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Revised Medical Criteria for Evaluating Immune System Disorders; Proposed Rule

SOCIAL SECURITY ADMINISTRATION

20 CFR Part 404

RIN 0960–AF33

Revised Medical Criteria for Evaluating Immune System Disorders

AGENCY: Social Security Administration.
ACTION: Proposed rule.

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

DATES: To be sure your comments are considered, we must receive them by October 3, 2006.

ADDRESSES: You may give us your comments by: using our Internet facility (i.e., Social Security Online) at *http://policy.ssa.gov/erm/rules.nsf/Rules+Open+To+Comment* or the Federal eRulemaking Portal at *http://www.regulations.gov;* e-mail to *regulations@ssa.gov;* telefax to (410) 966–2830; or, letter to the Commissioner of Social Security, P.O. Box 17703, Baltimore, MD 21235–7703. You may also deliver them to the Office of Regulations, Social Security Administration, 107 Altmeyer Building, 6401 Security Boulevard, Baltimore, MD 21235–6401, between 8 a.m. and 4:30 p.m. on regular business days. Comments are posted on our Internet site, or you may inspect them physically on regular business days by making arrangements with the contact person shown in this preamble.

FOR FURTHER INFORMATION CONTACT: Greg Zwitch, SSA Regulations Officer, Office of Regulations, Social Security Administration, 107 Altmeyer Building, 6401 Security Boulevard, Baltimore, Maryland 21235–6401, (410) 965–1887 or TTY (410) 966–5609. For information on eligibility or filing for benefits, call our national toll-free number, 1–800–772–1213 or TTY 1–800–325–0778, or visit our Internet Web site, Social Security Online, at *http://www.socialsecurity.gov/.*

SUPPLEMENTARY INFORMATION: *Electronic Access:* The electronic file of this document is available on the date of publication in the **Federal Register** at *http://www.gpoaccess.gov/fr/index.html.* It is also available on the Internet site for SSA (i.e., Social Security Online) at *http://policy.ssa.gov/pnpublic.nsf/LawsRegs.*

What programs would these proposed regulations affect?

These proposed regulations would affect disability determinations and decisions that we make for you under title II and title XVI of the Act. In addition, to the extent that Medicare entitlement and Medicaid eligibility are based on whether you qualify for disability benefits under title II and title XVI, these proposed regulations would also affect the Medicare and Medicaid programs.

Who can get disability benefits?

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

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

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

How do we define disability?

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

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

What are the listings?

The listings are examples of impairments that we consider severe enough to prevent you as an adult from doing any gainful activity. If you are a child seeking SSI payments based on disability, the listings describe impairments that we consider severe enough to result in ''marked and severe functional limitations.'' Although we publish the listings only in appendix 1 to subpart P of part 404 of our rules, we incorporate them by reference in the SSI program in § 416.925 of our regulations, and apply them to claims under both title II and title XVI of the Act.

How do we use the listings?

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

If you are a person under age 18, we first use the criteria in part B of the listings. If the listings in part B do not apply, and if the specific disease process(es) has a similar effect on adults and children, we then use the criteria in part A. (See §§ 404.1525 and 416.925.)

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

We use the listings only to decide that you are disabled or that you are still disabled. We will never deny your claim or decide that you no longer qualify for benefits because your impairment(s) does not meet or medically equal a listing. If you have a severe impairment(s) that does not meet or medically equal any listing, we may still find you disabled based on other rules in the ''sequential evaluation process'' that we use to evaluate all disability claims. (See §§ 404.1520, 416.920, and 416.924.)

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

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

Why are we proposing to revise the listings for immune system disorders?

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

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

How did we develop these proposed rules?

These proposed rules reflect our adjudicative experience and advances in medical knowledge, treatment, and methods of evaluating immune system disorders. They also reflect comments we asked you to provide to help us develop the proposals.

We published an Advance Notice of Proposed Rulemaking (ANPRM) in the **Federal Register** on May 9, 2003 (68 FR 24896). The purpose of the ANPRM was to inform the public that we were planning to update and revise the rules we use to evaluate immune system disorders and to invite interested individuals and organizations to send us comments and suggestions for updating and revising the immune system listings. In the ANPRM, we provided a 60-day period for comments and suggestions; that period ended on July 8, 2003. We received over 200 letters and e-mails in response to the notice, many from individuals who have immune system disorders or who have family members with such disorders. We also received comments from medical experts, advocates, and people who adjudicate claims for us. Although we are not summarizing or responding to the comments in this notice, we read and considered them carefully and are proposing changes in our rules based on some of the suggestions we received.

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

When will we start to use these rules?

We will not use these proposed rules until we evaluate the public comments we receive on them, determine whether they should be issued as final rules, and issue final rules in the **Federal Register**. If we publish final rules, we will explain in the preamble how we will apply them, and we will summarize and respond to the public comments. Until the effective date of any final rules, we will continue to use our current rules.

How long would these proposed rules be effective?

If we publish these proposed rules as final rules, they will remain in effect for 8 years after the date they become effective, unless we extend them, or revise and issue them again.

What revisions are we proposing to make?

We are proposing to:

- Expand and reorganize the introductory text in proposed 14.00 and 114.00 to provide more guidance for our adjudicators, to update it, and to reflect the revised listings.
- Add paragraph headings to the introductory text in proposed 14.00 and 114.00 for easier reference.
- Add proposed 14.00C and 114.00C to explain the meaning of key terms.
- Remove all reference listings. Reference listings are listings that are met by satisfying the criteria of another listing. For example, current listing 14.08G1 for human immunodeficiency virus (HIV) infection with anemia is a reference listing that requires evaluation under current listing 7.02 for chronic anemia. Therefore, it is redundant. Instead of using a reference listing, we propose to provide general guidance in the introductory text to the immune system listings (proposed 14.00J2g) stating that hematologic abnormalities, such as anemia, may be evaluated under 7.00ff. In some cases, we are also replacing reference listings with new specific listing criteria for the impairments. For example, current listing 14.06, for undifferentiated connective tissue disorders, is entirely a reference listing. In the proposed rules, we are replacing the reference listing criterion with criteria that are specific to these disorders.
- Add proposed listings 14.10 and 114.10 for evaluating Sjoögren's syndrome.
- Add criteria to the listings, similar to those in current HIV infection listings 14.08N and 114.08O, for each of the other listed immune system disorders (for example, systemic lupus erythematosus and systemic vasculitis).
- Make nonsubstantive editorial changes to update the medical terminology in the introductory text and the listings and to make their language simpler and clearer.

How are we proposing to change the introductory text to the adult immune system listings?

We propose to expand and reorganize the introductory text to these listings. There are four major sections in current 14.00, and the longest of those sections, 14.00D, addresses only the evaluation of HIV infection. In these proposed rules, we add more sections and expand the guidance we provide about evaluating other kinds of immune system disorders.

Some of the guidance in current 14.00D is useful for evaluating other kinds of immune system disorders in addition to HIV infection. We are proposing to move that guidance from current 14.00D to new sections that would have more general applicability to immune system disorders. We are not proposing to remove any substantive guidance about how we evaluate HIV infection, only to reorganize some of the

information now in 14.00D of the current rules and to give it broader applicability where appropriate. We are also proposing to update and expand some of the guidance we provide for evaluating HIV infection and its effects, as we describe in more detail below.

The four sections in the current rules are:

- Current 14.00A, a short paragraph that describes generally the kinds of disorders we include in this body system.
- Current 14.00B, a lengthy section that discusses the evaluation of connective tissue disorders; that is, autoimmune disorders. It includes six undesignated paragraphs that primarily explain the kinds of evidence we need to document the existence and severity of these disorders, including how we evaluate loss of function. These paragraphs are followed by six numbered sections that provide guidance about specific impairments in the listings.
- Current 14.00C, a single sentence that explains that we evaluate allergic disorders under the appropriate listing of the affected body system.
- Current 14.00D, a lengthy section that explains how we document the existence and severity of HIV infection, including how we evaluate loss of function under listing 14.08N. It includes eight numbered subsections and many paragraphs that are not designated with letters or numbers within those subsections.

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

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

- Proposed 14.00A: *What disorders do we evaluate under the immune system listings?*
- Proposed 14.00B: *What information do we need to show that you have an immune system disorder?*
- Proposed 14.00C: *Definitions*
- Proposed 14.00D: *What are the listed autoimmune disorders in these listings?*
- Proposed 14.00E: *How do we evaluate immune deficiency disorders, excluding HIV infection (14.07)?*
- Proposed 14.00F: *How do we evaluate human immunodeficiency virus (HIV) infection?*
- Proposed 14.00G: *How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?*
- Proposed 14.00H: *How do we consider your symptoms, including your constitutional symptoms or pain?*
- Proposed 14.00I: *How do we use the functional criteria in these listings?*
- Proposed 14.00J: *How do we evaluate your immune system disorder when it does not meet one of these listings?*

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

14.00 Immune System Disorders

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

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

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

We incorporate current 14.00A in the opening sentence of proposed 14.00A1. We propose to revise the sentence, which explains the kinds of immune system dysfunction that immune system disorders may cause, to update and simplify it. In proposed 14.00A1a and 14.00A1b, we incorporate the first sentence in the sixth paragraph of current 14.00B to explain that immune system disorders can cause dysfunction in one or more components of the immune system, and describe ways in which immune system disorders may result in loss of function. In the second sentence of 14.001b, we propose to add ''involuntary'' as a descriptor of weight loss to clarify that we mean weight loss due to an immune system disorder(s) or its treatment. We are adding ''involuntary'' as a descriptor of weight loss throughout the introductory text in part A and part B for this same reason. Proposed 14.00A1c is a new paragraph that explains how we have organized immune system disorders in the preface (introductory text) of these listings.

In proposed 14.00A2, *Autoimmune disorders,* we incorporate the first paragraph in current 14.00B to provide a brief description of autoimmune disorders. We propose to add an explanation that these disorders are sometimes referred to as ''rheumatic diseases,'' ''connective tissue disorders,'' or ''collagen vascular disorders'' and that some of the features of these disorders in adults differ from the features of the same disorders in children. We provide a cross-reference to proposed 14.00D, the section of the introductory text that addresses autoimmune disorders in detail. We also propose to remove the last sentence of the first paragraph of current 14.00B, which explains that connective tissue disorders generally evolve and persist over time, may result in functional loss, and may require long-term, repeated evaluation and management, because it does not provide useful adjudicative guidance. However, we do explain in proposed 14.00A1b that immune system disorders can cause limitation(s) that result in an ''extreme'' loss of function.

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

In proposed 14.00A4, *Human immunodeficiency virus (HIV) infection,* we provide a brief description of HIV infection. We propose to move the first sentence in current 14.00D1 to this section. The sentence explains that HIV infection is caused by a specific retrovirus and may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions.

We also provide a cross-reference to proposed 14.00F, the section of the introductory text that addresses HIV infection in detail.

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

In proposed 14.00B, we incorporate the first sentence of the second paragraph of current 14.00B to explain what information we need to show that you have an immune system disorder. We moved the second and third sentences of the second paragraph of current 14.00B, which define our term ''appropriate medically acceptable imaging,'' to proposed 14.00C, a new section that provides definitions of terms in these listings. We propose to remove the last two sentences of the current paragraph. They explain that we will not purchase tests that may involve significant risk; however, we already include this general policy in §§ 404.1519m and 416.919m of our regulations so it is not necessary to repeat them in this section.

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

We also propose to remove the third and fourth paragraphs of current 14.00B. The third paragraph of current 14.00B provides that we need a longitudinal clinical record of at least 3 months demonstrating active disease to assess the severity and duration of your impairment. However, this is not always the case, even under the current rules. For example, individuals with HIV infection and cryptococcal meningitis (current listing 14.08B4) or Kaposi's sarcoma (current listing 14.08B8), and individuals with ankylosing spondylitis with fixation (ankylosis) of the dorsolumbar spine at 45° (current listing 14.09B2) are disabled based on those findings alone. In that case, we do not need 3 months of evidence or evidence showing active disease. Other cases may be decided with less than 3 months of evidence, while others may require more than 3 months of evidence. Therefore, we are removing this guidance because each case should be decided on an individual basis.

