SOCIAL SECURITY ADMINISTRATION

20 CFR Part 404

[Docket No. SSA–2019–0013]

RIN 0960–AI43

Revised Medical Criteria for Evaluating Cardiovascular Disorders

AGENCY: Social Security Administration.

ACTION: Notice of proposed rulemaking (NPRM).

SUMMARY: We propose to revise some of the criteria in the Listing of Impairments (listings) that we use to evaluate claims involving cardiovascular disorders in adults and children under titles II and XVI of the Social Security Act (Act). The proposed revisions reflect advances in medical knowledge, our adjudicative experience, and comments we received from experts and the public in response to an advance notice of proposed rulemaking (ANPRM), and at an outreach policy conference.

DATES: To ensure that your comments are considered, we must receive them by no later than August 29, 2022.

ADDRESSES: You may submit comments by any one of three methods—internet, fax, or mail. Do not submit the same comments multiple times or by more than one method. Regardless of which method you choose, please state that your comments refer to Docket No. SSA–2019–0013, so that we may associate your comments with the correct regulation.

Caution: You should be careful to include in your comments only information that you wish to make publicly available. We strongly urge you not to include in your comments any personal information, such as Social Security numbers or medical information.

1. *Internet:* We strongly recommend that you submit your comments via the internet. Please visit the Federal eRulemaking portal at *http://www.regulations.gov.* Use the *search* function to find docket number SSA–2019–0013. The system will issue a tracking number to confirm your submission. You will not be able to view your comment immediately because we must post each comment manually. It may take up to a week for your comment to be viewable.

2. *Fax:* Fax comments to (410) 966–2830.

3. *Mail:* Address your comments to the Office of Regulations and Reports Clearance, Social Security Administration, 3100 West High Rise, 6401 Security Boulevard, Baltimore, Maryland 21235–6401.

Comments are available for public viewing on the Federal eRulemaking portal at *http://www.regulations.gov* or in person, during regular business hours, by arranging with the contact person identified below.

FOR FURTHER INFORMATION CONTACT: Michael J. Goldstein, Office of Disability Policy, Social Security Administration, 6401 Security Boulevard, Baltimore, Maryland 21235–6401, (410) 965–1020. 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 site, Social Security Online, at *http://www.socialsecurity.gov*.

SUPPLEMENTARY INFORMATION:

Background

For adults, the listings describe, for each of the major body systems, impairments that we consider to be severe enough to prevent an individual from doing any gainful activity regardless of his or her age, education, or work experience.[1] For children, the listings describe impairments we consider severe enough to cause marked and severe functional limitations.[2] We use the listings at step 3 of the sequential evaluation process to identify claims in which the individual is clearly disabled under our rules.[3] However, we do not deny any claim solely because a person's medical impairment(s) does not satisfy the criteria of a listing.

Why are we proposing to revise the listings for cardiovascular disorders?

We last published final rules that comprehensively revised the cardiovascular disorders listings on January 13, 2006, and the rules became effective on April 13, 2006.[4] We are now proposing targeted revisions to the cardiovascular disorders listings, as previously mentioned, to reflect advances in medical knowledge, our adjudicative experience, and comments we received from experts and the public in response to an ANPRM, and at an outreach policy conference.

How did we develop this proposed rule?

In developing this proposed rule:

- We published an ANPRM for cardiovascular disorders in the **Federal Register** on April 16, 2008.[5] We invited the public to send us written comments and suggestions about whether and how we should revise the cardiovascular disorders listings. We received five comments on the ANPRM. The commenters made several suggestions that we incorporated into the proposals, such as consideration for people with a single ventricle; clarifying in proposed 4.00D(4)(c)(ii) (*How do we evaluate CHF using 4.02?*) and 4.02B1 (*Chronic heart failure*) that when we evaluate a person's ability to perform activities of daily living, we will also consider the person's ability to perform them effectively;[6] and providing more examples of skin examination findings that might accompany venous insufficiency. Two commenters asked us to base our proposals on the American College of Cardiology/American Heart Association (ACC/AHA) clinical practice guidelines and one suggested we review and incorporate the American Society of Echocardiography (ASE) guidelines. As a result, we tasked a committee of medical experts with reviewing and analyzing the ACC/AHA and ASE guidelines to inform the recommendations in the Institute of Medicine (IOM)[7] report titled, ''*Cardiovascular Disability: Updating the Social Security Listings.*''[8] This report, which is discussed in more detail below, informed some of the proposed changes in this NPRM.
- On September 24 and 25, 2008, we hosted a policy conference titled ''*Cardiovascular Disorders in the Disability Programs*'' in Baltimore, Maryland. At this conference, we received public comments and suggestions from physicians and advocacy groups for updating and revising our criteria for evaluating cardiovascular disorders. Physicians and advocacy groups specifically discussed the evaluation of chronic heart failure, ischemic heart disease, peripheral artery disease, and chronic

[1] 20 CFR 404.1525(a) and 20 CFR 416.925(a).

[2] 20 CFR 416.925(a).

[3] 20 CFR 404.1520, 20 CFR 416.920, and 20 CFR 416.924.

[4] 71 FR 2312 (2006).

[5] 73 FR 20564 (2008).

[6] In current listing 4.02B1 (*Chronic heart failure*), we require persistent symptoms of heart failure ''which very seriously limit the person's ability to independently initiate, sustain or complete activities of daily living.'' Consistent with the commenter's suggestion and how we assess functional limitations in the adult mental disorders listings (*12.00*), in proposed 4.02B1, we require ''a very serious limitation in the ability to perform activities of daily living independently, appropriately, effectively, and on a sustained basis.''

[7] On April 28, 2015, the membership of the National Academy of Sciences voted to change the name of the IOM to the National Academy of Medicine. At that time, reports and studies of the IOM continued as activities of the Health and Medicine Division, a program unit operating under the direction of the National Academies of Sciences, Engineering, and Medicine.

[8] Institute of Medicine (IOM). (2010). *Cardiovascular Disability: Updating the Social Security Listings.* Washington, DC: The National Academies Press.

venous insufficiency. Participants made several suggestions that we researched and incorporated into the proposals, such as distinguishing cardiovascular disorders from pulmonary disorders by using biomarkers such as B-type natriuretic peptide (BNP).

• In 2009, we commissioned a study by an ad hoc committee of medical experts appointed by the Institute of Medicine (IOM). The committee: (1) conducted a comprehensive review of the relevant research literature and current professional practice guidelines developed jointly by the ACC/AHA; (2) assessed the current criteria in light of current research knowledge and evidence-based medical practice; and (3) produced a report with specific recommendations for revision of the criteria based on evidence and professional judgment. The committee provided its recommendations in a 2010 report titled, ''Cardiovascular Disability: Updating the Social Security Listings.'' [9] We recently sought guidance from our cardiologists and other medical experts, reviewed disability claims involving cardiovascular disorders, and reviewed current research to ensure the IOM recommendations are still relevant.

Recommendations we received from the IOM report, responses to the ANPRM, and the ''Cardiovascular Disorders in the Disability Programs'' policy conference informed the proposed changes in this NPRM. As with the IOM report, we have conducted independent medical research, consulted with agency cardiologists, and reviewed disability claims involving cardiovascular disorders to ensure the accuracy and relevance of these stated resources. In developing this proposed rule, we also considered information from several other sources, including:

• Medical experts in cardiology from SSA's Office of Medical Assistance [10] who assist in the development and evaluation of policy and whom we regularly consulted with in drafting these proposals;

• Advocacy groups for people with cardiovascular disorders and individuals with cardiovascular disorders and their families who submitted comments on the ANPRM or participated in the 2008 ''Cardiovascular Disorders in the Disability Program'' policy conference;

• Individuals who make and review disability determinations and decisions for us in State agencies; in our Office of Hearings Operations; and in our Office of Analytics, Review, and Oversight; and

• The published sources of medical literature and research we list in the references section at the end of this preamble.

What revisions are we proposing for cardiovascular disorders?

We propose to:

• Change the name of the body system from ''Cardiovascular System'' to ''Cardiovascular Disorders'' to be consistent with the nomenclature of all body systems in our listings;

• Reorganize and revise the introductory text (section 4.00 for adults and 104.00 for children) to provide guidance for using the revised criteria in the listings;

• Revise the adult and childhood listings for chronic heart failure (4.02 and 104.02), recurrent arrhythmias (4.05 and 104.05), symptomatic congenital heart disease (4.06) and congenital heart disease (104.06), and heart transplant (4.09 and 104.09);

• Revise the adult listings for ischemic heart disease (IHD) (4.04), chronic venous insufficiency (4.11), and peripheral arterial disease (4.12);

• Add adult listings for aortic valvular disease (4.07) and cardiomyopathy (4.08);

• Add adult and childhood listings for cardiac allograft vasculopathy (4.16 and 104.16);

• Remove childhood listing for rheumatic heart disease (104.13) and reserve listing number 104.13; and

• Make minor editorial revisions, including changes to conform to revised rules for evaluating medical evidence,[11] to the introductory text and to the listings for clarity.

Proposed Changes to the Adult Cardiovascular Disorders Introductory Text

The following table shows the heading of the current and proposed sections of the adult introductory text for cardiovascular disorders:

INTRODUCTORY TEXT 4.00

Current Sections of the Adult Introductory Text for Cardiovascular System.	Proposed Sections of the Adult Introductory Text for Cardiovascular Disorders.
4.00 Cardiovascular System	4.00 Cardiovascular Disorders.
A. General	A. How do we define cardiovascular disorders and cardiovascular terms?
B. Documenting Cardiovascular Impairment	B. What documentation do we need to evaluate cardiovascular disorders?
C. Using Cardiovascular Test Results	C. How do we use cardiovascular test results?
D. Evaluating Chronic Heart Failure	D. How do we evaluate chronic heart failure?
E. Evaluating Ischemic Heart Disease	E. How do we evaluate ischemic heart disease?
F. Evaluating Arrhythmias	F. How do we evaluate arrhythmias?
G. Evaluating Peripheral Vascular Disease	G. How do we evaluate peripheral vascular disease?
H. Evaluating Other Cardiovascular Impairments	H. How do we evaluate congenital heart disease?
I. Other Evaluation Issues	I. How do we evaluate other cardiovascular disorders?
	J. How do we evaluate issues that affect the cardiovascular system?
	K. How do we evaluate cardiovascular disorders that do not meet one of these listings?

[9] IOM. (2010).

[10] SSA's Office of Medical Assistance (OMA) provides medical and analytical support to ensure accurate and consistent disability policy and procedure application. In addition to a full complement of subject matter experts who are permanent SSA staff, OMA contracts with medical and psychological consultants to provide medical expertise in the development and evaluation of policy.

[11] 82 FR 5844 (2017).

Proposed 4.00—Introductory Text to the Adult Cardiovascular Disorders Listings

The following is a detailed description of the primary changes we are proposing to the introductory text. In addition to the changes we describe below, we are proposing minor changes to the introductory text to clarify how we use the proposed listings to evaluate cardiovascular disorders, changes to be consistent with current medical terminology, the language we use in other body system listings, and the revised rules for evaluating medical evidence.[12] We repeat much of the introductory text of proposed 4.00 in the introductory text of proposed 104.00 (the introductory text to the childhood cardiovascular disorders listings), making distinctions where needed. This is necessary because the same basic criteria for evaluating cardiovascular disorders apply to both adults and children.

Proposed 4.00A—How do we define cardiovascular disorders and cardiovascular terms?

To improve clarity and promote consistent understanding of the terms we use in these listings, we propose:

- In 4.00A3b (*Persistent*), to clarify that ''exceptions'' means brief periods when the required finding(s) is greatly reduced or gone. These periods are so brief or inconsequential, the required finding(s) remains a factor in the person's condition;
- In 4.00A3c (*Recurrent*), to clarify in our definition of ''recurrent'' that the term ''improvement of sufficient duration'' means the finding is greatly reduced (for example, treatment reduced a grade 3 chronic venous insufficiency (CVI) skin ulcer to a grade 1 CVI skin ulcer) or not present for long enough that the required finding(s) is no longer a factor in the person's condition.
- In 4.00A3f (*Uncontrolled*) we would remove the definition for the term ''uncontrolled'' because we propose to eliminate the term as a descriptor for recurrent episodes of cardiac syncope. The definition of ''uncontrolled'' was redundant after describing reoccurring episodes of cardiac syncope despite treatment.

Proposed 4.00C—How do we use cardiovascular test results?

We propose:

- In 4.00C8d(iv) (*When will we not purchase an exercise test or wait before we purchase an exercise test?*), to include the procedure percutaneous coronary intervention (PCI) to be consistent with medical advancements that indicate a PCI is a nonsurgical procedure to improve blood flow to the heart.[13]
- In 4.00C15 (*How do we evaluate cardiac catheterization evidence?*), to remove 4.00C15b and 4.00C15c and create a new 4.00C15b to simplify our explanation of cardiac catheterization reports and to include information about fractional flow reserve (FFR), which we use in the proposed 4.04D1 (*Ischemic heart disease*).

Proposed 4.00D—How do we evaluate chronic heart failure?

We propose:

- In 4.00D1a (*What is chronic heart failure (CHF)?*), to provide a more descriptive definition of ''ejection fraction'' for clarity;
- In 4.00D1b (*What is chronic heart failure (CHF)?*), to explain that high blood levels of the proteins B-type natriuretic peptide (BNP) and N-terminal pro-BNP (NT-pro-BNP) may help identify chronic heart failure as the cause of a person's symptoms (for example, shortness of breath);§ [14] [15] [16] [17]
- In 4.00D2a(i) (*What evidence of CHF do we need?*) and 4.00D2a(iii) (*What evidence of CHF do we need?*), to explain left atrial volume index (LAVi) and how it is calculated. LAVi is a new measurement used in criterion A2 of proposed listing 4.02 (*Chronic heart failure*) to provide a more precise representation of increased left atrial pressure;
- In 4.00D4c (*How do we evaluate CHF using 4.02?*),to describe in more detail the two-part process we use in criteria B1 of proposed listing 4.02 (*Chronic heart failure*) to evaluate chronic heart failure if a person cannot perform an exercise tolerance test (ETT);[18]; and
- Add 4.00D4e (*How do we evaluate CHF treated with a mechanical circulatory support device?*) to explain mechanical circulatory support devices (MCSD) and clarify how we would evaluate individuals treated for heart failure with a MCSD.[19] We propose to evaluate MCSDs under proposed 4.02D1 (*Chronic heart failure*)to account for cardiac bridge treatment which we currently evaluate under listing 4.09 (*Heart transplantation*).

Proposed 4.00E—How do we evaluate ischemic heart disease?

We propose:

- In 4.00E9b (*How do we evaluate IHD using 4.04?*), to consolidate and revise the guidance that is currently in 4.00E9b, c, d, and e for ease of reference;
- In 4.00E9b (*How do we evaluate IHD using 4.04?*), to explain that we will use an interpretation of electrocardiogram (ECG) findings by an acceptable medical source (AMS) [20] if the interpretation concludes that the ECG findings are positive for IHD. Interpreting ECG results requires a systematic review and analysis of several components, including relevant clinical details and raw data. Relying on an interpretation by an AMS is consistent with our rules for establishing a medically determinable impairment (MDI) and will ensure the accuracy of adjudication for claims involving IHD because interpretation of ECG findings requires medical judgment; [21]
- In 4.00E9c (*How do we evaluate IHD using 4.04?*), to clarify that revascularizations that result from unplanned hospitalizations must be an emergency and unplanned; [22]
- To redesignate current 4.00E9f, g, and h (*How do we evaluate IHD using 4.04?*) as proposed new sections 4.00E9c, d, and f, respectively; and
- In 4.00E9e (*How do we evaluate IHD using 4.04?*), to add a definition for

[12] Id.

[13] IOM. (2010), 109.

[14] Cohen-Solal, A., Laribi, S., Ishihara, S., Vergaro, G., Baudet, M., Logeart, D., . . . Seronde, M.-F. (2015). Prognostic markers of acute decompensated heart failure: The emerging roles of cardiac biomarkers and prognostic scores. *Archives of Cardiovascular Disease,* 108(1), 64–74. doi:10.1016/j.acvd.2014.10.002.

[15] Desai, A.S. (2013). Are serial BNP measurements useful in heart failure management? Serial natriuretic peptide measurements are not useful in heart failure management: The art of medicine remains long. Circulation, 127(4), 509–516. doi:10.1161/CIRCULATIONAHA.112.120493.

[16] Patterson, C.C., Blankenberg, S., Ben-Shlomo, Y., Heslop, L., Bayer, A., Lowe, G., . . . Yarnell, J. (2015). Which biomarkers are predictive specifically for cardiovascular or for non-cardiovascular mortality in men? Evidence from the Caerphilly Prospective Study (CaPS). International Journal of Cardiology, 201, 113–118. doi:10.1016/j.ijcard.2015.07.106.

[17] Uszko-Lencer, N.H., Frankenstein, L., Spruit, M.A., Maeder, M.T., Gutmann, M., Muzzarelli, S., . . . Brunner-La Rocca, H.-P. (2017). Predicting hospitalization and mortality in patients with heart failure: The BARDICHE-index. International Journal of Cardiology, 227, 901–907. doi:10.1016/j.ijcard.2016.11.122.

[18] We define an exercise tolerance test at 4.00C3b (*What are exercise tests and what are they used for?*) as ''a sign-or symptom-limited test in which you exercise while connected to an ECG until you develop a sign or symptom that indicates that you have exercised as much as is considered safe for you.'' This is an existing definition coming from existing regulation.

[19] Ciarka, A., Edwards, L., Nilsson, J., Stehlik, J., & Lund, L.H. (2017). Trends in the use of mechanical circulatory support as a bridge to heart transplantation across different age groups. *International Journal of Cardiology, 231,* 225–227. doi:10.1016/j.ijcard.2016.10.049.

[20] 20 CFR 404.1502(a) and 416.902(a).

[21] 20 CFR 404.1521 and 416.921.

[22] As we explain in 4.00E9c, revascularization means angioplasty (with or without stent placement) or bypass surgery.

the term ''fractional flow reserve (FFR),'' which is an objective measure of flow access across an obstruction that we use in criterion D1 of proposed listing 4.04 (*Ischemic heart disease*).[23]

Proposed 4.00G—How do we evaluate peripheral vascular disease?

In 4.00G6 (*Are there any other studies that are helpful in evaluating PAD?*), as IOM advised, we propose to provide more information about imaging and other tests used to diagnose peripheral arterial disease (PAD) to clearly convey our intent of the listing, which is to tie PAD to mobility.[24]

Proposed 4.00H—How do we evaluate congenital heart disease?

We propose:

- To redesignate and rename some paragraphs for ease of reference;
- In 4.00H (*How do we evaluate congenital heart disease?*), to add a definition for the term ''single ventricle,'' and include more detailed guidance on how we evaluate congenital heart disease as recommended by the IOM; and
- To add 4.00H1 (*What is congenital heart disease?*), 4.00H2 (*What is Eisenmenger syndrome?*), 4.00H3 (*What is single ventricle?*), and 4.004H4 (*How do we evaluate conditions associated with congenital heart disease?*).

Proposed 4.00I—How do we evaluate other cardiovascular disorders?

We propose:

- To rename and rearrange the content of 4.00I (*How do we evaluate other cardiovascular disorders?*), including redesignating some paragraphs, for ease of reference;
- In 4.00I2 (*What is cardiomyopathy and how will we evaluate it?*), to explain how we would evaluate cardiomyopathy under proposed new 4.08 (*Cardiomyopathy*);
- In 4.00I3 (*How do we evaluate valvular heart disease?*), to explain that we would evaluate valvular heart disease under the proposed new 4.07 (*Aortic valvular disease*);
- In 4.00I4 (*What do we consider when we evaluate heart transplant recipients?*), to explain that we would evaluate cardiac allograft vasculopathy under proposed new 4.16 (*Cardiac allograft vasculopathy*); and
- To add 4.00I5 (*What is cardiac allograft vasculopathy and how do we evaluate it?*), to explain proposed new 4.16 (*Cardiac allograft vasculopathy*).

Proposed 4.00J—How do we evaluate issues that affect the cardiovascular system?

We propose:

- To redesignate and rename some paragraphs for ease of reference;
- In 4.00J1 (*How do we consider the effects of obesity when we evaluate your cardiovascular disorder?*), to simplify and refocus our discussion of how we consider the effects of obesity more specifically on cardiovascular disorders;
- To add 4.00J3 (*How do we consider hospitalizations?*) to explain how we would evaluate hospitalizations for repeated exacerbations and complications of cardiovascular disorders under proposed 4.02B3 (*Chronic heart failure*), 4.04E (*Ischemic heart disease*), 4.06E (*Congenital heart disease*), and 4.08D (*Cardiomyopathy*).

Proposed 4.00K—How do we evaluate cardiovascular disorders that do not meet one of these listings?

- We propose to rename and redesignate 4.00I3 (*How do we evaluate impairments that do not meet one of the cardiovascular listings?*) as 4.00K and redesignate 4.00I3a and 4.00I3b as 4.00K1 and 4.00K2 for ease of reference.

Proposed Changes to the Adult Cardiovascular Disorders Listings

The following table shows the heading of the current and proposed sections of the adult listings for cardiovascular disorders:

ADULT CARDIOVASCULAR DISORDERS LISTINGS

Current	Proposed
4.02 Chronic heart failure	4.02 Chronic heart failure.
4.04 Ischemic heart disease	4.03 [Reserved].
4.05 Recurrent arrhythmias	4.04 Ischemic heart disease.
4.06 Symptomatic congenital heart disease	4.05 Recurrent arrhythmias.
4.09 Heart transplant	4.06 Congenital heart disease.
4.10 Aneurysm of aorta or major branches	4.07 Aortic valvular disease.
4.11 Chronic venous insufficiency	4.08 Cardiomyopathy.
4.12 Peripheral arterial disease	4.09 Heart transplantation.
	4.10 Dissecting aneurysm of the aorta or major branches.
	4.11 Chronic venous insufficiency.
	4.12 Peripheral arterial disease.
	4.13 [Reserved].
	4.14 [Reserved].
	4.15 [Reserved].
	4.16 Cardiac allograft vasculopathy.

