not be able to show an impairment that meets the criteria of one of the musculoskeletal listings, the child may have an impairment(s) that is either medically or, in the case of a claim for benefits under part 416 of this chapter, functionally equivalent in severity to one of the listed impairments.

4. *Evaluation when the criteria of a musculoskeletal listing are not met.* These listings are only examples of common musculoskeletal disorders that are severe enough to find a child disabled. Therefore, in any case in which a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will consider whether the child's impairment(s) is medically or, in the case of a claim for benefits under part 416 of this chapter, functionally equivalent in severity to the criteria of a listing. (See §§404.1526, 416.926, and 416.926a.) Individuals with claims for benefits under part 404, who have an impairment(s) with a level of severity that does not meet or equal the criteria of the musculoskeletal listings may or may not have the RFC that would enable them to engage

in substantial gainful activity. Evaluation of the impairment(s) of these individuals should proceed through the final steps of the sequential evaluation process in §404.1520 (or, as appropriate, the steps in the medical improvement review standard in §404.1594).

I. Effects of Treatment

1. *General.* Treatments for musculoskeletal disorders may have beneficial effects or adverse side effects. Therefore, medical treatment (including surgical treatment) must be considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the disorder, and in terms of any side effects that may further limit the child.

2. *Response to treatment.* Response to treatment and adverse consequences of treatment may vary widely. For example, a pain medication may relieve a child's pain completely, partially, or not at all. It may also result in adverse effects, e.g., drowsiness, dizziness, or disorientation, that compromise the child's ability to function. Therefore, each case must be considered on an individual basis, and include consideration of the effects of treatment on the child's ability to function.

3. *Documentation.* A specific description of the drugs or treatment given (including surgery), dosage, frequency of administration, and a description of the complications or response to treatment should be obtained. The effects of treatment may be temporary or long-term. As such, the finding regarding the impact of treatment must be based on a sufficient period of treatment to permit proper consideration or judgment about future functioning.

J. Orthotic, Prosthetic, or Assistive Devices

1. *General.* Consistent with clinical practice, children with musculoskeletal impairments may be examined with and without the use of any orthotic, prosthetic, or assistive devices as explained in this section.

2. *Orthotic devices.* Examination should be with the orthotic device in place and should include an evaluation of the child's maximum ability to function effectively with the orthosis. It is unnecessary to routinely evaluate the child's ability to function without the orthosis in place. If the child has difficulty with, or is unable to use, the orthotic device, the medical basis for the difficulty should be documented. In such cases, if the impairment involves a lower extremity or extremities, the examination should include information on the child's ability to ambulate effectively without the device in place unless contraindicated by the medical judgment of a physician who has treated or examined the child.

3. *Prosthetic devices.* Examination should be with the prosthetic device in place. In amputations involving a lower extremity or extremities, it is unnecessary to evaluate the child's ability to walk without the prosthesis in place. However, the child's medical ability to use a prosthesis to ambulate effectively, as defined in 101.00B2b, should be evaluated. The condition of the stump should be evaluated without the prosthesis in place.

4. *Hand-held assistive devices.* When a child with an impairment involving a lower extremity or extremities uses a hand-held assistive device, such as a cane, crutch or walker, examination should be with and without the use of the assistive device unless contraindicated by the medical judgment of a physician who has treated or examined the child. The child's ability to ambulate with and without the device provides information as to whether, or the extent to which, the child is able to ambulate without assistance. The medical basis for the use of any assistive device (e.g., instability, weakness) should be documented. The requirement to use a hand-held assistive device may also impact on the child's functional capacity by virtue of the fact that one or both upper extremities are not available for such activities as lifting, carrying, pushing, and pulling.

K. *Disorders of the spine,* listed in 101.04, result in limitations because of distortion of the bony and ligamentous architecture of the spine and associated impingement on nerve roots (including the cauda equina) or spinal cord. Such impingement on nerve tissue may result from a herniated nucleus pulposus or other miscellaneous conditions. Neurological abnormalities resulting from these disorders are to be evaluated by referral to the neurological listings in 111.00ff, as appropriate. (See also 101.00B and E.)

1. *Herniated nucleus pulposus* is a disorder frequently associated with the impingement of a nerve root, but occurs infrequently in children. Nerve root compression results in a specific neuro-anatomic distribution of symptoms and signs depending upon the nerve root(s) compromised.

2. *Other miscellaneous conditions* that may cause weakness of the lower extremities, sensory changes, areflexia, trophic ulceration, bladder or bowel incontinence, and that should be evaluated under 101.04 include, but are not limited to, lysosomal disorders, metabolic disorders, vertebral osteomyelitis, vertebral fractures and achondroplasia. Disorders such as spinal dysrhaphism, (e.g., spina bifida) diastematomyelia, and tethered cord syndrome may also cause such abnormalities. In these cases, there may be gait difficulty and deformity of the lower extremities based on neurological abnormalities, and the neurological effects are to be evaluated under the criteria in 111.00ff.

L. *Abnormal curvatures of the spine.* Abnormal curvatures of the spine (specifically, scoliosis, kyphosis and kyphoscoliosis) can result in impaired ambulation, but may also adversely affect functioning in body systems

other than the musculoskeletal system. For example, a child's ability to breathe may be affected; there may be cardiac difficulties (e.g., impaired myocardial function); or there may be disfigurement resulting in withdrawal or isolation. When there is impaired ambulation, evaluation of equivalence may be made by reference to 114.09A. When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 114.09C. When there is respiratory or cardiac involvement or an associated mental disorder, evaluation may be made under 103.00ff, 104.00ff, or 112.00ff, as appropriate. Other consequences should be evaluated according to the listing for the affected body system.

M. *Under continuing surgical management,* as used in 101.07 and 101.08, refers to surgical procedures and any other associated treatments related to the efforts directed toward the salvage or restoration of functional use of the affected part. It may include such factors as post-surgical procedures, surgical complications, infections, or other medical complications, related illnesses, or related treatments that delay the child's attainment of maximum benefit from therapy. When burns are not under continuing surgical management, see 108.00F.

N. *After maximum benefit from therapy has been achieved* in situations involving fractures of an upper extremity (101.07), or soft tissue injuries (101.08), *i.e.*, there have been no significant changes in physical findings or on appropriate medically acceptable imaging for any 6-month period after the last definitive surgical procedure or other medical intervention, evaluation must be made on the basis of the demonstrable residuals, if any. A finding that 101.07 or 101.08 is met must be based on a consideration of the symptoms, signs, and laboratory findings associated with recent or anticipated surgical procedures and the resulting recuperative periods, including any related medical complications, such as infections, illnesses, and therapies which impede or delay the efforts toward restoration of function. Generally, when there has been no surgical or medical intervention for 6 months after the last definitive surgical procedure, it can be concluded that maximum therapeutic benefit has been reached. Evaluation at this point must be made on the basis of the demonstrable residual limitations, if any, considering the child's impairment-related symptoms, signs, and laboratory findings, any residual symptoms, signs, and laboratory findings associated with such surgeries, complications, and recuperative periods, and other relevant evidence.

O. *Major function of the face and head,* for purposes of listing 101.08, relates to impact on any or all of the activities involving vision, hearing, speech, mastication, and the initiation of the digestive process.

P. *When surgical procedures have been performed,* documentation should include a copy of the operative notes and available pathology reports.

101.01 Category of Impairments, Musculoskeletal

101.02 *Major dysfunction of a joint(s) (due to any cause):* Characterized by gross anatomical deformity (e.g., subluxation, contracture, bony or fibrous ankylosis, instability) and chronic joint pain and stiffness with signs of limitation of motion or other abnormal motion of the affected joint(s), and findings on appropriate medically acceptable imaging of joint space narrowing, bony destruction, or ankylosis of the affected joint(s). With:

A. Involvement of one major peripheral weight-bearing joint (*i.e.*, hip, knee, or ankle), resulting in inability to ambulate effectively, as defined in 101.00B2b;

or

B. Involvement of one major peripheral joint in each upper extremity (*i.e.*, shoulder, elbow, or wrist-hand), resulting in inability to perform fine and gross movements effectively, as defined in 101.00B2c.

101.03 *Reconstructive surgery or surgical arthrodesis of a major weight-bearing joint,* with inability to ambulate effectively, as defined in 101.00B2b, and return to effective ambulation did not occur, or is not expected to occur, within 12 months of onset.

101.04 *Disorders of the spine* (e.g., lysosomal disorders, metabolic disorders, vertebral osteomyelitis, vertebral fracture, achondroplasia) resulting in compromise of a nerve root (including the cauda equina) or the spinal cord, with evidence of nerve root compression characterized by neuro-anatomic distribution of pain, limitation of motion of the spine, motor loss (atrophy with associated muscle weakness or muscle weakness) accompanied by sensory or reflex loss and, if there is involvement of the lower back, positive straight-leg raising test (sitting and supine).

101.05 *Amputation (due to any cause).*

A. Both hands;

or

B. One or both lower extremities at or above the tarsal region, with stump complications resulting in medical inability to use a prosthetic device to ambulate effectively, as defined in 101.00B2b, which have lasted or are expected to last for at least 12 months;

or

C. One hand and one lower extremity at or above the tarsal region, with inability to ambulate effectively, as defined in 101.00B2b;

or

D. Hemipelvectomy or hip disarticulation.

101.06 *Fracture of the femur, tibia, pelvis, or one or more of the tarsal bones.* With:

A. Solid union not evident on appropriate medically acceptable imaging, and not clinically solid;

and

B. Inability to ambulate effectively, as defined in 101.00B2b, and return to effective ambulation did not occur or is not expected to occur within 12 months of onset.

101.07 *Fracture of an upper extremity* with nonunion of a fracture of the shaft of the humerus, radius, or ulna, under continuing surgical management, as defined in 101.00M, directed toward restoration of functional use of the extremity, and such function was not restored or expected to be restored within 12 months of onset.

101.08 *Soft tissue injury (e.g., burns)* of an upper or lower extremity, trunk, or face and head, under continuing surgical management, as defined in 101.00M, directed toward the salvage or restoration of major function, and such major function was not restored or expected to be restored within 12 months of onset. Major function of the face and head is described in 101.00O.

102.00 Special Senses and Speech

A. *How do we evaluate visual disorders?*

1. *What are visual disorders?* Visual disorders are abnormalities of the eye, the optic nerve, the optic tracts, or the brain that may cause a loss of visual acuity or visual fields. A loss of visual acuity limits your ability to distinguish detail, read, do fine work, or perform other age-appropriate activities. A loss of visual fields limits your ability to perceive visual stimuli in the peripheral extent of vision.

2. *How do we define statutory blindness?* Statutory blindness is blindness as defined in sections 216(i)(1) and 1614(a)(2) of the Social Security Act (Act).

a. The Act defines blindness as central visual acuity of 20/200 or less in the better eye with the use of a correcting lens. We use your best-corrected central visual acuity for distance in the better eye when we determine if this definition is met. (For visual acuity testing requirements, see 102.00A5.)

b. The Act also provides that an eye that has a visual field limitation such that the widest diameter of the visual field subtends an angle no greater than 20 degrees is considered as having a central visual acuity of 20/200 or less. (For visual field testing requirements, see 102.00A6.)

c. You have statutory blindness only if your visual disorder meets the criteria of 102.02A, 102.02B, or 102.03A. You do not have statutory blindness if your visual disorder medically equals the criteria of 102.02A, 102.02B, or 102.03A or meets or medically equals the criteria of 102.03B, 102.03C, 102.04A, or 102.04B because your disability is based on criteria other than those in the statutory definition of blindness.

3. *What evidence do we need to establish statutory blindness under title XVI?* To establish that you have statutory blindness under title XVI, we need evidence showing only that your central visual acuity in your better eye or your visual field in your better eye meets the criteria in 102.00A2, provided that those measurements are consistent with the other evidence in your case record. We do not need documentation of the cause of your blindness. Also, there is no duration requirement for statutory blindness under title XVI (see §§ 416.981 and 416.983 of this chapter).

4. *What evidence do we need to evaluate visual disorders, including those that result in statutory blindness under title II?* To evaluate your visual disorder, we usually need a report of an eye examination that includes measurements of your best-corrected central visual acuity (see 102.00A5) or the extent of your visual fields (see 102.00A6), as appropriate. If you have visual acuity or visual field loss, we need documentation of the cause of the loss. A standard eye examination will usually indicate the cause of any visual acuity loss. A standard eye examination can also indicate the cause of some types of visual field deficits. Some disorders, such as cortical visual disorders, may result in abnormalities that do not appear on a standard eye examination. If the standard eye examination does not indicate the cause of your vision loss, we will request the information used to establish the presence of your visual disorder. If your visual disorder does not satisfy the criteria in 102.02, 102.03, or 102.04, we will request a description of how your visual disorder affects your ability to function.

5. *How do we measure your best-corrected central visual acuity?*

a. *Visual acuity testing.* When we need to measure your best-corrected central visual acuity, which is your optimal visual acuity attainable with the use of a corrective lens, we use visual acuity testing for distance that was carried out using Snellen methodology or any other testing methodology that is comparable to Snellen methodology.

(i) Your best-corrected central visual acuity for distance is usually measured by determining what you can see from 20 feet. If your visual acuity is measured for a distance other than 20 feet, we will convert it to a 20-foot measurement. For example, if your visual acuity is measured at 10 feet and is reported as 10/40, we will convert this measurement to 20/80.

(ii) A visual acuity recorded as CF (counts fingers), HM (hand motion only), LP or LPO (light perception or light perception only), or NLP (no light perception) indicates that no optical correction will improve your visual acuity. If your central visual acuity in an

eye is recorded as CF, HM, LP or LPO, or NLP, we will determine that your best-corrected central visual acuity is 20/200 or less in that eye.

(iii) We will not use the results of pinhole testing or automated refraction acuity to determine your best-corrected central visual acuity. These tests provide an estimate of potential visual acuity but not an actual measurement of your best-corrected central visual acuity.

(iv) Very young children, such as infants and toddlers, cannot participate in testing using Snellen methodology or other comparable testing. If you are unable to participate in testing using Snellen methodology or other comparable testing due to your young age, we will consider clinical findings of your fixation and visual-following behavior. If both these behaviors are absent, we will consider the anatomical findings or the results of neuroimaging, electroretinogram, or visual evoked response (VER) testing when this testing has been performed.

b. *Other test charts.*

(i) Children between the ages of 3 and 5 often cannot identify the letters on a Snellen or other letter test chart. Specialists with expertise in assessment of childhood vision use alternate methods for measuring visual acuity in young children. We consider alternate methods, for example, the Landolt C test or the tumbling-E test, which are used to evaluate young children who are unable to participate in testing using Snellen methodology, to be comparable to testing using Snellen methodology.

(ii) Most test charts that use Snellen methodology do not have lines that measure visual acuity between 20/100 and 20/200. Some test charts, such as the Bailey-Lovie or the Early Treatment Diabetic Retinopathy Study (ETDRS), used mostly in research settings, have such lines. If your visual acuity is measured with one of these charts, and you cannot read any of the letters on the 20/100 line, we will determine that you have statutory blindness based on a visual acuity of 20/200 or less. For example, if your best-corrected central visual acuity for distance in the better eye is 20/160 using an ETDRS chart, we will find that you have statutory blindness. Regardless of the type of test chart used, you do not have statutory blindness if you can read at least one letter on the 20/100 line. For example, if your best-corrected central visual acuity for distance in the better eye is 20/125 + 1 using an ETDRS chart, we will find that you do not have statutory blindness because you are able to read one letter on the 20/100 line.

c. *Testing using a specialized lens.* In some instances, you may have visual acuity testing performed using a specialized lens, such as a contact lens. We will use the visual acuity measurements obtained with a specialized lens only if you have demonstrated the ability to use the specialized lens on a sustained basis. We will not use visual acuity measurements obtained with telescopic lenses.

d. *Cycloplegic refraction* is an examination of the eye performed after administering cycloplegic eye drops capable of relaxing the ability of the pupil to become smaller and temporarily paralyzing the focusing muscles. If your case record contains the results of cycloplegic refraction, we may use the results to determine your best-corrected central visual acuity. We will not purchase cycloplegic refraction.

e. *VER testing* measures your response to visual events and can often detect dysfunction that is undetectable through other types of examinations. If you have an absent response to VER testing in your better eye, we will determine that your best-corrected central visual acuity is 20/200 or less in that eye and that your visual acuity loss satisfies the criterion in 102.02A or 102.02B4, as appropriate, when these test results are consistent with the other evidence in your case record. If you have a positive response to VER testing in an eye, we will not use that result to determine your best-corrected central visual acuity in that eye.

6. *How do we measure your visual fields?*

a. *General.* We generally need visual field testing when you have a visual disorder that could result in visual field loss, such as glaucoma, retinitis pigmentosa, or optic neuropathy, or when you display behaviors that suggest a visual field loss. When we need to measure the extent of your visual field loss, we use visual field testing (also referred to as perimetry) carried out using automated static threshold perimetry performed on an acceptable perimeter. (For perimeter requirements, see 102.00A9.)

b. *Automated static threshold perimetry requirements.*

(i) The test must use a white size III Goldmann stimulus and a 31.5 apostilb (asb) white background (or a 10 candela per square meter (cd/m^2) white background). The stimuli test locations must be no more than 6 degrees apart horizontally or vertically. Measurements must be reported on standard charts and include a description of the size and intensity of the test stimulus.

(ii) We measure the extent of your visual field loss by determining the portion of the visual field in which you can see a white III4e stimulus. The ''III'' refers to the standard Goldmann test stimulus size III (4 mm^2), and the ''4e'' refers to the standard Goldmann intensity filter (0 decibel (dB) attenuation, which allows presentation of the maximum luminance) used to determine the intensity of the stimulus.

(iii) In automated static threshold perimetry, the intensity of the stimulus varies. The intensity of the stimulus is expressed in

decibels (dB). A perimeter's maximum stimulus luminance is usually assigned the value 0 dB. We need to determine the dB level that corresponds to a 4e intensity for the particular perimeter being used. We will then use the dB printout to determine which points you see at a 4e intensity level (a ''seeing point''). For example:

A. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 10,000 asb, a 10 dB stimulus is equivalent to a 4e stimulus. Any point you see at 10 dB or greater is a seeing point.

B. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 4,000 asb, a 6 dB stimulus is equivalent to a 4e stimulus. Any point you see at 6 dB or greater is a seeing point.

C. When the maximum stimulus luminance (0 dB stimulus) on an acceptable perimeter is 1,000 asb, a 0 dB stimulus is equivalent to a 4e stimulus. Any point you see at 0 dB or greater is a seeing point.

c. *Evaluation under 102.03A.* To determine statutory blindness based on visual field loss in your better eye (102.03A), we need the results of a visual field test that measures the central 24 to 30 degrees of your visual field; that is, the area measuring 24 to 30 degrees from the point of fixation. Acceptable tests include the Humphrey Field Analyzer (HFA) 30–2, HFA 24–2, and Octopus 32.

d. *Evaluation under 102.03B.* To determine whether your visual field loss meets listing 102.03B, we use the mean deviation or defect (MD) from acceptable automated static threshold perimetry that measures the central 30 degrees of the visual field. MD is the average sensitivity deviation from normal values for all measured visual field locations. When using results from HFA tests, which report the MD as a negative number, we use the absolute value of the MD to determine whether your visual field loss meets listing 102.03B. We cannot use tests that do not measure the central 30 degrees of the visual field, such as the HFA 24–2, to determine if your impairment meets or medically equals 102.03B.

e. *Other types of perimetry.* If your case record contains visual field measurements obtained using manual or automated kinetic perimetry, such as Goldmann perimetry or the HFA ''SSA Test Kinetic,'' we can generally use these results if the kinetic test was performed using a white III4e stimulus projected on a white 31.5 asb (10 cd/m^2) background. Automated kinetic perimetry, such as the HFA ''SSA Test Kinetic,'' does not detect limitations in the central visual field because testing along a meridian stops when you see the stimulus. If your visual disorder has progressed to the point at which it is likely to result in a significant limitation in the central visual field, such as a scotoma (see 102.00A6h), we will not use *automated* kinetic perimetry to determine the extent of your visual field loss. Instead, we will determine the extent of your visual field loss using automated static threshold perimetry or manual kinetic perimetry.

f. *Screening tests.* We will not use the results of visual field screening tests, such as confrontation tests, tangent screen tests, or automated static screening tests, to determine that your impairment meets or medically equals a listing, or functionally equals the listings. We can consider normal results from visual field screening tests to determine whether your visual disorder is severe when these test results are consistent with the other evidence in your case record. (See §416.924(c) of this chapter.) We will not consider normal test results to be consistent with the other evidence if the clinical findings indicate that your visual disorder has progressed to the point that it is likely to cause visual field loss, or you have a history of an operative procedure for retinal detachment.

g. *Use of corrective lenses.* You must not wear eyeglasses during visual field testing because they limit your field of vision. You may wear contact lenses to correct your visual acuity during the visual field test to obtain the most accurate visual field measurements. For this single purpose, you do not need to demonstrate that you have the ability to use the contact lenses on a sustained basis.

h. *Scotoma.* A scotoma is a field defect or non-seeing area (also referred to as a ''blind spot'') in the visual field surrounded by a normal field or seeing area. When we measure your visual field, we subtract the length of any scotoma, other than the normal blind spot, from the overall length of any diameter on which it falls.

7. *How do we determine your visual acuity efficiency, visual field efficiency, and visual efficiency?*

a. *General. Visual efficiency,* a calculated value of your remaining visual function, is the combination of your *visual acuity efficiency* and your *visual field efficiency* expressed as a percentage.

b. *Visual acuity efficiency.* Visual acuity efficiency is a percentage that corresponds to the best-corrected central visual acuity for distance in your better eye. See Table 1.

TABLE 1—VISUAL ACUITY EFFICIENCY

Snellen best-corrected central visual acuity for distance		Visual acuity efficiency (%) (102.04A)
English	Metric	
20/16	6/5	100
20/20	6/6	100
20/25	6/7.5	95
20/30	6/9	90
20/40	6/12	85
20/50	6/15	75
20/60	6/18	70
20/70	6/21	65

TABLE 1—VISUAL ACUITY EFFICIENCY—Continued

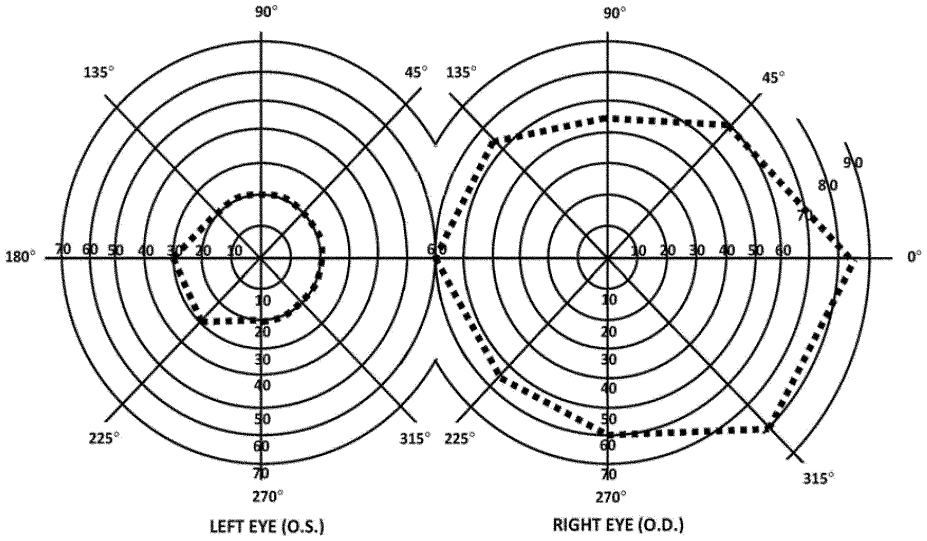
Snellen best-corrected central visual acuity for distance		Visual acuity efficiency (%) (102.04A)
English	Metric	
20/80	6/24	60
20/100	6/30	50

c. *Visual field efficiency.* Visual field efficiency is a percentage that corresponds to the visual field in your better eye. Under 102.03C, we require kinetic perimetry to determine your visual field efficiency percentage. We calculate the visual field efficiency percentage by adding the number of degrees you see along the eight principal meridians found on a visual field chart (0, 45, 90, 135, 180, 225, 270, and 315) in your better eye and dividing by 5. For example, in Figure 1:

A. The diagram of the left eye illustrates a visual field, as measured with a III4e stimulus, contracted to 30 degrees in two meridians (180 and 225) and to 20 degrees in the remaining six meridians. The visual efficiency percentage of this field is: $((2 \times 30) + (6 \times 20)) \div 5 = 36$ percent.

B. The diagram of the right eye illustrates the extent of a normal visual field as measured with a III4e stimulus. The sum of the eight principal meridians of this field is 500 degrees. The visual efficiency percentage of this field is $500 \div 5 = 100$ percent.

Figure 1:



d. *Visual efficiency.* Under 102.04A, we calculate the visual efficiency percentage by multiplying your visual acuity efficiency percentage (see 102.00A7b) by your visual field efficiency percentage (see 102.00A7c) and dividing by 100. For example, if your visual acuity efficiency percentage is 75 and your visual field efficiency percentage is 36, your visual efficiency percentage is: $(75 \times 36) \div 100 = 27$ percent.

8. *How do we determine your visual acuity impairment value, visual field impairment value, and visual impairment value?*

a. *General. Visual impairment value,* a calculated value of your loss of visual function, is the combination of your *visual acuity impairment value* and your *visual field impairment value.*

b. *Visual acuity impairment value.* Your visual acuity impairment value corresponds to the best-corrected central visual acuity for distance in your better eye. See Table 2.

TABLE 2—VISUAL ACUITY IMPAIRMENT VALUE

Snellen best-corrected central visual acuity for distance		Visual acuity impairment value (102.04B)
English	Metric	
20/16	6/5	0.00
20/20	6/6	0.00
20/25	6/7.5	0.10

TABLE 2—VISUAL ACUITY IMPAIRMENT VALUE—Continued

Snellen best-corrected central visual acuity for distance		Visual acuity impairment value (102.04B)
English	Metric	
20/30	6/9	0.18
20/40	6/12	0.30
20/50	6/15	0.40
20/60	6/18	0.48
20/70	6/21	0.54
20/80	6/24	0.60
20/100	6/30	0.70

c. *Visual field impairment value.* Your visual field impairment value corresponds to the visual field in your better eye. Using the MD from acceptable automated static threshold perimetry, we calculate the visual field impairment value by dividing the absolute value of the MD by 22. For example, if your MD on an HFA 30–2 is −16, your visual field impairment value is: $|-16| \div 22 = 0.73$.

d. *Visual impairment value.* Under 102.04B, we calculate the visual impairment value by adding your visual acuity impairment value (see 102.00A8b) and your visual field impairment value (see 102.00A8c). For example, if your visual acuity impairment value is 0.48 and your visual field impairment value is 0.73, your visual impairment value is: 0.48 + 0.73 = 1.21.

9. *What are our requirements for an acceptable perimeter?* We will use results from automated static threshold perimetry performed on a perimeter that:

a. Uses optical projection to generate the test stimuli.

b. Has an internal normative database for automatically comparing your performance with that of the general population.

c. Has a statistical analysis package that is able to calculate visual field indices, particularly mean deviation or mean defect.

d. Demonstrates the ability to correctly detect visual field loss and correctly identify normal visual fields.

e. Demonstrates good test-retest reliability.

f. Has undergone clinical validation studies by three or more independent laboratories with results published in peer-reviewed ophthalmic journals.

B. *How do we evaluate hearing loss?*

1. *What evidence do we need?*

a. We need evidence showing that you have a medically determinable impairment that causes your hearing loss and audiometric measurements of the severity of your hearing loss. We generally require both a complete otologic examination and audiometric testing to establish that you have a medically determinable impairment that causes your hearing loss. You should have this audiometric testing within 2 months of the complete otologic examination. Once we have evidence that you have a medically determinable impairment, we can use the results of later audiometric testing to assess the severity of your hearing loss without another complete otologic examination. We will consider your test scores together with any other relevant information we have about your hearing, including information from outside of the test setting.

b. The complete otologic examination must be performed by a licensed physician (medical or osteopathic doctor). It must include your medical history, your description of how your hearing loss affects you, and the physician's description of the appearance of the external ears (pinnae and external ear canals), evaluation of the tympanic membranes, and assessment of any middle ear abnormalities.

c. Audiometric testing must be performed by, or under the direct supervision of, an otolaryngologist or by an audiologist qualified to perform such tests. We consider an audiologist to be qualified if he or she is currently and fully licensed or registered as a clinical audiologist by the State or U.S. territory in which he or she practices. If no licensure or registration is available, the audiologist must be currently certified by the American Board of Audiology or have a Certificate of Clinical Competence (CCC–A) from the American Speech-Language-Hearing Association (ASHA).

2. *What audiometric testing do we need when you do not have a cochlear implant?*

a. *General.* We need either physiologic or behavioral testing (other than screening testing, see 102.00B2g) that is appropriate for your age at the time of testing. *See* 102.00B2c–102.00B2f. We will make every reasonable effort to obtain the results of physiologic testing that has been done; however, we will not purchase such testing.

b. *Testing requirements.* The testing must be conducted in accordance with the most recently published standards of the American National Standards Institute (ANSI). You must not wear hearing aids during the testing. Additionally, a person described in 102.00B1c must perform an otoscopic examination immediately before the audiometric testing. (An *otoscopic examination* provides a description of the appearance of your external ear canals and an evaluation of the tympanic membranes. In these rules, we use the term to include otoscopic examinations performed by physicians and otoscopic inspections performed by audiologists and others.) The otoscopic examination must show that there are no conditions that would prevent valid audiometric testing, such as fluid in the ear, ear infection, or obstruction in an ear canal. The person performing the test should also report on any other factors, such as your ability to maintain attention, that can affect the interpretation of the test results.

c. *Children from birth to the attainment of age 6 months.*

(i) We need physiologic testing, such as auditory brainstem response (ABR) testing.

(ii) To determine whether your hearing loss meets 102.10A, we will average your hearing thresholds at 500, 1000, 2000, and 4000 Hertz (Hz). If you do not have a response at a particular frequency, we will use a threshold of 5 decibels (dB) over the limit of the audiometer.

d. *Children from age 6 months to the attainment of age 2.*

(i) We need air conduction thresholds determined by a behavioral assessment, usually visual reinforcement audiometry (VRA). We can use ABR testing if the behavioral assessment cannot be completed or if the results are inconclusive or unreliable.

(ii) To determine whether your hearing loss meets 102.10A, we will average your hearing thresholds at 500, 1000, 2000, and 4000 Hz. If you do not have a response at a particular frequency, we will use a threshold of 5 dB over the limit of the audiometer.

(iii) For this age group, behavioral assessments are often performed in a sound field, and each ear is not tested separately. If each ear is not tested separately, we will consider the test results to represent the hearing in the better ear.

e. *Children from age 2 to the attainment of age 5.*

(i) We need air conduction thresholds determined by a behavioral assessment, such as conditioned play audiometry (CPA), tangible or visually reinforced operant conditioning audiometry (TROCA, VROCA), or VRA. If you have had ABR testing, we can use the results of that testing if the behavioral assessment cannot be completed or the results are inconclusive or unreliable.

(ii) To determine whether your hearing loss meets 102.10A, we will average your hearing thresholds at 500, 1000, 2000, and 4000 Hz. If you do not have a response at a particular frequency, we will use a threshold of 5 dB over the limit of the audiometer.

(iii) For this age group, behavioral assessments are often performed in a sound field and each ear is not tested separately. If each ear is not tested separately, we will consider the test results to represent the hearing in the better ear.

f. *Children from age 5 to the attainment of age 18.*

(i) We generally need pure tone air conduction and bone conduction testing, speech reception threshold (SRT) testing (also referred to as ''spondee threshold'' or ''ST'' testing), and word recognition testing (also referred to as ''word discrimination'' or ''speech discrimination'' testing). This testing must be conducted in a sound-treated booth or room and must be in accordance with the most recently published ANSI standards. Each ear must be tested separately.