Proposed 14.00C—Definitions

In proposed 14.00C, we define what we mean by important terms in these listings. As already noted, we include the definition of ''appropriate medically acceptable imaging'' from the second paragraph of current 14.00B. However, we propose to replace the word ''proper'' in the second sentence of this definition with the phrase ''generally accepted and consistent with the prevailing state of medical knowledge and clinical practice'' to more clearly explain what we mean. We also propose to include in this new section the definitions of the terms ''severe'' from the sixth paragraph of current 14.00B, ''inability to ambulate effectively'' and ''inability to perform fine and gross movements effectively'' from current 14.00B6b, and ''resistant to treatment,'' ''recurrent,'' and ''disseminated'' from the second, third, and fourth paragraphs of current 14.00D2. All of these terms will apply to several, and sometimes all, of the proposed listings in this body system.

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

We also propose to move and simplify the definitions of the terms ''resistant to treatment,'' ''recurrent,'' and ''disseminated'' in current 14.00D2, primarily to remove language that we believe is unnecessary. For example, we removed the explanation that the terms ''have the same general meaning as used by the medical community.'' These changes are only editorial. We do not intend the proposed definitions to be substantively different from the current rules.

In proposed 14.00C8, we reference current 1.00F for the definition of ''major peripheral joints'' instead of restating the definition as we do in current 14.00B6a. We also propose to add the definitions of several other important terms in these listings, including in proposed 14.00C2, the term ''constitutional symptoms or signs.'' In proposed 14.00C2, we also provide brief definitions for the constitutional symptoms ''severe fatigue'' and ''malaise.'' We propose to add these definitions in response to the many comments we received that indicated that the fatigue and malaise that people who have immune system disorders experience can be very limiting.

Proposed 14.00D—What are the listed autoimmune disorders in these listings?

In proposed 14.00D, we incorporate and expand upon the information in current 14.00B1 through 14.00B6, which describe features commonly associated with each of the listed autoimmune system disorders. Throughout these sections, we refer to ''autoimmune disorders'' instead of ''connective tissue disorders'' because the phrase ''autoimmune disorders'' is more medically accurate and more frequently used. We also propose to add a new section 14.00D7 for Sjögren's syndrome because we are proposing to add new listing 14.10 for that autoimmune disorder.

In proposed 14.00D1, *Systemic lupus erythematosus* (14.02), we expand and clarify the information in current 14.00B1. In proposed 14.00D1a, *General,* we explain that systemic lupus erythematosus (SLE) may involve any organ or body system and describe by body system some potential manifestations that may be involved. We expand our explanation of how SLE is frequently characterized clinically and propose to change ''fatigability'' used in current 14.00B1 to ''fatigue'' to be consistent with how we describe this symptom throughout the immune system listings. We also add ''involuntary'' as a descriptor of weight loss to clarify that we mean weight loss due to SLE or its treatment. In proposed 14.00D1b, *Documentation of SLE,* we propose to update our rules to explain that your medical evidence will generally, but not always, show that your SLE satisfies the criteria in the ''Criteria for the Classification of Systemic Lupus Erythematosus'' by the American College of Rheumatology, found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation. This is a more up-to-date reference than the 1982 reference in the current rules.

In proposed 14.00D2, *Systemic vasculitis (14.03)*, we clarify the information in the current rule. Proposed 14.00D2a, *General,* corresponds to the first three sentences of current 14.00B2. In it, we explain that vasculitis is an inflammation of blood vessels that may occur acutely in association with adverse drug reactions, certain chronic infections, and

occasionally malignancies, and that it may also be associated with other autoimmune disorders. We also give examples of several clinical patterns in which it may occur. We propose to remove the fourth sentence of current 14.00B2, which describes cutaneous vasculitis, because the impairment varies greatly in its manifestation, may not be associated with systemic involvement, and would not be expected to result in a listing-level impairment.

Proposed 14.00D2b, *Documentation of systemic vasculitis,* corresponds to the last two sentences of current 14.00B2. In it, we describe documentation that we use to confirm the diagnosis of systemic vasculitis.

Proposed 14.00D3, *Systemic sclerosis (scleroderma) (14.04),* corresponds to current 14.00B3. We propose to revise the heading and to expand the information in the section. Proposed 14.00D3a, *General,* corresponds to the first three sentences of current 14.00B3. We propose to change the term ''Raynaud's phenomena,'' which we use in the second and third sentences of current 14.00B3, to ''Raynaud's phenomenon'' because the latter is the correct term. We make this same change in proposed listing 14.04C. In proposed 14.00D3b, *Diffuse cutaneous systemic sclerosis,* we continue to explain that, in addition to skin or blood vessels, major organ or systemic involvement may include the gastrointestinal tract, lungs, heart, kidneys, and muscle. This guidance corresponds to the fourth sentence in the current rule.

Proposed 14.00D3c, *Localized scleroderma (linear scleroderma or morphea),* is new. We propose to add this section and appropriate listings in proposed 14.04 for these disorders that originate in childhood because their disabling effects can persist into adulthood. Proposed 14.00D3c is essentially the same as proposed 114.00D3c, which we describe in detail later in this preamble.

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

In proposed 14.00D4, *Polymyositis and dermatomyositis (14.05),* we clarify the information in current 14.00B4. Proposed 14.00D4a, *General,* corresponds to the first three sentences of current 14.00B4. It describes the characteristics of polymyositis and dermatomyositis. In proposed 14.00D4b, *Documentation of polymyositis or dermatomyositis,* we describe the findings that are generally used to document these impairments. The first sentence of the proposed rule corresponds to the last sentence of current 14.00B4. We propose minor editorial revisions, including the removal of the reference to ''myositis'' because there are multiple characteristic abnormalities on muscle biopsy that support the diagnosis of polymyositis or dermatomyositis. We also propose to add a sentence to explain that people with dermatomyositis have a characteristic skin rash.

In proposed 14.00D4c, *Additional information about how we evaluate polymyositis and dermatomyositis under the listings,* we explain how we evaluate commonly occurring limitations associated with these disorders. Proposed 14.00D4c(i) corresponds to the fourth and fifth sentences of current 14.00B4. We propose to delete the example of weakness of the anterior neck flexor muscles in the sixth sentence of current 14.00B4 because we are proposing to delete the reference to the cervical muscles from listing 14.05 for reasons we explain later in this preamble. We also propose to add an example of squatting. Squatting is a common means for evaluating weakness in the pelvic girdle muscles.

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

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

We propose to delete the last sentence of current 14.00B5. The current sentence indicates that the correct designation of an ''overlap'' disorder is important for the assessment of prognosis. We believe that this sentence, while useful in treatment settings, does not provide useful adjudicative guidance.

In proposed 14.00D6, *Inflammatory arthritis (14.09)*, we expand, reorganize, and clarify the rules in current 14.00B6. Proposed 14.00D6a, *General*, corresponds to the first and fourth sentences of current 14.00B6. We continue to explain that inflammatory arthritides include a vast array of disorders that differ in cause, course, and outcome and may result in difficulties of ambulation or fine and gross movements. We edited the fourth sentence of current 14.00B6 to break it up into three shorter sentences. However, we do not intend to change the meaning of the provision.

Proposed 14.00D6b, *Inflammatory arthritides involving the axial spine (spondyloarthropathies)*, and 14.00D6c, *Inflammatory arthritides involving the peripheral joints*, correspond to the second and third sentences of current 14.00B6. In these sections, we list some disorders that may be associated with inflammatory spondyloarthropathies involving the axial spine (proposed 14.00D6b) and inflammatory arthritides affecting the peripheral joints (proposed 14.00D6c). We propose to add inflammatory bowel disease (IBD) to the lists of examples in both sections because arthritis is the most common extra-intestinal complication of IBD. In proposed 14.00D6b, we remove the examples of ''other reactive arthropathies'' and ''undifferentiated spondylitis'' now included in the second sentence of current 14.00D6 because they are non-specific and the list is not intended to be complete, only to provide some examples. Finally, we propose to update some of the terminology in this section; for example, we refer to ''psoriatic arthritis'' instead of ''psoriatic arthropathy.''

Proposed 14.00D6d, *Documentation of inflammatory arthritides*, is new. In it, we explain that generally, but not always, the diagnosis of inflammatory arthritis is made by the clinical features and serologic findings described in the most recent edition of the *Primer on the Rheumatic Diseases.*

Proposed 14.00D6e, *How we evaluate the inflammatory arthritides under the*

listings, corresponds to the information in the last two sentences of current 14.00B6, current 14.00B6c, and current 14.00B6d. We are reorganizing the text to reflect the proposed reorganization of listing 14.09, which we explain later in this preamble, and to clarify it.

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

• Proposed 14.00D6e(ii) explains proposed listings 14.09B (current listing 14.09D), 14.09C2 (current listing 14.09E), and 14.09D. These listings do not describe a single impairment manifestation that results in an ''extreme'' limitation. Rather, they describe combinations of impairment manifestations that should result in an ''extreme'' limitation or in ''marked'' limitations in at least two areas of functioning. We also incorporate the provision in the first sentence of current 14.00B6d that extra-articular impairments may meet listings in other body systems.

• Proposed 14.00D6e(iii) corresponds to the third and fourth sentences of current 14.00B6d. It explains that extra-articular features of inflammatory arthritis may involve any body system and lists examples of commonly occurring extra-articular impairments by body system. We propose to reorganize and expand the list of examples of such impairments and to clarify the body systems to which they belong.

• Proposed 14.00D6e(iv) and 14.00D6e(v) correspond to the last sentence of current 14.00B6. In proposed 14.00D6e(iv), we replace ''persistent'' with ''permanent'' and remove ''without ongoing inflammation'' to clarify that we evaluate permanent deformity of a major peripheral joint under listing 1.02 when it is the dominant feature of your impairment. Proposed 14.00D6e(v) explains that we use listing 1.03 to evaluate surgical reconstruction of a major weight-bearing joint.

• Proposed 14.00D6e(vi) would clarify that we evaluate your impairment under any appropriate listing when you have both inflammation and chronic deformities.

We are not including the provisions of current 14.00B6e in proposed 14.00D6. Current 14.00B6e provides that the fact that an individual is dependent on steroids, or any other drug, for the control of inflammatory arthritis is insufficient in itself to establish disability. We added it to part A of our listings in 2002 for consistency with 114.00E6, a provision we added to part B of the listings at the same time (66 FR 58010, 58020 (2001)). We are proposing to remove that provision for reasons we explain below in our summary of the proposed rules in part B. Therefore, we are proposing to remove this provision in part A for consistency with that change. However, in proposed 14.00G3, we continue to state that we will consider the adverse side effects of treatment, including the adverse effects of corticosteroids, to ensure that our adjudicators remember to consider the side effects an individual might experience from steroids and any other treatment.

Proposed 14.00D7, *Sjögren's syndrome (14.10)*, is new. As already noted, we are proposing to add a new listing for Sjögren's syndrome. In connection with that proposed listing, proposed 14.00D7a, *General*, explains the features of the disorder, including its resulting symptoms and possible complications. We also list organ systems that may be involved and note that Sjögren's syndrome may be associated with other autoimmune disorders. In proposed 14.00D7b, *Documentation of Sjögren's syndrome*, we also explain that if you have Sjögren's syndrome, your medical evidence will generally, but not always, show that your disease satisfies the criteria in the ''Criteria for the Classification of Sjögren's Syndrome'' found in the most recent edition of the *Primer on the Rheumatic Diseases*.

Proposed 14.00E—How do we evaluate immune deficiency disorders, excluding HIV infection (14.07)?

In proposed 14.00E, we add a new section describing how immune deficiency disorders (excluding HIV infection) are classified, documented, and evaluated. This section has four subsections.