[23] Fearon, W.F. (2014). Percutaneous coronary intervention should be guided by fractional flow reserve measurement. *Circulation, 129*(18), 1860–1870. doi:10.1161/CIRCULATIONAHA.113.004300.

[24] IOM. (2010), 151.

The following table shows our proposed changes to the adult cardiovascular disorders listings criteria that involve changes to healthcare utilization and condition/episode requirements, the rationale for each change, and supporting resource. Following this table, we discuss all of our proposed changes to the adult cardiovascular disorders listings in more detail.

ADULT CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS

Current listing criterion	Proposed listing criterion	Rationale	Resources
Listing 4.02 Chronic heart failure			
4.02 A1—A. Medically documented presence of one of the following: 1. Systolic failure (see 4.00D1a(i)), with left ventricular end diastolic dimensions greater than 6.0 cm or ejection fraction of 30 percent or less during a period of stability (not during an episode of acute heart failure); or.	A. Medically documented presence of one of the following: 1. Systolic failure documented by appropriate medically acceptable imaging during a period of stability (not during an episode of exacerbation of heart failure), with left ventricular end diastolic dimension equal to or greater than 7.0 cm; or ejection fraction of 30 percent or less during a period of stability (not during an episode of acute heart failure).	Proposed criterion 4.02A1 requires an increased left ventricular end diastolic dimension (LVEDD) equal to or greater than 7.0 centimeters (cm) instead of the current criterion of an LVEDD greater than 6.0 cm. We followed the IOM's recommendation in determining that an increased LVEDD of greater than 6.0 cm but less than 7.0 cm indicates only a moderately enlarged heart, and an increased LVEDD of at least 7.0 cm more clearly establishes an enlarged heart with signs and symptoms indicating listing-level heart failure.	IOM. (2010), 88, 89.
4.02B2—Three or more separate episodes of acute congestive heart failure within a consecutive 12-month period (see 4.00A3e), with evidence of fluid retention (see 4.00D2b(ii)) from clinical and imaging assessments at the time of the episodes, requiring acute extended physician intervention such as hospitalization or emergency room treatment for 12 hours or more, separated by periods of stabilization (see 4.00D4c);	B.3. Exacerbations or complications of chronic heart failure (see 4.00D1b) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).	We propose to remove current 4.02B2 "three or more separate episodes of acute congestive heart failure" because we would evaluate these episodes under proposed 4.02B3. As recommended by the IOM, proposed 4.02B3 would evaluate exacerbations or complications of CHF, requiring three hospitalizations within a consecutive 12-month period and at least 30 days apart. An impairment resulting in exacerbations or complications that require this many hospitalizations in 12 months is a very severe impairment. We would require these hospitalizations to be at least 30 days apart to ensure we are evaluating separate episodes of exacerbations or complications.	IOM. (2010), 89, 91.
No current listing criteria	C. Heart failure with left ventricular ejection fraction of 20 percent or less while on a regimen of prescribed therapy, on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e) during a period of stability (not during an episode of exacerbation of heart failure);	The IOM recommended a criterion for chronic heart failure with an EF on a sustained basis of 20 percent or less. An EF of only 20 percent means the heart's pumping action is less than a third of normal, and critically affects a person's ability to perform gainful activity.	IOM. (2010), 84, 89. Desai, R.V., Guichard, J.L., Mujib, M., Ahmed, M.I., Feller, M.A., Fonarow, G.C., . . . Ahmed, A. (2013). Reduced right ventricular ejection fraction and increased mortality in chronic systolic heart failure patients receiving beta-blockers: Insights from the BEST trial. International Journal of Cardiology, 163(1), 61–67. doi:10.1016/j.ijcard.2011.05.051. Runge, M.S., Patterson, C., Stouffer, G.A., & Netter, F.H. (2010). Netter's Cardiology (2nd ed.). Philadelphia, PA: Saunders Elsevier.

ADULT CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS—Continued

Current listing criterion	Proposed listing criterion	Rationale	Resources
No current listing criteria	D. One of the following while hospitalized, at home, or both: 1. Mechanical circulatory support device except extracorporeal membrane oxygenation (ECMO) (see 4.00D4e). Consider under a disability for 1 year from the date of implantation; after that, evaluate any residual impairment(s) under the criteria for the affected body system. 2. Continuous intravenous administration of inotropic medication (for example, milrinone) for at least 30 consecutive days. Consider under a disability for 1 year from the date of initiation of the treatment; after that, evaluate any residual impairment(s) under the criteria for the affected body system.	Implanted MCSDs help the heart pump blood and may be used as a ''bridge'' while a person waits for a heart transplant. We currently use our medical equivalence rules to find someone with heart failure and an implanted MCSD disabled under listing 4.09 (Heart transplantation). Adding this new criterion will ensure that people with heart failure treated with MCSD are consistently identified. People who require continuous intravenous administration of inotropic medication (for example, milrinone) have very serious heart failure, and the length of this treatment can be an accurate predictor of impairment severity. Accordingly, proposed 4.02D2 would find people disabled if they require continuous intravenous inotropic medications for 30 or more consecutive days.	Malotte, K., Saguros, A., & Groninger, H. (2018). Continuous cardiac inotropes in patients with end-stage heart failure: An evolving experience. Journal of Pain Symptom Management, 55(1), 159–163. doi:10.1016/j.jpainsymman.2017.09.026. Bistola, V., Arfaras-Melainis, A., Polyzogopoulou, E., Ikonomidis, I., & Parissis, J. (2019). Inotropes in acute heart failure: From guidelines to practical use: Therapeutic options and clinical practice. Cardiac Failure Review, 5(3), 133–139. doi:10.15420/cfr.2019.11.2.
Listing 4.04 Ischemic heart disease			
4.04B—Three separate ischemic episodes, each requiring revascularization or not amenable to revascularization (see 4.00E9f), within a consecutive 12-month period (see 4.00A3e).	4.04C—Documentation of three separate ischemic episodes (see 4.00E9c) requiring unplanned hospitalization (inpatient or observation status) within a consecutive 12-month period (see 4.00A3e).	We would evaluate unplanned hospitalizations under this section to ensure we are only evaluating urgent ischemic episodes. Individuals who have ischemic episodes that result in unplanned hospitalizations may need intensive care, can have long hospital stays, may require multiple procedures, and can be at high risk for post-discharge morbidity and mortality.	Hua, M., Gong, M.N., Brady, J., & Wunsch, H. (2015). Early and late unplanned rehospitalizations for survivors of critical illness. Critical Care Medicine, 43(2), 430–438. doi:10.1097/CCM.0000000000000717.
No current criterion for repeated exacerbations or complications ischemic heart disease, 4.04B (above) only considers episodes requiring revascularization or that are not amenable to revascularization.	4.04E—Exacerbations or complications of ischemic heart disease (see 4.00E2–4.00E7) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00E9c).	An impairment resulting in exacerbations or complications that require three or more hospitalizations in 12 months is a very severe impairment. We would require these hospitalizations to be at least 30 days apart to ensure we are evaluating separate episodes of exacerbations or complications of ischemic heart disease.	Hua, M., Gong, M.N., Brady, J., & Wunsch, H. (2015). Early and late unplanned rehospitalizations for survivors of critical illness. Critical Care Medicine, 43(2), 430–438. doi:10.1097/CCM.0000000000000717.

ADULT CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS—Continued

Current listing criterion	Proposed listing criterion	Rationale	Resources
Listing 4.06 Congenital heart disease			
4.06A—A. Cyanosis at rest, and: 1. Hematocrit of 55 percent or greater; or. 2. Arterial O2 saturation of less than 90 percent in room air or resting arterial PO2 of 60 Torr or less.	A. Chronic hypoxemia, and 1, 2, or 3: 1. Hematocrit of 55 percent or greater on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e); or. 2. Arterial blood gas test measurement obtained at rest while breathing room air, as described in either a or b: a. SaO2 (arterial oxygen saturation) less than or equal to 89 percent; or. b. PO2 or PaO2 (partial pressure of oxygen) less than or equal to 60 mmHg; 3. SpO2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry either at rest, during a 6-minute walk test (6MWT), or after a 6MWT, while breathing room air, less than or equal to 87 percent on three evaluations at least 30 days apart within a consecutive 12-month period (see 4.00A3e).	We propose to revise current 4.06A to require ''hypoxemia'' rather than ''cyanosis or acyanosis''. Cyanosis is a more subjective assessment subject to misinterpretation due to by many factors, including skin complexion. Thus, the term ''hypoxemia'' relates more to the laboratory and pulse oximetry findings than the term ''cyanosis. We would require two hematocrit measurements instead of the current listing's single measurement. Two measurements, at least 90 days apart within a consecutive 12-month period will help ensure the person's hematocrit level is associated with chronic hypoxemia and not the result of a reversible condition. Proposed 4.06A3 would require three SpO2 measurements 30 days apart within a consecutive 12-month period showing hypoxemia. This will document that the condition is chronic and persistent, and the measurements are not related to a reversible condition or an inaccurate reading. We would add a criterion for SpO2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry, including measurements taken while the person is at rest or while doing a six-minute walk test (6MWT). Pulse oximetry measurements are a non-invasive alternative to invasive testing. A person's medical evidence often provides SpO2 findings, and SpO2 measured by pulse oximetry reflects an advance in medical technology that provides another way to establish listing-level severity.	Stout, K.K., Daniels, C.J., Aboulhosn, J.A., Bozkurt, B., Broberg, C.S., Colman, J.M., . . . Van Hare, G.F. (2019). 2018 AHA/ACC Guideline for the management of adults with congenital heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. Journal of the American College of Cardiology, 73(12), e81–e192. doi:10.1016/j.jacc.2018.08.1029. Stack, S.W, . . . & Berger, S.A. (2009). The effects of high hematocrit arterial flow—A phenomenological study of health risk implications. Chemical Engineering Science, 64(22), 4701–4706. doi:10/1016/j.ces.2009.07.017. IOM. (2010), 178. Oster, M.E, . . . & Kochilas, L.K. (2016). Screening for critical congenital heart disease. Clinics in Perinatology, 43(1), 73–80. doi:10.1016/j.clp.2015.11.005. Mechem, C.C. (2014). Pulse oximetry. In P.E. Parsons (Ed.), UpToDate (Jan. 2014). Retrieved from *https://www.uptodate.com/contents/pulse-oximetry*.
No current criteria	4.06E—Exacerbations or complications of congenital heart disease (see 4.00J3) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).	An impairment resulting in exacerbations or complications that require three or more hospitalizations in 12 months is a very severe impairment. We would require these hospitalizations to be at least 30 days apart to ensure we are evaluating separate episodes of exacerbations or complications.	Hua, M., Gong, M.N., Brady, J., & Wunsch, H. (2015). Early and late unplanned rehospitalizations for survivors of critical illness. Critical Care Medicine, 43(2), 430–438. doi:10.1097/CCM.0000000000000717.
Listing 4.08 Cardiomyopathy			
No current listing	4.08D—D. Exacerbations or complications of cardiomyopathy requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).	Consistent with IOM recommendations, we created this new cardiomyopathy listing to specifically address hypertrophic cardiomyopathy, endomyocardial fibrosis, and cardiac amyloidosis AL type, which are more serious types of cardiomyopathy.	IOM. (2010), 80. In addition, SSA has designated endomyocardial fibrosis and cardiac amyloidosis AL type as Compassionate Allowance (CAL) conditions. See Compassionate Allowances Website Home Page (*ssa.gov*).

ADULT CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS—Continued

Current listing criterion	Proposed listing criterion	Rationale	Resources
Listing 4.11 Chronic venous insufficiency			
4.11—Chronic venous insufficiency of a lower extremity with incompetency or obstruction of the deep venous system and one of the following:	4.11—Chronic venous insufficiency (see 4.00G) of a lower extremity with reflux or obstruction of the venous system documented by duplex ultrasound or other appropriate diagnostic technique, with A or B:	As recommended by IOM, we would require confirmation of CVI by duplex ultrasound or other appropriate diagnostic technique. The medical community considers the use of duplex ultrasound to be the best method for detecting reflux or obstruction.	IOM. (2010), 161.
4.11A—A. Extensive brawny edema (see 4.00G3) involving at least two-thirds of the leg between the ankle and knee or the distal one-third of the lower extremity between the ankle and hip.	A. Extensive trophic changes of skin (for example, hyperpigmentation, lipodermatosclerosis, brawny edema) involving at least two-thirds of the leg below the knee, on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e), with both 1 and 2:	We would adopt IOM recommendations and broaden the listing criteria we apply to trophic changes of the skin. For example, in addition to brawny edema, trophic changes evaluated under the proposed listing would include hyperpigmentation and lipodermatosclerosis We would revise the current requirement that these skin changes involve ''at least two-thirds of the leg between the ankle and knee or the distal one-third of the lower extremity between the ankle and hip.'' Instead, we would require extensive skin changes involving at least two-thirds of the leg below the knee, to make the requirement simpler to understand and apply. This revision is consistent with IOM's recommendation to require skin changes below the knee. We would require the skin changes under proposed 4.11A to be consistent with CVI, and we would document the skin changes over a period of at least 90 days to ensure they are chronic.	IOM. (2010), 157–161.
4.11B—Superficial varicosities, stasis dermatitis, and either recurrent ulceration or persistent ulceration that has not healed following at least 3 months of prescribed treatment.	4.11B—Two or more episodes of ulceration that has not healed following at least 6 months of prescribed treatment.	This requirement is more conclusive than the current requirement of 3 months of unsuccessful prescribed treatment as it demonstrates the condition has persisted despite treatment for a longer period of time. The CVI must be unresponsive to compression therapy, because this therapy usually enables people to return to a good level of functioning.	IOM. (2010), 161.

Proposed Listing 4.02—Chronic Heart Failure

We propose to revise the listing criteria for chronic heart failure (CHF) in 4.02A and 4.02B and add new listing criteria 4.02C and 4.02D. Proposed listing-level severity for CHF would be met when the person's CHF satisfies the criteria in 4.02A and 4.02B. Listing-level severity for CHF would also be met when the person's CHF satisfies either proposed 4.02C or 4.02D.

Proposed criterion 4.02A1 requires an increased left ventricular end diastolic dimension (LVEDD) equal to or greater than 7.0 centimeters (cm) instead of the current criterion of an LVEDD greater than 6.0 cm, because an LVEDD less than 5.6 cm is normal. We followed the IOM's recommendation in determining that an increased LVEDD of greater than 6.0 cm but less than 7.0 cm indicates only a moderately enlarged heart, and an increased LVEDD of at least 7.0 cm more clearly establishes an enlarged heart with signs and symptoms indicating listing-level heart failure and is comparable to an ejection fraction (EF) of 30 percent or less.[25]

In proposed 4.02A2, we would consider an elevated left atrial volume index (LAVi) measurement. An LAVi measurement provides a precise representation of increased left atrial pressure, making it a more accurate indicator of heart failure than considering left atrium size alone.[26]

To establish listing-level severity for CHF, in addition to satisfying the criteria in proposed 4.02A, a person's CHF must satisfy the criteria in proposed 4.02B. The criteria in proposed 4.02B are similar to the criteria in current 4.02B1 and 4.02B3, respectively, with some important changes. For proposed 4.02B1a, we can use a conclusion by a medical source that an exercise tolerance test (ETT) presents a significant risk to a person; for example, the person's cardiologist stating that an ETT would cause cardiac instability or injury. For proposed 4.02B1b, we would require findings showing that a person is very seriously limited in his or her ability to perform an ETT, similar to current 4.02B3. Consistent with the IOM's recommendations, proposed 4.02B2 would also include findings showing the inability to perform on an ETT at 15 milliliters/kilograms/minute (ml/kg/min) peak VO_2 (oxygen consumption).[27] Peak VO_2 at this level is comparable to the requirement in current 4.02B3 for an inability to perform on an ETT at a workload equivalent to 5 metabolic equivalents (METs) of task.

We propose to remove current 4.02B2 ''three or more separate episodes of acute congestive heart failure'' because we would evaluate these episodes under proposed 4.02B3. As recommended by the IOM, proposed 4.02B3 would evaluate exacerbations or complications

[25] IOM. (2010), 88, 89.

[26] Cacciapuoti, Fu., Scognamiglio, A., Paoli, V.D., Romano, C., & Cacciapuoti, Fe. (2012). Left atrial volume index as indicator of left ventricular diastolic dysfunction: Comparation between left atrial volume index and tissue myocardial performance index. Journal of Cardiovascular Ultrasound, 20(1), 25–29. doi:10.4250/jcu.2012.20.1.25.

[27] IOM. (2010), 85, 93.

of CHF, requiring three hospitalizations within a consecutive 12-month period and at least 30 days apart.[28]

Additionally, proposed 4.02B2 would not include the requirement in current 4.02B3b of frequent premature ventricular contractions (PVCs). Frequent PVCs do not necessarily reflect an inability to perform an ETT.[29] [30] [31] The proposed listing also would no longer include the criterion in current 4.02B3d requiring signs attributable to inadequate cerebral perfusion, such as ataxic gait or mental confusion. Such manifestations rarely occur during an ETT, even if the person has very serious CHF.[32]

The IOM recommended a criterion for chronic heart failure in people who are stable and receiving treatment but have an EF on a sustained basis of 20 percent or less. An EF of only 20 percent means the heart's pumping action is less than a third of normal, and critically affects a person's ability to perform gainful activity.[33] [34] [35] Most individuals with disease this advanced have greater risk of mortality and major functional limitations, such as shortness of breath or fatigue, even during mild exertion.[36] We propose 4.02C consistent with the IOM's recommendation. We would require at least two EF measurements equal to or less than 20 percent at least 90 days apart within a consecutive 12-month period to document chronic disease and to exclude heart failure resulting from reversible causes.

Under proposed 4.02D1, we would include a new criterion for heart failure treated with a mechanical circulatory support device (MCSD). Implanted MCSDs, such as a left ventricle assistive device (LVAD) or a right ventricle assistive device (RVAD), help the heart pump blood and may be used as a ''bridge'' while a person waits for a heart transplant. We currently use our medical equivalence rules [37] to find someone with heart failure and an implanted MCSD disabled under listing 4.09 (*Heart transplantation*). Adding this new criterion will ensure that people with heart failure treated with MCSD are consistently identified.

People who require continuous intravenous administration of inotropic medication (for example, milrinone) have very serious heart failure, and the length of this treatment can be an accurate predictor of impairment severity.[38] [39] Accordingly, proposed 4.02D2 would find people disabled if they require continuous intravenous inotropic medications for 30 or more consecutive days.

Proposed Listing 4.04—Ischemic Heart Disease

As noted earlier in this preamble, we propose to use reports from AMSs under proposed 4.04A to determine whether ECG findings are positive for IHD. We would also rely on such reports to determine whether systolic blood pressure measurements during ETTs are positive for IHD. Based on our program experience and consultation with agency medical experts, we expect that relying on these reports from AMSs will ensure the accuracy of disability claims adjudication.

We would replace current 4.04A4 with proposed 4.04A. Proposed 4.04B adds the requirement to consider imaging results derived from pharmacologic stress testing. The new proposed requirement will provide more specific findings than the current criterion, which generally requires only ''documented ischemia.'' [40] Because of these changes, we would redesignate current 4.04B as proposed 4.04C and revise the introductory text accordingly.

Proposed 4.04C (current 4.04B) would require three separate ischemic episodes that result in unplanned hospitalizations within a consecutive 12-month period (see 4.00A3e (*What do the following terms or phrases mean in these listings?*), including episodes requiring unplanned revascularization or treatment for myocardial infarction (heart attack), unstable angina, or an irregular heartbeat. We would evaluate unplanned hospitalizations under this section to ensure we are only evaluating urgent ischemic episodes. Individuals who have ischemic episodes that result in unplanned hospitalizations may need intensive care, can have long hospital stays, may require multiple procedures, and can be at high risk for post-discharge morbidity and mortality.[41] [42] [43] [44] Many also have serious co-occurring medical conditions (for example, heart failure, chronic kidney disease, and chronic obstructive pulmonary disease).[45] [46] [47]

We would redesignate current 4.04C as proposed 4.04D. We would reorganize the criteria in current 4.04C and follow IOM's recommendation to add new criteria to evaluate the severity of a person's IHD regardless of whether he or she has had a timely ETT or pharmacologic stress test or whether such tests are contraindicated. ETTs and pharmacologic stress test are commonly performed for diagnostic and prognostic purposes, and applicable to determine

[28] IOM. (2010), 89, 91.

[29] Cha, Y.-M., Lee, G.K., Klarich, K.W., & Grogan, M. (2012). Premature ventricular contraction-induced cardiomyopathy: A treatable condition. Circulation: Arrhythmia and Electrophysiology, 5(1), 229–236. doi:10.1161/CIRCEP.111.963348.

[30] Dukes, J.W., Dewland, T.A., Vittinghoff, E., Mandyam, M.C., Heckbert, S.R., Siscovick, D.S., . . . Marcus, G.M. (2015). Ventricular ectopy as a predictor of heart failure and death. Journal of the American College of Cardiology, 66(2), 101–109. doi:10.1016/j.jacc.2015.04.062.

[31] IOM. (2010), 87.

[32] IOM. (2010), 87, 88.

[33] Desai, R.V., Guichard, J.L., Mujib, M., Ahmed, M.I., Feller, M.A., Fonarow, G.C., . . . Ahmed, A. (2013). Reduced right ventricular ejection fraction and increased mortality in chronic systolic heart failure patients receiving beta-blockers: Insights from the BEST trial. International Journal of Cardiology, 163(1), 61–67. doi:10.1016/j.ijcard.2011.05.051.

