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**Revised Medical Criteria for Evaluating
Immune System Disorders; Final rule**

SOCIAL SECURITY ADMINISTRATION

20 CFR Part 404

[Docket No. SSA 2006-0070]

RIN 0960-AF33

Revised Medical Criteria for Evaluating Immune System Disorders

AGENCY: Social Security Administration.

ACTION: Final Rules.

SUMMARY: We are revising the criteria in the Listing of Impairments (the listings) that we use to evaluate claims involving immune system disorders. We apply these criteria when you claim benefits based on disability under title II and title XVI of the Social Security Act (the Act). The revisions reflect our adjudicative experience, as well as advances in medical knowledge, treatment, and methods of evaluating immune system disorders.

DATES: These rules are effective June 16, 2008.

FOR FURTHER INFORMATION CONTACT: Paul Scott, Office of Compassionate Allowances and Listings Improvement, Social Security Administration, 4422 Annex Building, 6401 Security Boulevard, Baltimore, Maryland 21235-6401, (410) 966-1192. For information on eligibility or filing for benefits, call our national toll-free number, 1-800-

772-1213 or TTY 1-800-325-0778, or visit our Internet Web site, Social Security Online at <http://www.socialsecurity.gov/>.

SUPPLEMENTARY INFORMATION:

Electronic Version

The electronic file of this document is available on the date of publication in the **Federal Register** at <http://www.gpoaccess.gov/fr/index.html>.

Background

We are revising and making final the rules we proposed for evaluating immune system disorders in the Notice of Proposed Rulemaking (NPRM) published in the **Federal Register** on August 4, 2006 (71 FR 44432, corrected at 71 FR 46983). We provide a summary of the provisions of the final rules below, with an explanation of the changes we have made from the text in the NPRM. We then provide summaries of the public comments on the NPRM and our reasons for adopting or not adopting the recommendations in those comments in the section "Public Comments on the NPRM." The final rule language follows that section.

What Programs Do These Final Rules Affect?

These final rules affect disability determinations and decisions that we

make under title II and title XVI of the Act. In addition, to the extent that Medicare entitlement and Medicaid eligibility are based on whether you qualify for disability benefits under title II and title XVI, these final rules also affect the Medicare and Medicaid programs.

Who Can Get Disability Benefits?

Under title II of the Act, we provide for the payment of disability benefits if you are disabled and belong to one of the following three groups:

- Workers insured under the Act,
- Children of insured workers, and
- Widows, widowers, and surviving divorced spouses (see § 404.336) of insured workers.

Under title XVI of the Act, we provide for Supplemental Security Income (SSI) payments on the basis of disability if you are disabled and have limited income and resources.

How do we define disability?

Under both the title II and title XVI programs, disability must be the result of any medically determinable physical or mental impairment or combination of impairments that is expected to result in death or which has lasted or is expected to last for a continuous period of at least 12 months. Our definitions of disability are shown in the following table:

If you file a claim under . . .	And you are . . .	Disability means you have a medically determinable impairment(s) as described above that results in . . .
title II	an adult or a child	the inability to do any substantial gainful activity (SGA).
title XVI	an individual age 18 or older	the inability to do any SGA.
title XVI	an individual under age 18	marked and severe functional limitations.

How do we decide whether you are disabled?

If you are applying for benefits under title II of the Act, or if you are an adult applying for payments under title XVI of the Act, we use a five-step "sequential evaluation process" to decide whether you are disabled. We describe this five-step process in our regulations at §§ 404.1520 and 416.920. We follow the five steps in order and stop as soon as we can make a determination or decision. The steps are:

1. Are you working, and is the work you are doing substantial gainful activity? If you are working and the work you are doing is substantial gainful activity, we will find that you are not disabled, regardless of your medical condition or your age, education, and work experience. If you are not, we will go on to step 2.

2. Do you have a "severe" impairment? If you do not have an

impairment or combination of impairments that significantly limits your physical or mental ability to do basic work activities, we will find that you are not disabled. If you do, we will go on to step 3.

3. Do you have an impairment(s) that meets or medically equals the severity of an impairment in the listings? If you do, and the impairment(s) meets the duration requirement, we will find that you are disabled. If you do not, we will go on to step 4.

4. Do you have the residual functional capacity (RFC) to do your past relevant work? If you do, we will find that you are not disabled. If you do not, we will go on to step 5.

5. Does your impairment(s) prevent you from doing any other work that exists in significant numbers in the national economy, considering your RFC, age, education, and work experience? If it does, and it meets the duration requirement, we will find that

you are disabled. If it does not, we will find that you are not disabled.

We use a different sequential evaluation process for children who apply for payments based on disability under title XVI of the Act. We describe that sequential evaluation process in § 416.924 of our regulations. If you are already receiving benefits, we also use a different sequential evaluation process when we decide whether your disability continues. See §§ 404.1594, 416.994, and 416.994a of our regulations. However, all of the processes include steps at which we consider whether your impairment(s) meets or medically equals one of our listings.

What are the listings?

The listings are examples of impairments that we consider severe enough to prevent you as an adult from doing any gainful activity. If you are a child seeking SSI payments based on disability, the listings describe

impairments that we consider severe enough to result in marked and severe functional limitations. Although the listings are contained only in appendix 1 to subpart P of part 404 of our regulations, we incorporate them by reference in the SSI program in § 416.925 of our regulations and apply them to claims under both title II and title XVI of the Act.

How do we use the listings?

The listings are in two parts. There are listings for adults (part A) and for children (part B). If you are an individual age 18 or over, we apply the listings in part A when we assess your claim, and we never use the listings in part B.

If you are an individual under age 18, we first use the criteria in part B of the listings. Part B contains criteria that apply only to individuals who are under age 18. If the criteria in part B do not apply, we may use the criteria in part A when those criteria give appropriate consideration to the effects of the impairment(s) in children. (See §§ 404.1525 and 416.925.)

If your impairment(s) does not meet any listing, we will also consider whether it medically equals any listing; that is, whether it is as medically severe as an impairment in the listings. (See §§ 404.1526 and 416.926.)

What if you do not have an impairment(s) that meets or medically equals a listing?

We use the listings only to decide that you are disabled or that you are still disabled. We will not deny your claim or decide that you no longer qualify for benefits because your impairment(s) does not meet or medically equal a listing. If you have a severe impairment(s) that does not meet or medically equal any listing, we may still find you disabled based on other rules in the “sequential evaluation process.” Likewise, we will not decide that your disability has ended only because your impairment(s) no longer meets or medically equals a listing.

Also, when we conduct reviews to determine whether your disability continues, we will not find that your disability has ended because we have changed a listing. Our regulations explain that, when we change our listings, we continue to use our prior listings when we review your case, if you qualified for disability benefits or SSI payments based on our determination or decision that your impairment(s) met or medically equaled a listing. In these cases, we determine whether you have experienced medical improvement and, if so, whether the

medical improvement is related to the ability to work. If your condition has medically improved so that you no longer meet or medically equal the prior listing, we evaluate your case further to determine whether you are currently disabled. We may find that you are currently disabled, depending on the full circumstances of your case. See §§ 404.1594(c)(3)(i) and 416.994(b)(2)(iv)(A). If you are a child who is eligible for SSI payments, we follow a similar rule when we decide that you have experienced medical improvement in your condition. See § 416.994a(b)(2).

Why are we revising the listings for immune system disorders?

We are making these revisions to update the medical criteria in the listings and to provide more information about how we evaluate immune system disorders. We first published these rules in 1993 (58 FR 36008). At that time, we established body system listings for immune system disorders in part A and part B. We made those rules effective for 5 years from the date of publication, unless we extended them, or revised and issued them again (58 FR at 36051). Since that time, we have extended the expiration date of the immune body system listings but we have not comprehensively revised them.

We have, however, made several changes to these listings over the years. On November 19, 2001, we published final rules in the **Federal Register** adding listings 14.09 and 114.09, for inflammatory arthritis, to the immune system listings, and adding introductory text for those listings in sections 14.00B6 and 114.00E (66 FR 58009). We published minor technical changes to the immune system listings on February 24, 2002 (67 FR 20018).

How did we develop these final rules?

These final rules reflect our adjudicative experience and advances in medical knowledge, treatment, and methods of evaluating immune system disorders. They also reflect comments on the NPRM we published in 2006.

Before we developed the NPRM, we published an Advance Notice of Proposed Rulemaking (ANPRM) in the **Federal Register** on May 9, 2003 (68 FR 24896). The purpose of the ANPRM was to inform the public that we were planning to update and revise the rules we use to evaluate immune system disorders and to invite interested individuals and organizations to send us comments and suggestions for updating and revising the immune system listings. In the ANPRM, we provided a 60-day period for comments and

suggestions; that period ended on July 8, 2003. We received over 200 letters and e-mails in response to the notice, many from individuals who have immune system disorders or who have family members with such disorders. We also received comments from medical experts, advocates, and people who adjudicate claims for us. Although we are not summarizing or responding to the ANPRM comments in these final rules, we read and considered them carefully.

We also hosted policy conferences on “Immune System Disorders in the Disability Programs” in Philadelphia, PA, on December 15, 2003, and in San Francisco, CA, on February 18 and 19, 2004. At these conferences, we heard comments and suggestions for updating and revising these rules from individuals who have immune system disorders and their family members, physicians who treat individuals with immune system disorders, other professionals who work with people who have immune system disorders, advocates who represent individuals with immune system disorders, and individuals who make disability determinations and decisions for us in the State agencies and the Office of Disability Adjudication and Review (formerly called the Office of Hearings and Appeals).

As already noted, these final rules also reflect comments we asked you to provide on the NPRM. We summarize and respond to those comments later in this preamble. Throughout this preamble, we refer to “public comments on the NPRM” whenever we refer to these comments to distinguish them from public comments we received on the ANPRM and at the outreach meetings.

What do we mean by “final rules” and “prior rules”?

Even though these rules will not go into effect until 90 days after publication of this notice, for clarity, we refer to the changes we are making here as the “final rules” and to the rules that will be changed by these final rules as the “prior rules.”

When will we start to use these final rules?

We will start to use these final rules on their effective date. We will continue to use our prior rules until the effective date of these final rules. When these final rules become effective, we will apply them to new applications filed on or after the effective date of these rules and to claims pending before us, as we describe below.

As is our usual practice when we make changes to our regulations, we will apply these final rules on or after their effective date whenever we make a determination or decision, including in those claims in which we make a determination or decision after a remand to us from a Federal court. With respect to claims in which we have made a final decision and that are pending judicial review in Federal court, we expect that the court would review the Commissioner's final decision in accordance with the rules in effect at the time the final decision of the Commissioner was issued. If a court reverses the Commissioner's final decision and remands the case for further administrative proceedings after the effective date of these final rules, we will apply the provisions of these final rules to the entire period at issue in the claim in our new decision issued pursuant to the court's remand.

How long will these final rules be effective?

These final rules will no longer be effective 8 years after the date on which they become effective, unless we extend them or revise and issue them again. However, we intend to monitor these rules, and if needed, will update the criteria for any impairment in these rules before the end of the 8-year period.

What revisions are we making with these final rules?

We are revising the prior rules to:

- Expand, reorganize, and update the introductory text in final 14.00 and 114.00 to provide more guidance for our adjudicators, and to reflect the revised listings.

- Add paragraph headings to the introductory text in final 14.00 and 114.00 for easier reference.

- Add final 14.00C and 114.00C to explain the meaning of key terms.

- Remove all reference listings.

Reference listings are listings that are met by satisfying the criteria of another listing. For example, prior listing 14.08G1 for human immunodeficiency virus (HIV) infection with anemia was a reference listing that required evaluation under current listing 7.02 for chronic anemia. Therefore, prior listing 14.08G1 was redundant. In some cases, instead of using reference listings, we provide general guidance in the introductory text for the immune system disorders listings (final 14.00J2g) stating that impairments in other body systems that result from immune system disorders should be evaluated under the criteria of the affected body system. In other cases, we are replacing reference listings with specific listing criteria that

are appropriate for evaluation under this body system. For example, prior listing 14.06, for undifferentiated connective tissue disorders, was entirely a reference listing. In the final rules, we are replacing the reference listing criterion with criteria that are specific to these disorders.

- Add final listings 14.10 and 114.10 for evaluating Sjögren's syndrome.

- Add functional criteria to the listings, similar to those in prior HIV infection listings 14.08N and 114.08O, for each of the other listed immune system disorders (for example, systemic lupus erythematosus and systemic vasculitis).

- Make nonsubstantive editorial changes to update the medical terminology in the introductory text and the listings and to make their language simpler and clearer.

How are we changing the introductory text for the immune system disorders listings for adults?

We are expanding and reorganizing the introductory text for these listings. There were four major sections in prior 14.00, and the longest of those sections, 14.00D, addressed only the evaluation of HIV infection. In these final rules, we are adding more sections and expanding the guidance we provide about evaluating other kinds of immune system disorders.

Some of the guidance in prior 14.00D was useful for evaluating other kinds of immune system disorders in addition to HIV infection. Therefore, we are moving that guidance from prior 14.00D to new sections that have more general applicability to immune system disorders. We are not removing any substantive guidance about how we evaluate HIV infection, only reorganizing some of the information that was in 14.00D of the prior rules and giving it broader applicability where appropriate. We are also updating and expanding some of the guidance for evaluating HIV infection and its effects that was in the prior rules, as we describe in more detail below.

The four sections in the prior rules were:

- Prior 14.00A, a short paragraph that described generally the kinds of disorders we include in this body system.

- Prior 14.00B, a lengthy section that discussed the evaluation of connective tissue disorders; that is, autoimmune disorders. It included six undesignated paragraphs that primarily explained the kinds of evidence we need to document the existence and severity of these disorders, including how we evaluate loss of function. These paragraphs were

followed by six numbered sections that provided guidance about specific impairments in the listings.

- Prior 14.00C, a single sentence that explained that we evaluate allergic disorders under the appropriate listing of the affected body system.

- Prior 14.00D, a lengthy section that explained how we documented the existence and severity of HIV infection, including how we evaluated loss of function under prior listing 14.08N. It included eight numbered subsections and many paragraphs that were not designated with letters or numbers within those subsections.

In the final rules, there are 10 sections in the introductory text. The first three sections (final 14.00A, B, and C) provide general information about this body system, including definitions of terms. Each of the next three sections describes a particular category or type of immune system disorder: Autoimmune disorders (final 14.00D); immune deficiency disorders, excluding HIV infection (final 14.00E); and HIV infection (final 14.00F). The next three sections explain how we consider the effects of your treatment (final 14.00G), your symptoms (final 14.00H), and the functional limitations from your immune system disorder under these listings (final 14.00I). The last section, final 14.00J, explains how we consider the effects of your immune system disorder when it does not meet the requirements of one of the immune system disorders listings. We are designating all paragraphs in the final rules with letters or numbers for easier reference. We are also providing headings for all of the major sections and many of the subsections.

The following are the names of the major sections in final 14.00. We describe each section in detail later in this preamble.

- Final 14.00A: *What disorders do we evaluate under the immune system disorders listings?*

- Final 14.00B: *What information do we need to show that you have an immune system disorder?*

- Final 14.00C: *Definitions*

- Final 14.00D: *How do we document and evaluate the listed autoimmune disorders?*

- Final 14.00E: *How do we document and evaluate immune deficiency disorders, excluding HIV infection?*

- Final 14.00F: *How do we document and evaluate human immunodeficiency virus (HIV) infection?*

- Final 14.00G: *How do we consider the effects of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?*

- Final 14.00H: *How do we consider your symptoms, including your pain, severe fatigue, and malaise?*

- Final 14.00I: *How do we use the functional criteria in these listings?*

- Final 14.00J: *How do we evaluate your immune system disorder when it does not meet one of these listings?*

The following is a detailed description of the changes in the introductory text.

14.00 Immune System Disorders

We are changing the name of this body system from “Immune System” to “Immune System Disorders” to more accurately reflect that we use these listings to evaluate immune system disorders in accordance with the requirements of the disability program.

Final 14.00A—What disorders do we evaluate under the immune system disorders listings?

In final 14.00A, we provide a brief overview of this body system. We explain the kinds of disorders we evaluate under the immune system disorders listings and that we organize these impairments under the categories of “autoimmune disorders,” “immune deficiency disorders, excluding HIV infection,” and “HIV infection.” Final 14.00A has four subsections.

We incorporate prior 14.00A in the opening sentence of final 14.00A1. We are revising the sentence, which explains the kinds of immune system dysfunction that immune system disorders may cause, to update and simplify it. In final 14.00A1a and 14.00A1b, we incorporate the first sentence in the sixth paragraph of prior 14.00B to explain that immune system disorders can cause dysfunction in one or more components of the immune system, and describe ways in which immune system disorders may result in loss of function. In the third sentence of final 14.00A1b, we are adding “involuntary” as a descriptor of weight loss to clarify that we mean weight loss due to an immune system disorder(s) or its treatment. We are adding “involuntary” as a descriptor of weight loss throughout the introductory text in part A and part B for this same reason. Final 14.00A1c is a new paragraph that explains how we have organized the discussions of immune system disorders in the introductory text for these listings.

In final 14.00A2, *Autoimmune disorders*, we incorporate the first paragraph in prior 14.00B to provide a brief description of autoimmune disorders. We are adding an explanation that these disorders are sometimes referred to as “rheumatic diseases,”

“connective tissue disorders,” or “collagen vascular disorders,” and that some of the features of these disorders in adults differ from the features of the same disorders in children. We provide a cross-reference to final 14.00D, the section of the introductory text that addresses autoimmune disorders in detail. We are also removing the last sentence of the first paragraph of prior 14.00B, which explained that connective tissue disorders generally evolve and persist over time, may result in functional loss, and may require long-term, repeated evaluation and management, because it did not provide useful adjudicative guidance. However, we do explain in final 14.00A1b that immune system disorders can cause “extreme” loss of function. We also explain parenthetically that “extreme” means “very serious” to make clear that we use the term “extreme” in the same way that we use it in other body systems; for example, see 1.00B2b1 and 1.00B2c in the musculoskeletal system.

Final 14.00A3, *Immune deficiency disorders, excluding HIV infection*, is new. We explain that these disorders can be classified as “primary” or “acquired,” are characterized by recurrent or unusual infections, and are associated with an increased risk of malignancies and of other autoimmune disorders. We also provide a cross-reference to final 14.00E, the section of the introductory text that addresses immune deficiency disorders in detail.

In final 14.00A4, *Human immunodeficiency virus (HIV) infection*, we provide a brief description of HIV infection. As in the NPRM, we include the first sentence from prior 14.00D1 in this section. However, in an editorial change from the prior rules and the NPRM, we have deleted the statement in the sentence that HIV infection is “caused by a specific retrovirus.” The change is not substantive, but only clarifies and updates our rules. It is now known that there are several forms of human immunodeficiency virus, therefore our statement that HIV infection is caused by “a specific” virus could be misleading. Also, since the “V” in the abbreviation “HIV” stands for “virus,” the sentence in the prior rules did not need to state that human immunodeficiency virus infection is caused by a virus. We have retained the rest of the sentence, which explains that HIV infection may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions. We also provide a cross-reference to final 14.00F, the section of the introductory text that addresses HIV infection in detail.

Final 14.00B—What information do we need to show that you have an immune system disorder?

In final 14.00B, we incorporate the first sentence of the second paragraph of prior 14.00B to explain what information we need to show that you have an immune system disorder. We moved the second and third sentences of the second paragraph of prior 14.00B, which define our term “appropriate medically acceptable imaging,” to final 14.00C, a new section that provides definitions of terms in these listings. We are removing the last two sentences of the prior paragraph, which explained that we would not purchase tests that may involve significant risk. Since we already include this general policy in §§ 404.1519m and 416.919m of our regulations, it is not necessary to repeat it in this section. However, as we explain below, we are including guidance about the purchase of certain tests in other sections of these final rules.

In the second sentence of final 14.00B, we provide that “we will make every reasonable effort” to obtain your medical history, medical findings, and the results of laboratory tests in documenting whether you have an immune system disorder. We included this requirement in prior 14.00D for HIV infection, but we did not include similar guidance in prior 14.00B for connective tissue disorders. We are adding this guidance under final 14.00B because it is appropriate for all immune system disorders.

We also are removing the third and fourth paragraphs of prior 14.00B. The third paragraph of prior 14.00B provided that we need a longitudinal clinical record of at least 3 months demonstrating active disease to assess the severity and duration of your impairment. This was not always the case, even under the prior rules. For example, individuals with HIV infection and cryptococcal meningitis (prior and final listing 14.08B4) or Kaposi’s sarcoma (prior and final listing 14.08E2), and individuals with ankylosing spondylitis with fixation (ankylosis) of the dorsolumbar spine at 45° (prior listing 14.09B2, final listing 14.09C1) are disabled based on those findings alone. In these cases, we do not need 3 months of evidence or evidence showing active disease. Other cases may be decided with less than 3 months of evidence, while others may require more than 3 months of evidence. Therefore, we are removing this guidance because we must decide each case on an individual basis.

Final 14.00C—Definitions

In final 14.00C, we define what we mean by important terms in these listings. As already noted, we include the definition of “appropriate medically acceptable imaging” from the second paragraph of prior 14.00B. However, in an editorial change from the NPRM, we are revising the definition of “appropriate” imaging from “one that is generally accepted and consistent with the prevailing state of medical knowledge and clinical practice” to “the proper one to support the evaluation and diagnosis of the impairment” to be consistent with the language used in other body system listings, for example, the musculoskeletal body system (see 1.00C1) and hematological disorders body system (see 7.00B). We are also including in this new section the definitions of the terms “severe” from the sixth paragraph of prior 14.00B, “inability to ambulate effectively” and “inability to perform fine and gross movements effectively” from prior 14.00B6b, and “resistant to treatment,” “recurrent,” and “disseminated” from the second, third, and fourth paragraphs of prior 14.00D2. All of these terms apply to several, and sometimes all, of the final listings in this body system.