(ii) To determine whether your hearing loss meets the air and bone conduction criterion in 102.10B1 or 102.10B3, we will average your hearing thresholds at 500, 1000, 2000, and 4000 Hz. If you do not have a response at a particular frequency, we will use a threshold of 5 dB over the limit of the audiometer.

(iii) The SRT is the minimum dB level required for you to recognize 50 percent of the words on a standard list of spondee words. (Spondee words are two-syllable words that have equal stress on each syllable.) The SRT is usually within 10 dB of the average pure tone air conduction hearing thresholds at 500, 1000, and 2000 Hz. If the SRT is not within 10 dB of the average pure tone air conduction threshold, the reason for the discrepancy must be documented. If we cannot determine that there is a medical basis for the discrepancy, we will not use the results of the testing to determine whether your hearing loss meets a listing.

(iv) Word recognition testing determines your ability to recognize an age-appropriate, standardized list of phonetically balanced monosyllabic words in the absence of any visual cues. This testing must be performed in quiet. The list may be recorded or presented live, but in either case, the words should be presented at a level of amplification that will measure your maximum ability to discriminate words, usually 35 to 40 dB above your SRT. However, the amplification level used in the testing must be medically appropriate, and you must be able to tolerate it. If you cannot be tested at 35 to 40 dB above your SRT, the person who performs the test should report your word recognition testing score at your highest comfortable level of amplification.

g. *Screening testing.* Physiologic testing, such as ABR and otoacoustic emissions (OAE), and pure tone testing can be used as hearing screening tests. We will not use these tests to determine that your hearing loss meets or medically equals a listing, or to assess functional limitations due to your hearing loss, when they are used only as screening tests. We can consider normal results from hearing screening tests to determine that your hearing loss is not ''severe'' when these test results are consistent with the other evidence in your case record. *See* §416.924(c).

3. *What audiometric testing do we need when you have a cochlear implant?*

a. If you have a cochlear implant, we will consider you to be disabled until age 5, or for 1 year after initial implantation, whichever is later.

b. After that period, we need word recognition testing performed with any age-appropriate version of the Hearing in Noise Test (HINT) or the Hearing in Noise Test for Children (HINT–C) to determine whether your

impairment meets 102.11B. This testing must be conducted in quiet in a sound field. Your implant must be functioning properly and adjusted to your normal settings. The sentences should be presented at 60 dB HL (Hearing Level) and without any visual cues.

4. *How do we evaluate your word recognition ability if you are not fluent in English?*

If you are not fluent in English, you should have word recognition testing using an appropriate word list for the language in which you are most fluent. The person conducting the test should be fluent in the language used for the test. If there is no appropriate word list or no person who is fluent in the language and qualified to perform the test, it may not be possible to measure your word recognition ability. If your word recognition ability cannot be measured, your hearing loss cannot meet 102.10B2 or 102.11B. Instead, we will consider the facts of your case to determine whether you have difficulty understanding words in the language in which you are most fluent, and if so, whether that degree of difficulty medically equals 102.10B2 or 102.11B. For example, we will consider how you interact with family members, interpreters, and other persons who speak the language in which you are most fluent.

5. *What do we mean by a marked limitation in speech or language as used in 102.10B3?*

a. We will consider you to have a marked limitation in speech if:

(i) Entire phrases or sentences in your conversation are intelligible to unfamiliar listeners at least 50 percent (half) of the time but no more than 67 percent (two-thirds) of the time on your first attempt; and

(ii) Your sound production or phonological patterns (the ways in which you combine speech sounds) are atypical for your age.

b. We will consider you to have a marked limitation in language when your current and valid test score on an appropriate comprehensive, standardized test of overall language functioning is at least two standard deviations below the mean. In addition, the evidence of your daily communication functioning must be consistent with your test score. If you are not fluent in English, it may not be possible to test your language performance. If we cannot test your language performance, your hearing loss cannot meet 102.10B3. Instead, we will consider the facts of your case to determine whether your hearing loss medically equals 102.10B3.

102.01 *Category of Impairments, Special Senses and Speech*

102.02 *Loss of central visual acuity.*

A. Remaining vision in the better eye after best correction is 20/200 or less.

OR

B. An inability to participate in visual acuity testing using Snellen methodology or other comparable testing, clinical findings that fixation and visual-following behavior are absent in the better eye, and one of the following:

1. Abnormal anatomical findings indicating a visual acuity of 20/200 or less in the better eye (such as the presence of Stage III or worse retinopathy of prematurity despite surgery, hypoplasia of the optic nerve, albinism with macular aplasia, or bilateral optic atrophy); or

2. Abnormal neuroimaging documenting damage to the cerebral cortex which would be expected to prevent the development of a visual acuity better than 20/200 in the better eye (such as neuroimaging showing bilateral encephalomyelitis or bilateral encephalomalacia); or

3. Abnormal electroretinogram documenting the presence of Leber's congenital amaurosis or achromatopsia in the better eye; or

4. An absent response to VER testing in the better eye.

102.03 *Contraction of the visual field in the better eye*, with:

A. The widest diameter subtending an angle around the point of fixation no greater than 20 degrees.

OR

B. An MD of 22 decibels or greater, determined by automated static threshold perimetry that measures the central 30 degrees of the visual field (see 102.00A6d.).

OR

C. A visual field efficiency of 20 percent or less, determined by kinetic perimetry (see 102.00A7c).

102.04 *Loss of visual efficiency, or visual impairment, in the better eye:*

A. A visual efficiency percentage of 20 or less after best correction (see 102.00A7d.).

OR

B. A visual impairment value of 1.00 or greater after best correction (see 102.00A8d).

102.10 *Hearing loss not treated with cochlear implantation.*

A. For children from birth to the attainment of age 5, an average air conduction hearing threshold of 50 decibels or greater in the better ear (see 102.00B2).

OR

B. For children from age 5 to the attainment of age 18:

1. An average air conduction hearing threshold of 70 decibels or greater in the better ear and an average bone conduction hearing threshold of 40 decibels or greater in the better ear (see 102.00B2f); or

2. A word recognition score of 40 percent or less in the better ear determined using a standardized list of phonetically balanced monosyllabic words (see 102.00B2f); or

3. An average air conduction hearing threshold of 50 decibels or greater in the better ear and a marked limitation in speech or language (see 102.00B2f and 102.00B5).

102.11 *Hearing loss treated with cochlear implantation.*

A. Consider under a disability until the attainment of age 5 or for 1 year after initial implantation, whichever is later.

OR

B. Upon the attainment of age 5 or 1 year after initial implantation, whichever is later, a word recognition score of 60 percent or less determined using the HINT or the HINT–C (*see* 102.00B3b).

103.00 RESPIRATORY SYSTEM

A. *Introduction.* The listings in this section describe impairments resulting from respiratory disorder based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of treatment prescribed by a treating source. Respiratory disorders, along with any associated impairment(s) must be established by medical evidence. Evidence must be provided in sufficient detail to permit an independent reviewer to evaluate the severity of the impairment. Reasonable efforts should be made to ensure evaluation by a program physician specializing in childhood respiratory impairments or a qualified pediatrician.

Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is such evidence, the longitudinal clinical record must include a description of the treatment prescribed by the treating source and response, in addition to information about the nature and severity of the impairment. It is important to document any prescribed treatment and response because this medical management may have improved the child's functional status. The longitudinal record should provide information regarding functional recovery, if any.

Some children will not have received ongoing treatment or have an ongoing relationship with the medical community, despite the existence of a severe impairment(s). A child who does not receive treatment may or may not be able to show an impairment that meets the criteria of these listings. Even if a child does not show that his or her impairment meets the criteria of these listings, the child may have an impairment(s) that medically or functionally equals the listings. Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the child's functioning, and the frequency, severity, and duration of symptoms. Also, the asthma listing specifically includes a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.

Evaluation should include consideration of adverse effects of respiratory impairment in all relevant body systems, and especially on the child's growth and development or mental functioning, as described under the growth impairment (100.00), neurological (111.00), and mental disorders (112.00) listings.

It must be remembered that these listings are only examples of common respiratory disorders that are severe enough to find a child disabled. When a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will make a determination whether the child's impairment(s) medically or functionally equals the listings. (See §§ 404.1526, 416.926, and 416.926a.)

B. *Documentation of Pulmonary Function Testing.* The results of spirometry that are used for adjudication, under the 103.02 A and B, 103.03, and 103.04 of these listings should be expressed in liters (L), body temperature and pressure saturated with water vapor (BTPS). The reported one-second forced expiratory volume (FEV_1) and forced vital capacity (FVC) should represent the largest of at least three satisfactory forced expiratory maneuvers. Two of the satisfactory spirograms should be reproducible for both pre-bronchodilator tests and, if indicated, post-bronchodilator tests. A value is considered reproducible if it does not differ from the largest value by more than 5 percent or 0.1 L, whichever is greater. The highest values of the FEV_1 and FVC, whether from the same or different tracings, should be used to assess the severity of the respiratory impairment. Peak flow should be achieved early in expiration, and the spirogram should have a smooth contour with gradually decreasing flow throughout expiration. The zero time for measurement of the FEV_1 and FVC, if not distinct, should be derived by linear back-extrapolation of peak flow to zero volume. A spirogram is satisfactory for measurement of the FEV_1 if the expiratory volume at the back-extrapolated zero time is less than 5 percent of the FVC or 0.1 L, whichever is greater. The spirogram is satisfactory for measurement of the FVC if maximal expiratory effort continues for at least 6 seconds, or if there is a plateau in the volume-time curve with no detectable change in expired volume (VE) during the last 2 seconds of maximal expiratory effort.

Spirometry should be repeated after administration of an aerosolized bronchodilator under supervision of the testing personnel if the pre-bronchodilator FEV_1 value is less than the appropriate reference value in table I or III, as appropriate. If a bronchodilator is not administered, the reason should be clearly stated in the report. Pulmonary function studies should not be performed unless the clinical status is stable (e.g., the child is not having an asthmatic attack or suffering from an acute respiratory

infection or other chronic illness). Wheezing is common in asthma, chronic bronchitis, or chronic obstructive pulmonary disease and does not preclude testing. Pulmonary function studies performed to assess airflow obstruction without testing after bronchodilators cannot be used to assess levels of impairment in the range that prevents a child from performing age-appropriate activities, unless the use of bronchodilators is contraindicated. Post-bronchodilator testing should be performed 10 minutes after bronchodilator administration. The dose and name of the bronchodilator administered should be specified. The values in 103.02 and 103.04 must only be used as criteria for the level of ventilatory impairment that exists during the child's most stable state of health (*i.e.*, any period in time except during or shortly after an exacerbation).

The appropriately labeled spirometric tracing, showing the child's name, date of testing, distance per second on the abscissa and distance per liter (L) on the ordinate, must be incorporated into the file. The manufacturer and model number of the device used to measure and record the spirogram should be stated. The testing device must accurately measure both time and volume, the latter to within 1 percent of a 3 L calibrating volume. If the spirogram was generated by any means other than direct pen linkage to a mechanical displacement-type spirometer, the testing device must have had a recorded calibration performed previously on the day of the spirometric measurement.

If the spirometer directly measures flow, and volume is derived by electronic integration, the linearity of the device must be documented by recording volume calibrations at three different flow rates of approximately 30 L/min (3 L/6 sec), 60 L/min (3 L/3 sec), and 180 L/min (3 L/sec). The volume calibrations should agree to within 1 percent of a 3 L calibrating volume. The proximity of the flow sensor to the child should be noted, and it should be stated whether or not a BTPS correction factor was used for the calibration recordings and for the child's actual spirograms.

The spirogram must be recorded at a speed of at least 20 mm/sec and the recording device must provide a volume excursion of at least 10 mm/L. If reproductions of the original spirometric tracings are submitted, they must be legible and have a time scale of at least 20 mm/sec and a volume scale of at least 10 mm/L to permit independent measurements. Calculation of FEV_1 from a flow volume tracing is not acceptable, *i.e.*, the spirogram and calibrations must be presented in a volume-time format at a speed of at least 20 mm/sec and a volume excursion of at least 10 mm/L to permit independent evaluation.

A statement should be made in the pulmonary function test report of the child's ability to understand directions, as well as his or her efforts and cooperation in performing the pulmonary function tests.

Purchase of a pulmonary function test is appropriate only when the child is capable of performing reproducible forced expiratory maneuvers. This capability usually occurs around age 6. Purchase of a pulmonary function test may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

The pulmonary function tables in 103.02 and 103.04 are based on measurement of standing height without shoes. If a child has marked spinal deformities (e.g., kyphoscoliosis), the measured span between the fingertips with the upper extremities abducted 90 degrees should be substituted for height when this measurement is greater than the standing height without shoes.

C. *Documentation of chronic impairment of gas exchange.*

1. *Arterial blood gas studies (ABGS).* An ABGS performed at rest (while breathing room air, awake and sitting or standing) should be analyzed in a laboratory certified by a State or Federal agency. If the laboratory is not certified, it must submit evidence of participation in a national proficiency testing program as well as acceptable quality control at the time of testing. The report should include the altitude of the facility and the barometric pressure on the date of analysis.

Purchase of resting ABGS may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided. Before purchasing resting ABGS, a program physician, preferably one experienced in the care of children with pulmonary disease, must review the clinical and laboratory data short of this procedure, including spirometry, to determine whether obtaining the test would present a significant risk to the child.

2. *Oximetry.* Pulse oximetry may be substituted for arterial blood gases in children under 12 years of age. The oximetry unit should employ the basic technology of spectrophotometric plethysmography as described in Taylor, M.B., and Whitwain, J.G., ''Current Status of Pulse Oximetry,'' ''Anesthesia,'' Vol. 41, No. 9, pp. 943–949, 1986. The unit should provide a visual display of the pulse signal and the corresponding oxygen saturation. A hard copy of the readings (heart rate and saturation) should be provided. Readings should be obtained for a minimum of 5 minutes. The written report should describe patient activity during the recording, *i.e.*, sleep rate, feeding, or exercise. Correlation between the actual heart rate determined by a trained observer and that displayed by the oximeter should be provided. A statement should be made in the

report of the child's effort and cooperation during the test.

Purchase of oximetry may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

D. *Cystic fibrosis* is a disorder that affects either the respiratory or digestive body systems or both and may impact on a child's growth and development. It is responsible for a wide and variable spectrum of clinical manifestations and complications. Confirmation of the diagnosis is based upon an elevated sweat sodium concentration or chloride concentration accompanied by one or more of the following: the presence of chronic obstructive pulmonary disease, insufficiency of exocrine pancreatic function, meconium ileus, or a positive family history. The quantitative pilocarpine iontophoresis procedure for collection of sweat content must be utilized. Two methods are acceptable: the ''Procedure for the Quantitative Iontophoretic Sweat Test for Cystic Fibrosis,'' published by the Cystic Fibrosis Foundation and contained in, ''A Test for Concentration of Electrolytes in Sweat in Cystic Fibrosis of the Pancreas Utilizing Pilocarpine Iontophoresis,'' Gibson, I.E., and Cooke, R.E., ''Pediatrics,'' Vol 23: 545, 1959; or the ''Wescor Macroduct System.'' To establish the diagnosis of cystic fibrosis, the sweat sodium or chloride content must be analyzed quantitatively using an acceptable laboratory technique. Another diagnostic test is the ''CF gene mutation analysis'' for homozygosity of the cystic fibrosis gene. The pulmonary manifestations of this disorder should be evaluated under 103.04. The nonpulmonary aspects of cystic fibrosis should be evaluated under the listings for the digestive system (105.00) or growth impairments (100.00). Because cystic fibrosis may involve the respiratory and digestive body systems, as well as impact on a child's growth and development, the combined effects of this involvement must be considered in case adjudication.

Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

E. *Bronchopulmonary dysplasia (BPD).* Bronchopulmonary dysplasia is a form of chronic obstructive pulmonary disease that arises as a consequence of acute lung injury in the newborn period and treatment of hyaline membrane disease, meconium aspiration, neonatal pneumonia and apnea of prematurity. The diagnosis is established by the requirement for continuous or nocturnal supplemental oxygen for more than 30 days, in association with characteristic changes on medically acceptable imaging and clinical signs of respiratory dysfunction, including retractions, rales, wheezing, and tachypnea.

F. *How do we evaluate growth failure due to any chronic respiratory disorder?*

1. To evaluate growth failure due to any chronic respiratory disorder, we require documentation of the oxygen supplementation described in 103.06A and the growth measurements in 103.06B within the same consecutive 12-month period. The dates of oxygen supplementation may be different from the dates of growth measurements.

2. Under 103.06B, we use the appropriate table(s) under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile.

a. For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

b. For children age 2 to attainment of age 18, we use the body mass index (BMI)-for-age table corresponding to the child's gender (Table III or Table IV).

c. BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

103.01 Category of Impairments, Respiratory System

103.02 *Chronic pulmonary insufficiency.* With:

A. Chronic obstructive pulmonary disease, due to any cause, with the FEV_1 equal to or less than the value specified in table I corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

TABLE I

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
119 or less	46 or less	0.65
120–129	47–50	0.75
130–139	51–54	0.95
140–149	55–58	1.15
150–159	59–62	1.35
160–164	63–64	1.45
165–169	65–66	1.55
170 or more	67 or more	1.65

Or

B. Chronic restrictive ventilatory disease, due to any cause, with the FVC equal to or less than the value specified in table II corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

TABLE II

Height without shoes (centimeters)	Height without shoes (inches)	FVC equal to or less than (L, BTPS)
119 or less	46 or less	0.65
120–129	47–50	0.85
130–139	51–54	1.05
140–149	55–58	1.25
150–159	59–62	1.45
160–164	63–64	1.65
165–169	65–66	1.75
170 or more	67 or more	2.05

Or

C. Frequent need for:

1. Mechanical ventilation; or

2. Nocturnal supplemental oxygen as required by persistent or recurrent episodes of hypoxemia;

Or

D. The presence of a tracheostomy in a child under 3 years of age;

Or

E. Bronchopulmonary dysplasia characterized by two of the following:

1. Prolonged expirations; or

2. Intermittent wheezing or increased respiratory effort as evidenced by retractions, flaring and tachypnea; or

3. Hyperinflation and scarring on a chest radiograph or other appropriate imaging techniques; or

4. Bronchodilator or diuretic dependency; or

5. A frequent requirement for nocturnal supplemental oxygen; or

6. Weight disturbance with:

a. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from established growth curve (on standard growth charts) which persists for 2 months or longer; or

b. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall to below the third percentile from established growth curve (on standard growth charts) which persists for 2 months or longer;

Or

F. Two required hospital admissions (each longer than 24 hours) within a 6-month period for recurrent lower respiratory tract infections or acute respiratory distress associated with:

1. Chronic wheezing or chronic respiratory distress; or

2. Weight disturbance with:

a. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from established growth curve (on standard growth charts) which persists for 2 months or longer; or

b. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall to below the third percentile from established growth curve (on standard growth charts) which persists for 2 months or longer;

Or

G. Chronic hypoventilation ($PaCO_2$ greater than 45 mm Hg) or chronic cor pulmonale as described under the appropriate criteria in 104.02;

Or

H. Growth impairment as described under the criteria in 100.00.

103.03 *Asthma*. With:

A. FEV_1 equal to or less than the value specified in table I of 103.02A;

Or

B. Attacks (as defined in 3.00C), in spite of prescribed treatment and requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for control of asthma counts as two attacks, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of attacks;

Or

C. Persistent low-grade wheezing between acute attacks or absence of extended symptom-free periods requiring daytime and nocturnal use of sympathomimetic bronchodilators with one of the following:

1. Persistent prolonged expiration with radiographic or other appropriate imaging techniques evidence of pulmonary hyperinflation or peribronchial disease; or

2. Short courses of corticosteroids that average more than 5 days per month for at least 3 months during a 12-month period;

Or

D. Growth impairment as described under the criteria in 100.00.

103.04 *Cystic fibrosis*. With:

A. An FEV_1 equal to or less than the appropriate value specified in table III corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Or

B. For children in whom pulmonary function testing cannot be performed, the presence of two of the following:

1. History of dyspnea on exertion or accumulation of secretions as manifested by repetitive coughing or cyanosis; or

2. Persistent bilateral rales and rhonchi or substantial reduction of breath sounds related to mucous plugging of the trachea or bronchi; or

3. Appropriate medically acceptable imaging evidence of extensive disease, such as

thickening of the proximal bronchial airways or persistence of bilateral peribronchial infiltrates;

Or

C. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased bacterial infection occurring at least once every 6 months and requiring intravenous or nebulization antimicrobial treatment;

Or

D. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;

Or

E. Growth impairment as described under the criteria in 100.00.

TABLE III

[Applicable only for evaluation under 103.04A—cystic fibrosis]

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
119 or less	46 or less	0.75
120–129	47–50	0.85
130–139	51–54	1.05
140–149	55–58	1.35
150–159	59–62	1.55
160–164	63–64	1.85
165–169	65–66	2.05
170 or more	67 or more	2.25

103.05 *Lung transplant.* Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment(s).

103.06 *Growth failure due to any chronic respiratory disorder* (see 103.00F), documented by:

A. Hypoxemia with the need for at least 1.0 L/min of oxygen supplementation for at least 4 hours per day and for at least 90 consecutive days.

AND

B. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2,* three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or

2. *For children age 2 to attainment of age 18,* three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.

104.00 CARDIOVASCULAR SYSTEM

A. General

1. *What do we mean by a cardiovascular impairment?*

a. We mean any disorder that affects the proper functioning of the heart or the circulatory system (that is, arteries, veins, capillaries, and the lymphatic drainage). The disorder can be congenital or acquired.

b. Cardiovascular impairment results from one or more of four consequences of heart disease:

(i) Chronic heart failure or ventricular dysfunction.

(ii) Discomfort or pain due to myocardial ischemia, with or without necrosis of heart muscle.

(iii) Syncope, or near syncope, due to inadequate cerebral perfusion from any cardiac cause, such as obstruction of flow or disturbance in rhythm or conduction resulting in inadequate cardiac output.

(iv) Central cyanosis due to right-to-left shunt, reduced oxygen concentration in the arterial blood, or pulmonary vascular disease.

c. Disorders of the veins or arteries (for example, obstruction, rupture, or aneurysm) may cause impairments of the lower extremities (peripheral vascular disease), the central nervous system, the eyes, the kidneys, and other organs. We will evaluate peripheral vascular disease under 4.11 or 4.12 in part A, and impairments of another body system(s) under the listings for that body system(s).

2. *What do we consider in evaluating cardiovascular impairments?* The listings in this section describe cardiovascular impairments based on symptoms, signs, laboratory findings, response to a regimen of prescribed treatment, and functional limitations.

3. *What do the following terms or phrases mean in these listings?*

a. *Medical consultant* is an individual defined in §§404.1616(a) and 416.1016(a). This term does not include medical sources who provide consultative examinations for us. We use the abbreviation ''MC'' throughout this section to designate a medical consultant.

b. *Persistent* means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been present, or is expected to be present, for a continuous period of at least 12 months, such that a pattern of continuing severity is established.

c. *Recurrent* means that the longitudinal clinical record shows that, within a consecutive 12-month period, the finding(s) occurs at

least three times, with intervening periods of improvement of sufficient duration that it is clear that separate events are involved.

d. *Appropriate medically acceptable imaging* means that the technique used is the proper one to evaluate and diagnose the impairment and is commonly recognized as accurate for assessing the cited finding.

e. *A consecutive 12-month period* means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with an application or continuing disability review.

f. *Currently present* means that the finding is present at the time of adjudication.

g. *Uncontrolled* means the impairment does not respond adequately to standard prescribed medical treatment.

B. Documenting Cardiovascular Impairment

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular impairment. A longitudinal clinical record covering a period of not less than 3 months of observations and treatment is usually necessary, unless we can make a determination or decision based on the current evidence.

2. *Why is a longitudinal clinical record important?* We will usually need a longitudinal clinical record to assess the severity and expected duration of your impairment(s). If you have a listing-level impairment, you probably will have received medically prescribed treatment. Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your treating or other medical source. It should also include your response to this medical management, as well as information about the nature and severity of your impairment. The record will provide us with information on your functional status over an extended period of time and show whether your ability to function is improving, worsening, or unchanging.

3. *What if you have not received ongoing medical treatment?*

a. You may not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In this situation, we will base our evaluation on the current objective medical evidence and the other evidence we have. If you do not receive treatment, you cannot show an impairment that meets the criteria of these listings. However, we may find you disabled because you have another impairment(s) that in combination with your cardiovascular impairment medically equals the severity of a listed impairment or that functionally equals the listings.

b. Unless we can decide your claim favorably on the basis of the current evidence, a longitudinal record is still important. In rare instances where there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the severity and duration of your impairment.

4. *When will we wait before we ask for more evidence?*

a. We will wait when we have information showing that your impairment is not yet stable and the expected change in your impairment might affect our determination or decision. In these situations, we need to wait to properly evaluate the severity and duration of your impairment during a stable period. Examples of when we might wait are:

(i) If you have had a recent acute event; for example, acute rheumatic fever.

(ii) If you have recently had a corrective cardiac procedure; for example, open-heart surgery.

(iii) If you have started new drug therapy and your response to this treatment has not yet been established; for example, beta-blocker therapy for dilated congestive cardiomyopathy.

b. In these situations, we will obtain more evidence 3 months following the event before we evaluate your impairment. However, we will not wait if we have enough information to make a determination or decision based on all of the relevant evidence in your case.

5. *Will we purchase any studies?* In appropriate situations, we will purchase studies necessary to substantiate the diagnosis or to document the severity of your impairment, generally after we have evaluated the medical and other evidence we already have. We will not purchase studies involving exercise testing if there is significant risk involved or if there is another medical reason not to perform the test. We will follow sections 4.00C6, 4.00C7, 4.00C8, and 104.00B7 when we decide whether to purchase exercise testing. We will make a reasonable effort to obtain any additional studies from a qualified medical source in an office or center experienced in pediatric cardiac assessment. (See §416.919g.)

6. *What studies will we not purchase?* We will not purchase any studies involving cardiac catheterization, such as coronary angiography, arteriograms, or electrophysiological studies. However, if the results of catheterization are part of the existing evidence we have, we will consider them together with the other relevant evidence. See 4.00C15a in part A.

7. *Will we use exercise tolerance tests (ETTs) for evaluating children with cardiovascular impairment?*

a. ETTs, though increasingly used, are still less frequently indicated in children than in adults, and can rarely be performed successfully by children under 6 years of age. An ETT may be of value in the assessment of

some arrhythmias, in the assessment of the severity of chronic heart failure, and in the assessment of recovery of function following cardiac surgery or other treatment.

b. We will purchase an ETT in a childhood claim only if we cannot make a determination or decision based on the evidence we have and an MC, preferably one with experience in the care of children with cardiovascular impairments, has determined that an ETT is needed to evaluate your impairment. We will not purchase an ETT if you are less than 6 years of age. If we do purchase an ETT for a child age 12 or younger, it must be performed by a qualified medical source in a specialty center for pediatric cardiology or other facility qualified to perform exercise tests of children.

c. For full details on ETT requirements and usage, see 4.00C in part A.

C. Evaluating Chronic Heart Failure

1. *What is chronic heart failure (CHF)?*

a. *CHF* is the inability of the heart to pump enough oxygenated blood to body tissues. This syndrome is characterized by symptoms and signs of pulmonary or systemic congestion (fluid retention) or limited cardiac output. Certain laboratory findings of cardiac functional and structural abnormality support the diagnosis of CHF.

b. CHF is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, or rheumatic, congenital, or other heart disease. However, if the CHF is the result of primary pulmonary hypertension secondary to disease of the lung (cor pulmonale), we will evaluate your impairment using 3.09 in the respiratory system listings in part A.

2. *What evidence of CHF do we need?*

a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization.

(i) Cardiomegaly is present when:

(*A*) Left ventricular diastolic dimension or systolic dimension is greater than 2 standard deviations above the mean for the child's body surface area;

(*B*) Left ventricular mass is greater than 2 standard deviations above the mean for the child's body surface area; or

(*C*) Chest x-ray (6 foot PA film) is indicative of cardiomegaly if the cardiothoracic ratio is over 60 percent at 1 year of age or less, or 55 percent or greater at more than 1 year of age.

(ii) Ventricular dysfunction is present when indices of left ventricular function, such as fractional shortening or ejection fraction (the percentage of the blood in the ventricle actually pumped out with each contraction), are greater than 2 standard deviations below the mean for the child's age. (Fractional shortening, also called shortening fraction, reflects the left ventricular systolic function in the absence of segmental wall motion abnormalities and has a linear correlation with ejection fraction. In children, fractional shortening is more commonly used than ejection fraction.)

(iii) However, these measurements alone do not reflect your functional capacity, which we evaluate by considering all of the relevant evidence.

(iv) Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema. These findings need not be present on each report, since CHF may be controlled by prescribed treatment.

b. To establish that you have *chronic* heart failure, we require that your medical history and physical examination describe characteristic symptoms and signs of pulmonary or systemic congestion or of limited cardiac output associated with abnormal findings on appropriate medically acceptable imaging. When a remediable factor, such as arrhythmia, triggers an acute episode of heart failure, you may experience restored cardiac function, and a chronic impairment may not be present.

(i) Symptoms of congestion or of limited cardiac output include easy fatigue, weakness, shortness of breath (dyspnea), cough, or chest discomfort at rest or with activity. Children with CHF may also experience shortness of breath on lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). They may also experience cardiac arrhythmias resulting in palpitations, lightheadedness, or fainting. Fatigue or exercise intolerance in an infant may be manifested by prolonged feeding time, often associated with excessive respiratory effort and sweating.

(ii) During infancy, other manifestations of chronic heart failure may include repeated lower respiratory tract infections.

(iii) Signs of congestion may include hepatomegaly, ascites, increased jugular venous distention or pressure, rales, peripheral edema, rapid shallow breathing (tachypnea), or rapid weight gain. However, these signs need not be found on all examinations because fluid retention may be controlled by prescribed treatment.

3. *How do we evaluate growth failure due to CHF?*

a. To evaluate growth failure due to CHF, we require documentation of the clinical findings of CHF described in 104.00C2 and the growth measurements in 104.02C within the same consecutive 12-month period. The dates of clinical findings may be different from the dates of growth measurements.

b. Under 104.02C, we use the appropriate table(s) under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile.

(i) For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

(ii) For children age 2 to attainment of age 18, we use the body mass index (BMI)-for-age table corresponding to the child's gender (Table III or Table IV).

(iii) BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

D. Evaluating Congenital Heart Disease

1. *What is congenital heart disease?* Congenital heart disease is any abnormality of the heart or the major blood vessels that is present at birth. Examples include:

a. *Abnormalities of cardiac septation,* including ventricular septal defect or atrioventricular canal;

b. *Abnormalities resulting in cyanotic heart disease,* including tetralogy of Fallot or transposition of the great arteries;

c. *Valvular defects or obstructions to ventricular outflow,* including pulmonary or aortic stenosis or coarctation of the aorta; and

d. *Major abnormalities of ventricular development,* including hypoplastic left heart syndrome or pulmonary tricuspid atresia with hypoplastic right ventricle.

2. *How will we evaluate symptomatic congenital heart disease?*

a. Because of improved treatment methods, more children with congenital heart disease are living longer. Although some types of congenital heart disease may be corrected by surgery, many children with treated congenital heart disease continue to have problems throughout their lives (symptomatic congenital heart disease). If you have congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias, we will evaluate your impairment under 104.02 or 104.05. Otherwise, we will evaluate your impairment under 104.06.

b. For 104.06A2, we will accept pulse oximetry measurements instead of arterial O_2, but the arterial O_2 values are preferred, if available.

c. For 104.06D, examples of impairments that in most instances will require life-saving surgery or a combination of surgery and other major interventional procedures (for example, multiple ''balloon'' catheter procedures) before age 1 include, but are not limited to, the following:

(i) Hypoplastic left heart syndrome,

(ii) Critical aortic stenosis with neonatal heart failure,

(iii) Critical coarctation of the aorta, with or without associated anomalies,

(iv) Complete atrioventricular canal defects,

(v) Transposition of the great arteries,

(vi) Tetralogy of Fallot,

(vii) Pulmonary atresia with intact ventricular septum,

(viii) Single ventricle,

(ix) Tricuspid atresia, and

(x) Multiple ventricular septal defects.