[34] IOM. (2010), 84, 89.

[35] Runge, M.S., Patterson, C., Stouffer, G.A., & Netter, F.H. (2010). Netter's Cardiology (2nd ed.). Philadelphia, PA: Saunders Elsevier.

[36] IOM. (2010), 78, 81, 82, 94.

[37] 20 CFR 404.1526 and 416.926.

[38] Malotte, K., Saguros, A., & Groninger, H. (2018). Continuous cardiac inotropes in patients with end-stage heart failure: An evolving experience. Journal of Pain Symptom Management, 55(1), 159–163. doi:10.1016/j.jpainsymman.2017.09.026.

[39] Bistola, V., Arfaras-Melainis, A., Polyzogopoulou, E., Ikonomidis, I., & Parissis, J. (2019). Inotropes in acute heart failure: From guidelines to practical use: Therapeutic options and clinical practice. Cardiac Failure Review, 5(3), 133–139. doi:10.15420/cfr.2019.11.2.

[40] IOM. (2010), 120.

[41] Hua, M., Gong, M.N., Brady, J., & Wunsch, H. (2015). Early and late unplanned rehospitalizations for survivors of critical illness. Critical Care Medicine, 43(2), 430–438. doi:10.1097/CCM.0000000000000717.

[42] Kim, Y., Gani, F., Canner, J.K., Margonis G.A., Makary, M.A., Schneider, E.B., & Pawlik, T.M. (2016). Hospital readmission after multiple major operative procedures among patients with employer provided health insurance. Surgery, 160(1), 178–190. doi:10.1016/j.surg.2016.01.025.

[43] Reynolds, K., Butler, M.G., Kimes, T.M., Rosales, A.G., Chan, W., & Nichols, G.A. (2015). Relation of acute heart failure hospital length of stay to subsequent readmission and all-cause mortality. American Journal of Cardiology, 116(3), 400–405. doi:10.1016/j.amjcard.2015.04.052.

[44] Yu, P.-J., Cassiere, H.A., Fishbein, J., Esposito, R.A., & Hartman, A.R. (2016). Outcomes of patients with prolonged intensive care unit length of stay after cardiac surgery. Journal of Cardiothoracic and Vascular Anesthesia, 30(6), 1550–1554. doi:10.1053/j.jvca.2016.03.145.

[45] Frigola-Capell, E., Comin-Colet, J., Davins-Miralles, J., Gich-Saladich, I., Wensing, M., & Verdú-Rotellar, J.M. (2012). Trends and predictors of hospitalization, readmissions and length of stay in ambulatory patients with heart failure. Revista Clinica Espanola, 213(1), 1–7. doi:10.1016/j.rce.2012.10.006.

[46] Nombela-Franco, L., del Trigo, M., Morrison-Polo, G., Veiga, G., Jimenez-Quevedo, P., Altisent, O.A.-J., . . . Rodés-Cabau, J. (2015). Incidence, causes, and predictors of early (<30 days) and late unplanned hospital readmissions after transcatheter aortic valve replacement. Journal of the American College of Cardiology: Cardiovascular Interventions, 8(13), 1748–1757. doi:10.1016/j.jcin.2015.07.022.

[47] Versteeg, H., Hoogwegt, M.T., Hansen, T.B., Pedersen, S.S., Zwisler, A.-D., & Thygesen, L.C. (2013). Depression, not anxiety, is independently associated with 5-year hospitalizations and mortality in patients with ischemic heart disease. Journal of Psychosomatic Research, 75(6), 518–525. doi:10.1016/j.jpsychores.2013.10.005.

functional capacity in persons with IHD.[48]

We would create proposed new 4.04D1 based upon blood flow in the coronary arteries expressed as fractional flow reserve (FFR). Updated medical science has shown FFR is a more objective and medically updated measure of the severity of stenosis.[49] A clinician may measure FFR in a stenotic (obstructed) artery to determine whether it requires revascularization. A normal (patent) artery has an FFR equal to 1.0. If a stenotic artery has an FFR equal to or less than 0.80, and the artery is amenable to revascularization, the clinician will use revascularization to restore the vessel, because the obstructed artery is causing significant ischemia. Accordingly, an FFR measurement less than or equal to 0.80 in the proximal or mid segment of an artery that is *not* amenable to revascularization is consistent with the requirements of proposed 4.04D1.[50] [51] [52]

In proposed 4.04D2 we would evaluate IHD by taking into consideration a person's history of coronary artery bypass graft surgery. In proposed 4.04D3, we would evaluate IHD by taking into consideration that a person has a decreased EF (*i.e.,* EF of less than 50 percent while medically stable and on a regimen of prescribed treatment).

In both proposed 4.04D2 and 4.04D3, we would require the person to have symptoms of myocardial ischemia, as described in current 4.00E3 through 4.00E7 and be on a regimen of prescribed treatment. Proposed 4.04D2 and 4.04D3 would include the criteria in current 4.04C1a, b, and d with minor editorial changes. Proposed 4.04D2 includes criteria in current 4.04C1e. We would not include the criteria in current 4.04C1c in the proposed listings 4.04D2 and 4.04D3 because based on our program experience, we determined that these criteria do not consistently correlate with listing-level severity.

We would remove current 4.04C2 with its requirement that a person's IHD result in very serious limitations in his or her ability to independently initiate, sustain, or complete activities of daily living. The criteria in proposed listings 4.04D1, 4.04D2, and 4.04D3 alone provide a description of listing-level IHD.

We are proposing new 4.04E to evaluate exacerbations or complications of IHD requiring three hospitalizations within a consecutive 12-month period and at least 30 days apart. These hospitalizations may be planned or unplanned. The hospitalizations required under 4.04E differ from those required in proposed 4.04C, which requires that hospitalizations be unplanned.

Proposed Listing 4.05—Recurrent Arrhythmias

We propose to reorganize the basic structure and presentation of current 4.05 (*Recurrent arrhythmias*) to improve its clarity and ease of reference. We would breakdown the current criteria into two parts: recurrent episodes of syncope (or near syncope) and findings and documentation by a medically acceptable test, to demonstrate that both parts must be satisfied to document listing-level severity. We would also remove ''uncontrolled'' as a descriptor for recurrent episodes of cardiac syncope because, inherently, these episodes are uncontrolled if they recur while a person is on a regimen of prescribed treatment. Because we would remove ''uncontrolled'' as a descriptor in the listing, we would also remove the definition for this term from current 4.00A3 in the introductory text. Symptoms associated with arrhythmia include: anxiety, chest pain, fatigue, sweating, near-syncope, and fainting (syncope). Syncope and near-syncope are two of the more serious symptoms; individuals with arrhythmias and recurrent episodes of syncope have a higher risk of mortality and sudden cardiac death. Furthermore, syncope and near-syncope are more quantifiable and objective than other symptoms like anxiety and fatigue. For these reasons, recurrent episodes of syncope and near syncope continue to be an appropriate indicator of listing-level severity for individuals with recurrent arrhythmias.[53]

Proposed Listing 4.06—Congenital Heart Disease

For the reason discussed below, we propose to remove the parenthetical reference to ''cyanotic or acyanotic'' congenital heart disease from the heading in current 4.06 in order to focus on hypoxemia.

Accordingly, we propose to revise current 4.06A to require ''hypoxemia'' rather than ''cyanosis or acyanosis.'' ''Hypoxemia'' reflects abnormalities in the blood, such as an increased hematocrit level or a low blood oxygen level, which are detected through laboratory analysis or pulse oximetry. On the other hand, the term ''cyanosis'' refers to skin discoloration observed during a physical examination. Cyanosis is a more subjective assessment subject to misinterpretation due to by many factors, including skin complexion. Thus, the term ''hypoxemia'' relates more to the laboratory and pulse oximetry findings than the term ''cyanosis.'' [54]

To establish listing-level severity for individuals with congenital heart disease, IOM recommended documentation of chronic and persistent hypoxemia. Therefore to demonstrate the chronic and persistent nature, in proposed 4.06A1, we would require two hematocrit measurements instead of the current listing's single measurement. Two measurements, at least 90 days apart within a consecutive 12-month period will help ensure the person's hematocrit level is associated with chronic hypoxemia and not the result of a reversible condition, such as dehydration.[55] The proposed requirement that the two measurements be 90 days apart is consistent with the time period requirement used in our other body system listings, and consistent with instructions providers receive for scheduling patients [56] and established check-up intervals for adults with congenital heart disease.[57]

[48] IOM. (2010), 70.

[49] Shlofmitz, E., and Jeremias, A. (2017). *FFR in 2017: Current Status in PCI Management—American College of Cardiology.* Available online at: *https://www.acc.org/latest-in-cardiology/articles/2017/05/25/08/34/ffr-in-2017-current-status-in-pci-management* (accessed September 17, 2021).

[50] Fearon, W.F. (2014). Percutaneous coronary intervention should be guided by fractional flow reserve measurement. Circulation, 129(18), 1860–1870. doi:10.1161/CIRCULATIONAHA.113.004300.

[51] Fearon, W.F., De Bruyne, B., & Pijlis, N.H. (2016). Fractional flow reserve in acute coronary syndromes. Journal of the American College of Cardiology, 68(11), 1192–1194. doi:10.1016/j.jacc.2016.07.713.

[52] Tebaldi, M., Biscaglia, S., Pecoraro, A., Fineschi, M., & Campo, G. (2016). Fractional flow reserve implementation in daily clinical practice: A European survey. International Journal of Cardiology, 207, 206–207. doi:10.1016/j.ijcard.2016.01.097.

[53] Koene, R.J., Adkisson, W.O., & Benditt, D.G. (2017). Syncope and the risk of sudden cardiac death: Evaluation, management, and prevention. *Journal of arrhythmia, 33*(6), 533–544. *https://doi.org/10.1016/j.joa.2017.07.005.*

[54] Stout, K.K. (2019).

[55] Stack, S.W. . . ., & Berger, S.A. (2009). The effects of high hematocrit arterial flow—A phenomenological study of health risk implications. Chemical Engineering Science, 64(22), 4701–4706. doi:10/1016/j.ces.2009.07.017.

[56] Bavafa, H., Savin, S., & Terwiesch, C. (2019). Redesigning Primary Care Delivery: Customized Office Revisit Intervals and E-Visits. *https://dx.doi.org/10.21con39/ssrn.2363685.*

Paper referenced by Bavafa: Schectman, G., G. Barnas, P. Laud, L. Cantwell, M. Horton, E.J. Zarling. 2005. Prolonging the return visit interval in primary care. *The American Journal of Medicine, 118*(4) 393–39.

[57] According to the University of Washington's Adult Congenital Heart Disease Clinic's *Information for Patients and Families* (2016), most people with congenital heart disease require regular check-ups with their cardiologist ''at intervals ranging from

Continued

Furthermore, requiring two measurements at least 90 days apart is consistent with the current (and proposed) childhood congenital heart disease criterion (104.06A1) and will assist with documenting duration and establishing that the persistent nature of the person's condition.[58]

We would include the medical abbreviation ''S_aO_2'' in 4.06A2. This abbreviation frequently appears in the medical evidence to indicate arterial oxygen (O_2) saturation determined by arterial blood gas testing. We would also include the medical abbreviation ''P_aO_2'' (partial pressure of O_2), because medical reports may use it interchangeably with the abbreviation ''PO_2'' that we use in current 4.06A for arterial partial pressure of oxygen. Additionally, we would express P_aO_2 and PO_2 in millimeters of mercury (mmHg) instead of Torr units to make the listing consistent with current medical practice and terminology.[59]

In proposed 4.06A3, we would add a criterion for S_pO_2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry, including measurements taken while the person is at rest or while doing a six-minute walk test (6MWT). Pulse oximetry measurements are a non-invasive alternative to invasive testing. A person's medical evidence often provides S_pO_2 findings, and S_pO_2 measured by pulse oximetry reflects an advance in medical technology that provides another way to establish listing-level severity.[60] [61] [62]

Proposed 4.06A3 would require three S_pO_2 measurements 30 days apart within a consecutive 12-month period showing hypoxemia. We explain in the introductory text of proposed 4.00H4c that these measurements must be documented by a medical source using methods consistent with the prevailing state of medical knowledge and clinical practice, and also must be consistent with the other evidence in the person's case record. We would require an S_pO_2 of 87 percent or less because this finding is comparable in severity to an S_aO_2 of less than 90 percent in current 4.06A2.[63] By requiring several measurements at least 30 days apart, we ensure that the required findings span a period of at least 90 days. Similar to the requirement for repeated hematocrit measurements under 4.06A1, this will document that the condition is chronic and persistent, and the measurements are not related to a reversible condition or an inaccurate reading.

In proposed 4.06B, we would include an additional option of taking an S_aO_2 measurement for determining the level of hypoxemia during exertion. This change would provide an additional way of evaluating hypoxemia. Similarly, we would include an oxygen uptake measurement as another option.

We are proposing several changes to current 4.06C. We would use the term ''pulmonary hypertension'' to describe the impairment instead of the term ''pulmonary vascular obstructive disease.'' ''Pulmonary hypertension'' is the term more commonly used by clinicians and, therefore, the most likely to appear in the medical evidence.[64] We are also proposing to delete ''secondary'' from the listing's heading, because pulmonary hypertension can be disabling regardless of whether it is a ''primary'' or ''secondary'' condition.

We would include medical findings in proposed 4.06C that are expressed in millimeters of mercury (mmHg). Findings of pulmonary artery pressure are expressed in mmHg more often than they are expressed as a percentage of ''systemic arterial systolic pressure,'' as in current 4.06C. Pulmonary hypertension may be reported in the medical evidence as either pulmonary artery pressure or *mean* pulmonary artery pressure, so we would include both types of findings in the proposed listing.

We would add a new criterion—proposed 4.06D—to evaluate adults with ''single ventricle,'' which is also known as ''single ventricle physiology'' or ''functional single ventricle.'' Children born with single ventricle have a severe, medically determinable impairment (MDI) that will usually need to be corrected by staged surgery called ''Fontan procedures.'' [65] These procedures enable an increasing percentage of affected children to survive into adulthood. As adults, they have significantly reduced functional capacities that steadily decline. We would find adults disabled under proposed 4.06D if objective medical evidence shows the person has single ventricle, regardless of whether or not they had Fontan or other surgical procedures.[66] [67] We provide information in the introductory text in proposed 4.00H3 (*What is single ventricle?*) about single ventricle and these surgical procedures.

Proposed Listing 4.07—Aortic Valvular Disease

We currently evaluate aortic valvular disease under other cardiovascular disorders listings, which include requirements for ETT or repeated hospitalization. According to the IOM report, due to the risk associated with exercise testing for individuals with symptomatic aortic stenosis, ETT is not advised.[68] Furthermore, very serious symptomatic aortic stenosis is ''universally fatal'' if left untreated and there are few effective, long-term medical therapies for individuals with this level of disease.[69] Therefore, we followed IOM recommendations to provide evaluation criteria for aortic valvular disease and propose to add new listing 4.07 to evaluate aortic valvular disease. The medical community considers an aortic valve area equal to or less than 1.0 cm^2 indicative of advanced stenotic disease associated with significant dyspnea, fatigue, angina, and other serious

every several months to every several years.'' Accessed May 4, 2022, from *HeartInstitute_AdultCongenitalHeartDiseaseClinic.pdf* (*uwmedicine.org*).

People with listing-level congenital heart disease are expected to require more frequent checkups than those who are asymptomatic or have less severe disease.

[58] See 20 CFR 404.1505(a) and 416.905(a). The law defines disability as the inability to do any substantial gainful activity by reason of any medically determinable physical or mental impairment which 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.

[59] Castro D, Patil SM, Keenaghan M. Arterial Blood Gas. 2021 Jan 27. In: StatPearls [internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan–. PMID: 30725604.

[60] IOM. (2010), 178.

[61] Oster, M. E. . . ., & Kochilas, L.K. (2016). Screening for critical congenital heart disease. Clinics in Perinatology, 43(1), 73–80. doi:10.1016/j.clp.2015.11.005.

[62] Mechem, C.C. (2014). Pulse oximetry. In P.E. Parsons (Ed.), UpToDate (Jan. 2014). Retrieved from *https://www.uptodate.com/contents/pulse-oximetry.*

[63] IOM. (2010), 178.

[64] A review of the website for the Journal of the American Medical Association (JAMA), a peer-reviewed medical journal published 48 times a year by the American Medical Association, found that the exact term ''pulmonary hypertension'' came back with more than 800 results. A search for the exact term ''pulmonary vascular obstructive disease'' came back with zero results. The search was conducted on September 8, 2021.

[65] People with single ventricle will generally undergo staged reconstructive ''Fontan procedures,'' ultimately resulting in a ''Fontan circulation.'' Fontan circulation describes the state in which virtually all systemic venous return-blood passively flows directly into the pulmonary arteries via surgical or catheter-placed shunts, without the blood passing through a ventricle.

[66] Cohen, S., & Marelli, A. (2016). Evolving heart transplantation across the lifespan: A growing population of adults with congenital heart disease. Archives of Cardiovascular Disease, 109(10), 511–513. doi:10.1016/j.acvd.2016.05.001.

[67] IOM. (2010), 169, 178.

[68] IOM. (2010), 195.

[69] IOM. (2010), 194, 195.

symptoms.[70] [71] [72] [73] Proposed 4.07 would require appropriate testing that documents the aortic valve area.

Proposed Listing 4.08—Cardiomyopathy

Consistent with IOM recommendations, we would add 4.08 to evaluate cardiomyopathies, such as hypertrophic cardiomyopathy (HCM). We currently evaluate cardiomyopathy under 4.02 (*Chronic heart failure*), 4.04 (*Ischemic heart disease*), 4.05 (*Recurrent arrhythmias*), or 11.04 (*Vascular insult to the brain*), depending on its effects. Depending on the underlying cause of the person's cardiomyopathy or its effects, we may continue to evaluate cardiomyopathy under 4.02, 4.04, 4.05, and 11.04. We created this new cardiomyopathy listing to specifically address HCM, endomyocardial fibrosis, and cardiac amyloidosis AL type, which are more serious types of cardiomyopathy.[74]

HCM with severe left ventricular or septal wall thickness can cause serious problems, including chest pain, dyspnea, syncope, and arrhythmias.[75] As recommended by IOM, we would evaluate HCM under proposed 4.08A by requiring the heart to have a left ventricular or septal wall thickness equal to or greater than 20 millimeters. Proposed 4.08 also requires the individual to be seriously limited in the ability to perform an ETT, or a medical source has concluded that the performance of an ETT would present a significant risk.

Proposed 4.08B would evaluate endomyocardial fibrosis, a form of cardiomyopathy with a generally poor prognosis despite treatment. Under proposed 4.08B, we would require endomyocardial fibrosis resulting in a loss of heart chamber volume, atrial dilatation, and mitral or tricuspid valve regurgitation.

Proposed 4.08C would evaluate cardiac amyloidosis AL (light-chain) type, another form of cardiomyopathy with a poor prognosis. We would need objective medical evidence, such as biopsy findings, echocardiogram, cardiac MRI, and PET scan to establish listing-level severity.

Proposed 4.08D would evaluate exacerbations or complications of cardiomyopathy, requiring three hospitalizations within a consecutive 12-month period and at least 30 days apart.

Proposed Listing 4.09—Heart Transplantation

We are proposing editorial changes in the heading and text of current 4.09, which would not be substantive but would clarify the guidance. We have changed from ''1 year following surgery'' to ''1 year from the date of the transplant'' consistent with transplantation listings in other body systems such as 6.04 (*Chronic kidney disease*) and 7.17 (*Hematological disorders treated by bone marrow or stem cell transplantation*).

Proposed Listing 4.10—Dissecting Aneurysm of the Aorta or Major Branches

We propose to revise the heading for listing 4.10 to specify that we evaluate only ''dissecting'' aneurysms under the listing consistent with IOM recommendations.[76]

Proposed Listing 4.11—Chronic Venous Insufficiency

We propose to revise the heading in current listing 4.11 by replacing the outdated term ''incompetency'' with the term ''reflux''—the term the medical community currently uses to describe decrease blood flow and pooling of blood in the veins.[77] We would delete the word ''deep'' in the heading so that the listing covers reflux or obstruction associated with superficial and perforating veins. Reflux or obstruction in these veins may result in the required level of CVI.[78] Additionally, as recommended by IOM, we would require confirmation of CVI by duplex ultrasound or other appropriate diagnostic technique. The medical community considers the use of duplex ultrasound to be the best method for detecting reflux or obstruction.[79]

In proposed 4.11A, we would adopt IOM recommendations and broaden the listing criteria we apply to trophic changes (changes resulting from interruption of nerve supply) of the skin. For example, in addition to brawny edema, trophic changes evaluated under the proposed listing would include hyperpigmentation and lipodermatosclerosis.

We would revise the current requirement that these skin changes involve ''at least two-thirds of the leg between the ankle and knee or the distal one-third of the lower extremity between the ankle and hip.'' Instead, we would require extensive skin changes involving at least two-thirds of the leg below the knee, to make the requirement simpler to understand and apply. This revision is consistent with IOM's recommendation to require skin changes below the knee.[80]

We would require the skin changes under proposed 4.11A to be consistent with CVI, and we would document the skin changes over a period of at least 90 days to ensure they are chronic. Additionally, the CVI must be unresponsive to compression therapy, because this therapy usually enables people to return to a good level of functioning.[81]

In proposed 4.11B, we would remove findings in the current listing that no longer demonstrate required severity. For example, we would remove superficial varicosities, which indicate venous disease but not necessarily CVI. We would follow IOM's recommendation and also remove stasis dermatitis, because it is ''a generic term referring to the trophic changes,'' and it is unreliable because it may be a sign of other unrelated conditions including aging.[82]

Proposed 4.11B would require recurrent or persistent skin ulceration that has not healed after 6 or more months of prescribed treatment. In regard to documenting duration and severity of CVI, this requirement is more conclusive than the current requirement of 3 months of unsuccessful prescribed treatment as it demonstrates the condition has persisted despite treatment for a longer period of time.[83]

Proposed Listing 4.12—Peripheral Arterial Disease

We propose to revise the heading of the current listing to evaluate peripheral arterial disease (PAD) while the person is on a regimen of prescribed treatment. PAD often improves with angioplasty, supervised physical rehabilitation, and other prescribed therapies.[84]

[70] Berthelot-Richer, M., Pibarot, P., Capoulade, R., Dumesnil, J.G., Dahou, A., Thebault, C., . . . Clavel, M.-A. (2016). Discordant grading of aortic stenosis severity: Echocardiographic predictors of survival benefit associated with aortic valve replacement. Journal of the American College of Cardiology: Cardiovascular Imaging, 9(7), 797–805. doi:10.1016/j.jcmg.2015.09.026.