In final 14.00C, we do not include the phrase “must have lasted, or be expected to last, for at least 12 months” from the definitions of “inability to ambulate effectively” and “inability to perform fine and gross movements effectively” that was in prior 14.00B6b because we believe it is unnecessary. Unless an impairment is expected to result in death, it must have lasted or must be expected to last for a continuous period of at least 12 months to meet the definition of disability. This change also makes the definitions of the terms consistent with the definitions of the same terms in 1.00B2b and 1.00B2c in the musculoskeletal body system.

We are also including, but simplifying, the definitions of the terms “resistant to treatment,” “recurrent,” and “disseminated” that were in prior 14.00D2, primarily to remove language that we believe was unnecessary. For example, we removed the explanation that the terms “have the same general meaning as used by the medical community.” These changes are editorial only, and the final definitions are not substantively different from the prior rules.

In final 14.00C2, we are adding the definitions of several other important terms in these listings, including the term “constitutional symptoms or signs.” We are revising this definition slightly in response to a public

comment on the NPRM to indicate that for purposes of these listings the constitutional symptoms or signs are severe fatigue, fever, malaise, and involuntary weight loss. In the proposed rules, we inadvertently referred to “fatigue” in our definition of constitutional symptoms or signs, rather than “severe fatigue.” We did, however, include a separate definition for “severe fatigue” because it is the criterion we use in all of the listings that include criteria for constitutional symptoms or signs. The change in the definition we are making in these final rules makes no substantive difference to the application of the listings, makes this definition consistent with the criteria of the listings, and more accurately reflects our intent.

As in the NPRM, we are also providing a definition for the term “malaise.” We are adding the definitions for severe fatigue and malaise in response to the many comments we received before we developed the proposed rules that indicated that the fatigue and malaise that people who have immune system disorders experience can be very limiting.

In final 14.00C8, we reference current 1.00F for the definition of “major peripheral joints” instead of restating the definition as we did in prior 14.00B6a.

In final 14.00C12, we change “describes” to “means.” This is an editorial change from the NPRM for consistency with the other definitions in this section.

Final 14.00D—How do we document and evaluate the listed autoimmune disorders?

We are changing the heading of proposed 14.00D in response to a public comment on the NPRM that we describe in the public comments section of this preamble. In final 14.00D, we are incorporating and expanding upon the information in prior 14.00B1 through 14.00B6, which described features commonly associated with each of the listed autoimmune system disorders. Throughout these sections, we refer to “autoimmune disorders” instead of “connective tissue disorders” because the phrase “autoimmune disorders” is more medically accurate and more frequently used by medical professionals. We are also adding section 14.00D7 for Sjögren’s syndrome because we are adding listing 14.10 for that autoimmune disorder.

In final 14.00D1, *Systemic lupus erythematosus (14.02)*, we expand and clarify the information in prior 14.00B1. In final 14.00D1a, *General*, we explain

that systemic lupus erythematosus (SLE) may involve any organ or body system and describe by body system some potential manifestations of SLE. We expand our explanation of how SLE is frequently characterized clinically. We are changing the reference to “fatigability” used in prior 14.00B1 to “severe fatigue” to be consistent with how we describe the constitutional symptoms throughout the final immune system disorders listings. We are also adding “involuntary” as a descriptor of weight loss to clarify that we mean weight loss due to SLE or its treatment, and to be consistent with our addition of this word throughout the introductory text and listings, as we have already explained.

In final 14.00D1b, *Documentation of SLE*, we are updating our rules to explain that your medical evidence will generally, but not always, show that your SLE satisfies the criteria in the “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. This is a more up-to-date reference than the 1982 reference in the prior rules.

In final 14.00D2, *Systemic vasculitis (14.03)*, we clarify the information in the prior rule. Final 14.00D2a, *General*, corresponds to the first three sentences of prior 14.00B2. In it, we explain what vasculitis is, and that it may be associated with other autoimmune disorders. We also give examples of several clinical patterns in which it may occur. We are removing the fourth sentence of prior 14.00B2, which described cutaneous vasculitis, because the impairment varies greatly in its manifestation, may not be associated with systemic involvement, and would not be expected to result in a listing-level impairment.

Final 14.00D2b, *Documentation of systemic vasculitis*, corresponds to the last two sentences of prior 14.00B2. In it, we describe the documentation that is used to confirm the diagnosis of systemic vasculitis. In response to a comment described later in this preamble, we are expanding the guidance we provide in this section to explain that we will make “every reasonable effort” to obtain reports of angiography or tissue biopsy when they are part of your medical records. However, we will not purchase these invasive and costly procedures.

Final 14.00D3, *Systemic sclerosis (scleroderma) (14.04)*, corresponds to prior 14.00B3. We are revising the heading and expanding the information that was in the prior section. Final

14.00D3a, *General*, corresponds to the first three sentences of prior 14.00B3. We are changing the term “Raynaud’s phenomena,” which we used in the second and third sentences of prior 14.00B3, to “Raynaud’s phenomenon” because the latter is the correct term. We make this same change in final listing 14.04C. In final 14.00D3b, *Diffuse cutaneous systemic sclerosis*, we continue to explain that, in addition to skin or blood vessels, major organ or systemic involvement may include the gastrointestinal tract, lungs, heart, kidneys, and muscle. This guidance corresponds to the fourth sentence in prior 14.00B3.

Final 14.00D3c, *Localized scleroderma (linear scleroderma or morphea)*, is new. We are adding this section and appropriate listings in final 14.04 for these disorders that originate in childhood because their disabling effects can persist into adulthood. Final 14.00D3c is essentially the same as final 114.00D3c, which we describe in detail later in this preamble. We are also making minor editorial changes from the language we proposed in the NPRM for clarity.

Final 14.00D3d, *Documentation of systemic sclerosis (scleroderma)*, is also new. In it, we explain what documenting systemic sclerosis (scleroderma) involves and that there may be an overlap with other autoimmune disorders.

In final 14.00D4, *Polymyositis and dermatomyositis (14.05)*, we clarify the information in prior 14.00B4. Final 14.00D4a, *General*, corresponds to the first three sentences of prior 14.00B4. It describes the characteristics of polymyositis and dermatomyositis. In the final rule, we have made minor editorial changes from the language we proposed in the NPRM.

In final 14.00D4b, *Documentation of polymyositis or dermatomyositis*, we describe the findings that are generally used to document these impairments. The first sentence of the final rule corresponds to the last sentence of prior 14.00B4. We are making minor editorial revisions to the prior rules, including the removal of the reference to “myositis,” because there are multiple characteristic abnormalities on muscle biopsy that support the diagnosis of polymyositis or dermatomyositis. We also are adding a sentence to explain that people with dermatomyositis have characteristic skin findings. In response to a comment described later in this preamble, we are expanding the guidance we provide in this section to explain that we will make “every reasonable effort” to obtain reports of electromyography or muscle biopsy

when they are part of your medical records. However, we will not purchase these procedures.

In final 14.00D4c, *Additional information about how we evaluate polymyositis and dermatomyositis under the listings*, we explain how we evaluate commonly occurring limitations associated with these disorders. Final 14.00D4c(i) corresponds to the fourth and fifth sentences of prior 14.00B4. We are deleting the example of weakness of the anterior neck flexor muscles in the sixth sentence of prior 14.00B4 because we are deleting the reference to the cervical muscles from listing 14.05 for reasons we explain later in this preamble. We are adding an example of rising independently from a squatting position because this is a common means for evaluating weakness in the pelvic girdle muscles.

In final 14.00D4c(ii), we explain that we will evaluate malignancies (which may be associated with these disorders) under the malignant neoplastic diseases listings (13.00). (We do not provide this guidance in final 114.00D4c in the part B (childhood) section for polymyositis or dermatomyositis because malignancies are not commonly associated with these disorders in children.) We also explain that we evaluate the involvement of other organs or body systems under the affected body system.

In final 14.00D5, *Undifferentiated and mixed connective tissue disease (14.06)*, we reorganize and clarify the information from prior 14.00B5. In the final rules, we are adding an explicit reference to mixed connective tissue disease (MCTD) to clarify what we meant in the prior rules when we referred to “overlap” syndromes. This is not a substantive change, but a clarification of our prior rules to update medical terminology. In final 14.00D5a, *General*, we describe what we mean by undifferentiated and mixed connective tissue disease. In final 14.00D5b, *Documentation of undifferentiated and mixed connective tissue disease*, we explain when clinical features and serologic findings may be used to diagnose undifferentiated and mixed connective tissue disease. These provisions in final 14.00D5a and 14.00D5b are not substantively different from the provisions in the first three sentences of prior 14.00B5.

We are removing the last sentence of prior 14.00B5. The sentence indicated that the correct designation of an “overlap” disorder is important for the assessment of prognosis. While the correct designation of an “overlap” disorder is useful in treatment settings, in our experience the requirement in

our prior rules was not useful for adjudication.

In final 14.00D6, *Inflammatory arthritis (14.09)*, we expand, reorganize, and clarify the rules in prior 14.00B6. Throughout final 14.00D6, we are simplifying the language of the NPRM, in which we used the rarely encountered word “arthritides”; that is, the plural form of “arthritis.” Instead, we use the terms “arthritis,” and in final 14.00D6a, “the spectrum of inflammatory arthritis.”

Final 14.00D6a, *General*, corresponds to the first and fourth sentences of prior 14.00B6. We continue to explain that inflammatory arthritis includes a vast array of disorders that differ in cause, course, and outcome, and that may result in difficulties with ambulation or fine and gross movements. We edited the fourth sentence of prior 14.00B6 to break it into three shorter sentences. However, we did not change the meaning of the provision. In addition to changing the term “arthritides” from the NPRM, we also made minor editorial changes in the final paragraph for clarity.

Final 14.00D6b, *Inflammatory arthritis involving the axial spine (spondyloarthritis)*, and final 14.00D6c, *Inflammatory arthritis involving the peripheral joints*, correspond to the second and third sentences of prior 14.00B6. In these sections, we list some disorders that may be associated with inflammatory arthritis involving the axial spine (final 14.00D6b) and inflammatory arthritis affecting the peripheral joints (final 14.00D6c). We are including inflammatory bowel disease (IBD) in the lists of examples of specific disorders in these sections because arthritis is the most common extra-intestinal complication of IBD. In final 14.00D6b, we are not including the examples of “other reactive arthropathies” and “undifferentiated spondylitis,” which were in the second sentence of prior 14.00D6, because they are non-specific and we do not intend to provide a complete list, only some examples. Finally, we are updating some of the terminology in this section. For example, we refer to “psoriatic arthritis” instead of “psoriatic arthropathy.”

Final 14.00D6d, *Documentation of inflammatory arthritis*, is new. In it, we explain that 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*.

Final 14.00D6e, *How we evaluate inflammatory arthritis under the listings*, corresponds to the information

in the last two sentences of prior 14.00B6, prior 14.00B6c, and prior 14.00B6d. We are reorganizing the text to reflect the reorganization of listing 14.09, which we explain later in this preamble, and to clarify it. We are also making changes to 14.00D6e in response to a public comment on the NPRM, as explained below and in the public comments section of this preamble.

- Final 14.00D6e(i) explains that final listings 14.09A and 14.09C1 (prior listings 14.09A and 14.09B) are met by showing an impairment that results in an “extreme” limitation. This is how we describe “inability to ambulate effectively” in 1.00B2b in our musculoskeletal listings and, therefore, it is only a clarification of the prior rule. In the final rule, we retain the provision from prior 14.00B6c that the inability to ambulate effectively is implicit in final listing 14.09C1 (prior listing 14.09B), the listing for ankylosis of the spine with fixation at a 45° angle, even though individuals who have the degree of ankylosis described in the listing ordinarily do not require the use of bilateral upper limb assistance.

A public commenter on the NPRM pointed out that proposed (and prior) listing 14.09 did not account for individuals who are unable to ambulate effectively because of involvement of a major peripheral joint in one lower extremity, requiring our adjudicators to refer to listings 1.02 and 1.03 in those cases. In response to this comment, we decided to simplify our rules so that there is no longer a need to cross-refer to the listings in the musculoskeletal system. We revised listing 14.09 (and listing 114.09) so that all individuals with inflammatory arthritis who are unable to ambulate effectively or to use their upper extremities effectively can qualify under the inflammatory arthritis listing. As a consequence, we revised this section to reflect the revised listing criteria. We also removed proposed 14.00D6e(iv) and 14.00D6e(v) as explained below. (For clarity, we are also revising a sentence in 1.00B1 and 101.00B1 in the musculoskeletal system listings. We describe this and the public comment that led to these changes in the public comments section of this preamble.)

- Final 14.00D6e(ii) explains final listings 14.09B (prior listing 14.09D), 14.09C2 (prior listing 14.09E), and 14.09D. We revised the language in the NPRM to more clearly explain that listing-level severity can result from various combinations of complications from inflammatory arthritis. This is not a substantive change, only a clarification. In this section, we also incorporate the provision in the first

sentence of prior 14.00B6d that extra-articular impairments may meet listings in other body systems.

- Final 14.00D6e(iii) corresponds to the third and fourth sentences of prior 14.00B6d. It explains that extra-articular features of inflammatory arthritis may involve any body system and lists examples of commonly occurring extra-articular impairments by body system. We are reorganizing and expanding the list of examples of such impairments from the prior rules and clarifying the body systems to which they belong. We are also making a minor editorial change to the sentence we proposed. In the NPRM, we introduced the list of examples with the statement “Commonly occurring extra-articular impairments include * * *.” However, the list that followed was actually a list of body systems, each of which contained parenthetical examples of specific impairments. In the final rules, we are providing a more accurate introduction to the list of examples of body systems and their parenthetical examples.

- As indicated above, we removed proposed 14.00D6e(iv) and 14.00D6e(v) in response to a public comment. These sections corresponded to the last sentence of prior 14.00B6, which explained that we used listing 1.02 or 1.03 in the musculoskeletal system when the dominant feature of the impairment was persistent deformity without ongoing inflammation or when there had been surgical reconstruction.

- Final 14.00D6e(iv) (proposed 14.00D6e(vi)) clarifies that we evaluate your impairment under any appropriate listing when you have both inflammation and chronic deformities.

We are not including the provisions of prior 14.00B6e in these final rules. Prior 14.00B6e provided that the fact that an individual is dependent on steroids, or any other drug, for the control of inflammatory arthritis is insufficient in itself to establish disability. We added it to part A of our listings in 2002 for consistency with 114.00E6, a provision we added to part B of the listings at the same time (66 FR at 58020 (2001)). We are removing that provision for reasons we explain below in our summary of the final rules in part B. Therefore, we are removing this provision in part A for consistency with that change. However, in final 14.00G3, we continue to state that we will consider the adverse side effects of treatment, including the adverse effects of corticosteroids, to ensure that our adjudicators consider the side effects an individual might experience from steroids and any other treatment.

Final 14.00D7, *Sjögren’s syndrome (14.10)*, is new. As already noted, we are adding a listing for Sjögren’s syndrome. In connection with that final listing, final 14.00D7a, *General*, explains the features of the disorder, including its resulting symptoms and possible complications. We also list organ systems that may be involved and note that Sjögren’s syndrome may be associated with other autoimmune disorders. In final 14.00D7b, *Documentation of Sjögren’s syndrome*, we also explain that if you have Sjögren’s syndrome, your 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” found in the most recent edition of the *Primer on the Rheumatic Diseases*.

Final 14.00E—How do we document and evaluate immune deficiency disorders, excluding HIV infection?

We changed the heading of proposed 14.00E in response to a public comment on the NPRM that we describe in the public comments section of this preamble. In final 14.00E, we add a section describing how immune deficiency disorders (excluding HIV infection) are classified, documented, and evaluated. This section has four subsections.

- In final 14.00E1, *General*, we explain that immune deficiency disorders are classified as either “primary” or “acquired.” Primary disorders are mainly seen in children but, due to recent advances in treatment, many affected children survive into adulthood.

- In final 14.00E2, *Documentation of immune deficiency disorders*, we explain that documentation of these disorders may be based on laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

- In final 14.00E3, *Immune deficiency disorders treated by stem cell transplantation*, we explain how we evaluate immune deficiency disorders that are treated in this way. In final 14.00E3a, *Evaluation in the first 12 months*, we explain that if you undergo stem cell transplantation, we will consider you disabled until at least 12 months from the date of the transplant. This is the same provision that we use for most malignancies treated by bone marrow or stem cell transplants in the neoplastic listings. In 13.00L3b of the malignant neoplastic diseases body system, we also include a special provision for autologous bone marrow transplants—transplants using your own

stem cells. We do not include such an alternative provision in these final rules because people with immune deficiency disorders receive allogeneic transplants—that is, stem cells taken from other people. Also, unlike in the rules in the malignant neoplastic diseases body system, we use the phrase “stem cell transplantation” instead of “bone marrow or stem cell transplantation” in this final section and in final listing 14.07B because “stem cell transplantation” is a broader term that encompasses different sites for obtaining hematopoietic (blood-forming) stem cells, including bone marrow, peripheral blood, and umbilical cord blood. In final 14.00E3b, *Evaluation after the 12-month period has elapsed*, we explain that after this period has elapsed, we consider any demonstrable residuals of your immune deficiency disorder including any residual impairment(s) resulting from your treatment. The provision is based on 13.00L4 in our malignant neoplastic diseases listings.

- In final 14.00E4, *Medication-induced immune suppression*, we explain that medication can result in immune suppression that will usually resolve once the medication is ceased. However, if you take prescribed medications for long-term immune suppression, such as after an organ transplant, we will look at the frequency and severity of any infections you get, residuals from the organ transplant itself, and whether there has been any significant deterioration of other organ systems.

Final 14.00F—How do we document and evaluate human immunodeficiency virus (HIV) infection?

We changed the heading of proposed 14.00F in response to a public comment on the NPRM that we describe in the public comments section of this preamble. In final 14.00F, we incorporate, update, and expand information on HIV infection that was contained in prior 14.00D3 through 14.00D7. We also make nonsubstantive editorial changes.

As already noted, we moved the first sentence of prior 14.00D1 to final 14.00A4. Therefore, we begin final 14.00F with the second sentence of prior 14.00D1. It is a reminder that an individual’s HIV infection need not meet the Centers for Disease Control and Prevention (CDC) definition of acquired immune deficiency syndrome (AIDS) to meet or medically equal the criteria of listing 14.08. We made minor editorial changes to the sentence, but did not change its meaning.

We do not require an individual’s HIV infection to meet the CDC definition of AIDS because in evaluating disability claims, our concern is to determine whether an individual’s impairment(s) is severe enough to prevent him or her from engaging in any substantial gainful activity. The CDC’s definition is designed to enhance its capability for activities such as disease reporting and surveillance, epidemiologic studies, prevention and control activities, and public health policy and planning. This definition is not intended to determine whether any statutory or regulatory requirements for disability are met.

We moved the provisions of prior 14.00D2 to other sections in the final rules. In the first four paragraphs of prior 14.00D2, we defined the terms “resistant to treatment,” “recurrent,” and “disseminated,” and we now define those terms in final 14.00C. In the fifth paragraph of prior 14.00D2, we defined “significant involuntary weight loss” for purposes of prior listing 14.08I (final listing 14.08H). In the final rules, we include this definition in 14.00F5.

Like prior 14.00D3, final 14.00F1 is in two major sections: A section explaining how we document the diagnosis of HIV infection definitively (14.00F1a) and a section explaining how we document the diagnosis of HIV infection when we do not have definitive evidence (14.00F1b). In final 14.00F1, *Documentation of HIV infection*, we incorporate and update the information in prior 14.00D3 to explain the laboratory tests or other evidence we accept as documentation of HIV infection. In response to a public comment on the NPRM, we have also added a statement, similar to the statements we added in final 14.00D2b and 14.00D4b, explaining that we will not purchase laboratory testing to establish whether you have HIV infection.

Final 14.00F1a, *Definitive documentation of HIV infection*, corresponds to prior 14.00D3a. We updated and expanded this section to include newer laboratory diagnostic techniques that did not exist or were not widely used when we published the prior rules in 1993.

- Final 14.00F1a(i), for HIV antibody tests, corresponds to prior 14.00D3a(i). We made only nonsubstantive editorial changes.

- Final 14.00F1a(ii) is new from our prior rules. It adds positive “viral load” tests for HIV infection, such as quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT-PCR), that were not

widely available when we published the prior rules.

- Final 14.00F1a(iii) is for HIV DNA detection by polymerase chain reaction (PCR). We included it as an example of an “other test” in prior 14.00D3a(iii) because it was not widely available when we published the prior rules.

- Final 14.00F1a(iv), for HIV antigen, corresponds to prior 14.00D3a(ii).

- Final 14.00F1a(v) is new from our prior rules. It adds a positive viral culture for HIV from peripheral blood mononuclear cells (PBMC) as another test that definitively documents HIV infection. Even though it is not commonly used, we will accept it as definitive evidence if it is in your medical records.

- Final 14.00F1a(vi), for other tests that are highly specific for detection of HIV, corresponds to the first paragraph in prior 14.00D3a(iii).

Final 14.00F1b, *Other acceptable documentation of HIV infection*, corresponds to prior 14.00D3b. It explains what documentation of HIV infection we will accept instead of definitive laboratory testing. The final rule is essentially the same as the prior rule except for nonsubstantive editorial changes. However, in response to a public comment on the NPRM, we removed the word “*carinii*” and refer now only to “*Pneumocystis pneumonia*” (PCP) in this section and others in these final rules. We explain the reason for this change in the public comments section of this preamble.

In final 14.00F2, *CD4 tests*, we combine the provisions in the second undesignated paragraph after prior 14.00D3a(iii) and the second paragraph in prior 14.00D4a. We specify that, even though a reduced CD4 count or percent alone does not establish a definitive diagnosis of HIV infection, a count below 200/mm³ (or below 14 percent of the total lymphocyte count) along with clinical findings does offer supportive evidence of the existence of HIV infection without a definitive diagnosis. This is because a CD4 count below 200 is an indicator of an increased susceptibility to developing opportunistic infections.

In the final rules, we slightly revised the language we proposed to correct minor inconsistencies in the NPRM. In the fourth sentence of proposed 14.00F2, we referred to a CD4 count “below 200.” However, in the third sentence, we referred to a CD4 count that is “200 mm³ or less,” which is not precisely the same thing. In these final rules, we are correcting the third sentence to also say “below 200” for consistency. Likewise, we revised the parenthetical reference to “below 14

percent” and clarified that the reference is to the percentage of CD4 cells to the total lymphocyte count. We made the same changes throughout these final rules for consistency with these corrections. We also made nonsubstantive editorial changes in this paragraph.