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

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

In proposed 14.00E3, *Immune deficiency disorders treated by stem cell transplantation*, we explain how we evaluate immune deficiency disorders that are treated in this way. In proposed 14.00E3a, *Evaluation in the first 12 months*, we explain that if you undergo stem cell transplantation we will consider you disabled until at least 12 months from the date of the transplant. This is the same provision that we use for most malignancies treated by bone marrow or stem cell transplants in the neoplastic listings. In 13.00L4 of those listings, we also included a special provision for autologous bone marrow transplants—transplants using your own stem cells (69 FR 67034). We do not include such an alternative provision in these proposed rules because people with immune deficiency disorders receive allogeneic transplants—that is, stem cells taken from other people. Also, we propose to use ''stem cell transplantation'' instead of ''bone marrow or stem cell transplantation'' in this proposed section and in proposed listing 14.07B because ''stem cell transplantation'' is a broader term that encompasses different sites for obtaining hematopoetic (blood-forming) stem cells, including bone marrow, peripheral blood, and umbilical cord blood. In proposed 14.00E3b, *Evaluation after the 12-month period has elapsed*, we explain that, after that period has elapsed, we consider any demonstrable residuals of your immune deficiency disorder including any residual impairment(s) resulting from your treatment. The provision also is based on 13.00L4 in our malignant neoplastic diseases listings.

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

Proposed 14.00F—How do we evaluate human immunodeficiency virus (HIV) infection?

In proposed 14.00F, we incorporate, update, and expand information on HIV infection contained in current 14.00D3 through 14.00D7. We also make nonsubstantive editorial changes.

As already noted, we propose to move the first sentence of current 14.00D1 to proposed 14.00A4. Therefore, we begin proposed 14.00F with what is now the second sentence of current 14.00D1. It is a reminder that an individual with HIV infection need not meet the Centers for Disease Control definition of acquired immune deficiency syndrome (AIDS) to meet or medically equal the criteria of listing 14.08. We have made minor editorial changes to the sentence, but we do not intend to change its meaning.

We propose to move the provisions of current 14.00D2 to other sections in the proposed rules. In the first four paragraphs of current 14.00D2, we define the terms ''resistant to treatment,'' ''recurrent,'' and ''disseminated,'' and we would now define those terms in proposed 14.00C. In the fifth paragraph of current 14.00D2, we define ''significant involuntary weight loss'' for purposes of current listing 14.08I (which has become listing 14.08H in these proposed rules). In the proposed rules, we include this definition in 14.00F5.

Like current 14.00D3, proposed 14.00F1 is in two major sections: A section explaining how we document the diagnosis of HIV infection definitively (14.00F1a) and a section explaining how we document the diagnosis of HIV infection when we do not have definitive evidence (14.00F1b). In proposed 14.00F1, *Documentation of HIV infection*, we incorporate and update the information in current 14.00D3 to explain the laboratory tests or other evidence we accept as documentation of HIV infection. Proposed 14.00F1a, *Documentation of HIV infection by definitive diagnosis*, corresponds to current 14.00D3a. We propose to update and expand this section to include newer laboratory diagnostic techniques that did not exist or were not widely used when we published the current rules in 1993.

- Proposed 14.00F1a(i), for HIV antibody tests, corresponds to current 14.00D3a(i). We propose only nonsubstantive editorial changes.
- Proposed 14.00F1a(ii) is new. It would add positive ''viral load'' tests for HIV infection, such as quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT–PCR), that were not widely available when we published the current rules.
- Proposed 14.00F1a(iii) is for HIV DNA detection by polymerase chain reaction (PCR). We include it as an example of an ''other test'' in current 14.00D3a(iii) because it was not widely available when we published the current rules.
- Proposed 14.00F1a(iv), for HIV antigen, corresponds to current 14.00D3a(ii).
- Proposed 14.00F1a(v) is new. It would add a positive viral culture for HIV from peripheral blood mononuclear cells (PBMC) as another test that definitively documents HIV infection. Even though it is not commonly used, we will accept it as definitive evidence if it is in your medical records.
- Proposed 14.00F1a(vi), for other tests that are highly specific for detection of HIV, corresponds to the first paragraph in current 14.00D3a(iii).

Proposed 14.00F1b, *Other acceptable documentation of HIV infection*, corresponds to current 14.00D3b. It explains what documentation of HIV infection we will accept instead of definitive laboratory testing. The proposed rule is essentially the same as the current rule except for nonsubstantive editorial changes.

In proposed 14.00F2, *CD4 tests*, we combine the provisions in the second undesignated paragraph after current 14.00D3a(iii) and the second paragraph in current 14.00D4a. We specify that, even though a reduced CD4 count or percent alone does not establish a definitive diagnosis of HIV infection, a CD4 count below 200/mm^3 or 14 percent along with clinical findings does offer supportive evidence of opportunistic infections without a definitive diagnosis. This is because a CD4 count below 200 or 14 percent is an indicator of an increased susceptibility to developing opportunistic infections. We also make nonsubstantive editorial changes.

In proposed 14.00F3, *Documentation of the manifestations of HIV infection*, we incorporate the information in current 14.00D4 with nonsubstantive editorial changes. Like proposed 14.00F1 and current 14.00D4, proposed 14.00F3 is divided into two main parts. The first section explains how we document manifestation of HIV infection definitively (14.00F3a), and the second section explains how we document manifestations of HIV infection when we do not have definitive evidence (14.00F3b).

Proposed 14.00F3a, *Documentation of the manifestations of HIV by definitive diagnosis*, incorporates the first paragraph in current 14.00D4a.

In proposed 14.00F3b, *Other acceptable documentation of the manifestations of HIV infection*, we incorporate information that is in the first paragraph of current 14.00D4b. We propose to revise the language of this paragraph both editorially and to clarify our original intent. In the current rule, we indicate that ''if no definitive laboratory evidence is available, manifestations of HIV infection may be documented by medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence.'' The sentence may imply that we need to have all of the things listed (medical history *and* clinical findings *and* laboratory findings *and* diagnosis(es)) to determine that you have a manifestation of HIV infection when we do not have definitive laboratory findings. That is not our intent, so we are clarifying in the proposed rule that we may need only some of this information to make a finding that you have a manifestation of HIV infection, depending on the prevailing state of medical knowledge and clinical practice. We also propose to clarify what we mean by ''laboratory findings'' in this context; that is, laboratory findings that do not in themselves definitively establish the existence of a diagnosis of an HIV-related manifestation.

In 14.00D4 of the current rules we provide specific guidance for documenting one particular manifestation of HIV infection without definitive evidence: cytomegalovirus (CMV) disease. In proposed 14.00F3b, we expand the section to include two additional manifestations. In proposed 14.00F3b(i), we add guidance to explain that Pneumocystis carinii pneumonia (PCP) is frequently diagnosed presumptively without definitive evidence and to provide examples of evidence that is supportive of a presumptive diagnosis of PCP. We also note that Pneumocystis carinii is now known as Pneumocystis jiroveci; however, ''PCP'' remains in common usage for the pneumonia caused by this organism.

In proposed 14.00F3b(ii), we incorporate and expand the information now in the second paragraph of current 14.00D4b, regarding the documentation of CMV disease. We propose to clarify that a positive serology test for CMV identifies a ''history'' of infection but does not confirm an ''active'' disease process. We do not include ''documentation of CMV disease requires confirmation by biopsy'' as in the last sentence of the second

paragraph of current 14.00D4 because we are providing information on documentation other than definitive laboratory findings. Also, instead of stating that we can use generally acceptable methods to confirm the diagnosis of CMV, we provide examples of evidence, such as fever and positive CMV serology test, that are supportive evidence of a presumptive diagnosis of CMV disease.

In proposed 14.00F3b(iii), we add guidance on how toxoplasmosis of the brain is presumptively diagnosed since the definitive method of diagnosing toxoplasmosis of the brain by biopsy is not commonly performed.

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

In proposed 14.00F5, *involuntary weight loss*, we incorporate the last paragraph of current 14.00D2 with nonsubstantive editorial changes, including a change that reflects our proposal to redesignate listing 14.08I to listing 14.08H.

Proposed 14.00G—How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

In the current rules, we refer to treatment and its effects in four places.

- In the third paragraph of 14.00B, we provide that, for connective tissue diseases, we need a longitudinal clinical record of at least 3 months demonstrating active disease despite prescribed treatment, with the expectation that the disease will remain active for 12 months.
- In the fifth paragraph of 14.00B, we explain that ''the chronic adverse effects of treatment (e.g., corticosteroid-related ischemic necrosis of bone) may result in functional loss'' in individuals with connective tissue disease.
- In 14.00B6e, we explain that the fact that an individual with inflammatory arthritis is dependent on steroids or any other drug for the control of the arthritis is not in itself sufficient to establish that the individual is disabled. We also explain that we must evaluate each case on its own merits, taking into consideration any adverse effects of treatment.
- In 14.00D7, *Effect of treatment,* we provide three paragraphs discussing how we consider treatment in people with HIV infection. This section explains that we must consider both the positive effects and negative side effects of treatment for HIV infection and its manifestations, special considerations in evaluating treatment in individuals with HIV infection and, briefly, the kinds of evidence we need.

We are proposing to remove the provisions in the third paragraph of 14.00B and paragraph 14.00B6e. Neither of those sections nor the other current rules we will continue to use contain provisions that explain in detail how we evaluate the positive effects and negative side effects of treatment in individuals who have autoimmune disorders and immune deficiency disorders apart from HIV infection. Also, most current treatments for HIV infection came into use, or came into wide use, after we first published listing 14.08 in 1993. As a consequence, we believe that current 14.00D7 needs to be updated to reflect the newer and more widely used treatments and treatment protocols for HIV infection and to reflect the considerable medical experience that has been gained since 1993 about the long-term effects, usefulness, and limitations of such treatments.

Therefore, we propose to add a new separate section 14.00G—*How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?* The new section would address in one place issues of treatment that are common to all three types of immune system disorders as well as issues of treatment that are unique to each type of disorder, including treatment that is specifically for HIV infection. We do not propose to remove any guidance about treatment for HIV infection that is still relevant, only to move it to this new section. In fact, we propose to expand and update our rules to reflect what has been learned in applying different treatments for HIV infection since we published the current rules more than a decade ago. The provisions for addressing both the positive effects and negative side effects of treatment in individuals who have autoimmune disorders and immune deficiency disorders other than HIV infection would be new in these listings and, we believe, would provide useful adjudicative guidance that is lacking in our current rules.

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

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

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

In proposed 14.00G2, *Variability of your response to treatment,* we explain what we mean by this factor in terms of both HIV infection and other immune

system disorders. This proposed rule is based on the language of the second paragraph in current 14.00D7 and the second sentence of the third paragraph of that section. However, we propose to expand that guidance and to apply it to all other immune system disorders in addition to HIV infection. For example, we explain in a general way applicable to all immune system disorders that some individuals may show an initial positive response to drug treatment (or a combination of drugs), but the initial positive response may be followed by a decrease in the effectiveness of the medication.

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

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

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

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

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

Proposed 14.00H—How do we consider your symptoms, including your constitutional symptoms or pain?

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

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

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

Many people said that we should recognize how immune system disorders can affect an individual's functioning. Many people described physical symptoms, such as pain, fatigue, and malaise, as well as mental symptoms, including loss of memory, loss of concentration, and depression. Commenters stressed that these symptoms could be very severe. A number of people indicated that the fatigue associated with these disorders was not merely a feeling of tiredness but a more profound and debilitating experience. Many people also noted that the impairments could be both episodic and variable in intensity, with some people experiencing ''good'' or relatively good days interspersed with days in which they were unable to function. They pointed out that there was a need for the rules to recognize the longitudinal effect of these episodic limitations on the ability to work. Other people pointed out that there is often comorbidity of immune system disorders; that is, many people have features of more than one immune system disorder. In those cases, the symptoms and limitations are multiplied to an effect that is worse than simply adding them up. These commenters said that under the current listings there is no adequate way to assess these multiplied effects. Many people also pointed out the effect that stress can have on the medical condition and symptomatology of individuals who have immune system disorders. Other people described the debilitating effects of treatment, not only the side effects, but sometimes the need to follow a very rigorous and time-

consuming schedule of treatment that in itself can be limiting.

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

We carefully considered these comments and are proposing a number of changes throughout the introductory text to the immune system listings to address them. We are proposing to significantly expand our guidance about specific immune system disorders and the effects of treatment. We agree with those commenters who suggested that we include the same kind of criteria for evaluating the overall functional impact of other immune system disorders as we provide in current listing 14.08N for people who have HIV infection. Therefore, we are proposing to add criteria similar to those in current listing 14.08N for each of the listed impairments in this body system. The proposed listings for evaluating functioning for other immune system disorders would be 14.02B, 14.03B, 14.04D, 14.05E, 14.06B, 14.07C, 14.09D, and 14.10B. We are also proposing to redesignate current listing 14.08N as 14.08K for reasons we explain below.