E. Evaluating Arrhythmias

1. *What is an arrhythmia?* An *arrhythmia* is a change in the regular beat of the heart. Your heart may seem to skip a beat or beat irregularly, very quickly (tachycardia), or very slowly (bradycardia).

2. *What are the different types of arrhythmias?*

a. There are many types of arrhythmias. Arrhythmias are identified by where they occur in the heart (atria or ventricles) and by what happens to the heart's rhythm when they occur.

b. Arrhythmias arising in the cardiac atria (upper chambers of the heart) are called atrial or supraventricular arrhythmias. Ventricular arrhythmias begin in the ventricles (lower chambers). In general, ventricular arrhythmias caused by heart disease are the most serious.

3. *How do we evaluate arrhythmias using 104.05?*

a. We will use 104.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator and you have uncontrolled recurrent episodes of syncope or near syncope. If your arrhythmias are controlled, we will evaluate your underlying heart disease using the appropriate listing. For other considerations when we evaluate arrhythmias in the presence of an implanted cardiac defibrillator, see 104.00E4.

b. We consider *near syncope* to be a period of altered consciousness, since syncope is a loss of consciousness or a faint. It is not merely a feeling of light-headedness, momentary weakness, or dizziness.

c. For purposes of 104.05, there must be a documented association between the syncope or near syncope and the recurrent arrhythmia. The recurrent arrhythmia, not some other cardiac or non-cardiac disorder, must be established as the cause of the associated symptom. This documentation of the association between the symptoms and the arrhythmia may come from the usual diagnostic methods, including Holter monitoring (also called ambulatory electrocardiography) and tilt-table testing with a concurrent ECG. Although an arrhythmia may be a coincidental finding on an ETT, we will not purchase an ETT to document the presence of a cardiac arrhythmia.

4. *What will we consider when you have an implanted cardiac defibrillator and you do not*

have arrhythmias that meet the requirements of 104.05?

a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in children who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group of children at risk for sudden cardiac death consists of children with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in children with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator is a unique form of treatment; it rescues a child from what may have been cardiac arrest. However, as a consequence of the shock(s), children may experience psychological distress, which we may evaluate under the mental disorders listings in 112.00ff.

b. Most implantable cardiac defibrillators have rhythm-correcting and pacemaker capabilities. In some children, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. (The shock is like being kicked in the chest.) Implanted cardiac defibrillators may deliver inappropriate shocks, often repeatedly, in response to benign arrhythmias or electrical malfunction. Also, exposure to strong electrical or magnetic fields, such as from MRI (magnetic resonance imaging), can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. We must consider the frequency of, and the reason(s) for, the shocks when evaluating the severity and duration of your impairment.

c. In general, the exercise limitations imposed on children with an implanted cardiac defibrillator are those dictated by the underlying heart impairment. However, the exercise limitations may be greater when the implanted cardiac defibrillator delivers an inappropriate shock in response to the increase in heart rate with exercise, or when there is exercise-induced ventricular arrhythmia.

F. Evaluating Other Cardiovascular Impairments

1. *What is ischemic heart disease (IHD) and how will we evaluate it in children? IHD* results when one or more of your coronary arteries is narrowed or obstructed or, in rare situations, constricted due to vasospasm, interfering with the normal flow of blood to your heart muscle (ischemia). The obstruction may be the result of an embolus, a thrombus, or plaque. When heart muscle tissue dies as a result of the reduced blood supply, it is called a myocardial infarction (heart attack). Ischemia is rare in children, but when it occurs, its effects on children are the same as on adults. If you have IHD, we will evaluate it under 4.00E and 4.04 in part A.

2. *How will we evaluate hypertension?* Because *hypertension* (high blood pressure) generally causes disability through its effects on other body systems, we will evaluate it by reference to the specific body system(s) affected (heart, brain, kidneys, or eyes) when we consider its effects under the listings. We will also consider any limitations imposed by your hypertension when we consider whether you have an impairment that functionally equals the listings.

3. *What is cardiomyopathy and how will we evaluate it? Cardiomyopathy* is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy). There are various types of cardiomyopathy, which fall into two major categories: *Ischemic* and *nonischemic* cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: Dilated, hypertrophic, and restrictive. We will evaluate cardiomyopathy under 4.04 in part A, 104.02, 104.05, or 111.06, depending on its effects on you.

4. *How will we evaluate valvular heart disease?* We will evaluate valvular heart disease under the listing appropriate for its effect on you. Thus, we may use 4.04 in part A, 104.02, 104.05, 104.06, or an appropriate neurological listing in 111.00ff.

5. *What do we consider when we evaluate heart transplant recipients?*

a. After your heart transplant, we will consider you disabled for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year.

b. However, heart transplant patients generally meet our definition of disability before they undergo transplantation. We will determine the onset of your disability based on the facts in your case.

c. We will not assume that you became disabled when your name was placed on a transplant waiting list. This is because you may be placed on a waiting list soon after diagnosis of the cardiac disorder that may eventually require a transplant. Physicians recognize that candidates for transplantation often have to wait months or even years before a suitable donor heart is found, so they place their patients on the list as soon as permitted.

d. When we do a continuing disability review to determine whether you are still disabled, we will evaluate your residual impairment(s), as shown by symptoms, signs, and laboratory findings, including any side effects of medication. We will consider any remaining symptoms, signs, and laboratory findings indicative of cardiac dysfunction in

deciding whether medical improvement (as defined in §416.994a) has occurred.

6. *How will we evaluate chronic rheumatic fever or rheumatic heart disease?* The diagnosis should be made in accordance with the current revised Jones criteria for guidance in the diagnosis of rheumatic fever. We will evaluate persistence of rheumatic fever activity under 104.13. If you have evidence of chronic heart failure or recurrent arrhythmias associated with rheumatic heart disease, we will use 104.02 or 104.05.

7. *What is hyperlipidemia and how will we evaluate it? Hyperlipidemia* is the general term for an elevation of any or all of the lipids (fats or cholesterol) in the blood; for example, hypertriglyceridemia, hypercholesterolemia, and hyperlipoproteinemia. These disorders of lipoprotein metabolism and transport can cause defects throughout the body. The effects most likely to interfere with function are those produced by atherosclerosis (narrowing of the arteries) and coronary artery disease. We will evaluate your lipoprotein disorder by considering its effects on you.

8. *How will we evaluate Kawasaki disease?* We will evaluate Kawasaki disease under the listing appropriate to its effects on you, which may include major coronary artery aneurysm or heart failure. A major coronary artery aneurysm may cause ischemia or arrhythmia, which we will evaluate under 4.04 in part A or 104.05. We will evaluate chronic heart failure under 104.02.

9. *What is lymphedema and how will we evaluate it?*

a. *Lymphedema* is edema of the extremities due to a disorder of the lymphatic circulation; at its worst, it is called elephantiasis. Primary lymphedema is caused by abnormal development of lymph vessels and may be present at birth (congenital lymphedema), but more often develops during the teens (lymphedema praecox). Secondary lymphedema is due to obstruction or destruction of normal lymphatic channels due to tumor, surgery, repeated infections, or parasitic infection such as filariasis. Lymphedema most commonly affects one extremity.

b. Lymphedema does not meet the requirements of 4.11 in part A, although it may medically equal the severity of that listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular listing, such as 4.11, or a musculoskeletal listing, such as 101.02A or 101.03. If no listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we consider whether you have an impairment that functionally equals the listings.

10. *What is Marfan syndrome and how will we evaluate it?*

a. Marfan syndrome is a genetic connective tissue disorder that affects multiple body systems, including the skeleton, eyes, heart, blood vessels, nervous system, skin, and lungs. There is no specific laboratory test to diagnose Marfan syndrome. The diagnosis is generally made by medical history, including family history, physical examination, including an evaluation of the ratio of arm/leg size to trunk size, a slit lamp eye examination, and a heart test(s), such as an echocardiogram. In some cases, a genetic analysis may be useful, but such analyses may not provide any additional helpful information.

b. The effects of Marfan syndrome can range from mild to severe. In most cases, the disorder progresses as you age. Most individuals with Marfan syndrome have abnormalities associated with the heart and blood vessels. Your heart's mitral valve may leak, causing a heart murmur. Small leaks may not cause symptoms, but larger ones may cause shortness of breath, fatigue, and palpitations. Another effect is that the wall of the aorta may be weakened and stretch (aortic dilation). This aortic dilation may tear, dissect, or rupture, causing serious heart problems or sometimes sudden death. We will evaluate the manifestations of your Marfan syndrome under the appropriate body system criteria, such as 4.10 in part A, or if necessary consider the functional limitations imposed by your impairment.

G. Other Evaluation Issues

1. *What effect does obesity have on the cardiovascular system and how will we evaluate it?* Obesity is a medically determinable impairment that is often associated with disorders of the cardiovascular system. Disturbance of this system can be a major cause of disability in children with obesity. Obesity may affect the cardiovascular system because of the increased workload the additional body mass places on the heart. Obesity may make it harder for the chest and lungs to expand. This can mean that the respiratory system must work harder to provide needed oxygen. This in turn would make the heart work harder to pump blood to carry oxygen to the body. Because the body would be working harder at rest, its ability to perform additional work would be less than would otherwise be expected. Thus, the combined effects of obesity with cardiovascular impairments can be greater than the effects of each of the impairments considered separately. We must consider any additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular impairment or a listing-level cardiovascular impairment (or a combination of impairments that medically equals a listing), and when we determine whether your impairment(s) functionally equals the listings.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, we may need to defer evaluation of the impairment for a period of up to 3 months from the date treatment began to permit consideration of treatment effects, unless we can make a determination or decision using the evidence we have. See 104.00B4.

3. *How do we evaluate impairments that do not meet one of the cardiovascular listings?*

a. These listings are only examples of common cardiovascular disorders that we consider severe enough to result in marked and severe functional limitations. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

b. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926.) If you have a severe impairment(s) that does not meet or medically equal the criteria of a listing, we will consider whether it functionally equals the listings. (See §416.926a.) When we decide whether you continue to be disabled, we use the rules in §416.994a.

104.01 Category of Impairments, Cardiovascular System

104.02. *Chronic heart failure* while on a regimen of prescribed treatment, with symptoms and signs described in 104.00C2, and with one of the following:

A. Persistent tachycardia at rest (see Table I);

OR

B. Persistent tachypnea at rest (see Table II) or markedly decreased exercise tolerance (see 104.00C2b);

OR

C. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2,* three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or

2. *For children age 2 to attainment of age 18,* three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.

104.05 *Recurrent arrhythmias,* not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled (see 104.00A3g), recurrent (see 104.00A3c) episodes of cardiac syncope or near syncope (see 104.00E3b), despite prescribed treatment (see 104.00B3 if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography, or by other appropriate medically acceptable testing, coincident with the occurrence of syncope or near syncope (see 104.00E3c).

104.06 *Congenital heart disease,* documented by appropriate medically acceptable imaging (see 104.00A3d) or cardiac catheterization, with one of the following:

A. Cyanotic heart disease, with persistent, chronic hypoxemia as manifested by:

1. Hematocrit of 55 percent or greater on two evaluations 3 months or more apart within a consecutive 12-month period (see 104.00A3e); or

2. Arterial O_2 saturation of less than 90 percent in room air, or resting arterial PO_2 of 60 Torr or less; or

3. Hypercyanotic spells, syncope, characteristic squatting, or other incapacitating symptoms directly related to documented cyanotic heart disease; or

4. Exercise intolerance with increased hypoxemia on exertion.

OR

B. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure.

OR

C. Symptomatic acyanotic heart disease, with ventricular dysfunction interfering very seriously with the ability to independently initiate, sustain, or complete activities.

OR

D. For infants under 12 months of age at the time of filing, with life-threatening congenital heart impairment that will require or already has required surgical treatment in the first year of life, and the impairment is expected to be disabling (because of residual impairment following surgery, or the recovery time required, or both) until the attainment of at least 1 year of age, consider the infant to be under disability until the attainment of at least age 1; thereafter, evaluate impairment severity with reference to the appropriate listing.

104.09 *Heart transplant*. Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under the appropriate listing.

104.13 *Rheumatic heart disease*, with persistence of rheumatic fever activity manifested by significant murmurs(s), cardiac enlargement or ventricular dysfunction (see 104.00C2a), and other associated abnormal laboratory findings; for example, an elevated sedimentation rate or ECG findings, for 6 months or more in a consecutive 12-month period (see 104.00A3e). Consider under a disability for 18 months from the established onset of impairment, then evaluate any residual impairment(s).

105.00 Digestive System

A. *What kinds of disorders do we consider in the digestive system?* Disorders of the digestive system include gastrointestinal hemorrhage, hepatic (liver) dysfunction, inflammatory bowel disease, short bowel syndrome, and malnutrition. They may also lead to complications, such as obstruction, or be accompanied by manifestations in other body systems. Congenital abnormalities involving the organs of the gastrointestinal system may interfere with the ability to maintain adequate nutrition, growth, and development.

B. *What documentation do we need?* We need a record of your medical evidence, including clinical and laboratory findings. The documentation should include appropriate medically acceptable imaging studies and reports of endoscopy, operations, and pathology, as appropriate to each listing, to document the severity and duration of your digestive disorder. We may also need assessments of your growth and development. Medically acceptable imaging includes, but is not limited to, x-ray imaging, sonography, computerized axial tomography (CAT scan), magnetic resonance imaging (MRI), and radionuclide scans. *Appropriate* means that the technique used is the proper one to support the evaluation and diagnosis of the disorder. The findings required by these listings must occur within the period we are considering in connection with your application or continuing disability review.

C. *How do we consider the effects of treatment?*

1. Digestive disorders frequently respond to medical or surgical treatment; therefore, we generally consider the severity and duration of these disorders within the context of the prescribed treatment.

2. We assess the effects of treatment, including medication, therapy, surgery, or any other form of treatment you receive, by determining if there are improvements in the symptoms, signs, and laboratory findings of your digestive disorder. We also assess any side effects of your treatment that may further limit your functioning.

3. To assess the effects of your treatment, we may need information about:

a. The treatment you have been prescribed (for example, the type of medication or therapy, or your use of parenteral (intravenous) nutrition or supplemental enteral nutrition via a gastrostomy);

b. The dosage, method, and frequency of administration;

c. Your response to the treatment;

d. Any adverse effects of such treatment; and

e. The expected duration of the treatment.

4. Because the effects of treatment may be temporary or long-term, in most cases we need information about the impact of your treatment, including its expected duration and side effects, over a sufficient period of time to help us assess its outcome. When adverse effects of treatment contribute to the severity of your impairment(s), we will consider the duration or expected duration of the treatment when we assess the duration of your impairment(s).

5. If you need parenteral (intravenous) nutrition or supplemental enteral nutrition via a gastrostomy to avoid debilitating complications of a digestive disorder, this treatment will not, in itself, indicate that you have marked and severe functional limitations. The exceptions are 105.07, short bowel syndrome, and 105.10, for children who have not attained age 3 and who require supplemental daily enteral feedings via a gastrostomy (see 105.00F and 105.00H).

6. If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the severity and duration of your digestive impairment on the basis of current medical and other evidence in your case record. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the digestive system listings, but your digestive impairment may medically equal a listing or functionally equal the listings.

D. *How do we evaluate chronic liver disease?*

1. *General. Chronic liver disease* is characterized by liver cell necrosis, inflammation, or scarring (fibrosis or cirrhosis), due to any cause, that persists for more than 6 months. Chronic liver disease may result in portal hypertension, cholestasis (suppression of bile flow), extrahepatic manifestations, or liver cancer. (We evaluate liver cancer under 113.03.) Significant loss of liver function may be manifested by hemorrhage from varices or portal hypertensive gastropathy, ascites (accumulation of fluid in the abdominal cavity), hydrothorax (ascitic fluid in the chest cavity), or encephalopathy. There can also be progressive deterioration of laboratory findings that are indicative of liver dysfunction. Liver transplantation is the only definitive cure for end stage liver disease (ESLD).

2. *Examples of chronic liver disease* include, but are not limited to, biliary atresia, chronic hepatitis, non-alcoholic steatohepatitis (NASH), primary biliary cirrhosis (PBC), primary sclerosing cholangitis (PSC), autoimmune hepatitis, hemochromatosis, drug-induced liver disease, Wilson's disease, and serum alpha-1 antitrypsin deficiency. Children can also have congenital abnormalities of abdominal organs or inborn metabolic disorders that result in chronic liver disease. Acute hepatic injury is frequently reversible as in viral, drug-induced, toxin-induced, and ischemic hepatitis. In the absence of evidence of a chronic impairment, episodes of acute liver disease do not meet 105.05.

3. *Manifestations of chronic liver disease.*

a. *Symptoms* may include, but are not limited to, pruritis (itching), fatigue, nausea, loss of appetite, or sleep disturbances. Children can also have associated developmental delays or poor school performance. Symptoms of chronic liver disease may have a poor correlation with the severity of liver disease and functional ability.

b. *Signs* may include, but are not limited to, jaundice, enlargement of the liver and spleen, ascites, peripheral edema, and altered mental status.

c. *Laboratory findings* may include, but are not limited to, increased liver enzymes, increased serum total bilirubin, increased ammonia levels, decreased serum albumin, and abnormal coagulation studies, such as increased International Normalized Ratio (INR) or decreased platelet counts. Abnormally low serum albumin or elevated INR levels indicate loss of synthetic liver function, with increased likelihood of cirrhosis and associated complications. However, other abnormal lab tests, such as liver enzymes, serum total bilirubin, or ammonia levels, may have a poor correlation with the severity of liver disease and functional ability. A liver biopsy may demonstrate the degree of liver cell necrosis, inflammation, fibrosis, and cirrhosis. If you have had a liver biopsy, we will make every reasonable effort to obtain the results; however, we will not purchase a liver biopsy. Imaging studies (CAT scan, ultrasound, MRI) may show the size and consistency (fatty liver, scarring) of the liver and document ascites (see 105.00D6).

4. *Chronic viral hepatitis infections.*

a. *General.*

(i) *Chronic viral hepatitis* infections are commonly caused by hepatitis C virus (HCV), and to a lesser extent, hepatitis B virus (HBV). Usually, these are slowly progressive disorders that persist over many years during which the symptoms and signs are typically nonspecific, intermittent, and mild (for example, fatigue, difficulty with concentration, or right upper quadrant pain). Laboratory findings (liver enzymes, imaging studies, liver biopsy pathology) and complications are generally similar in HCV and HBV. The spectrum of these chronic viral hepatitis infections ranges widely and includes an asymptomatic state; insidious disease with mild to moderate symptoms associated with fluctuating liver tests; extrahepatic manifestations; cirrhosis, both compensated and decompensated; ESLD with the need for liver transplantation; and liver cancer. Treatment for chronic viral hepatitis infections varies considerably based on age, medication tolerance, treatment response, adverse effects of treatment, and duration of the treatment. Comorbid disorders, such as HIV infection, may affect the clinical course of viral hepatitis infection(s) or may alter the response to medical treatment.

(ii) We evaluate all types of chronic viral hepatitis infections under 105.05 or any listing in an affected body system(s). If your impairment(s) does not meet or medically equal a listing, we will consider the effects of your hepatitis when we assess whether your impairment(s) functionally equals the listings.

b. *Chronic hepatitis B virus (HBV) infection.*

(i) *Chronic HBV* infection is diagnosed by the detection of hepatitis B surface antigen (HBsAg) in the blood for at least 6 months. In addition, detection of the hepatitis B envelope antigen (HBeAg) suggests an increased likelihood of progression to cirrhosis and ESLD.

(ii) The therapeutic goal of treatment is to suppress HBV replication and thereby prevent progression to cirrhosis and ESLD. Treatment usually includes a combination of interferon injections and oral antiviral agents. Common adverse effects of treatment are the same as noted in 105.00D4c(ii) for HCV, and generally end within a few days after treatment is discontinued.

c. *Chronic hepatitis C virus (HCV) infection.*

(i) *Chronic HCV infection* is diagnosed by the detection of hepatitis C viral RNA in the blood for at least 6 months. Documentation of the therapeutic response to treatment is also monitored by the quantitative assay of serum HCV RNA (''HCV viral load''). Treatment usually includes a combination of interferon injections and oral ribavirin; whether a therapeutic response has occurred is usually assessed after 12 weeks of treatment by checking the HCV viral load. If there has been a substantial reduction in HCV viral load (also known as early viral response, or EVR), this reduction is predictive of a sustained viral response with completion of treatment. Combined therapy is commonly discontinued after 12 weeks when there is no early viral response, since in that circumstance there is little chance of obtaining a sustained viral response (SVR). Otherwise, treatment is usually continued for a total of 48 weeks.

(ii) Combined interferon and ribavirin treatment may have significant adverse effects that may require dosing reduction,

planned interruption of treatment, or discontinuation of treatment. Adverse effects may include: Anemia (ribavirin-induced hemolysis), neutropenia, thrombocytopenia, fever, cough, fatigue, myalgia, arthralgia, nausea, loss of appetite, pruritis, and insomnia. Behavioral side effects may also occur. Influenza-like symptoms are generally worse in the first 4 to 6 hours after each interferon injection and during the first weeks of treatment. Adverse effects generally end within a few days after treatment is discontinued.

d. *Extrahepatic manifestations of HBV and HCV*. In addition to their hepatic manifestations, both HBV and HCV may have significant extrahepatic manifestations in a variety of body systems. These include, but are not limited to: Keratoconjunctivitis (sicca syndrome), glomerulonephritis, skin disorders (for example, lichen planus, porphyria cutanea tarda), neuropathy, and immune dysfunction (for example, cryoglobulinemia, Sjögren's syndrome, and vasculitis). The extrahepatic manifestations of HBV and HCV may not correlate with the severity of your hepatic impairment. If your impairment(s) does not meet or medically equal a listing in an affected body system(s), we will consider the effects of your extrahepatic manifestations when we determine whether your impairment(s) functionally equals the listings.

5. *Gastrointestinal hemorrhage* (105.02 and 105.05A). Gastrointestinal hemorrhaging can result in hematemesis (vomiting of blood), melena (tarry stools), or hematochezia (bloody stools). Under 105.02, the required transfusions of at least 10 cc of blood/kg of body weight must be at least 30 days apart and occur at least three times during a consecutive 6-month period. Under 105.05A, *hemodynamic instability* is diagnosed with signs such as pallor (pale skin), diaphoresis (profuse perspiration), rapid pulse, low blood pressure, postural hypotension (pronounced fall in blood pressure when arising to an upright position from lying down) or syncope (fainting). Hemorrhaging that results in hemodynamic instability is potentially life-threatening and therefore requires hospitalization for transfusion and supportive care. Under 105.05A, we require only one hospitalization for transfusion of at least 10 cc of blood/kg of body weight.

6. *Ascites or hydrothorax* (105.05B) indicates significant loss of liver function due to chronic liver disease. We evaluate ascites or hydrothorax that is not attributable to other causes under 105.05B. The required findings must be present on at least two evaluations at least 60 days apart within a consecutive 6-month period and despite continuing treatment as prescribed.

7. *Spontaneous bacterial peritonitis* (105.05C) is an infectious complication of chronic liver disease. It is diagnosed by ascitic peritoneal fluid that is documented to contain an absolute neutrophil count of at least 250 cells/mm^3. The required finding in 105.05C is satisfied with one evaluation documenting peritoneal fluid infection. We do not evaluate other causes of peritonitis that are unrelated to chronic liver disease, such as tuberculosis, malignancy, and perforated bowel, under this listing. We evaluate these other causes of peritonitis under the appropriate body system listings.

8. *Hepatorenal syndrome* (105.05D) is defined as functional renal failure associated with chronic liver disease in the absence of underlying kidney pathology. Hepatorenal syndrome is documented by elevation of serum creatinine, marked sodium retention, and oliguria (reduced urine output). The requirements of 105.05D are satisfied with documentation of any one of the three laboratory findings on one evaluation. We do not evaluate known causes of renal dysfunction, such as glomerulonephritis, tubular necrosis, drug-induced renal disease, and renal infections, under this listing. We evaluate these other renal impairments under 106.00ff.

9. *Hepatopulmonary syndrome* (105.05E) is defined as arterial deoxygenation (hypoxemia) that is associated with chronic liver disease due to intrapulmonary arteriovenous shunting and vasodilatation, in the absence of other causes of arterial deoxygenation. Clinical manifestations usually include dyspnea, orthodeoxia (increasing hypoxemia with erect position), platypnea (improvement of dyspnea with flat position), cyanosis, and clubbing. The requirements of 105.05E are satisfied with documentation of any one of the findings on one evaluation. In 105.05E1, we require documentation of the altitude of the testing facility because altitude affects the measurement of arterial oxygenation. We will not purchase the specialized studies described in 105.05E2; however, if you have had these studies at a time relevant to your claim, we will make every reasonable effort to obtain the reports for the purpose of establishing whether your impairment meets 105.05E2.

10. *Hepatic encephalopathy* (105.05F).

a. *General*. Hepatic encephalopathy usually indicates severe loss of hepatocellular function. We define hepatic encephalopathy under 105.05F as a recurrent or chronic neuropsychiatric disorder, characterized by abnormal behavior, cognitive dysfunction, altered state of consciousness, and ultimately coma and death. The diagnosis is established by changes in mental status associated with fleeting neurological signs, including ''flapping tremor'' (asterixis), characteristic electroencephalographic (EEG) abnormalities, or abnormal laboratory values that indicate loss of synthetic liver function. We will not purchase the EEG testing described in 105.05F3b. However, if you have had this test at a time relevant to your claim, we will make every reasonable effort

to obtain the report for the purpose of establishing whether your impairment meets 105.05F.

b. *Acute encephalopathy.* We will not evaluate your acute encephalopathy under 105.05F if it results from conditions other than chronic liver disease, such as vascular events and neoplastic diseases. We will evaluate these other causes of acute encephalopathy under the appropriate body system listings.

11. *End stage liver disease (ESLD) documented by scores from the SSA Chronic Liver Disease (SSA CLD) calculation (105.05G1) and SSA Chronic Liver Disease-Pediatric (SSA CLD–P) calculation (105.05G2).*

a. *SSA CLD score.*

(i) If you are age 12 or older, we will use the SSA CLD score to evaluate your ESLD under 105.05G1. We explain how we calculate the SSA CLD score in a(ii) through a(vii) of this section.

(ii) To calculate the SSA CLD score, we use a formula that includes three laboratory values: Serum total bilirubin (mg/dL), serum creatinine (mg/dL), and International Normalized Ratio (INR). The formula for the SSA CLD score calculation is:

$9.57 \times [\text{Log}_e \text{ (serum creatinine mg/dL)}]$
$+ 3.78 \times [\text{Log}_e \text{ (serum total bilirubin mg/dL)}]$
$+ 11.2 \times [\text{Log}_e \text{ (INR)}]$
$+ 6.43$

(iii) When we indicate ''Log_e'' in the formula for the SSA CLD score calculation, we mean the ''base *e* logarithm'' or ''natural logarithm'' (ln) of a numerical laboratory value, not the ''base 10 logarithm'' or ''common logarithm'' (log) of the laboratory value, and not the actual laboratory value. For an example of SSA CLD calculation, see 5.00D11c.

(iv) For any SSA CLD score calculation, all of the required laboratory values must have been obtained within 30 days of each other. If there are multiple laboratory values within the 30-day interval for any given laboratory test (serum total bilirubin, serum creatinine, or INR), we will use the highest value for the SSA CLD score calculation. We will round all laboratory values less than 1.0 up to 1.0.

(v) Listing 105.05G requires two SSA CLD scores. The laboratory values for the second SSA CLD score calculation must have been obtained at least 60 days after the latest laboratory value for the first SSA CLD score and within the required 6-month period. We will consider the date of each SSA CLD score to be the date of the first laboratory value used for its calculation.

(vi) If you are in renal failure or on dialysis within a week of any serum creatinine test in the period used for the SSA CLD calculation, we will use a serum creatinine of 4, which is the maximum serum creatinine level allowed in the calculation, to calculate your SSA CLD score.

(vii) If you have the two SSA CLD scores required by 105.05G1, we will find that your impairment meets the criteria of the listing from at least the date of the first SSA CLD score.

b. *SSA CLD–P score.*

(i) If you have not attained age 12, we will use the SSA CLD–P score to evaluate your ESLD under 105.05G2. We explain how we calculate the SSA CLD–P score in b(ii) through b(vii) of this section.

(ii) To calculate the SSA CLD–P score, we use a formula that includes four parameters: Serum total bilirubin (mg/dL), International Normalized Ratio (INR), serum albumin (g/dL), and whether growth failure is occurring. The formula for the SSA CLD–P score calculation is:

$4.80 \times [\text{Log}_e \text{ (serum total bilirubin mg/dL)}]$
$+ 18.57 \times [\text{Log}_e \text{ (INR)}]$
$- 6.87 \times [\text{Log}_e \text{ (serum albumin g/dL)}]$
$+ 6.67$ if the child has growth failure (< -2 standard deviations for weight or height)

(iii) When we indicate ''Log_e'' in the formula for the SSA CLD–P score calculation, we mean the ''base *e* logarithm'' or ''natural logarithm'' (ln) of a numerical laboratory value, not the ''base 10 logarithm'' or ''common logarithm'' (log) of the laboratory value, and not the actual laboratory value. For example, if a female child is 4.0 years old, has a current weight of 13.5 kg (10th percentile for age) and height of 92 cm (less than the third percentile for age), and has laboratory values of serum total bilirubin 2.2 mg/dL, INR 1.0, and serum albumin 3.5 g/dL, we will compute the SSA CLD–P score as follows:

$4.80 \times [\text{Log}_e + \text{(serum total bilirubin 2.2 mg/dL)} = 0.788]$
$+ 18.57 \times [\text{Log}_e \text{ (INR 1.0)} = 0]$
$- 6.87 \times [\text{Log}_e + \text{(serum albumin 3.5 g/dL)} = 1.253]$
$+ 6.67$

$= 3.78 + 0 - 8.61 + 6.67$
$= 1.84$, which is then rounded to an SSA CLD–P score of 2

(iv) For any SSA CLD–P score calculation, all of the required laboratory values (serum total bilirubin, INR, or serum albumin) must have been obtained within 30 days of each other. We will not purchase INR values for children who have not attained age 12. If there is no INR value for a child under 12 within the applicable time period, we will use an INR value of 1.1 to calculate the SSA CLD–P score. If there are multiple laboratory values within the 30-day interval for any given laboratory test, we will use the highest serum total bilirubin and INR values and the lowest serum albumin value for the SSA CLD–P score calculation. We will round all laboratory values less than 1.0 up to 1.0.

(v) The weight and length/height measurements used for the calculation must be obtained from one evaluation within the same 30-day period as in D11b(iv).

(vi) Listing 105.05G2 requires two SSA CLD–P scores. The laboratory values for the second SSA CLD–P score calculation must have been obtained at least 60 days after the latest laboratory value for the first SSA CLD–P score and within the required 6-month period. We will consider the date of each SSA CLD–P score to be the date of the first laboratory value used for its calculation.

(vii) If you have the two SSA CLD–P scores required by listing 105.05G2, we will find that your impairment meets the criteria of the listing from at least the date of the first SSA CLD–P score.

12. *Extrahepatic biliary atresia (EBA)* (105.05H) usually presents in the first 2 months of life with persistent jaundice. The impairment meets 105.05H if the diagnosis of EBA is confirmed by liver biopsy or intraoperative cholangiogram that shows obliteration of the extrahepatic biliary tree. EBA is usually surgically treated by portoenterostomy (for example, Kasai procedure). If this surgery is not performed in the first months of life or is not completely successful, liver transplantation is indicated. If you have had a liver transplant, we will evaluate your impairment under 105.09.