In final 14.00F3, *Documentation of the manifestations of HIV infection*, we incorporate the information in prior 14.00D4 with nonsubstantive editorial changes. Like final 14.00F1 and prior 14.00D4, final 14.00F3 is divided into two main parts:

- Final 14.00F3a, *Definitive documentation of the manifestations of HIV infection*, incorporates the first paragraph in prior 14.00D4a and explains how we document manifestations of HIV infection definitively.

- Final 14.00F3b, *Other acceptable documentation of the manifestations of HIV infection*, incorporates information that was in the first paragraph of prior 14.00D4b and explains how we document manifestations of HIV infection when we do not have definitive evidence.

We are revising the language of proposed 14.00F3b to clarify our original intent. In the prior rule, we indicated that “if no definitive laboratory evidence is available, manifestations of HIV infection may be documented by medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence.” The sentence may have implied that we needed to have all of the things listed (medical history and clinical findings and laboratory findings and diagnosis(es)) to determine that you have a manifestation of HIV infection when we do not have definitive laboratory findings. That was not our intent, so we are clarifying in the final rule that we may need only some of this information to make a finding that you have a manifestation of HIV infection, depending on the prevailing state of medical knowledge and clinical practice. We are also clarifying what we mean by “laboratory findings” in this context; that is, laboratory findings that do not in themselves definitively establish the existence of an HIV-related manifestation. In response to a public comment on the NPRM, we are also clarifying in final 14.00F3b that the manifestations that are listed are only examples of manifestations that can be diagnosed without definitive evidence. We will accept a presumptive diagnosis of any manifestation of HIV infection so long as the method used to make the diagnosis is consistent with the

prevailing state of medical knowledge and clinical practice.

In 14.00D4 of the prior rules we provided specific guidance for documenting one particular manifestation of HIV infection without definitive evidence: *Cytomegalovirus* (CMV) disease. In final 14.00F3b, we expand the section to include three additional manifestations, including a manifestation we added in response to a public comment on the NPRM. The revised guidance is as follows:

- In final 14.00F3b(i), we explain that PCP is frequently diagnosed presumptively without definitive evidence and provide examples of evidence that is supportive of a presumptive diagnosis of PCP. Because we removed the word “*carinii*” in a change we made in final 14.00F1b, we no longer need the parenthetical note we proposed to include in 14.00F3b(i); therefore, we have not included it in these final rules. In response to a public comment on the NPRM, we also added “no evidence of bacterial pneumonia” to the list of evidence that is supportive of a presumptive diagnosis of PCP. For consistency with a change we made in final 14.00F3b(ii) in response to a public comment on the NPRM, we also indicate that supportive evidence of a presumptive diagnosis of PCP “may” include the items we list. This is not a change in the meaning of the proposed rule, only a clarification.

- In final 14.00F3b(ii), we incorporate and expand the information now in the second paragraph of prior 14.00D4b, regarding the documentation of CMV disease. However, in an editorial change from the NPRM, we revised the second and fourth sentences and removed the third sentence in proposed 14.00F3b(ii). In the NPRM, we stated that a serology test “identifies a history of infection with CMV, but it does not confirm an active disease process.” We revised this to state that a serology test “does not establish a definitive diagnosis of CMV disease, but it does offer supportive evidence of a presumptive diagnosis of CMV disease.” Due to this revision, we removed a positive CMV serology test from the list of examples of clinical findings that are supportive of a presumptive diagnosis of CMV that were in the fourth sentence of the proposed section, and revised the sentence to indicate that the examples provided are other clinical findings that support a presumptive diagnosis of CMV. We removed the third sentence because it was unnecessary. These changes are not substantive, only a clarification of the proposed rules. As in the NPRM, we do not include “documentation of CMV disease

requires confirmation by biopsy” as in the last sentence of the second paragraph of prior 14.00D4b because we are providing information on documentation other than definitive laboratory findings. Also, instead of stating that we can use generally acceptable methods to confirm the diagnosis of CMV, we provide examples of evidence, such as fever and a positive CMV serology test, that is supportive of a presumptive diagnosis of CMV disease. In response to a public comment on the NPRM, we are clarifying that an individual need not have all of the findings we list by indicating that supporting evidence “may” include these findings.

- In final 14.00F3b(iii), we explain how toxoplasmosis of the brain is presumptively diagnosed since the definitive method of diagnosing toxoplasmosis of the brain by biopsy is not commonly performed.

- In final 14.00F3b(iv) we provide guidance about how candidiasis of the esophagus may be presumptively diagnosed. We explain our reasons for making this addition and the other changes summarized above in the public comments section of this preamble.

We are also making a minor change from the NPRM in the opening paragraph of 14.00F3. The last sentence explained that we will make every reasonable effort to obtain reports of the results of laboratory testing you have had for a manifestation of HIV infection. We are not including that sentence in final 14.00F3 because it is repetitive of other provisions in these final rules and in our other regulations. See, for example, final 14.00B and current §§ 404.1512 and 416.912. Therefore, this revision is only editorial, simplifying the proposed rule without changing any requirements.

In final 14.00F4, *HIV infection manifestations specific to women*, we incorporate the information in prior 14.00D5. In final 14.00F4a, *General*, we incorporate the first paragraph of prior 14.00D5, while in final 14.00F4b, *Additional considerations for evaluating HIV infection in women*, we incorporate the second paragraph of prior 14.00D5. Except for adding paragraph designations and headings and minor editorial changes (including changes that are reflected in the paragraph designations of the listings explained below), the final provisions are the same as in the prior rules.

In final 14.00F5, *Involuntary weight loss*, we incorporate the last paragraph of prior 14.00D2 with nonsubstantive editorial changes, including a change that reflects the redesignation of prior

listing 14.08I as final listing 14.08H. In a change from the NPRM, we are not including the first sentence we had proposed, which was also in the prior rules. The sentence said, “[S]ignificant involuntary weight loss’ does not correspond to a specific minimum amount or percentage of weight loss.” The sentence could have been confusing because the very next sentence (what is now the first sentence in the final rule) explains that a 10 percent weight loss is always “significant”; therefore, in some cases “significant weight loss” does correspond to a specific percentage. It was also unnecessary because the next sentence (the second sentence in the final rule) explains that a weight loss of less than 10 percent may or may not be “significant,” which has essentially the same meaning as the sentence we removed.

Final 14.00G—How do we consider the effects of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

In final 14.00G, we explain how we consider the effects of treatment for all three categories of immune system disorders; that is, autoimmune disorders, immune deficiency disorders, and HIV infection. The new section addresses in one place issues of treatment that are common to all three types of immune system disorders as well as issues of treatment that are unique to each type of disorder, including treatment that is specifically for HIV infection. We did not remove any guidance about treatment for HIV infection that is still relevant, but instead we moved it to this new section. In fact, we expanded and updated our rules to reflect what has been learned in applying different treatments for HIV infection since we published the prior rules. The provisions for addressing both the positive effects and negative side effects of treatment in individuals who have autoimmune disorders and immune deficiency disorders, other than HIV infection, are new in these final listings and, we believe, provide useful adjudicative guidance that was lacking in the prior rules.

Final section 14.00G has six subsections. The first two (final 14.00G1 and 14.00G2) and the last one (final 14.00G6) are applicable to all immune system disorders. Final 14.00G3–14.00G5 provide guidance specific to each of the three main types of immune system disorders: Autoimmune disorders (final 14.00G3), immune deficiency disorders, excluding HIV infection (final 14.00G4), and HIV infection (final 14.00G5).

In final 14.00G1, *General*, we incorporate the first and fifth sentences of prior 14.00D7. We believe that this guidance has general applicability to all immune system disorders, not just HIV infection. We first explain that we consider the effectiveness of your treatment on your signs, symptoms, and laboratory findings, and the negative side effects of your treatment on your functioning. We also explain that we will make every reasonable effort to obtain a specific description of the treatment you receive. Then, we list eight factors we consider when we evaluate your treatment. They are mostly based on factors we mentioned in the prior rule, but we expanded the list, and in some cases clarified the factors that were in the prior rules. For example, instead of referring only to the “dosage [and] frequency of administration” of your treatment, we refer to “the intrusiveness and complexity of your treatment (for example, dosing schedule, need for injections).” In final 14.00G1e, we also introduce the term “variability of your response to treatment,” a concept we addressed for HIV infection in prior 14.00D7 but that we believe is of particular importance in considering the effects of treatment in all individuals with immune system disorders. We explain this concept in more detail in final 14.00G2.

Final 14.00G1f is new. It describes the interactive and cumulative effects of treatments for immune system disorders and other disorders that persons with immune system disorders may also have. We explain that the effects of these treatments taken together may be greater than they would be if we considered them separately, and we provide an example of treatment for HIV infection together with treatment for hepatitis C. Final 14.00G1g is also new. It explains that we will also consider the duration of your treatment. Final 14.00G1h is a catchall for other relevant factors we have not listed in 14.00G1a–14.00G1g.

In final 14.00G2, *Variability of your response to treatment*, we explain what we mean by this factor in terms of both HIV infection and other immune system disorders. The final rule is based on the language of the second paragraph in prior 14.00D7 and the second sentence of the third paragraph of that sentence. However, we are expanding that guidance and applying it to all other immune system disorders in addition to HIV infection. For example, we explain in a general way applicable to all immune system disorders that some individuals may show an initial positive response to drug treatment (or a

combination of drugs), but the initial positive response may be followed by a decrease in the effectiveness of the medication.

We provide more specific information about treatment of autoimmune disorders in final 14.00G3, *How we evaluate the effects of treatment for autoimmune disorders on your ability to function*. This final rule repeats the rule in the fifth paragraph of prior 14.00B that we consider the adverse effects that may result in loss of function when we evaluate the effects of your treatment for your autoimmune disorder(s). We expanded this guidance to include more examples of potential chronic adverse effects of steroid treatment and to explain that the side effects of some medications may be acute or long-term. We add a provision that recognizes that the medications used in the treatment of autoimmune disorders may have effects on mental function, including cognition (memory), concentration, and mood.

Final 14.00G4, *How we evaluate the effects of treatment for immune deficiency disorders, excluding HIV infection, on your ability to function*, is new. As in final 14.00G3, we repeat the principle that we will consider the side effects of your treatment when we evaluate your ability to function. We cite intravenous immunoglobulin and gamma interferon therapy as examples of treatment you may be receiving. We also provide examples of side effects of treatment for immune deficiency disorders, including physical symptoms (such as severe fatigue and headaches), clinical signs (such as high blood pressure and joint swelling), or limitations in mental function, including cognition, concentration, and mood.

Final 14.00G5, *How we evaluate the effects of treatment for HIV infection on your ability to function*, is in two parts. In final 14.00G5a, *General*, as in final 14.00G3 and 14.00G4, we repeat the principle from prior 14.00D7 that we consider the side effects of antiretroviral treatment and treatment for the manifestation of HIV infection on your ability to function. We expand the guidance to provide examples of the physical and mental side effects of antiretroviral drugs. We also note that the symptoms of HIV infection and the side effects of medications may be indistinguishable, and that we will consider your functional limitations whether they are a result of your symptoms or signs of HIV infection or the side effects of your treatment.

We made two changes in final 14.00G5a in response to a public comment on the NPRM. We added a parenthetical reference to “fat

redistribution, such as ‘buffalo hump.’” “Fat redistribution” is another name for lipodystrophy, which we had included in the proposed rule, and “buffalo hump” is a kind of lipodystrophy. We also expanded the last sentence of the paragraph to explain that we consider functional limitations from signs of HIV infection as well as from symptoms. We explain our reasons for these changes in the public comments section of this preamble.

In final 14.00G5b, *Structured treatment interruptions*, we provide new guidance specifically about structured treatment interruptions (STIs, also called drug holidays) in individuals with HIV infection. The guidance explains that STIs are part of a prescribed treatment plan; therefore, they do not show that an individual is failing to follow treatment or in themselves establish that an individual’s impairment is not as severe as alleged.

In final 14.00G6, *When there is no record of ongoing treatment*, we explain how we evaluate the medical severity and duration of your immune system disorder when you have not received ongoing treatment or have not had an ongoing relationship with any treatment source despite the existence of a severe impairment(s). The provision is based on a standard provision we include in most other body system listings; for example, 1.00H3 in the musculoskeletal system, the third paragraph of 3.00A in the respiratory system, and the third paragraph of 4.00B3 in the cardiovascular system. We also explain that if you have just begun treatment and we cannot decide whether you are disabled based on the evidence we have, we may need to wait to determine the effect of your treatment. We explain that there is no set period because how long we may need to wait will depend on the facts of your individual case. This is consistent with the guidance we provided in the last two sentences of the third paragraph in prior 14.00D7, which explained that decisions about the impact of treatment should be based on a sufficient period of treatment to permit proper consideration of the temporary or long-term effects of the treatment.

Final 14.00H—How do we consider your symptoms, including your pain, severe fatigue, and malaise?

Final 14.00H is new. In it, we explain that we will evaluate the impact your symptoms have on your ability to function when the evidence of your immune system disorder(s) shows that you have a medically determinable

impairment that could reasonably be expected to produce your symptoms.

We added a sentence in the final rule in response to a public comment we describe later in this preamble. The sentence explains that we will not draw any inferences about your symptoms and their functional effects from the fact that you do not receive treatment or 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. As we explain in more detail later, the sentence is based on a provision in Social Security Ruling (SSR) 96–7p. We also clarified the heading in the final rule by listing the two constitutional symptoms, severe fatigue and malaise, instead of referring to “constitutional symptoms.”

Final 14.00I—How do we use the functional criteria in these listings?

We indicated in the ANPRM that we would not summarize or respond to the public comments (68 FR 24897). However, there was one theme that was common to many of the letters and e-mails and that was raised repeatedly at our two outreach meetings by the medical specialists, advocates for persons who have immune system disorders, and individuals with immune system disorders: The functional impact of immune system disorders, and the inadequacy of the immune system rules to address that impact, especially for immune system disorders other than HIV infection. This issue was raised so often, and as a matter of such great public interest, that we believe that it will be helpful to summarize briefly what commenters said to help explain why we are adding new rules for evaluating functioning in these listings.

Many commenters said that we should recognize how immune system disorders can affect an individual’s functioning. Many individuals described physical symptoms, such as pain, fatigue, and malaise, as well as mental symptoms, including loss of memory, loss of concentration, and depression. Commenters stressed that these symptoms could be very severe. A number of persons indicated that the fatigue associated with these disorders was not merely a feeling of tiredness but a more profound and debilitating experience. Many individuals also noted that the impairments could be both episodic and variable in intensity, with some individuals experiencing “good” or relatively good days interspersed with days in which they were unable to function. They pointed out that there was a need for the rules

to recognize the longitudinal effect of these episodic limitations on the ability to work. Other persons pointed out that there is often comorbidity of immune system disorders, that is, many persons have features of more than one immune system disorder. In those cases, the combination of symptoms and limitations have a multiplication effect in the individual’s overall condition that is worse than simply adding the individual effects of the symptoms and limitations to each other. These commenters said that under the prior listings there is no adequate way to assess these multiplied effects. Many commenters also pointed out the effect that stress can have on the medical condition and symptomatology of individuals who have immune system disorders. Other individuals described the debilitating effects of treatment, not only the side effects, but sometimes the need to follow a very rigorous and time-consuming schedule of treatment that in itself can be limiting.

A number of the commenters pointed with approval to the provisions of prior listing 14.08N and the text in prior 14.00D8 that explains that listing. These individuals thought that the provisions should not be confined to persons who have HIV infection but should be extended to individuals with other kinds of immune system disorders who may be continuously limited by their symptoms and other manifestations, frequently become ill, have periodic manifestations, or have the kinds of serious limitations described in those rules. They urged us to consider extending such criteria to all listed immune system disorders to ensure that we do not overlook individuals who do not necessarily have the objective evidence needed to meet the other criteria in the listings but who may still be disabled.

As we have noted, in these final rules we are significantly expanding our guidance about specific immune system disorders and the effects of treatment. We also agree with those commenters on the ANPRM and at the public outreach meetings who suggested that we include the same kind of criteria for evaluating the overall functional impact of other immune system disorders as we provided in prior listing 14.08N for persons who have HIV infection. Therefore, we are adding criteria similar to those in prior listing 14.08N (final listing 14.08K) for each of the listed impairments in this body system. The final listings for evaluating functioning for other immune system disorders are 14.02B, 14.03B, 14.04D, 14.05E, 14.06B, 14.07C, 14.09D, and 14.10B. We are also redesignating prior listing 14.08N as

final 14.08K for reasons we explain below.

Final 14.00I is the section of the introductory text that explains the listings that include functional criteria. It corresponds to prior 14.00D8, but we revised it so that it applies to all of the new final listings that include functional criteria, not just the listing for HIV infection (prior listing 14.08N).

Like prior 14.00D8, final 14.00I includes eight paragraphs. Except as described below, we revised each paragraph so that it applies not only to HIV infection but to the other immune system disorders as well. For example, in the first paragraph of prior 14.00D8 we explained that prior listing 14.08N (final listing 14.08K) established standards for evaluating manifestations of HIV infection that do not meet the criteria of any of the preceding listings within 14.08; that is, prior listings 14.08A–14.08M. We also explained that we used prior listing 14.08N both for manifestations that were listed in the preceding listings within 14.08 and for manifestations that were not listed at all. We have modified this language so that it applies to all of the immune system disorders within this body system. We also made minor editorial changes throughout the paragraphs.

The following are other changes we are making in this section.

In final 14.00I2, we are removing the first sentence in the second paragraph of prior 14.00D8. That sentence explained that, for individuals with HIV infection, we assessed listing-level severity under prior listing 14.08N based on the functional limitations imposed by the impairment. We believe that this point is already made in final 14.00I1 and that it is unnecessary to repeat it in final 14.00I2. We are revising the second sentence, which said that we must consider the full impact of “signs, symptoms, and laboratory findings” on the individual’s ability to function. We believe that this guidance may not have clearly explained what we intended. Therefore, we are revising it to explain that when we use one of the listings cited in final 14.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder(s) on your ability to function on a sustained basis.

In final 14.00I3–14.00I8, which correspond to the last six paragraphs in prior 14.00D, we are updating our rules to make their language more consistent with our other rules that define the term “marked” and the areas of functioning. However, these changes are not intended to be substantively different from the prior rules. We are also

including references to both pain and severe fatigue throughout final 14.00I6–14.00I8 as symptoms that may cause limitations. The prior rules were not consistent in this regard.

We added guidance in final 14.00I3 in response to public comments on the NPRM. The guidance clarifies that your impairment will satisfy the criterion for “repeated” manifestations regardless of whether you have the same kind of manifestation repeatedly, all different manifestations, or a combination of some manifestations that are the same and some different; for example, two of the same kind of manifestation and one different one. You must only have the required number of manifestations with the frequency and duration required in this section. This is not a change in meaning from the proposed rules, but a clarification of our intent. In response to another comment, we also clarify that the manifestations must occur within the period covered by your claim.

Final 14.00J—How do we evaluate your immune system disorder when it does not meet one of these listings?

Final 14.00J1 and 14.00J3 replace the guidance we provided in the first and third paragraphs of prior 14.00D6. As in other provisions throughout the introductory text, we are revising the language to make it apply generally to all immune system disorders, not just HIV infection. Also, we are removing guidance that is already covered in other sections in the introductory text, such as the guidance that individuals may have signs or symptoms of a mental impairment or of another physical impairment.

Final 14.00J2 is a new section in this body system. For reasons we have already explained, we are removing reference listings—that is, listings that are met or equaled by meeting or equaling the criteria of another listing—from this body system. However, immune system disorders can have effects in virtually every body system, and we believe it is important to include guidance about those effects in the introductory text so that they are not overlooked.

Therefore, we are adding section 14.00J2 to explain that immune system disorders can have effects in other body systems; we also provide a list of examples of those effects in each of the relevant body systems with references to other body system listings. These provisions are based on language in the second paragraph of prior 14.00D6, which was relevant only to the evaluation of HIV infection, and on the reference listings we are removing. We are expanding the information that was

in that paragraph to provide specific examples of impairments that may be caused by autoimmune disorders.

For example, prior listings 14.02A6 and 14.04A4 were met with evidence of SLE, systemic sclerosis, or scleroderma with “Digestive involvement, as described under the criteria in 5.00ff.” Apart from the fact that these listings were unnecessary because any individual who meets the criteria of a listing in the digestive body system (5.00) would be disabled under that listing, the guidance was not very specific. Also, in the prior rules, we included these criteria only under prior listings 14.02 and 14.04. However, other immune system disorders can have effects in the digestive system. Therefore, in final 14.00J2e, we provide that any immune system disorder can have effects in the digestive system, and we include an example of hepatitis C in addition to providing a reference to 5.00.

In these final rules, we are adding a reference to weight loss as a result of HIV infection that affects the digestive system in final 14.00J2e. We explain later in this preamble that our reason for adding this reference is to respond to public comments we received on the NPRM about HIV wasting syndrome.

Final 14.00J2k provides examples of allergic disorders (including skin disorders) that individuals with immune system disorders may have. It replaces prior 14.00C.

How are we changing the criteria in the immune system disorders listings for adults?

14.01—Category of Impairments, Immune System Disorders

The following is a detailed explanation of the significant changes in the final listings. Some changes are common to several listings, so we describe them first.

1. We are removing all of the reference listings from this body system for reasons we have already explained.

2. We are revising prior listings 14.02B, 14.03B, 14.04B, and 14.09D (final listings 14.02A, 14.03A, 14.04A, and 14.09B) as follows:

- We are removing the criterion for “significant, documented” constitutional symptoms or signs in each of these listings because we define the constitutional symptoms and signs in final 14.00C2. Moreover, it is unnecessary to specify “documented” because we always need to document the existence of any symptom or sign in any disability claim.