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

Like current 14.00D8, proposed 14.00I includes eight paragraphs. Except as described below, we propose to revise each paragraph so that it applies not only to HIV infection but to the other immune system disorders as well. For example, in the first paragraph of current 14.00D8 we explain that current listing 14.08N (proposed listing 14.08K) establishes standards for evaluating manifestations of HIV infection that do not meet the criteria of any of the preceding listings within 14.08; that is, current listings 14.08A–14.08M. We also explain that we use listing 14.08N both for manifestations that are listed in the preceding listings within 14.08 and for manifestations that are not listed at all. We propose to modify this language so that it applies to all of the immune system disorders within this body system. We also propose minor editorial changes throughout the paragraphs.

The following are other changes we propose to make in this section.

In proposed 14.00I2, we propose to remove the first sentence in the second paragraph of current 14.00D8, which explains that for individuals with HIV infection, we assess listing-level severity under current listing 14.08N based on the functional limitations imposed by the impairment. We believe that this point is already made in proposed 14.00I1 and that it is unnecessary to repeat it in proposed 14.00I2. We propose to revise the second sentence, which says that we must consider the full impact of ''signs, symptoms, and laboratory findings'' on the individual's ability to function. We believe that this guidance may not clearly explain what we intend. Therefore, we propose to revise it to explain that when we use one of the listings cited in 14.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder(s) on your ability to function on a sustained basis.

In proposed 14.00I3–14.00I8, which correspond to the last six paragraphs in current 14.00D, we propose to update our rules to make their language more consistent with our other rules that define the term ''marked'' and the domains of functioning. We do not intend these changes to be substantively different from the current rules. We also propose to include references to both pain and fatigue throughout proposed 14.00I6–14.00I8 as symptoms that may cause limitations. The current rules are not consistent in this regard.

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

Proposed 14.00J1 and 14.00J3 would replace the guidance we now provide in the first and third paragraphs of current 14.00D6. As in other provisions throughout the introductory text, we propose to revise the language to make it apply generally to all immune system disorders, not just HIV infection. Also, we propose to remove guidance that is already covered in other sections in the introductory text of these proposed rules, such as the guidance that individuals may have signs or symptoms of a mental impairment or of another physical impairment.

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

Therefore, we propose to add new section 14.00J2 to explain that immune system disorders can have effects in other body systems; we also provide a list of examples of those effects in each of the relevant body systems with references to other body systems listings. The proposed provisions are based on language in the second paragraph of current 14.00D6, which is currently relevant only to the evaluation of HIV infection, and on the reference listings we are proposing to remove. In the latter case, we are also expanding the information to provide specific examples of impairments that may be caused by autoimmune disorders.

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

Proposed 14.00J2k provides examples of allergic disorders (including skin disorders) that individuals with immune system disorders may have. It would replace current 14.00C.

How are we proposing to change the criteria in the listings for evaluating immune system impairments in adults?

14.01 Category of Impairments, Immune System Disorders

The following is a detailed explanation of the significant changes in the proposed listings. Some changes are

common to several listings so we describe them first.

1. We propose to remove all of the reference listings from this body system. Every current listing section in this body system, except listing 14.07, includes reference listings. Reference listings are listings that are met by satisfying the criteria of another listing. For example, current listing 14.02A1, Joint involvement, is met when the resulting impairment meets the criteria of any appropriate listing in the musculoskeletal body system, 1.00ff. Current listing 14.08G1, for HIV infection with anemia, requires evaluation under current listing 7.02. Therefore, these listings are redundant because impairments that meet these listings must meet the requirements of other listings. We are removing reference listings from all of the body systems as we revise them. As already noted, instead of using reference listings, we propose to provide guidance in 14.00J of the introductory text stating that we may evaluate the resulting impairment of an immune system disorder under any affected body system.

2. We propose to revise current listings 14.02B, 14.03B, 14.04B, and 14.09D (proposed listings 14.02A, 14.03A, 14.04A, and 14.09B) as follows:

- We propose to remove the criterion for ''significant, documented'' constitutional symptoms or signs in each of these listings because we define the constitutional symptoms and signs in proposed 14.00C2. Moreover, it is unnecessary to specify ''documented'' because we always need to document the existence of any symptom or sign in any disability claim.
- Each of these current listings, except current listing 14.09D, also requires you to have all four of the constitutional symptoms or signs: Severe fatigue, fever, malaise, and involuntary weight loss. We propose to revise this requirement to ''at least two'' of the constitutional symptoms or signs instead of all four, because we believe that the requirement in the current listing is too severe. We believe that any individual with an autoimmune disorder involving two or more organs/body systems with one organ/body system involved to at least a moderate level of severity and who has at least two of the constitutional symptoms and signs in these listings will have an impairment that precludes any gainful activity. We also have added ''involuntary'' as a descriptor of weight loss in proposed listings 14.02A, 14.03A, 14.04A, 14.05E, 14.06A, 14.07C, 14.08K, 14.09B, and 14.10A for the same reason we explained earlier in the preamble.
- In proposed listings 14.02A, 14.03A, and 14.04A, which correspond to current listings 14.02B, 14.03B, and 14.04B, we propose to remove the reference to ''lesser involvement'' because we propose to remove the current reference listings to which these rules refer. We also believe the phrase is unnecessary—the severity of the impairment is demonstrated by the remaining criteria.

3. As we have already noted under the explanation of proposed 14.00I, we propose to add listings based on repeated manifestations accompanied by functional limitations and modeled after current listing 14.08N for each of the other immune system disorders. The proposed new listings are

- 14.02B for SLE,
- 14.03B for systemic vasculitis,
- 14.04D for systemic sclerosis (scleroderma),
- 14.05E for polymyositis and dermatomyositis,
- 14.06B for undifferentiated and mixed connective tissue disease,
- 14.07C for immune deficiency disorders (other than HIV infection),
- 14.09D for inflammatory arthritides, and
- 14.10B for Sjögren's syndrome.

Each listing requires you to have:

- The specified immune system disorder for that listing,
- Repeated manifestations that do not satisfy the requisite findings of another listing for the specified immune system disorder,
- At least two of the constitutional symptoms or signs, and
- ''Marked'' limitation in one of three domains of functioning: Activities of daily living, social functioning, or completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

We explain what we mean by ''repeated'' in proposed 14.00I3 and by ''marked'' in proposed 14.00I4–5.

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

Proposed Listing 14.04—Systemic sclerosis (scleroderma)

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

We also propose minor clarifications in the language of the current listing. Current listing 14.04C describes ''[g]eneralized scleroderma with digital contractures.'' We propose to clarify that ''digital'' refers to either the toes or the fingers, and to list the effects in the toes separately from the effects in the fingers, in proposed listings 14.04B1 and 14.04B2, respectively. We also propose to remove the requirement for ''generalized'' scleroderma (that is, systemic sclerosis) because the very serious digital contractures described in the proposed listings would in themselves be disabling regardless of whether the scleroderma is generalized.

Proposed listing 14.04C corresponds to current listing 14.04D. We propose to change ''Raynaud's phenomena'' in current listing 14.04D to ''Raynaud's phenomenon'' for the same reason already described in the explanation of proposed 14.00D3. We propose to remove the word ''[s]evere'' as a descriptor of Raynaud's phenomenon in this listing because it is unnecessary given the severity of the impairment demonstrated by the remaining criteria, such as ischemia with ulcerations of fingers or toes, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively. As in proposed listing 14.04B, we also propose to clarify that ''digital'' refers to fingers or toes.

In proposed listing 14.04C, we also propose to revise the criteria in current listing 14.04D to provide a better description of listing-level Raynaud's phenomenon. The criteria in current listing 14.04D require severe Raynaud's phenomenon that is characterized by digital ulcerations, ischemia, or gangrene. We believe that this does not describe an impairment that precludes any gainful activity in every case. Therefore, in proposed listing 14.04C1 we would provide criteria for Raynaud's phenomenon characterized by gangrene of a toe or finger in at least two extremities, or a toe and finger to indicate an impairment that would preclude any gainful activity. We do not propose to require that the gangrene result in the inability to ambulate effectively or to perform fine and gross movements effectively because the presence of gangrene of a toe or finger

in at least two extremities or in a toe and finger by itself is an indication of a very serious impairment. In proposed listing 14.04C2, we provide criteria for ischemia with ulcerations of the toes or fingers that results in the inability to ambulate effectively or to perform fine and gross movements effectively; Raynaud's phenomenon characterized only by ischemia with ulcerations does not by itself describe an impairment that would necessarily result in an extreme loss of function. Also, ulcerations are an outcome of ischemia, so we propose to revise the language so that ischemia and ulcerations are not listed as though they are separate entities, as in the current rule.

Proposed Listing 14.05—Polymyositis and Dermatomyositis

Proposed listing 14.05A corresponds to current listing 14.05A. We propose to replace the word ''severe'' as a descriptor of proximal limb-girdle weakness with the more accurate ''resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 14.00C6 and 14.00C7.'' We also propose to change ''shoulder and/or pelvic'' muscle weakness to ''pelvic or shoulder'' muscle weakness because pelvic muscle weakness can result in the inability to ambulate effectively and shoulder muscle weakness can result in the inability to perform fine and gross movements effectively. Therefore, either one of these findings could be sufficient in itself to show disability and the ''and'' is unnecessary.

Proposed listing 14.05B corresponds to current listing 14.05B1. We propose to remove the requirements in the opening paragraph for less severe limb-girdle muscle weakness than in 14.05A, associated with cervical muscle weakness, because impaired swallowing or impaired respiration may result in listing-level limitations without the presence of either of those findings. We also propose to remove the phrase ''to at least a moderate level of severity'' because the criterion in proposed 14.05B is of at least a moderate level of severity, making this language unnecessary. We propose to revise ''impaired swallowing with dysphagia'' to ''impaired swallowing (dysphagia)'' because dysphagia means impaired swallowing. We propose to revise ''episodes of aspiration'' to ''aspiration'' because of the progressive nature of muscle weakness that results from polymyositis or dermatomyositis. Once an episode of aspiration is documented, further documentation of multiple episodes is unnecessary. In addition, we propose to replace ''cricopharyngeal weakness'' with ''muscle weakness'' in proposed 14.05B because impaired swallowing with dysphagia and aspiration may result from muscles other than the cricopharyngeal muscles.

Proposed listing 14.05C corresponds to current listing 14.05B2. We propose to remove the requirements in the opening paragraph of current 14.05B2 for the same reasons as in the above paragraph for proposed listing 14.05B.

Proposed listing 14.05D, Diffuse calcinosis, is a new adult listing and has the same criteria as in proposed listing 114.05D for children, which we describe in detail later in this preamble. We propose to add this listing for individuals who had a form of the disorder as children and who still have listing-level functional limitations as adults.

Proposed Listing 14.06—Undifferentiated and Mixed Connective Tissue Disease

We propose to change the heading of current 14.06 to update it and to more accurately describe the disorders we evaluate under this listing.

Current listing 14.06 is entirely a reference listing, requiring evaluation under current listings 14.02A, 14.02B, or 14.04. We propose to change it to a stand-alone listing containing its own criteria. Proposed listing 14.06A uses the same criteria as in proposed listings 14.02A, 14.03A, and 14.04A for involvement of two or more body systems to at least a moderate level of severity and at least two constitutional symptoms or signs. Proposed listing 14.06B incorporates the same functional criteria for the evaluation of repeated manifestations of undifferentiated and mixed connective tissue disease as the other listings in this body system.

Proposed Listing 14.07—Immune Deficiency Disorders, Excluding HIV Infection

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

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

Proposed listing 14.07A is essentially the same as current listing 14.08M (proposed listing 14.08J) which describes individuals with HIV infection whose immune systems are so compromised that they frequently become ill. However, unlike current listing 14.07, current listing 14.08M provides that the infections must occur three times in a 12-month period, not three times in only a 5-month period. Current listing 14.08M is also more precise. It explains how severe the infections need to be by reference to resistance to treatment or a requirement for hospitalization or intravenous treatment. It also specifies six types of infections. We believe that the criteria in current listing 14.08M for people with HIV infection are equally as applicable to individuals with other kinds of immune deficiency disorders, and that they would be more inclusive than the criteria in current listing 14.07.