13. *Liver transplantation* (105.09) may be performed for metabolic liver disease, progressive liver failure, life-threatening complications of liver disease, hepatic malignancy, and acute fulminant hepatitis (viral, drug-induced, or toxin-induced). We will consider you to be disabled for 1 year from the date of the transplantation. Thereafter, we will evaluate your residual impairment(s) by considering the adequacy of post-transplant liver function, the requirement for post-transplant antiviral therapy, the frequency and severity of rejection episodes, comorbid complications, and all adverse treatment effects.

E. *How do we evaluate inflammatory bowel disease (IBD)?*

1. *Inflammatory bowel disease* (105.06) includes, but is not limited to, Crohn's disease and ulcerative colitis. These disorders, while distinct entities, share many clinical, laboratory, and imaging findings, as well as similar treatment regimens. Remissions and exacerbations of variable duration are the hallmark of IBD. Crohn's disease may involve the entire alimentary tract from the mouth to the anus in a segmental, asymmetric fashion. Obstruction, stenosis, fistulization, perineal involvement, and extraintestinal manifestations are common. Crohn's disease is rarely curable and recurrence may be a lifelong problem, even after surgical resection. In contrast, ulcerative colitis only affects the colon. The inflammatory process may be limited to the rectum, extend proximally to include any contiguous segment, or involve the entire colon. Ulcerative colitis may be cured by total colectomy.

2. Symptoms and signs of IBD include diarrhea, fecal incontinence, rectal bleeding, abdominal pain, fatigue, fever, nausea, vomiting, arthralgia, abdominal tenderness, palpable abdominal mass (usually inflamed loops of bowel) and perineal disease. You may also have signs or laboratory findings indicating malnutrition, such as weight loss, edema, anemia, hypoalbuminemia, hypokalemia, hypocalcemia, or hypomagnesemia.

3. IBD may be associated with significant extraintestinal manifestations in a variety of body systems. These include, but are not limited to, involvement of the eye (for example, uveitis, episcleritis, iritis); hepatobiliary disease (for example, gallstones, primary sclerosing cholangitis); urologic disease (for example, kidney stones, obstructive hydronephrosis); skin involvement (for example, erythema nodosum, pyoderma gangrenosum); or non-destructive inflammatory arthritis. You may also have associated thromboembolic disorders or vascular disease. These manifestations may not correlate with the severity of your IBD. If your impairment does not meet any of the criteria of 105.06, we will consider the effects of your extraintestinal manifestations in determining whether you have an impairment(s) that meets or medically equals another listing, and we will also consider the effects of your extraintestinal manifestations when we determine whether your impairment(s) functionally equals the listings.

4. Surgical diversion of the intestinal tract, including ileostomy and colostomy, does not very seriously interfere with age-appropriate functioning if you are able to maintain adequate nutrition and function of the stoma. However, if you are not able to maintain adequate nutrition, we will evaluate your impairment under 105.08.

F. *How do we evaluate short bowel syndrome (SBS)?*

1. *Short bowel syndrome* (105.07) is a disorder that occurs when congenital intestinal abnormalities, ischemic vascular insults (for example, necrotizing enterocolitis, volvulus), trauma, or IBD complications require surgical resection of more than one-half of the small intestine, resulting in the loss of intestinal absorptive surface and a state of chronic malnutrition. The management of SBS requires long-term parenteral nutrition via an indwelling central venous catheter (central line); the process is often referred to as *hyperalimentation* or *total parenteral nutrition* (TPN). Children with SBS can also feed orally, with variable amounts of nutrients being absorbed through their remaining intestine.

Over time, some of these children can develop additional intestinal absorptive surface, and may ultimately be able to be weaned off their parenteral nutrition.

2. Your impairment will continue to meet 105.07 as long as you remain dependent on daily parenteral nutrition via a central venous catheter for most of your nutritional requirements. Long-term complications of SBS and parenteral nutrition include abnormal growth rates, central line infections (with or without septicemia), thrombosis, hepatotoxicity, gallstones, and loss of venous access sites. Intestinal transplantation is the only definitive treatment for children with SBS who remain chronically dependent on parenteral nutrition.

3. To document SBS, we need a copy of the operative report of intestinal resection, the summary of the hospitalization(s) including: Details of the surgical findings, medically appropriate postoperative imaging studies that reflect the amount of your residual small intestine, or if we cannot get one of these reports, other medical reports that include details of the surgical findings. We also need medical documentation that you are dependent on daily parenteral nutrition to provide most of your nutritional requirements.

G. *How do we evaluate growth failure due to any digestive disorder?*

1. To evaluate growth failure due to any digestive disorder, we require documentation of the laboratory findings of chronic nutritional deficiency described in 105.08A and the growth measurements in 105.08B within the same consecutive 12-month period. The dates of laboratory findings may be different from the dates of growth measurements.

2. Under 105.08B, we evaluate a child's growth failure by using the appropriate table for age and gender.

a. For children from birth to attainment of age 2, we use the weight-for-length table (see Table I or Table II).

b. For children age 2 to attainment of age 18, we use the body mass index (BMI)-for-age table (see Tables III or IV).

c. BMI is the ratio of a child's weight to the square of the child's height. We calculate BMI using one of the following formulas:

English Formula

BMI = [Weight in Pounds/(Height in Inches × Height in Inches)] × 703

Metric Formulas

BMI = Weight in Kilograms/(Height in Meters × Height in Meters)

BMI = [Weight in Kilograms/(Height in Centimeters × Height in Centimeters)] × 10,000

H. *How do we evaluate the need for supplemental daily enteral feeding via a gastrostomy?*

1. *General.* Infants and young children may have anatomical, neurological, or developmental disorders that interfere with their ability to feed by mouth, resulting in inadequate caloric intake to meet their growth needs. These disorders frequently result in the medical necessity to supplement caloric intake and to bypass the anatomical feeding route of mouth-throat-esophagus into the stomach.

2. Children who have not attained age 3 and who require supplemental daily enteral nutrition via a feeding gastrostomy meet 105.10 regardless of the medical reason for the gastrostomy. Thereafter, we evaluate growth impairment under 100.02, malnutrition under 105.08, or other medical or developmental disorder(s) (including the disorder(s) that necessitated gastrostomy placement) under the appropriate listing(s).

I. *How do we evaluate esophageal stricture or stenosis?* Esophageal stricture or stenosis (narrowing) from congenital atresia (absence or abnormal closure of a tubular body organ) or destructive esophagitis may result in malnutrition or the need for gastrostomy placement, which we evaluate under 105.08 or 105.10. Esophageal stricture or stenosis may also result in complications such as pneumonias due to frequent aspiration, or difficulty in maintaining nutritional status short of listing-level severity. While none of these complications may be of such severity that they would meet the criteria of another listing, the combination of impairments may medically equal the severity of a listing or functionally equal the listings.

J. *What do we mean by the phrase ''consider under a disability for 1 year''?* We use the phrase ''consider under a disability for 1 year'' following a specific event in 105.02, 105.05A, and 105.09 to explain how long your impairment can meet the requirements of those particular listings. This phrase does not refer to the date on which your disability began, only to the date on which we must reevaluate whether your impairment continues to meet a listing or is otherwise disabling. For example, if you have received a liver transplant, you may have become disabled before the transplant because of chronic liver disease. Therefore, we do not restrict our determination of the onset of disability to the date of the specified event. We will establish an onset date earlier than the date of the specified event if the evidence in your case record supports such a finding.

K. *How do we evaluate impairments that do not meet one of the digestive disorder listings?*

1. These listings are only examples of common digestive disorders that we consider severe enough to result in marked and severe functional limitations. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system. For example:

a. If you have hepatitis B or C and you are depressed, we will evaluate your impairment under 112.04.

b. If you have multiple congenital abnormalities, we will evaluate your impairment(s) under the criteria in the listings for impairments that affect multiple body systems (110.00) or the criteria of listings in other affected body systems.

c. If you have digestive disorders that interfere with intake, digestion, or absorption of nutrition, and result in a reduction in your rate of growth, and your impairment does not satisfy the criteria in the malnutrition listing (105.08), we will evaluate your impairment under the growth impairment listings (100.00).

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926.) If your impairment(s) does not meet or medically equal a listing, you may or may not have an impairment(s) that functionally equals the listings. (See §416.926a.) When we decide whether you continue to be disabled, we use the rules in §416.994a.

105.01 *Category of Impairments, Digestive System*

105.02 *Gastrointestinal hemorrhaging from any cause, requiring blood transfusion* (with or without hospitalization) of at least 10 cc of blood/kg of body weight, and occurring at least three times during a consecutive 6-month period. The transfusions must be at least 30 days apart within the 6-month period. Consider under a disability for 1 year following the last documented transfusion; thereafter, evaluate the residual impairment(s).

105.03–105.04 [Reserved]

105.05 *Chronic liver disease*, with:

A. Hemorrhaging from esophageal, gastric, or ectopic varices or from portal hypertensive gastropathy, demonstrated by endoscopy, x-ray, or other appropriate medically acceptable imaging, resulting in hemodynamic instability as defined in 105.00D5, and requiring hospitalization for transfusion of at least 10 cc of blood/kg of body weight. Consider under a disability for 1 year following the last documented transfusion; thereafter, evaluate the residual impairment(s).

OR

B. Ascites or hydrothorax not attributable to other causes, despite continuing treatment as prescribed, present on at least two evaluations at least 60 days apart within a consecutive 6-month period. Each evaluation must be documented by:

1. Paracentesis or thoracentesis; or

2. Appropriate medically acceptable imaging or physical examination and one of the following:

a. Serum albumin of 3.0 g/dL or less; or

b. International Normalized Ratio (INR) of at least 1.5.

OR

C. Spontaneous bacterial peritonitis with peritoneal fluid containing an absolute neutrophil count of at least 250 cells/mm^3.

OR

D. Hepatorenal syndrome as described in 105.00D8, with one of the following:

1. Serum creatinine elevation of at least 2 mg/dL; or

2. Oliguria with 24-hour urine output less than 1 mL/kg/hr; or

3. Sodium retention with urine sodium less than 10 mEq per liter.

OR

E. Hepatopulmonary syndrome as described in 105.00D9, with:

1. Arterial oxygenation (P_aO_2,) on room air of:

a. 60 mm Hg or less, at test sites less than 3000 feet above sea level, or

b. 55 mm Hg or less, at test sites from 3000 to 6000 feet, or

c. 50 mm Hg or less, at test sites above 6000 feet; or

2. Documentation of intrapulmonary arteriovenous shunting by contrast-enhanced echocardiography or macroaggregated albumin lung perfusion scan.

OR

F. Hepatic encephalopathy as described in 105.00D10, with 1 and either 2 or 3:

1. Documentation of abnormal behavior, cognitive dysfunction, changes in mental status, or altered state of consciousness (for example, confusion, delirium, stupor, or coma), present on at least two evaluations at least 60 days apart within a consecutive 6-month period; and

2. History of transjugular intrahepatic portosystemic shunt (TIPS) or any surgical portosystemic shunt; or

3. One of the following occurring on at least two evaluations at least 60 days apart within the same consecutive 6-month period as in F1:

a. Asterixis or other fluctuating physical neurological abnormalities; or

b. Electroencephalogram (EEG) demonstrating triphasic slow wave activity; or

c. Serum albumin of 3.0 g/dL or less; or

d. International Normalized Ratio (INR) of 1.5 or greater.

OR

G. End Stage Liver Disease, with:

1. For children 12 years of age or older, SSA CLD scores of 22 or greater calculated as described in 105.00D11a. Consider under a disability from at least the date of the first score.

2. For children who have not attained age 12, SSA CLD–P scores of 11 or greater calculated as described in 105.00D11b. Consider under a disability from at least the date of the first score.

OR

H. Extrahepatic biliary atresia as diagnosed on liver biopsy or intraoperative cholangiogram. Consider under a disability for 1 year following the diagnosis; thereafter, evaluate the residual liver function.

105.06 *Inflammatory bowel disease (IBD)* documented by endoscopy, biopsy, appropriate medically acceptable imaging, or operative findings with:

A. Obstruction of stenotic areas (not adhesions) in the small intestine or colon with proximal dilatation, confirmed by appropriate medically acceptable imaging or in surgery, requiring hospitalization for intestinal decompression or for surgery, and occurring on at least two occasions at least 60 days apart within a consecutive 6-month period;

OR

B. Two of the following despite continuing treatment as prescribed and occurring within the same consecutive 6-month period:

1. Anemia with hemoglobin less than 10.0 g/dL, present on at least two evaluations at least 60 days apart; or

2. Serum albumin of 3.0 g/dL or less, present on at least two evaluations at least 60 days apart; or

3. Clinically documented tender abdominal mass palpable on physical examination with abdominal pain or cramping that is not completely controlled by prescribed narcotic medication, present on at least two evaluations at least 60 days apart; or

4. Perineal disease with a draining abscess or fistula, with pain that is not completely controlled by prescribed narcotic medication, present on at least two evaluations at least 60 days apart; or

5. Need for supplemental daily enteral nutrition via a gastrostomy or daily parenteral nutrition via a central venous catheter. (See 105.10 for children who have not attained age 3.)

105.07 *Short bowel syndrome (SBS)*, due to surgical resection of more than one-half of the small intestine, with dependence on daily parenteral nutrition via a central venous catheter (see 105.00F).

105.08 *Growth failure due to any digestive disorder* (see 105.00G), documented by A and B:

A. Chronic nutritional deficiency present on at least two evaluations at least 60 days apart within a consecutive 12-month period documented by one of the following:

1. Anemia with hemoglobin less than 10.0 g/dL; or

2. Serum albumin of 3.0 g/dL or less;

AND

B. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2*, three weight-for-length measurements that are:

a. Within a 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on Table I or Table II; or

TABLE I—MALES BIRTH TO ATTAINMENT OF AGE 2

[Third Percentile Values for Weight-for-Length]

Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight (kilograms)
45.0	1.597	64.5	6.132	84.5	10.301
45.5	1.703	65.5	6.359	85.5	10.499
46.5	1.919	66.5	6.584	86.5	10.696
47.5	2.139	67.5	6.807	87.5	10.895
48.5	2.364	68.5	7.027	88.5	11.095
49.5	2.592	69.5	7.245	89.5	11.296
50.5	2.824	70.5	7.461	90.5	11.498
51.5	3.058	71.5	7.674	91.5	11.703
52.5	3.294	72.5	7.885	92.5	11.910
53.5	3.532	73.5	8.094	93.5	12.119
54.5	3.771	74.5	8.301	94.5	12.331
55.5	4.010	75.5	8.507	95.5	12.546
56.5	4.250	76.5	8.710	96.5	12.764
57.5	4.489	77.5	8.913	97.5	12.987
58.5	4.728	78.5	9.113	98.5	13.213
59.5	4.966	79.5	9.313	99.5	13.443
60.5	5.203	80.5	9.512	100.5	13.678
61.5	5.438	81.5	9.710	101.5	13.918
62.5	5.671	82.5	9.907	102.5	14.163
63.5	5.903	83.5	10.104	103.5	14.413

TABLE II—FEMALES BIRTH TO ATTAINMENT OF AGE 2

[Third percentile values for weight-for-length]

Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight kilograms)
45.0	1.613	64.5	5.985	84.5	10.071
45.5	1.724	65.5	6.200	85.5	10.270

TABLE II—FEMALES BIRTH TO ATTAINMENT OF AGE 2—Continued

[Third percentile values for weight-for-length]

Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight (kilograms)	Length (centimeters)	Weight kilograms)
46.5	1.946	66.5	6.413	86.5	10.469
47.5	2.171	67.5	6.625	87.5	10.670
48.5	2.397	68.5	6.836	88.5	10.871
49.5	2.624	69.5	7.046	89.5	11.074
50.5	2.852	70.5	7.254	90.5	11.278
51.5	3.081	71.5	7.461	91.5	11.484
52.5	3.310	72.5	7.667	92.5	11.691
53.5	3.538	73.5	7.871	93.5	11.901
54.5	3.767	74.5	8.075	94.5	12.112
55.5	3.994	75.5	8.277	95.5	12.326
56.5	4.220	76.5	8.479	96.5	12.541
57.5	4.445	77.5	8.679	97.5	12.760
58.5	4.669	78.5	8.879	98.5	12.981
59.5	4.892	79.5	9.078	99.5	13.205
60.5	5.113	80.5	9.277	100.5	13.431
61.5	5.333	81.5	9.476	101.5	13.661
62.5	5.552	82.5	9.674	102.5	13.895
63.5	5.769	83.5	9.872	103.5	14.132

2. *For children age 2 to attainment of age 18*, three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on Table III or Table IV.

TABLE III—MALES AGE 2 TO ATTAINMENT OF AGE 18

[Third Percentile Values for BMI-for-Age]

Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI
2.0 to 2.1	14.5	10.11 to 11.2	14.3	14.9 to 14.10	16.1
2.2 to 2.4	14.4	11.3 to 11.5	14.4	14.11 to 15.0	16.2
2.5 to 2.7	14.3	11.6 to 11.8	14.5	15.1 to 15.3	16.3
2.8 to 2.11	14.2	11.9 to 11.11	14.6	15.4 to 15.5	16.4
3.0 to 3.2	14.1	12.0 to 12.1	14.7	15.6 to 15.7	16.5
3.3 to 3.6	14.0	12.2 to 12.4	14.8	15.8 to 15.9	16.6
3.7 to 3.11	13.9	12.5 to 12.7	14.9	15.10 to 15.11	16.7
4.0 to 4.5	13.8	12.8 to 12.9	15.0	16.0 to 16.1	16.8
4.6 to 5.0	13.7	12.10 to 13.0	15.1	16.2 to 16.3	16.9
5.1 to 6.0	13.6	13.1 to 13.2	15.2	16.4 to 16.5	17.0
6.1 to 7.6	13.5	13.3 to 13.4	15.3	16.6 to 16.8	17.1
7.7 to 8.6	13.6	13.5 to 13.7	15.4	16.9 to 16.10	17.2
8.7 to 9.1	13.7	13.8 to 13.9	15.5	16.11 to 17.0	17.3
9.2 to 9.6	13.8	13.10 to 13.11	15.6	17.1 to 17.2	17.4
9.7 to 9.11	13.9	14.0 to 14.1	15.7	17.3 to 17.5	17.5
10.0 to 10.3	14.0	14.2 to 14.4	15.8	17.6 to 17.7	17.6
10.4 to 10.7	14.1	14.5 to 14.6	15.9	17.8 to 17.9	17.7
10.8 to 10.10	14.2	14.7 to 14.8	16.0	17.10 to 17.11	17.8

TABLE IV—FEMALES AGE 2 TO ATTAINMENT OF AGE 18

[Third Percentile Values for BMI-for-Age]

Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI
2.0 to 2.2	14.1	10.8 to 10.10	14.0	14.3 to 14.5	15.6
2.3 to 2.6	14.0	10.11 to 11.2	14.1	14.6 to 14.7	15.7
2.7 to 2.10	13.9	11.3 to 11.5	14.2	14.8 to 14.9	15.8
2.11 to 3.2	13.8	11.6 to 11.7	14.3	14.10 to 15.0	15.9
3.3 to 3.6	13.7	11.8 to 11.10	14.4	15.1 to 15.2	16.0
3.7 to 3.11	13.6	11.11 to 12.1	14.5	15.3 to 15.5	16.1
4.0 to 4.4	13.5	12.2 to 12.4	14.6	15.6 to 15.7	16.2
4.5 to 4.11	13.4	12.5 to 12.6	14.7	15.8 to 15.10	16.3
5.0 to 5.9	13.3	12.7 to 12.9	14.8	15.11 to 16.0	16.4
5.10 to 7.6	13.2	12.10 to 12.11	14.9	16.1 to 16.3	16.5
7.7 to 8.4	13.3	13.0 to 13.2	15.0	16.4 to 16.6	16.6

TABLE IV—FEMALES AGE 2 TO ATTAINMENT OF AGE 18—Continued

[Third Percentile Values for BMI-for-Age]

Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI	Age (yrs. and mos.)	BMI
8.5 to 8.10	13.4	13.3 to 13.4	15.1	16.7 to 16.9	16.7
8.11 to 9.3	13.5	13.5 to 13.7	15.2	16.10 to 17.0	16.8
9.4 to 9.8	13.6	13.8 to 13.9	15.3	17.1 to 17.3	16.9
9.9 to 10.0	13.7	13.10 to 14.0	15.4	17.4 to 17.7	17.0
10.1 to 10.4	13.8	14.1 to 14.2	15.5	17.8 to 17.11	17.1
10.5 to 10.7	13.9				

105.09 *Liver transplantation.* Consider under a disability for 1 year following the date of transplantation; thereafter, evaluate the residual impairment(s) (see 105.00D13 and 105.00J).

105.10 *Need for supplemental daily enteral feeding via a gastrostomy* due to any cause, for children who have not attained age 3; thereafter, evaluate the residual impairment(s) (see 105.00H).

106.00 GENITOURINARY DISORDERS

A. Which disorders do we evaluate under these listings?

We evaluate genitourinary disorders resulting in chronic kidney disease (CKD). Examples of such disorders include chronic glomerulonephritis, hypertensive nephropathy, diabetic nephropathy, chronic obstructive uropathy, and hereditary nephropathies. We also evaluate nephrotic syndrome due to glomerular dysfunction, and congenital genitourinary disorders, such as ectopic ureter, exstrophic urinary bladder, urethral valves, and Eagle-Barrett syndrome (prune belly syndrome), under these listings.

B. What evidence do we need?

1. We need evidence that documents the signs, symptoms, and laboratory findings of your CKD. This evidence should include reports of clinical examinations, treatment records, and documentation of your response to treatment. Laboratory findings, such as serum creatinine or serum albumin levels, may document your kidney function. We generally need evidence covering a period of at least 90 days unless we can make a fully favorable determination or decision without it.

2. *Estimated glomerular filtration rate (eGFR).* The eGFR is an estimate of the filtering capacity of the kidneys that takes into account serum creatinine concentration and other variables, such as your age, gender, and body size. If your medical evidence includes eGFR findings, we will consider them when we evaluate your CKD under 106.05.

3. *Kidney or bone biopsy.* If you have had a kidney or bone biopsy, we need a copy of the pathology report. When we cannot get a copy of the pathology report, we will accept a statement from an acceptable medical source verifying that a biopsy was performed and describing the results.

C. What other factors do we consider when we evaluate your genitourinary disorder?

1. *Chronic hemodialysis or peritoneal dialysis.*

a. Dialysis is a treatment for CKD that uses artificial means to remove toxic metabolic byproducts from the blood. Hemodialysis uses an artificial kidney machine to clean waste products from the blood; peritoneal dialysis uses a dialyzing solution that is introduced into and removed from the abdomen (peritoneal cavity) either continuously or intermittently. Under 106.03, your ongoing dialysis must have lasted or be expected to last for a continuous period of at least 12 months. To satisfy the requirement in 106.03, we will accept a report from an acceptable medical source that describes your CKD and your current dialysis, and indicates that your dialysis will be ongoing.

b. If you are undergoing chronic hemodialysis or peritoneal dialysis, your CKD may meet our definition of disability before you started dialysis. We will determine the onset of your disability based on the facts in your case record.

2. *Kidney transplant.*

a. If you receive a kidney transplant, we will consider you to be disabled under 106.04 for 1 year from the date of transplant. After that, we will evaluate your residual impairment(s) by considering your post-transplant function, any rejection episodes you have had, complications in other body systems, and any adverse effects related to ongoing treatment.

b. If you received a kidney transplant, your CKD may meet our definition of disability before you received the transplant. We will determine the onset of your disability based on the facts in your case record.

3. *Anasarca* (generalized massive edema or swelling). Under 106.06B, we need a description of the extent of edema, including pretibial (in front of the tibia), periorbital (around the eyes), or presacral (in front of the sacrum) edema. We also need a description of any ascites, pleural effusion, or pericardial effusion.

4. *Congenital genitourinary disorder.* Procedures such as diagnostic cystoscopy or circumcision do not satisfy the requirement for urologic surgical procedures in 106.07.

5. *Growth failure due to any chronic renal disease.*

a. To evaluate growth failure due to any chronic renal disease, we require documentation of the laboratory findings described in 106.08A and the growth measurements in 106.08B within the same consecutive 12-month period. The dates of laboratory findings may be different from the dates of growth measurements.

b. Under 106.08B, we use the appropriate table(s) under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile.

(i) For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

(ii) For children age 2 to attainment of age 18, we use the body mass index (BMI)-for-age table corresponding to the child's gender (Table III or Table IV).

(iii) BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

6. *Complications of CKD.* The hospitalizations in 106.09 may be for different complications of CKD. Examples of complications from CKD that may result in hospitalization include stroke, congestive heart failure, hypertensive crisis, or acute kidney failure requiring a short course of hemodialysis. If the CKD complication occurs during a hospitalization that was initially for a co-occurring condition, we will evaluate it under our rules for determining medical equivalence. (See §416.926 of this chapter.) We will evaluate co-occurring conditions, including those that result in hospitalizations, under the listings for the affected body system or under our rules for medical equivalence.

D. How do we evaluate disorders that do not meet one of the genitourinary listings?

1. The listed disorders are only examples of common genitourinary disorders that we consider severe enough to result in marked and severe functional limitations. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926 of this chapter.) Genitourinary disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, we will also consider whether it functionally equals the listings. (See §416.926a of this chapter.) We use the rules in §416.994a of this chapter when we decide whether you continue to be disabled.

106.01 Category of Impairments, Genitourinary Disorders

106.03 *Chronic kidney disease,* with chronic hemodialysis or peritoneal dialysis (see 106.00C1).

106.04 *Chronic kidney disease,* with kidney transplant. Consider under a disability for 1 year following the transplant; thereafter, evaluate the residual impairment (see 106.00C2).

106.05 *Chronic kidney disease,* with impairment of kidney function, with one of the following documented on at least two occasions at least 90 days apart during a consecutive 12-month period:

A. Serum creatinine of 3 mg/dL or greater;

OR

B. Creatinine clearance of 30 ml/min/1.73m^2 or less;

OR

C. Estimated glomerular filtration rate (eGFR) of 30 ml/min/1.73m^2 or less.

106.06 *Nephrotic syndrome,* with A and B:

A. Laboratory findings as described in 1 or 2, documented on at least two occasions at least 90 days apart during a consecutive 12-month period:

1. Serum albumin of 3.0 g/dL or less, or

2. Proteinuria of 40 mg/m^2/hr or greater;

AND

B. Anasarca (see 106.00C3) persisting for at least 90 days despite prescribed treatment.

106.07 *Congenital genitourinary disorder* (see 106.00C4) requiring urologic surgical procedures at least three times in a consecutive 12-month period, with at least 30 days between procedures. Consider under a disability for 1 year following the date of the last surgery; thereafter, evaluate the residual impairment.

106.09 *Complications of chronic kidney disease* (see 106.00C5) requiring at least three hospitalizations within a consecutive 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

106.08 *Growth failure due to any chronic renal disease* (see 106.00C5), with:

A. Serum creatinine of 2 mg/dL or greater, documented at least two times within a consecutive 12-month period with at least 60 days between measurements.

AND

B. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2*, three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or

2. *For children age 2 to attainment of age 18*, three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.

107.00 HEMATOLOGICAL DISORDERS

A. What hematological disorders do we evaluate under these listings?

1. We evaluate non-malignant (non-cancerous) hematological disorders, such as hemolytic anemias (107.05), disorders of thrombosis and hemostasis (107.08), and disorders of bone marrow failure (107.10). These disorders disrupt the normal development and function of white blood cells, red blood cells, platelets, and clotting-factor proteins (factors).

2. We evaluate malignant (cancerous) hematological disorders, such as lymphoma, leukemia, and multiple myeloma under the appropriate listings in 113.00, except for lymphoma associated with human immunodeficiency virus (HIV) infection, which we evaluate under 114.08E.

B. What evidence do we need to document that you have a hematological disorder?

We need the following evidence to document that you have a hematological disorder:

1. A laboratory report of a definitive test that establishes a hematological disorder, signed by a physician; or

2. A laboratory report of a definitive test that establishes a hematological disorder that is not signed by a physician and a report from a physician that states you have the disorder; or

3. When we do not have a laboratory report of a definitive test, a persuasive report from a physician that a diagnosis of your hematological disorder was confirmed by appropriate laboratory analysis or other diagnostic method(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test or tests for diagnosing your disorder and provide the results, or explain how your diagnosis was established by other diagnostic method(s) consistent with the prevailing state of medical knowledge and clinical practice.

4. We will make every reasonable effort to obtain the results of appropriate laboratory testing you have had. We will not purchase complex, costly, or invasive tests, such as tests of clotting-factor proteins, and bone marrow aspirations.

C. What are hemolytic anemias, and how do we evaluate them under 107.05?

1. *Hemolytic anemias, both congenital and acquired*, are disorders that result in premature destruction of red blood cells (RBCs). Hemolytic anemias include abnormalities of hemoglobin structure (hemoglobinopathies), abnormal RBC enzyme content and function, and RBC membrane (envelope) defects that are congenital or acquired. The diagnosis of hemolytic anemia is based on hemoglobin electrophoresis or analysis of the contents of the RBC (enzymes) and membrane. Examples of congenital hemolytic anemias include sickle cell disease, thalassemia, and their variants, and hereditary spherocytosis. Acquired hemolytic anemias may result from autoimmune disease (for example, systemic lupus erythematosus) or mechanical devices (for example, heart valves, intravascular patches).

2. The hospitalizations in 107.05B do not all have to be for the same complication of the hemolytic anemia. They may be for three different complications of the disorder. Examples of complications of hemolytic anemia that may result in hospitalization include dactylitis, osteomyelitis, painful (vaso-occlusive) crisis, pulmonary infections or infarctions, acute chest syndrome, pulmonary hypertension, chronic heart failure, gallbladder disease, hepatic (liver) failure, renal (kidney) failure, nephrotic syndrome, aplastic crisis, and strokes. We will count the hours you receive emergency treatment in a comprehensive sickle cell disease center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

3. For 107.05C, we do not require hemoglobin to be measured during a period in which you are free of pain or other symptoms of your disorder. We will accept hemoglobin measurements made while you are experiencing complications of your hemolytic anemia.

4. 107.05D refers to the most serious type of beta thalassemia major in which the bone marrow cannot produce sufficient numbers of normal RBCs to maintain life. The only available treatments for beta thalassemia major are life-long RBC transfusions (sometimes called hypertransfusion) or bone marrow transplantation. For purposes of 107.05D, we do not consider prophylactic RBC transfusions to prevent strokes or other complications in sickle cell disease and its variants to be of equal significance to life-saving RBC transfusions for beta thalassemia major. However, we will consider the functional limitations associated with prophylactic

RBC transfusions and any associated side effects (for example, iron overload) under functional equivalence and any affected body system(s). We will also evaluate strokes and resulting complications under 111.00 and 112.00.

D. What are disorders of thrombosis and hemostasis, and how do we evaluate them under 107.08?

1. *Disorders of thrombosis and hemostasis* include both clotting and bleeding disorders, and may be congenital or acquired. These disorders are characterized by abnormalities in blood clotting that result in hypercoagulation (excessive blood clotting) or hypocoagulation (inadequate blood clotting). The diagnosis of a thrombosis or hemostasis disorder is based on evaluation of plasma clotting-factor proteins (factors) and platelets. Protein C or protein S deficiency and Factor V Leiden are examples of hypercoagulation disorders. Hemophilia, von Willebrand disease, and thrombocytopenia are examples of hypocoagulation disorders. Acquired excessive blood clotting may result from blood protein defects and acquired inadequate blood clotting (for example, acquired hemophilia A) may be associated with inhibitor autoantibodies.