- Each of these prior listings, except prior listing 14.09D, also required you to

have all four of the constitutional symptoms or signs: Severe fatigue, fever, malaise, and involuntary weight loss. We are revising this requirement to “at least two” of the constitutional symptoms or signs, instead of all four, because we believe that the requirement in the prior listings was too severe. We believe that any individual with an autoimmune disorder involving two or more organs/body systems with one organ/body system involved to at least a moderate level of severity and who has at least two of the constitutional symptoms and signs in these listings will have an impairment that precludes any gainful activity. We have also added “involuntary” as a descriptor of weight loss in final listings 14.02A, 14.03A, 14.04A, 14.05E, 14.06A, 14.07C, 14.08K, 14.09B, and 14.10A for reasons we explained earlier in this preamble.

- In final listings 14.02A, 14.03A, and 14.04A, which correspond to prior listings 14.02B, 14.03B, and 14.04B, we are removing the reference to “lesser involvement” because we are removing the prior reference listings to which these rules refer. We also believe the phrase is unnecessary—the severity of the impairment is demonstrated by the remaining criteria.

3. As we have already noted under the explanation of final 14.00I, we are adding listings based on repeated manifestations accompanied by functional limitations and modeled after prior listing 14.08N (final listing 14.08K) for each of the other immune system disorders. The final listings are:

- 14.02B for SLE,
 - 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,
 - 14.09D for inflammatory arthritis, and
 - 14.10B for Sjögren’s syndrome.
- Each listing requires you to have:
- The specified immune system disorder for that listing,
 - Repeated manifestations of the specified immune system disorder,
 - At least two of the constitutional symptoms or signs, and
 - A “marked” limitation in one of three domains of functioning: Activities of daily living, maintaining social functioning, or completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

We explain what we mean by “repeated” in final 14.00I3 and by “marked” in final 14.00I4–5.

In the final rules, we made a number of changes from the proposed rules in response to public comments on the NPRM. Chiefly, we removed from several listings the requirement that there must be manifestations “without the requisite findings in” a specified paragraph earlier in the listing; for example, proposed listing 14.02B said “without the requisite findings in [14.02]A.” Our only intent was to explain that we would use the listing criterion (for example, listing 14.02B) when you have an impairment that does not meet the requirements of the previously specified listing section (for example, listing 14.02A). However, a public comment pointed out that our language could have been confusing, and we determined that it was not necessary to have it at all. We explain in detail the public comment and our reasons for making this change throughout the final listings in the public comments section of this preamble.

The following is an explanation of the other significant changes we are making. We are also making minor editorial changes in some listings and changes to cross-references to the introductory text throughout the listings to reflect the changes to the introductory text for the final rules. We do not describe all of those changes below.

Final Listing 14.04—Systemic Sclerosis (Scleroderma)

Final listing 14.04B corresponds to prior listing 14.04C. As we have already noted, we are expanding this listing to include provisions for individuals who had a form of the disorder as children and who still have listing-level functional limitations as adults. The final listing is essentially identical to final listing 114.04, which we describe in detail later in this preamble, except that it includes references to appropriate adult rules defining “inability to ambulate effectively” and “inability to perform fine and gross movements effectively.”

We are also making minor clarifications in the language of the prior listing. Prior listing 14.04C described “[g]eneralized scleroderma with digital contractures.” We are clarifying that “digital” refers to either the toes or the fingers and are listing the effects in the toes separately from the effects in the fingers in final listings 14.04B1 and 14.04B2, respectively. We also are removing the requirement for “generalized” scleroderma (that is, systemic sclerosis) because the very serious digital contractures described in the final listings would in themselves be

disabling regardless of whether the scleroderma is generalized.

Final listing 14.04C corresponds to prior listing 14.04D. We are changing “Raynaud’s phenomena” in prior listing 14.04D to “Raynaud’s phenomenon” for the same reason already described in the explanation of final 14.00D3. We are removing the word “[s]evere” as a descriptor of Raynaud’s phenomenon in this listing because it is unnecessary given the severity of the impairment demonstrated by the remaining criteria, such as ischemia with ulcerations of toes or fingers, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively. As in final listing 14.04B, we also are clarifying that “digital” refers to fingers or toes.

In final listing 14.04C, we are also revising the criteria in prior listing 14.04D to provide a better description of listing-level Raynaud’s phenomenon. The criteria in prior listing 14.04D required severe Raynaud’s phenomenon characterized by digital ulcerations, ischemia, or gangrene. As we noted in the NPRM, we believe that this included some individuals who did not have impairments of listing-level severity.

Therefore, in final listing 14.04C1, we provide criteria for Raynaud’s phenomenon characterized by gangrene involving “at least two extremities” to establish an impairment that would preclude any gainful activity. The final rule is somewhat different from the proposed rule, which referred to fingers and toes. We clarified it in response to a public comment on the NPRM that we describe in the public comments section of this preamble. As in the NPRM, we do not require that the gangrene result in the inability to ambulate effectively or to perform fine and gross movements effectively because the presence of gangrene involving at least two extremities by itself demonstrates a very serious impairment.

In final listing 14.04C2, we provide criteria for ischemia with ulcerations of the toes or fingers that results in the inability to ambulate effectively or to perform fine and gross movements effectively; Raynaud’s phenomenon characterized only by ischemia with ulcerations does not, by itself, describe an impairment that would necessarily result in an extreme loss of function. Also, ulcerations are an outcome of ischemia, so we are revising the language of the prior rule so that ischemia and ulcerations are not listed as though they are separate entities.

Final Listing 14.05—Polymyositis and Dermatomyositis

Final listing 14.05A corresponds to prior listing 14.05A. We are replacing the word “severe” as a descriptor of proximal limb-girdle weakness with the more accurate “resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 14.00C6 and 14.00C7.” We are also changing “shoulder and/or pelvic” muscle weakness to “pelvic or shoulder” muscle weakness because either pelvic muscle weakness that results in the inability to ambulate effectively or shoulder muscle weakness that results in the inability to perform fine and gross movements effectively is sufficient in itself to show disability, and the “and” is unnecessary.

Final listing 14.05B corresponds to prior listing 14.05B1. We are removing a number of the requirements from the prior rule because we have determined that impaired swallowing with aspiration due to muscle weakness establishes a listing-level impairment. We are revising the requirement for “episodes of aspiration” to only “aspiration” because of the progressive nature of muscle weakness that results from polymyositis or dermatomyositis. Once an episode of aspiration is documented, further documentation of multiple episodes is unnecessary. In addition, we are replacing “cricopharyngeal weakness” with “muscle weakness” in final 14.05B because impaired swallowing and aspiration may result from muscles other than the cricopharyngeal muscles. Finally, we are revising the phrase “impaired swallowing with dysphagia” to “impaired swallowing (dysphagia)” because “dysphagia” means impaired swallowing.

Final listing 14.05C corresponds to prior listing 14.05B2, for individuals who have polymyositis or dermatomyositis with impaired respiration due to intercostal and diaphragmatic muscle weakness.

Final listing 14.05D, Diffuse calcinosis, is a new listing for adults that has the same criteria as final listing 14.05D for children, which we describe in detail later in this preamble. We are adding this listing for individuals who had a form of the disorder as children and who still have listing-level functional limitations as adults.

Final Listing 14.06—Undifferentiated and Mixed Connective Tissue Disease

We are changing the heading of prior 14.06 to update it and to more

accurately describe the disorders we evaluate under this listing.

Prior listing 14.06 was entirely a reference listing, requiring evaluation under prior listings 14.02A, 14.02B, or 14.04. We are changing it to a stand-alone listing. Final listing 14.06A contains the same criteria as final listings 14.02A, 14.03A, and 14.04A; that is, involvement of two or more body systems to at least a moderate level of severity and at least two of the constitutional symptoms or signs. Final listing 14.06B contains the same functional criteria for the evaluation of repeated manifestations of undifferentiated and mixed connective tissue disease as the other listings in this body system.

Final Listing 14.07—Immune Deficiency Disorders, Excluding HIV Infection

We are changing the heading of listing 14.07 to update its terminology and to more accurately describe the disorders we evaluate under this listing.

The prior listing was met with documented, recurrent severe infections occurring three or more times within a 5-month period. We are replacing this criterion with a more accurate and up-to-date listing. The listing is in three parts.

Final listing 14.07A is essentially the same as final listing 14.08J (prior listing 14.08M), which describes individuals with HIV infection whose immune systems are so compromised that they frequently become ill. We believe that these criteria for individuals with HIV infection are equally as applicable to individuals with other kinds of immune deficiency disorders, and that they are more inclusive than the criteria in prior listing 14.07.

As in final listing 14.08J, final listing 14.07A provides that the infections must occur three times in a 12-month period, not three times in only a 5-month period. It also more precisely explains how severe the infections need to be by requiring either resistance to treatment or a need for hospitalization or intravenous treatment. It also specifies six types of infections.

Final listing 14.07B is new. We are adding this listing to recognize that some immune system disorders are treated by stem cell transplantation. In final listing 14.07B, we state that we will consider you to be under a disability until at least 12 months from the date of transplantation and, thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

Final listing 14.07C incorporates the same functional criteria for the evaluation of repeated manifestations of

immune deficiency disorders (excluding HIV infection) as in the other final listings in this body system and for the same reasons as described above.

Final Listing 14.08—Human Immunodeficiency Virus (HIV) Infection

Except as described below, we are not making any changes to the criteria in listing 14.08. As noted in the NPRM, we carefully considered the advances in treatment and consequent increases in longevity that have occurred since we published the prior rules in 1993. Based on this review, we did not believe that there had been sufficient progress in the treatment and control of HIV infection to warrant any change in these rules at that time. However, as a result of public comments we received on the NPRM, we now believe that some changes may be appropriate. Therefore, while final listing 14.08 is substantively the same as proposed listing 14.08, we are publishing separately an ANPRM in today’s edition of the **Federal Register** inviting comments and suggestions on how to update and revise our listing for HIV infection. We will consider the comments and suggestions that we receive in response to the ANPRM, as well as our adjudicative experience and additional information about advances in medical knowledge, treatment, and methods of evaluating HIV infection. If we determine that listing 14.08 should be revised, we will publish for public comment an NPRM that will propose specific revisions to the listing.

As already noted, we are removing reference listings throughout this body system, including the reference listings in listing 14.08. This results in the removal of several specific listings within 14.08 and the redesignation of some of the prior listings; for example, prior listing 14.08N has become final listing 14.08K. Where we are removing a reference listing, however, we have ensured that we provide guidance in the introductory text about where to evaluate the impairment. For example, prior listing 14.08A4, for HIV infection with syphilis or neurosyphilis, was a reference listing that said only to consider the impairment under the criteria for the affected body system, such as 2.00 (special senses and speech), 4.00 (cardiovascular system), or 11.00 (neurological). Although we are removing this reference listing, we include this same guidance in final 14.00J21.

We are also clarifying some of the rules. In final listing 14.08B2, we are reorganizing the language from prior listing 14.08B2 to make it clearer that we evaluate under this listing candidiasis involving the esophagus,

trachea, bronchi, or lungs, or at another site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes. We are moving prior listing 14.08C2, for PCP, from the listing for protozoan and helminthic infections to the listing for fungal infections because the organism that causes PCP is now known to be a fungus. We redesignate it as final listing 14.08B7.

We are redesignating prior listing 14.08N as final listing 14.08K. We are expanding our guidance on manifestations we evaluate under final listing 14.08K by adding “pancreatitis, hepatitis, peripheral neuropathy, glucose intolerance, muscle weakness, cognitive or other mental limitation” as new examples. We are also expanding our list of signs and symptoms by adding “nausea, vomiting, headaches, or insomnia.”

We made minor changes to the language of the functional criteria in final listing 14.08K from the language in prior listing 14.08N. For example, we replaced the words “restriction” in prior listing 14.08N1 and “difficulties” in prior listings 14.08N2 and 14.08N3 with the word “limitation” in final listings 14.08K1, 14.08K2, and 14.08K3. We made this change because “limitation” is a more accurate description for the functional criteria in these listings.

We are making a number of changes from the proposed rule in response to public comments on the NPRM and for editorial reasons. The changes are in:

- Final listing 14.08B2, in which we made a minor editorial correction to remove a redundant word;
- Final listing 14.08B7, in which we removed the word “*carinii*” and the parenthetical “*jiroveci*” from the name of “*Pneumocystis pneumonia*” in response to a public comment on the NPRM;
- Final listing 14.08E4, in which we revised the criterion from “squamous cell carcinoma of the anus” to “squamous cell carcinoma of the anal canal or anal margin” in response to a public comment on the NPRM;¹
- Final listing 14.08H, in which we clarified that the 10 percent loss of weight from baseline may be calculated in pounds, kilograms, or by body mass index (BMI) in response to a public comment on the NPRM;
- Final listing 14.08J, in which we removed an unnecessary comma; and
- Final listing 14.08K, in which we changed the reference to “fatigue” to “severe fatigue” and a reference to a

“mental impairment” to a “mental limitation” in response to public comments on the NPRM, and removed the proposed cross-reference to 14.00I5. The removal of the cross-reference is only editorial. The reference was unnecessary, incomplete (the term “marked” for the various domains is also defined in final 14.00I6, 14.00I7, and 14.00I8), and inconsistent with other sections of the proposed immune disorder listings which contained the same severity criteria but did not include this cross-reference.

We provide detailed explanations of the changes we made in response to public comments on the NPRM and our reasons for making them in the public comments section of this preamble.

Final Listing 14.09—Inflammatory Arthritis

We are redesignating prior listing 14.09D as final listing 14.09B, prior listing 14.09B as final listing 14.09C1, and prior listing 14.09E as final listing 14.09C2 to put these listings in a more logical order. In the final rules, listing 14.09A describes persistent inflammation or deformity of major peripheral joints that alone is disabling, while listing 14.09B describes disability with lesser inflammation or deformity of major peripheral joints together with organ involvement and constitutional symptoms or signs. Final listing 14.09C describes listing-level inflammatory arthritis of the spine. Final listing 14.09C1 describes disability based only on fixation (ankylosis) of the spine, while final listing 14.09C2 describes disability based on a lesser degree of ankylosis of the spine with organ involvement. Final listing 14.09D is the same functional listing we include in all of the final immune system disorders listings and applies to inflammatory arthritis affecting any joints.

Final listing 14.09A corresponds to prior listing 14.09A. We are removing the requirement for a history of joint pain, swelling, and tenderness from this listing because it is unnecessary. (We do refer to joint pain, swelling, and tenderness in final 14.00D6a as possible signs and symptoms of the disorder.) Persistent joint inflammation or deformity in one or more major peripheral weight-bearing joints resulting in the inability to ambulate effectively, or persistent joint inflammation or deformity of major peripheral joints in both upper extremities resulting in inability to perform fine and gross movements effectively, is in itself indicative of an impairment that would preclude any gainful activity. For the same reasons, we are also removing the requirement

for “signs on current physical examination.” We do not need signs of joint inflammation on a current physical examination when we have medical evidence documenting that you have inflammatory arthritis that results in the inability to ambulate effectively or inability to perform fine and gross movements effectively. Also, because of the episodic nature of inflammatory arthritis, a current physical examination could show a brief period of improvement for a few days even though your longitudinal medical records may show persistent joint inflammation that results in the inability to ambulate effectively or inability to perform fine and gross movements effectively.

As we noted under the explanation of final 14.00D6e, we are revising listing 14.09A in response to a public comment on the NPRM so that there is no longer a need to use listing 1.02 or 1.03 in cases involving inflammatory arthritis. Final listing 14.09 (and final listing 114.09) will apply to all individuals who have listing-level limitations as a result of inflammatory arthritis. The revised listing includes essentially the same requirements as listings 1.02 and 1.03 of the musculoskeletal listings.

Because of this, we are changing the structure of final listing 14.09A to provide separate criteria for inflammatory arthritis that involves one or more major peripheral weight-bearing joints (final listing 14.09A1) and inflammatory arthritis involving one or more major peripheral joints in both upper extremities (final listing 14.09A2), with appropriate severity criteria for each. We define the “major peripheral joints” in final 14.00C8.

Final listing 14.09B corresponds to prior listing 14.09D. The revisions in final 14.09B are similar to those in final listing 14.09A for the same reasons and to make it clearer that this listing requires joint inflammation in one or more major peripheral joints. Final 14.09B continues to require less joint involvement than in A, but we no longer require “lesser extra-articular features than in C” because “C” refers to prior reference listing 14.09C, which we have removed. Final listing 14.09B1 corresponds to prior listing 14.09D2 with nonsubstantive editorial changes to make it consistent with how we present this criterion throughout these listings. Final listing 14.09B2 corresponds to prior listing 14.09D1 except that we have removed the phrase “significant, documented” for reasons we have already explained. We are also correcting an error in prior listing 14.09D1. The explanatory abbreviation, “e.g.” (for example) in prior listing

¹ We also made minor conforming changes in prior 13.00A and 113.00A of the malignant neoplastic diseases listings to reflect this change.

14.09D1 inaccurately indicated that the four constitutional symptoms or signs, that is, severe fatigue, fever, malaise, and involuntary weight loss, were only examples when they are in fact a complete list. Consistent with changes in other final listings, we are requiring at least two of the constitutional symptoms or signs because we believe that the criteria in final listing 14.09B are indicative of an impairment that precludes any gainful activity.

Final listing 14.09C1 corresponds to prior listing 14.09B. We are reorganizing the criteria and removing the requirements for “diagnosis established by findings of unilateral or bilateral sacroiliitis (e.g., erosions or fusions)” and “[h]istory of back pain, tenderness, and stiffness” because these findings are unnecessary. We believe ankylosing spondylitis or other spondyloarthropathies with ankylosis of the dorsolumbar or cervical spines at 45° or more of flexion documented as required in final listing 14.09C1 are in themselves indicative of an impairment that precludes any gainful activity.

Final listing 14.09C2 corresponds to prior listing 14.09E. We are reorganizing this listing to make it more consistent with the structure and criteria that we use in the final listings for other autoimmune disorders. We are removing the phrase “with lesser deformity than in B,” which describes a deformity that is less than the fixation “of the dorsolumbar or cervical spine at 45° or more of flexion” under prior listing 14.09B, and replacing it with fixation “at 30° or more of flexion (but less than 45°).” We believe that this is a clearer and more specific criterion that helps to provide greater uniformity in adjudications under this listing. We are removing the phrase “lesser extra-articular features than in C” because it refers to prior reference listing 14.09C, which we are removing. We also are removing the phrase “with signs of unilateral or bilateral sacroiliitis” because the criteria in the final listing would be sufficient to show listing-level severity without this requirement, and the phrase “with the extra-articular features described in 14.09D” because it is unnecessary.

Final Listing 14.10—Sjögren’s Syndrome

Final listing 14.10 is new. We are adding it in response to comments we received before we developed the NPRM indicating that Sjögren’s syndrome is distinct from other immune system disorders and that it has unique aspects that the prior immune system listings did not address.

Although individuals with Sjögren’s syndrome were able to qualify under

prior listings 14.03 and 14.09 and other listings, we believe that it is now appropriate to list Sjögren’s syndrome separately in these listings. We are using the same two listing criteria for establishing listing-level severity as in the other final listings for autoimmune disorders because Sjögren’s syndrome is an autoimmune disorder that can cause the same kinds of constitutional symptoms and signs as other autoimmune disorders, and because it can be as functionally limiting as other autoimmune disorders. Final listing 14.10A is the same as final listings 14.02A, 14.03A, 14.04A, and 14.06A, and final listing 14.10B is the same as final listings 14.02B, 14.03B, 14.04D, 14.05E, 14.06B, and 14.09D. As already noted, we also provide a new separate section in the introductory text that describes the unique features of Sjögren’s syndrome, final 14.00D7.

How are we changing the introductory text for the immune system disorders listings for children?

As in final 14.00 in the adult rules, we are changing the name of this body system to “Immune System Disorders.”

Except for minor editorial changes, we have repeated much of the introductory text of final 14.00 in the introductory text of final 114.00. This is because the same basic rules for establishing and evaluating the existence and severity of immune system disorders in adults also apply to children. Because we have already described these provisions under the explanation of final 14.00, the following discussions describe only those provisions that are unique to the childhood rules or that require further explanation. We describe only the major provisions. For example, we do not summarize minor editorial changes that refer to “children” instead of adults or to the policy of “functional equivalence” instead of RFC assessment and steps in the adult sequential evaluation process.

Also, where appropriate in the introductory text of final 114.00, we have made an editorial change from the prior rules in the terms we use to identify the age categories of children in the introductory text of prior 114.00 to be consistent with the terms we use in the introductory text of current 112.00, Mental disorders. For example, in final 114.00F1b(ii), we use “newborn and younger infants (birth to attainment of age 1)” instead of “an infant 12 months of age or less” as in prior 114.00D3b(i).

Finally, we have changed the part B final rules from the NPRM in the same way that we changed the part A final

rules from the NPRM whenever those proposed rules were the same.

Final 114.00A—What disorders do we evaluate under the immune system disorders listings?

In final 114.00A1b, we incorporate the first sentence in the last paragraph of prior 114.00B, which explains that immune system disorders may affect growth, development, attainment of age-appropriate skills, and performance of age-appropriate activities in children. We are revising the sentence by adding the phrase “or their treatment.” We are also removing the phrase “attainment of age-appropriate skills” because it is redundant of “development.”

Final 114.00A2 is essentially the same as final 14.00A2 and similar to the first and second paragraphs of prior 114.00B. We are expanding and clarifying the guidance in the second paragraph to explain that autoimmune disorders or their treatment may have a considerable impact on the physical, psychological, and developmental growth of pre-pubertal children that often differs from that of post-pubertal children or adults. We are also removing the last sentences from both the first and second paragraphs of prior 114.00B because they cross-referred to 14.00 in the part A listings. In part B of these final rules, we are repeating criteria from part A when they are appropriate for evaluating children so it should rarely be necessary to refer back to 14.00 in part A.

Final 114.00D—How do we document and evaluate the listed autoimmune disorders?

Final 114.00D parallels the structure and content of final 14.00D in the adult rules, except where the features commonly associated with the autoimmune disorders in these listings differ in children from adults.