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

Proposed listing 14.07C would incorporate the same functional criteria for the evaluation of repeated manifestations of immune deficiency disorders (excluding HIV infection) as in the other proposed listings in this body system and for the same reasons as described above.

Proposed Listing 14.08—Human Immunodeficiency Virus (HIV) Infection

We do not propose any substantive changes to the criteria in listing 14.08. We have carefully considered the advances in treatment and consequent longevity that have occurred since we published the current rules in 1993. However, we do not believe that there has been sufficient progress in the treatment and control of HIV infection to warrant any change in these rules. Moreover, even as some problems of people who have HIV infection appear to be improved, new problems have arisen to take their place. Advances in treatment are a case in point. While there have been significant strides in the treatment of HIV infection that have improved mortality, the treatment itself is often disabling both in terms of its side effects and its administration. Many people must structure their days and nights around their treatment, and any lapse can have dire consequences. Some people respond to treatment initially but become unresponsive without warning. Others have only limited success with their treatments. Relatively few people with HIV infection are considered ''well.'' Therefore, from the standpoint of Social Security disability policy and efficient

administration of the disability programs, we have not seen sufficient evidence to persuade us to propose any significant changes in this listing.

As already noted, we propose to remove current reference listings throughout this body system, including the reference listings in listing 14.08. This would result in the removal of several specific listings within 14.08 and the redesignation of some of the current listings; for example, current listing 14.08N would become listing 14.08K. Where we propose to remove a reference listing, however, we have ensured that we provide guidance in the introductory text about where to evaluate the impairment. For example, current listing 14.08A4, for HIV infection with syphilis or neurosyphilis, is a reference listing that says only to consider the impairment under the criteria for the affected body system, such as 2.00 (special senses and speech), 4.00 (cardiovascular system), or 11.00 (neurological). Although we propose to remove this reference listing, we include this same guidance in proposed 14.00J2l.

We also propose to clarify some of the rules. We propose to reorganize the language in listing 14.08B2 to make it clearer that we evaluate under this listing candidiasis involving the esophagus, trachea, bronchi, or lungs, or at another site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes. We propose to move current listing 14.08C2, for PCP, from the listing for protozoan and helminthic infections to the listing for fungal infections because the organism that causes PCP is now known to be a fungus. We redesignate it as proposed listing 14.08B7.

We propose to redesignate current listing 14.08N as proposed listing 14.08K. We propose to expand our guidance on manifestations we evaluate under proposed listing 14.08K by adding ''pancreatitis, hepatitis, peripheral neuropathy, glucose intolerance, muscle weakness, and cognitive or other mental impairments'' as new examples. We also expand our list of signs or symptoms by adding ''nausea, vomiting, headaches, or insomnia.''

We propose minor changes to the language of the functional criteria in proposed listing 14.08K from the current language in listing 14.08N. For example, we would replace the words ''restriction'' in current listing 14.08N1 and ''difficulties'' in current listings 14.08N2 and 14.08N3 with the word ''limitation'' in proposed listings 14.08K1, 14.08K2, and 14.08K3. We propose to make this change because ''limitation'' is a clearer term that we use throughout our rules.

Proposed Listing 14.09—Inflammatory Arthritis

We are redesignating current listing 14.09D as proposed listing 14.09B, current listing 14.09B as proposed listing 14.09C1, and current listing 14.09E as proposed listings 14.09C2 to put them in a more logical order. In the proposed rules, listing 14.09A would describe persistent inflammation or deformity of major peripheral joints that alone is disabling, while listing 14.09B would describe disability with lesser inflammation or deformity of major peripheral joints, organ involvement, and constitutional symptoms. Listing 14.09C would describe listing-level inflammatory arthritis of the spine. Proposed listing 14.09C1 would describe disability based only on fixation (ankylosis) of the spine, while listing 14.09C2 would describe disability based on a lesser degree of ankylosis of the spine with organ involvement. Proposed listing 14.09D would be the same functional listing we include in all of the proposed immune system listings and would apply to inflammatory arthritis affecting any joints.

Proposed listing 14.09A corresponds to current listing 14.09A. We propose to remove the requirement for a history of joint pain, swelling, and tenderness from this listing because it is unnecessary and to provide only that joint inflammation must be ''persistent.'' (We do refer to joint pain, swelling, and tenderness in proposed 14.00D6a.) Persistent joint inflammation or deformity in two or more major peripheral joints resulting in the inability to ambulate effectively or inability to perform fine and gross movements effectively is in itself indicative of an impairment that would preclude any gainful activity. For the same reasons, we also propose to remove the requirement for ''signs on current physical examination.'' We would not need signs of joint inflammation on a current physical examination when we have medical evidence documenting that you have inflammatory arthritis that results in the inability to ambulate effectively or inability to perform fine and gross movements effectively. Also, because of the episodic nature of inflammatory arthritis a current physical examination could show a brief period of improvement for a few days even though your longitudinal medical records may show persistent joint inflammation that results in the inability to ambulate effectively or inability to perform fine and gross movements. We propose to change ''two or more major joints'' to ''two or more major peripheral joints'' to distinguish these joints from the joints of the spine. We define ''major peripheral joints'' in proposed 14.00C8.

Proposed listing 14.09B corresponds to current listing 14.09D. The revisions in proposed 14.09B are similar to those in proposed listing 14.09A for the same reasons and to make it clearer that this listing requires joint inflammation in one or more major peripheral joints. Proposed 14.09B continues to require less joint involvement than in A, but we would no longer require ''lesser extra-articular features than in C'' because ''C'' refers to current reference listing 14.09C which we are proposing to remove. Instead, we require ''extra-articular features that do not satisfy the criteria of a listing.'' Proposed listing 14.09B1 corresponds to current listing 14.09D2 with nonsubstantive editorial changes to make it consistent with how we present this criterion throughout these listings. Proposed listing 14.09B2 corresponds to current listing 14.09D1 except that we have removed the phrase ''significant, documented'' for reasons we have already explained. We also propose to correct an error in current listing 14.09D1. The explanatory abbreviation, ''e.g.'' (for example) in current listing 14.09D1 inaccurately indicates that the four constitutional symptoms or signs, that is, fatigue, fever, malaise, and involuntary weight loss, are only examples when they are in fact a complete list. Consistent with changes in other proposed listings, we propose to require at least two of the constitutional symptoms or signs because we believe that the criteria in proposed listing 14.09B are indicative of an impairment that precludes any gainful activity.

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

Proposed listing 14.09C2 corresponds to current listing 14.09E. We propose to reorganize this listing to make it more consistent with the structure and

criteria that we use in the proposed listings for other autoimmune disorders. We propose to remove the phrase ''with lesser deformity than in B,'' which describes a deformity that is less than the fixation ''of the dorsolumbar or cervical spine at 45° or more of flexion'' under current listing 14.09B, and to replace it with fixation ''at 30° or more of flexion (but less than 45°).'' We believe that this would be a clearer and more specific criterion that would help to provide greater uniformity in adjudications under this listing. We propose to remove the phrase ''lesser extra-articular features than in C'' because it refers to current reference listing 14.09C, which we are proposing to remove. We also propose to remove the phrase ''with signs of unilateral or bilateral sacroiliitis'' because the criteria in the proposed listing would be sufficient to show listing-level severity without this requirement, and the phrase ''with the extra-articular features described in 14.09D'' because it is unnecessary language.

Proposed Listing 14.10—Sjögren's Syndrome

Proposed listing 14.10 is new. We are proposing to add it in response to comments we received that Sjögren's syndrome is distinct from other immune system disorders with unique aspects that the current immune system listings do not address.

Although individuals with Sjögren's syndrome can qualify under current listings 14.03 and 14.09, and other listings, we believe that it is now appropriate to list Sjögren's syndrome separately in these listings. We propose to use the same two listing criteria for establishing listing-level severity as in the other proposed listings for autoimmune disorders because Sjögren's syndrome is an autoimmune disorder that can cause the same kinds of constitutional symptoms and signs as other autoimmune disorders, and because it can be as functionally limiting as other autoimmune disorders. Proposed listing 14.10A is the same as proposed listings 14.02A, 14.03A, 14.04A, and 14.06A, and proposed listing 14.10B is the same as proposed listings 14.02B, 14.03B, 14.04D, 14.05E, 14.06B, and 14.09D. We also provide a new separate section in the introductory text that describes the unique features of Sjögren's syndrome, proposed 14.00D7.

What revisions are we proposing to make in the immune system disorder listings for children—114.00?

As in proposed 14.00 in the adult rules, we propose to change the name of this body system to ''Immune System Disorders.''

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

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

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

In proposed 114.00A1b, we incorporate the first sentence in the last paragraph of current 114.00B, which explains that immune system disorders may affect growth, development, attainment of age-appropriate skills, and performance of age-appropriate activities in children. We propose to revise the sentence by adding the phrase ''or their treatment.'' We also propose to remove the phrase ''attainment of age-appropriate skills'' because it is redundant of ''development.''

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

Proposed 114.00D—What are the listed autoimmune disorders in these listings?

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

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

In proposed 114.00D3c, *Localized scleroderma (linear scleroderma or morphea)*, we describe features of focal forms of scleroderma in children. These disorders occur primarily in children and are more common than systemic sclerosis in children. In proposed 114.00D3c(i), we explain that the extent of involvement and the location of lesions are important factors in determining the limitations resulting from scleroderma. We also note that it may be appropriate to evaluate the limitations resulting from these impairments under the musculoskeletal (101.00) listings. In proposed 114.00D3c(ii), we describe features of isolated morphea of the face and explain that it may be more appropriate to evaluate the limitations from these disorders under the affected body system, such as the special senses listings (102.00) or mental disorders listings (112.00). In 114.00D3c(iii) we describe features of chronic variants of these syndromes and explain that it is appropriate to evaluate the limitations from these disorders under the affected body system, such as the musculoskeletal listings (101.00) or respiratory system listings (103.00).

In proposed 114.00D4, *Polymyositis and dermatomyositis (114.05)*, we note (in 114.00D4a, *General*) that polymyositis occurs rarely in children and describe the features of dermatomyositis that occur differently in children than in adults. In children, polymyositis and dermatomyositis usually do not occur in association with

malignancies. For this reason, we do not include a reference to malignancy or provide guidance that we will evaluate malignancies under the malignant neoplastic diseases listings (113.00ff) in proposed 114.00D4, as we do for adults in proposed 14.00D4. However, unlike in the adult rules, we include a reference to calcinosis for children because some children develop calcinosis late in the disease. Also, when dermatomyositis involves other organs or body systems, we evaluate the involvement under the affected body system. In proposed 114.00D4b, *Documentation of polymyositis or dermatomyositis*, we note that magnetic resonance imaging (MRI) showing muscle inflammation or vasculitis provides additional evidence of childhood dermatomyositis. We did not provide this guidance in proposed 14.00D4b because MRI findings are not considered diagnostic of dermatomyositis in adults. In proposed 114.00D4c(i), we explain how to evaluate polymyositis and dermatomyositis under the listings in newborn and younger infants.

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

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

Current 114.00E6 provides that the fact that a child is dependent on steroids, or any other drug, for the control of inflammatory arthritis is, in and of itself, insufficient to find disability. It explains that advances in the treatment of inflammatory connective tissue disease and in the administration of steroids for its treatment have corrected some of the previously disabling consequences of continuous steroid use. Although this statement is still true, we are not including this provision of current 114.00E6 in these proposed rules because we believe we no longer need it in the introductory text of the listings.