2. The hospitalizations in 107.08 do not all have to be for the same complication of a disorder of thrombosis and hemostasis. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include anemias, thromboses, embolisms, and uncontrolled bleeding requiring multiple factor concentrate infusions or platelet transfusions. We will also consider any surgery that you have, even if it is not related to your hematological disorder, to be a complication of your disorder of thrombosis and hemostasis if you require treatment with clotting-factor proteins (for example, factor VIII or IX) or anticoagulant medication to control bleeding or coagulation in connection with your surgery. We will count the hours you receive emergency treatment in a comprehensive hemophilia treatment center immediately before the hospitalization if this treatment is comparable to the treatment provided in a hospital emergency department.

E. What are disorders of bone marrow failure, and how do we evaluate them under 107.10?

1. *Disorders of bone marrow failure* may be congenital or acquired, characterized by bone marrow that does not make enough healthy RBCs, platelets, or granulocytes (specialized types of white blood cells); there may also be a combined failure of these bone marrow-producing cells. The diagnosis is based on peripheral blood smears and bone marrow aspiration or bone marrow biopsy, but not peripheral blood smears alone. Examples of these disorders are myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis. Acquired disorders of bone marrow failure may result from viral infections, chemical exposure, or immunologic disorders.

2. The hospitalizations in 107.10A do not all have to be for the same complication of bone marrow failure. They may be for three different complications of the disorder. Examples of complications that may result in hospitalization include uncontrolled bleeding, anemia, and systemic bacterial, viral, or fungal infections.

3. For 107.10B, the requirement of life-long RBC transfusions to maintain life in myelodysplastic syndromes or aplastic anemias has the same meaning as it does for beta thalassemia major. (See 107.00C4.)

F. How do we evaluate bone marrow or stem cell transplantation under 107.17?

We will consider you to be disabled for 12 months from the date of bone marrow or stem cell transplantation, or we may consider you to be disabled for a longer period if you are experiencing any serious post-transplantation complications, such as graft-versus-host (GVH) disease, frequent infections after immunosuppressive therapy, or significant deterioration of organ systems. We do not restrict our determination of the onset of disability to the date of the transplantation in 107.17. We may establish an earlier onset of disability due to your transplantation if evidence in your case record supports such a finding.

G. How do we consider your symptoms, including your pain, severe fatigue, and malaise?

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your hematological disorder meets or medically equals a listing, or in our determination whether you otherwise have marked and severe functional limitations. We cannot consider your symptoms unless you have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 107.00 and in our other regulations. (See sections 416.928 and 416.929 of this chapter.) Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide on why

you are not receiving or following treatment.

H. How do we evaluate episodic events in hematological disorders?

Some of the listings in this body system require a specific number of events within a consecutive 12-month period. (See 107.05, 107.08, and 107.10A.) When we use such criteria, a consecutive 12-month period means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with your application or continuing disability review. These events must occur at least 30 days apart to ensure that we are evaluating separate events.

I. How do we evaluate hematological disorders that do not meet one of these listings?

1. These listings are only common examples of hematological disorders that we consider severe enough to result in marked and severe functional limitations. If your disorder does not meet the criteria of any of these listings, we must consider whether you have a disorder that satisfies the criteria of a listing in another body system. For example, we will evaluate hemophilic joint deformity under 101.00; polycythemia vera under 103.00, 104.00, or 111.00; chronic iron overload resulting from repeated RBC transfusion (transfusion hemosiderosis) under 103.00, 104.00, or 105.00; and the effects of intracranial bleeding or stroke under 111.00 or 112.00.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See section 416.926 of this chapter.) Hematological disorders may be associated with disorders in other body systems, and we consider the combined effects of multiple impairments when we determine whether they medically equal a listing. If your impairment(s) does not medically equal a listing, we will also consider whether it functionally equals the listings. (See section 416.926a of this chapter.) We use the rules in § 416.994a of this chapter when we decide whether you continue to be disabled.

107.01 Category of Impairments, Hematological Disorders

107.05 *Hemolytic anemias,* including sickle cell disease, thalassemia, and their variants (see 107.00C), with:

A. Documented painful (vaso-occlusive) crises requiring parenteral (intravenous or intramuscular) narcotic medication, occurring at least six times within a 12-month period with at least 30 days between crises.

OR

B. Complications of hemolytic anemia requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive sickle cell disease center immediately before the hospitalization (see 107.00C2).

OR

C. Hemoglobin measurements of 7.0 grams per deciliter (g/dL) or less, occurring at least three times within a 12-month period with at least 30 days between measurements.

OR

D. Beta thalassemia major requiring lifelong RBC transfusions at least once every 6 weeks to maintain life (see 107.00C4).

107.08 *Disorders of thrombosis and hemostasis,* including hemophilia and thrombocytopenia (see 107.00D), with complications requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department or comprehensive hemophilia treatment center immediately before the hospitalization (see 107.00D2).

107.10 *Disorders of bone marrow failure,* including myelodysplastic syndromes, aplastic anemia, granulocytopenia, and myelofibrosis (see 107.00E), with:

A. Complications of bone marrow failure requiring at least three hospitalizations within a 12-month period and occurring at least 30 days apart. Each hospitalization must last at least 48 hours, which can include hours in a hospital emergency department immediately before the hospitalization (see 107.00E2).

OR

B. Myelodysplastic syndromes or aplastic anemias requiring life-long RBC transfusions at least once every 6 weeks to maintain life (see 107.00E3).

107.17 *Hematological disorders treated by bone marrow or stem cell transplantation* (see 107.00F). Consider under a disability for at least 12 consecutive months from the date of transplantation. After that, evaluate any residual impairment(s) under the criteria for the affected body system.

108.00 SKIN DISORDERS

A. *What skin disorders do we evaluate with these listings?* We use these listings to evaluate skin disorders that may result from hereditary, congenital, or acquired pathological processes. The kinds of impairments covered by these listings are: Ichthyosis, bullous diseases, chronic infections of the skin or mucous membranes, dermatitis, hidradenitis suppurativa, genetic photosensitivity disorders, and burns.

B. *What documentation do we need?* When we evaluate the existence and severity of your skin disorder, we generally need information about the onset, duration, frequency of flareups, and prognosis of your skin disorder;

the location, size, and appearance of lesions; and, when applicable, history of exposure to toxins, allergens, or irritants, familial incidence, seasonal variation, stress factors, and your ability to function outside of a highly protective environment. To confirm the diagnosis, we may need laboratory findings (for example, results of a biopsy obtained independently of Social Security disability evaluation or blood tests) or evidence from other medically acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

C. *How do we assess the severity of your skin disorders(s)?* We generally base our assessment of severity on the extent of your skin lesions, the frequency of flareups of your skin lesions, how your symptoms (including pain) limit you, the extent of your treatment, and how your treatment affects you.

1. *Extensive skin lesions.* Extensive skin lesions are those that involve multiple body sites or critical body areas, and result in a very serious limitation. Examples of extensive skin lesions that result in a very serious limitation include but are not limited to:

a. Skin lesions that interfere with the motion of your joints and that very seriously limit your use of more than one extremity; that is, two upper extremities, two lower extremities, or one upper and one lower extremity.

b. Skin lesions on the palms of both hands that very seriously limit your ability to do fine and gross motor movements.

c. Skin lesions on the soles of both feet, the perineum, or both inguinal areas that very seriously limit your ability to ambulate.

2. *Frequency of flareups.* If you have skin lesions, but they do not meet the requirements of any of the listings in this body system, you may still have an impairment that results in marked and severe functional limitations when we consider your condition over time, especially if your flareups result in extensive skin lesions, as defined in C1 of this section. Therefore, if you have frequent flareups, we may find that your impairment(s) is medically equal to one of these listings even though you have some periods during which your condition is in remission. We will consider how frequent and serious your flareups are, how quickly they resolve, and how you function between flareups to determine whether you have marked and severe functional limitations that have lasted for a continuous period of at least 12 months or that can be expected to last for a continuous period of at least 12 months. We will also consider the frequency of your flareups when we determine whether you have a severe impairment and when we need to assess functional equivalence.

3. *Symptoms (including pain).* Symptoms (including pain) may be important factors contributing to the severity of your skin disorder(s). We assess the impact of symptoms as explained in §§ 404.1528, 404.1529, 416.928, and 416.929 of this chapter.

4. *Treatment.* We assess the effects of medication, therapy, surgery, and any other form of treatment you receive when we determine the severity and duration of your impairment(s). Skin disorders frequently respond to treatment; however, response to treatment can vary widely, with some impairments becoming resistant to treatment. Some treatments can have side effects that can in themselves result in limitations.

a. We assess the effects of continuing treatment as prescribed by determining if there is improvement in the symptoms, signs, and laboratory findings of your disorder, and if you experience side effects that result in functional limitations. To assess the effects of your treatment, we may need information about:

i. The treatment you have been prescribed (for example, the type, dosage, method and frequency of administration of medication or therapy);

ii. Your response to the treatment;

iii. Any adverse effects of the treatment; and

iv. The expected duration of the treatment.

b. Because treatment itself or the effects of treatment may be temporary, in most cases sufficient time must elapse to allow us to evaluate the impact and expected duration of treatment and its side effects. Except under 108.07 and 108.08, you must follow continuing treatment as prescribed for at least 3 months before your impairment can be determined to meet the requirements of a skin disorder listing. (See 108.00H if you are not undergoing treatment or did not have treatment for 3 months.) We consider your specific response to treatment when we evaluate the overall severity of your impairment.

D. *How do we assess impairments that may affect the skin and other body systems?* When your impairment affects your skin and has effects in other body systems, we first evaluate the predominant feature of your impairment under the appropriate body system. Examples include, but are not limited to the following.

1. *Tuberous sclerosis* primarily affects the brain. The predominant features are seizures, which we evaluate under the neurological listings in 111.00, and developmental delays or other mental disorders, which we evaluate under the mental disorders listings in 112.00.

2. *Malignant tumors of the skin* (for example, malignant melanoma) are cancers, or neoplastic diseases, which we evaluate under the listings in 113.00.

3. *Autoimmune disorders and other immune system disorders* (for example, systemic lupus erythematosus (SLE), scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than

one body system. We first evaluate these disorders under the immune system disorders listings in 114.00. We evaluate SLE under 114.02, scleroderma under 114.04, HIV infection under 114.08, and Sjögren's syndrome under 114.10.

4. *Disfigurement or deformity* resulting from skin lesions may result in loss of sight, hearing, speech, and the ability to chew (mastication). We evaluate these impairments and their effects under the special senses and speech listings in 102.00 and the digestive system listings in 105.00. Facial disfigurement or other physical deformities may also have effects we evaluate under the mental disorders listings in 112.00, such as when they affect mood or social functioning.

5. We evaluate *erythropoietic porphyrias* under the hemic and lymphatic listings in 107.00.

6. We evaluate *hemangiomas associated with thrombocytopenia and hemorrhage* (for example, Kasabach-Merritt syndrome) involving coagulation defects, under the hemic and lymphatic listings in 107.00. But, when hemangiomas impinge on vital structures or interfere with function, we evaluate their primary effects under the appropriate body system.

E. *How do we evaluate genetic photosensitivity disorders?*

1. *Xeroderma pigmentosum (XP).* When you have XP, your impairment meets the requirements of 108.07A if you have clinical and laboratory findings showing that you have the disorder. (See 108.00E3.) People who have XP have a lifelong hypersensitivity to all forms of ultraviolet light and generally lead extremely restricted lives in highly protective environments in order to prevent skin cancers from developing. Some people with XP also experience problems with their eyes, neurological problems, mental disorders, and problems in other body systems.

2. *Other genetic photosensitivity disorders.* Other genetic photosensitivity disorders may vary in their effects on different people, and may not result in marked and severe functional limitations for a continuous period of at least 12 months. Therefore, if you have a genetic photosensitivity disorder other than XP (established by clinical and laboratory findings as described in 108.00E3), you must show that you have either extensive skin lesions or an inability to function outside of a highly protective environment to meet the requirements of 108.07B. You must also show that your impairment meets the duration requirement. By *inability to function outside of a highly protective environment* we mean that you must avoid exposure to ultraviolet light (including sunlight passing through windows and light from unshielded fluorescent bulbs), wear protective clothing and eyeglasses, and use opaque broad-spectrum sunscreens in order to avoid skin cancer or other serious effects. Some genetic photosensitivity disorders can have very serious effects in other body systems, especially special senses and speech (102.00), neurological (111.00), mental (112.00), and neoplastic (113.00). We will evaluate the predominant feature of your impairment under the appropriate body system, as explained in 108.00D.

3. *Clinical and laboratory findings.*

a. *General.* We need documentation from an acceptable medical source, as defined in §§ 404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment. In general, we must have evidence of appropriate laboratory testing showing that you have XP or another genetic photosensitivity disorder. We will find that you have XP or another genetic photosensitivity disorder based on a report from an acceptable medical source indicating that you have the impairment, supported by definitive genetic laboratory studies documenting appropriate chromosomal changes, including abnormal DNA repair or another DNA or genetic abnormality specific to your type of photosensitivity disorder.

b. *What we will accept as medical evidence instead of the actual laboratory report.* When we do not have the actual laboratory report, we need evidence from an acceptable medical source that includes appropriate clinical findings for your impairment and that is persuasive that a positive diagnosis has been confirmed by appropriate laboratory testing at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive genetic laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

F. *How do we evaluate burns?* Electrical, chemical, or thermal burns frequently affect other body systems; for example, musculoskeletal, special senses and speech, respiratory, cardiovascular, renal, neurological, or mental. Consequently, we evaluate burns the way we evaluate other disorders that can affect the skin and other body systems, using the listing for the predominant feature of your impairment. For example, if your soft tissue injuries are under continuing surgical management (as defined in 101.00M), we will evaluate your impairment under 101.08. However, if your burns do not meet the requirements of 101.08 and you have extensive skin lesions that result in a very serious limitation (as defined in 108.00C1) that has lasted or can be expected to last for a continuous period of at least 12 months, we will evaluate them under 108.08.

G. *How do we determine if your skin disorder(s) will continue at a disabling level of severity in order to meet the duration requirement?* For all of these skin disorder listings except 108.07 and 108.08, we will find that

your impairment meets the duration requirement if your skin disorder results in extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. By *persist*, we mean that the longitudinal clinical record shows that, with few exceptions, your lesions have been at the level of severity specified in the listing. For 108.07A, we will presume that you meet the duration requirement. For 108.07B and 108.08, we will consider all of the relevant medical and other information in your case record to determine whether your skin disorder meets the duration requirement.

H. *How do we assess your skin disorder(s) if your impairment does not meet the requirements of one of these listings?*

1. These listings are only examples of common skin disorders that we consider severe enough to result in marked and severe functional limitations. For most of these listings, if you do not have continuing treatment as prescribed, if your treatment has not lasted for at least 3 months, or if you do not have extensive skin lesions that have persisted for at least 3 months, your impairment cannot meet the requirements of these skin disorder listings. (This provision does not apply to 108.07 and 108.08.) However, we may still find that you are disabled because your impairment(s) meets the requirements of a listing in another body system, medically equals (see §§ 404.1526 and 416.926 of this chapter) the severity of a listing, or functionally equals the severity of the listings.

2. If you have not received ongoing treatment or do not have an ongoing relationship with the medical community despite the existence of a severe impairment(s), or if your skin lesions have not persisted for at least 3 months but you are undergoing continuing treatment as prescribed, you may still have an impairment(s) that meets a listing in another body system or that medically equals a listing. If you do not have an impairment(s) that meets or medically equals a listing, we will consider whether your impairment(s) functionally equals the listings. (See § 416.924 of this chapter.) When we decide whether you continue to be disabled, we use the rules in § 416.994a of this chapter.

108.01 Category of Impairments, Skin Disorders

108.02 *Ichthyosis*, with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.03 *Bullous disease* (for example, pemphigus, erythema multiforme bullosum, epidermolysis bullosa, bullous pemphigoid, dermatitis herpetiformis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.04 *Chronic infections of the skin or mucous membranes*, with extensive fungating or extensive ulcerating skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.05 *Dermatitis* (for example, psoriasis, dyshidrosis, atopic dermatitis, exfoliative dermatitis, allergic contact dermatitis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.06 *Hidradenitis suppurativa*, with extensive skin lesions involving both axillae, both inguinal areas, or the perineum that persist for at least 3 months despite continuing treatment as prescribed.

108.07 *Genetic photosensitivity disorders*, established as described in 108.00E.

A. Xeroderma pigmentosum. Consider the individual disabled from birth.

B. Other genetic photosensitivity disorders, with:

1. Extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months, or

2. Inability to function outside of a highly protective environment for a continuous period of at least 12 months (see 108.00E2).

108.08 *Burns*, with extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months. (See 108.00F).

109.00 Endocrine Disorders

A. *What is an endocrine disorder?*

An endocrine disorder is a medical condition that causes a hormonal imbalance. When an endocrine gland functions abnormally, producing either too much of a specific hormone (hyperfunction) or too little (hypofunction), the hormonal imbalance can cause various complications in the body. The major glands of the endocrine system are the pituitary, thyroid, parathyroid, adrenal, and pancreas.

B. *How do we evaluate the effects of endocrine disorders?* The only listing in this body system addresses children from birth to the attainment of age 6 who have diabetes mellitus (DM) and require daily insulin. We evaluate other impairments that result from endocrine disorders under the listings for other body systems. For example:

1. *Pituitary gland disorders* can disrupt hormone production and normal functioning in other endocrine glands and in many body systems. The effects of pituitary gland disorders vary depending on which hormones are involved. For example, when pituitary growth hormone deficiency in growing children limits bone maturation and results in pathological short stature, we evaluate this linear growth impairment under 100.00. When pituitary hypofunction affects water and electrolyte balance in the kidney and leads to diabetes insipidus, we evaluate the effects of recurrent dehydration under 106.00.

2. *Thyroid gland disorders* affect the sympathetic nervous system and normal metabolism. We evaluate thyroid-related changes in

linear growth under 100.00; thyroid-related changes in blood pressure and heart rate that cause cardiac arrhythmias or other cardiac dysfunction under 104.00; thyroid-related weight loss under 105.00; and cognitive limitations, mood disorders, and anxiety under 112.00.

3. *Parathyroid gland disorders* affect calcium levels in bone, blood, nerves, muscle, and other body tissues. We evaluate parathyroid-related osteoporosis and fractures under 101.00; abnormally elevated calcium levels in the blood (hypercalcemia) that lead to cataracts under 102.00; kidney failure under 106.00; and recurrent abnormally low blood calcium levels (hypocalcemia) that lead to increased excitability of nerves and muscles, such as tetany and muscle spasms, under 111.00.

4. *Adrenal gland disorders* affect bone calcium levels, blood pressure, metabolism, and mental status. We evaluate adrenal-related linear growth impairments under 100.00; adrenal-related osteoporosis with fractures that compromises the ability to walk or to use the upper extremities under 101.00; adrenal-related hypertension that worsens heart failure or causes recurrent arrhythmias under 104.00; adrenal-related weight loss under 105.00; and mood disorders under 112.00.

5. *Diabetes mellitus and other pancreatic gland disorders* disrupt the production of several hormones, including insulin, that regulate metabolism and digestion. Insulin is essential to the absorption of glucose from the bloodstream into body cells for conversion into cellular energy. The most common pancreatic gland disorder is *diabetes mellitus* (DM). There are two major types of DM: type 1 and type 2. Both type 1 and type 2 DM are chronic disorders that can have serious, disabling complications that meet the duration requirement. Type 1 DM—previously known as ''juvenile diabetes'' or ''insulin-dependent diabetes mellitus'' (IDDM)—is an absolute deficiency of insulin secretion that commonly begins in childhood and continues throughout adulthood. Treatment of type 1 DM always requires lifelong daily insulin. With type 2 DM—previously known as ''adult-onset diabetes mellitus'' or ''non-insulin-dependent diabetes mellitus'' (NIDDM)—the body's cells resist the effects of insulin, impairing glucose absorption and metabolism. Type 2 is less common than type 1 DM in children, but physicians are increasingly diagnosing type 2 DM before age 18. Treatment of type 2 DM generally requires lifestyle changes, such as increased exercise and dietary modification, and sometimes insulin in addition to other medications. While both type 1 and type 2 DM are usually controlled, some children do not achieve good control for a variety of reasons including, but not limited to, hypoglycemia unawareness, other disorders that can affect blood glucose levels, inability to manage DM due to a mental disorder, or inadequate treatment.

a. *Hyperglycemia.* Both types of DM cause hyperglycemia, which is an abnormally high level of blood glucose that may produce acute and long-term complications. Acute complications of hyperglycemia include diabetic ketoacidosis. Long-term complications of chronic hyperglycemia include many conditions affecting various body systems but are rare in children.

b. *Diabetic ketoacidosis (DKA).* DKA is an acute, potentially life-threatening complication of DM in which the chemical balance of the body becomes dangerously hyperglycemic and acidic. It results from a severe insulin deficiency, which can occur due to missed or inadequate daily insulin therapy or in association with an acute illness. It usually requires hospital treatment to correct the acute complications of dehydration, electrolyte imbalance, and insulin deficiency. You may have serious complications resulting from your treatment, which we evaluate under the affected body system. For example, we evaluate cardiac arrhythmias under 104.00, intestinal necrosis under 105.00, and cerebral edema and seizures under 111.00. Recurrent episodes of DKA in adolescents may result from mood or eating disorders, which we evaluate under 112.00.

c. *Hypoglycemia.* Children with DM may experience episodes of hypoglycemia, which is an abnormally low level of blood glucose. Most children age 6 and older recognize the symptoms of hypoglycemia and reverse them by consuming substances containing glucose; however, some do not take this step because of hypoglycemia unawareness. Severe hypoglycemia can lead to complications, including seizures or loss of consciousness, which we evaluate under 111.00, or altered mental status, cognitive deficits, and permanent brain damage, which we evaluate under 112.00.

C. *How do we evaluate DM in children?*

Listing 109.08 is only for children with DM who have not attained age 6 and who require daily insulin. For all other children (that is, children with DM who are age 6 or older and require daily insulin, and children of any age with DM who do not require daily insulin), we follow our rules for determining whether the DM is severe, alone or in combination with another impairment, whether it meets or medically equals the criteria of a listing in another body system, or functionally equals the listings under the criteria in §416.926a, considering the factors in §416.924a. The management of DM in children can be complex and variable from day to day, and all children with DM require some level of adult supervision. For example, if a child age 6 or older has a medical need for 24-hour-a-day adult supervision of insulin treatment, food intake, and physical activity to ensure

survival, we will find that the child's impairment functionally equals the listings based on the example in §416.926a(m)(5).

D. *How do we evaluate other endocrine disorders that do not have effects that meet or medically equal the criteria of any listing in other body systems?* If your impairment(s) does not meet or medically equal a listing in another body system, we will consider whether your impairment(s) functionally equals the listings under the criteria in §416.926a, considering the factors in §416.924a. When we decide whether you continue to be disabled, we use the rules in §416.994a.

109.01 *Category of Impairments, Endocrine*

109.08 *Any type of diabetes mellitus in a child who requires daily insulin and has not attained age 6.* Consider under a disability until the attainment of age 6. Thereafter, evaluate the diabetes mellitus according to the rules in 109.00B5 and C.

110.00 Congenital Disorders That Affect Multiple Body Systems

A. *Which disorders do we evaluate under this body system?* We evaluate non-mosaic Down syndrome and catastrophic congenital disorders under this body system.

B. *What is non-mosaic Down syndrome?* Non-mosaic Down syndrome is a genetic disorder. Most children with non-mosaic Down syndrome have three copies of chromosome 21 in all of their cells (chromosome 21 trisomy); some have an extra copy of chromosome 21 attached to a different chromosome in all of their cells (chromosome 21 translocation). Virtually all children with non-mosaic Down syndrome have characteristic facial or other physical features, delayed physical development, and intellectual disability. Children with non-mosaic Down syndrome may also have congenital heart disease, impaired vision, hearing problems, and other disorders. We evaluate non-mosaic Down syndrome under 110.06. If you have non-mosaic Down syndrome documented as described in 110.00C, we consider you disabled from birth.

C. What evidence do we need to document non-mosaic Down syndrome under 110.06?

1. Under 110.06A, we will find you disabled based on laboratory findings.

a. To find that your disorder meets 110.06A, we need a copy of the laboratory report of karyotype analysis, which is the definitive test to establish non-mosaic Down syndrome. We will not purchase karyotype analysis. We will not accept a fluorescence in situ hybridization (FISH) test because it does not distinguish between the mosaic and non-mosaic forms of Down syndrome.

b. If a physician (see §§404.1513(a)(1) and 416.913(a)(1) of this chapter) has not signed the laboratory report of karyotype analysis, the evidence must also include a physician's statement that you have Down syndrome.

c. For purposes of 110.06A, we do not require evidence stating that you have the distinctive facial or other physical features of Down syndrome.

2. If we do not have a laboratory report of karyotype analysis documenting that you have non-mosaic Down syndrome, we may find you disabled under 110.06B or 110.06C.

a. Under 110.06B, we need a physician's report stating: (i) your karyotype diagnosis or evidence that documents your type of Down syndrome that is consistent with prior karyotype analysis (for example, reference to a diagnosis of ''trisomy 21'') and (ii) that you have the distinctive facial or other physical features of Down syndrome. We do not require a detailed description of the facial or other physical features of the disorder. However, we will not find that your disorder meets 110.06B if we have evidence—such as evidence of functioning inconsistent with the diagnosis—that indicates that you do not have non-mosaic Down syndrome.

b. If we do not have evidence of prior karyotype analysis (you did not have testing, or you had testing but we do not have information from a physician about the test results), we will find that your disorder meets 110.06C if we have: (i) a physician's report stating that you have the distinctive facial or other physical features of Down syndrome and (ii) evidence that your functioning is consistent with a diagnosis of non-mosaic Down syndrome. This evidence may include medical or nonmedical information about your physical and mental abilities, including information about your development, education, work history, or the results of psychological testing. However, we will not find that your disorder meets 110.06C if we have evidence—such as evidence of functioning inconsistent with the diagnosis—that indicates that you do not have non-mosaic Down syndrome.

D. *What are catastrophic congenital disorders?* Some catastrophic congenital disorders, such as anencephaly, cyclopia, chromosome 13 trisomy (Patau syndrome or trisomy D), and chromosome 18 trisomy (Edwards' syndrome or trisomy E), are usually expected to result in early death. Others such as cri du chat syndrome (chromosome 5p deletion syndrome) and the infantile onset form of Tay-Sachs disease interfere very seriously with development. We evaluate catastrophic congenital disorders under 110.08. The term ''very seriously'' in 110.08 has the same meaning as in the term ''extreme'' in §416.926a(e)(3) of this chapter.

E. *What evidence do we need under 110.08?*

We need one of the following to determine if your disorder meets 110.08A or B:

1. A laboratory report of the definitive test that documents your disorder (for example,

genetic analysis or evidence of biochemical abnormalities) signed by a physician.

2. A laboratory report of the definitive test that documents your disorder that is not signed by a physician *and* a report from a physician stating that you have the disorder.

3. A report from a physician stating that you have the disorder with the typical clinical features of the disorder and that you had definitive testing that documented your disorder. In this case, we will find that your disorder meets 110.08A or B unless we have evidence that indicates that you do not have the disorder.

4. If we do not have the definitive laboratory evidence we need under E1, E2, or E3, we will find that your disorder meets 110.08A or B if we have: (i) a report from a physician stating that you have the disorder and that you have the typical clinical features of the disorder, *and* (ii) other evidence that supports the diagnosis. This evidence may include medical or nonmedical information about your development and functioning.

5. For obvious catastrophic congenital anomalies that are expected to result in early death, such as anencephaly and cyclopia, we need evidence from a physician that demonstrates that the infant has the characteristic physical features of the disorder. In these rare cases, we do not need laboratory testing or any other evidence that confirms the disorder.

F. *How do we evaluate mosaic Down syndrome and other congenital disorders that affect multiple body systems?*

1. *Mosaic Down syndrome.* Approximately 2 percent of children with Down syndrome have the mosaic form. In mosaic Down syndrome, there are some cells with an extra copy of chromosome 21 and other cells with the normal two copies of chromosome 21. Mosaic Down syndrome can be so slight as to be undetected clinically, but it can also be profound and disabling, affecting various body systems.

2. *Other congenital disorders that affect multiple body systems.* Other congenital disorders, such as congenital anomalies, chromosomal disorders, dysmorphic syndromes, inborn metabolic syndromes, and perinatal infectious diseases, can cause deviation from, or interruption of, the normal function of the body or can interfere with development. Examples of these disorders include both the juvenile and late-onset forms of Tay-Sachs disease, trisomy X syndrome (XXX syndrome), fragile X syndrome, phenylketonuria (PKU), caudal regression syndrome, and fetal alcohol syndrome. For these disorders and other disorders like them, the degree of deviation, interruption, or interference, as well as the resulting functional limitations and their progression, may vary widely from child to child and may affect different body systems.

3. *Evaluating the effects of mosaic Down syndrome or another congenital disorder under the listings.* When the effects of mosaic Down syndrome or another congenital disorder that affects multiple body systems are sufficiently severe we evaluate the disorder under the appropriate affected body system(s), such as musculoskeletal, special senses and speech, neurological, or mental disorders. Otherwise, we evaluate the specific functional limitations that result from the disorder under our other rules described in 110.00G.

G. *What if your disorder does not meet a listing?* If you have a severe medically determinable impairment(s) that does not meet a listing, we will consider whether your impairment(s) medically equals a listing. See §416.926 of this chapter. If your impairment(s) does not meet or medically equal a listing, we will consider whether it functionally equals the listings. See §§416.924a and 416.926a of this chapter. We use the rules in §416.994a of this chapter when we decide whether you continue to be disabled.

110.01 CATEGORY OF IMPAIRMENTS, CONGENITAL DISORDERS THAT AFFECT MULTIPLE BODY SYSTEMS

110.06 *Non-mosaic Down syndrome* (chromosome 21 trisomy or chromosome 21 translocation), documented by:

A. A laboratory report of karyotype analysis signed by a physician, or both a laboratory report of karyotype analysis not signed by a physician *and* a statement by a physician that the child has Down syndrome (see 110.00C1), or

B. A physician's report stating that the child has chromosome 21 trisomy or chromosome 21 translocation consistent with karyotype analysis with the distinctive facial or other physical features of Down syndrome (see 110.00C2a), or

C. A physician's report stating that the child has Down syndrome with the distinctive facial or other physical features *and* evidence demonstrating that the child is functioning at the level of a child with non-mosaic Down syndrome (see 110.00C2b).

110.08 *A catastrophic congenital disorder* (see 110.00D and 110.00E) with:

A. Death usually expected within the first months of life, or

B. Very serious interference with development or functioning.

111.00 NEUROLOGICAL

A. *Convulsive epilepsy* must be substantiated by at least one detailed description of a typical seizure. Report of recent documentation should include a neurological examination with frequency of episodes and any associated phenomena substantiated.

Young children may have convulsions in association with febrile illnesses. Proper use

of 111.02 and 111.03 requires that epilepsy be established. Although this does not exclude consideration of seizures occurring during febrile illnesses, it does require documentation of seizures during nonfebrile periods.

There is an expected delay in control of epilepsy when treatment is started, particularly when changes in the treatment regimen are necessary. Therefore, an epileptic disorder should not be considered to meet the requirements of 111.02 or 111.03 unless it is shown that convulsive episodes have persisted more than three months after prescribed therapy began.

B. *Nonconvulsive epilepsy.* Classical petit mal seizures must be documented by characteristic EEG pattern, plus information as to age at onset and frequency of clinical seizures.

C. *Motor dysfunction.* As described in 111.06, motor dysfunction may be due to any neurological disorder. It may be due to static or progressive conditions involving any area of the nervous system and producing any type of neurological impairment. This may include weakness, spasticity, lack of coordination, ataxia, tremor, athetosis, or sensory loss. Documentation of motor dysfunction must include neurologic findings and description of type of neurologic abnormality (e.g., spasticity, weakness), as well as a description of the child's functional impairment (*i.e.*, what the child is unable to do because of the abnormality). Where a diagnosis has been made, evidence should be included for substantiation of the diagnosis (e.g., blood chemistries and muscle biopsy reports), wherever applicable.