In final 114.00D2, *Systemic vasculitis (114.03)*, as in prior 114.00C3, we provide guidance (in final 114.00D2a(ii)) on how we evaluate Kawasaki disease and add guidance about anaphylactoid purpura (Henoch-Schoenlein purpura). Also, in final 114.00D2a(ii), we do not use the example of giant cell arteritis (temporal arteritis) that is in final 14.00D2a(ii) because this disorder occurs almost exclusively in individuals over 50 years of age.

In final 114.00D3c, *Localized scleroderma (linear scleroderma or morphea)*, we describe features of focal forms of scleroderma in children. These disorders occur primarily in children and are more common than systemic sclerosis in children. In final

114.00D3c(i), we explain that the extent of involvement and the location of the lesions are important factors in determining the limitations resulting from scleroderma. We also note that it may be appropriate to evaluate the limitations resulting from these impairments under the musculoskeletal listings (101.00).

In final 114.00D3c(ii), we describe features of isolated morphea of the face and explain that it may be more appropriate to evaluate the limitations from these disorders under the affected body system, such as special senses and speech (102.00) or mental disorders (112.00). We have made a minor correction in the final rule. In the NPRM, we indicated that it would be more appropriate to evaluate the limitations from these disorders only under the special senses or mental disorders listings. However, we explained in the preamble that these body systems were only examples of body systems that might be affected. In the final rule, we are clarifying that the body systems we cite are only examples. We have made the same correction in part A.

In final 114.00D3c(iii), we describe musculoskeletal and respiratory features of chronic variants of these syndromes and explain that it is appropriate to evaluate the limitations from these disorders under the musculoskeletal listings (101.00) or respiratory system listings (103.00).

In final 114.00D4, *Polymyositis and dermatomyositis (114.05)*, we note (in final 114.00D4a, *General*) that polymyositis occurs rarely in children and describe the features of dermatomyositis that occur differently in children than in adults.

In children, polymyositis and dermatomyositis usually do not occur in association with malignancies. For this reason, we do not include a reference to malignancy or provide guidance that we will evaluate malignancies under the malignant neoplastic diseases listings (113.00) in final 114.00D4, as we do for adults in final 14.00D4. However, unlike in the adult rules, we include a reference to calcinosis for children in this section. Calcinosis is primarily an outcome of juvenile dermatomyositis; when adults with dermatomyositis have calcinosis, it is generally because they have had the condition since childhood. For this reason, we refer to calcinosis only in the introductory text for children, final 114.00D4. However, we include a criterion for diffuse calcinosis in final listing 14.05D (as well as final listing 114.05D) for adults who have the condition. Also, when dermatomyositis involves other organs or body systems,

we evaluate the involvement under the affected body system.

In final 114.00D4b, *Documentation of polymyositis or dermatomyositis*, we note that magnetic resonance imaging (MRI) showing muscle inflammation or vasculitis provides additional evidence of childhood dermatomyositis. We did not provide this guidance in final 14.00D4b because MRI findings are not considered diagnostic of dermatomyositis in adults. Similar to final 14.00D4b, we added two sentences to the final rule to indicate that when the results of electromyography, muscle biopsy, or MRI are in your medical records we will make every reasonable effort to obtain them, but that we will not purchase any of these tests.

In final 114.00D4c(i), we explain how to evaluate polymyositis and dermatomyositis under the listings in newborn and younger infants.

In final 114.00D5, *Undifferentiated and mixed connective tissue disease (114.06)*, we note (in final 114.00D5a, *General*) that the most common pattern of undifferentiated autoimmune disorders in children is mixed connective tissue disease (MCTD). In final 114.00D5b, *Documentation of undifferentiated and mixed connective tissue disease*, we note diagnostic laboratory findings specifically for children with MCTD and that the clinical findings are often suggestive of SLE or childhood dermatomyositis. We also note that many children later develop features of scleroderma.

In final 114.00D6, *Inflammatory arthritis (114.09)*, we incorporate (in final 114.00D6a, *General*) guidance from prior 114.00C2 and 114.00E. We explain that we evaluate growth impairment resulting from inflammatory arthritis under the criteria in 100.00. In final 114.00D6b, *Inflammatory arthritis involving the axial spine (spondyloarthropathy)*, we incorporate the second sentence in prior 114.00E and revise some of the examples of disorders that may be associated with inflammatory spondyloarthropathies involving the axial spine with disorders that are more common in children.

Prior 114.00E6 provided that the fact that a child is dependent on steroids, or any other drug, for the control of inflammatory arthritis is, in and of itself, insufficient to find disability. It explained that advances in the treatment of inflammatory connective tissue disease and in the administration of steroids for its treatment have corrected some of the previously disabling consequences of continuous steroid use. Although this statement is still true, we are not including this provision of prior 114.00E6 in these

final rules because we believe we no longer need it in the introductory text for the listings.

We added prior 114.00E6 in 2002 (66 FR at 58022 and 58045). It was important when we added it because the listings prior to the revisions we made in 2002 included a listing (prior listing 101.02B) that said that all children with rheumatoid arthritis who were dependent on steroids were disabled. We removed that listing in 2002, explaining that, although the prior listing was appropriate when we first published it, advances in treatment and other reasons had made it obsolete (66 FR at 58022). Thus, the paragraph in the introductory text served as a reminder that we no longer had that listing and that it was no longer appropriate to presume disability based on steroid use alone. Now that several years have passed since we removed the prior listing, we do not believe that we need this reminder any longer. However, in final 114.00G3, we continue to state that we will consider the adverse side effects of treatment, including the effects of corticosteroids, to ensure that our adjudicators remember to consider the side effects of steroids and any other treatment an individual might have.

Final 114.00F—How do we document and evaluate human immunodeficiency virus (HIV) infection?

Final 114.00F parallels the structure and content of final 14.00F in the adult rules, except where the features commonly associated with HIV infection differ in children from adults.

Final 114.00F1a, *Definitive documentation of HIV infection*, corresponds to 114.00D3a in the prior rules and 14.00F1a in the final rules. In final 114.00F1a(i), we are lowering the age for using HIV antibody tests from the 24 months of age or older that was in prior 114.00D3a(i) to 18 months or older. Current clinical practice now accepts these tests beginning at 18 months of age.

In final 114.00F1a(iv), we clarify the provision in prior 114.00D3a(ii) by explaining that a specimen that contains HIV antigen may be used to establish the diagnosis of HIV infection in a child age 1 month or older.

Final 114.00F1b, *Definitive documentation of HIV infection in children from birth to the attainment of 18 months*, corresponds to the second paragraph in prior 114.00D3b, *Other acceptable documentation of HIV infection in children*. We are moving this information and revising the age cutoff to 18 months to recognize that laboratory values we previously considered to be “other acceptable

documentation” of HIV infection are now considered definitively diagnostic in children from birth to age 18 months who have tested positive for HIV antibodies.

In final 114.00F1b(i), we add “One or more of the tests listed in F1a(ii)–F1a(vii)” of final 114.00F1a because these tests are accepted as diagnostic of HIV infection.

In final 114.00F1b(iii), we change “12 to 24 months of age” in current 114.00D3b(ii) to “12 to 18 months of age” based on how these findings are used in current clinical practice.

In final 114.00F1b(v), we specify that a severely diminished immunoglobulin G (IgG) level is “< 4g/l or 400 mg/dl.” However, we do not provide an IgG level for greater than normal range for age due to the variability in the higher normal range of IgG level in children by age. There is consistency in the normal lower average range in children, so we are able to specify levels for severely diminished IgG.

Final 114.00F1c, *Other acceptable documentation of HIV infection*, corresponds to prior 114.00D3b and final 14.00F1b. We are removing the first paragraph in prior 114.00D3b, which explained that HIV infection is not documented in children under 24 months of age by a serum specimen containing HIV antibodies. All infants who have HIV antibodies are now tested to determine definitively whether they have HIV infection.

In final 114.00F2, *CD4 tests*, we add more detailed guidance to the second paragraph of prior 114.00D4a by specifying that the extent of immune depression correlates with the level of CD4 counts (relative to the age of the child), and that by age 6, CD4 levels become comparable to adult CD4 levels.

In final 114.00F3b, *Other acceptable documentation of the manifestations of HIV infection*, we explain, in 114.00F3b(i) for PCP and in 114.00F3b(ii) for CMV disease, that a CD4 count below 200 in children 6 years of age or older is supportive evidence of a presumptive diagnosis of these manifestations.

Final 114.00F4, *HIV manifestations specific to children*, corresponds to prior 114.00D5, *HIV in children*. In final 114.00F4a, *General*, we are removing the second sentence in prior 114.00D5. That sentence explained that survival times were shorter for children who were infected in the first year of life than they were for older children and adults. However, due to advances in medical treatment this is no longer the case. The second sentence of final 114.00F4a is based on the first paragraph in prior 114.00D5.

In final 114.00F4b, *Neurologic abnormalities*, we make some nonsubstantive editorial changes to the second paragraph in prior 114.00D5 in which we explained that the methods of identifying and evaluating neurological abnormalities vary depending on a child’s age. We also replace “acquisition” with “onset” in the last sentence of final 114.00F4b because a sudden “onset” of a new learning disability is medically a more accurate description of how this neurologic abnormality would manifest in a child with HIV infection.

In final 114.00F4c, *Bacterial infections*, we incorporate the last two paragraphs in prior 114.00D5. We make only nonsubstantive editorial changes, including removing text that only repeats criteria from the listings.

Final 114.00G—How do we consider the effects of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

In final 114.00G2, *Variability of your response to treatment*, we use an example of a child who develops otitis media instead of pneumonia or tuberculosis as we do in final 14.00G2 for an adult because otitis media is more common in children.

In final 114.00G3, *How we evaluate the effects of treatment for autoimmune disorders on your ability to function*, we use examples of impaired growth and osteopenia instead of osteoporosis as we do in final 14.00G3 because impaired growth and osteopenia are more common in children.

Final 114.00I—How do we use the functional criteria in these listings?

As in the adult rules, we are adding listings based on functional criteria to each of the listings in the immune system in addition to those that are already in listing 114.08. Final 114.00I—*How do we use the functional criteria in these listings?*—corresponds to prior 114.00D8 and provides guidance for applying the listings based on functional criteria in all of the final childhood listings. We revised the prior language to reflect the fact that there are now functional listings for each of the listed impairments in this body system and for consistency with adult rules where appropriate.

Final 114.00J—How do we evaluate your immune system disorder when it does not meet one of these listings?

In final 114.00J2, we repeat the guidance in final 14.00J but with appropriate references to childhood listings in part B, including an example of growth impairment under 100.00.

How are we changing the criteria in the immune system disorders listings for children?

Final 114.01—Category of Impairments, Immune System Disorders

As in the adult listings in part A, we are removing all reference listings from part B. We also add listings like final listing 114.08L (prior listing 114.08O) for each of the other listed impairments in this body system. (As in the NPRM, we are redesignating prior listing 114.08O as final listing 114.08L because of the deletion of reference listings.) The new listings are final listings 114.02B, 114.03B, 114.04D, 114.05E, 114.06B, 114.07C, 114.09D, and 114.10B. The functional criteria in the final listings for children are the same as in prior listing 114.08O, using the functional criteria in listings 112.02 and 112.12. They are different from the functional criteria in part A because the childhood functional criteria vary depending on the age of the child and are a better way to measure broad functional limitations in children.

The following is a description of the significant changes in part B when they are different from the changes we made in part A or require additional explanation.

Final Listing 114.04—Systemic Sclerosis (Scleroderma)

Final listings 114.04B1 and 114.04B2 correspond to prior listing 114.04B1. We are changing the requirement in prior listing 114.04B1 for fixed deformity of “both feet” to “one or both feet” and adding “inability to ambulate effectively” to the listing criteria. This will allow some children with a serious deformity in only one foot to qualify based on the functional limitation we use to define listing-level severity throughout these listings. We are also adding a criterion for “toe contractures” to final 114.04B1, even though toe contractures of listing-level severity would be rare in children, to make it consistent with the criterion in final 14.04B1. We are retaining the requirement for involvement of both hands in final listing 114.04B2, because inability to perform fine and gross movements effectively can occur only when both upper extremities are affected. We are adding the criterion of “finger contractures” to final 114.04B2 for the same reason we are adding “toe contractures” to final 114.04B1.

Final listings 114.04B3 and 114.04B4 correspond to prior listing 114.04B2, the listing for “[m]arked destruction or marked atrophy of an extremity.” We are revising the prior rules to:

- Remove the word “marked.”

- Change the criterion for “destruction” to “irreversible damage,”
- Require both atrophy and irreversible damage in one or both lower extremities or both upper extremities, and
- Require either inability to ambulate effectively or to perform fine and gross movements effectively.

We are removing the word “marked” because we use it in various other listings and other regulations to describe a particular measure of functional limitations, and it does not describe what we intend in this listing. We are replacing the criterion for “marked destruction” with a criterion for “irreversible damage” because it is a more accurate medical description of this complication of systemic sclerosis. We are requiring both atrophy and irreversible damage because we would not expect either of these findings alone to establish an impairment that results in marked and severe functional limitations in every case. Finally, we are requiring “inability to ambulate effectively” or “inability to perform fine or gross movements effectively” to establish an impairment that is of listing-level severity, consistent with other listings.

Final listing 114.04C, Raynaud’s phenomenon, is a new childhood listing and has the same criteria as in final listing 14.04C for adults.

Final Listing 114.05—Polymyositis and Dermatomyositis

We are removing prior listing 114.05B1 because multiple joint contractures are not typically a part of the disease process of polymyositis or dermatomyositis in children. However, if this should occur, we would evaluate whether your polymyositis or dermatomyositis with multiple joint contractures meets or medically equals the criteria in final listing 114.05E, medically equals the criteria in another listing, such as final listing 114.05A, or functionally equals the listings.

In final listing 114.05D, we are revising prior listing 114.05B2 by replacing “cutaneous calcification” with “calcinosis.” We are making this change because “calcification” describes the normal process by which calcium salts are deposited in bone, and “calcinosis” describes the abnormal deposits of calcium salt in body tissues as we intend by this criterion. We are also replacing “formation of an exoskeleton” with “limitation of joint mobility or intestinal motility” because it is a better description of the known complications of dermatomyositis in children.

Final Listing 114.07—Immune Deficiency Disorders, Excluding HIV Infection

We are removing prior listing 114.07B because of advances in medical knowledge that now allow the identification of different subgroups of thymic dysplastic syndromes. The subgroups of these disorders vary in severity, and therefore, we will evaluate them under final listing 114.07A, B, or C, as appropriate to the particular immune deficiency disorder and its effects.

Final Listing 114.08—Human Immunodeficiency Virus (HIV) Infection

In final listing 114.08A4, we have added a reference to final 114.00F4c in response to a public comment on the NPRM about children who are age 13 or older, whose impairments cannot meet but can medically equal this listing. In final listing 114.08A5, we incorporate prior listing 114.08A6 except to remove “Other” as a descriptor to make it consistent with the final adult listing. We replace “acquisition” as used in prior listing 114.08H1 with “onset” in final listing 114.08G1 because a sudden “onset” of a new learning disability is medically a more accurate description of how this neurologic abnormality would manifest in a child with HIV infection. We are also redesignating a number of listings to reflect the removal of reference listings.

Final Listing 114.10— Sjögren’s Syndrome

We are adding listing 114.10 to evaluate Sjögren’s syndrome in children for the same reasons we are adding a Sjögren’s syndrome listing for adults in part A.

Other Changes

We are making minor conforming changes in prior 1.00B and 101.00B, and 1.00L and 101.00L to reflect changes in the final immune body system listings.

We are also making minor conforming changes in prior 8.00D3 and 108.00D3 of the skin disorders listings. We are revising these sections to indicate that we evaluate Sjögren’s syndrome under the new listing for that disorder, final listings 14.10 and 114.10.

We are also making minor conforming changes in prior 13.00A and 113.00A of the malignant neoplastic diseases listings. We are revising these sections to reflect changes in final listings 14.08E and 114.08E.

Throughout these final rules, we are also making a number of minor editorial changes from the NPRM that we have not summarized above. For example, we have corrected unintentional language

inconsistencies between part A and part B, changed sentences to use active voice instead of passive voice, and removed some repetitive statements and unnecessary words. None of these revisions are substantive, and they do not change the meaning of what we originally proposed in the NPRM.

Public Comments on the NPRM

In the NPRM, we published in the **Federal Register** on August 04, 2006 (71 FR 44432, corrected at 71 FR 46983), we provided the public with a 60-day comment period that ended on October 13, 2006. In addition to our notice to the public, we invited comments from national medical organizations and professionals, advocacy groups, and legal services organizations.

We received 55 comment letters. The commenters included advocacy groups, legal services organizations, State agencies that make disability determinations for us, medical organizations, and individuals, including individuals who have immune system disorders or relatives with immune system disorders. One of the comment letters reflected the comments from 40 organizations. We carefully considered all of the comments and provide our reasons for adopting or not adopting the comments in our responses below. Because some of the comments were long, we have condensed, summarized, and paraphrased them. We believe we have presented the commenters’ views accurately, and have responded to all of the significant issues raised by the commenters that were within the scope of these rules.

Some commenters also wrote in about issues that were not related to the proposed rules, and in some cases not to Social Security disability benefits. Although we did read those letters, we did not respond to them.

Also, some commenters sent comments supporting the rules changes and noting provisions with which they agreed without suggestions for changes in those provisions. In most cases, we have not summarized or responded to those comments below because they do not require a response. However, we appreciate receiving them.

Use of Functional Criteria in the Immune System Disorders Listings

Comment: Several commenters supported our proposal to add functional criteria to each of the listings in this body system. However, three other commenters expressed concerns about the proposal. One commenter suggested that we should avoid introducing functional criteria into

these listings. The commenter observed that, while the consideration of functional impacts may result in greater latitude among adjudicators and more flexibility in decisionmaking, there is also an element of subjectivity that could result in greater inconsistency in our decisions. The second commenter, who generally agreed that "functioning should be considered in ratings," said that the addition of functional criteria to the listings for immune system disorders other than HIV infection would not make the evaluation of these disorders any easier. This commenter said that considering functional information in claimant and third party reports of activities of daily living, and treating physician and other source statements would make evaluating these disorders more difficult. The commenter also believed that more evidence would be needed to support the decisions.

We address the third commenter's concern in the next comment and response.

Response: As we explained in the NPRM (71 FR at 44440) and earlier in this preamble, we are adding the functional criteria in response to many comments we received on the ANPRM and in public outreach meetings. As many commenters pointed out, the debilitating effects of immune system disorders are often "invisible"; that is, outward signs of the disorders and objective severity markers often are not obvious and we cannot describe them in a listing. Because of this, the proposal received support from many individuals (or their family members) who received disability benefits only after going through a long appeals process. We also received comments about inconsistencies in our adjudications because we did not provide the kinds of guidance about evaluating the functional impact of immune system disorders that we do in these final rules.

Therefore, we do not agree with the commenters who thought that adding the functional criteria would have the negative effects they described or that we should not add functional criteria to these listings. To the contrary, we believe that these final listings will result in more consistent adjudications, and in some cases, faster adjudications, a need for less development, and fewer cases in which appeals are necessary, as we explain in more detail below.

The final listings describe individuals who are very ill. To qualify under one of these listings, an individual must first establish with objective medical evidence that he or she has the type of immune system disorder described by a given listing. Second, the individual must show that he or she repeatedly

becomes ill as a result of the impairment. These two findings alone establish that the individual has a significant medical problem. The third requirement, to show a "marked" limitation in at least one of the areas of functioning, establishes that the overall impairment causes serious limitations.

A "marked" limitation as we define it is an obvious, serious limitation that affects all aspects of the individual's life (activities of daily living, social functioning) or the ability to do tasks (deficiencies in concentration, persistence, or pace). Therefore, it can be easier for an adjudicator to assess whether there is a "marked" limitation in an area of functioning, and to justify that assessment, than it is to assess and justify a residual functional capacity assessment. Residual functional capacity is more detailed, requiring evaluation of specific physical and mental work-related functions, what we often call a "function-by-function" assessment.

Because of this, without these final listings, our adjudicators would have to do more work in most, if not all, cases of individuals who have immune system disorders that will meet these final listings only to reach the same decision. Under the prior rules, virtually all of the individuals who could now qualify under the new functional listings required a residual functional capacity assessment. Our adjudicators not only had to do additional work to provide this more detailed assessment of functioning, but they also had to do the additional work associated with making findings about the ability to do past relevant work at step 4 of the sequential evaluation process, and to make an adjustment to other work at step 5. Each of these determinations—function-by-function residual functional capacity assessment, assessment of the ability to do past relevant work, and ability to make an adjustment to other work—required development of information. We believe that in some cases adjudications under these final listings will be easier, faster, and more consistent.

Finally, we have significant experience applying these and similar functional criteria in many claims. We began using these functional criteria in listing 14.08 in 1993. We used some of the same criteria to evaluate physical impairments in children when we first implemented the policy of functional equivalence for children in 1991,² and have used similar kinds of criteria for evaluating functional equivalence in physical impairment claims since 2000

under § 416.926a of our rules (65 FR 54747 (2000)). Many of our listings, including most of our musculoskeletal listings, several of our cardiovascular listings, and most of the neurological listings, contain functional criteria.

Comment: The third commenter (whose comment was about the functional criteria in proposed listing 14.08) suggested that limitations in maintaining social functioning and in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace are basic issues for evaluating mental impairments under 12.00, for mental disorders, and should be removed from the listing. Similarly, one of the two commenters whose comments we summarized in the preceding comment summary expressed concern that adjudicators could assume that the functional criteria in listing 14.08 pertain only to the evaluation of mental impairments because they are similar to those considered in the context of the mental listings.

Response: We do not agree that maintaining social functioning or completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace describe only mental functioning and should be removed from listing 14.08K or any of the other corresponding final listings. We addressed this issue at length in 1993 when we first published these rules. In the preamble to the 1993 publication of the rules, we explained in responding to public comments:

We do not agree that it is inappropriate to apply these functional criteria to physical disorders because the criteria are generic; they do not describe mental functions, but broad areas of functioning that are relevant to any adult's ability to work or any child's ability to independently, appropriately and effectively engage in age-appropriate activity. * * * [T]hese activities describe what people do and how well they do it on a day-to-day basis. For our purposes, it is immaterial whether an individual has difficulty doing chores or maintaining concentration because of a mental disorder or because of fatigue, weakness, pain, headaches, frequent diarrhea, or any other physical problem; the person still has the limitation that results from a medically determinable impairment(s).