We added current 114.00E6 in 2002 (66 FR 58010, 58022 and 58045 (2001)). It was important when we added it because the listings prior to the revisions we made in 2002 included a listing (prior listing 101.02B) that said that all children with rheumatoid arthritis who were dependent on steroids were disabled. We removed that listing in 2002, explaining that, although the prior listing was appropriate when we first published it, advances in treatment and other reasons had made it obsolete (66 FR 58022). Thus, the paragraph in the introductory text served as a reminder that we no longer had that listing and that it was no longer appropriate to presume disability based on steroid use alone. Now that several years have passed since we removed the prior listing, we do not believe that we need this reminder any longer. However, in proposed 114.00G3, we continue to state that we will consider the adverse side effects of treatment, including the effects of corticosteroids, to ensure that our adjudicators remember to consider the side effects of steroids and any other treatment an individual might have.

Proposed 114.00F—How do we evaluate human immunodeficiency virus (HIV) infection?

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

Proposed 114.00F1a, *Documentation of HIV infection by definitive diagnosis*, corresponds to 114.00D3a in the current rules and 14.00F1a in the proposed rules. In this section, we propose to lower the age for using HIV antibody tests from 24 months of age or older that is in current 114.00D3a(i) to 18 months or older in proposed 114.00F1a(i) because current clinical practice now accepts these tests beginning at 18 months of age.

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

Proposed 114.00F1b, *Documentation of HIV infection in children from birth to the attainment of 18 months* is new and corresponds to the second paragraph in current 114.00D3b, *Other acceptable documentation of HIV infection in children*. However, we are proposing to move this information under proposed 114.00F1b to provide documentation of HIV infection by definitive diagnosis in children from birth to the attainment of 18 months of age who have tested positive for HIV antibodies. We also propose to lower the age for children testing positive for HIV antibodies from 24 months of age that is in the second paragraph of current 114.00D3b to 18 months in proposed 114.00F1b. We are proposing to make these changes because current clinical practice now accepts these positive test results as diagnostic of HIV infection in children beginning at 18 months of age who have tested positive for HIV antibodies.

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

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

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

Proposed 114.00F1c, *Other acceptable documentation of HIV infection*, corresponds to current 114.00D3b and proposed 14.00F1b. We propose to remove the first paragraph in current 114.00D3b because all infants who have HIV antibodies are now tested to determine definitively whether they have HIV infection. This makes the first paragraph in current 114.00D3b unnecessary.

In proposed 114.00F2, *CD4 tests*, we add more detailed guidance to the second paragraph of current 114.00D4a by specifying that the extent of immune depression correlates with the level of CD4 counts in children at 6 years of age or older, the age at which CD4 levels become comparable to adult CD4 levels.

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

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

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

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

Proposed 114.00G—How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?

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

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

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

As in the adult rules, we propose to add listings based on functional criteria to each of the listings in the immune system in addition to listing 114.08. Current listing 114.08O is the childhood listing that corresponds to current adult listing 14.08N, and we are proposing to use essentially the same criteria in the other listings as we do in this listing. (In the proposed rules, current listing 114.08O would become listing 114.08L.) Proposed 114.00I—*How do we use the functional criteria in these listings*?—corresponds to current 114.00D8 and provides guidance for applying the listings based on functional criteria. We propose to revise the current language to reflect the fact that there would now be functional listings for each of the listed impairments in this body system and for consistency with adult rules where appropriate.

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

In proposed 114.00J2, we repeat the guidance in proposed 14.00J but with appropriate references to listings in part B, and we include growth impairment under 100.00ff as an example.

How are we proposing to change the criteria in the listings for evaluating immune system impairments in children?

Proposed 114.01 Category of Impairments, Immune System Disorders

As in the adult listings in part A, we propose to remove all reference listings from part B. We also propose to add listings like 114.08O to each of the other listings in this body system. The new listings would be proposed listings 114.02B, 114.03B, 114.04D, 114.05E, 114.06B, 114.07C, 114.09D, and 114.10B. In addition, current listing 114.08O would be redesignated as listing 114.08L because of the deletion of reference listings. The functional criteria in the new proposed listings for children would be the same as in current listing 114.08O (proposed listing 114.08L). They are different from the functional criteria in part A because the functional criteria for adults are not applicable to the evaluation of functioning in children. The childhood functional criteria are the same as in current listing 114.08O (proposed listing 114.08L); they use the functional criteria in listings 112.02 and 112.12.

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

Proposed Listing 114.04—Systemic Sclerosis (Scleroderma)

Proposed listings 114.04B1 and 114.04B2 correspond to current listing 114.04B1. We propose to change the requirement in current listing 114.04B1 for fixed deformity of ''both feet'' to ''one or both feet'' and to add ''inability to ambulate effectively'' to the listing criteria. This will allow some children with a serious deformity in only one foot to qualify based on the functional limitation we use to define listing-level severity throughout these listings. We also propose to add the criterion of ''toe contractures'' to proposed 114.04B1 even though toe contractures of listing-level severity would be rare in children to make it consistent with the criteria in proposed 14.04B1. We are retaining the requirement for involvement of both hands in proposed listing 114.04B2, because inability to use fine and gross movements effectively can only occur when both upper extremities are affected. We propose to add the criterion of ''finger contractures'' to proposed 114.004B2 for the same reason we are proposing to add ''toe contractures'' to proposed 114.04B1.

Proposed listings 114.04B3 and 114.04B4 correspond to current listing 114.04B2, the listing for ''[m]arked destruction or marked atrophy of an extremity.'' We propose to revise the rules to

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

We propose to remove the word ''marked'' because we use it in various other listings and other regulations to describe a particular measure of functional limitations, and it does not describe what we intend in this listing. We propose to replace the criterion for ''marked destruction'' with a criterion for ''irreversible damage'' because it is a more accurate medical description of this complication of systemic sclerosis. We propose to require both atrophy and irreversible damage because we would not expect either of these findings alone to establish an impairment that results in marked and severe functional limitations in every case. Finally, we propose to require ''inability to ambulate effectively'' or ''inability to

perform fine or gross movements effectively'' to establish an impairment that is of listing-level severity, consistent with other existing and proposed listings.

Proposed listing 114.04C, Raynaud's phenomenon, is a new childhood listing and has the same criteria as in proposed listing 14.04C for adults. Even though listing-level severity would be rare in children with Raynaud's phenomenon, it can occur.

Proposed Listing 114.05—Polymyositis and Dermatomyositis

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

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

Proposed Listing 114.07—Immune deficiency disorders, excluding HIV infection

We propose to remove current listing 114.07B because of advances in medical knowledge that now allow us to identify different subgroups of thymic dysplastic syndromes. The subgroups of these disorders vary in severity, and therefore, they should be evaluated under proposed listing 114.07A, B, or C, as appropriate to the particular immune deficiency disorder and its effects.

Proposed Listing 114.08—Human Immunodeficiency Virus (HIV) Infection

In proposed listing 114.08A5, we incorporate current listing 114.08A6 except to remove ''Other'' as a descriptor to make it consistent with the proposed adult listing. We propose to replace ''acquisition'' as used in current listing 114.08H1 with ''onset'' in proposed listing 114.08G1 because a sudden ''onset'' of a new learning disability is medically a more accurate description of how this neurologic abnormality would manifest in a child with HIV infection. We are also redesignating a number of listings to reflect the proposed removal of reference listings.

Proposed Listing 114.10— Sjögren's Syndrome

We propose to add a new listing 114.10 to evaluate Sjögren's syndrome in children for the same reasons we propose to add a Sjögren's syndrome listing for adults in part A.

Other Changes

We propose to make minor conforming changes in current 1.00B and 101.00B, and 1.00L and 101.00L to reflect changes in the proposed immune body system listings.

We also propose to make minor conforming changes in current 8.00D3 and 108.00D3 of the skin disorders listings. We would revise these sections to indicate that we evaluate Sjögren's syndrome under the new listing for that disorder, listings 14.10 and 114.10.

Clarity of These Proposed Rules

Executive Order 12866, as amended by Executive Order 13258, requires each agency to write all rules in plain language. In addition to your substantive comments on these proposed rules, we invite your comments on how to make these proposed rules easier to understand.

For example:

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

Regulatory Procedures

Executive Order 12866

We have consulted with the Office of Management and Budget (OMB) and determined that these proposed rules meet the requirements for a significant regulatory action under Executive Order 12866, as amended by Executive Order 13258. Thus, they were subject to OMB review.

Regulatory Flexibility Act

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

Paperwork Reduction Act

These proposed rules contain reporting requirements at 14.00B, 14.00D, 14.00E, 14.00F, 114.00B, 114.00D, 114.00E, 114.00F, 114.08 and 114.09. The public reporting burden is accounted for in the Information Collection Requests for the various forms that the public uses to submit the information to SSA. Consequently, a 1-hour placeholder burden is being assigned to the specific reporting requirement(s) contained in these rules. We are seeking clearance of the burdens referenced in these rules because they were not considered during the clearance of the forms. An Information Collection Request has been submitted to OMB. We are soliciting comments on the burden estimate; the need for the information; its practical utility; ways to enhance its quality, utility and clarity; and on ways to minimize the burden on respondents, including the use of automated collection techniques or other forms of information technology. Comments should be submitted and/or faxed to the Office of Management and Budget and to the Social Security Administration at the following addresses/numbers:

Office of Management and Budget, Attn: Desk Officer for SSA, New Executive Office Building, Room 10230, 725 17th St., NW., Washington, DC 20530. Fax Number: 202–395–6974.

Social Security Administration, Attn: SSA Reports Clearance Officer, Rm. 1338 Annex Building, 6401 Security Boulevard, Baltimore, MD 21235–6401. Fax Number: 410–965–6400.

Comments can be received for up to 60 days after publication of this notice, and your comments will be most useful if received by SSA within 30 days of publication. To receive a copy of the OMB clearance package, you may call the SSA Reports Clearance Officer on 410–965–0454.

References

We consulted the following sources when developing these proposed rules:

Bartlett, J.G. and Gallant, J.E., Medical Management of HIV Infection (Johns Hopkins University 2003).

Burchett, S.A. and Pizzo, P.A., HIV Infection in Infants, Children, and Adolescents, Pediatrics in Review, 24(6), 186–194 (2001).
Davidson, A. and Diamond, B., Autoimmune Diseases, The New England Journal of Medicine, 345(5), 1–21 (2001).
Furst, D.E., Stem Cell Transplantation for Autoimmune Disease: Progress and Problems, Current Opinion in Rheumatology, 14(3), 220–224 (2002).
Harris, E.D., *et al.*, Eds., Kelley's Textbook of Rheumatology, (Elsevier, 7th ed. 2005).
Klippel, J.H., *et al.*, Eds., Primer on the Rheumatic Diseases, (Arthritis Foundation, 12th ed. 2001).
Sicherer, S.H., *et al.*, Primary Immunodeficiency Diseases in Adults, Journal of American Medical Association, 279:58 (1998).
Tyndall, A. and Koike, T., High-dose immunoablative therapy with hematopoietic stem cell support in the treatment of severe autoimmune disease: current status and future direction, Internal Medicine, 41(8), 608–12 (2002).

These references are included in the rulemaking record for these proposed rules and are available for inspection by interested persons by making arrangements with the contact person shown in this preamble.

(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 20 CFR Part 404

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

Dated: July 28, 2006.

Jo Anne B. Barnhart,

Commissioner of Social Security.

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

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

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

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

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

2. Appendix 1 to subpart P of part 404 is amended as follows:

a. Revise the expiration date in item 15 of the introductory text before part A of appendix 1.

b. Revise the second sentence of section 1.00B1 of part A of appendix 1.

c. Revise the fourth sentence of section 1.00L of part A of appendix 1.

d. Revise section 8.00D3 of part A of appendix 1.

e. Revise section 14.00 of part A of appendix 1.

f. Revise the second sentence of section 101.00B1 of part B of appendix 1.

g. Revise the fourth sentence of section 101.00L of part B of appendix 1.

h. Revise section 108.00D3 of part B of appendix 1.

i. Revise section 114.00 of part B of appendix 1.

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

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15. Immune System Disorders (14.00 and 114.00): (date 8 years from the effective date of the final rules.)