[71] IOM. (2010), 195.

[72] Nombela-Franco, L. (2015).

[73] Ziberszac, R., Gabriel, H., Schemper, M., Laufer, G., Maurer, G., & Rosenhek, R. (2017). Asymptomatic severe aortic stenosis in the elderly. Journal of the American College of Cardiology: Cardiovascular Imaging, 10(1), 43–50. doi:10.1016/j.jcmg.2016.05.015.

[74] SSA has designated endomyocardial fibrosis and cardiac amyloidosis AL type as Compassionate Allowance (CAL) conditions. See *Compassionate Allowances website Home Page (ssa.gov).*

[75] IOM. (2010), 80.

[76] IOM. (2010), 217.

[77] IOM. (2010), 160.

[78] IOM. (2010), 161.

[79] Id.

[80] IOM. (2010), 157–161.

[81] IOM. (2010), 159.

[82] IOM. (2010), 161.

[83] IOM. (2010), 161.

[84] Poredoš P, Jezovnik M, Kalodiki E. Medical management of patients with peripheral arterial

Continued

We would add leg pain in the heading as a serious and potentially debilitating symptom of PAD. People who have PAD with intermittent leg pain may be impaired to a similar extent as a person with PAD with intermittent claudication.[85]

Consistent with IOM recommendations, we would require a person's intermittent leg pain or claudication to interfere with his or her mobility. This proposed change clarifies our original intent in the listing, which is to tie PAD to functioning. Finally, we would replace the term ''appropriate medically acceptable imaging'' in the listing heading with ''appropriate test(s)'' (4.00G6—*Are there any other studies that are helpful in evaluating PAD?*) to acknowledge that non-imaging procedures such as physical examination and blood tests may also help detect PAD.[86]

Proposed Listing 4.16—Cardiac Allograft Vasculopathy

We propose to add new listing 4.16 to evaluate a person who received a heart transplant (allograft) and subsequently developed cardiac allograft vasculopathy (CAV). Currently, we evaluate CAV through medical equivalence to listing 4.09 (*Heart transplant*). CAV results in stenosis of the heart's blood vessels that may progress quickly and cause significant heart dysfunction. CAV with moderate stenosis, as defined in the medical literature,[87] may also result in a listing-level impairment, depending on the extent and seriousness of dysfunction. To establish the required level of CAV, we would require a cardiac index (cardiac output) of less than 2 liters/minute/meter square (L/min/m^2), an ejection fraction equal to or less than 45 percent, right atrial pressure greater than 12 mmHg, or pulmonary capillary wedge pressure greater than 15 mmHg. Individuals who have any of these findings have a poor prognosis and are limited in their activities and ability to work.[88] [89] [90] [91]

Other Proposed Changes

As mentioned, we are proposing new criteria to evaluate exacerbations or complications of several categories of cardiovascular disorders. These new criteria include proposed 4.02B3 for evaluating chronic heart failure, proposed 4.04E for evaluating ischemic heart disease, proposed 4.06E for evaluating congenital heart disease, and proposed 4.08D for evaluating cardiomyopathies. Consistent with IOM recommendations, we are proposing these new criteria for evaluating chronic heart failure and cardiomyopathies.[92] In addition, we are proposing these new criteria for evaluating ischemic heart disease (4.04) and congenital heart disease (4.06). Our adjudicative experience shows that these cardiovascular disorders are prone to exacerbations and serious complications. These proposed criteria would require exacerbations or complications causing a person to be hospitalized three or more times within a consecutive 12-month period.[93] An impairment resulting in exacerbations or complications that require this many hospitalizations in 12 months will prevent a person from engaging in any gainful activity.[94] [95] [96] [97] [98] [99] [100] We would require these hospitalizations to be at least 30 days apart and to last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization, to ensure we are evaluating separate listing-level episodes of exacerbations or complications. Our proposal to require that each hospitalization last at least 48 hours is generally consistent with data showing that the average length of hospital stays for serious cardiac conditions like primary heart failure and adult congenital heart disease is at least 48 hours.[101] [102]

Another revision we are proposing would affect chronic venous insufficiency evaluated under 4.11. Under our proposed changes, we would follow IOM's recommendations and require documentation that certain manifestations of chronic venous insufficiency (for example, trophic changes of the skin) occurred at least twice within a consecutive 12-month period, instead of only once under the current listings.[103] This change is based on the IOM recommendation that two occurrences per year more accurately and consistently demonstrates listing-level severity. We would also require documentation that these manifestations occurred at least 90 days apart. These requirements ensure we are appropriately documenting the chronicity and persistence of these conditions and evaluating people who have very serious chronic conditions.

Proposed Changes to the Childhood Cardiovascular Disorders Introductory Text

Proposed 104.00—Introductory Text to the Childhood Cardiovascular Disorders Listings

We repeat much of the introductory text of proposed 4.00 in the introductory text of proposed 104.00, because the same basic criteria for evaluating cardiovascular disorders apply to both adults and children. Because we have already described these proposed criteria above, the following discussion describes only those criteria that are unique to children or that require further explanation in how they will be applied to children.

The following table shows the heading of the current and proposed sections of the childhood introductory text for cardiovascular disorders:

disease. Int Angiol. 2015 Feb;34(1):75–93. Epub 2014 Jun 11. PMID: 24916346.

[85] IOM. (2010), 151.

[86] Id.

[87] The International Society of Heart and Lung Transplantation's grading classification defines mild cardiac allograft vasculopathy (CAV_1) as having left main artery stenosis of less than 50 percent, primary vessel stenosis greater than 70 percent (including the right coronary artery), or any branch stenosis greater than 70 percent (see Mehra, M.R., Crespo-Leiro, M.G., Dipchand, A., Ensminger, S.M., Hiemann, N.E., Kobashigawa, J.A., . . . Uber, P.A. (2010). International Society of Heart and Lung Transplantation working formulation of a standardized nomenclature for cardiac allograft vasculopathy—2010. Journal of Heart and Lung Transplantation, 29(7), 717–727. doi:10.1016/j.healun.2010.05.017).

[88] Kindel, S.J., & Pahl, E. (2011). Cardiac allograft vasculopathy in children—treatment challenges. Progress in Pediatric Cardiology, 32(1), 37–42. doi:10.1016/j.ppedcard.2011.06.008.

[89] Kobashingawa, J.A. (2015). The changing face of first-year intravascular ultrasonography in heart transplantation. Journal of the American College of Cardiology: Heart Failure, 3(12), 954–955. doi:10.1016/j.jchf.2015.09.004.

[90] Mehra, M.R. (2010).

[91] Okada, K., Kitahara, H., Yang, H., Tanaka, S., Kobayashi, Y., Kimura, T., . . . Fearon, W.F. (2015). Paradoxical vessel remodeling of the proximal segment of the left anterior descending artery predicts long-term mortality after heart transplantation. Journal of the American College of Cardiology: Heart Failure, 3(12), 945–952. doi:10.1016/j.jchf.2015.07.013.

[92] IOM. (2010), 89, 172.

[93] Id.

[94] Hua, M. (2015).

[95] IOM. (2010), 30, 90, 196.

[96] Kim, Y. (2016).

[97] Kyriakou, M., & Kiff, P.F. (2016). Prognosis of the comorbid heart failure and anemia: A systematic review and meta-analysis. Clinical Trials and Regulatory Science in Cardiology, 16, 12–21. doi:10.1016/j.ctrsc.2016.01.008.

[98] Reynolds, K. (2015).

[99] Versteeg, H. (2013).

[100] Yu, P.-J. (2016).

[101] Jackson SL, Tong X, King RJ, et al. National Burden of Heart Failure Events in the United States, 2006 to 2014. Circulation. Heart Failure. 2018 Dec;11(12):e004873. DOI: 10.1161/circheartfailure.117.004873. PMID: 30562099; PMCID: PMC6424109.

[102] Cedars, A., Benjamin, L., Burns, S.V., Novak, E., & Amin, A. (2017). Clinical predictors of length of stay in adults with congenital heart disease. *Heart (British Cardiac Society), 103*(16), 1258–1263. *https://doi.org/10.1136/heartjnl-2016-310841.*

[103] IOM. (2010), 161.

Current sections of the childhood introductory text and listings for cardiovascular system	Proposed sections of the childhood introductory text and listings for cardiovascular disorders
104.00 Cardiovascular System	104.00 Cardiovascular Disorders.
A. General	A. How do we define cardiovascular disorder and cardiovascular terms?
B. Documenting Cardiovascular Impairment	B. What documentation do we need to evaluate cardiovascular disorders?
C. Evaluating Chronic Heart Failure	C. How do we evaluate chronic heart failure?
D. Evaluating Congenital Heart Disease	D. How do we evaluate congenital heart disease?
E. Evaluating Arrhythmias	E. How do we evaluate arrhythmias?
F. Evaluating Other Cardiovascular Impairments	F. How do we evaluate other cardiovascular disorders?
G. Other Evaluation Issues	G. How do we evaluate issues that affect the cardiovascular system?
	H. How do we evaluate cardiovascular disorders that do not meet one of these listings?

Proposed 104.00C—How do we evaluate chronic heart failure?

We are proposing changes in current 104.00C consistent with changes we are proposing in the adult listings for chronic heart failure. We would extensively revise current 104.00C2 by removing specific findings for documenting cardiomegaly. These findings are often not provided in a child's case record and, therefore, have presented difficulty in adjudication. Proposed 104.00C2a would describe the types of imaging provided in a child's case record for documenting cardiomegaly. We explain at 104.00C2b(iii) that signs of congestion need not be found on all examinations because congestion may be controlled by prescribed treatment or may not be present at the time of evaluation. We have added 104.00C4 to explain how we propose to evaluate chronic heart failure treated with a mechanical circulatory support device under proposed 104.02D (*Chronic heart failure*).

Proposed 104.00D—How do we evaluate congenital heart disease?

We plan to significantly expand the information in current 104.00D. In proposed 104.00D2 (*How do we evaluate conditions associated with congenital heart disease?*), we would explain how we evaluate conditions associated with congenital heart disease. Proposed 104.00D2 includes additional means of measuring oxygen saturation in 104.06 (*Congenital heart disease*), because these measurements are readily found in the medical evidence. We are proposing new 104.00D3 (*What is Eisenmenger syndrome?*) to explain Eisenmenger syndrome in children, and we propose new 104.00D4 (*What is a single ventricle?*) to include a definition for the term ''single ventricle.''

Proposed 104.00F—How do we evaluate other cardiovascular disorders?

We propose revisions to 104.00F6 (*How will we evaluate chronic rheumatic fever or rheumatic heart disease?*) consistent with the removal of current listing 104.13, rheumatic heart disease. These revisions would explain that we evaluate rheumatic heart disease under 104.02 (*Chronic heart failure*) or 104.05 (*Recurrent arrhythmias*). We propose adding 104.00F11 (*What is cardiac allograft vasculopathy and how do we evaluate it?*) consistent with proposed 104.16 (*Cardiac allograft vasculopathy*).

Proposed 104.00G—How do we evaluate issues that affect the cardiovascular system?

We propose revisions to 104.00G consistent with those proposed to the adult listings. We propose to revise 104.00G1 (*How do we consider the effects of obesity when we evaluate your cardiovascular disorder?*) to simplify and refocus our discussion of how we consider the effects of obesity more specifically on cardiovascular disorders. We propose adding 104.00G3 (*How do we consider hospitalizations?*), consistent with new 104.02E (*Chronic heart failure*) and 104.06E (*Congenital heart disease*). We propose to redesignate current 104.00G3 (*How do we evaluate impairments that do not meet one of the cardiovascular listings?*) as 104.00H (*How do we evaluate cardiovascular disorders that do not meet one of these listings?*).

Proposed Changes to the Childhood Cardiovascular Disorders Listings

We are proposing some changes to the childhood listings that correspond with changes we are proposing to the adult listings. Other changes are specific to how we evaluate cardiovascular disorders in children. The reasons provided above for changing or removing current criteria for adults also apply to the criteria for children. Because we have already described these proposed criteria above, the following discussion describes only those criteria that are unique to children or that require further explanation in how we will specifically apply them to children. Additionally, the numbering of the childhood listings would conform to that of the adult listings.

The following table shows the heading of the current and proposed sections of the childhood listings for cardiovascular disorders:

CHILDHOOD CARDIOVASCULAR DISORDERS LISTINGS

Current	Proposed
104.02 Chronic heart failure	104.02 Chronic heart failure.
104.05 Recurrent arrhythmias	104.03 [Reserved].
104.06 Congenital heart disease	104.04 [Reserved].
104.09 Heart transplant	104.05 Recurrent arrhythmias.
104.13 Rheumatic heart disease	104.06 Congenital heart disease.
	104.07 [Reserved].
	104.08 [Reserved].
	104.09 Heart transplantation.
	104.10 [Reserved].
	104.11 [Reserved].
	104.12 [Reserved].

CHILDHOOD CARDIOVASCULAR DISORDERS LISTINGS—Continued

Current	Proposed
	104.13 [Reserved]. 104.14 [Reserved]. 104.15 [Reserved]. 104.16 Cardiac allograft vasculopathy.

The following table shows our proposed changes to the childhood cardiovascular disorders listings criteria that involve changes to healthcare utilization and condition/episode requirements, the rationale for each change, and supporting resource. Following this table, we discuss all of our proposed changes to the childhood cardiovascular disorders listings in more detail.

CHILDHOOD CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS

Current listing criterion	Proposed listing criterion	Rationale	Resources
Listing 104.02 Chronic heart failure			
104.02A Persistent tachycardia at rest (see Table I);	A. Persistent tachycardia at rest measured at least twice within a consecutive 12-month period and at least 90 days apart documented by apical heart rate greater than or equal to the value in Table I.	We would clarify that to satisfy 104.02A, we would require two or more tachycardia measurements in a consecutive 12-month period. Our intent is to ensure that the child has persistent tachycardia despite treatment. We would also require that readings of tachycardia occur at least 90 days apart to further document chronic disease.	IOM. (2010), 171,173, 176.
104.02B ..... Persistent tachypnea at rest (see Table II) or markedly decreased exercise tolerance (see 104.00C2b);	B. Persistent tachypnea at rest measured at least twice within a consecutive12-month period and at least 90 days apart documented by respiratory rate greater than or equal to the value in Table II or markedly decreased exercise tolerance (see 104.00C2b).	To satisfy 104.02B, we would require two or more tachypnea measurements in a consecutive 12-month period. Our intent is to ensure that the child has persistent tachypnea, despite treatment. We would also require that readings of tachypnea occur at least 90 days apart to further document chronic disease.	IOM. (2010), 171,173, 176.
Listing 104.06 Congenital heart disease			
104.06 ..... A. 2. Arterial O2 saturation of less than 90 percent in room air, or resting arterial PO2 of 60 Torr or less; or 3. Hypercyanotic spells, syncope, characteristic squatting, or other incapacitating symptoms directly related to documented cyanotic heart disease; or 4. Exercise intolerance with increased hypoxemia on exertion.	104.06 ..... A. 2. Arterial blood gas test measurement obtained at rest while breathing room air, as described in either a or b: a. SaO2 (arterial oxygen saturation) less than or equal to 89 percent; or b. PO2 or PaO2 (partial pressure of oxygen) less than or equal to 60 mmHg; or 3. SpO2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry either at rest, or after activity, while breathing room air, less than or equal to 87 percent on three evaluations at least 30 days apart within a consecutive 12-month period (see 104.00A3e).	In 104.06A2, we would use the measurement of millimeters of mercury, ''mmHg,'' instead of the measurement of ''Torr'' that is used in current 104.06A2, and we would note that arterial PO_2 is normally measured in room air. We would remove current 104.06A3 and 104.06A4, because Agency medical experts indicated they are less objective and more difficult to document than the other criteria and they are used infrequently. We would add another criterion to 104.06A by adding S_pO_2 (percentage of oxygen saturation of blood hemoglobin), measured by pulse oximetry equal to or less than 87 percent. Consistent with the proposed adult listing (4.06), this criterion would become the new 104.06A3.	Based on SSA administrative data from FY 2019–2021, of all childhood claims with a primary impairment of congenital heart disease that met or medically equaled listing 104.06, approximately .2 percent cited 104.06A3 or 104.06A4 criteria. See Table A and B in supporting and related materials to this Docket for more information.

CHILDHOOD CARDIOVASCULAR DISORDERS LISTINGS CRITERIA—CHANGES IN HEALTHCARE UTILIZATION AND CONDITION/EPISODE REQUIREMENTS—Continued

Current listing criterion	Proposed listing criterion	Rationale	Resources
No current criteria	E. Exacerbations or complications of congenital heart disease (see 104.00D) requiring three hospitalizations within a consecutive 12-month period (see 104.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 104.00G3).	We would add 104.06E to evaluate exacerbations or complications of congenital heart disease occurring at least 30 days apart and resulting in at least three hospitalizations within a consecutive 12-month period. An impairment resulting in exacerbations or complications that require this many hospitalizations in 12 months will result in marked and severe functional limitations for children. We would require these hospitalizations to be at least 30 days apart to ensure we are evaluating separate episodes of exacerbations or complications.	IOM. (2010), 179.

Proposed Listing 104.02—Chronic Heart Failure

We would clarify that to satisfy 104.02A, we would require two or more tachycardia measurements in a consecutive 12-month period, and to satisfy 104.02B, we would require two or more tachypnea measurements in a consecutive 12-month period.[104] Our intent is to ensure that the child has persistent tachycardia or persistent tachypnea, despite treatment. We would also require that readings of tachycardia or tachypnea occur at least 90 days apart to further document chronic disease.[105]

When we last published final rules for growth disorders and weight loss in children,[106] we inadvertently removed Table I for tachycardia at rest and Table II for tachypnea at rest in listing 104.02. We would restore these tables to the proposed 104.02.

We would add new 104.02D to describe how we will evaluate chronic heart failure treated with a mechanical circulatory support device.

We would add new 104.02E(), to describe how we will evaluate exacerbations and complications of heart failure requiring extended medical intervention in the hospital or emergency department, as explained above.

Proposed Listing 104.06—Congenital Heart Disease

In 104.06A2, we would use the measurement of millimeters of mercury, ''mmHg,'' instead of the measurement of ''Torr'' that is used in current 104.06A2, and we would note that arterial PO_2 is normally measured in room air.

We would add another criterion to 104.06A by adding S_pO_2 (percentage of oxygen saturation of blood hemoglobin), measured by pulse oximetry equal to or less than 87 percent. This criterion would become the new criterion 104.06A3. As we are proposing in the adult criteria, we would explain in the introductory text to the childhood listings that we need pulse oximetry measurements documented by medical sources using methods consistent with the prevailing state of medical knowledge and clinical practice. These measurements must be consistent with the other evidence in the case record.

We would remove current 104.06A3 and 104.06A4, because they are used infrequently.[107] Our adjudicative experience shows that children with impairments meeting these listings would be evaluated under current and proposed 104.06.

We would add multiple medical readings for pulmonary hypertension in 104.06B. We propose adding laboratory findings expressed in millimeters of mercury (mmHG) in 104.06B2, and we would add mean pulmonary artery pressure readings in 104.06B3.

We are proposing 104.06C, similar to the adult criterion 4.06D (*Congenital heart disease*), to evaluate children born with a single ventricle. Adding consideration of single ventricle to listing 104.06 enables seriously limited children to be identified earlier in the sequential evaluation process.

We would add 104.06E to evaluate exacerbations or complications of congenital heart disease occurring at least 30 days apart and resulting in at least three hospitalizations within a consecutive 12-month period. An impairment resulting in exacerbations or complications that require this many hospitalizations in 12 months will result in marked and severe functional limitations for children.[108] We would require these hospitalizations to be at least 30 days apart to ensure we are evaluating separate episodes of exacerbations or complications.

Proposed Removal of Listing 104.13—Rheumatic Heart Disease

We would remove and reserve listing 104.13 because rheumatic heart disease is a complication of rheumatic fever, which is rare in the United States due to widely available treatment with antibiotics.[109] [110] When complications of rheumatic fever result in rheumatic heart disease, and these complications last for 12 months or more, we would evaluate the complications under other cardiovascular listings, such as 104.02 (*Chronic heart failure*) or 104.05

[104] IOM. (2010), 171, 176.

[105] IOM. (2010), 173.

[106] 80 FR 19522 (2015).

[107] Based on SSA administrative data from FY 2019–2021, of all childhood claims with a primary impairment of congenital heart disease that met or medically equaled listing 104.06, approximately .2 percent cited these criteria. See Table A and B in supporting and related materials to this Docket for more information.

[108] IOM. (2010), 179.

[109] Beaudoin, A., Edison, L., Introcaso, C.E., Goh, L., Marrone, J., Mejia, A. . . ., & Van Beneden, C. (2015). Acute rheumatic fever and rheumatic heart disease among children—America Samoa, 2011–2012. Morbidity and Mortality Weekly Report, 64(20), 555–558. Retrieved from *https://www.cdc.gov/mmwr/pdf/wk/mm6420.pdf.*

[110] Yandrapalli, S., Tariq, S., Vuddanda, V.L.K., Sanaani, A., Solangi, Z., Anugu, V.R., . . . Aronow, W. (2017). In-hospital outcomes and hospitalizations for acute rheumatic heart disease: A United States national study. Journal of the American College of Cardiology, 69(11)(Suppl.), 1742. doi:10.1016/S0735–1097(17)35131–8.