58 FR at 36040. We also explained that we had modified the language of the introductory text to make it more specific to individuals with HIV infection. Those modifications remain in these final rules with even further clarifications.

A number of commenters on the 1993 rules specifically commented that the area of social functioning is meant to measure an individual's psychiatric

² See generally 56 FR 5534 (1991).

condition and is not appropriate for the evaluation of HIV. We responded that:

* * * the ability to interact with other people can be affected by a physical impairment. For instance, an individual who is fatigued may have difficulty going out or sustaining conversation. * * *

58 FR at 36041.

In addition, and as we noted in the response immediately preceding this one, over the almost 15 years since we first published listing 14.08, we have gained considerable experience applying functional criteria such as these to physical impairments.

In final 14.00I, as in the NPRM, we provide that functional limitations may result from the impact of the disorder on mental functioning, physical functioning, or both mental and physical functioning. As we indicated in the NPRM, we revised 14.00I so that it applies to all of the listed impairments and more consistently refers to symptoms that are related to physical impairments. We believe that these revisions will help our adjudicators to better understand and remember that the areas of functioning should be applied to physical, as well as mental, limitations. However, we will provide training on the new functional criteria in these final rules.

Comment: One commenter said that adjudicators will need guidance on how to determine whether to use the immune system disorders listings alone versus completing the typical full documentation required for the mental disorders listings. The commenter remarked that doing additional mental development such as obtaining a consultative examination for a mental status examination could potentially delay a claimant's determination.

Response: We agree that guidance is needed and plan to address this issue in the training that we will conduct on these final rules. We do not believe that mental consultative examinations will be required as a result of these final listings because we are not trying to document mental impairments. Rather, we are determining any functional limitations and restrictions that a person may have as a result of his or her immune system disorder(s). As we do for other impairments, such as HIV infection, we would expect adjudicators and reviewers to assess functioning by evaluating objective medical evidence and evidence from other sources as described in §§ 404.1512 and 416.912.

Comment: One commenter suggested that we provide more concrete guidance on how to evaluate the severity of limitations in activities of daily living and more structure on the application of

terms such as "moderate, marked, and extreme" to reduce the likelihood of inconsistent interpretation of these terms.

Response: We did not adopt this comment because the application of these terms is often dependent on specific case facts, and because we believe that any additional detail would be better presented in training and other instructions. Our adjudicators have considerable experience evaluating "marked" and "extreme" limitations and have used the functional criteria in prior listing 14.08N which are similar to the criteria we include in these final rules. However, we will remind adjudicators of our guidance in these areas when we conduct training on these final rules.

Comment: One commenter referred to proposed 14.00I and said that it "introduce[d] the concept of 'repeated manifestations accompanied by functional limitations'" and the application of this concept to eight listings. The commenter observed that this "new way of evaluating the impact of repetitive episodes" was "sound in theory" but "may be difficult to apply in practice" because of the implicit need to document activities of daily living during periods sometimes well in the past. The commenter suggested that we clarify that the intent of the listings that include standards for evaluating functional limitations resulting from repeated manifestations of immune system disorders is to document functional limitations occurring in the present and does not require extensive documentation of the impact on activities of daily living during earlier episodes. The commenter indicated that evaluating the impact of repetitive episodes may be difficult because of the extended time period for which we may need to develop documentation of activities of daily living.

Response: We believe we accommodated this comment by adding language in final 14.00I3 explaining that the manifestation episodes must occur within the period covered by the claim. As we already do, for example, whenever we need to assess residual functional capacity, we will develop evidence about the individual's functioning for the entire period covered by the claim. The final rules do not impose any additional burden in that regard, as we have explained in our responses to the preceding comments.

Also, we must note that the concept of repeated manifestations accompanied by functional limitations is not new. We have used the criterion in the HIV infection listings since 1993. The innovation in these final rules is to

apply the same kind of criterion to the other listed immune system disorders.

Systemic Lupus Erythematosus (SLE)

Comment: One commenter thought that the terms "repeated," "marked," and "manifestation" in the SLE listing could cause confusion for physicians and adjudicators. The commenter recommended that we clarify the definition of each term or replace the section in the SLE listing with a different rule, which the commenter also proposed. (We address the proposal to replace the SLE listing in a later comment and response.)

With regard to the term "marked," the commenter believed that our proposed definition was ambiguous. The commenter suggested that we add more examples of "marked" and define it, giving examples of "moderate" for comparison. The commenter also said that physicians do not use the term "marked" in describing limitations resulting from SLE.

The commenter also suggested that we provide a definition of "manifestation" with examples because it was not defined in the proposed rule.

Response: We do not expect physicians and other medical sources to use our terminology. We only need for them to provide us with medical evidence that we will use to determine whether an individual's impairment meets the requirements of a listing. For example, a physician does not need to tell us that a flare of his or her patient's SLE was a "manifestation," only report to us what occurred in medical terms, and if necessary, provide an opinion that it was related to the SLE.

Likewise, we realize that physicians may not use the term "marked" in describing limitations resulting from SLE. However, for the purpose of determining disability, the issue of whether an individual has a "marked" limitation is an administrative finding that we make based upon consideration of all relevant evidence in the individual's case record, which may include information that the treating source does not have. We only need evidence describing the individual's limitations, and we will determine whether those limitations meet our definition of "marked."

The definitions of the terms "repeated" and "marked" in these final rules are substantively the same as the definitions of these terms in our prior rules, and our adjudicators have been using these definitions since 1993, when we issued the prior rules. As we have already noted, we use the term "marked" in a number of our other rules as well.

Comment: With regard to the term “repeated,” the same commenter indicated that patients might not see their physicians often enough to satisfy the criterion in the proposed rule, or physicians might not record the required information in a patient’s chart. The commenter said that physicians may not spend time documenting their records because of time constraints, and this would be a problem if the individual later applies for disability benefits.

Response: We understand the commenter’s concern. However, such individuals with SLE can still qualify under final listing 14.02A, which does not require a showing of repeated manifestations, and in other ways; for example, with impairment manifestations that meet other listings, based on our policy of “medical equivalence,” or based on residual functional capacity. We address the latter issues in final 14.00G6 for individuals who have not received ongoing treatment or do not have an ongoing relationship with the medical community, and final 14.00J, for individuals whose impairments do not meet the requirements of one of these listings.

Comment: The commenter also said that the requirement for repeated manifestations did not recognize that SLE can cause permanent damage that remains chronic after the manifestations have stopped. As an example, the commenter described an individual who had a severe heart attack caused by lupus, who does not experience any new manifestations, but who is disabled from permanent heart damage.

Response: The example of an individual who has permanent, disabling heart damage that the commenter provided is an example of the principles we discussed in the response immediately above. If the heart damage is sufficiently severe, it would meet or medically equal one of our cardiac listings in 4.00, the cardiovascular body system. Even if it does not meet or medically equal a listing in the cardiovascular body system, it could be the basis for a finding of disability at the last step of the sequential evaluation process because of the functional limitations it causes.

Also, our criteria for evaluating repeated manifestations of SLE do not require repetition of the same manifestation. For example, an individual who has experienced three different manifestations of SLE (for example, heart problems, leukopenia, and pleuritis) with the frequency and duration required in final 14.00I3 would

have an impairment that satisfies the criterion in final listing 14.02B. In response to this comment, we have added language to final 14.00I3 to make this clear. This is not a change in what we proposed, only a clarification of our intent.

Comment: The same commenter also suggested that we use the term “flare” instead of “manifestation” because that is the word physicians treating SLE use to describe increased symptoms and disease activity.

Response: We are aware that physicians who treat SLE often use the term “flare” to describe increased symptoms and disease activity. However, “flare” implies a temporary state, and our term “manifestation” does not necessarily mean that. We believe that many medical professionals who do not work for us will understand our term, but it is not critical that they do.

Comment: The same commenter provided a suggested replacement for the criteria in proposed listing 14.02B that included language such as “severe impairment” in one of the domains and the “opinion” of a specialist regarding prognosis for improvement in functional capacity. The commenter indicated that the proposed criteria were medically accurate for evaluating lupus, could be documented through a claimant’s medical records, and could be easily applied by adjudicators.

Response: We did not adopt the recommendation for a number of reasons. The commenter’s criteria included essentially the same criteria we had proposed. However, the commenter would have also required medical evidence that shows that treatment has not significantly reduced the severity of the disorder and is not likely to restore the capacity to work. This would have made the listing stricter than what we had proposed and stricter than the prior listing.

Comment: One commenter suggested that we add “intense generalized muscle aches and pains” to the constitutional symptoms and signs of severe fatigue, fever, malaise, or weight loss in proposed listing 14.02 because it is the most common symptom that rheumatologists who treat individuals with lupus hear from their patients.

Response: We agree that intense generalized muscle aches and pains is a common complaint of individuals with SLE. However, these symptoms generally respond to treatment. If the muscle aches and pains persist or do not respond to treatment, they may be the result of a secondary disorder other than SLE. Therefore, we did not adopt this comment.

Systemic Sclerosis (Scleroderma)

Comment: One commenter suggested that we should make the criterion for toe contractures in listing 14.04B1 more specific to make it more comparable with the criteria for finger contractures in proposed listing 14.04B2, atrophy of the lower extremities in proposed listing 14.04B3, and atrophy of the upper extremities in proposed listing 14.04B4. The commenter remarked that ordinary hammer toes are contractures and only the most severe result in significant incapacity.

Response: We did not adopt the comment because we believe that it is clear that listing 14.04B1 cannot be met with simple hammer toes. The listing requires that the toe contractures be so serious that they result in the inability to ambulate effectively. This is consistent with listings 14.04B2, 14.04B3, and 14.04B4, which require contractures or atrophy with irreversible damage resulting in either the inability to ambulate effectively or the inability to perform fine and gross motor movements effectively.

Comment: One commenter pointed out that our inclusion of the phrase “or of a toe and finger” in proposed listing 14.04C1 was redundant because we also required that the gangrene must be present in at least two extremities. The commenter said that the intent to require two extremity involvement is clear and suggested that we remove the rest of the language in proposed listing 14.04C1.

Response: We adopted the comment.

Immune Deficiency Disorders, Excluding HIV Infection

Comment: One commenter suggested that when we give examples of primary immune deficiency disorders in these proposed rules we use “Common Variable Immunodeficiency Disorder (CVID)” instead of the word “agammaglobulinemia” because it would be less confusing.

Response: We did not adopt this comment because the example we use in these rules is of “X-linked agammaglobulinemia” and the term CVID does not include this disorder.

Comment: One commenter suggested that we clarify what constitutes “sepsis” as required in proposed listing 114.07A1 for immune deficiency disorders. The commenter remarked that it is not uncommon for clinicians to inappropriately label someone as having sepsis or urosepsis when the more correct diagnosis was bacteremia with a urinary tract infection.

Response: We did not adopt this comment because we do not agree that

sepsis is commonly misdiagnosed as bacteremia. Additionally, sepsis is such a serious condition that we believe that it will be clear from the medical records when bacteremia is incorrectly labeled as sepsis.

Human Immunodeficiency Virus (HIV) Infection

General

Comment: Many commenters suggested that the final rules should include enough general language to accommodate the inevitable changes in understanding and treatment of HIV infection that will occur during the anticipated 8-year life of the rules. The commenters believed that we would unfairly deny individuals if we did not include such general language and if the individuals' medical records did not include the clinical markers required by these listings. The commenters recommended that we add a criterion for "an infection that is systemic or disseminated" to listings 14.08A through F in recognition of these anticipated changes. The commenters also suggested that the rules should accurately and comprehensively reflect the current understanding of HIV disease and treatment.

Response: The final rules, like the prior rules, do include general language that will allow our adjudicators to establish the existence of HIV infection and identify manifestations of HIV infection based on future advances in medicine and changes in medical science.

- With regard to definitive diagnosis of HIV infection, we include in final 14.00F1a(vi) a catchall criterion for "[o]ther tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge." This criterion is similar to prior 14.00D3a(iii), and we include it specifically to allow for future advances or changes in the methods for diagnosing HIV infection.

- Likewise, as in 14.00D3b of the prior rules, we include in final 14.00F1b a provision that allows our adjudicators to 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 [the individual's] case record." This permits our adjudicators to establish the existence of HIV infection based on current prevailing medical practice and even in the absence of laboratory testing. (For an additional explanation of this provision when we originally

published it in 1993, see 58 FR at 36019 and 36033.)

- With regard to the manifestations of HIV infection, the language in these final rules is general. For example, final 14.00F3a requires only definitive documentation "by culture, serologic test, or microscopic examination of biopsied tissue or other material." Final 14.00F3b contains virtually the same language as in final 14.00F1b regarding other acceptable documentation of the manifestations of HIV infection.

Additionally, we did not add the recommended listing criterion for two reasons. First, the listings are only examples of impairments that we consider severe enough to prevent any gainful activity and are not meant to be an all-inclusive list of such impairments. If an individual with HIV infection has an opportunistic disease or other condition that is not listed, we will consider whether it medically equals any listing; that is, whether it is as medically severe as an impairment in the listings. Second, if we added the language proposed by the commenters we might inadvertently include some persons who do not have listing-level impairments.

It is also important to remember that we do not deny benefits to anyone simply because his or her impairment(s) does not meet or medically equal the severity of a listing. We may still find such an individual disabled based on other rules in the appropriate sequential evaluation process for adults or children.

We do, however, agree that the listings should reflect the latest medical knowledge of HIV infection. As noted earlier, we are publishing separately an ANPRM in today's edition of the **Federal Register** inviting comments and suggestions on how to update and revise our listings for HIV infection. We believe that we need additional information before considering whether to propose additional changes to the criteria in the HIV infection listings.

Comment: Many commenters suggested that we add guidance to acknowledge that disability may result from conditions that are not specified in these final listings or that may emerge as a result of new or sustained HIV treatment by adding the following guidance: "Special consideration should be given to other conditions, signs and symptoms deemed by the primary care provider as contributing to substantial functional limitations."

Response: We did not adopt these comments. The final listing—like the prior listings—already allows for the consideration of conditions that are not specified and that may arise in the

future. The opening paragraph of final 14.08K explains that HIV manifestations considered under this listing can be the manifestations listed in 14.08A–J "or other manifestations," and then provides a parenthetical list of examples of such other manifestations. Since the parenthetical list says "for example," the listing does include any other manifestations of HIV infection, including new manifestations that may arise in the future. The nature of the manifestation is less important than the fact that the individual repeatedly experiences them.

We did not include the phrase "deemed by the primary care provider as contributing to substantial functional limitations" because the statement is not an accurate characterization of how we determine the existence and severity of impairments, impairment manifestations, and functional limitations, or of how we consider medical opinions from treating sources. We have other, general rules that explain these policies, and it would not be appropriate to repeat them in a listing.

Also, if a new manifestation should arise in the coming years, we will still be able to tell our adjudicators about it through internal guidelines we can issue. We can also provide training if necessary.

Comment: Many commenters suggested that these rules should address the interplay between HIV and mental health. The commenters said that the rules should recognize that mental health conditions can be a manifestation of HIV infection which, even if they do not meet or medically equal mental disorders listings, should be considered as repeated manifestations of HIV infection. They also said that the rules should indicate that attention must be paid to the signs and limitations that stem from mental and emotional deficits when evaluating the severity and level of progression of HIV disease.

Many commenters remarked that HIV medications can themselves cause mental impairments, such as significant memory loss, cognitive deficits, depression, anxiety, paranoia, and hypervigilance. These commenters also indicated that mental illness may become more pronounced as the HIV disease progresses and can interfere with self-care, activities of daily living, and adherence to treatment regimens and appointment schedules. The commenters suggested that primary care providers and infectious disease specialists may prescribe compensatory medications, such as anti-depressants and anti-anxiety medication, to their

patients without referring them for psychiatric care or counseling. They said that, in such cases, there will be no longitudinal history of psychiatric care or assessment, but that we should recognize these manifestations of HIV infection which contribute to the disabling nature of the disease. The commenters suggested that we add another subsection to final 14.00F to make these points and that we revise listings 14.08K and 114.08L to recognize specifically that mental health conditions can be a manifestation of HIV infection that can be considered under those listings.

Response: We did not agree with these comments, but we clarified a phrase in the final rules in response to them. The proposed rules did, and these final rules do, recognize the interplay between HIV infection and mental health, and that mental health conditions can be manifestations of HIV infection. While we did indicate in proposed 14.00J2 that individuals with immune system disorders “including HIV infection” may manifest signs or symptoms of a mental impairment that could be evaluated under the mental disorders listings, we also made provision throughout the immune system disorders listings for individuals whose mental impairments would not meet or medically equal a mental disorders listing, and recognized that mental limitations could result from HIV infection or its treatment.

First and foremost, we included “cognitive or other mental impairment” as an example of a manifestation of HIV infection that would satisfy the requirement for repeated manifestations in proposed listing 14.08K. We also provided in proposed 14.00G1, 14.00G5, and their corresponding childhood sections that limitations in mental functioning can be a side effect of treatment for immune system disorders, while in proposed 14.00I4 and 114.00I3 we indicated that mental limitations can result from the impact of the disease process itself. All of these provisions are in the final rules.

We did not add some of the other information the commenters suggested because we believe that it is too detailed for inclusion in our listings, and some of the proposals also would apply to our evaluation of other immune system disorders as well as HIV infection. However, we will consider including this guidance in the training we provide for our adjudicators on these listings.

However, in response to these comments, we changed the phrase “cognitive or other mental impairment” in proposed 14.08K to “cognitive or other mental limitation” in final 14.08K.

This should help to clarify that we will consider cognitive or other mental limitations as manifestations under this listing regardless of whether the existence of a “mental impairment” (that is, a mental condition) has been established.

Comment: Many commenters suggested that we make it clear throughout the proposed rules that each claimant is entitled to an individualized assessment of his or her HIV infection.

Response: We did not make any changes in response to this comment. The commenters did not provide examples of sections of the rules that they thought should be improved and did not recommend specific revisions, and we believe these final rules do make clear that we require an individualized assessment of an individual’s HIV infection or any other immune system disorder. For example, the rules stress the importance of considering the individual’s symptoms and limitations caused by the disease or its treatment. Also, individualized assessment is a general principle that applies throughout all of our disability rules.

Comment: Two commenters questioned our decision to not make any substantive changes to the proposed HIV infection listings that require HIV infection and certain opportunistic infections, such as the listing for PCP. The commenters indicated that there have been advances in the understanding and treatment of HIV infections since these listings were originally published. One commenter remarked that the widespread availability of highly active antiretroviral therapy (HAART) has changed the occurrence and progression of complications of HIV infection and that scientific advances have permitted the dosing of much fewer pills than previously required. Other commenters, including a medical association representing HIV medical providers, supported our decision not to change the stand-alone listings contained in listing 14.08.

Response: As noted in the NPRM, we carefully considered the advances in treatment and consequent increases in longevity that have occurred since we published the prior rules in 1993. Based on this review, we did not believe that there had been sufficient progress in the treatment and control of HIV infection to warrant any change in these rules at that time. However, as a result of public comments on the NPRM, we now believe that some changes may be appropriate. Therefore, as noted above, we are publishing separately an ANPRM in today’s edition of the **Federal Register** inviting comments and

suggestions on how we might update and revise our listings for HIV infection. We will consider the comments and suggestions that we receive in response to the ANPRM, as well as our adjudicative experience and additional information about advances in medical knowledge, treatment, and methods of evaluating HIV infection. If we determine that listing 14.08 should be further revised, we will publish for public comment an NPRM that will propose specific revisions to the listing.

Comment: Three commenters suggested that there should be a time period for reviewing claims allowed under proposed listing 14.08, such as a period of 12 months or 3 years, similar to the time period we have in some other listings, such as organ transplants and malignant neoplastic diseases.

Response: We did not adopt this comment. The disease process for HIV infection is not the same as it is for disorders such as organ transplants or malignant neoplastic diseases, and we do not believe the use of timeframes for the HIV infection listings would be appropriate at this time.

Manifestations of HIV Infection

Comment: One commenter suggested, without explanation, that we modify the criteria in proposed listing 14.08A1 by eliminating the requirement that pulmonary tuberculosis be “resistant to treatment.”

Response: We did not adopt this comment. We added pulmonary tuberculosis resistant to treatment in 1993 in response to public comments. (58 FR at 36021) We are unaware of changes in medical science or treatment since then that would indicate that we should consider pulmonary tuberculosis that is responsive to treatment to be of listing-level severity, and the commenter did not provide a reason for the recommendation.

Comment: One commenter suggested that we include esophageal candidiasis in the examples of those conditions in final 14.00F3b for which a presumptive diagnosis can be made. The commenter indicated that, like PCP, CMV diseases, and toxoplasmosis of the brain, esophageal candidiasis is typically diagnosed based on clinical manifestations, history, and treatment response, and that when it is, it will meet listing 14.08B2. Another commenter made a similar comment and suggested that we include information about medical and other evidence that could be used to presumptively diagnose *Candida* esophagitis, similar to the guidance in 14.00F3b(i) for PCP. This commenter suggested that such guidance would

remind our adjudicators that a diagnosis of “*Candida esophagitis*” without supporting medical evidence is insufficient to meet or medically equal listing 14.08B2.

Response: We adopted these comments by adding new paragraphs 14.00F3b(iv) and 114.00F3b(iv). They describe other acceptable evidence that we may use to document the presence of candidiasis of the esophagus, also known as *Candida esophagitis*. We agree with the first commenter that presumptively diagnosed *Candida* of the esophagus meets the requirements of the listing. We also agree with the second commenter that a diagnosis alone is not sufficient to establish disability under the listing; we must have medical evidence to support the diagnosis. We did not state this in the new paragraph because it is a basic principle in our disability programs, applicable to any impairment.