D. *Impairment of communication.* The documentation should include a description of a recent comprehensive evaluation, including all areas of affective and effective communication, performed by a qualified professional.

E. *Brain tumors.* We evaluate malignant brain tumors under the criteria in 113.13. For benign brain tumors, we determine the severity and duration of the impairment on the basis of symptoms, signs, and laboratory findings (111.05).

111.01 Category of Impairment, Neurological

111.02 *Major motor seizure disorder.*

A. *Convulsive epilepsy.* In a child with an established diagnosis of epilepsy, the occurrence of more than one major motor seizure per month despite at least three months of prescribed treatment. With:

1. Daytime episodes (loss of consciousness and convulsive seizures); or

2. Nocturnal episodes manifesting residuals which interfere with activity during the day.

B. *Convulsive epilepsy syndrome.* In a child with an established diagnosis of epilepsy, the occurrence of at least one major motor seizure in the year prior to application despite at least three months of prescribed treatment. And one of the following:

1. IQ of 70 or less; or

2. Significant interference with communication due to speech, hearing, or visual defect; or

3. Significant mental disorder; or

4. Where significant adverse effects of medication interfere with major daily activities.

111.03 *Nonconvulsive epilepsy.* In a child with an established seizure disorder, the occurrence of more than one minor motor seizure per week, with alteration of awareness or loss of consciousness, despite at least three months of prescribed treatment.

111.05 *Benign brain tumors.* Evaluate under 111.02, 111.03, 111.06, 111.09 or the criteria of the affected body system.

111.06 *Motor dysfunction (due to any neurological disorder).* Persistent disorganization or deficit of motor function for age involving two extremities, which (despite prescribed therapy) interferes with age-appropriate major daily activities and results in disruption of:

A. Fine and gross movements; or

B. Gait and station.

111.07 *Cerebral Palsy.* With:

A. Motor dysfunction meeting the requirements of 101.02 or 111.06; or

B. Less severe motor dysfunction (but more than slight) and one of the following:

1. IQ of 70 or less; or

2. Seizure disorder, with at least one major motor seizure in the year prior to application; or

3. Significant interference with communication due to speech, hearing or visual defect; or

4. Significant emotional disorder.

111.08 *Meningomyelocele (and related disorders).* With one of the following despite prescribed treatment:

A. Motor dysfunction meeting the requirements of 101.02 or 111.06; or

B. Less severe motor dysfunction (but more than slight), and:

1. Urinary or fecal incontinence when inappropriate for age; or

2. IQ of 70 or less; or

C. Four extremity involvement; or

D. Noncompensated hydrocephalus producing interference with mental or motor developmental progression.

111.09 *Communication impairment, associated with documented neurological disorder.* And one of the following:

A. Documented speech deficit which significantly affects the clarity and content of the speech; or

B. Documented comprehension deficit resulting in ineffective verbal communication for age; or

C. Impairment of hearing as described under the criteria in 102.10 or 102.11.

112.00 MENTAL DISORDERS

A. *Introduction:* The structure of the mental disorders listings for children under age 18 parallels the structure for the mental disorders listings for adults but is modified to reflect the presentation of mental disorders in children. The listings for mental disorders in children are arranged in 11 diagnostic categories: Organic mental disorders (112.02); schizophrenic, delusional (paranoid), schizoaffective, and other psychotic disorders (112.03); mood disorders (112.04); intellectual disability (112.05); anxiety disorders (112.06); somatoform, eating, and tic disorders (112.07); personality disorders (112.08); psychoactive substance dependence disorders (112.09); autistic disorder and other pervasive developmental disorders (112.10); attention deficit hyperactivity disorder (112.11); and developmental and emotional disorders of newborn and younger infants (112.12).

There are significant differences between the listings for adults and the listings for children. There are disorders found in children that have no real analogy in adults; hence, the differences in the diagnostic categories for children. The presentation of mental disorders in children, particularly the very young child, may be subtle and of a character different from the signs and symptoms found in adults. For example, findings such as separation anxiety, failure to mold or bond with the parents, or withdrawal may serve as findings comparable to findings that mark mental disorders in adults. The activities appropriate to children, such as learning, growing, playing, maturing, and school adjustment, are also different from the activities appropriate to the adult and vary widely in the different childhood stages.

Each listing begins with an introductory statement that describes the disorder or disorders addressed by the listing. This is followed (except in listings 112.05 and 112.12) by paragraph A criteria (a set of medical findings) and paragraph B criteria (a set of impairment-related functional limitations). An individual will be found to have a listed impairment when the criteria of both paragraphs A and B of the listed impairment are satisfied.

The purpose of the criteria in paragraph A is to substantiate medically the presence of a particular mental disorder. Specific symptoms and signs under any of the listings 112.02 through 112.12 cannot be considered in isolation from the description of the mental disorder contained at the beginning of each listing category. Impairments should be analyzed or reviewed under the mental category(ies) indicated by the medical findings.

Paragraph A of the listings is a composite of medical findings which are used to substantiate the existence of a disorder and may or may not be appropriate for children at specific developmental stages. However, a range of medical findings is included in the listings so that no age group is excluded. For example, in listing 112.02A7, emotional lability and crying would be inappropriate criteria to apply to older infants and toddlers, age 1 to attainment of age 3; whereas in 112.02A1, developmental arrest, delay, or regression are appropriate criteria for older infants and toddlers. Whenever the adjudicator decides that the requirements of paragraph A of a particular mental listing are satisfied, then that listing should be applied regardless of the age of the child to be evaluated.

The purpose of the paragraph B criteria is to describe impairment-related functional limitations which are applicable to children. Standardized tests of social or cognitive function and adaptive behavior are frequently available and appropriate for the evaluation of children and, thus, such tests are included in the paragraph B functional parameters. The functional restrictions in paragraph B must be the result of the mental disorder which is manifested by the medical findings in paragraph A.

We did not include separate C criteria for listings 112.02, 112.03, 112.04, and 112.06, as are found in the adult listings, because for the most part we do not believe that the residual disease processes described by these listings are commonly found in children. However, in unusual cases where these disorders are found in children and are comparable to the severity and duration found in adults, we may use the adult listings 12.02C, 12.03C, 12.04C, and 12.06C criteria to evaluate such cases.

The structure of the listings for Intellectual Disability (112.05) and Developmental and Emotional Disorders of Newborn and Younger Infants (112.12) is different from that of the other mental disorders. Listing 112.05 (Intellectual Disability) contains six sets of criteria. If an impairment satisfies the diagnostic description in the introductory paragraph and any one of the six sets of criteria, we will find that the child's impairment meets the listing. For listings 112.05D and 112.05F, we will assess the degree of functional limitation the additional impairment(s) imposes to determine if it causes more than minimal functional limitations, *i.e.*, is a ''severe'' impairment(s), as defined in § 416.924(c). If the additional impairment(s) does not cause limitations that are ''severe'' as defined in § 416.924(c), we will not find that the additional impairment(s) imposes an additional and significant limitation of function. Listing 112.12 (Developmental and Emotional Disorders of Newborn and Younger Infants) contains five criteria, any one of which, if satisfied, will result in a finding that the infant's impairment meets the listing.

It must be remembered that these listings are only examples of common mental disorders that are severe enough to find a child disabled. When a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will make a determination whether the child's impairment(s) medically or functionally equals the listings. (See §§ 404.1526, 416.926, and 416.926a.) This determination can be especially important in older infants and toddlers (age 1 to attainment of age 3), who may be too young for identification of a specific diagnosis, yet demonstrate serious functional limitations. Therefore, the determination of equivalency is necessary to the evaluation of any child's case when the child does not have an impairment that meets a listing.

B. *Need for Medical Evidence:* The existence of a medically determinable impairment of the required duration must be established by medical evidence consisting of symptoms, signs, and laboratory findings (including psychological or developmental test findings). Symptoms are complaints presented by the child. Psychiatric signs are medically demonstrable phenomena that indicate specific psychological abnormalities, e.g., abnormalities of behavior, mood, thought, memory, orientation, development, or perception, as described by an appropriate medical source. Symptoms and signs generally cluster together to constitute recognizable mental disorders described in paragraph A of the listings. These findings may be intermittent or continuous depending on the nature of the disorder.

C. *Assessment of Severity:* In childhood cases, as with adults, severity is measured according to the functional limitations imposed by the medically determinable mental impairment. However, the range of functions used to assess impairment severity for children varies at different stages of maturation. The functional areas that we consider are: Motor function; cognitive/communicative function; social function; personal function; and concentration, persistence, or pace. In most functional areas, there are two alternative methods of documenting the required level of severity: (1) Use of standardized tests alone, where appropriate test instruments are available, and (2) use of other medical findings. (See 112.00D for explanation of these documentation requirements.) The use of standardized tests is the preferred method of documentation if such tests are available.

Newborn and younger infants (birth to attainment of age 1) have not developed sufficient personality differentiation to permit formulation of appropriate diagnoses. We have, therefore, assigned listing 112.12 for Developmental and Emotional Disorders of Newborn and Younger Infants for the evaluation of mental disorders of such children. Severity of these disorders is based on measures of development in motor, cognitive/communicative, and social functions. When older infants and toddlers (age 1 to attainment of age 3) do not clearly satisfy the paragraph A criteria of any listing because of insufficient developmental differentiation, they must be evaluated under the rules for equivalency. The principles for assessing the severity of impairment in such children, described in the following paragraphs, must be employed.

Generally, when we assess the degree of developmental delay imposed by a mental impairment, we will use an infant's or toddler's chronological age; *i.e.*, the child's age based on birth date. If the infant or toddler was born prematurely, however, we will follow the rules in § 416.924b(b) to determine whether we should use the infant's or toddler's corrected chronological age; *i.e.*, the chronological age adjusted by the period of gestational prematurity.

In defining the severity of functional limitations, two different sets of paragraph B criteria corresponding to two separate age groupings have been established, in addition to listing 112.12, which is for children who have not attained age 1. These age groups are: older infants and toddlers (age 1 to attainment of age 3) and children (age 3 to attainment of age 18). However, the discussion below in 112.00C1, 2, 3, and 4, on the age-appropriate areas of function, is broken down into four age groupings: older infants and toddlers (age 1 to attainment of age 3), preschool children (age 3 to attainment of age 6), primary school children (age 6 to attainment of age 12), and adolescents (age 12 to attainment of age 18). This was done to provide specific guidance on the age group variances in disease manifestations and methods of evaluation.

Where ''marked'' is used as a standard for measuring the degree of limitation it means more than moderate but less than extreme. A marked limitation may arise when several activities or functions are impaired, or even when only one is impaired, as long as the degree of limitation is such as to interfere seriously with the ability to function (based upon age-appropriate expectations) independently, appropriately, effectively, and on a sustained basis. When standardized tests are used as the measure of functional parameters, a valid score that is two standard deviations below the norm for the test will be considered a marked restriction.

1. *Older infants and toddlers (age 1 to attainment of age 3).* In this age group, impairment severity is assessed in three areas: (a) Motor development, (b) cognitive/communicative function, and (c) social function.

a. *Motor development.* Much of what we can discern about mental function in these children frequently comes from observation of

the degree of development of fine and gross motor function. Developmental delay, as measured by a good developmental milestone history confirmed by medical examination, is critical. This information will ordinarily be available in the existing medical evidence from the claimant's treating sources and other medical sources, supplemented by information from nonmedical sources, such as parents, who have observed the child and can provide pertinent historical information. It may also be available from standardized testing. If the delay is such that the older infant or toddler has not achieved motor development generally acquired by children no more than one-half the child's chronological age, the criteria are satisfied.

b. *Cognitive/communicative function.* Cognitive/communicative function is measured using one of several standardized infant scales. Appropriate tests for the measure of such function are discussed in 112.00D. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases, the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

For older infants and toddlers, alternative criteria covering disruption in communication as measured by their capacity to use simple verbal and nonverbal structures to communicate basic needs are provided.

c. *Social function.* Social function in older infants and toddlers is measured in terms of the development of relatedness to people (e.g., bonding and stranger anxiety) and attachment to animate or inanimate objects. Criteria are provided that use standard social maturity scales or alternative criteria that describe marked impairment in socialization.

2. *Preschool children (age 3 to attainment of age 6).* For the age groups including preschool children through adolescence, the functional areas used to measure severity are: (a) Cognitive/communicative function, (b) social function, (c) personal function, and (d) deficiencies of concentration, persistence, or pace resulting in frequent failure to complete tasks in a timely manner. After 36 months, motor function is no longer felt to be a primary determinant of mental function, although, of course, any motor abnormalities should be documented and evaluated.

a. *Cognitive/communicative function.* In the preschool years and beyond, cognitive function can be measured by standardized tests of intelligence, although the appropriate instrument may vary with age. A primary criterion for limited cognitive function is a valid verbal, performance, or full scale IQ of 70 or less. The listings also provide alternative criteria, consisting of tests of language development or bizarre speech patterns.

b. *Social function.* Social functioning refers to a child's capacity to form and maintain relationships with parents, other adults, and peers. Social functioning includes the ability to get along with others (e.g., family members, neighborhood friends, classmates, teachers). Impaired social functioning may be caused by inappropriate externalized actions (e.g., running away, physical aggression—but not self-injurious actions, which are evaluated in the personal area of functioning), or inappropriate internalized actions (e.g., social isolation, avoidance of interpersonal activities, mutism). Its severity must be documented in terms of intensity, frequency, and duration, and shown to be beyond what might be reasonably expected for age. Strength in social functioning may be documented by such things as the child's ability to respond to and initiate social interaction with others, to sustain relationships, and to participate in group activities. Cooperative behaviors, consideration for others, awareness of others' feelings, and social maturity, appropriate to a child's age, also need to be considered. Social functioning in play and school may involve interactions with adults, including responding appropriately to persons in authority (e.g., teachers, coaches) or cooperative behaviors involving other children. Social functioning is observed not only at home but also in preschool programs.

c. *Personal function.* Personal functioning in preschool children pertains to self-care; *i.e.*, personal needs, health, and safety (feeding, dressing, toileting, bathing; maintaining personal hygiene, proper nutrition, sleep, health habits; adhering to medication or therapy regimens; following safety precautions). Development of self-care skills is measured in terms of the child's increasing ability to help himself/herself and to cooperate with others in taking care of these needs. Impaired ability in this area is manifested by failure to develop such skills, failure to use them, or self-injurious actions. This function may be documented by a standardized test of adaptive behavior or by a careful description of the full range of self-care activities. These activities are often observed not only at home but also in preschool programs.

d. *Concentration, persistence, or pace.* This function may be measured through observations of the child in the course of standardized testing and in the course of play.

3. *Primary school children (age 6 to attainment of age 12).* The measures of function here are similar to those for preschool-age children except that the test instruments may change and the capacity to function in the school setting is supplemental information. Standardized measures of academic achievement, e.g., Wide Range Achievement

Test-Revised, Peabody Individual Achievement Test, etc., may be helpful in assessing cognitive impairment. Problems in social functioning, especially in the area of peer relationships, are often observed firsthand by teachers and school nurses. As described in 112.00D, *Documentation,* school records are an excellent source of information concerning function and standardized testing and should always be sought for school-age children.

As it applies to primary school children, the intent of the functional criterion described in paragraph B2d, *i.e.*, deficiencies of concentration, persistence, or pace resulting in failure to complete tasks in a timely manner, is to identify the child who cannot adequately function in primary school because of a mental impairment. Although grades and the need for special education placement are relevant factors which must be considered in reaching a decision under paragraph B2d, they are not conclusive. There is too much variability from school district to school district in the expected level of grading and in the criteria for special education placement to justify reliance solely on these factors.

4. *Adolescents (age 12 to attainment of age 18).* Functional criteria parallel to those for primary school children (cognitive/communicative; social; personal; and concentration, persistence, or pace) are the measures of severity for this age group. Testing instruments appropriate to adolescents should be used where indicated. Comparable findings of disruption of social function must consider the capacity to form appropriate, stable, and lasting relationships. If information is available about cooperative working relationships in school or at part-time or full-time work, or about the ability to work as a member of a group, it should be considered when assessing the child's social functioning. Markedly impoverished social contact, isolation, withdrawal, and inappropriate or bizarre behavior under the stress of socializing with others also constitute comparable findings. (Note that self-injurious actions are evaluated in the personal area of functioning.)

a. Personal functioning in adolescents pertains to self-care. It is measured in the same terms as for younger children, the focus, however, being on the adolescent's ability to take care of his or her own personal needs, health, and safety without assistance. Impaired ability in this area is manifested by failure to take care of these needs or by self-injurious actions. This function may be documented by a standardized test of adaptive behavior or by careful descriptions of the full range of self-care activities.

b. In adolescents, the intent of the functional criterion described in paragraph B2d is the same as in primary school children, However, other evidence of this functional impairment may also be available, such as from evidence of the child's performance in work or work-like settings.

D. *Documentation:* 1. The presence of a mental disorder in a child must be documented on the basis of reports from acceptable sources of medical evidence. See §§404.1513 and 416.913. Descriptions of functional limitations may be available from these sources, either in the form of standardized test results or in other medical findings supplied by the sources, or both. (Medical findings consist of symptoms, signs, and laboratory findings.) Whenever possible, a medical source's findings should reflect the medical source's consideration of information from parents or other concerned individuals who are aware of the child's activities of daily living, social functioning, and ability to adapt to different settings and expectations, as well as the medical source's findings and observations on examination, consistent with standard clinical practice. As necessary, information from nonmedical sources, such as parents, should also be used to supplement the record of the child's functioning to establish the consistency of the medical evidence and longitudinality of impairment severity.

2. For some newborn and younger infants, it may be very difficult to document the presence or severity of a mental disorder. Therefore, with the exception of some genetic diseases and catastrophic congenital anomalies, it may be necessary to defer making a disability decision until the child attains age 3 months of age in order to obtain adequate observation of behavior or affect. See, also, 110.00 of this part. This period could be extended in cases of premature infants depending on the degree of prematurity and the adequacy of documentation of their developmental and emotional status.

3. For infants and toddlers, programs of early intervention involving occupational, physical, and speech therapists, nurses, social workers, and special educators, are a rich source of data. They can provide the developmental milestone evaluations and records on the fine and gross motor functioning of these children. This information is valuable and can complement the medical examination by a physician or psychologist. A report of an interdisciplinary team that contains the evaluation and signature of an acceptable medical source is considered acceptable medical evidence rather than supplemental data.

4. In children with mental disorders, particularly those requiring special placement, school records are a rich source of data, and the required reevaluations at specified time periods can provide the longitudinal data needed to trace impairment progression over time.

5. In some cases where the treating sources lack expertise in dealing with mental disorders of children, it may be necessary to obtain evidence from a psychiatrist, psychologist, or pediatrician with experience and skill in the diagnosis and treatment of mental disorders as they appear in children. In these cases, however, every reasonable effort must be made to obtain the records of the treating sources, since these records will help establish a longitudinal picture that cannot be established through a single purchased examination.

6. Reference to a ''standardized psychological test'' indicates the use of a psychological test measure that has appropriate validity, reliability, and norms, and is individually administered by a qualified specialist. By ''qualified,'' we mean the specialist must be currently licensed or certified in the State to administer, score, and interpret psychological tests and have the training and experience to perform the test.

7. Psychological tests are best considered as standardized sets of tasks or questions designed to elicit a range of responses. Psychological testing can also provide other useful data, such as the specialist's observations regarding the child's ability to sustain attention and concentration, relate appropriately to the specialist, and perform tasks independently (without prompts or reminders). Therefore, a report of test results should include both the objective data and any clinical observations.

8. The salient characteristics of a good test are: (1) Validity, *i.e.*, the test measures what it is supposed to measure; (2) reliability, *i.e.*, the consistency of results obtained over time with the same test and the same individual; (3) appropriate normative data, *i.e.*, individual test scores can be compared to test data from other individuals or groups of a similar nature, representative of that population; and (4) wide scope of measurement, *i.e.*, the test should measure a broad range of facets/aspects of the domain being assessed. In considering the validity of a test result, we should note and resolve any discrepancies between formal test results and the child's customary behavior and daily activities.

9. Identical IQ scores obtained from different tests do not always reflect a similar degree of intellectual functioning. The IQ scores in listing 112.05 reflect values from tests of general intelligence that have a mean of 100 and a standard deviation of 15, e.g., the Wechsler series. IQs obtained from standardized tests that deviate significantly from a mean of 100 and standard deviation of 15 require conversion to a percentile rank so that the actual degree of limitation reflected by the IQ scores can be determined. In cases where more than one IQ is customarily derived from the test administered, e.g., where verbal, performance, and full scale IQs are provided in the Wechsler series, the lowest of these is used in conjunction with listing 112.05.

10. IQ test results must also be sufficiently current for accurate assessment under 112.05. Generally, the results of IQ tests tend to stabilize by the age of 16. Therefore, IQ test results obtained at age 16 or older should be viewed as a valid indication of the child's current status, provided they are compatible with the child's current behavior. IQ test results obtained between ages 7 and 16 should be considered current for 4 years when the tested IQ is less than 40, and for 2 years when the IQ is 40 or above. IQ test results obtained before age 7 are current for 2 years if the tested IQ is less than 40 and 1 year if at 40 or above.

11. Standardized intelligence test results are essential to the adjudication of all cases of intellectual disability that are not covered under the provisions of listings 112.05A, 112.05B, and 112.05F. Listings 112.05A, 112.05B, and 112.05F may be the bases for adjudicating cases where the results of standardized intelligence tests are unavailable, e.g., where the child's young age or condition precludes formal standardized testing.

12. In conjunction with clinical examinations, sources may report the results of screening tests, *i.e.*, tests used for gross determination of level of functioning. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

13. Where reference is made to developmental milestones, this is defined as the attainment of particular mental or motor skills at an age-appropriate level, *i.e.*, the skills achieved by an infant or toddler sequentially and within a given time period in the motor and manipulative areas, in general understanding and social behavior, in self-feeding, dressing, and toilet training, and in language. This is sometimes expressed as a developmental quotient (DQ), the relation between developmental age and chronological age as determined by specific standardized measurements and observations. Such tests include, but are not limited to, the Cattell Infant Intelligence Scale, the Bayley Scales of Infant Development, and the Revised Stanford-Binet. Formal tests of the attainment of developmental milestones are generally used in the clinical setting for determination of the developmental status of infants and toddlers.

14. Formal psychological tests of cognitive functioning are generally in use for preschool children, for primary school children, and for adolescents except for those instances noted below.

15. Generally, it is preferable to use IQ measures that are wide in scope and include

items that test both verbal and performance abilities. However, in special circumstances, such as the assessment of children with sensory, motor, or communication abnormalities, or those whose culture and background are not principally English-speaking, measures such as the Test of Nonverbal Intelligence, Third Edition (TONI–3), Leiter International Performance Scale-Revised (Leiter-R), or Peabody Picture Vocabulary Test—Third Edition (PPVT-III) may be used.

16. We may consider exceptions for formal standardized psychological testing when an individual qualified by training and experience to perform such an evaluation is not available, or in cases where appropriate standardized measures for the child's social, linguistic, and cultural background are not available. In these cases, the best indicator of severity is often the level of adaptive functioning and how the child performs activities of daily living and social functioning.

17. Comprehensive neuropsychological examinations may be used to establish the existence and extent of compromise of brain function, particularly in cases involving organic mental disorders. Normally these examinations include assessment of cerebral dominance, basic sensation and perception, motor speed and coordination, attention and concentration, visual-motor function, memory across verbal and visual modalities, receptive and expressive speech, higher-order linguistic operations, problem-solving, abstraction ability, and general intelligence. In addition, there should be a clinical interview geared toward evaluating pathological features known to occur frequently in neurological disease and trauma, e.g., emotional lability, abnormality of mood, impaired impulse control, passivity and apathy, or inappropriate social behavior. The specialist performing the examination may administer one of the commercially available comprehensive neuropsychological batteries, such as the Luria-Nebraska or Halstead-Reitan, or a battery of tests selected as relevant to the suspected brain dysfunction. The specialist performing the examination must be properly trained in this area of neuroscience.

E. *Effect of Hospitalization or Residential Placement:* As with adults, children with mental disorders may be placed in a variety of structured settings outside the home as part of their treatment. Such settings include, but are not limited to, psychiatric hospitals, developmental disabilities facilities, residential treatment centers and schools, community-based group homes, and workshop facilities. The reduced mental demands of such structured settings may attenuate overt symptomatology and superficially make the child's level of adaptive functioning appear better than it is. Therefore, the capacity of the child to function outside highly structured settings must be considered in evaluating impairment severity. This is done by determining the degree to which the child can function (based upon age-appropriate expectations) independently, appropriately, effectively, and on a sustained basis outside the highly structured setting.

On the other hand, there may be a variety of causes for placement of a child in a structured setting which may or may not be directly related to impairment severity and functional ability. Placement in a structured setting in and of itself does not equate with a finding of disability. The severity of the impairment must be compared with the requirements of the appropriate listing.

F. *Effects of Medication:* Attention must be given to the effect of medication on the child's signs, symptoms, and ability to function. While drugs used to modify psychological functions and mental states may control certain primary manifestations of a mental disorder, e.g., hallucinations, impaired attention, restlessness, or hyperactivity, such treatment may not affect all functional limitations imposed by the mental disorder. In cases where overt symptomatology is attenuated by the use of such drugs, particular attention must be focused on the functional limitations that may persist. These functional limitations must be considered in assessing impairment severity.

Psychotropic medicines used in the treatment of some mental illnesses may cause drowsiness, blunted affect, or other side effects involving other body systems. Such side effects must be considered in evaluating overall impairment severity.

112.01 Category of Impairments, Mental

112.02 *Organic Mental Disorders:* Abnormalities in perception, cognition, affect, or behavior associated with dysfunction of the brain. The history and physical examination or laboratory tests, including psychological or neuropsychological tests, demonstrate or support the presence of an organic factor judged to be etiologically related to the abnormal mental state and associated deficit or loss of specific cognitive abilities, or affective changes, or loss of previously acquired functional abilities.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence of at least one of the following:

1. Developmental arrest, delay or regression; or

2. Disorientation to time and place; or

3. Memory impairment, either short-term (inability to learn new information), intermediate, or long-term (inability to remember information that was known sometime in the past); or

4. Perceptual or thinking disturbance (e.g., hallucinations, delusions, illusions, or paranoid thinking); or

5. Disturbance in personality (e.g., apathy, hostility); or

6. Disturbance in mood (e.g., mania, depression); or

7. Emotional lability (e.g., sudden crying); or

8. Impairment of impulse control (e.g., disinhibited social behavior, explosive temper outbursts); or

9. Impairment of cognitive function, as measured by clinically timely standardized psychological testing; or

10. Disturbance of concentration, attention, or judgment;

AND

B. Select the appropriate age group to evaluate the severity of the impairment:

1. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the following:

a. Gross or fine motor development at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings (see 112.00C); or

b. Cognitive/communicative function at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings of equivalent cognitive/communicative abnormality, such as the inability to use simple verbal or nonverbal behavior to communicate basic needs or concepts; or

c. Social function at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings of an equivalent abnormality of social functioning, exemplified by serious inability to achieve age-appropriate autonomy as manifested by excessive clinging or extreme separation anxiety; or

d. Attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in two or more areas covered by a., b., or c., as measured by an appropriate standardized test or other appropriate medical findings.

2. For children (age 3 to attainment of age 18), resulting in at least two of the following:

a. Marked impairment in age-appropriate cognitive/communicative function, documented by medical findings (including consideration of historical and other information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, the results of appropriate standardized psychological tests, or for children under age 6, by appropriate tests of language and communication; or

b. Marked impairment in age-appropriate social functioning, documented by history and medical findings (including consideration of information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, the results of appropriate standardized tests; or

c. Marked impairment in age-appropriate personal functioning, documented by history and medical findings (including consideration of information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, appropriate standardized tests; or

d. Marked difficulties in maintaining concentration, persistence, or pace.

112.03 *Schizophrenic, Delusional (Paranoid), Schizoaffective, and Other Psychotic Disorders:* Onset of psychotic features, characterized by a marked disturbance of thinking, feeling, and behavior, with deterioration from a previous level of functioning or failure to achieve the expected level of social functioning.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence, for at least 6 months, either continuous or intermittent, of one or more of the following:

1. Delusions or hallucinations; or

2. Catatonic, bizarre, or other grossly disorganized behavior; or

3. Incoherence, loosening of associations, illogical thinking, or poverty of content of speech; or

4. Flat, blunt, or inappropriate affect; or

5. Emotional withdrawal, apathy, or isolation;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.04 *Mood Disorders:* Characterized by a disturbance of mood (referring to a prolonged emotion that colors the whole psychic life, generally involving either depression or elation), accompanied by a full or partial manic or depressive syndrome.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one of the following:

1. Major depressive syndrome, characterized by at least five of the following, which must include either depressed or irritable mood or markedly diminished interest or pleasure:

a. Depressed or irritable mood; or
b. Markedly diminished interest or pleasure in almost all activities; or
c. Appetite or weight increase or decrease, or failure to make expected weight gains; or
d. Sleep disturbance; or
e. Psychomotor agitation or retardation; or
f. Fatigue or loss of energy; or
g. Feelings of worthlessness or guilt; or
h. Difficulty thinking or concentrating; or
i. Suicidal thoughts or acts; or
j. Hallucinations, delusions, or paranoid thinking;

OR

2. Manic syndrome, characterized by elevated, expansive, or irritable mood, and at least three of the following:
a. Increased activity or psychomotor agitation; or
b. Increased talkativeness or pressure of speech; or
c. Flight of ideas or subjectively experienced racing thoughts; or
d. Inflated self-esteem or grandiosity; or
e. Decreased need for sleep; or
f. Easy distractibility; or
g. Involvement in activities that have a high potential of painful consequences which are not recognized; or
h. Hallucinations, delusions, or paranoid thinking;

OR

3. Bipolar or cyclothymic syndrome with a history of episodic periods manifested by the full symptomatic picture of both manic and depressive syndromes (and currently or most recently characterized by the full or partial symptomatic picture of either or both syndromes);

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.05 *Intellectual Disability:* Characterized by significantly subaverage general intellectual functioning with deficits in adaptive functioning.

The required level of severity for this disorder is met when the requirements in A, B, C, D, E, or F are satisfied.

A. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02;

OR

B. Mental incapacity evidenced by dependence upon others for personal needs (grossly in excess of age-appropriate dependence) and inability to follow directions such that the use of standardized measures of intellectual functioning is precluded;

OR

C. A valid verbal, performance, or full scale IQ of 59 or less;

OR

D. A valid verbal, performance, or full scale IQ of 60 through 70 and a physical or other mental impairment imposing an additional and significant limitation of function;

OR

E. A valid verbal, performance, or full scale IQ of 60 through 70 and:
1. For older infants and toddlers (age 1 to attainment of age 3), resulting in attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in either paragraphs B1a or B1c of 112.02; or
2. For children (age 3 to attainment of age 18), resulting in at least one of paragraphs B2b or B2c or B2d of 112.02;

OR

F. Select the appropriate age group:
1. For older infants and toddlers (age 1 to attainment of age 3), resulting in attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in paragraph B1b of 112.02, and a physical or other mental impairment imposing an additional and significant limitation of function;
OR
2. For children (age 3 to attainment of age 18), resulting in the satisfaction of 112.02B2a, and a physical or other mental impairment imposing an additional and significant limitation of function.