(*Recurrent arrhythmias*). Rheumatic heart disease will still be addressed in the introductory text under 104.00F6 (*How will we evaluate chronic rheumatic fever or rheumatic heart disease?*).

Proposed Listing 104.16—Cardiac Allograft Vasculopathy

We propose to add listing 104.16 (*Cardiac allograft vasculopathy*) to evaluate a child who received a heart transplant and developed cardiac allograft vasculopathy (CAV). CAV may develop after heart transplantation and progress to a very serious condition with significant functional effects. The medical literature indicates that CAV is a leading cause of graft failure and mortality in pediatric heart transplant recipients.[111] To establish listing-level CAV for children, we would require only CAV documented by appropriate medically acceptable imaging, because pediatric CAV alone is disabling enough to result in marked and severe functional limitations for children

Specific Questions for the Public

While the public is welcome to comment on any aspect of this proposed rule, we are also seeking input on the following topics:

- Should any of the proposed listings for cardiovascular disorders be combined into one listing, or divided into multiple listings, to enable our adjudicators to more easily identify adults or children with impairments that are of listing-level severity? If you believe our listing categories create unnecessary administrative barriers for impairments that meet listing level severity, please tell us by submitting your comments and any supporting research or data.
- Are there changes in the medical terminology related to cardiovascular disorders that we should consider incorporating or clarifying in future revisions to the cardiovascular disorders listings? If you believe we should consider updating the medical terminology we use in our cardiovascular disorders listings, please tell us by submitting your comments and any supporting research or data.
- Do the frequencies and durations of exacerbations of cardiovascular disorders in this proposed rule adequately represent listing level severity for cardiovascular disorders? Are there other treatments and evidence we should consider when assessing listing-level severity including additional objective medical tests, for any of the proposed cardiovascular disorders listings? We encourage you to cite relevant research or data to support your comments.
- Are the proposed functional criteria for cardiovascular disorders sufficient for assessing listing level severity? Please provide specific suggestions along with supporting research and data for different criteria you would like SSA to consider.
- Did we not include any valuable information that should be included in the introductory text of the cardiovascular disorders listings? This text is intended to ease administrative burden for adjudicators, claimants, claimant representatives, and the public. Please submit specific comments, along with supporting research or data, about additional information to include in the introductory text.
- In proposed 4.02A1 (*Chronic heart failure*), we require systolic failure documented by appropriate medically acceptable imaging during a period of stability (not during an episode of exacerbation of heart failure), with left ventricular end diastolic dimension equal to or greater than 7.0 cm; or ejection fraction of 30 percent or less. If you believe we should require more than one evaluation to document the duration of an individual's chronic heart failure, please tell us by submitting your comments and any supporting research or data.
- In proposed 4.02A2 (*Chronic heart failure*), we require diastolic failure documented by appropriate medically acceptable imaging during a period of stability (not during an episode of exacerbation of heart failure), with left ventricular posterior wall plus septal thickness totaling 2.5 cm or greater, with an enlarged left atrium greater than or equal to 4.5 cm, *OR* left atrial volume index (LAVi) greater than or equal to 40 ml, BSA/m2 (milliliters to body surface area in squared meters). If you believe we should consider other measurements of chronic heart failure, please tell us by submitting your comments and any supporting research or data.
- In proposed 104.02A (*Chronic heart failure*), we require persistent tachycardia at rest measured twice within a consecutive 12-month period and at least 90 days apart documented by apical heart rate greater than or equal to the value in Table I. In proposed 104.02B, we require persistent tachypnea measured at least twice within a consecutive 12-month period and at least 90 days apart documented by respiratory rate greater than or equal to the value in Table II or markedly decreased exercise tolerance. If you believe our proposed requirement for at least two measurements of apical heart rate and respiratory rate under this listing is inconsistent with current medical practice or standards of care (*i.e.,* medical providers do not routinely repeat these measurements), please tell us by submitting your comments and any supporting research or data.
- In proposed 4.06A1 (*Congenital heart disease*), we require two measurements of hematocrit at least 90 days apart within a consecutive 12-month period instead of the current requirement for one measurement. If you would like to propose a different time frame during which these measures should occur, please submit comments and any supporting research or data.
- Are there alternatives to pulse oximetry testing that are reliable, non-invasive, and commonly used to measure chronic hypoxemia that we should consider incorporating into proposed listing criterion 4.06A3 (*Congenital heart disease*) and 104.06A3 (*Congenital heart disease*)? If you believe there are tests that fit into this category, please tell us by submitting your comments and any supporting research or data.
- At IOM's recommendation, we are proposing to add listing 4.07 (*Aortic valvular disease*) to provide evaluation criteria for symptomatic adult individuals with aortic valvular disease.[112] We currently evaluate aortic valvular disease under other cardiovascular disorders listings, which include requirements for exercise testing or repeated hospitalizations. If you disagree with proposed 4.07 (*Aortic valvular disease*), please tell us by submitting your comments and any supporting research or data.

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

The Act authorizes us to make rules and regulations and to establish necessary and appropriate procedures to implement them.[113]

How long would this proposed rule be effective?

If we publish this proposed rule as a final rule, it will remain in effect for five years after the date it becomes effective, unless we extend it, or revise and issue it again.

Rulemaking Analyses and Notices

We will consider all comments we receive on or before the close of

[111] Kindel, S.J. (2011).

[112] IOM. (2010), 195.

[113] Sections 205(a), 702(a)(5), and 1631(d)(1) of the Social Security Act.

business on the comment closing date indicated above. The comments will be available for examination in the rulemaking docket for this rule at the above address. We will file comments received after the comment closing date in the docket and will consider those comments to the extent practicable. However, we will not address untimely comments. We may publish a final rule at any time after close of the comment period.

Clarity of This Proposed Rule

Executive Order 12866, as supplemented by Executive Order 13563, requires each agency to write all rules in plain language. In addition to your substantive comments on this proposed rule, we invite your comments on how to make them easier to understand.

For example:

- Would more, but shorter, sections be better?
- Are the requirements in the rule clearly stated?
- Have we organized the material to suit your needs?
- Could we improve clarity by adding tables, lists, or diagrams?
- What else could we do to make the rule easier to understand?
- Does the rule contain technical language or jargon that is not clear?
- Would a different format make the rule easier to understand such as using different groupings and order of sections, headings, or paragraphing?

When will we start to use this rule?

We will not use this proposed rule until we evaluate public comments and publish a final rule in the **Federal Register**. All final rules we issue include an effective date. We will continue to use our current rule until that date. If we publish a final rule, we will include a summary of the relevant comments we received and an explanation of how we will apply the new rule.

Regulatory Procedures

Executive Order 12866, as Supplemented by Executive Order 13563

We consulted with the Office of Management and Budget (OMB) and determined that this proposed rule meets the criteria for a significant regulatory action under Executive Order 12866, as supplemented by Executive Order 13563. Therefore, OMB reviewed the rule.

We also determined that this proposed rule meets the plain language requirement of Executive Order 12866.

Executive Order 13132 (Federalism)

We analyzed this proposed rule in accordance with the principles and criteria established by Executive Order 13132 and determined that this proposed rule will not have sufficient federalism implications to warrant the preparation of a federalism assessment. We also determined that this proposed rule will not preempt any State law or State regulation or affect the States' abilities to discharge traditional State governmental functions.

Regulatory Flexibility Act

We certify that this proposed rule would not have a significant economic impact on a substantial number of small entities because it affects individuals only. Therefore, a regulatory flexibility analysis is not required under the Regulatory Flexibility Act, as amended.

Executive Order 13771

Based upon the criteria established in Executive Order 13771 and M–17–21 (Guidance Implementing E.O. 13771), we consider this rule a transfer rule with no more than *de minimis* costs. As such, it is exempt from requirements under E.O. 13771.

Anticipated Accounting Costs of This Proposed Rule

Anticipated Costs to Our Programs

Our Office of the Chief Actuary has developed estimates of the effects of implementing this proposed rule, which are presented in a memorandum attached to this NPRM as a supplementary document. The memorandum indicates the estimated annual changes in Old-Age, Survivors and Disability Insurance (OASDI) benefit payments and Federal Supplemental Security Income (SSI) payments over the 10-year period of fiscal years (FY) 2022–2031. The memorandum also provides details about the case study developed for the purpose of making these estimates, as well as changes since the time the case study was originally developed and conducted, that may have impacted the case study results.

In summary, based on the best available data, our Office of the Chief Actuary estimates that this proposed rule, assuming it is finalized and implemented for all disability decisions completed on or after April 1, 2023, would result in net increases of $308 million in scheduled OASDI benefit payments and $71 million in Federal SSI payments over the 10-year period of fiscal years (FY) 2022–2031.

Anticipated Administrative Costs to the Social Security Administration

The Office of Budget, Finance, and Management estimates a net administrative savings of less than 15 work years and $2 million annually.

Paperwork Reduction Act

This rule does not create any new or affect any existing collections and, therefore, does not require OMB approval under the Paperwork Reduction Act.

References

We consulted the following references when we developed this proposed rule:

Adams, E.E., & Wrightson, M.L. (2018). Quality of life with an LVAD: A misunderstood concept. *Heart & Lung, 47*(3), 177–183. doi:10.1016/j.hrtlng.2018.02.003.

Agarwal, S., Sud, K., & Shishehbor, M.H. (2016). Nationwide trends of hospital admission and outcomes among critical limb ischemia patients: From 2003–2011. *Journal of the American College of Cardiology, 67*(16), 1901–1913. doi:10.1016/j.jacc.2016.02.040.

Bavafa, H., Savin, S., & Terwiesch, C. (2019). Redesigning Primary Care Delivery: Customized Office Revisit Intervals and E-Visits. *https://dx.doi.org/10.21con39/ssrn.2363685.* Paper referenced by Bavafa: Schectman, G., G. Barnas, P. Laud, L. Cantwell, M. Horton, E.J. Zarling. 2005. Prolonging the return visit interval in primary care. *The American Journal of Medicine, 118*(4) 393–39.

Beaudoin, A., Edison, L., Introcaso, C.E., Goh, L., Marrone, J., Mejia, A., & Van Beneden, C. (2015). Acute rheumatic fever and rheumatic heart disease among children—America Samoa, 2011–2012. *Morbidity and Mortality Weekly Report, 64*(20), 555–558. Retrieved from *https://www.cdc.gov/mmwr/pdf/wk/mm6420.pdf.*

Berthelot-Richer, M., Pibarot, P., Capoulade, R., Dumesnil, J.G., Dahou, A., Thebault, C., . . . Clavel, M.-A. (2016). Discordant grading of aortic stenosis severity: Echocardiographic predictors of survival benefit associated with aortic valve replacement. *Journal of the American College of Cardiology: Cardiovascular Imaging, 9*(7), 797–805. doi:10.1016/j.jcmg.2015.09.026.

Bistola, V., Arfaras-Melainis, A., Polyzogopoulou, E., Ikonomidis, I., & Parissis, J. (2019). Inotropes in acute heart failure: From guidelines to practical use: Therapeutic options and clinical practice. *Cardiac Failure Review, 5*(3), 133–139. doi:10.15420/cfr.2019.11.2.

Blume, E.D., Rosenthal, D.N., Rossano, J.W., Baldwin, J.T., Eghtesady, P., Morales, D.L., . . . Kirklin, J.K.. (2016). Outcomes of children implanted with ventricular assist devices in the United States: First analysis of the pediatric interagency registry of mechanical circulatory support (PediMACS). *Journal of Heart*

and Lung Transplantation, 35(5), 578–584. doi:10.1016/j.healun.2016.01.1227.

Cacciapuoti, Fu., Scognamiglio, A., Paoli, V.D., Romano, C., & Cacciapuoti, Fe. (2012). Left atrial volume index as indicator of left ventricular diastolic dysfunction: Comparation between left atrial volume index and tissue myocardial performance index. *Journal of Cardiovascular Ultrasound, 20*(1), 25–29. doi:10.4250/jcu.2012.20.1.25.

Cedars, A., Benjamin, L., Burns, S.V., Novak, E., & Amin, A. (2017). Clinical predictors of length of stay in adults with congenital heart disease. *Heart (British Cardiac Society), 103*(16), 1258–1263. *https://doi.org/10.1136/heartjnl-2016-310841.*

Cha, Y.-M., Lee, G.K., Klarich, K.W., & Grogan, M. (2012). Premature ventricular contraction-induced cardiomyopathy: A treatable condition. *Circulation: Arrhythmia and Electrophysiology, 5*(1), 229–236. doi:10.1161/CIRCEP.111.963348.

Chen, J., Dharmarajan, K., Wang, Y., & Krumholz, H.M. (2013). National trends in heart failure hospital stay rates, 2001 to 2009. *Journal of the American College of Cardiology, 61*(10), 1078–1088. doi:10.1016/j.jacc.2012.11.057.

Cheng, R.K., Cox, M., Neely, M.L., Heidenreich, P.A., Bhatt, D.L., Eapen, Z.J., . . . Fonarow, G.C. (2014). Outcomes in patients with heart failure with preserved, borderline, and reduced ejection fraction in the Medicare population. *American Heart Journal, 168*(5), 721–730. doi:10.1016/j.ahj.2014.07.008.

Ciarka, A., Edwards, L., Nilsson, J., Stehlik, J., & Lund, L.H. (2017). Trends in the use of mechanical circulatory support as a bridge to heart transplantation across different age groups. *International Journal of Cardiology, 231,* 225–227. doi:10.1016/j.ijcard.2016.10.049.

Cohen, S., & Marelli, A. (2016). Evolving heart transplantation across the lifespan: A growing population of adults with congenital heart disease. *Archives of Cardiovascular Disease, 109*(10), 511–513. doi:10.1016/j.acvd.2016.05.001.

Cohen-Solal, A., Laribi, S., Ishihara, S., Vergaro, G., Baudet, M., Logeart, D., . . . Seronde, M.-F. (2015). Prognostic markers of acute decompensated heart failure: The emerging roles of cardiac biomarkers and prognostic scores. *Archives of Cardiovascular Disease, 108*(1), 64–74. doi:10.1016/j.acvd.2014.10.002.

Desai, A.S. (2013). Are serial BNP measurements useful in heart failure management? Serial natriuretic peptide measurements are not useful in heart failure management: The art of medicine remains long. *Circulation, 127*(4), 509–516. doi:10.1161/CIRCULATIONAHA.112.120493.

Desai, R.V., Guichard, J.L., Mujib, M., Ahmed, M.I., Feller, M.A., Fonarow, G.C., . . . Ahmed, A. (2013). Reduced right ventricular ejection fraction and increased mortality in chronic systolic heart failure patients receiving beta-blockers: Insights from the BEST trial. *International Journal of Cardiology, 163*(1), 61–67. doi:10.1016/j.ijcard.2011.05.051.

Dukes, J.W., Dewland, T.A., Vittinghoff, E., Mandyam, M.C., Heckbert, S.R., Siscovick, D.S., . . . Marcus, G.M. (2015). Ventricular ectopy as a predictor of heart failure and death. *Journal of the American College of Cardiology, 66*(2), 101–109. doi:10.1016/j.jacc.2015.04.062.

Fearon, W.F. (2014). Percutaneous coronary intervention should be guided by fractional flow reserve measurement. *Circulation, 129*(18), 1860–1870. doi:10.1161/CIRCULATIONAHA.113.004300.

Fearon, W.F., De Bruyne, B., & Pijlis, N.H. (2016). Fractional flow reserve in acute coronary syndromes. *Journal of the American College of Cardiology, 68*(11), 1192–1194. doi:10.1016/j.jacc.2016.07.713.

Fihn, S.D., Gardin, J.M., Abrams, J., Berra, K., Blankenship, J.C., Dallas, A.P., . . . Williams, S.V. (2012). 2012 ACCF/AHA/ACP/AATS/PCNA/SCAI/STS Guideline for the diagnosis and management of patients with stable ischemic heart disease: Executive summary: A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, and the American College of Physicians, American Association for Thoracic Surgery, Preventive Cardiovascular Nurses Association, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *Circulation, 126*(25), 3097–3137. doi:10.1161/CIR.0b013e3182776f83.

Fletcher, G.F., Ades, P.A., Kligfield, P., Arena, R., Balady, G.J., Bittner, V.A., . . . Williams, M.A. (2013). Exercise standards for testing and training: A scientific statement from the American Heart Association. *Circulation, 128*(8), 873–934. doi:10.1161/CIR.0b013e31829b5b44.

Frigola-Capell, E., Comin-Colet, J., Davins-Miralles, J., Gich-Saladich, I., Wensing, M., & Verdú-Rotellar, J.M. (2012). Trends and predictors of hospitalization, readmissions and length of stay in ambulatory patients with heart failure. *Revista Clinica Espanola, 213*(1), 1–7. doi:10.1016/j.rce.2012.10.006.

Henry, T.D., Satran, D., Hodges, J.S., Johnson, R.K., Poulose, A.K., Campbell, A.R., . . . Traverse, J.H. (2013). Long-term survival in patients with refractory angina. *European Heart Journal, 34*(34), 2683–2688. doi:10.1093/eurheartj/eht165.

Höbaus, C., Roller-Wimsberger, Schernthaner, G.-H. (2016). Peripheral arterial disease and loss of physical function: Just two old friends? *Atherosclerosis, 257,* 246–247. doi:10.1016/j.atherosclerosis.2016.12.015.

Hua, M., Gong, M.N., Brady, J., & Wunsch, H. (2015). Early and late unplanned rehospitalizations for survivors of critical illness. *Critical Care Medicine, 43*(2), 430–438. doi:10.1097/CCM.0000000000000717.

Institute of Medicine. (2010). *Cardiovascular Disability: Updating the Social Security Listings.* Washington, DC: The National Academies Press.

Jackson SL, Tong X, King RJ, et al. (2018). National Burden of Heart Failure Events in the United States, 2006 to 2014. *Circulation: Heart Failure.* 2018 Dec;11(12):e004873. DOI: 10.1161/circheartfailure.117.004873. PMID: 30562099; PMCID: PMC6424109.

Januzzi, J.L., Chen-Tournoux, A.A., Christenson, R.H., Doros, G., Hollander, J.E., Levy, P.D., . . . Gaggin, H.K. (2018). N-terminal in pro-B-type natriuretic peptide in the emergency department: The ICON–RELOADED Study. *Journal of the American College of Cardiology, 71*(11), 1191–1200. doi:10.1016/j.jacc.2018.01.021.

Koene, R.J., Adkisson, W.O., & Benditt, D.G. (2017). Syncope and the risk of sudden cardiac death: Evaluation, management, and prevention. *Journal of arrhythmia, 33*(6), 533–544. *https://doi.org/10.1016/j.joa.2017.07.005.*

Khan, T.Z., Pottle, A., Pennell, D.J., & Barbir, M.S. (2015). The impact of lipoprotein apheresis in patients with refractory angina and raised lipoprotein(a): Objectives and methods of a randomized controlled trial. *Atherosclerosis Supplements, 18,* 103–108. doi:10.1016/j.atherosclerosissup.2015.02.019.

Kim, Y., Gani, F., Canner, J.K., Margonis G.A., Makary, M.A., Schneider, E.B., & Pawlik, T.M. (2016). Hospital readmission after multiple major operative procedures among patients with employer provided health insurance. *Surgery, 160*(1), 178–190. doi:10.1016/j.surg.2016.01.025.

Kindel, S.J., & Pahl, E. (2011). Cardiac allograft vasculopathy in children—treatment challenges. *Progress in Pediatric Cardiology, 32*(1), 37–42. doi:10.1016/j.ppedcard.2011.06.008.

Kindel, S.J., Law, Y.M., Chin, C., Burch, M., Kirklin. J.K., Naftel, D.C., . . . Pahl, E. (2015). Improved detection of cardiac allograft vasculopathy: A multi-institutional analysis of functional parameters in pediatric heart transplant recipients. *Journal of the American College of Cardiology, 66*(5), 547–557. doi:10.1016/j.jacc.2015.05.063.

Kobashingawa, J.A. (2015). The changing face of first-year intravascular ultrasonography in heart transplantation. *Journal of the American College of Cardiology: Heart Failure, 3*(12), 954–955. doi:10.1016/j.jchf.2015.09.004.

Khoo, C., Chakrabarti, S., Arbour, L. & Krahn, A.D. (2013). Recognizing life-threatening causes of syncope. *Cardiology Clinics, 31*(1), 51–66. doi:10/1016/j.cci.2012.10.005.

Kyriakou, M., & Kiff, P.F. (2016). Prognosis of the comorbid heart failure and anemia: A systematic review and meta-analysis. *Clinical Trials and Regulatory Science in Cardiology, 16,* 12–21. doi:10.1016/j.ctrsc.2016.01.008.