In the new paragraphs, we provide guidance indicating that typical treatment response “can be supportive of the diagnosis,” consistent with the first commenter’s recommendation. For consistency, we added the same guidance in final 14.00F3b(i) and 114.00F3b(i) in the statement about treatment response for PCP.

Comment: One commenter suggested that the guidance in proposed 14.00F3b(i) for documenting the diagnosis of PCP without definitive laboratory evidence was questionable and insufficient. The commenter remarked that the diagnosis of PCP should be documented on the basis of prevailing and accepted medical knowledge, and that the discussion in this proposed section should otherwise be deleted.

Response: We did not agree with this comment. The criteria we included in the NPRM and these final rules are appropriate examples of medically accepted supportive evidence of PCP infection.³ However, in response to this comment we are adding “no evidence of bacterial pneumonia” in final 14.00F3b(i) and 114.00F3b(i) as another piece of supportive evidence that may be used to diagnose PCP presumptively.

Comment: One commenter suggested that we change the reference to “*Pneumocystis carinii* pneumonia (PCP)” in proposed 14.00F1b to “*PneumoCystis* Pneumonia (PCP) caused by infection with *Pneumocystis jiroveci*” to be more consistent with prevailing medical knowledge. The commenter also suggested that we

change the criteria of “*Pneumocystis carinii* (*jiroveci*) pneumonia or extrapulmonary *pneumocystis carinii* (*jiroveci*) infection” in proposed listing 14.08B7 to “*PneumoCystis* Pneumonia (PCP) or extrapulmonary *pneumocystis* infection caused by *Pneumocystis jiroveci*.”

Response: We partially adopted the comment. In final 14.00F1b and final listing 14.08B7, we now refer to “*Pneumocystis* pneumonia (PCP)” to reflect current medical terminology. Because of this change, we also removed the note we had proposed to include in 14.00F3b(i) which explained that “*Pneumocystis carinii*” is now known as “*Pneumocystis jiroveci*” and that “PCP” remains in common usage for the pneumonia caused by this organism. We no longer need the note because we no longer refer to *Pneumocystis carinii* or *Pneumocystis jiroveci* in these rules. We also made corresponding changes in the childhood introductory text.

Comment: One commenter suggested that we include an authoritative source for moving prior listing 14.08B7 for PCP from the section of the listings for protozoan and helminthic infections to the section of the listings for fungal infections.

Response: When we published the NPRM, we listed the references that we consulted when we were developing the proposed rules (71 FR at 44448). This list included “Medical Management of HIV Infection” (Johns Hopkins University 2003) by J.G. Bartlett and J.E. Gallant, which classifies *Pneumocystis carinii* as a fungal infection.

Comment: One commenter suggested that we modify the language in the next to the last sentence in proposed 14.00F3b(ii) to clarify that we do not require the presence of all of the signs noted in this sentence to support a presumptive diagnosis of *Cytomegalovirus* by indicating that the supporting evidence “may” include the findings we listed.

Response: We adopted the comment. As we noted in the summary of the final rules earlier in this preamble, we are also adding the word “may” in final 14.00F3b(i), for PCP, to be consistent with this change.

Comment: One commenter suggested that we clarify whether the intent of proposed listing 14.08E4, for squamous cell carcinoma of the anus, was to include both anal canal cancers and anal margin tumors or to limit the listing solely to anal canal cancers (developing from mucosa).

Response: We adopted the comment by changing the criterion to “Squamous cell carcinoma of the anal canal or anal margin” in final 14.08E4 and 114.08E4.

This is not a substantive change, but only clarifies our intent.

Comment: Many commenters said that we should revise the criteria in proposed listing 14.08H for evaluating HIV wasting syndrome to reflect more current medical knowledge about this condition. They said that we should provide that body mass index (BMI) and body cell mass (BCM) can be relied upon as accurate indicators of the severity of wasting in a given individual. They also said that this listing is too restrictive in its documentation requirements, and that involuntary weight loss as low as 5 percent has been associated with increased risk of death. Another commenter suggested that we revise the criteria for this listing to “HIV wasting syndrome, characterized by involuntary weight loss of 5 percent or more below ideal body weight within six months and, in the absence of concurrent illness that could explain the findings.” The commenter said that this would reflect medical guidelines for diagnosing the condition and the significance of rapid, unintentional weight loss.

Most of the commenters also said that the prior requirements for diarrhea were too restrictive because a person with HIV infection who experiences wasting is functionally unable to work if he or she experiences diarrhea for 2 weeks and protein deficiency. They also said that, although a documented fever is a useful clinical indicator of wasting syndrome, the listing should not require the individual to have “many temperature readings throughout a month or for a longer period.” They said that HIV wasting syndrome can be disabling even in the absence of the listing requirement when it is accompanied by constitutional symptoms, such as weakness, lack of muscle strength, fatigue, malaise, or inability to lift. They suggested that as an alternative to evidence of diarrhea or fever, the listing could contain language comparable to that in proposed 14.00F; that is, “documented by other generally acceptable methods consistent with the prevailing state of medical knowledge or clinical practice.”

Response: We agreed with the commenters who suggested that we include a reference to BMI in the listing, and have clarified final listing 14.08H by explaining that we can compute the 10 percent loss of weight in pounds, kilograms, or by BMI. We did not add a reference to BCM because BCM is more of a research concept, involves calculations of body composition, and is not in wide usage in the general medical community.

³ See *Cecil Textbook of Medicine* at 2059–2064 (Lee Goldman and Dennis Ausiello, eds. 22nd ed., 2004).

We also added guidance in final 14.00F5 to remind adjudicators that they can evaluate HIV infection that affects the digestive system and results in malnutrition under listing 5.08. Even though there is no listing for “wasting syndrome” in part B, there is a criterion in final listing 114.08H3, the growth disturbance listing, for a loss of 10 percent of body weight. We have added the same statement about pounds, kilograms, and BMI in that final rule as well, and a statement referring to listing 105.08 in the digestive system at the end of final 114.00F4a.

We did not make other changes in these final listings in response to the comments. We use listings to find individuals whose impairments are so severe that we do not need to consider their age, education, and previous work experience to decide that they are disabled. We believe that, while some individuals with the findings recommended by the commenters will be disabled under our rules, and some will be at risk of dying, others will not, so we cannot presume disability based on those findings in all, or even most, individuals. Even if they are initially unable to work, we believe that many individuals with the findings suggested by the commenters will not have impairments that meet the duration requirement in the Act and our regulations, that is, have an impairment that is expected to result in death or that has lasted or can be expected to last for a continuous period of not less than 12 months.

However, some individuals with a 5 percent weight loss will have impairments that meet the requirements of listing 14.08H; in some individuals, a 5 percent weight loss will be a “significant involuntary weight loss.” As we explain in final 14.00F5, final listing 14.08H does not require a specific minimum amount or percentage of weight loss. We always consider an involuntary weight loss of at least 10 percent of baseline “significant,” but an involuntary weight loss of less than 10 percent may also be “significant” depending on the individual’s baseline weight and body habitus. We also provide examples in final 14.00F5 of when weight loss of less than 10 percent of body weight may and may not be significant.

Likewise, although we agree that an individual with HIV infection who experiences diarrhea for 2 weeks with protein deficiency would have work-related limitations, and may be unable to work for a time, we do not believe that this finding by itself would necessarily be indicative of an impairment that would be expected to

result in death or prevent the ability to work for a continuous period of at least 12 months. We must consider the specific facts of such individuals’ cases to decide whether they are disabled.

With regard to the comment about fever, we did not include a requirement in the prior rule or proposed rule, nor do we include one in the final rule, for the number of times during the course of a month in which the individual’s temperature must be taken. We must only have sufficient information to determine that the individual has had a persistent fever throughout most of a month. More importantly, the criterion for fever in final listing 14.08H2 is only one of two criteria in listing 14.08H by which an individual may qualify, so an individual could qualify under this listing without fever. We believe that the fever criterion is medically supportable as an indicator of an HIV infection of listing-level severity when considered in the context of the other criteria of involuntary weight loss and chronic weakness. Also, an individual with wasting syndrome could qualify without a finding of fever and with the kinds of constitutional symptoms and signs suggested by the commenters under final listing 14.08K.

We also did not add language that is comparable to that in proposed 14.00F as an alternative to the evidence of diarrhea or fever because the criteria in final listings 14.08H1 and 14.08H2 are severity criteria. The language proposed by the commenters would only help to establish the diagnosis of wasting syndrome and would not be sufficient to establish severity or duration under the listings.

However, as we noted earlier, we are publishing separately an ANPRM in today’s edition of the **Federal Register** inviting comments and suggestions on how we might update and revise our listings for HIV infection. We believe that we need additional information before determining whether to propose any substantive changes to the criteria in the HIV infection listings.

Comment: Many commenters said that we should modify proposed listing 14.08I to reflect current medical views regarding diarrhea and its treatment. They said that many patients with disabling diarrhea do not require hydration and therefore are not treated with intravenous hydration, and that “tube feeding” is rarely used now to treat diarrhea.

The commenters said that diarrhea can rise to the level of being disabling without the objective findings in proposed listing 14.08I. They suggested that this listing should include individuals who have multiple loose

stools each day, bowel incontinence, or a combination of the two, despite modifications in HAART and anti-diarrheals. They also suggested that we should allow documentation by other objective evidence, such as reports of a rectal examination, stool culture, or fecal occult blood test. Finally, they recommended that we add language comparable to that in proposed 14.00F; that is, “documented by other generally acceptable methods consistent with the prevailing state of medical knowledge or clinical practice.”

Response: We did not adopt the comments in these final rules. While we agree that many individuals with chronic diarrhea do not need hydration and that tube feeding is rare, these criteria provide some objective verification of the chronicity and severity of the diarrhea and our adjudicative experience shows that individuals do qualify based on the criteria. We did not adopt the criteria the commenters proposed because we believe that they are not sufficient to reliably document the severity, frequency, and chronicity of the diarrhea for our disability evaluation purposes. We also believe that the other objective evidence the commenters proposed (that is, rectal examination, stool culture, and fecal occult blood testing) would not be sufficient for this purpose. Lastly, we did not adopt the comment asking us to add language to proposed 14.00F because it would only help to establish the existence of the impairment, not its frequency and chronicity.

Comment: One commenter suggested that we should characterize the symptom of “fatigue” in listing 14.08K as “severe fatigue” to reflect a symptom at listing-level severity and to be consistent with the other immune system disorders listings.

Response: We adopted the comment. The change is not substantive, but only a clarification. Like the prior rule and the proposed rule, the final rule specifies that the symptoms listed must be “significant.” Therefore, adding “severe” does not change its meaning. For consistency, we added the word “severe” before the word “fatigue” throughout these final rules.

Comment: One commenter asked why we limited proposed listing 114.08A4 to children less than 13 years of age, particularly when proposed 114.00F4c said that children age 13 and older may have an impairment that medically equals this listing. The commenter noted that there is nothing in the listing to alert one to the possibility of a medical equals for older children.

Response: We partially adopted the comment. The age 13 cutoff has been in this listing since we first published it in 1993. When we first published it, we explained in the preamble to the regulations that these types of infections are more serious and more indicative of a rapid decline in younger children, that we had considered a younger age cutoff, but that we decided on age 13 as a medically appropriate dividing line. See 58 FR at 36047.

The impact of pyogenic bacterial infections in children who are under the age of 13 is usually more harmful than in older children, and there is general medical acceptance for evaluating the severity of these infections differently depending on the age of the child. Therefore, we did not change the age requirement in this listing. However, in response to this comment, we added a reference to 114.00F4c in final listing 114.08A4 to remind adjudicators that children age 13 and older may medically equal this listing.

Suggested Additional Criteria for the Listing for HIV Infection

Comment: One commenter suggested that we “acknowledge” in final 14.00F2 that a CD4 count of 100 or less would document the severity or functional limitations of HIV infection and establish disability. The commenter remarked that the CDC classifies a person with HIV and a T-cell (CD4) count below 200 as having AIDS and that the susceptibility to illness for such individuals increases dramatically. The commenter also indicated that a person with HIV and a CD4 count below 100 is likely to exhibit an extreme susceptibility to opportunistic infections and disabling illnesses, have difficulty tolerating medication, experience graver physical conditions, and exhibit lower functional capacities than individuals with stronger immune responses.

Response: We did not adopt this comment. We agree that a CD4 count of 100 or less indicates an increased susceptibility to developing opportunistic infections and is an important finding when considering treatment options. However, we do not agree that CD4 counts are a good indicator of disability. We continue to have the same opinion we had when we published the prior rules in 1993. In the preamble to those rules, we explained that:

while a low CD4 count (and especially a rapidly declining CD4 count) is an indicator of a compromised immune system and a valuable tool for determining when to institute prophylactic treatment, there is no consistent correlation between a given CD4

count and how or whether an individual is functionally impaired by HIV infection. Individuals with high CD4 counts may be quite severely limited, while others with very low counts may be able to continue normal activities. One individual who commented on our proposed rules related his own story of living with HIV infection, noting that he continued to feel well and to work until his CD4 count was well below 100. He argued that to base our rules on such an unreliable indicator would be to unfairly stigmatize individuals who are able to function well despite low CD4 counts.

58 FR at 36018.⁴

There have been significant advances in treatment and monitoring of individuals with HIV infection since we published the prior rules in 1993. Therefore, we believe that what we said in 1993 is, if anything, even more relevant to our disability adjudications today.

Comment: One commenter suggested, without explanation, that we add “*Rhodococcus*” to the criteria of listing 14.08A for bacterial infections, “*Blastomycosis*” and “*Penicillium marneffei*” to the criteria of listing 14.08B for fungal infections, and “*Leishmaniasis*” and “*Microsporidiosis*” to listing 14.08C (protozoan or helminthic infections).

Response: We did not adopt these comments. We did include “*microsporidiosis*” in proposed, now final, listings 14.08C1 and 114.08C1; it was also in prior listings 14.08C1 and 114.08C1. We did not add the other suggested manifestations because the listings are only examples of impairments that we consider severe enough to prevent any gainful activity and are not meant to be all-inclusive. Also, if an individual with HIV infection has an opportunistic disease or other condition that is not listed, we will consider whether it medically equals a listing.

Comment: Many commenters suggested that the criteria in proposed listing 14.08D, for viral infections, should include individuals who have both HIV infection and hepatitis B or hepatitis C under listing 14.08D. The commenters said that individuals who are infected with both HIV and hepatitis are more prone to illness, more difficult to treat, and less able to function than individuals who are only infected with a hepatitis virus. They also indicated that co-infection with HIV and hepatitis B or C complicates the treatment of both conditions.

Response: We did not adopt this comment. While we agree that co-

infection with HIV infection and hepatitis B or C may complicate the treatment of these conditions, increase susceptibility to illness, and impact functioning, we also believe that the severity of the co-infection will vary from individual to individual and may not result in disability. Because of this, we believe that each claim involving this co-infection must be evaluated on a case-by-case basis. This includes evaluating whether the co-infection results in manifestations that would satisfy the criteria in final listings 14.08K or 114.08L.

However, we do provide in final 14.00G1f and 114.00G1f that the interactive and cumulative effects of treatments for co-occurring impairments, such as treatment for HIV infection and hepatitis C, may be greater than the effects of each treatment considered separately.

Comment: Many commenters said that we should add a stand-alone listing for chronic or severe acute pancreatitis under proposed listing 14.08. The commenters indicated that pancreatitis is frequently associated with HIV infection, can be caused by HIV infection or medications used to treat HIV infection, and may severely impair an individual’s ability to function. They also said that pancreatitis can cause severe and recurring manifestations, such as abdominal pain, nausea, vomiting, fever, chills, and shortness of breath, that can result in a hospital admission for 2 or 3 weeks at a time or in profound weight loss and long-term food intolerance.

One commenter suggested that we specify under this listing that an individual with HIV infection is disabled if he or she requires hospitalization for pancreatitis twice in a 1-year period. Other commenters suggested that we include a listing that is satisfied by evidence of one or more episodes of pancreatitis from which clinical recovery is incomplete after 6 months and is accompanied with disabling symptoms such as, but not limited to, abdominal pain, diarrhea, significant weight loss, nausea, anorexia, and glucose intolerance requiring frequent monitoring or treatment.

Response: We did not adopt the comments. Generally, pancreatitis in individuals with HIV infection is caused by HAART and is acute; the pancreatitis usually resolves after HAART is suspended briefly. Because of this, it would not be appropriate to add a stand-alone listing for episodes of pancreatitis or the other criteria recommended by the commenters. The criteria recommended by the

⁴ See also 58 FR at 36038, where we provided the same information in our response to the public comments about this issue.

commenters would not necessarily result in the inability to do any gainful activity for a continuous period of at least 12 months as required by the Act.

However, individuals with pancreatitis can qualify under these listings. As we did in the NPRM, we include pancreatitis as an example of an "other manifestation" under final listing 14.08K. (We do not refer to it in 114.08L because pancreatitis is not as frequent a problem in children as it is in adults. However, since the list of other manifestations is only a list of examples, pancreatitis is still included.) Many individuals who experience pancreatitis with the significant accompanying problems described by the commenters will also have serious functional limitations and will be able to qualify under final listing 14.08K. Individuals with problems such as profound weight loss with prolonged food intolerance may have impairments that meet or medically equal the requirements of other HIV infection listings or listings in other body systems; for example, listings 5.08 and 105.08 for weight loss. We may also find that they qualify based on an individualized assessment of residual functional capacity if there is an inability to work or, for children, functional equivalence.

Effects of Treatment for HIV Infection

Comment: Many commenters suggested that in proposed 14.00G5 and 114.00G5 we should directly address the issue of a claimant's non-responsiveness to HIV treatments and specifically state that the mere fact that an individual fails to respond to HAART does not indicate that he or she is not disabled or is not credible. They also suggested that we add a subsection addressing the fragility of persons who do not respond to prescribed treatment and the impact of reduced treatment options on them. The commenters noted that we addressed these issues in the "general section" on response to treatment (that is, 14.00G2 and 114.00G2) but thought that we should address these issues specifically for HIV infection in 14.00G5 and 114.00G5.

Response: We did not adopt these comments. As the commenters noted, we provide guidance in 14.00G2 and 114.00G2 that response to treatment and adverse or beneficial consequences of treatment may vary widely. These sections explain that we consider a variety of factors when evaluating response to treatment, including the limited number of drug combinations that may be available for treatment, and that we must consider the effects of treatment on an individual basis. We also provide a specific example of an

individual with HIV infection whose impairment does not respond to antibiotics or who develops a resistance to treatment that had worked in the past.

We included this new guidance in our rules to address the major issues that are raised in these comments, and we believe that it will help to respond to the concerns that the commenters raised, not only for individuals who have HIV infection but for individuals with other kinds of immune system disorders who experience the same kinds of problems. Therefore, we do not believe that there is a need to repeat this guidance specifically for HIV infection in final 14.00G5 and 114.00G5 at this time.

Comment: Many commenters suggested that we revise proposed 14.00G5 to address the difficulty of adhering to HIV treatment regimens, and to acknowledge that there are many valid reasons why individuals with HIV infection do not strictly adhere to their prescribed treatment regimens. They also suggested that the rules state that a claimant's admitted lack of adherence to HAART should neither reflect on the claimant's credibility nor indicate that his or her functional capacity is "artificially low." They indicated that claimants should not be penalized for their failure to adhere to complicated medication regimens.

Response: We partially adopted the comment. We agree that some individuals may have difficulty adhering to their treatment regimens for HIV infection, such as HAART, and that there may be valid reasons for their lack of adherence, such as side effects of treatment (for example, diarrhea, nausea, vomiting, neuropathy, or severe fatigue). We addressed this in proposed, now final, 14.00G to an extent, especially in 14.00G1 and 14.00G2, in which we provided a list of things that we consider when we evaluate the effects of treatment. We also have other rules that tell our adjudicators not to make the kinds of presumptions that concerned the commenters. For example, our regulations on evaluating residual functional capacity, §§ 404.1545 and 416.945, provide that adjudicators must consider all relevant evidence in determining a person's functional abilities; this means that they cannot draw conclusions only from the fact that an individual is not receiving or following treatment. In Social Security Ruling (SSR) 96-7p, we provide that, when we consider treatment in assessing an individual's statements about symptoms, "adjudicator[s] must not draw any inferences about an individual's

symptoms and their functional effects from a failure to seek or pursue regular medical treatment without first considering any explanations that the individual may provide, or other information in the case record, that may explain infrequent or irregular medical visits or failure to seek medical treatment." One of the examples of a good explanation that we provide in the SSR is "[t]he individual may not take prescription medication because the side effects are less tolerable than the symptoms."⁵

However, in response to this comment, we added a sentence to final 14.00H and 114.00H that is based on the sentence from SSR 96-7p quoted above. We chose this section for the new sentence because we believe that the issue that concerned the commenters will arise most often when we are evaluating symptoms and their functional effects. We did not add the more detailed information the commenters asked us to include because we determined that it would be too extensive to include in the final listing. However, we will address the issue in training and consider whether to provide written guidance in our internal instructions as well.

Comment: One commenter suggested that we expand proposed 14.00G5a to discuss the disfiguring aspects of treatment as an adverse effect of treatment. The commenter remarked that adverse reactions to treatment, such as "buffalo hump" and other fat redistribution can have a significant impact on the ability of a claimant who is HIV positive to function physically, as well as on his or her emotional well-being.

Response: We partially adopted this comment. We added a parenthetical statement in final 14.00G5a and 114.00G5a to clarify that "lipodystrophy" means fat redistribution. We also cite "buffalo hump" as an example of fat redistribution.

In addressing this comment, we also noticed that in the last sentence of the paragraph, where we referred to limitations from HIV infection, we mentioned only limitations that result from symptoms. Since the objective effects of HIV infection can also cause limitations, we expanded this sentence to include "signs" of HIV infection. We do not believe other changes are needed because the sentence also refers to the

⁵ See "Titles II and XVI: Evaluation of Symptoms in Disability Claims: Assessing the Credibility of an Individual's Statements," 61 FR 34483 (1996). Also available at: http://www.socialsecurity.gov/OP_Home/rulings/di/01/SSR96-07-di-01.html.

side effects of treatment, which includes “buffalo hump.”