* * * * *

Part A

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1.00 Musculoskeletal System

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B. *Loss of function.*

1. *General.* * * * For inflammatory arthritides that may result in loss of function because of inflammatory peripheral joint or axial arthritis or sequelae, or because of extra-articular features, see 14.00D6. * * *

* * * * *

L. *Abnormal curvatures of the spine.* * * * When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 14.09C. * * *

* * * * *

8.00 Skin Disorders

* * * * *

D. *How do we assess impairments that may affect the skin and other body systems?*

* * * * *

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

* * * * *

14.00 Immune System Disorders

A. *What disorders do we evaluate under the immune system listings?*

1. *We evaluate immune system disorders that cause dysfunction in one or more components of your immune system.*

a. These listings are examples of immune system disorders that are severe enough to prevent you from doing any gainful activity. The dysfunction may be due to problems in antibody production, impaired cell-mediated immunity, a combined type of antibody/cellular deficiency, impaired phagocytosis, or complement deficiency.

b. Immune system disorders may result in recurrent and unusual infections, or inflammation and dysfunction of the body's own tissues. Immune system disorders can cause a deficit in a single organ or body system that results in extreme (that is, very serious) loss of function. They can also cause lesser degrees of limitations in two or more organs or body systems, and when associated with symptoms or signs such as fatigue, fever, malaise, diffuse musculoskeletal pain, or involuntary weight loss, can also result in extreme limitation.

c. In this preface, we organize the discussions of immune system disorders in three categories: Autoimmune disorders; Immune deficiency disorders, excluding human immunodeficiency virus (HIV) infection; and HIV infection.

2. *Autoimmune disorders (14.00D).* Autoimmune disorders are caused by dysfunctional immune responses directed against the body's own tissues, resulting in chronic, multisystem impairments that differ in clinical manifestations, course, and outcome. They are sometimes referred to as rheumatic diseases, connective tissue disorders, or collagen vascular disorders. Some of the features of autoimmune disorders in adults differ from the features of the same disorders in children.

3. *Immune deficiency disorders, excluding HIV infection (14.00E).* Immune deficiency disorders are characterized by recurrent or unusual infections that respond poorly to treatment, and are often associated with complications affecting other parts of the body. Immune deficiency disorders are classified as either *primary* (congenital) or *acquired.* Individuals with immune deficiency disorders also have an increased risk of malignancies and of having autoimmune disorders.

4. *Human immunodeficiency virus (HIV) infection (14.00F).* HIV infection is caused by a specific retrovirus and may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions as described in 14.08.

B. *What information do we need to show that you have an immune system disorder?* Generally, we need your medical history, report(s) of physical examination, report(s) of laboratory findings, and in some instances, appropriate medically acceptable imaging or tissue biopsy reports to show that you have an immune system disorder. Therefore, we will make every reasonable effort to obtain your medical history, medical findings, and results of laboratory tests. We explain the information we need in more detail in the sections below.

C. *Definitions.*

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is one that is generally accepted and consistent with the

prevailing state of medical knowledge and clinical practice to support the evaluation and diagnosis of the impairment.

2. *Constitutional symptoms or signs* means fatigue, fever, malaise, or involuntary weight loss. *Severe fatigue* means a frequent sense of exhaustion that results in significantly reduced physical activity or mental function. *Malaise* means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.

3. *Disseminated* means that a condition is spread over a considerable area. The type and extent of the spread will depend on your specific disease.

4. *Dysfunction* means that one or more of the body regulatory mechanisms are impaired, causing either an excess or deficiency of immunocompetent cells or their products.

5. *Extra-articular* means ''other than the joints''; for example, the effect is in an organ(s) such as the heart, lungs, kidneys, or skin.

6. *Inability to ambulate effectively* has the same meaning as in 1.00B2b.

7. *Inability to perform fine and gross movements effectively* has the same meaning as in 1.00B2c.

8. *Major peripheral joints* has the same meaning as in 1.00F.

9. *Persistent* means that a sign(s) or symptom(s) has continued over time. The precise meaning will depend on the specific immune system disorder, the usual course of the disorder, and the other circumstances of your clinical course.

10. *Recurrent* means that a condition that previously responded adequately to an appropriate course of treatment returns after a period of remission or regression. The precise meaning, such as the extent of response or remission and the time periods involved, will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

11. *Resistant to treatment* means that a condition did not respond adequately to an appropriate course of treatment. Whether a response is adequate or a course of treatment is appropriate will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

12. *Severe* describes medical severity as used by the medical community. The term does not have the same meaning as it does when we use it in connection with a finding at the second step of the sequential evaluation processes in §§ 404.1520, 416.920, and 416.924.

D. *What are the listed autoimmune disorders in these listings?*

1. *Systemic lupus erythematosus (14.02).*

a. *General.* Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that can affect any organ or body system. It is frequently, but not always, accompanied by constitutional symptoms or signs (fatigue, fever, malaise, involuntary weight loss). Major organ or body system involvement can include: Respiratory (pleuritis, pneumonitis), cardiovascular (endocarditis, myocarditis, pericarditis, vasculitis), renal (glomerulonephritis), hematologic (anemia, leukopenia, thrombocytopenia), skin (photosensitivity), neurologic (seizures), mental (anxiety), fluctuating cognition (''lupus fog''), mood disorders, organic brain syndrome, psychosis), or immune system (inflammatory arthritis) disorders. Immunologically, there is an array of circulating serum auto-antibodies and pro- and anti-coagulant proteins that may occur in a highly variable pattern.

b. *Documentation of SLE.* Generally, but not always, the medical evidence will show that your SLE satisfies the criteria in the current ''Criteria for the Classification of Systemic Lupus Erythematosus'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

2. *Systemic vasculitis (14.03).*

a. *General.* (i) Vasculitis is an inflammation of blood vessels. It may occur acutely in association with adverse drug reactions, certain chronic infections, and occasionally, malignancies. More often, it is chronic and the cause is unknown. Symptoms vary depending on which blood vessels are involved. Systemic vasculitis may also be associated with other autoimmune disorders; for example, SLE or dermatomyositis.

(ii) There are several clinical patterns, including but not limited to polyarteritis nodosa, Takayasu's arteritis (aortic arch arteritis), giant cell arteritis (temporal arteritis), and Wegener's granulomatosis.

b. *Documentation of systemic vasculitis.* Angiography or tissue biopsy confirms a diagnosis of systemic vasculitis when the disease is suspected clinically. Usually the results will be in your medical records.

3. *Systemic sclerosis (scleroderma) (14.04).*

a. *General.* Systemic sclerosis (scleroderma) constitutes a spectrum of disease in which thickening of the skin is the clinical hallmark. Raynaud's phenomenon, often medically severe and progressive, is present frequently and may be the peripheral manifestation of a vasospastic abnormality in the heart, lungs, and kidneys. The CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) is a variant that may slowly progress over years to the generalized process, systemic sclerosis.

b. *Diffuse cutaneous systemic sclerosis.* In diffuse cutaneous systemic sclerosis (also known as diffuse scleroderma), major organ or systemic involvement can include the gastrointestinal tract, lungs, heart, kidneys, and muscle in addition to skin or blood vessels. Although arthritis can occur, joint dysfunction results primarily from soft tissue/cutaneous thickening, fibrosis, and contractures.

c. *Localized scleroderma (linear scleroderma and morphea).*

(i) Localized scleroderma (linear scleroderma and morphea) is more common in children than in adults; however, this type of scleroderma can persist into adulthood. The extent of involvement of linear scleroderma and a description of the lesions are important in assessing the severity of the impairment. For example, linear scleroderma involving the arm but not crossing any joints is not as functionally limiting as sclerodactyly (scleroderma localized to the fingers). Linear scleroderma of a lower extremity involving skin thickening and atrophy of underlying muscle or bone can result in contracture(s) and leg length discrepancies. In such cases, evaluation under the musculoskeletal (1.00ff) listing may be appropriate.

(ii) When there is isolated morphea of the face causing facial disfigurement from unilateral hypoplasia of the mandible, maxilla, zygoma, or orbit, adjudication may be more appropriate under the criteria in the special senses listings (2.00ff) or mental disorders listings (12.00ff).

(iii) Chronic variants of these syndromes include disseminated morphea, Shulman's disease (diffuse fasciitis with eosinophilia), and eosinophilia-myalgia syndrome (often associated with toxins such as toxic oil or contaminated tryptophan), all of which can impose medically severe musculoskeletal dysfunction and may also lead to restrictive pulmonary disease. We evaluate these variants of the disease under the criteria in the musculoskeletal listings (1.00ff) or respiratory system listings (3.00ff).

d. *Documentation of systemic sclerosis (scleroderma).* Documentation involves differentiating the clinical features of systemic sclerosis (scleroderma) from other autoimmune disorders; however, there may be an overlap.

4. *Polymyositis and dermatomyositis (14.05).*

a. *General.* Polymyositis and dermatomyositis are related disorders that are characterized by an inflammatory process in striated muscle, occurring alone or in association with other autoimmune disorders or malignancy. Symmetric weakness, and less frequently pain and tenderness of the proximal limb-girdle (shoulder or pelvic) musculature, are the most common manifestations. There may also be involvement of the cervical, cricopharyngeal, esophageal, intercostal, and diaphragmatic muscles.

b. *Documentation of polymyositis and dermatomyositis.* Generally, but not always, polymyositis is associated with elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, aldolase), and characteristic abnormalities on electromyography and muscle biopsy. In dermatomyositis there are characteristic skin findings in addition to the findings of polymyositis.

c. *Additional information about how we evaluate polymyositis and dermatomyositis under the listings.*

(i) Weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to ambulate effectively. Weakness of your shoulder girdle muscles may result in your inability to perform lifting, carrying, and reaching overhead, and also may seriously affect your ability to perform activities requiring fine movements. We evaluate these limitations under 14.05A.

(ii) We use the malignant neoplastic diseases listings (13.00ff) to evaluate malignancies associated with polymyositis or dermatomyositis. We evaluate the involvement of other organs/body systems under the criteria for the listings in the affected body system.

5. *Undifferentiated and mixed connective tissue disease (14.06).*

a. *General.* This listing includes syndromes with clinical and immunologic features of several autoimmune disorders, but which do not satisfy the criteria for any of the specific disorders described. For example, you may have clinical features of systemic lupus erythematosus and systemic vasculitis, and the serologic (blood test) findings of rheumatoid arthritis.

b. *Documentation of undifferentiated and mixed connective tissue disease.* Undifferentiated connective tissue disease is diagnosed when clinical features and serologic (blood test) findings, such as rheumatoid factor or antinuclear antibody (consistent with an autoimmune disorder) are present but do not satisfy the criteria for a specific disease. Mixed connective tissue disease (MCTD) is diagnosed when clinical features and serologic findings of two or more autoimmune diseases overlap.

6. *Inflammatory arthritis (14.09).*

a. *General.* The inflammatory arthritides include a vast array of disorders that differ in cause, course, and outcome. Clinically, inflammation of major peripheral joints may be the dominant manifestation causing difficulties with ambulation or fine and gross movements; there may be joint pain, swelling, and tenderness. The arthritis may affect other joints, or cause less functional limitations in ambulation or performance of fine and gross movements. However, in combination with extra-articular features, including constitutional symptoms or signs (fatigue, fever, malaise, involuntary weight loss), inflammatory arthritis may result in an extreme limitation.

b. *Inflammatory arthritides involving the axial spine (spondyloarthropathies).* In adults, inflammatory arthritides involving the axial spine may be associated with heterogeneous disorders such as:

(i) Reiter's syndrome;

(ii) Ankylosing spondylitis;

(iii) Psoriatic arthritis;

(iv) Whipple's disease;

(v) Behçet's disease; and

(vi) Inflammatory bowel disease.

c. *Inflammatory arthritides involving the peripheral joints.* The inflammatory arthropathies involving peripheral joints may be associated with disorders such as:

(i) Rheumatoid arthritis;

(ii) Sjögren's syndrome;

(iii) Psoriatic arthritis;

(iv) Crystal deposition disorders (gout and pseudogout);

(v) Lyme disease; and

(vi) Inflammatory bowel disease.

d. *Documentation of inflammatory arthritides.* Generally, but not always, the diagnosis of inflammatory arthritis is made by the clinical features and serologic findings described in the most recent edition of the *Primer on Rheumatic Diseases* published by the Arthritis Foundation.

e. *How we evaluate the inflammatory arthritides under the listings.*

(i) Listing-level severity in 14.09A and 14.09C1 is shown by an impairment that results in an ''extreme'' (very serious) limitation. In 14.09A, the criterion is satisfied with persistent inflammation or deformity in two or more major peripheral joints resulting in the inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 14.00C6 and 14.00C7. In 14.09C1, if you have the required ankylosis (fixation) of your cervical or dorsolumbar spine, we will find that you have an extreme limitation in your ability to see in front of you, above you, and to the side. Therefore, inability to ambulate effectively is implicit in 14.09C1, even though you might not require bilateral upper limb assistance.