Marine, J.E. (2013). ECG features that suggest a potentially life-threatening arrhythmia as the cause of syncope. *Journal of*

Electrocardiology, 46(6), 561–568. doi:10.1016/j.jelectrocard.2013.07.008.
Malotte, K., Saguros, A., & Groninger, H. (2018). Continuous cardiac inotropes in patients with end-stage heart failure: An evolving experience. *Journal of Pain Symptom Management, 55*(1), 159–163. doi:10.1016/j.jpainsymman.2017.09.026.
Mechem, C.C. (2014). Pulse oximetry. In P.E. Parsons (Ed.), *UpToDate* (Jan. 2014). Retrieved from *https://www.uptodate.com/contents/pulse-oximetry.*
Mehra, M.R., Crespo-Leiro, M.G., Dipchand, A., Ensminger, S.M., Hiemann, N.E., Kobashigawa, J.A., . . . Uber, P.A. (2010). International Society of Heart and Lung Transplantation working formulation of a standardized nomenclature for cardiac allograft vasculopathy—2010. *Journal of Heart and Lung Transplantation, 29*(7), 717–727. doi:10.1016/j.healun.2010.05.017.
Nakano, Y. & Wataru, S. (2017). Syncope in patients with inherited arrhythmias. *Journal of Arrhythmia, 33*(6), 572–578. doi:10.1016/j.joa.2017.07.007.
National Academies of Sciences, Engineering, and Medicine. (2018). *Health-care utilization as a proxy in disability determination.* Washington, DC: The National Academies Press. doi:10.17226/24969.
Nieminen, M.S., Dickstein, K., Fonseca, C., Serrano, J.M., Parissis, J., Fedele, F., . . . Zymliński, R. (2015). The patient perspective: Quality of life in advanced heart failure with frequent hospitalizations. *International Journal of Cardiology, 191,* 256–264. doi:10.1016.j.ijcard.2015.04.235.
Nixon, J.L., Benuzillo, J., McCubrey, R.O., Budge, D., Rasmusson, K., Brush, S., . . . Kfoury, A. (2013). Is 30-day readmission a valid measure for quality care or simply an indicator of severe disease in patients discharged after heart failure hospitalization? *Journal of Cardiac Failure, 19*(8)(Suppl.), S80–S81. doi:10.1016/j.cardfail.2013.06.257.
Nombela-Franco, L., del Trigo, M., Morrison-Polo, G., Veiga, G., Jimenez-Quevedo, P., Altisent, O. A.-J., . . . Rodés-Cabau, J. (2015). Incidence, causes, and predictors of early (<30 days) and late unplanned hospital readmissions after transcatheter aortic valve replacement. *Journal of the American College of Cardiology: Cardiovascular Interventions, 8*(13), 1748–1757. doi:10.1016/j.jcin.2015.07.022.
Okada, K., Kitahara, H., Yang, H., Tanaka, S., Kobayashi, Y., Kimura, T., . . . Fearon, W.F. (2015). Paradoxical vessel remodeling of the proximal segment of the left anterior descending artery predicts long-term mortality after heart transplantation. *Journal of the American College of Cardiology: Heart Failure, 3*(12), 945–952. doi:10.1016/j.jchf.2015.07.013.
Oster, M.E., & Kochilas, L.K. (2016). Screening for critical congenital heart disease. *Clinics in Perinatology, 43*(1), 73–80. doi:10.1016/j.clp.2015.11.005.
Park, S.H., Jeon, K.-H., Lee, J.M., Nam, C.-W., Doh, J.-H., Lee, B.-K., . . . Koo, B.-K. (2015). Long-term clinical outcomes of fractional flow reserve-guided versus routine drug-eluting stent implantation in patients with intermediate coronary stenosis: Five-year clinical outcomes of DEFER–DES trial. *Circulation: Cardiovascular Interventions, 8*(12), e002442. doi:10.1161/CIRCINTERVENTIONS.115.002442.
Patel, D.A., Lavie, C.J., Milani, R.V., Shah, S., & Gilliland, Y. (2009). Clinical implications of left atrial enlargement: A review. *The Ochsner Journal, 9*(4), 191–196.
Patterson, C.C., Blankenberg, S., Ben-Shlomo, Y., Heslop, L., Bayer, A., Lowe, G., . . . Yarnell, J. (2015). Which biomarkers are predictive specifically for cardiovascular or for non-cardiovascular mortality in men? Evidence from the Caerphilly Prospective Study (CaPS). *International Journal of Cardiology, 201,* 113–118. doi:10.1016/j.ijcard.2015.07.106.
Pijls, N.H., Fearon, W.F., Tonino, P.A., Siebert, U., Ikeno, F., Bornschein, B., . . . De Bruyne, B. (2010). Fractional flow reserve versus angiography for guiding percutaneous coronary intervention in patients with multivessel coronary artery disease: 2-year follow-up of the FAME (Fractional Flow Reserve Versus Angiography for Multivessel Evaluation) study. *Journal of the American College of Cardiology, 56*(3), 177–184. doi:10.1016/j.jacc.2010.04.012.
Poredoš P, Jezovnik M, Kalodiki E. Medical management of patients with peripheral arterial disease. Int Angiol. 2015 Feb;34(1):75–93. Epub 2014 Jun 11. PMID: 24916346.
Reynolds, K., Butler, M.G., Kimes, T.M., Rosales, A.G., Chan, W., & Nichols, G.A. (2015). Relation of acute heart failure hospital length of stay to subsequent readmission and all-cause mortality. *American Journal of Cardiology, 116*(3), 400–405. doi:10.1016/j.amjcard.2015.04.052.
Rodés-Cabau, J. (2015). Incidence, causes, and predictors of early (<30 days) and late unplanned hospital readmissions after transcatheter aortic valve replacement. *Journal of the American College of Cardiology: Cardiovascular Interventions, 8*(13), 1748–1757. doi:10.1016/j.jcin.2015.07.02.
Rosenthal, D.N., Almond, C.S., Jaquiss, R.D., Peyton, C.E., Auerbach, S.R., Morales, D.R., . . . Blume, E.D. (2016). Adverse events in children implanted with ventricular assist devices in the United States: Data from the Pediatric Interagency Registry for Mechanical Circulatory Support (PediMACS). *The Journal of Heart and Lung Transplantation, 35*(5), 569–577. doi:10.1016/j.healun.2016.03.005.
Roston, T.M., Tran, D.T., Sanatani, S., Sandhu, R., Sheldon, R., & Kaul, P. (2017). A population-based study of syncope in the young. *Canadian Journal of Cardiology, 34*(2), 195–201. doi:10.1016/j.cjca.2017.12.006.
Runge, M.S., Patterson, C., Stouffer, G.A., & Netter, F. H. (2010). *Netter's Cardiology* (2nd ed.). Philadelphia, PA: Saunders Elsevier.
Salamanca-Bautista, P., Conde-Martel, A., Aramburu-Bodas, Ó., Formiga, F., Trullàs, J.C., Quesada-Simón, M.A., . . . Montero-Pérez-Barquero, M. (2016). Precipitating factors of heart failure admission: Differences related to age and left ventricular ejection fraction. *International Journal of Cardiology, 219,* 150–155. doi:10.1016/j.ijcard.2016.06.040.
Santangeli, P., & Marchlinski, F.E. (2015). Ventricular ectopy as a modifiable risk factor for heart failure and death: ''Déjà vu all over again'' may be a good thing. *Journal of the American College of Cardiology, 66*(2), 110–112. doi:10.1016/j.jacc.2015.05.031.
Shlofmitz, E., and Jeremias, A. (2017). *FFR in 2017: Current Status in PCI Management–American College of Cardiology.* Available online at: *https://www.acc.org/latest-in-cardiology/articles/2017/05/25/08/34/ffr-in-2017-current-status-in-pci-management* (accessed September 17, 2021).
Sheikh, A., Patel, N.J., Nalluri, N., Agnihotri, K., Spagnola, J., Patel, A., . . . Paydak, H. (2015). Trends in hospitalization for atrial fibrillation: Epidemiology, cost, and implications for the future. *Progress in Cardiovascular Diseases, 58*(2), 105–116. doi:10.1016/j.pcad.2015.07.002.
Siracuse, J.J., Gill, H.L., Jones, D.W., Schneider, D.B, Connolly, P.H., Parrack, I., . . . Meltzer, A.J. (2014). Risk factors for protracted preoperative length of stay after lower extremity bypass for critical limb ischemia. *Annals of Vascular Surgery, 28*(6), 1432–1438. doi:10.1016/j.avsg.2013.12.027.
Sorensen, L.L, Liang, H.-L., Pinheiro, A., Hilser, A., Dimaano, V., Olsen, N.T., . . . Abraham, T.P. (2017). Safety profile and utility of treadmill exercise in patients with high-gradient hypertrophic cardiomyopathy. *American Heart Journal, 184,* 47–54. doi:10.1016/j.ahj.2016.10.010.
Stack, S.W., & Berger, S.A. (2009). The effects of high hematocrit arterial flow—A phenomenological study of health risk implications. *Chemical Engineering Science, 64*(22), 4701–4706. doi:10/1016/j.ces.2009.07.017.
Stout, K.K., Daniels, C.J., Aboulhosn, J.A., Bozkurt, B., Broberg, C.S., Colman, J.M., . . . Van Hare, G.F. (2019). 2018 AHA/ACC Guideline for the management of adults with congenital heart disease: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *Journal of the American College of Cardiology, 73*(12), e81-e192. doi:10.1016/j.jacc.2018.08.1029.
Tebaldi, M., Biscaglia, S., Pecoraro, A., Fineschi, M., & Campo, G. (2016). Fractional flow reserve implementation in daily clinical practice: A European survey. *International Journal of Cardiology, 207,* 206–207. doi:10.1016/j.ijcard.2016.01.097.
Thrush, P.T., & Canter, C.E. (2015). Looking to the future of ventricular assist devices in pediatric cardiomyopathy. *Progress in Pediatric Cardiology, 39*(1), 21–23. doi:10.1016/j.ppedcard.2015.01.003.

Uszko-Lencer, N.H., Frankenstein, L., Spruit, M.A., Maeder, M.T., Gutmann, M., Muzzarelli, S., . . . Brunner-La Rocca, H.-P. (2017). Predicting hospitalization and mortality in patients with heart failure: The BARDICHE-index. *International Journal of Cardiology, 227,* 901–907. doi:10.1016/j.ijcard.2016.11.122.

Versteeg, H., Hoogwegt, M.T., Hansen, T.B., Pedersen, S.S., Zwisler, A.-D., & Thygesen, L.C. (2013). Depression, not anxiety, is independently associated with 5-year hospitalizations and mortality in patients with ischemic heart disease. *Journal of Psychosomatic Research, 75*(6), 518–525. doi:10.1016/j.jpsychores.2013.10.005.

Wei, B., Qian, C., Fang, Q., & Wang, Y. (2016). The prognostic value of peripheral artery disease in heart failure: Insights from a meta-analysis. *Heart, Lung and Circulation, 25*(12), 1195–1202. doi:10.1016/j.hlc.2016.04.002.

Yandrapalli, S., Tariq, S., Vuddanda, V.L.K., Sanaani, A., Solangi, Z., Anugu, V.R., . . . Aronow, W. (2017). In-hospital outcomes and hospitalizations for acute rheumatic heart disease: A United States national study. *Journal of the American College of Cardiology, 69*(11)(Suppl.), 1742. doi:10.1016/S0735–1097(17)35131–8.

Yu, P.-J., Cassiere, H.A., Fishbein, J., Esposito, R.A., & Hartman, A.R. (2016). Outcomes of patients with prolonged intensive care unit length of stay after cardiac surgery. Journal of Cardiothoracic and Vascular Anesthesia, 30(6), 1550–1554. doi:10.1053/j.jvca.2016.03.145.

Ziberszac, R., Gabriel, H., Schemper, M., Laufer, G., Maurer, G., & Rosenhek, R. (2017). Asymptomatic severe aortic stenosis in the elderly. *Journal of the American College of Cardiology: Cardiovascular Imaging, 10*(1), 43–50. doi:10.1016/j.jcmg.2016.05.015.

We will make these references available to you for inspection if you are interested in reading them. Please make arrangements with the contact person shown in this preamble if you would like to review any reference materials.

(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.

The Acting Commissioner of Social Security, Kilolo Kijakazi, Ph.D., M.S.W., having reviewed and approved this document, is delegating the authority to electronically sign this document to Faye I. Lipsky, who is the primary Federal Register Liaison for the Social Security Administration, for purposes of publication in the **Federal Register**.

Faye I. Lipsky,

Federal Register Liaison, Office of Legislation and Congressional Affairs, Social Security Administration.

For the reasons set forth in the preamble, we propose to amend subpart P of part 404 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 (h)–(j), 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 (h)–(j), 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).

■ 2. Amend appendix 1 to subpart P of part 404 by:
■ a. Revising item 5 of the introductory text before part A;
■ b. Revising the body system name for section 4.00 in the table of contents;
■ c. Revising the heading of 4.00A, the heading of 4.00A1, the introductory text of 4.00A1b, and 4.00A1b(iv), 4.00A2, and the first sentence of 4.00A3a; and adding two sentences to 4.00A3b, and two sentences to 4.00A3c; and removing 4.00A3f;
■ d. Revising the heading of 4.00B, the first sentence of 4.00B1, the third sentence of 4.00B2, the second and fourth sentences of 4.00B3(a), the second sentence of 4.00B3(b), and the first sentence of 4.00B5;
■ e. Revising the heading of 4.00C, the second sentence of 4.00C3(c), 4.00C6(a)(i), 4.00C7(b) through (d), 4.00C8(d)(iv), 4.00C8(e), and 4.00C9(a); removing the third sentence of 4.00C15(a), revising 4.00C15(b), removing 4.00C15(c); and revising the first two sentences of 4.00C16;
■ f. Revising the heading of 4.00D; adding new fourth and fifth sentences to 4.00D1a; revising the second sentence of 4.00D1b; adding a third sentence to 4.00D1b; revising 4.00D2(a)(i) through (iii), the first sentence of 4.00D2(b), the second sentence of 4.00D2(b)(i), the second sentence of 4.00D2(b)(ii), the third sentence of 4.00D3, 4.00D4(b) and 4.00D4(c) and the first, second, and fifth sentences of 4.00D4(d); and adding 4.00D4(e);
■ g. Revising the heading of 4.00E, 4.00E2b; the first sentence of 4.00E5, 4.00E7(b)(i)–(ii); and adding 4.00E7(b)(iii);
■ h. Revising the first three sentences of 4.00E8, and4.00E9(b) through (f), and removing 4.00E9(g) and (h);
■ i. Revising the heading of 4.00F; adding a new sentence to 4.00F1, revising the first sentence of 4.00F3(a), 4.00F4(a), and the second and fourth sentences of 4.00F4(b);
■ j. Revising the heading of 4.00G; adding a fourth sentence to 4.00G1, revising 4.00G2, the first two sentences of 4.00G4(b), 4.00G6, and the fourth sentence of 4.00G7(b);
■ k. Redesignating 4.00H and I as 4.00I and J, respectively
■ l. Adding a new 4.00H;
■ m. Revising the heading of 4.00I; 4.00I1, removing 4.00I2, and redesignating 4.00I3 through 5 as 4.00I2 through 4;
■ n. Revising 4.00I2, 4.00I3, 4.00I4(a) and (d), adding a new 4.00I5, and revising the third sentence of 4.00I8b;
■ o. Revising the heading of 4.00J, 4.00J1, the first sentence of 4.00J2, redesignating 4.00J3 as 4.00K, and adding new 4.00J3;
■ p. Revising 4.00K;
■ q. Revising listings 4.01 and 4.02, adding and reserving listing 4.03, revising listings 4.05 and 4.06, adding listing 4.07, adding listing 4.08, revising 4.09 through 4.12, adding and reserving listings 4.13 through 4.15, and adding listing 4.16.
■ r. Revising the heading of 104.00A; the heading of 104.00A1; the introductory text of 104.00A1(b), 104.00A1(b)(iv), and 104.00A2; and the first sentence of 104.00A3(a), and adding two sentences to 104.00A3(b), adding two sentences to 104.00A3(c), and removing 104.00A3(f) and (g);
■ s. Revising the heading of 104.00B; the first sentence of 104.00B1; the third sentence of 104.00B2; the second and fourth sentences of 104.00B3(a); the second sentence of 104.00B3(b); 104.00B4(a)(i); the first and third sentences of 104.00B5; the heading of 100.04B7; the second sentence of 100.04B7(a); and the first sentence of 104.00B7(b);
■ t. Revising the heading of 104.00C; the heading of 104.00C1, and the first sentence of 104.00C1a; adding two sentences to 104.00C1a; revising 104.00C1b; 104.00C2(a);.00C2(b), and the second sentence of 104.00C2(b)(iii); and adding 104.00C4;
■ u. Revising the heading of 104.00D; 104.00D1, 104.00D1d, and 104.00D2; and adding 104.00D3 and 104.00D4;
■ v. Revising the heading of 104.00E; adding a new sentence to the end of 104.00E1; revising the fourth and fifth sentences of 104.00E4(a), and the fourth sentence of 104.00E4(b);

■ w. Revising the heading of 104.00F; the last sentence of 104.00F1, the first sentence of 104.00F2; removing the fourth through seventh sentences of 104.00F3; adding a new 104.00F3a and 104.00F3b; revising 104.00F4, 104.00F5(a), 104.00F5(d), 104.00F6, and 104.00F9b; and adding 104.00F11;
■ x. Revising the heading of 104.00G; 104.00G1, the first sentence of 104.00G2; redesignating 104.00G3 as 104.00H, 104.00G3(a) as 104.00H1, and 104.00G3(b) as 104.00H2; and adding a new 104.00G3;
■ y. Revising 104.00H; and
■ z. Revising listings 104.01,104.02; adding and reserving 104.03 and 104.04; revising 104.05 and 104.06; adding and reserving 104.07 and 104.08; revising 104.09; adding and reserving listings 104.10 through 104.12; removing and reserving listing 104.13; adding and reserving listings 104.14 and 104.15: and adding listing 104.16.

The additions and revision to read as follows:

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

* * * * *

5. Cardiovascular Disorders (4.00 and 104.00) [DATE 5 YEARS FROM THE EFFECTIVE DATE OF THE FINAL RULE].

* * * * *

Part A

* * * * *

4.00 Cardiovascular Disorders.

* * * * *

4.00 Cardiovascular Disorders

A. *How do we define cardiovascular disorders and cardiovascular terms?*

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

a. * * *

b. Cardiovascular disorders result from one or more of four consequences of heart disease:

(i) * * *

(ii) * * *

(iii) * * *

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

c. * * *

2. *What do we consider in evaluating cardiovascular disorders?* The listings in this section describe cardiovascular disorders based on the medical and other evidence, including response to a regimen of prescribed treatment and functional limitations.

3. * * *

a. *Medical consultant* is a person defined in §§ 404.1616(a) and 416.1016(a) of this chapter. * * *

b. * * * By ''exceptions,'' we mean brief periods when the required finding(s) is greatly reduced or gone. These periods are so brief or inconsequential, the required finding(s) remains a factor in the person's condition.

c. * * * By ''improvement of sufficient duration,'' we mean the finding is greatly reduced or not present for long enough that the required finding(s) is no longer a factor in the person's condition.

* * * * *

f. [Removed]

B. *What documentation do we need to evaluate cardiovascular disorders?*

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular disorder.

* * * * *

2. *Why is a longitudinal clinical record important?* * * * Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your medical source(s). * * *

3. * * *

a. * * * In this situation, we will base our evaluation on the current evidence we have. * * * However, we may find you disabled because you have another impairment(s) that, in combination with your cardiovascular disorder, medically equals a listing or based on consideration of your residual functional capacity and age, education, and work experience.

b. * * * In rare instances when there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the existence, severity, and duration of your impairment.

* * * * *

5. *Will we purchase any studies?* In appropriate situations, we may purchase studies necessary to substantiate the existence of a medically determinable impairment or to document the severity of your impairment, generally after we have evaluated the evidence we already have.

* * *

* * * * *

C. *How do we use cardiovascular test results?*

* * * * *

3. * * *

c. * * * In this test, you walk on a treadmill, usually for a specified period of time, and the person who administers the test measures the effect of exercise on the flow of blood in your legs, usually by using ultrasound. * * *

* * * * *

6. * * *

a. * * *

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

* * * * *

7. * * *

a. * * *

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

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

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

* * * * *

8. * * *

d. * * *

(iv) Percutaneous transluminal coronary angioplasty (PTCA) or percutaneous coronary intervention (PCI) with or without stenting.

e. If you are deconditioned after an extended period of bedrest or inactivity and could improve with activity, or if you are in acute heart failure and are expected to improve with treatment, we will wait an appropriate period of time until you are ready and there are no medical reasons that prevent us from purchasing an exercise test.

9. * * *

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

* * * * *

15. * * *

a. * * *

b. Cardiac catheterization reports commonly include evaluation of coronary artery size and flow patterns, pressures in the left and right side of the heart, and evaluation of wall motion and ejection fraction, as well as chamber size. Also more routinely included in the catheterization report is fractional flow reserve (FFR), which is an objective measure of flow across across an obstruction. FFR also helps define the adequacy of collateral flow that directly affects function in ischemic heart disease.

16. *What details should exercise Doppler test reports contain?* The reports of exercise Doppler tests must describe the level of exercise; for example, the speed and grade of the treadmill settings, the duration of exercise, changes in the person's condition during exercise, and the reasons for stopping exercise if the expected level of exercise was not attained. These reports must also provide the blood pressures at the ankle and other pertinent sites measured after exercise, and also provide the time required for the systolic blood pressure to return toward or to the pre-exercise level. * * *

* * * * *

D. *How do we evaluate chronic heart failure?*

1. * * *

a. * * * Ejection fraction in heart failure is a continuum ranging from low ejection

fraction due to muscle dysfunction to preserved ejection fraction resulting from high intracardiac pressures. We consider heart failure to be chronic when the condition persists or recurs over time despite treatment. * * *

b. * * * If the CHF is the result of primary pulmonary hypertension secondary to disease of the lung, we evaluate your impairment under the listings in 3.00 (for example, 3.09) or 4.00, as appropriate. For the purposes of 4.02B3, a finding of elevated B-type natriuretic peptide (BNP) or N-terminal pro-B-type natriuretic peptide (NT-pro-BNP) in the blood assists in differentiating chronic heart failure from non-heart failure symptoms.

2. * * *

a. * * *

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

(ii) An LVEDD equal to or greater than 7.0 cm, *or* an EF of 30 percent or less during a period of stability (that is, not during an episode of acute heart failure) may be associated clinically with systolic dysfunction.