Inflammatory Arthritis

Comment: One commenter recognized that we had removed reference listings and that we provided guidance for using appropriate listings in the introductory text. Nevertheless, the commenter suggested that in listing 14.09 we refer adjudicators to listings 1.02 and 1.03 when involvement of only one major lower extremity joint results in ineffective ambulation.

Response: We adopted the comment by revising the listing so that it is no longer necessary for adjudicators to refer to listing 1.02 or 1.03. As a consequence of this change, we also removed proposed 14.00D6e(iv) and 14.00D6e(v).

The commenter was referring to an anomaly in our prior rules. Like the prior listing, proposed listing 14.09A required inflammatory arthritis with involvement of two or more peripheral weight-bearing joints that resulted in an inability to ambulate effectively. However, some individuals who have involvement of only one major peripheral weight-bearing joint have an inability to ambulate effectively. Under the proposed listing and our prior rules, these individuals qualified under listing 1.02 in the musculoskeletal system, which specifies that the listing is met with “involvement of one major peripheral weight-bearing joint.” In reviewing this comment, we determined that it would be simpler if we included a provision similar to that in listing 1.02 under listing 14.09A. This inclusion allows our adjudicators to use the inflammatory arthritis listing for all individuals who have inflammatory arthritis that results in an inability to ambulate effectively.

Likewise, the proposed rules and our prior rules made a distinction between individuals with inflammatory arthritis who had persistent deformity without ongoing inflammation (evaluated under listing 1.02) and those who had ongoing inflammation (evaluated under prior listing 14.09). In reviewing the proposed rules in light of the comment letter, we realized that there is no practical reason to maintain that distinction.

We also realized that there was no reason to maintain the guidance in the prior and proposed rules that required the use of listing 1.03 when there had been reconstructive surgery. Final listing 14.09A1 is sufficient to cover the situation described in listing 1.03 for individuals with inflammatory arthritis who have had reconstructive surgery of a major peripheral weight-bearing joint and have been unable to ambulate effectively for at least 12 months or can

be expected to be unable to ambulate effectively for at least 12 months.

As already noted in the summary of the changes in these rules, we revised the second sentence in 1.00B1, in the introductory text of the musculoskeletal system listings, to reflect these changes. We also made corresponding changes in part B of the listings, in 101.00B, 114.00D6, and 114.09A.

Comment: One commenter suggested that the term “dorsolumbar” ankylosis in proposed listing 14.09C should indicate that “dorsolumbar” means dorsal and lumbar, not either one.

Response: We did not adopt the comment. The term “dorsolumbar” is a common medical term that is generally recognized to mean the area of the spine relating to the lower thoracic and upper lumbar vertebral region of the back. We used this term in prior listing 14.09B2 (final listing 14.09C1), and we are not aware that it caused any confusion. However, we will reinforce the definition when we conduct training on these final rules.

Other Disorders

Comment: One commenter noted that in proposed 14.00D6c(v) we mentioned Lyme disease only by name and only as an impairment that we evaluate under listing 14.09 for inflammatory arthritis. The commenter said that the symptoms of Lyme disease are the same as for SLE, and suggested that we provide criteria for evaluating the disorder similar to the criteria for SLE and Sjögren’s syndrome. The commenter also noted that Lyme disease with co-infections can be fatal.

This commenter and a second commenter noted that, like other immune system disorders, the symptoms of Lyme disease can be “invisible,” making it difficult to evaluate disability. One of the commenters suggested that we should not focus on the name of the disease but on its effects and made recommendations for how we could better adjudicate cases; for example, by giving more weight to reports from treating physicians. This commenter also noted that the symptoms of the impairment can improve at times but that we should not assume that an individual is not disabled just because he or she is able to function well for a short period. Both commenters also described difficulties in our adjudication system.

Response: We agree with the commenters that some individuals with Lyme disease have symptoms that are the same as or similar to the symptoms of SLE, Sjögren’s syndrome, and other immune system disorders we include in these listings. However, there are

hundreds of disorders that affect the immune system, and we are not able to list all of them in our listing of impairments. In proposed (now final) 14.00D6c, we included Lyme disease as an example of a disorder that could cause inflammatory arthritis because that is the most frequent disabling outcome of Lyme disease.

Some individuals with disabling Lyme disease will have impairments that meet the requirements of final listings 14.09B and especially 14.09D. Final listing 14.09D recognizes that individuals with Lyme disease and other disorders that can cause inflammatory arthritis can have serious functional limitations as a result of their symptoms, including the kinds of symptoms described by the commenters. The functional criteria in final listing 14.09D and throughout the final immune system disorders listings recognize the “invisible” nature of most immune system disorders. As we noted in the preamble to the NPRM, we also consider the variable nature of the symptoms of immune system disorders. (See 71 FR at 44441)

As in the proposed rule, final 14.00J also provides that individuals with immune system disorders that do not meet the criteria of one of these listings can have impairments resulting from their immune system disorders that meet the requirements of listings in other body systems, such as neurological or mental disorders. In final 14.00D6e(iii), as in the NPRM, we list such extra-articular features of immune disorders that can cause inflammatory arthritis by body system to provide guidance about such other effects that these disorders, including Lyme disease, may have. However, in reviewing these comment letters and the proposed rules, we realized that we had inadvertently omitted reference to possible mental signs and symptoms in this section. Therefore, we are including the phrase “mental (cognitive dysfunction, poor memory)” in final 14.00D6e(iii) in response to these comments. The phrase is the same one that we use in final 14.00D7a(ii) for Sjögren’s syndrome. We also added the same language in final 114.00D6e(iii) in part B.

Individuals who have Lyme disease but who do not have repeated manifestations of inflammatory arthritis can also qualify under the listings for SLE, Sjögren’s syndrome, or other appropriate listings in the immune disorders body system or any other appropriate body system based on our policy of medical equivalence.

Finally, we carefully considered the recommendations of the commenter

who suggested ways to improve our evaluations of cases involving Lyme disease. These suggestions were covered by other general regulations and policy statements we have, such as our policies for evaluating symptoms and treating source opinions. Therefore, we decided not to adopt those comments.

Comment: Several commenters suggested that we add additional disorders to the listings, including myasthenia gravis, multiple sclerosis, colon cancer, chronic fatigue syndrome, and fibromyalgia.

Response: We have not added the specific disorders suggested by the commenters. In some instances the disorders are already included in our rules:

- Multiple sclerosis, listing 11.09 (neurological body system),
- Myasthenia gravis, listing 11.12 (neurological body system), and
- Stage IV colon cancer, listing 13.18 (malignant neoplastic diseases body system).

You can see all of our listings at: http://www.socialsecurity.gov/OP_Home/cfr20/404/404-ap10.htm and <http://www.socialsecurity.gov/disability/professionals/bluebook/index.htm>.

In other instances, such as fibromyalgia and chronic fatigue syndrome, we did not add the suggested disorders. Although we recognize fibromyalgia and chronic fatigue syndrome as medically determinable impairments, we do not list them, in part because there is not sufficient agreement in the medical community about the nature of these impairments. However, we may find that fibromyalgia and chronic fatigue syndrome medically equal a listing or that they are disabling at a later step of the sequential evaluation process for adults or children. See, for example, Social Security Ruling (SSR) 99–2p, *Titles II and XVI: Evaluating Cases Involving Chronic Fatigue Syndrome (CFS)*, 64 FR 23380 (1999), available at http://www.socialsecurity.gov/OP_Home/rulings/di/01/SSR99-02-di-01.html.

Comment: Several commenters who have multiple immune disorders or family members with immune disorders noted that having multiple immune system disorders can significantly limit an individual's ability to function and to work. One commenter suggested that we include other autoimmune diseases that affect only one organ, such as Hashimoto's or Graves disease, as an additional disease entity to support one of the other listed immune system disorders in a disability claim.

Response: We agree that an individual with multiple immune system disorders

may have significant limitations in the ability to function. However, we did not adopt this comment because we believe that the new functional criteria in each of the final listings will help individuals like the commenters and their family members without additional changes to the listings.

Other Comments

Comment: One commenter addressed our proposal to change the requirement throughout the listings in this body system that an individual have all four of the constitutional symptoms and signs to a requirement for only two of the constitutional symptoms and signs. The commenter noted that fatigue and malaise are both symptoms, and therefore, that an individual could meet this requirement of several of the immune system disorders listings with two symptoms. The commenter also indicated that these symptoms are "exceedingly common" in the general population and said that they are poor discriminators of severity. Therefore, the commenter suggested that we consider fatigue and malaise as one criterion, that is, fatigue/malaise, rather than two separate criteria.

Response: We did not adopt this comment. As we define them in final 14.00C2 and 114.00C2, the symptoms of fatigue and malaise are quite severe and not at all common in the general population. As we indicated in the preamble to the NPRM, we proposed to add these definitions "in response to the many comments we received [on the ANPRM and in the outreach meetings] that indicated that the fatigue and malaise that people who have immune system disorders experience can be very limiting." (71 FR at 44435) In discussing the proposed functional criteria, we also reported that "[a] number of people indicated that the fatigue associated with these disorders was not merely a feeling of tiredness but a more profound and debilitating experience." (71 FR at 44440) This is consistent with information we received from medical specialists in immune system disorders at the outreach meetings and our own review of the medical literature. (See 71 FR at 44448 for a list of the medical references we consulted when we were preparing the proposed rules.) Moreover, the presence of two of the constitutional symptoms and signs is only one criterion in the listings. To meet any of the listings that include this criterion, the individual must also have an established immune system disorder and involvement of at least two organs or body systems. As we explained in the preamble to the NPRM, we proposed to revise the requirement for all four

constitutional symptoms and signs to "at least two" of the constitutional symptoms or signs:

because we believe that the requirement in the current listing is too severe. We believe that any individual with an autoimmune disorder involving two or more organs/body systems with one organ/body system involved to at least a moderate level of severity and who has at least two of the constitutional symptoms and signs in these listings will have an impairment that precludes any gainful activity.

(71 FR at 44442)

Comment: One commenter noted that multiple listings (for example, proposed listings 14.02B, 14.03B, and 14.06B) used the phrase "without the requisite findings in A." The commenter thought that the phrase was unclear, and that it was not clear when this listing criterion would apply. For example, the commenter asked whether this meant in proposed listing 14.02B that the individual had involvement of only one organ or that there was involvement of two organs but neither to a "moderate" degree.

Response: We adopted the comment by deleting the phrase "but without the requisite findings in" from the proposed listings that included that phrase, except in listings 14.08K and 114.08L. Because of their structure, some proposed listings referred only to paragraph A, while others referred to additional paragraphs. For example, proposed listing 14.04D included the phrase "but without the requisite findings in A, B, or C." We removed all of these references. We also made conforming editorial changes to the first sentence in final 14.00I1 and 114.00I1.

In considering the comment, we realized that the phrase was unnecessary and that deleting it would not change our intent. For example, an individual's SLE meets final listing 14.02A if there is involvement of at least two organs/body systems with one of the organs/body systems involved to at least a moderate level of severity and with at least two of the constitutional symptoms and signs. An individual's SLE meets listing 14.02B if it causes repeated manifestations of SLE, at least two of the constitutional symptoms and signs, and a "marked" limitation in one of the listed areas of functioning. There is no need for a reference to listing 14.02A in listing 14.02B.

The same can be said about final listings 14.08K and 114.08L. However, we decided to keep the phrase in those listings because it has been in the prior versions of those listings for many years, is clear in the context of those listings, and is followed by parenthetical

examples that we do not want to remove.

We also realized that related language we proposed in the listings was unclear in other ways. The phrase "Repeated manifestations of [the listed immune disorder] * * * resulting in at least two of the constitutional symptoms or signs" could have been misinterpreted. It could have been read to mean that we would need evidence demonstrating that the constitutional symptoms or signs were the result of the *manifestations* of the immune system disorder, not the immune system disorder itself. We revised the language to clarify our intent, which is that the constitutional symptoms and signs can be the result of either the immune disorder itself or any of its manifestations. Also, some of the listings, for example, proposed listing 14.02A2 (which was referenced by proposed listing 14.02B), used the unclear phrase "At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss." (Emphasis added.) This could have been misinterpreted to mean that there are other constitutional symptoms and signs. Therefore, we revised all of the listings that included this statement to say "At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss)." For consistency with this change, we also revised our definition of "constitutional symptoms or signs" in proposed 14.00C and 114.00C to explain that the fatigue must be "severe fatigue" for purposes of these listings. This is not a substantive change in the proposed rules because in fact all of the proposed listings required "severe fatigue" when they referred to constitutional symptoms or signs.

Comment: One commenter suggested that we specify in these rules which tests we will not purchase, such as angiography and tissue biopsy. The commenter noted that this would also make the immune system disorders listings consistent with the most recent revision of the cardiovascular system listings, which we issued in early 2006.

Response: We adopted the comment. The new guidance is in final 14.00D2b, 14.00D4b, and 14.00F1 and the corresponding childhood sections. We considered adding the same language in final 14.00F3 and 114.00F3 but decided not to because there are some manifestations for which we may purchase tests, such as routine types of blood tests.

Comment: One commenter noted that the heading in proposed 14.00D was different than the headings in proposed 14.00E and 14.00F. The commenter

suggested revisions to the headings of 14.00D, 14.00E, and 14.00F that would make them consistent with each other.

Response: We adopted the comment. The commenter recommended that we change the headings to declarative statements, but we retained the question form to be consistent with most of the other headings in this body system. Otherwise, we used the same language the commenter recommended.

Comment: One commenter suggested that we use simple terms in these rules.

Response: We have simplified the language as much as we can given the complexity of these disorders. However, to provide useful adjudicative guidance, our rules need to reference the technical terms that are used in medical records and severity terms we use in our regulations. When appropriate, we have provided definitions of these terms in final 14.00C and 114.00C and elsewhere in these final rules.

Comment: One commenter questioned how we can give benefits to some and deny others when an autoimmune disease is a disabling disease with no hope of getting better.

Response: While we understand the concern of the commenter, we also recognize that many individuals who are diagnosed with autoimmune disorders lead reasonably normal lives, including regular employment. We can pay benefits only to individuals who are under a disability as defined in the Act and in our regulations. The issue in a disability determination under the listings is whether the individual has an impairment that prevents him or her from engaging in any gainful activity (or in a child, that causes "marked and severe functional limitations"), and that can be expected to result in death or which has lasted or can be expected to last for a continuous period of not less than 12 months. If the impairment does not meet or medically equal the listings, we may still find that the impairment is disabling based on an assessment of the individual's residual functional capacity (or the child's ability to function).

Comment: One commenter suggested that it will be essential to provide a training program for all workers who are involved in the disability process, particularly those who make the initial determination. The commenter indicated that it will be necessary for adjudicators to understand all of the information in the introductory text and that this will be difficult for them. The commenter also remarked that we should be aware that it will be more burdensome and time-consuming for treating physicians to understand the nuances of these rules and that physicians have less and less time to

deal with extensive reading in order to complete a form or to write letters for their patients' disability claims.

Response: We agree that training on these final rules will be needed. We will conduct training that will provide adjudicators with guidance on applying these listings.

We do not believe the expanded guidance in these final rules imposes additional burdens on treating physicians. It is our responsibility to decide whether individuals meet the criteria of these rules, and the information we need from treating sources so that we can make our decision is no different under these rules than it was before. As we have already explained, we expect that in some cases we will need even less information than we did in the past because of additional medical and functional criteria in these listings that will permit us to allow individuals who should be allowed under the listings instead of at a later step in the sequential evaluation process.

Even the new functional criteria in the listings will not impose a new burden on treating sources. This is because when we ask for information from treating and other medical sources we also ask them for opinions about how their patients' medical conditions limit functioning in case we need to consider residual functional capacity or, for children, functional equivalence. See, for example, §§ 404.1513 and 416.913 of our regulations. We will be able to use the same information treating sources provide for residual functional capacity assessments or determinations about functional equivalence to make our determinations about limitations under the new listings and, in some cases, will need even less information when the functional limitations are clearly as serious as the listings describe.

What is our authority to make rules and set procedures for determining whether a person is disabled under the statutory definition?

Section 205(a) of the Act and, by reference to section 205(a), section 1631(d)(1) provide that:

The Commissioner of Social Security shall have full power and authority to make rules and regulations and to establish procedures, not inconsistent with the provisions of this title, which are necessary or appropriate to carry out such provisions, and shall adopt reasonable and proper rules and regulations to regulate and provide for the nature and extent of the proofs and evidence and the method of taking and furnishing the same in order to establish the right to benefits hereunder.

Regulatory Procedures

Executive Order 12866

We have consulted with the Office of Management and Budget (OMB) and determined that these final rules meet the requirements for a significant regulatory action under Executive Order 12866, as amended. Thus, they were subject to OMB review.

Regulatory Flexibility Act

We certify that these final rules do not have a significant economic impact on a substantial number of small entities because they affect only individuals. Thus, a regulatory flexibility analysis as provided in the Regulatory Flexibility Act, as amended, is not required.

Paperwork Reduction Act

The Paperwork Reduction Act (PRA) of 1995 says that no persons are required to respond to a collection of information unless it displays a valid OMB control number. In accordance with the PRA, SSA is providing notice that OMB has approved the information collection requirements contained in sections 14.00B, 14.00D, 14.00E, 14.00F, 114.00B, 114.00D, 114.00E, 114.00F, 114.08 and 114.09 of these final rules. The OMB Control Number for this collection is 0960–0642, expiring March 31, 2008.

(Catalog of Federal Domestic Assistance Program Nos. 96.001, Social Security-Disability Insurance; 96.002, Social Security-Retirement Insurance; 96.004, Social Security-Survivors Insurance; and 96.006, Supplemental Security Income)

List of Subjects in 20 CFR Part 404

Administrative practice and procedure, Blind, Disability benefits, Old-Age, Survivors, and Disability Insurance, Reporting and recordkeeping requirements, Social Security.

Michael J. Astrue,

Commissioner of Social Security.

■ For the reasons set out in the preamble, we are amending subpart P of part 404 of chapter III of title 20 of the Code of Federal Regulations as set forth below:

PART 404—FEDERAL OLD-AGE, SURVIVORS AND DISABILITY INSURANCE (1950–)

■ 1. The authority citation for subpart P of part 404 continues to read as follows:

Authority: Secs. 202, 205(a), (b), and (d)–(h), 216(i), 221(a) and (i), 222(c), 223, 225, and 702(a)(5) of the Social Security Act (42 U.S.C. 402, 405(a), (b), and (d)–(h), 416(i), 421(a) and (i), 422(c), 423, 425, and 902(a)(5)); sec. 211(b), Pub. L. 104–193, 110 Stat. 2105, 2189; sec. 202, Pub. L. 108–203, 118 Stat. 509 (42 U.S.C. 902 note).

Appendix 1 to Subpart P of Part 404— [Amended]

- 2. Appendix 1 to subpart P of Part 404 is amended as follows:
 - a. Revise the body system name and the expiration date in item 15 of the introductory text before part A of appendix 1.
 - b. Amend the table of contents for part A of appendix 1 by revising the body system name for section 14.00.
 - c. Revise the second sentence of section 1.00B1 of part A of appendix 1.
 - d. Revise the fourth sentence of section 1.00L of part A of appendix 1.
 - e. Revise section 8.00D3 of part A of appendix 1.
 - f. Revise the second sentence of section 13.00A of part A of appendix 1.
 - g. Revise section 14.00 of part A of appendix 1.
 - h. Amend the table of contents for part B of appendix 1 by revising the body system name for section 14.00.
 - i. Revise the second sentence of section 101.00B1 of part B of appendix 1.
 - j. Revise the fourth sentence of section 101.00L of part B of appendix 1.
 - k. Revise section 108.00D3 of part B of appendix 1.
 - l. Revise the second sentence of section 113.00A of part B of appendix 1.
 - m. Revise section 114.00 of part B of appendix 1.

The revised text is set forth as follows:

Appendix 1 to Subpart P of Part 404— Listing of Impairments

* * * * *
 15. Immune System Disorders (14.00 and 114.00): June 16, 2016.

Part A

* * * * *

14.00 Immune System Disorders.

1.00 MUSCULOSKELETAL SYSTEM

* * * * *

B. * * *

1 * * * The provisions of 1.02 and 1.03 notwithstanding, inflammatory arthritis is evaluated under 14.09 (see 14.00D6). * * *

L. * * * 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. * * *

* * * * *

8.00 SKIN DISORDERS

* * * * *

D. * * *

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.

* * * * *

13.00 MALIGNANT NEOPLASTIC DISEASES

A. * * * We use the criteria in 14.08E to evaluate carcinoma of the cervix, Kaposi’s sarcoma, lymphoma, and squamous cell carcinoma of the anal canal and anal margin if you also have HIV infection.

* * * * *

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 (creatinine 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 (spondyloarthritis).* 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 (iritidocyclitis, 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 (Felt'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/mm³ (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 fundoscopic 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.

1. 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 nontyphoid; 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

*	*	*	*	*
	114.00			Immune System Disorders.
*	*	*	*	*

101.00				MUSCULOSKELETAL SYSTEM
*	*	*	*	*

B. * * *

1. * * * The provisions of 101.02 and 101.03 notwithstanding, inflammatory arthritis is evaluated under 114.09 (see 114.00D6). * * *

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L. * * * 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. * * *

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108.00 SKIN DISORDERS
* * * * *

D. * * *

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.

* * * * *

113.00 MALIGNANT NEOPLASTIC DISEASES

A. * * * We use the criteria in 114.08E to evaluate carcinoma of the cervix, Kaposi's sarcoma, lymphoma, and squamous cell carcinoma of the anal canal and anal margin if you also have HIV infection.

* * * * *

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) 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 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/mm³ 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/mm³ 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/mm³ (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 funduscopy 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.

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 nontyphoid; 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. Growth disturbance, with:

1. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from an established growth curve (on standard growth charts) that persists for 2 months or longer; or

2. 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 an established growth curve (on standard growth charts) that persists for 2 months or longer; or

3. Involuntary weight loss of 10 percent or more of baseline (computed based on pounds, kilograms, or body mass index (BMI)) that persists for 2 months 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. 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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