112.06 *Anxiety Disorders:* In these disorders, anxiety is either the predominant disturbance or is experienced if the individual attempts to master symptoms, e.g., confronting the dreaded object or situation in a phobic disorder, attempting to go to school in a separation anxiety disorder, resisting the obsessions or compulsions in an obsessive compulsive disorder, or confronting strangers or peers in avoidant disorders.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of at least one of the following:
1. Excessive anxiety manifested when the child is separated, or separation is threatened, from a parent or parent surrogate; or
2. Excessive and persistent avoidance of strangers; or
3. Persistent unrealistic or excessive anxiety and worry (apprehensive expectation), accompanied by motor tension, autonomic hyperactivity, or vigilance and scanning; or

4. A persistent irrational fear of a specific object, activity, or situation which results in a compelling desire to avoid the dreaded object, activity, or situation; or

5. Recurrent severe panic attacks, manifested by a sudden unpredictable onset of intense apprehension, fear, or terror, often with a sense of impending doom, occurring on the average of at least once a week; or

6. Recurrent obsessions or compulsions which are a source of marked distress; or

7. Recurrent and intrusive recollections of a traumatic experience, including dreams, which are a source of marked distress;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.07 *Somatoform, Eating, and Tic Disorders:* Manifested by physical symptoms for which there are no demonstrable organic findings or known physiologic mechanisms; or eating or tic disorders with physical manifestations.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of one of the following:

1. An unrealistic fear and perception of fatness despite being underweight, and persistent refusal to maintain a body weight which is greater than 85 percent of the average weight for height and age, as shown in the most recent edition of the *Nelson Textbook of Pediatrics*, Richard E. Behrman and Victor C. Vaughan, III, editors, Philadelphia: W. B. Saunders Company; or

2. Persistent and recurrent involuntary, repetitive, rapid, purposeless motor movements affecting multiple muscle groups with multiple vocal tics; or

3. Persistent nonorganic disturbance of one of the following:

a. Vision; or

b. Speech; or

c. Hearing; or

d. Use of a limb; or

e. Movement and its control (e.g., coordination disturbance, psychogenic seizures); or

f. Sensation (diminished or heightened); or

g. Digestion or elimination; or

4. Preoccupation with a belief that one has a serious disease or injury;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.08 *Personality Disorders:* Manifested by pervasive, inflexible, and maladaptive personality traits, which are typical of the child's long-term functioning and not limited to discrete episodes of illness.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Deeply ingrained, maladaptive patterns of behavior, associated with one of the following:

1. Seclusiveness or autistic thinking; or

2. Pathologically inappropriate suspiciousness or hostility; or

3. Oddities of thought, perception, speech, and behavior; or

4. Persistent disturbances of mood or affect; or

5. Pathological dependence, passivity, or aggressiveness; or

6. Intense and unstable interpersonal relationships and impulsive and exploitative behavior; or

7. Pathological perfectionism and inflexibility;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.09 *Psychoactive Substance Dependence Disorders:* Manifested by a cluster of cognitive, behavioral, and physiologic symptoms that indicate impaired control of psychoactive substance use with continued use of the substance despite adverse consequences.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of at least four of the following:

1. Substance taken in larger amounts or over a longer period than intended and a great deal of time is spent in recovering from its effects; or

2. Two or more unsuccessful efforts to cut down or control use; or

3. Frequent intoxication or withdrawal symptoms interfering with major role obligations; or

4. Continued use despite persistent or recurring social, psychological, or physical problems; or

5. Tolerance, as characterized by the requirement for markedly increased amounts of substance in order to achieve intoxication; or

6. Substance taken to relieve or avoid withdrawal symptoms;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least

one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.10 *Autistic Disorder and Other Pervasive Developmental Disorders:* Characterized by qualitative deficits in the development of reciprocal social interaction, in the development of verbal and nonverbal communication skills, and in imaginative activity. Often, there is a markedly restricted repertoire of activities and interests, which frequently are stereotyped and repetitive.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of the following:

1. For autistic disorder, all of the following:

a. Qualitative deficits in the development of reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity; and

c. Markedly restricted repertoire of activities and interests;

OR

2. For other pervasive developmental disorders, both of the following:

a. Qualitative deficits in the development of reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraphs B2 of 112.02.

112.11 *Attention Deficit Hyperactivity Disorder:* Manifested by developmentally inappropriate degrees of inattention, impulsiveness, and hyperactivity.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of all three of the following:

1. Marked inattention; and

2. Marked impulsiveness; and

3. Marked hyperactivity;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.12 *Developmental and Emotional Disorders of Newborn and Younger Infants (Birth to attainment of age 1):* Developmental or emotional disorders of infancy are evidenced by a deficit or lag in the areas of motor, cognitive/communicative, or social functioning. These disorders may be related either to organic or to functional factors or to a combination of these factors.

The required level of severity for these disorders is met when the requirements of A, B, C, D, or E are satisfied.

A. Cognitive/communicative functioning generally acquired by children no more than one-half the child's chronological age, as documented by appropriate medical findings (e.g., in infants 0–6 months, markedly diminished variation in the production or imitation of sounds and severe feeding abnormality, such as problems with sucking swallowing, or chewing) including, if necessary, a standardized test;

OR

B. Motor development generally acquired by children no more than one-half the child's chronological age, documented by appropriate medical findings, including if necessary, a standardized test;

OR

C. Apathy, over-excitability, or fearfulness, demonstrated by an absent or grossly excessive response to one of the following:

1. Visual stimulation; or

2. Auditory stimulation; or

3. Tactile stimulation;

OR

D. Failure to sustain social interaction on an ongoing, reciprocal basis as evidenced by:

1. Inability by 6 months to participate in vocal, visual, and motoric exchanges (including facial expressions); or

2. Failure by 9 months to communicate basic emotional responses, such as cuddling or exhibiting protest or anger; or

3. Failure to attend to the caregiver's voice or face or to explore an inanimate object for a period of time appropriate to the infant's age;

OR

E. Attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in two or more areas (*i.e.*, cognitive/communicative, motor, and social), documented by appropriate medical findings, including if necessary, standardized testing.

113.00 Cancer (Malignant Neoplastic Diseases)

A. *What impairments do these listings cover?* We use these listings to evaluate all cancers (malignant neoplastic diseases), except certain cancers associated with human immunodeficiency virus (HIV) infection. If you have HIV infection, we use the criteria in

114.08E to evaluate carcinoma of the cervix, Kaposi sarcoma, lymphoma, and squamous cell carcinoma of the anal canal and anal margin.

B. *What do we consider when we evaluate cancer under these listings?* We will consider factors including:

1. Origin of the cancer.

2. Extent of involvement.

3. Duration, frequency, and response to anticancer therapy.

4. Effects of any post-therapeutic residuals.

C. *How do we apply these listings?* We apply the criteria in a specific listing to a cancer originating from that specific site.

D. *What evidence do we need?*

1. We need medical evidence that specifies the type, extent, and site of the primary, recurrent, or metastatic lesion. When the primary site cannot be identified, we will use evidence documenting the site(s) of metastasis to evaluate the impairment under 13.27 in part A.

2. For operative procedures, including a biopsy or a needle aspiration, we generally need a copy of both the:

a. Operative note, and

b. Pathology report.

3. When we cannot get these documents, we will accept the summary of hospitalization(s) or other medical reports. This evidence should include details of the findings at surgery and, whenever appropriate, the pathological findings.

4. In some situations, we may also need evidence about recurrence, persistence, or progression of the cancer, the response to therapy, and any significant residuals. (See 113.00G.)

E. *When do we need longitudinal evidence?*

1. *Cancer with distant metastases.* Most cancer of childhood consists of a local lesion with metastases to regional lymph nodes and, less often, distant metastases. We generally do not need longitudinal evidence for cancer that has metastasized beyond the regional lymph nodes because this cancer usually meets the requirements of a listing. Exceptions are for cancer with distant metastases that we expect to respond to anticancer therapy. For these exceptions, we usually need a longitudinal record of 3 months after therapy starts to determine whether the therapy achieved its intended effect, and whether this effect is likely to persist.

2. *Other cancers.* When there are no distant metastases, many of the listings require that we consider your response to initial anticancer therapy; that is, the initial planned treatment regimen. This therapy may consist of a single modality or a combination of modalities; that is, multimodal therapy (see 113.00I3).

3. *Types of treatment.*

a. Whenever the initial planned therapy is a single modality, enough time must pass to allow a determination about whether the therapy will achieve its intended effect. If the treatment fails, the failure often happens within 6 months after treatment starts, and there will often be a change in the treatment regimen.

b. Whenever the initial planned therapy is multimodal, we usually cannot make a determination about the effectiveness of the therapy until we can determine the effects of all the planned modalities. In some cases, we may need to defer adjudication until we can assess the effectiveness of therapy. However, we do not need to defer adjudication to determine whether the therapy will achieve its intended effect if we can make a fully favorable determination or decision based on the length and effects of therapy, or the residuals of the cancer or therapy (see 113.00G).

F. *How do we evaluate impairments that do not meet one of the cancer listings?*

1. These listings are only examples of cancers that we consider severe enough to result in marked and severe functional limitations. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that meets the criteria of a listing in another body system.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§404.1526 and 416.926 of this chapter.) If your impairment(s) does not meet or medically equal a listing, we will also consider whether you have an impairment(s) that functionally equals the listings. (See §416.926a of this chapter.) We use the rules in §416.994a of this chapter when we decide whether you continue to be disabled.

G. *How do we consider the effects of anticancer therapy?*

1. *How we consider the effects of anticancer therapy under the listings.* In many cases, cancers meet listing criteria only if the therapy is not effective and the cancer persists, progresses, or recurs. However, as explained in the following paragraphs, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in the case record.

2. *Effects can vary widely.*

a. We consider each case on an individual basis because the therapy and its toxicity may vary widely. We will request a specific description of the therapy, including these items:

i. Drugs given.

ii. Dosage.

iii. Frequency of drug administration.

iv. Plans for continued drug administration.

v. Extent of surgery.

vi. Schedule and fields of radiation therapy.

b. We will also request a description of the complications or adverse effects of therapy, such as the following:

i. Continuing gastrointestinal symptoms.

ii. Persistent weakness.

iii. Neurological complications.

iv. Cardiovascular complications.

v. Reactive mental disorders.

3. *Effects of therapy may change.* The severity of the adverse effects of anticancer therapy may change during treatment; therefore, enough time must pass to allow us to evaluate the therapy's effect. The residual effects of treatment are temporary in most instances; however, on occasion, the effects may be disabling for a consecutive period of at least 12 months. In some situations, very serious adverse effects may interrupt and prolong multimodal anticancer therapy for a continuous period of almost 12 months. In these situations, we may determine there is an expectation that your impairment will preclude you from engaging in any age-appropriate activities for at least 12 months.

4. *When the initial anticancer therapy is effective.* We evaluate any post-therapeutic residual impairment(s) not included in these listings under the criteria for the affected body system. We must consider any complications of therapy. When the residual impairment(s) does not meet a listing, we must consider whether it medically equals a listing, or, as appropriate, functionally equals the listings.

H. *How long do we consider your impairment to be disabling?*

1. In some listings, we specify that we will consider your impairment to be disabling until a particular point in time (for example, until at least 12 months from the date of transplantation). We may consider your impairment to be disabling beyond this point when the medical and other evidence justifies it.

2. When a listing does not contain such a specification, we will consider an impairment(s) that meets or medically equals a listing in this body system to be disabling until at least 3 years after onset of complete remission. When the impairment(s) has been in complete remission for at least 3 years, that is, the original tumor or a recurrence (or relapse) and any metastases have not been evident for at least 3 years, the impairment(s) will no longer meet or medically equal the criteria of a listing in this body system.

3. Following the appropriate period, we will consider any residuals, including residuals of the cancer or therapy (see 113.00G), in determining whether you are disabled. If you have a recurrence or relapse of your cancer, your impairment may meet or medically equal one of the listings in this body system again.

I. *What do we mean by the following terms?*

1. *Anticancer therapy* means surgery, radiation, chemotherapy, hormones, immunotherapy, or bone marrow or stem cell transplantation. When we refer to surgery as an anticancer treatment, we mean surgical excision for treatment, not for diagnostic purposes.

2. *Metastases* means the spread of cancer cells by blood, lymph, or other body fluid. This term does not include the spread of cancer cells by direct extension of the cancer to other tissues or organs.

3. *Multimodal therapy* means anticancer therapy that is a combination of at least two types of treatment given in close proximity as a unified whole and usually planned before any treatment has begun. There are three types of treatment modalities: Surgery, radiation, and systemic drug therapy (chemotherapy, hormone therapy, and immunotherapy or biological modifier therapy). Examples of multimodal therapy include:

a. Surgery followed by chemotherapy or radiation.

b. Chemotherapy followed by surgery.

c. Chemotherapy and concurrent radiation.

4. *Persistent* means the planned initial anticancer therapy failed to achieve a complete remission of your cancer; that is, your cancer is evident, even if smaller, after the therapy has ended.

5. *Progressive* means the cancer becomes more extensive after treatment; that is, there is evidence that your cancer is growing after you have completed at least half of your planned initial anticancer therapy.

6. *Recurrent or relapse* means the cancer that was in complete remission or entirely removed by surgery has returned.

J. *Can we establish the existence of a disabling impairment prior to the date of the evidence that shows the cancer satisfies the criteria of a listing?* Yes. We will consider factors such as:

1. The type of cancer and its location.

2. The extent of involvement when the cancer was first demonstrated.

3. Your symptoms.

K. *How do we evaluate specific cancers?*

1. *Lymphoma.*

a. We provide criteria for evaluating lymphomas that are disseminated or have not responded to anticancer therapy in 113.05.

b. Lymphoblastic lymphoma is treated with leukemia-based protocols, so we evaluate this type of cancer under 113.06.

2. *Leukemia.*

a. *Acute leukemia.* The initial diagnosis of acute leukemia, including the accelerated or blast phase of chronic myelogenous (granulocytic) leukemia, is based on definitive bone marrow examination. Additional diagnostic information is based on chromosomal analysis, cytochemical and surface marker studies on the abnormal cells, or

other methods consistent with the prevailing state of medical knowledge and clinical practice. Recurrent disease must be documented by peripheral blood, bone marrow, or cerebrospinal fluid examination, or by testicular biopsy. The initial and follow-up pathology reports should be included.

b. *Chronic myelogenous leukemia (CML).* We need a diagnosis of CML based on documented granulocytosis, including immature forms such as differentiated or undifferentiated myelocytes and myeloblasts, and a chromosomal analysis that demonstrates the Philadelphia chromosome. In the absence of a chromosomal analysis, or if the Philadelphia chromosome is not present, the diagnosis may be made by other methods consistent with the prevailing state of medical knowledge and clinical practice. The requirement for CML in the accelerated or blast phase is met in 113.06B if laboratory findings show the proportion of blast (immature) cells in the peripheral blood or bone marrow is 10 percent or greater.

c. *Juvenile chronic myelogenous leukemia (JCML).* JCML is a rare, Philadelphia-chromosome-negative childhood leukemia that is aggressive and clinically similar to acute myelogenous leukemia. We evaluate JCML under 113.06A.

d. *Elevated white cell count.* In cases of chronic leukemia (either myelogenous or lymphocytic), an elevated white cell count, in itself, is not a factor in determining the severity of the impairment.

3. *Malignant solid tumors.* The tumors we consider under 113.03 include the histiocytosis syndromes except for solitary eosinophilic granuloma. We do not evaluate thyroid cancer (see 113.09), retinoblastomas (see 113.12), primary central nervous system (CNS) cancers (see 113.13), neuroblastomas (see 113.21), or malignant melanoma (see 113.29) under this listing.

4. *Primary central nervous system (CNS) cancers.* We use the criteria in 113.13 to evaluate cancers that originate within the CNS (that is, brain and spinal cord cancers).

a. The CNS cancers listed in 113.13A are highly malignant and respond poorly to treatment, and therefore we do not require additional criteria to evaluate them. We do not list pituitary gland cancer (for example, pituitary gland carcinoma) in 113.13A, although this CNS cancer is highly malignant and responds poorly to treatment. We evaluate pituitary gland cancer under 113.13A and do not require additional criteria to evaluate it.

b. We consider a CNS tumor to be malignant if it is classified as Grade II, Grade III, or Grade IV under the World Health Organization (WHO) classification of tumors of the CNS (*WHO Classification of Tumours of the Central Nervous System*, 2007).

c. We evaluate benign (for example, WHO Grade I) CNS tumors under 111.05. We evaluate metastasized CNS cancers from non-CNS sites under the primary cancers (see 113.00C). We evaluate any complications of CNS cancers, such as resultant neurological or psychological impairments, under the criteria for the affected body system.

5. *Retinoblastoma.* The treatment for bilateral retinoblastoma usually results in a visual impairment. We will evaluate any resulting visual impairment under 102.02.

6. *Melanoma.* We evaluate malignant melanoma that affects the skin (cutaneous melanoma), eye (ocular melanoma), or mucosal membranes (mucosal melanoma) under 113.29. We evaluate melanoma that is not malignant that affects the skin (benign melanocytic tumor) under the listings in 108.00 or other affected body systems.

L. *How do we evaluate cancer treated by bone marrow or stem cell transplantation, including transplantation using stem cells from umbilical cord blood?* Bone marrow or stem cell transplantation is performed for a variety of cancers. We require the transplantation to occur before we evaluate it under these listings. We do not need to restrict our determination of the onset of disability to the date of transplantation (113.05 or 113.06). We may be able to establish an earlier onset date of disability due to your transplantation if the evidence in your case record supports such a finding.

1. *Acute leukemia (including all types of lymphoblastic lymphomas and JCML) or accelerated or blast phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of transplantation, whichever is later.

2. *Lymphoma or chronic phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 12 months from the date of transplantation.

3. *Evaluating disability after the appropriate time period has elapsed.* We consider any residual impairment(s), such as complications arising from:

a. Graft-versus-host (GVH) disease.

b. Immunosuppressant therapy, such as frequent infections.

c. Significant deterioration of other organ systems.

113.01 Category of Impairments, Cancer (Malignant Neoplastic Diseases)

113.01 Category of Impairments, Malignant Neoplastic Diseases

113.03 *Malignant solid tumors.* Consider under a disability:

A. For 24 months from the date of initial diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. For 24 months from the date of recurrence of active disease. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

113.05 *Lymphoma (excluding all types of lymphoblastic lymphomas—113.06).* (See 113.00K1.)

A. Non-Hodgkin lymphoma (including Burkitt's and anaplastic large cell), with either 1 or 2:

1. Bone marrow, brain, spinal cord, liver, or lung involvement at initial diagnosis. Consider under a disability for 24 months from the date of diagnosis. Thereafter, evaluate under 113.05A2, or any residual impairments(s) under the criteria for the affected body system.

2. Persistent or recurrent following initial anticancer therapy.

OR

B. Hodgkin lymphoma, with either 1 or 2:

1. Bone marrow, brain, spinal cord, liver, or lung involvement at initial diagnosis. Consider under a disability for 24 months from the date of diagnosis. Thereafter, evaluate under 113.05B2, or any residual impairment(s) under the criteria for the affected body system.

2. Persistent or recurrent following initial anticancer therapy.

OR

C. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria of the affected body system.

OR

D. Mantle cell lymphoma.

113.06 *Leukemia.* (See 113.00K2.)

A. Acute leukemia (including all types of lymphoblastic lymphomas and juvenile chronic myelogenous leukemia (JCML)). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Chronic myelogenous leukemia (except JCML), as described in 1 or 2:

1. Accelerated or blast phase (see 113.00K2b). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

2. Chronic phase, as described in a or b:

a. Consider under a disability until at least 12 months from the date of bone marrow or stem cell transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

b. Progressive disease following initial antineoplastic therapy.

113.09 *Thyroid gland.*

A. Anaplastic (undifferentiated) carcinoma.

OR

B. Carcinoma with metastases beyond the regional lymph nodes progressive despite radioactive iodine therapy.

OR

C. Medullary carcinoma with metastases beyond the regional lymph nodes.

113.12 *Retinoblastoma.*

A. With extension beyond the orbit.

OR

B. Persistent or recurrent following initial anticancer therapy.

OR

C. With regional or distant metastases.

113.13 *Nervous system.* (See 113.00K4.) Primary central nervous system (CNS; that is, brain and spinal cord) cancers, as described in A, B, or C:

A. Glioblastoma multiforme, ependymoblastoma, and diffuse intrinsic brain stem gliomas (see 113.00K4a).

B. Any Grade III or Grade IV CNS cancer (see 113.00K4b), including astrocytomas, sarcomas, and medulloblastoma and other primitive neuroectodermal tumors (PNETs).

C. Any primary CNS cancer, as described in 1 or 2:

1. Metastatic.

2. Progressive or recurrent following initial anticancer therapy.

113.21 *Neuroblastoma.*

A. With extension across the midline.

OR

B. With distant metastases.

OR

C. Recurrent.

OR

D. With onset at age 1 year or older.

113.29 *Malignant melanoma* (including skin, ocular, or mucosal melanomas), as described in either A, B, or C:

A. Recurrent (except an additional primary melanoma at a different site, which is not considered to be recurrent disease) following either 1 or 2:

1. Wide excision (skin melanoma).

2. Enucleation of the eye (ocular melanoma).

OR

B. With metastases as described in 1, 2, or 3:

1. Metastases to one or more clinically apparent nodes; that is, nodes that are detected by imaging studies (excluding lymphoscintigraphy) or by clinical evaluation (palpable).

2. If the nodes are not clinically apparent, with metastases to four or more nodes.

3. Metastases to adjacent skin (satellite lesions) or distant sites (for example, liver, lung, or brain).

OR

C. Mucosal melanoma.

114.00 IMMUNE SYSTEM DISORDERS

A. What disorders do we evaluate under the immune system disorders listings?

1. *We evaluate immune system disorders that cause dysfunction in one or more components of your immune system.*

a. The dysfunction may be due to problems in antibody production, impaired cell-mediated immunity, a combined type of antibody/cellular deficiency, impaired phagocytosis, or complement deficiency.

b. Immune system disorders may result in recurrent and unusual infections, or inflammation and dysfunction of the body's own tissues. Immune system disorders can cause a deficit in a single organ or body system that results in extreme (that is, very serious) loss of function. They can also cause lesser degrees of limitations in two or more organs or body systems, and when associated with symptoms or signs, such as severe fatigue, fever, malaise, diffuse musculoskeletal pain, or involuntary weight loss, can also result in extreme limitation. In children, immune system disorders or their treatment may also affect growth, development, and the performance of age-appropriate activities.

c. We organize the discussions of immune system disorders in three categories: Autoimmune disorders; Immune deficiency disorders, excluding human immunodeficiency virus (HIV) infection; and HIV infection.

2. *Autoimmune disorders (114.00D).* Autoimmune disorders are caused by dysfunctional immune responses directed against the body's own tissues, resulting in chronic, multisystem impairments that differ in clinical manifestations, course, and outcome. They are sometimes referred to as rheumatic diseases, connective tissue disorders, or collagen vascular disorders. Some of the features of autoimmune disorders in children differ from the features of the same disorders in adults. The impact of the disorders or their treatment on physical, psychological, and developmental growth of pre-pubertal children may be considerable, and often differs from that of post-pubertal adolescents or adults.

3. *Immune deficiency disorders, excluding HIV infection (114.00E).* Immune deficiency disorders are characterized by recurrent or unusual infections that respond poorly to treatment, and are often associated with complications affecting other parts of the body. Immune deficiency disorders are classified as either *primary* (congenital) or *acquired*. Children with immune deficiency disorders also have an increased risk of malignancies and of having autoimmune disorders.

4. *Human immunodeficiency virus (HIV) infection (114.00F).* HIV infection may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions, as described in 114.08.

B. What information do we need to show that you have an immune system disorder?

Generally, we need your medical history, a report(s) of a physical examination, a report(s) of laboratory findings, and in some instances, appropriate medically acceptable imaging or tissue biopsy reports to show that you have an immune system disorder. Therefore, we will make every reasonable effort to obtain your medical history, medical findings, and results of laboratory tests. We explain the information we need in more detail in the sections below.

C. Definitions

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Constitutional symptoms or signs,* as used in these listings, means severe fatigue, fever, malaise, or involuntary weight loss. *Severe fatigue* means a frequent sense of exhaustion that results in significantly reduced physical activity or mental function. *Malaise* means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.

3. *Disseminated* means that a condition is spread over a considerable area. The type and extent of the spread will depend on your specific disease.

4. *Dysfunction* means that one or more of the body regulatory mechanisms are impaired, causing either an excess or deficiency of immunocompetent cells or their products.

5. *Extra-articular* means ''other than the joints''; for example, an organ(s) such as the heart, lungs, kidneys, or skin.

6. *Inability to ambulate effectively* has the same meaning as in 101.00B2b.

7. *Inability to perform fine and gross movements effectively* has the same meaning as in 101.00B2c.

8. *Major peripheral joints* has the same meaning as in 101.00F.

9. *Persistent* means that a sign(s) or symptom(s) has continued over time. The precise meaning will depend on the specific immune system disorder, the usual course of the disorder, and the other circumstances of your clinical course.

10. *Recurrent* means that a condition that previously responded adequately to an appropriate course of treatment returns after a period of remission or regression. The precise meaning, such as the extent of response or remission and the time periods involved, will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

11. *Resistant to treatment* means that a condition did not respond adequately to an appropriate course of treatment. Whether a response is adequate or a course of treatment is appropriate will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

12. *Severe* means medical severity as used by the medical community. The term does not have the same meaning as it does when we use it in connection with a finding at the second step of the sequential evaluation process in §416.924.

D. How do we document and evaluate the listed autoimmune disorders?

1. *Systemic lupus erythematosus (114.02).*

a. *General.* Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that can affect any organ or body system. It is frequently, but not always, accompanied by constitutional symptoms or signs (severe fatigue, fever, malaise, involuntary weight loss). Major organ or body system involvement can include: Respiratory (pleuritis, pneumonitis), cardiovascular (endocarditis, myocarditis, pericarditis, vasculitis), renal (glomerulonephritis), hematologic (anemia, leukopenia, thrombocytopenia), skin (photosensitivity), neurologic (seizures), mental (anxiety, fluctuating cognition (''lupus fog''), mood disorders, organic brain syndrome, psychosis), or immune system disorders (inflammatory arthritis). Immunologically, there is an array of circulating serum auto-antibodies and pro- and anti-coagulant proteins that may occur in a highly variable pattern.

b. *Documentation of SLE.* Generally, but not always, the medical evidence will show that your SLE satisfies the criteria in the current ''Criteria for the Classification of Systemic Lupus Erythematosus'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

2. *Systemic vasculitis (114.03).*

a. *General.*

(i) Vasculitis is an inflammation of blood vessels. It may occur acutely in association with adverse drug reactions, certain chronic infections, and occasionally, malignancies. More often, it is chronic and the cause is unknown. Symptoms vary depending on which blood vessels are involved. Systemic vasculitis may also be associated with other autoimmune disorders; for example, SLE or dermatomyositis.

(ii) Children can develop the vasculitis of Kawasaki disease, of which the most serious manifestation is formation of coronary artery aneurysms and related complications. We evaluate heart problems related to Kawasaki disease under the criteria in the cardiovascular listings (104.00). Children can also develop the vasculitis of anaphylactoid purpura (Henoch-Schoenlein purpura), which may cause intestinal and renal disorders. We evaluate intestinal and renal disorders related to vasculitis of anaphylactoid purpura under the criteria in the digestive (105.00) or genitourinary (106.00) listings. Other clinical patterns include, but are not limited to, polyarteritis nodosa, Takayasu's arteritis (aortic arch arteritis), and Wegener's granulomatosis.

b. *Documentation of systemic vasculitis.* Angiography or tissue biopsy confirms a diagnosis of systemic vasculitis when the disease is suspected clinically. When you have had angiography or tissue biopsy for systemic vasculitis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase angiography or tissue biopsy.

3. *Systemic sclerosis (scleroderma) (114.04).*

a. *General.* Systemic sclerosis (scleroderma) constitutes a spectrum of disease in which thickening of the skin is the clinical hallmark. Raynaud's phenomenon, often medically severe and progressive, is present frequently and may be the peripheral manifestation of a vasospastic abnormality in the heart, lungs, and kidneys. The CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) is a variant that may slowly progress over years to the generalized process, systemic sclerosis.

b. *Diffuse cutaneous systemic sclerosis.* In diffuse cutaneous systemic sclerosis (also known as diffuse scleroderma), major organ or systemic involvement can include the gastrointestinal tract, lungs, heart, kidneys, and muscle in addition to skin or blood vessels. Although arthritis can occur, joint dysfunction results primarily from soft tissue/cutaneous thickening, fibrosis, and contractures.

c. *Localized scleroderma (linear scleroderma and morphea).*

(i) Localized scleroderma (linear scleroderma and morphea) is more common in children than systemic scleroderma. To assess the severity of the impairment, we need a description of the extent of involvement of linear scleroderma and the location of the lesions. For example, linear scleroderma involving the arm but not crossing any joints is not as functionally limiting

as sclerodactyly (scleroderma localized to the fingers). Linear scleroderma of a lower extremity involving skin thickening and atrophy of underlying muscle or bone can result in contractures and leg length discrepancy. In such cases, we may evaluate your impairment under the musculoskeletal listings (101.00).

(ii) When there is isolated morphea of the face causing facial disfigurement from unilateral hypoplasia of the mandible, maxilla, zygoma, or orbit, adjudication may be more appropriate under the criteria in the affected body system, such as special senses and speech (102.00) or mental disorders (112.00).

(iii) Chronic variants of these syndromes include disseminated morphea, Shulman's disease (diffuse fasciitis with eosinophilia), and eosinophilia-myalgia syndrome (often associated with toxins such as toxic oil or contaminated tryptophan), all of which can impose medically severe musculoskeletal dysfunction and may also lead to restrictive pulmonary disease. We evaluate these variants of the disease under the criteria in the musculoskeletal listings (101.00) or respiratory system listings (103.00).

d. *Documentation of systemic sclerosis (scleroderma).* Documentation involves differentiating the clinical features of systemic sclerosis (scleroderma) from other autoimmune disorders. However, there may be an overlap.

4. *Polymyositis and dermatomyositis (114.05).*

a. *General.*

(i) Polymyositis and dermatomyositis are related disorders that are characterized by an inflammatory process in striated muscle, occurring alone or in association with other autoimmune disorders. The most common manifestations are symmetric weakness, and less frequently, pain and tenderness of the proximal limb-girdle (shoulder or pelvic) musculature. There may also be involvement of the cervical, cricopharyngeal, esophageal, intercostal, and diaphragmatic muscles.

(ii) Polymyositis occurs rarely in children; the more common presentation in children is dermatomyositis with symmetric proximal muscle weakness and characteristic skin findings. The clinical course of dermatomyositis can be more severe when it is accompanied by systemic vasculitis rather than just localized to striated muscle. Late in the disease, some children with dermatomyositis develop calcinosis of the skin and subcutaneous tissues, muscles, and joints. We evaluate the involvement of other organs/body systems under the criteria for the listings in the affected body system.

b. *Documentation of polymyositis and dermatomyositis.* Generally, but not always, polymyositis is associated with elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, and aldolase), and characteristic abnormalities on electromyography and muscle biopsy. In children, the diagnosis of dermatomyositis is supported largely by medical history, findings on physical examination that include the characteristic skin findings, and elevated serum muscle enzymes. Muscle inflammation or vasculitis depicted on MRI is additional evidence supporting the diagnosis of childhood dermatomyositis. When you have had electromyography, muscle biopsy, or MRI for polymyositis or dermatomyositis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase electromyography, muscle biopsy, or MRI.

c. *Additional information about how we evaluate polymyositis and dermatomyositis under the listings.*

(i) In newborn and younger infants (birth to attainment of age 1), we consider muscle weakness that affects motor skills, such as head control, reaching, grasping, taking solids, or self-feeding, under 114.05A. In older infants and toddlers (age 1 to attainment of age 3), we also consider muscle weakness affecting your ability to roll over, sit, crawl, or walk under 114.05A.

(ii) If you are of preschool age through adolescence (age 3 to attainment of age 18), weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to ambulate effectively. Weakness of your shoulder girdle muscles may result in your inability to perform lifting, carrying, and reaching overhead, and also may seriously affect your ability to perform activities requiring fine movements. We evaluate these limitations under 114.05A.