(iii) LAVi is measured in milliliters (ml) indexed to body surface area (BSA) measured in squared meters (m^2). Indexing is a method of standardizing measurements to different body sizes. Diastolic dysfunction may be clinically associated with LAVi of 40 ml, BSA/m^2 or greater. The imaging report will contain a measurement for the left atrium volume. The index is calculated by dividing the left atrium volume by BSA.

* * * * *

b. Your medical history and physical examination should describe characteristic symptoms and signs of pulmonary or systemic congestion (fluid retention) or of limited cardiac output associated with the abnormal findings on appropriate medically acceptable imaging. * * *

(i) * * * People with CHF may also experience shortness of breath upon lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). * * *

(ii) * * * However, these signs need not be found on all examinations because congestion may be controlled by prescribed treatment or may not be present at the time of evaluation.

3. *Is it safe for you to have an ETT if you have CHF?* * * * ETT has been used safely in people with CHF. Therefore, we may purchase an ETT for evaluation under 4.02B2 if an MC, preferably one experienced in the care of patients with cardiovascular disease, determines that the test poses no significant risk to you. * * *

4. * * *

a. * * *

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

c. In 4.02B1, we follow a two-part process to evaluate your impairment. Your impairment must satisfy the requirements in the first part of this process before we will move to the second part.

(i) Your impairment satisfies the first part if a medical source has concluded that the performance of an ETT would present a significant risk to you. This medical source, such as a cardiologist, may be providing your care. If your case record does not include a conclusion from a medical source that an ETT would present a significant risk to you, an MC as defined in 4.00A3a may make such a conclusion if evidence in your case record supports it.

(ii) In the second part of the process, we will evaluate activities of daily living (ADL). ADLs include, but are not limited to, such activities as doing household chores, grooming and hygiene, shopping at a grocery store, taking public transportation, or paying bills. We will assess whether you have persistent symptoms of chronic heart failure (for example, easy fatigue, weakness, shortness of breath, or chest discomfort) at rest or with activity that very seriously limit your ability to perform ADLs independently, appropriately, effectively, and on a sustained basis. Even if you are able to perform some ADLs, we may find your ability is very seriously limited and that your impairment satisfies the second part of the evaluation.

d. Listing 4.02B2b requires a decrease in systolic blood pressure below the baseline level or below any systolic pressure reading recorded during exercise. We have this requirement because, normally, systolic blood pressure and heart rate increase gradually with exercise. * * * Also, some people with increased sympathetic responses because of deconditioning or apprehension may increase their systolic blood pressure and heart rate above their baseline level just before and early into exercise. * * *

e. *How do we evaluate CHF treated with a mechanical circulatory support device?* We use 4.02D1 to evaluate CHF treated with an implanted mechanical circulatory support device (MCSD), such as a left ventricle assistive device (LVAD) or a right ventricle assistive device (RVAD). Implanted MCSDs are intended for long-term circulatory support in helping the heart pump blood. For the purposes of 4.02D1, an MCSD does not include extracorporeal membrane oxygenation (ECMO). Although ECMO is a form of mechanical circulatory support, we do not include it in 4.02D1 because ECMO is intended only for short-term circulatory support (maximum 30 days), used in a setting of imminent or actual cardiac arrest.

E. *How do we evaluate ischemic heart disease*?

1. * * *

2. * * *

a. * * *

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

* * * * *

5. *What is anginal equivalent?* Often, people with IHD will complain of shortness of breath (dyspnea) on exertion without chest pain or discomfort. * * *

* * * * *

7. * * *

a. * * *

b. * * *

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

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

(iii) People who have diabetes mellitus with neuropathy. People with diabetes mellitus can have a higher threshold for pain because of the neuropathy and may not feel chest pain or discomfort from cardiac ischemia.

* * * * *

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

9. * * *

a. * * *

b. In 4.04A, we need evidence, such as an ECG interpretation, from an acceptable medical source who reviewed your ETT findings and found them positive for ischemia. These ETT findings may include ECG tracings or systolic blood pressure measurements. If your case record does not have such an interpretation from an acceptable medical source, an MC, as defined in 4.00A3a, may review your ETT findings and interpret them as being positive for ischemia if evidence in your case record supports it.

(i) ETT findings may show the classically accepted changes in ECG tracings of horizontal or down sloping ST depression or of ST elevation. For example, ECG tracings may show horizontal or down sloping depression, in the absence of digitalis glycoside treatment or hypokalemia, of the ST segment of at least –0.10 millivolts (-1.0 mm) in at least three consecutive complexes that are on a level baseline in any lead other than a VR, and depression of at least –0.10 millivolts last for at least 1 minute of

recovery. Alternatively, the ECG tracings may show at least 0.10 millivolt (1 mm) ST elevation above resting baseline in non-infarct leads during both exercise and 1 or more minutes of recovery.

(ii) ETT findings may also show a decrease of 10 mmHg or more in systolic pressure below the baseline systolic blood pressure or the preceding systolic pressure measured during exercise due to left ventricular dysfunction, despite an increase in workload. This finding is the same finding required in 4.02B2b. See 4.00D4d for full details.

c. In 4.04C, each ischemic episode must result in an unplanned hospitalization. Examples of ischemic episodes that may result in unplanned hospitalizations include unplanned revascularizations, myocardial infarctions, unstable angina, or dysrhythmias. *Revascularization* means angioplasty (with or without stent placement) or bypass surgery.

(i) How do we calculate separate ischemic episodes? Reocclusion that occurs after a revascularization procedure but during the same hospitalization and that requires a second procedure during the same hospitalization will not be counted as another ischemic episode. If you are hospitalized for documented myocardial infarction and have a revascularization procedure during the same hospitalization, this event will be counted as one ischemic episode.

(ii) How do we evaluate ischemic episodes not amenable to revascularizations? If your ischemic episodes are not amenable to revascularization, we will evaluate them using the appropriate listing (for example, 4.04D). *Not amenable* means that the revascularization procedure could not be done because of another medical impairment or because the vessel was not suitable for revascularization.

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

e. In 4.04D1, the term fractional flow reserve (FFR) is a measurement of the pressure differences across an obstructive lesion, giving an estimate of the severity of stenosis. An FFR measurement of 1.0 indicates normal blood flow. An FFR measurement equal to or less than 0.80 indicates stenosis capable of producing serious myocardial ischemia in an artery appropriate for revascularization. An FFR measurement that is greater than 0.80 indicates stenosis not likely to produce significant ischemia.

f. In 4.04D2 and 4.04D3, the term nonbypassed means that the blockage is in a vessel that is potentially bypassable; that is, large enough to be bypassed and considered to be a cause of your ischemia. These vessels are usually major arteries or one of a major artery's major branches. A vessel that has become obstructed again after angioplasty or stent placement and has remained obstructed or is not amenable to another revascularization is considered a nonbypassed vessel for purposes of the listings. When you have had revascularization, we will not use the pre-operative findings to assess the current severity of your coronary artery disease under 4.04D, although we will consider the severity and duration of your impairment before your surgery in making our determination or decision.

F. *How do we evaluate arrhythmias?*

1. *What is an arrhythmia?* * * * Although we use the term ''arrhythmia'' in the listings, the term ''dysrhythmia'' may also be used in the medical evidence to describe this condition.

* * * * *

3. * * *

a. We will use 4.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator, and you have recurrent episodes of syncope or near syncope. * * *

* * * * *

4. * * *

a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in people who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group at risk for sudden cardiac death consists of people with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in people with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator rescues a person from what may have been cardiac arrest. However, as a consequence of the shock(s), similar to the effects of treatments for other cardiovascular disease, a person may experience psychological distress, which we may evaluate under the listings in 12.00.

b. * * * In some people, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. * * * Also, exposure to strong electrical or magnetic fields, such as from magnetic resonance imaging, can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. * * *

* * * * *

G. *How do we evaluate peripheral vascular disease?*

1. *What is peripheral vascular disease (PVD)?* * * * Neuropathy may mask these typical symptoms. * * *

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

* * * * *

4. * * *

a. * * *

b. Lymphedema does not meet the requirements of 4.11, although it may medically equal the listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing, or whether the lymphedema medically equals a cardiovascular disorders listing such as 4.11 or a listing in 1.00. * * *

5. * * *

6. *Are there any other studies that are helpful in evaluating PAD?* Doppler studies done using a recording ultrasonic Doppler unit and strain-gauge plethysmography are other useful tools for evaluating PAD. A recording Doppler, which prints a tracing of the arterial pulse wave in the femoral, popliteal, dorsalis pedis, and posterior tibia arteries, is an evaluation tool that compares waveforms in normal and compromised peripheral blood flow. Qualitative analysis of the pulse wave is helpful in the overall assessment of the severity of the occlusive disease. Tracings help in assessing severity if you have small vessel disease related to diabetes mellitus or other diseases with similar vascular changes, or diseases causing medial calcifications when ankle pressure is either normal or falsely high. When there is evidence of medial calcification of the ankle arteries or the ankle-brachial index is 0.50 or greater, other appropriate tests for PAD include magnetic resonance angiography, computed tomography angiography, contrast angiography, and graded treadmill tests.

7. * * *

a. * * *

b. * * * The criterion in 4.12A is met when your resting ankle/brachial systolic blood pressure ratio is less than 0.50. * * *

* * * * *

H. *How do we evaluate congenital heart disease?*

1. *What is congenital heart disease?* Congenital heart disease is any abnormality of the heart or the major blood vessels that is present at birth. Congenital heart disease includes abnormal structure of the individual heart chambers, valves, and blood vessels, and abnormal relative relationship of the chambers to each other that alters the normal pattern of blood flow. Surgery in childhood is the usual treatment, and with improving surgical techniques and medical management, more children with congenital heart disease are surviving into adulthood. Rarely, a person with congenital heart disease may not have received the usual surgery in childhood, and later, as an adult, he or she is no longer a surgical candidate, as for example, in Eisenmenger syndrome.

2. *What is Eisenmenger syndrome?* Eisenmenger syndrome refers to any surgically untreated congenital heart defect with intracardiac communication that over time leads to pulmonary hypertension, reversal of blood flow, and hypoxemia.

a. Lesions in Eisenmenger syndrome, such as large septal defects, are characterized by elevated pulmonary pressures or a high pulmonary flow rate. In response, the pulmonary blood vessels pathologically change, leading eventually to pulmonary hypertension. Development of Eisenmenger syndrome represents a point at which pulmonary hypertension is irreversible and the cardiac lesion is likely inoperable.

b. Examples of congenital heart disease that if untreated may cause pulmonary vascular disease leading to Eisenmenger syndrome include atrial septal defect (ASD), ventricular septal defect (VSD), and large patent ductus arteriosus (PDA).

3. *What is single ventricle?* The term ''single ventricle'' (also known as single ventricle physiology or functional single ventricle) describes a diverse group of congenital cardiac anomalies sharing the common feature that only one of the two heart ventricles is adequately developed. At birth, one ventricle must functionally do the work of two, pumping blood for both the body (systemic) and the lungs (pulmonary). Because of this feature, the ultimate plan for cardiac reconstruction is similar for most of these anomalies. People with single ventricle will generally undergo staged reconstructive ''Fontan procedures,'' ultimately resulting in a ''Fontan circulation.'' Fontan circulation describes the hemodynamic state in which virtually all systemic venous return-blood passively flows directly into the pulmonary arteries via surgical or catheter-placed shunts, without the blood passing through a ventricle. Some of the anomalies described as single ventricle include the following:

(a) Hypoplastic left heart syndrome;

(b) Hypoplastic right ventricle;

(c) Tricuspid valve atresia;

(d) Double inlet left ventricle; and

(e) Some variations of double outlet right ventricle.

4. *How do we evaluate conditions associated with congenital heart disease?*

a. We evaluate congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias under 4.02 or 4.05, respectively. Otherwise, we evaluate your impairment under 4.06.

b. We evaluate pulmonary hypertension due to congenital heart disease under 4.06B or 4.06C. We evaluate pulmonary hypertension not due to congenital heart disease under the listings in 3.00 (for example, 3.09).

c. We need pulse oximetry measurements documented by medical sources using methods consistent with the prevailing state of medical knowledge and clinical practice to evaluate chronic hypoxemia in congenital heart disease under 4.06A3. These pulse oximetry measurements also must be consistent with the other evidence in the case record.

d. We evaluate single ventricle physiology under 4.06D and will consider you disabled if your medical evidence documents that you have any congenital heart disorder that results in single ventricle physiology (functional single ventricle). In addition to the above congenital heart disorders, examples of palliative surgical procedures that indicate single ventricle physiology include the Glenn, Fontan, and Norwood procedures.

* * * * *

I. *How do we evaluate other cardiovascular disorders?*

1. *How do we evaluate hypertension? Hypertension* (high blood pressure) over time may significantly raise the pressures in the heart to the point of ineffective heart muscle function known generally as hypertensive heart disease that we can evaluate under 4.02. Other body systems, such as the brain, kidneys, or eyes may also be affected. We evaluate these impairments by reference to the specific body system(s) that is affected. We will also consider any limitations imposed by your hypertension when we assess your residual functional capacity.

2. *What is cardiomyopathy and how will we evaluate it?* Cardiomyopathy is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy).

a. There are various types of cardiomyopathy, which fall into two major categories: ischemic and nonischemic cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: dilated, hypertensive, hypertrophic, and restrictive. Cardiomyopathy includes hypertrophic cardiomyopathy, endomyocardial fibrosis, or cardiac amyloidosis AL type.

b. We evaluate cardiomyopathy under 4.08. Depending on the underlying cause of the cardiomyopathy or its effects on you, we may also evaluate your cardiomyopathy under 4.02, 4.04, or 4.05. If your cardiomyopathy results in vascular insult to the brain, we may also evaluate it under 11.04.

c. Under 4.08A2, we need a conclusion from a medical source that the performance of an exercise test would present a significant risk to you. If your case record does not have a conclusion from a medical source that an exercise test would present a significant risk to you, an MC defined in 4.00A3a may make such a conclusion if evidence in your case record supports it.

3. *How do we evaluate valvular heart disease?* We evaluate aortic valvular disease under 4.07. We may also evaluate aortic valvular disease, as well as other forms of valvular disease, under 4.02, 4.04, 4.05, 4.06, or a listing in 11.00, depending on its effects on you.

4. *What do we consider when we evaluate heart transplant recipients?* a. After your heart transplant, we will consider you disabled under 4.09 for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year. If you develop cardiac allograft vasculopathy after your transplant, we will evaluate this impairment under 4.16.

b. * * *

c. * * *

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

5. *What is cardiac allograft vasculopathy and how do we evaluate it?* Cardiac allograft vasculopathy (CAV) may affect a person who has received a heart transplant and involves thickening in the walls of the coronary arteries that may progress quickly into serious vascular stenosis and heart dysfunction. Stenosis in CAV is caused by a pathological process different from classic atherosclerosis and treatment often is only palliative. We evaluate CAV under 4.16.

* * * * *

8. * * *

a. * * *

b. * * * Most people with Marfan syndrome have abnormalities associated with the heart and blood vessels. * * * * *

J. *How do we evaluate issues that affect the cardiovascular system?* 1. *How do we consider the effects of obesity when we evaluate your cardiovascular disorder?* Obesity is a medically determinable impairment that may be associated with cardiovascular disorders. The additional body mass may make it harder for the chest and lungs to expand or may cause the heart to work harder to pump blood to carry oxygen to the body. The combined effects of obesity with a cardiovascular disorder can be greater than the effects of each of the impairments considered separately. We consider the additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular disorder, a listing-level cardiovascular disorder, a combination of impairments that medically equals the severity of a listed impairment, and when we assess your residual functional capacity.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular disorder cannot be made on the basis of the type of treatment rendered or anticipated. * * *

3. *How do we consider hospitalizations?* When we evaluate hospitalizations for chronic heart failure (4.02B3), ischemic heart disease (4.04E), congenital heart disease (4.06E), and cardiomyopathy (4.08D), the hospitalizations do not all have to be for the same cardiovascular disorder(s). They may be for three different exacerbations or complications resulting from your cardiovascular disorder. The hospitalizations must be at least 30 days apart, and each one must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

K. *How do we evaluate cardiovascular disorders that do not meet one of these listings?*

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

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

when we decide whether you continue to be disabled.

4.01 Category of Impairments, Cardiovascular Disorders

4.02 *Chronic heart failure* (see 4.00D) while on a regimen of prescribed treatment, with symptoms and signs described in 4.00D2. The required level of severity for this impairment is met when the requirements are satisfied by A and B; or C alone; or D alone.

A. Medically documented presence of one of the following:

1. Systolic failure documented by appropriate medically acceptable imaging during a period of stability (not during an episode of exacerbation of heart failure), with left ventricular end diastolic dimension equal to or greater than 7.0 cm; or ejection fraction of 30 percent or less during a period of stability (not during an episode of acute heart failure); OR

2. Diastolic failure documented by appropriate medically acceptable imaging during a period of stability (not during an episode of exacerbation of heart failure), with left ventricular posterior wall plus septal thickness totaling 2.5 cm or greater, with an enlarged left atrium greater than or equal to 4.5 cm, *OR* left atrial volume index (LAVi) greater than or equal to 40 ml, BSA/m2 (milliliters to body surface area in squared meters).

AND

B. Resulting in one of the following:

1. Recurrent (see 4.00A3c) symptoms of heart failure, resulting in both a and b:

a. A medical source (see 4.00D4c(i)) has concluded that the performance of an exercise test would present a significant risk to the person; and

b. Very serious limitation in the ability to perform activities of daily living independently, appropriately, effectively, and on a sustained basis; or

2. Inability to perform on an exercise tolerance test at a workload equivalent to 5 METs or less if using a standard treadmill (or bicycle) test without gas exchange, or at 15 ml/kg/min peak VO2 (oxygen consumption) on a cardiopulmonary exercise test, due to either a or b:

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

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

3. Exacerbations or complications of chronic heart failure (see 4.00D1b) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3);

OR

C. Heart failure with left ventricular ejection fraction of 20 percent or less while on a regimen of prescribed therapy, on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e) during a period of stability (not during an episode of exacerbation of heart failure);

OR

D. One of the following while hospitalized, at home, or both:

1. Mechanical circulatory support device except extracorporeal membrane oxygenation (ECMO) (see 4.00D4e). Consider under a disability for 1 year from the date of implantation; after that, evaluate any residual impairment(s) under the criteria for the affected body system.

2. Continuous intravenous administration of inotropic medication (for example, milrinone) for at least 30 consecutive days. Consider under a disability for 1 year from the date of initiation of the treatment; after that, evaluate any residual impairment(s) under the criteria for the affected body system.

4.03 [Reserved]

4.04 *Ischemic heart disease* (see 4.00E), with symptoms due to myocardial ischemia, while on a regimen of prescribed treatment (see 4.00B3 if there is no regimen of prescribed treatment), with A, B, C, D, or E:

A. Inability to perform on an exercise tolerance test at a workload equivalent to 5 METs or less with findings interpreted by an acceptable medical source as positive for ischemia (see 4.00E9b).

OR

B. Ischemic response with exercise or pharmacological (drug-induced) stress testing (see 4.00C14) on medically appropriate imaging, with either 1 or 2:

1. At least two reversible or fixed regional myocardial perfusion defects and either a or b:

a. Transient ischemic dilatation; or

b. Resting left ventricular ejection fraction of less than 50 percent; or

2. At least two reversible or fixed regional wall motion abnormalities and either a or b:

a. Decrease in left ventricular ejection fraction during testing; or

b. Resting left ventricular ejection fraction of less than 50 percent.

OR

C. Documentation of three separate ischemic episodes (see 4.00E9c) requiring unplanned hospitalization (inpatient or observation status) within a consecutive 12-month period (see 4.00A3e).

OR

D. Coronary artery disease, documented by coronary angiography (obtained independently of Social Security disability evaluation) with 1, 2, or 3:

1. Fractional flow reserve (see 4.00E9e) measurement of less than or equal to 0.80 of a proximal segment or mid segment coronary artery not amenable to revascularization (see 4.00E9c(ii)).

2. History of coronary artery bypass graft surgery with manifestations of ischemia, as described in 4.00E3–4.00E7, while on a regimen of prescribed treatment (see 4.00B3 if there is no regimen of prescribed treatment) with a, b, c, or d:

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

b. 70 percent or more stenosis in the proximal segment or mid segment of another nonbypassed coronary artery; or

c. 50 percent or more stenosis in the proximal segment or mid segment of at least two nonbypassed coronary arteries; or

d. 70 percent or more stenosis of a bypass graft vessel.

3. Resting left ventricular ejection fraction of less than 50 percent while medically stable (see 4.00B4) with manifestations of ischemia, as described in 4.00E3–4.00E7, while on a regimen of prescribed treatment (see 4.00B3 if there is no regimen of prescribed treatment) with a, b, or c:

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

b. 70 percent stenosis in the proximal segment or mid segment of another nonbypassed coronary artery; or

c. 50 percent or more stenosis in the proximal segment or mid segment of at least two nonbypassed coronary arteries.

OR

E. Exacerbations or complications of ischemic heart disease (see 4.00E2–4.00E7) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).

4.05 *Recurrent arrhythmias* (see 4.00F), not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, while on a regimen of prescribed treatment (see 4.00B3 if there is no prescribed treatment), demonstrated by both A and B:

A. Coincident with recurrent (see 4.00A3c) episodes of cardiac syncope or near syncope (see 4.00F3b).

AND

B. Documented by either 1 or 2:

1. Resting or ambulatory (Holter) electrocardiography; or

2. Other appropriate medically acceptable testing.

4.06 *Congenital heart disease* (see 4.00H), documented by appropriate medically acceptable imaging (see 4.00A3d) or cardiac catheterization, with A, B, C, D, or E:

A. Chronic hypoxemia, and 1, 2, or 3:

1. Hematocrit of 55 percent or greater on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e); or

2. Arterial blood gas test measurement obtained at rest while breathing room air, as described in either a or b:

a. S_aO_2 (arterial oxygen saturation) less than or equal to 89 percent; or

b. PO_2 or P_aO_2 (partial pressure of oxygen) less than or equal to 60 mmHg;

3. S_pO_2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry either at rest, during a 6-minute walk test (6MWT), or after a 6MWT, while breathing room air, less than or equal to 87 percent on three evaluations at least 30 days apart within a consecutive 12-month period (see 4.00A3e).

OR

B. Intermittent right-to-left shunting (for example, Eisenmenger syndrome; see 4.00H2) during cardiopulmonary exercise testing while breathing room air, resulting in oxygen desaturation on exertion at a workload equivalent to 5 METs or less, or peak VO_2 (oxygen uptake) of 15.0 ml/kg/min or less, *and* arterial blood gas test measurement, with either 1 or 2:

1. S_aO_2 less than or equal to 89 percent; or
2. PO_2 or P_aO_2 less than or equal to 60 mmHg.

OR

C. Pulmonary hypertension documented by cardiac catheterization while medically stable, as described in 1, 2, or 3:

1. Pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure; or

2. Pulmonary arterial systolic pressure equal to or greater than 70 mmHg; or

3. Mean pulmonary artery pressure equal to or greater than 40mmHg.

OR

D. Single ventricle (with or without Fontan procedures) (see 4.00H4).

OR

E. Exacerbations or complications of congenital heart disease (see 4.00J3) requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).

4.07 *Aortic valvular disease* (see 4.00I3), with symptoms due to stenosis, determined by appropriate test or tests showing an aortic valve area of less than 1.0 cm^2.

4.08 *Cardiomyopathy* (see 4.00I2) while on a regimen of prescribed treatment, with A, B, C, or D:

A. Hypertrophic cardiomyopathy documented by appropriate medically acceptable imaging, with left ventricular or septal wall thickness equal to or greater than 20 mm in the absence of other causes of left ventricular hypertrophy (for example, hypertension or aortic valvular disease) and either 1 or 2:

1. Inability to perform on an exercise tolerance test at a workload equivalent to 5 METs or less if using a standard treadmill (or bicycle) test without gas exchange, or at 15 ml/kg/min peak VO_2 (oxygen consumption) on a cardiopulmonary exercise test; or

2. A medical source (see 4.00I2c) has concluded that the performance of an exercise tolerance test would present a significant risk to the person.

OR

B. Endomyocardial fibrosis documented by appropriate medically acceptable imaging, with 1, 2, and 3:

1. Loss of chamber volume due to fibrosis of the endocardium of at least one ventricle; and

2. Right or left atrial dilatation (chamber enlargement); and

3. Regurgitant (backward) blood flow through the mitral or tricuspid valve.

OR

C. Cardiac amyloidosis AL (light-chain) type documented by biopsy.

OR

D. Exacerbations or complications of cardiomyopathy requiring three hospitalizations within a consecutive 12-month period (see 4.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 4.00J3).

4.09 *Heart transplantation* (see 4.00I4). Consider under a disability for 1 year from the date of the transplant; after that, evaluate the residual impairment(s).

4.10 *Dissecting aneurysm of the aorta or major branches* (see 4.00I6), due to any cause (for example, atherosclerosis, cystic medical necrosis Marfan syndrome, or trauma), demonstrated by appropriate medically acceptable imaging, with dissection not controlled by prescribed treatment.

4.11 *Chronic venous insufficiency* (see 4.00G) of a lower extremity with reflux or obstruction of the venous system documented by duplex ultrasound or other appropriate diagnostic technique, with A or B:

A. Extensive trophic changes of skin (for example, hyperpigmentation, lipodermatosclerosis, brawny edema) involving at least two-thirds of the leg below the knee, on two evaluations at least 90 days apart within a consecutive 12-month period (see 4.00A3e), with both 1 and 2:

1. Consistent with chronic venous insufficiency; and

2. Unresponsive to compression therapy.

OR

B. Two or more episodes of ulceration that has not healed following at least 6 months of prescribed treatment.

4.12 *Peripheral arterial disease* (see 4.00G7) while on a regimen of prescribed treatment resulting in intermittent claudication or leg pain that interferes with mobility (see 4.00G1), with A, B, C, or D, as determined by an appropriate test(s) (see 4.00G5–4.00G6):

A. Resting ankle/brachial systolic blood pressure ratio of less than 0.50 (see 4.00G7a).

OR

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

OR

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

OR

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

4.13–4.15 [Reserved]

4.16 *Cardiac allograft vasculopathy* (see 4.00I5), documented by appropriate medically acceptable imaging (for example, intravascular ultrasonography or coronary angiography) (see 4.00A3d), with A, B, C, or D:

A. Cardiac index (CI) or cardiac output (CO) less than 2 l/min/m^2.

OR

B. Left ventricular ejection fraction equal to or less than 45 percent.

OR

C. Right atrial pressure (RAP) greater than 12 mmHg.

OR

D. Pulmonary capillary wedge pressure (PCWP) greater than 15 mmHg.

* * * * *

5. Amend part B of appendix 1 to subpart P of part 404 by revising the body system name for section 104.00 in the table of contents to read as follows:

* * * * *

Part B

* * * * *

104.00 Cardiovascular Disorders

* * * * *

104.00 Cardiovascular Disorders

A. *How do we define cardiovascular disorders and cardiovascular terms?*

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

a. * * *

b. Cardiovascular disorders result from one or more of four consequences of heart disease: * * *

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

* * * * *

2. *What do we consider in evaluating cardiovascular disorders?* The listings in this section describe cardiovascular disorders based on the medical and other evidence, including response to a regimen of prescribed treatment and functional limitations.

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

a. *Medical consultant* is a person defined in § 416.1016(a) of this chapter. * * *

b. * * * By ''exceptions,'' we mean brief periods when the required finding(s) is greatly reduced or gone. These periods are so brief or inconsequential, the required finding(s) remains a factor in the person's condition.

c. * * * By ''improvement of sufficient duration,'' we mean the finding is greatly reduced or not present for long enough that the required finding(s) is no longer a factor in the person's condition.

* * * * *

B. *What documentation do we need to evaluate cardiovascular disorders?*

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular disorder.

* * * * *

2. *Why is a longitudinal clinical record important?* * * * Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your medical source(s). * * *

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

a. * * * In this situation, we will base our evaluation on the current evidence we have. * * * However, we may find you disabled because you have another impairment(s) that, in combination with your cardiovascular disorder, medically equals a listing or functionally equals the listings.

b. * * * In rare instances when there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the existence, severity, and duration of your impairment.

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

a. * * *

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

* * * * *

5. *Will we purchase any studies?* In appropriate situations, we may purchase studies necessary to substantiate the existence of a medically determinable impairment or to document the severity of your impairment, generally after we have evaluated the evidence we already have. * * * We will follow sections 4.00C6, 4.00C7, 4.00C8 in part A, and 104.00B7, when we decide whether to purchase exercise testing. * * *

* * * * *

7. *Will we use exercise tolerance tests (ETT) for evaluating children with cardiovascular disorders?*

a. * * * An ETT may be of value in the assessment of some arrhythmias, as indicated in 104.05B2. ETTs may also be used in the assessment of the severity of chronic heart failure and in the assessment of recovery of function following cardiac surgery or other treatment.

b. We will purchase an ETT only if we cannot make a determination or decision based on the evidence we have and an MC, preferably one with experience in the care of children with cardiovascular disorders, has determined that an ETT is needed to evaluate your impairment. * * *

c. For full details on ETT requirements and usage, see 4.00C3 in part A.

C. *How do we evaluate chronic heart failure?*

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

a. *Heart failure* is the inability of the heart to pump enough oxygenated blood to body tissues. * * * Ejection fraction in heart failure is a continuum ranging from low ejection fraction due to muscle dysfunction to preserved ejection fraction resulting from high intracardiac pressures. We consider heart failure to be chronic when the condition persists or recurs over time despite treatment.

b. *CHF* is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, congenital, or other heart disease. If the CHF is the result of primary pulmonary hypertension secondary to disease of the lung, we will evaluate your impairment under the listings in 3.00 (for example, 3.09) or 4.00, as appropriate.

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

a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization. Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema.

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

(i) * * *

(ii) * * *

(iii) * * * However, these signs need not be found on all examinations because congestion may be controlled by prescribed treatment or may not be present at the time of evaluation.

* * * * *

4. *How do we evaluate CHF treated with a mechanical circulatory support device?* We use 104.02D to evaluate CHF treated with an implanted mechanical circulatory support device (MCSD), such as a left ventricle assistive device (LVAD) or a right ventricle assistive device (RVAD). Implanted MCSDs are intended for long-term circulatory support in helping the heart pump blood. For the purposes of 104.02D, an MCSD does not include extracorporeal membrane oxygenation (ECMO). Although ECMO is a form of mechanical circulatory support, we do not include it in 104.02D because ECMO is intended only for short-term circulatory support (maximum 30 days), used in a setting of imminent or actual cardiac arrest.

D. *How do we evaluate congenital heart disease?*

1. *What is congenital heart disease? Congenital heart disease* is any abnormality of the heart or the major blood vessels that is present at birth. Congenital heart disease includes abnormal structure of the individual heart chambers, valves, and blood vessels, and abnormal relative relationship of the chambers to each other that alters the normal pattern of blood flow. Surgery is the usual treatment, and with improving surgical techniques and medical management, more children with congenital heart disease are surviving into adulthood. Examples of congenital heart disease include:

a. * * *

b. * * *

c. * * *

d. Major abnormalities of ventricular development, including hypoplastic left heart syndrome or tricuspid atresia with hypoplastic right ventricle.

2. *How do we evaluate conditions associated with congenital heart disease?*

a. We will evaluate congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias under 104.02 or 104.05, respectively. Otherwise, we will evaluate your impairment under 104.06.

b. We need pulse oximetry measurements documented by medical sources using methods consistent with the prevailing state of medical knowledge and clinical practice to evaluate chronic hypoxemia in congenital heart disease under 104.06A3. These pulse oximetry measurements also must be consistent with the other evidence in the case record.

c. 104.06D, life-threatening congenital heart disease does not include single ventricle; we evaluate single ventricle physiology separately under 104.06C. When we evaluate life-threatening congenital heart disease under 104.06D, we consider whether it responds to surgical treatment and, therefore, may not meet the 12-month duration requirement. Examples of impairments that in most instances will require life-saving surgery or a combination of surgery and other major interventional procedures (for example, multiple ''balloon'' catheter procedures) before age 1 include, but are not limited to, the following:

(i) Critical aortic stenosis with neonatal heart failure,

(ii) Critical coarctation of the aorta, with associated anomalies,

(iii) Complete atrioventricular canal defects,

(iv) Transposition of the great arteries,

(v) Tetralogy of Fallot, and

(vi) Multiple ventricular septal defects.

3. *What is Eisenmenger syndrome?* Eisenmenger syndrome refers to any surgically untreated congenital heart defect with intracardiac communication that over time leads to pulmonary hypertension, reversal of blood flow, and hypoxemia.

a. Lesions in Eisenmenger syndrome, such as large septal defects, are characterized by elevated pulmonary pressures or a high pulmonary flow rate. In response, the pulmonary blood vessels pathologically change, leading eventually to pulmonary hypertension. Development of Eisenmenger syndrome represents a point at which pulmonary hypertension is irreversible and the cardiac lesion is likely inoperable.

b. Examples of congenital heart disease that if untreated may cause pulmonary vascular disease leading to Eisenmenger syndrome include atrial septal defect (ASD), ventricular septal defect (VSD), and large patent ductus arteriosus (PDA). We evaluate Eisenmenger syndrome under 104.06A or 104.06B.

4. *What is single ventricle?* The term ''single ventricle'' (also known as single ventricle physiology or functional single ventricle) describes a diverse group of congenital cardiac anomalies sharing the common feature that only one of the two heart ventricles is adequately developed. At birth, one ventricle must functionally do the work of two, pumping blood for both the body (systemic) and the lungs (pulmonary). Because of this feature, the ultimate plan for cardiac reconstruction is similar for most of these anomalies. People with single ventricle will generally undergo staged reconstructive ''Fontan procedures,'' ultimately resulting in a ''Fontan circulation.'' Fontan circulation describes the hemodynamic state in which virtually all systemic venous return-blood passively flows directly into the pulmonary arteries via surgical or catheter-placed shunts, without (the blood) passing through a ventricle. Some of the anomalies described as single ventricle include the following:

(i) Hypoplastic left heart syndrome;

(ii) Hypoplastic right ventricle;

(iii) Tricuspid valve atresia;

(iv) Double inlet left ventricle; and

(v) Some variations of double outlet right ventricle.

E. *How do we evaluate arrhythmias?*

1. *What is an arrhythmia?* * * * Although we use the term ''arrhythmia'' in the listings, the term ''dysrhythmia'' may also be used in the medical evidence to describe this condition.

* * * * *

4. *What will we consider when you have an implanted cardiac defibrillator and you do*

not have arrhythmias that meet the requirements of 104.05?

a. * * * The shock from the implanted cardiac defibrillator rescues a child from what may have been cardiac arrest. However, as a consequence of the shock(s), similar to the effects of treatments for other cardiovascular disease, a child may experience psychological distress, which we may evaluate under the listings in 112.00.

b. * * * Also, exposure to strong electrical or magnetic fields, such as from magnetic resonance imaging, can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. * * *

* * * * *

F. *How do we evaluate other cardiovascular disorders?*

1. *What is ischemic heart disease (IHD) and how will we evaluate it in children?* * * * If you have IHD, we will evaluate it under 4.04 in part A.

2. *How will we evaluate hypertension?* Hypertension (high blood pressure) generally causes disability in children through its effects on other body systems, such as the brain, kidneys, or eyes, and we will evaluate these impairments by reference to the specific body system(s) that is affected. * * *

3. *What is cardiomyopathy and how will we evaluate it?*

a. There are various types of cardiomyopathy, which fall into two major categories: ischemic and nonischemic cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: dilated, hypertensive, hypertrophic, and restrictive.

b. We will evaluate cardiomyopathy under 4.04 in part A, 104.02, or 104.05, depending on its effects on you.

4. *How will we evaluate valvular heart disease?* We will evaluate aortic valvular disease under 4.07 in part A. We may also evaluate aortic valvular disease, as well as other forms of valvular disease, under 4.04 in part A, 104.02, 104.05, 104.06, or a listing in 111.00, depending on its effects on you.

5. * * *

a. After your heart transplant, we will consider you disabled under 104.09 for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year. If you develop cardiac allograft vasculopathy after your transplant, we will evaluate this impairment under 104.16.

* * * * *

d. When we do a continuing disability review to determine whether you are still disabled, we will evaluate your residual impairment(s), as shown by the evidence in your case record, including any side effects of medication. We will consider all evidence indicative of cardiac dysfunction in deciding whether medical improvement (as defined in § 416.994a of this chapter) has occurred.

6. *How will we evaluate chronic rheumatic fever or rheumatic heart disease?* We will evaluate rheumatic fever or rheumatic heart disease under the listing appropriate to its effects on you, which may include heart failure or recurrent arrhythmias. If you have evidence of chronic heart failure or recurrent arrhythmias associated with rheumatic heart disease, we will evaluate these disorders under 104.02 or 104.05, respectively.

* * * * *

9. * * *

a. * * *

b. Lymphedema does not meet the requirements of 4.11 in part A, although it may medically equal the listing. We evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular disorders listing, such as 4.11 in part A, or a listing in 101.00. If no listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we consider whether you have an impairment(s) that functionally equals the listings.

* * * * *

11. *What is cardiac allograft vasculopathy and how do we evaluate it?* Cardiac allograft vasculopathy (CAV) may affect a person who has received a heart transplant and involves thickening in the walls of the coronary arteries that may progress quickly into serious vascular stenosis and heart dysfunction. Stenosis in CAV is caused by a pathological process different from classic atherosclerosis and treatment often is only palliative. We evaluate CAV under 104.16.

G. *How do we evaluate issues that affect the cardiovascular system?*

1. How do we consider the effects of obesity when we evaluate your cardiovascular disorder? Obesity is a medically determinable impairment that may be associated with cardiovascular disorders. The additional body mass may make it harder for the chest and lungs to expand or may cause the heart to work harder to pump blood to carry oxygen to the body. The combined effects of obesity with a cardiovascular disorder can be greater than the effects of each of the impairments considered separately. We consider the additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular disorder, a listing-level cardiovascular disorder, a combination of impairments that medically equals the severity of a listed impairment, and when we determine whether your impairment(s) functionally equals the listings.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular disorder cannot be made on the basis of the type of treatment rendered or anticipated. * * *

3. *How do we consider hospitalizations?* The hospitalizations in 104.02E and 104.06E do not all have to be for the same exacerbation or complication of your cardiovascular disorder(s). They may be for three different exacerbations or complications resulting from your cardiovascular disorder. The hospitalizations must be at least 30 days apart, and each one must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

H. *How do we evaluate cardiovascular disorders that do not meet one of these listings?*

1. These listings are only examples of common cardiovascular disorders that we consider severe enough to result in marked and severe functional limitations. If your impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an impairment(s) that satisfies the criteria of a listing in another body system.

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. See § 416.926 of this chapter. If your impairment(s) does not meet or medically equal a listing, we will also consider whether it functionally equals the listings. See § 416.926a of this chapter. We will use the rules in § 416.994a of this chapter when we decide whether you continue to be disabled.

104.01 Category of Impairments, Cardiovascular Disorders

104.02 *Chronic heart failure* (see 104.00C) while on a regimen of prescribed treatment with symptoms and signs described in 104.00C2, and with A, B, C, D, or E:

A. Persistent tachycardia at rest measured at least twice within a consecutive 12-month period and at least 90 days apart documented by apical heart rate greater than or equal to the value in Table I.

TABLE I—TACHYCARDIA AT REST

Age	Apical heart rate (beats per minute)
Under 1 year	150
1 through 3 years	130
4 through 9 years	120
10 through 15 years	110
Over 15 years	100

OR

B. Persistent tachypnea at rest measured at least twice within a consecutive 12-month period and at least 90 days apart documented by respiratory rate greater than or equal to the value in Table II *or* markedly decreased exercise tolerance (see 104.00C2b).

TABLE II—TACHYPNEA AT REST

Age	Respiratory rate (per minute)
Under 1 year	40
1 through 5 years	35
6 through 9 years	30
Over 9 years	25

OR

C. Growth failure as required in 1 or 2:

1. *For children from birth to attainment of age 2,* three weight-for-length measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or

2. *For children age 2 to attainment of age 18,* three BMI-for-age measurements that are:

a. Within a consecutive 12-month period; and

b. At least 60 days apart; and

c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.

OR

D. Mechanical circulatory support device (except an extracorporeal membrane oxygenation (ECMO) while hospitalized, at home, or both (see 104.00C4). Consider under a disability for 12 months from the date of implantation; after that, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

E. Exacerbations or complications of chronic heart failure (see 104.00C1b) requiring three hospitalizations within a consecutive 12-month period and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 104.00G3).

104.03–104.04 [Reserved]

104.05 *Recurrent arrhythmias* (see 104.00E), not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, while on a regimen of prescribed treatment (see 104.00B3 if there is no prescribed treatment), demonstrated by both A and B:

A. Coincident with recurrent (see 104.00A3c) episodes of cardiac syncope or near syncope (see 104.00E3b).

AND

B. Documented by either 1 or 2:

1. Resting or ambulatory (Holter) electrocardiography; or

2. Other appropriate medically acceptable testing.

104.06 *Congenital heart disease* (see 104.00D), documented by appropriate medically acceptable imaging (see 104.00A3d) or cardiac catheterization, with A, B, C, D, or E:

A. Chronic hypoxemia, and 1, 2, or 3:

1. Hematocrit of 55 percent or greater on two evaluations at least 90 days apart within a consecutive 12-month period (see 104.00A3e); or

2. Arterial blood gas test measurement obtained at rest while breathing room air, as described in either a or b:

a. S_aO_2 (arterial oxygen saturation) less than or equal to 89 percent; or

b. PO_2 or P_aO_2 (partial pressure of oxygen) less than or equal to 60 mmHg; or

3. S_pO_2 (percentage of oxygen saturation of blood hemoglobin) measured by pulse oximetry either at rest, or after activity, while breathing room air, less than or equal to 87 percent on three evaluations at least 30 days apart within a consecutive 12-month period (see 104.00A3e).

OR

B. Pulmonary hypertension documented by cardiac catheterization while medically stable, as described in 1, 2, or 3:

1. Pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure; or

2. Pulmonary arterial systolic pressure equal to or greater than 70 mmHg; or

3. Mean pulmonary artery pressure equal to or greater than 40 mmHg.

OR

C. Single ventricle (for example, hypoplastic left or right ventricle) that has or will require Fontan procedures (see 104.00D5).

OR

D. For infants under 1 year of age at the time of filing, with life-threatening congenital heart disease (see 104.00D3c) that will require or already has required surgical treatment in the first year of life, and the impairment is expected to be disabling (because of residual impairment following surgery, or the recovery time required, or both) until the attainment of at least 1 year of age, consider under a disability until the attainment of at least age 1; after that, evaluate impairment severity with the appropriate listing.

OR

E. Exacerbations or complications of congenital heart disease (see 104.00D) requiring three hospitalizations within a consecutive 12-month period (see 104.00A3e) and at least 30 days apart. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization (see 104.00G3).

104.07–104.08 [Reserved]

104.09 *Heart transplantation* (see 104.00F5). Consider under a disability for 1 year from the date of the transplant; after that, evaluate the residual impairment(s).

104.10–104.15 [Reserved]

104.16 *Cardiac allograft vasculopathy* (see 104.00F11), documented by appropriate medically acceptable imaging (for example, intravascular ultrasonography or coronary angiography).

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