5. *Undifferentiated and mixed connective tissue disease (114.06).*

a. *General.* This listing includes syndromes with clinical and immunologic features of several autoimmune disorders, but which do not satisfy the criteria for any of the specific disorders described. For example, you may have clinical features of SLE and systemic vasculitis, and the serologic (blood test) findings of rheumatoid arthritis. The most common pattern of undifferentiated autoimmune disorders in children is mixed connective tissue disease (MCTD).

b. *Documentation of undifferentiated and mixed connective tissue disease.* Undifferentiated connective tissue disease is diagnosed when clinical features and serologic (blood test) findings, such as rheumatoid factor or antinuclear antibody (consistent with an autoimmune disorder) are present but do not satisfy the criteria for a specific disease. Children with MCTD have laboratory findings of extremely high antibody titers to extractable nuclear antigen (ENA) or ribonucleoprotein (RNP) without high titers of anti-dsDNA or anti-SM antibodies. There are often clinical findings suggestive of SLE

or childhood dermatomyositis. Many children later develop features of scleroderma.

6. *Inflammatory arthritis (114.09).*

a. *General.* The spectrum of inflammatory arthritis includes a vast array of disorders that differ in cause, course, and outcome. Clinically, inflammation of major peripheral joints may be the dominant manifestation causing difficulties with ambulation or fine and gross movements; there may be joint pain, swelling, and tenderness. The arthritis may affect other joints, or cause less limitation in ambulation or the performance of fine and gross movements. However, in combination with extra-articular features, including constitutional symptoms or signs (severe fatigue, fever, malaise, involuntary weight loss), inflammatory arthritis may result in an extreme limitation. You may also have impaired growth as a result of the inflammatory arthritis because of its effects on the immature skeleton, open epiphyses, and young cartilage and bone. We evaluate any associated growth impairment under the criteria in 100.00.

b. *Inflammatory arthritis involving the axial spine (spondyloarthropathy).* In children, inflammatory arthritis involving the axial spine may be associated with disorders such as:

(i) Reactive arthropathies;

(ii) Juvenile ankylosing spondylitis;

(iii) Psoriatic arthritis;

(iv) SEA syndrome (seronegative enthesopathy arthropathy syndrome);

(v) Behçet's disease; and

(vi) Inflammatory bowel disease.

c. *Inflammatory arthritis involving the peripheral joints.* In children, inflammatory arthritis involving peripheral joints may be associated with disorders such as:

(i) Juvenile rheumatoid arthritis;

(ii) Sjöogren's syndrome;

(iii) Psoriatic arthritis;

(iv) Crystal deposition disorders (gout and pseudogout);

(v) Lyme disease; and

(vi) Inflammatory bowel disease.

d. *Documentation of inflammatory arthritis.* Generally, but not always, the diagnosis of inflammatory arthritis is based on the clinical features and serologic findings described in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

e. *How we evaluate inflammatory arthritis under the listings.*

(i) Listing-level severity in 114.09A and 114.09C1 is shown by an impairment that results in an ''extreme'' (very serious) limitation. In 114.09A, the criterion is satisfied with persistent inflammation or deformity in one major peripheral weight-bearing joint resulting in the inability to ambulate effectively (as defined in 114.00C6) or one major peripheral joint in each upper extremity resulting in the inability to perform fine and gross movements effectively (as defined in 114.00C7). In 114.09C1, if you have the required ankylosis (fixation) of your cervical or dorsolumbar spine, we will find that you have an extreme limitation in your ability to see in front of you, above you, and to the side. Therefore, inability to ambulate effectively is implicit in 114.09C1, even though you might not require bilateral upper limb assistance.

(ii) Listing-level severity is shown in 114.09B, 114.09C2, and 114.09D by inflammatory arthritis that involves various combinations of complications of one or more major peripheral joints or involves other joints, such as inflammation or deformity, extra-articular features, repeated manifestations, and constitutional symptoms and signs. Extra-articular impairments may also meet listings in other body systems.

(iii) Extra-articular features of inflammatory arthritis may involve any body system; for example: Musculoskeletal (heel enthesopathy), ophthalmologic (iridocyclitis, keratoconjunctivitis sicca, uveitis), pulmonary (pleuritis, pulmonary fibrosis or nodules, restrictive lung disease), cardiovascular (aortic valve insufficiency, arrhythmias, coronary arteritis, myocarditis, pericarditis, Raynaud's phenomenon, systemic vasculitis), renal (amyloidosis of the kidney), hematologic (chronic anemia, thrombocytopenia), neurologic (peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss), mental (cognitive dysfunction, poor memory), and immune system (Felty's syndrome (hypersplenism with compromised immune competence)).

(iv) If both inflammation and chronic deformities are present, we evaluate your impairment under the criteria of any appropriate listing.

7. *Sjögren's syndrome (114.10).*

a. *General.*

(i) Sjögren's syndrome is an immune-mediated disorder of the exocrine glands. Involvement of the lacrimal and salivary glands is the hallmark feature, resulting in symptoms of dry eyes and dry mouth, and possible complications, such as corneal damage, blepharitis (eyelid inflammation), dysphagia (difficulty in swallowing), dental caries, and the inability to speak for extended periods of time. Involvement of the exocrine glands of the upper airways may result in persistent dry cough.

(ii) Many other organ systems may be involved, including musculoskeletal (arthritis, myositis), respiratory (interstitial fibrosis), gastrointestinal (dysmotility, dysphagia, involuntary weight loss), genitourinary (interstitial cystitis, renal tubular acidosis), skin (purpura, vasculitis,), neurologic (central nervous system disorders, cranial and peripheral neuropathies), mental (cognitive dysfunction, poor memory), and neoplastic

(lymphoma). Severe fatigue and malaise are frequently reported. Sjögren's syndrome may be associated with other autoimmune disorders (for example, rheumatoid arthritis or SLE); usually the clinical features of the associated disorder predominate.

b. *Documentation of Sjögren's syndrome.* If you have Sjögren's syndrome, the medical evidence will generally, but not always, show that your disease satisfies the criteria in the current ''Criteria for the Classification of Sjögren's Syndrome'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

E. How do we document and evaluate immune deficiency disorders, excluding HIV infection?

1. *General.*

a. Immune deficiency disorders can be classified as:

(i) *Primary* (congenital); for example, X-linked agammaglobulinemia, thymic hypoplasia (DiGeorge syndrome), severe combined immunodeficiency (SCID), chronic granulomatous disease (CGD), C1 esterase inhibitor deficiency.

(ii) *Acquired;* for example, medication-related.

b. Primary immune deficiency disorders are seen mainly in children. However, recent advances in the treatment of these disorders have allowed many affected children to survive well into adulthood. Occasionally, these disorders are first diagnosed in adolescence or adulthood.

2. *Documentation of immune deficiency disorders.* The medical evidence must include documentation of the specific type of immune deficiency. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

3. *Immune deficiency disorders treated by stem cell transplantation.*

a. *Evaluation in the first 12 months.* If you undergo stem cell transplantation for your immune deficiency disorder, we will consider you disabled until at least 12 months from the date of the transplant.

b. *Evaluation after the 12-month period has elapsed.* After the 12-month period has elapsed, we will consider any residuals of your immune deficiency disorder as well as any residual impairment(s) resulting from the treatment, such as complications arising from:

(i) Graft-versus-host (GVH) disease.

(ii) Immunosuppressant therapy, such as frequent infections.

(iii) Significant deterioration of other organ systems.

4. *Medication-induced immune suppression.* Medication effects can result in varying degrees of immune suppression, but most resolve when the medication is ceased. However, if you are prescribed medication for long-term immune suppression, such as after an organ transplant, we will evaluate:

a. The frequency and severity of infections.

b. Residuals from the organ transplant itself, after the 12-month period has elapsed.

c. Significant deterioration of other organ systems.

F. *How do we document and evaluate human immunodeficiency virus (HIV) infection?* Any child with HIV infection, including one with a diagnosis of acquired immune deficiency syndrome (AIDS), may be found disabled under 114.08 if his or her impairment meets the criteria in that listing or is medically equivalent to the criteria in that listing.

1. *Documentation of HIV infection.* The medical evidence must include documentation of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for HIV infection, we will make every reasonable effort to obtain reports of the results of that testing. However, we will not purchase laboratory testing to establish whether you have HIV infection.

a. *Definitive documentation of HIV infection.* A definitive diagnosis of HIV infection is documented by one or more of the following laboratory tests:

(i) HIV antibody tests. HIV antibodies are usually first detected by an ELISA screening test performed on serum. Because the ELISA can yield false positive results, confirmation is required using a more definitive test, such as a Western blot or an immunofluorescence assay. Positive results on these tests are considered to be diagnostic of HIV infection in a child age 18 months or older. (See b. below for information about HIV antibody testing in children younger than 18 months of age.)

(ii) Positive ''viral load'' (VL) tests. These tests are normally used to quantitate the amount of the virus present but also document HIV infection. Such tests include the quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT–PCR).

(iii) HIV DNA detection by polymerase chain reaction (PCR).

(iv) A specimen that contains HIV antigen (for example, serum specimen, lymphocyte culture, or cerebrospinal fluid) in a child age 1 month or older.

(v) A positive viral culture for HIV from peripheral blood mononuclear cells (PBMC).

(vi) An immunoglobulin A (IgA) serological assay that is specific for HIV.

(vii) Other tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge.

b. *Definitive documentation of HIV infection in children from birth to the attainment of 18 months.* For children from birth to the attainment of 18 months of age, and who have tested positive for HIV antibodies, HIV infection is documented by:

(i) One or more of the tests listed in F1a(ii)–F1a(vii).

(ii) For newborn and younger infants (birth to attainment of age 1), a CD4 (T4) count of 1500/mm^3 or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iii) For older infants and toddlers from 12 to 18 months of age, a CD4 (T4) count of 750/mm^3 or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iv) An abnormal CD4/CD8 ratio.

(v) A severely diminished immunoglobulin G (IgG) level (<4 g/l or 400 mg/dl), or significantly greater than normal range for age.

c. *Other acceptable documentation of HIV infection.* We may also document HIV infection without the definitive laboratory evidence described in 114.00F1a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. If no definitive laboratory evidence is available, we may document HIV infection by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence. For example, we will accept a diagnosis of HIV infection without definitive laboratory evidence of the HIV infection if you have an opportunistic disease that is predictive of a defect in cell-mediated immunity (for example, *Pneumocystis* pneumonia (PCP)), and there is no other known cause of diminished resistance to that disease (for example, long-term steroid treatment, lymphoma). In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

2. *CD4 tests.* Children who have HIV infection or other disorders of the immune system may have tests showing a reduction of either the absolute count or the percentage of their T-helper lymphocytes (CD4 cells). The extent of immune suppression correlates with the level or rate of decline of the CD4 count (relative to the age of the young child). By age 6, children have CD4 counts comparable to those levels found in adults. Generally, in these children when the CD4 count is below 200/mm^3 (or below 14 percent of the total lymphocyte count) the susceptibility to opportunistic infection is greatly increased. Although a reduced CD4 count alone does not establish a definitive diagnosis of HIV infection, a CD4 count below 200 does offer supportive evidence when there are clinical findings, but not a definitive diagnosis of an opportunistic infection(s). However, a reduced CD4 count *alone* does not document the severity or functional consequences of HIV infection.

3. *Documentation of the manifestations of HIV infection.* The medical evidence must also include documentation of the manifestations of HIV infection. Documentation may be by laboratory evidence or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

a. *Definitive documentation of the manifestations of HIV infection.* The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serologic test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). We will make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histologic or other test has been performed, the evidence should include a copy of the appropriate report. If we cannot obtain the report, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including appropriate medically acceptable imaging studies) or microscopic examination of the appropriate tissues or body fluids.

b. *Other acceptable documentation of the manifestations of HIV infection.* We may also document manifestations of HIV infection without the definitive laboratory evidence described in 114.00F3a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. For example, many conditions are now commonly diagnosed based on some or all of the following: Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing. The following are examples of how we may document manifestations of HIV infection with other appropriate evidence.

(i) Although a definitive diagnosis of PCP requires identifying the organism in bronchial washings, induced sputum, or lung biopsy, these tests are frequently bypassed if PCP can be diagnosed presumptively. Supportive evidence may include: Fever, dyspnea, hypoxia, CD4 count below 200 in children 6 years of age or older, and no evidence of bacterial pneumonia. Also supportive are bilateral lung interstitial infiltrates on x-ray, a typical pattern on CAT scan, or a gallium scan positive for pulmonary uptake. Response to anti-PCP therapy usually requires 5–7 days, and such a response can be supportive of the diagnosis.

(ii) Documentation of *Cytomegalovirus* (CMV) disease (114.08D) may present special problems because definitive diagnosis (except for chorioretinitis, which may be diagnosed by an ophthalmologist or optometrist on funduscopic examination) requires identification of viral inclusion bodies or a positive culture from the affected organ and the absence of any other infectious agent likely to be causing the disease. A positive serology test does not establish a definitive diagnosis of CMV disease, but does offer supportive evidence of a presumptive diagnosis of CMV disease. Other clinical findings that support a presumptive diagnosis of CMV may include: Fever, urinary culture positive for CMV, and CD4 count below 200 in children 6 years of age or older. A clear response to anti-CMV therapy also supports a diagnosis.

(iii) A definitive diagnosis of toxoplasmosis of the brain is based on brain biopsy, but this procedure carries significant risk and is not commonly performed. This condition is usually diagnosed presumptively based on symptoms or signs of fever, headache, focal neurologic deficits, seizures, typical lesions on brain imaging, and a positive serology test.

(iv) Candidiasis of the esophagus (also known as *Candida* esophagitis) may be presumptively diagnosed based on symptoms of retrosternal pain on swallowing (odynophagia) and either oropharyngeal thrush (white patches or plaques) diagnosed on physical examination or by microscopic documentation of *Candida* fungal elements from a noncultured specimen scraped from the oral mucosa. Treatment with oral (systemic) antifungal agents usually produces improvement after 5 or more days of therapy, and such a response can be supportive of the diagnosis.

4. *HIV infection manifestations specific to children.*

a. *General.* The clinical manifestation and course of disease in children who become infected with HIV perinatally or in the first 12 years of life may differ from that in adolescents (age 12 to attainment of age 18) and adults. Newborn and younger infants (birth to attainment of age 1) and older infants and toddlers (age 1 to attainment of age 3) may present with failure to thrive or PCP; preschool children (age 3 to attainment of age 6) and primary school children (age 6 to attainment of age 12) may present with recurrent infections, neurological problems, or developmental abnormalities. Adolescents may also exhibit neurological abnormalities, such as HIV encephalopathy, or have growth problems. HIV infection that affects the digestive system and results in malnutrition also may be evaluated under 105.08.

b. *Neurologic abnormalities.* The methods of identifying and evaluating neurologic abnormalities may vary depending on a child's age. For example, in an infant, impaired brain growth can be documented by a decrease in the growth rate of the head. In an older child, impaired brain growth may be documented by brain atrophy on a CAT scan or MRI. Neurologic abnormalities in infants and young children may present as serious developmental delays or in the loss of previously acquired developmental milestones. In school-age children and adolescents, this type of neurologic abnormality generally presents as the loss of previously acquired intellectual abilities. This may be evidenced in a child by a decrease in intelligence quotient (IQ) scores, by forgetting information previously learned, by inability to learn new information, or by a sudden onset of a new learning disability.

c. *Bacterial infections.* Children with HIV infection may contract any of a broad range of bacterial infections. Certain major infections caused by pyogenic bacteria (for example, some pneumonias) can be severely limiting, especially in pre-adolescent children. We evaluate these major bacterial infections under 114.08A4. Although 114.08A4 applies only to children under 13 years of age, children age 13 and older may have an impairment that medically equals this listing if the circumstances of the case warrant; for example, if there is delayed puberty. We will evaluate pelvic inflammatory disease in older girls under 114.08A5.

d. *Growth failure due to HIV immune suppression.*

(i) To evaluate growth failure due to HIV immune suppression, we require documentation of the laboratory values described in 114.08H1 and the growth measurements in 114.08H2 or 114.08H3 within the same consecutive 12-month period. The dates of laboratory findings may be different from the dates of growth measurements.

(ii) Under 114.08H2 and 114.08H3, we use the appropriate table under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile.

(A) For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

(B) For children age 2 to attainment of age 18, we use the body mass index (BMI)-for-age table corresponding to the child's gender (Table III or Table IV).

(C) BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

G. How do we consider the effects of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

1. *General.* If your impairment does not otherwise meet the requirements of a listing, we will consider your medical treatment in terms of its effectiveness in improving the signs, symptoms, and laboratory abnormalities of your specific immune system disorder

or its manifestations, and in terms of any side effects that limit your functioning. We will make every reasonable effort to obtain a specific description of the treatment you receive (including surgery) for your immune system disorder. We consider:

a. The effects of medications you take.

b. Adverse side effects (acute and chronic).

c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).

d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).

e. Variability of your response to treatment (see 114.00G2).

f. The interactive and cumulative effects of your treatments. For example, many children with immune system disorders receive treatment both for their immune system disorders and for the manifestations of the disorders or co-occurring impairments, such as treatment for HIV infection and hepatitis C. The interactive and cumulative effects of these treatments may be greater than the effects of each treatment considered separately.

g. The duration of your treatment.

h. Any other aspects of treatment that may interfere with your ability to function.

2. *Variability of your response to treatment.* Your response to treatment and the adverse or beneficial consequences of your treatment may vary widely. The effects of your treatment may be temporary or long term. For example, some children may show an initial positive response to a drug or combination of drugs followed by a decrease in effectiveness. When we evaluate your response to treatment and how your treatment may affect you, we consider such factors as disease activity before treatment, requirements for changes in therapeutic regimens, the time required for therapeutic effectiveness of a particular drug or drugs, the limited number of drug combinations that may be available for your impairment(s), and the time-limited efficacy of some drugs. For example, a child with HIV infection or another immune deficiency disorder who develops otitis media may not respond to the same antibiotic regimen used in treating children without HIV infection or another immune deficiency disorder, or may not respond to an antibiotic that he or she responded to before. Therefore, we must consider the effects of your treatment on an individual basis, including the effects of your treatment on your ability to function.

3. *How we evaluate the effects of treatment for autoimmune disorders on your ability to function.* Some medications may have acute or long-term side effects. When we consider the effects of corticosteroids or other treatments for autoimmune disorders on your ability to function, we consider the factors in 114.00G1 and 114.00G2. Long-term corticosteroid treatment can cause ischemic necrosis of bone, posterior subcapsular cataract, impaired growth, weight gain, glucose intolerance, increased susceptibility to infection, and osteopenia that may result in a loss of function. In addition, medications used in the treatment of autoimmune disorders may also have effects on mental functioning, including cognition (for example, memory), concentration, and mood.

4. *How we evaluate the effects of treatment for immune deficiency disorders, excluding HIV infection, on your ability to function.* When we consider the effects of your treatment for your immune deficiency disorder on your ability to function, we consider the factors in 114.00G1 and 114.00G2. A frequent need for treatment such as intravenous immunoglobulin and gamma interferon therapy can be intrusive and interfere with your ability to function. We will also consider whether you have chronic side effects from these or other medications, including severe fatigue, fever, headaches, high blood pressure, joint swelling, muscle aches, nausea, shortness of breath, or limitations in mental function including cognition (for example, memory) concentration, and mood.

5. *How we evaluate the effects of treatment for HIV infection on your ability to function.*

a. *General.* When we consider the effects of antiretroviral drugs (including the effects of highly active antiretroviral therapy (HAART)) and the effects of treatments for the manifestations of HIV infection on your ability to function, we consider the factors in 114.00G1 and 114.00G2. Side effects of antiretroviral drugs include, but are not limited to: Bone marrow suppression, pancreatitis, gastrointestinal intolerance (nausea, vomiting, diarrhea), neuropathy, rash, hepatotoxicity, lipodystrophy (fat redistribution, such as ''buffalo hump''), glucose intolerance, and lactic acidosis. In addition, medications used in the treatment of HIV infection may also have effects on mental functioning, including cognition (for example, memory), concentration, and mood, and may result in malaise, severe fatigue, joint and muscle pain, and insomnia. The symptoms of HIV infection and the side effects of medication may be indistinguishable from each other. We will consider all of your functional limitations, whether they result from your symptoms or signs of HIV infection or the side effects of your treatment.

b. *Structured treatment interruptions.* A structured treatment interruption (STI, also called a ''drug holiday'') is a treatment practice during which your treating source advises you to stop taking your medications temporarily. An STI in itself does not imply that your medical condition has improved; nor does it imply that you are noncompliant with your treatment because you are following your treating source's advice. Therefore, if you have stopped taking medication

because your treating source prescribed or recommended an STI, we will not find that you are failing to follow treatment or draw inferences about the severity of your impairment on this fact alone. We will consider why your treating source has prescribed or recommended an STI and all the other information in your case record when we determine the severity of your impairment.

6. *When there is no record of ongoing treatment.* If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the medical severity and duration of your immune system disorder on the basis of the current objective medical evidence and other evidence in your case record, taking into consideration your medical history, symptoms, clinical and laboratory findings, and medical source opinions. If you have just begun treatment and we cannot determine whether you are disabled based on the evidence we have, we may need to wait to determine the effect of the treatment on your ability to develop and function in an age-appropriate manner. The amount of time we need to wait will depend on the facts of your case. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the immune system disorders listings, but your immune system disorder may medically equal a listing or functionally equal the listings.

H. How do we consider your symptoms, including your pain, severe fatigue, and malaise?

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your immune system disorder(s) meets or medically equals a listing or in our determination whether you otherwise have marked and severe functional limitations. In order for us to consider your symptoms, you must have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 114.00 and in our other regulations. See §§ 416.928, and 416.929. Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

I. How do we use the functional criteria in these listings?

1. The following listings in this body system include standards for evaluating the functional limitations resulting from immune system disorders: 114.02B, for systemic lupus erythematosus; 114.03B, for systemic vasculitis; 114.04D, for systemic sclerosis (scleroderma); 114.05E, for polymyositis and dermatomyositis; 114.06B, for undifferentiated and mixed connective tissue disease; 114.07C, for immune deficiency disorders, excluding HIV infection; 114.08L, for HIV infection; 114.09D, for inflammatory arthritis; and 114.10B, for Sjögren's syndrome.

2. When we use one of the listings cited in 114.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder on your ability to function. Important factors we will consider when we evaluate your functioning under these listings include, but are not limited to: Your symptoms, the frequency and duration of manifestations of your immune system disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

3. To satisfy the functional criterion in a listing, your immune system disorder must result in an ''extreme'' limitation in one domain of functioning or a ''marked'' limitation in two domains of functioning depending on your age. (See 112.00C for additional discussion of these areas of functioning and §§ 416.924a and 416.926a for additional guidance on the evaluation of functioning in children.) Functional limitation may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This could result from persistent or intermittent symptoms, such as depression, severe fatigue, or pain, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. You may also have limitations because of your treatment and its side effects (see 114.00G).

J. How do we evaluate your immune system disorder when it does not meet one of these listings?

1. These listings are only examples of immune system disorders that we consider severe enough to result in marked and severe functional limitations. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. Individuals with immune system disorders, including HIV infection, may manifest signs or symptoms of a mental impairment or of another physical impairment. We

may evaluate these impairments under any affected body system. For example, we will evaluate:

a. Growth impairment under 100.00.

b. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 101.00.

c. Ocular involvement, such as dry eye, under 102.00.

d. Respiratory impairments, such as pleuritis, under 103.00.

e. Cardiovascular impairments, such as cardiomyopathy, under 104.00.

f. Digestive impairments, such as hepatitis (including hepatitis C) or weight loss as a result of HIV infection that affects the digestive system, under 105.00.

g. Genitourinary impairments, such as nephropathy, under 106.00.

h. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 107.00.

i. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 108.00.

j. Neurologic impairments, such as neuropathy or seizures, under 111.00.

k. Mental disorders, such as depression, anxiety, or cognitive deficits, under 112.00.

l. Allergic disorders, such as asthma or atopic dermatitis, under 103.00 or 108.00 or under the criteria in another affected body system.

m. Syphilis or neurosyphilis under the criteria for the affected body system, for example, 102.00 Special senses and speech, 104.00 Cardiovascular system, or 111.00 Neurological.

3. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926.) If it does not, we will also consider whether you have an impairment(s) that functionally equals the listings. (See §416.926a.) We use the rules in §416.994a when we decide whether you continue to be disabled.

114.01 *Category of Impairments, Immune System Disorders.*

114.02 *Systemic lupus erythematosus.* As described in 114.00D1. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Any other manifestation(s) of SLE resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.03 *Systemic vasculitis.* As described in 114.00D2. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Any other manifestation(s) of systemic vasculitis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.04 *Systemic sclerosis (scleroderma).* As described in 114.00D3. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. With one of the following:

1. Toe contractures or fixed deformity of one or both feet, resulting in the inability to ambulate effectively as defined in 114.00C6; or

2. Finger contractures or fixed deformity in both hands, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7; or

3. Atrophy with irreversible damage in one or both lower extremities, resulting in the inability to ambulate effectively as defined in 114.00C6; or

4. Atrophy with irreversible damage in both upper extremities, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7.

or

C. Raynaud's phenomenon, characterized by:

1. Gangrene involving at least two extremities; or

2. Ischemia with ulcerations of toes or fingers, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively as defined in 114.00C6 and 114.00C7;

or

D. Any other manifestation(s) of systemic sclerosis (scleroderma) resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.05 *Polymyositis and dermatomyositis.* As described in 114.00D4. With:

A. Proximal limb-girdle (pelvic or shoulder) muscle weakness, resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively as defined in 114.00C6 and 114.00C7.

or

B. Impaired swallowing (dysphagia) with aspiration due to muscle weakness.

or

C. Impaired respiration due to intercostal and diaphragmatic muscle weakness.

or

D. Diffuse calcinosis with limitation of joint mobility or intestinal motility.

or

E. Any other manifestation(s) of polymyositis or dermatomyositis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12;

or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.06 *Undifferentiated and mixed connective tissue disease.* As described in 114.00D5. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

B. Any other manifestation(s) of undifferentiated or mixed connective tissue disease resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.07 *Immune deficiency disorders, excluding HIV infection.* As described in 114.00E. With:

A. One or more of the following infections. The infection(s) must either be resistant to treatment or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

or

B. Stem cell transplantation as described under 114.00E3. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

or

C. Any other manifestation(s) of an immune deficiency disorder resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.08 *Human immunodeficiency virus (HIV) infection.* With documentation as described in 114.00F and one of the following:

A. Bacterial infections:

1. Mycobacterial infection (for example, caused by *M. avium-intracellulare*, *M. kansasii*, or *M. tuberculosis*) at a site other than the lungs, skin, or cervical or hilar lymph nodes, or pulmonary tuberculosis resistant to treatment; or

2. Nocardiosis; or

3. *Salmonella* bacteremia, recurrent non-typhoid; or

4. In a child less than 13 years of age, multiple or recurrent pyogenic bacterial infections (sepsis, pneumonia, meningitis, bone or joint infection, or abscess of an internal organ or body cavity, but not otitis media or superficial skin or mucosal abscesses) occurring two or more times in 2 years (for children age 13 and older, see 114.00F4c); or

5. Multiple or recurrent bacterial infections, including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment three or more times in a 12-month period.

or

B. Fungal infections:

1. Aspergillosis; or

2. Candidiasis involving the esophagus, trachea, bronchi, or lungs, or at a site other

than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes; or

3. Coccidioidomycosis, at a site other than the lungs or lymph nodes; or

4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis); or

5. Histoplasmosis, at a site other than the lungs or lymph nodes; or

6. Mucormycosis; or

7. *Pneumocystis* pneumonia or extrapulmonary *Pneumocystis* infection.

or

C. Protozoan or helminthic infections:

1. Cryptosporidiosis, isosporiasis, or microsporidiosis, with diarrhea lasting for 1 month or longer; or

2. Strongyloidiasis, extra-intestinal; or

3. Toxoplasmosis of an organ other than the liver, spleen, or lymph nodes.

or

D. Viral infections:

1. *Cytomegalovirus* disease (documented as described in 114.00F3b(ii)) at a site other than the liver, spleen, or lymph nodes; or

2. Herpes simplex virus causing:

a. Mucocutaneous infection (for example, oral, genital, perianal) lasting for 1 month or longer; or

b. Infection at a site other than the skin or mucous membranes (for example, bronchitis, pneumonitis, esophagitis, or encephalitis); or

c. Disseminated infection; or

3. Herpes zoster:

a. Disseminated; or

b. With multidermatomal eruptions that are resistant to treatment; or

4. Progressive multifocal leukoencephalopathy.

or

E. Malignant neoplasms:

1. Carcinoma of the cervix, invasive, FIGO stage II and beyond; or

2. Kaposi's sarcoma with:

a. Extensive oral lesions; or

b. Involvement of the gastrointestinal tract, lungs, or other visceral organs; or

3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease); or

4. Squamous cell carcinoma of the anal canal or anal margin.

or

F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above), with extensive fungating or ulcerating lesions not responding to treatment (for example, dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal *Candida*, condyloma caused by human *Papillomavirus*, genital ulcerative disease).

or

G. Neurological manifestations of HIV infection (for example, HIV encephalopathy, peripheral neuropathy) resulting in one of the following:

1. Loss of previously acquired, or marked delay in achieving, developmental milestones or intellectual ability (including the sudden onset of a new learning disability);

or

2. Impaired brain growth (acquired microcephaly or brain atrophy—see 114.00F4b); or

3. Progressive motor dysfunction affecting gait and station or fine and gross motor skills.

or

H. *Immune suppression and growth failure* (see 114.00F4d) documented by 1 and 2, or by 1 and 3.

1. CD4 measurement:

a. *For children from birth to attainment of age 5*, CD4 percentage of less than 20 percent; or

b. *For children age 5 to attainment of age 18*, absolute CD4 count of less than 200 cells/mm^3, or CD4 percentage of less than 14 percent; and

2. *For children from birth to attainment of age 2*, three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or

3. *For children age 2 to attainment of age 18*, three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.

I. Diarrhea, lasting for 1 month or longer, resistant to treatment and requiring intravenous hydration, intravenous alimentation, or tube feeding.

or

J. Lymphoid interstitial pneumonia/pulmonary lymphoid hyperplasia (LIP/PLH complex), with respiratory symptoms that significantly interfere with age-appropriate activities, and that cannot be controlled by prescribed treatment.

or

K. One or more of the following infections (other than described in A–J, above). The infection(s) must either be resistant to treatment or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

or

L. Any other manifestation(s) of HIV infection, including those listed in 114.08A–K, but without the requisite findings for those listings (for example, oral candidiasis not meeting the criteria in 114.08F, diarrhea not meeting the criteria in 114.08I), or other manifestation(s) (for example, oral hairy leukoplakia, hepatomegaly), resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.09 *Inflammatory arthritis.* As described in 114.00D6. With:

A. Persistent inflammation or persistent deformity of:

1. One or more major peripheral weight-bearing joints resulting in the inability to ambulate effectively (as defined in 114.00C6); or

2. One or more major peripheral joints in each upper extremity resulting in the inability to perform fine and gross movements effectively (as defined in 114.00C7).

or

B. Inflammation or deformity in one or more major peripheral joints with:

1. Involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

or

C. Ankylosing spondylitis or other spondyloarthropathies, with:

1. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 45° or more of flexion from the vertical position (zero degrees); or

2. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 30° or more of flexion (but less than 45°) measured from the vertical position (zero degrees), and involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity.

or

D. Any other manifestation(s) of inflammatory arthritis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.10 *Sjögren's syndrome.* As described in 114.00D7. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity; and

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

B. Any other manifestation(s) of Sjögren's syndrome resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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EDITORIAL NOTE: For FEDERAL REGISTER citations affecting appendix 1, see the List of CFR Sections Affected, which appears in the Finding Aids section of the printed volume and at *www.fdsys.gov*.

APPENDIX 2 TO SUBPART P OF PART 404—MEDICAL-VOCATIONAL GUIDELINES

Sec.

200.00 *Introduction.* (a) The following rules reflect the major functional and vocational patterns which are encountered in cases