(ii) Listing-level severity is shown in 14.09B, 14.09C2, and 14.09D when the arthritis does not result in the extreme limitation in 14.09A or 14.09C1, involves one or more major peripheral joints, or involves other joints, but is complicated by extra-articular features that cumulatively result in an ''extreme'' (very serious) limitation or ''marked'' (serious) limitations in at least two areas of functioning. Extra-articular impairments may also meet listings in other body systems.

(iii) Extra-articular features of inflammatory arthritis may involve any body system. Commonly occurring extra-articular impairments include: Musculoskeletal (heel enthesopathy), ophthalmologic (iridocyclitis, keratoconjunctivitis sicca, uveitis), pulmonary (pleuritis, pulmonary fibrosis or nodules, restrictive lung disease), cardiovascular (aortic valve insufficiency, arrhythmias, coronary arteritis, myocarditis, pericarditis, Raynaud's phenomenon, systemic vasculitis), renal (amyloidosis of the kidney), hematologic (chronic anemia, thrombocytopenia), neurologic (peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss), and immune system (Felty's syndrome (hypersplenism with compromised immune competence)) disorders.

(iv) If permanent deformity of a major peripheral joint is the dominant feature of your impairment, we evaluate your impairment under 1.02.

(v) If there has been surgical reconstruction of a major weight-bearing joint, we evaluate your impairment under 1.03.

(vi) If both inflammation and chronic deformities are present, we evaluate your impairment under the criteria of any appropriate listing.

7. *Sjögren's syndrome (14.10).*

a. *General.* (i) Sjögren's syndrome is an immune-mediated disorder of the exocrine glands. Involvement of the lacrimal and salivary glands is the hallmark feature, resulting in symptoms of dry eyes and dry mouth, and possible complications such as corneal damage, blepharitis (eyelid inflammation), dysphagia (difficulty in swallowing), dental caries, and the inability to speak for extended periods of time. Involvement of the exocrine glands of the upper airways may result in persistent dry cough.

(ii) Many other organ systems may be involved, including musculoskeletal (arthritis, myositis), respiratory (interstitial fibrosis), gastrointestinal (dysmotility, dysphagia, involuntary weight loss), genitourinary (interstitial cystitis, renal tubular acidosis), skin (purpura, vasculitis), neurologic (central nervous system disorders, cranial and peripheral neuropathies), mental (cognitive dysfunction, poor memory), and neoplastic (lymphoma). Fatigue and malaise are frequently reported. Sjögren's syndrome may be associated with other autoimmune disorders (for example, rheumatoid arthritis or SLE); usually the clinical features of the associated disorder predominate.

b. *Documentation of Sjögren's syndrome.* If you have Sjögren's syndrome, the medical evidence will generally, but not always, show that your disease satisfies the criteria in the current ''Criteria for the Classification of Sjögren's Syndrome'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

E. *How do we evaluate immune deficiency disorders, excluding HIV infection (14.07)?*

1. *General.*

a. Immune deficiency disorders can be classified as:

(i) *Primary* (congenital); for example, X-linked agammaglobulinemia, thymic hypoplasia (DiGeorge syndrome), severe combined immunodeficiency (SCID), chronic granulomatous disease (CGD), C1 esterase inhibitor deficiency.

(ii) *Acquired;* for example, medication-related.

b. Primary immune deficiency disorders are seen mainly in children. However, recent advances in the treatment of these disorders have allowed many affected children to survive well into adulthood. Occasionally, these disorders are first diagnosed in adolescence or adulthood.

2. *Documentation of immune deficiency disorders.* The medical evidence must include documentation of the specific type of immune deficiency. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

3. *Immune deficiency disorders treated by stem cell transplantation.*

a. *Evaluation in the first 12 months.* If you undergo stem cell transplantation for your immune deficiency disorder, we will consider you disabled until at least 12 months from the date of the transplant.

b. *Evaluation after the 12-month period has elapsed.* After the 12-month period has elapsed, we will consider any residuals of your immune deficiency disorder as well as any residual impairment(s) resulting from the treatment, such as complications arising from:

(i) Graft-versus-host (GVH) disease.

(ii) Immunosuppressant therapy, such as frequent infections.

(iii) Significant deterioration of other organ systems.

4. *Medication-induced immune suppression.* Medication effects can result in varying degrees of immune suppression, but

most resolve when the medication is ceased. However, if you are prescribed medication for long-term immune suppression, such as after an organ transplant, we will evaluate:

a. The frequency and severity of infections.

b. Residuals from the organ transplant itself, after the 12-month period has elapsed.

c. Significant deterioration of other organ systems.

F. *How do we evaluate human immunodeficiency virus (HIV) infection?* Any individual with HIV infection, including one with a diagnosis of acquired immune deficiency syndrome (AIDS), may be found disabled under 14.08 if his or her impairment meets the criteria in that listing or is medically equivalent to the criteria in that listing.

1. *Documentation of HIV infection.* The medical evidence must include documentation of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for HIV infection, we will make every reasonable effort to obtain reports of the results of that testing.

a. *Documentation of HIV infection by definitive diagnosis.* A definitive diagnosis of HIV infection is documented by one or more of the following laboratory tests:

(i) HIV antibody tests. HIV antibodies are usually first detected by an ELISA screening test performed on serum. Because the ELISA can yield false positive results, confirmation is required using a more definitive test, such as a Western blot or an immunofluorescence assay.

(ii) Positive ''viral load'' (VL) tests. These tests are normally used to quantitate the amount of the virus present but also document HIV infection. Such tests include the quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT–PCR).

(iii) HIV DNA detection by polymerase chain reaction (PCR).

(iv) A specimen that contains HIV antigen (for example, serum specimen, lymphocyte culture, or cerebrospinal fluid).

(v) A positive viral culture for HIV from peripheral blood mononuclear cells (PBMC).

(vi) Other tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge.

b. *Other acceptable documentation of HIV infection.* We may also document HIV infection without the definitive laboratory evidence described in 14.00F1a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice, and is consistent with the other evidence in your case record. If no definitive laboratory evidence is available, we may document HIV infection by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence. For example, we will accept a diagnosis of HIV infection without definitive laboratory evidence if you have an opportunistic disease that is predictive of a defect in cell-mediated immunity (for example, toxoplasmosis of the brain, Pneumocystis carinii pneumonia (PCP)), and there is no other known cause of diminished resistance to that disease (for example, long-term steroid treatment, lymphoma). In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

2. *CD4 tests.* Individuals who have HIV infection or other disorders of the immune system may have tests showing a reduction of either the absolute count or the percentage of their T-helper lymphocytes (CD4 cells). The extent of immune suppression correlates with the level or rate of decline of the CD4 count. Generally, when the CD4 count is 200/mm^e or less (14 percent or less) the susceptibility to opportunistic infection is greatly increased. Although a reduced CD4 count *alone* does not establish a definitive diagnosis of HIV infection, a CD4 count below 200 does offer supportive evidence when there are clinical findings, but not a definitive diagnosis of an opportunistic infection(s). However, a reduced CD4 count alone does not document the severity or functional consequences of HIV infection.

3. *Documentation of the manifestations of HIV infection.* The medical evidence must also include documentation of the manifestations of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for a manifestation of HIV infection, we will make every reasonable effort to obtain reports of the results of that testing.

a. *Documentation of the manifestations of HIV infection by definitive diagnosis.* The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serologic test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). We will make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histologic or other test has been performed, the evidence should include a copy of the appropriate report. If we cannot obtain the report, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including appropriate medically acceptable imaging studies) or microscopic examination of the appropriate tissues or body fluids.

b. *Other acceptable documentation of the manifestations of HIV infection.* We may also document manifestations of HIV infection without the definitive laboratory evidence described in 14.00F3a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice, and is consistent with the other evidence in your case record. If no definitive evidence is available, we may document the manifestations of HIV infection with other appropriate evidence. For example, many conditions are now commonly diagnosed based on some or all of the following: Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

(i) Although a definitive diagnosis of PCP requires identifying the organism in bronchial washings, induced sputum, or lung biopsy, these tests are frequently bypassed if PCP can be diagnosed presumptively. (Note: *Pneumocystis carinii* is now known as *Pneumocystis jiroveci*; however, ''PCP'' remains in common usage for the pneumonia caused by this organism.) Supportive evidence includes: Fever, dyspnea, hypoxia, and CD4 count below 200. Also supportive are bilateral lung interstitial infiltrates on x-ray, or a typical pattern on CT scan, or a gallium scan positive for pulmonary uptake. Response to anti-PCP therapy usually requires 5–7 days.

(ii) Documentation of cytomegalovirus (CMV) disease (14.08D) may present special problems because definitive diagnosis (except for chorioretinitis, which may be diagnosed by an ophthalmologist on funduscopic exam) requires identification of viral inclusion bodies or a positive culture from the affected organ and the absence of any other infectious agent likely to be causing the disease. A positive serology test identifies a history of infection with CMV, but it does not confirm an active disease process. Therefore, a presumptive diagnosis of CMV disease requires corroborating evidence that CMV is causing the disease. Supportive evidence includes: Fever, positive CMV serology test, urinary culture positive for CMV, and CD4 count below 200. A clear response to anti-CMV therapy also supports a diagnosis.

(iii) A definitive diagnosis of toxoplasmosis of the brain is made by brain biopsy, but this procedure carries significant risk and is not commonly performed. This condition is usually diagnosed presumptively based on symptoms or signs of fever, headache, focal neurologic deficits, seizures, typical lesions on brain imaging, and a positive serology test.

4. *Manifestations specific to women.*

a. *General.* Most women with severe immunosuppression secondary to HIV infection exhibit the typical opportunistic infections and other conditions, such as PCP, candida esophagitis, wasting syndrome, cryptococcosis, and toxoplasmosis. However, HIV infection may have different manifestations in women than in men. Adjudicators must carefully scrutinize the medical evidence and be alert to the variety of medical conditions specific to, or common in, women with HIV infection that may affect their ability to function in the workplace.

b. *Additional considerations for evaluating HIV infection in women.* Many of these manifestations (for example, vulvovaginal candidiasis, pelvic inflammatory disease) occur in women with or without HIV infection, but can be more severe or resistant to treatment, or occur more frequently in a woman whose immune system is suppressed. Therefore, when evaluating the claim of a woman with HIV infection, it is important to consider gynecologic and other problems specific to women, including any associated

symptoms (for example, pelvic pain), in assessing the severity of the impairment and resulting functional limitations. We may evaluate manifestations of HIV infection in women under the specific criteria (for example, cervical cancer under 14.08E), under an applicable general category (for example, pelvic inflammatory disease under 14.08A4) or, in appropriate cases, under 14.08K.

5. *Involuntary weight loss.* As used in 14.08H, ''significant involuntary weight loss'' does not correspond to a specific minimum amount or percentage of weight loss. For purposes of this listing, an involuntary weight loss of at least 10 percent of baseline is always considered significant. Loss of less than 10 percent may or may not be significant, depending on the individual's baseline weight and body habitus. (For example, a 7-pound weight loss in a 100-pound woman who is 63 inches tall might be considered significant; but a 14-pound weight loss in a 200-pound woman who is the same height might not be significant.)

G. *How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?*

1. *General.* If your impairment does not otherwise meet the requirements of a listing we will consider your medical treatment both in terms of its effectiveness in improving the signs, symptoms, and laboratory abnormalities of your specific immune system disorder or its manifestations, and in terms of any side effects that limit your functioning. We will make every reasonable effort to obtain a specific description of the treatment you receive (including surgery) for your immune system disorder. We consider:

a. The effects of medications you take.

b. Adverse side effects (acute and chronic).

c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).

d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).

e. Variability of your response to treatment (see 14.00G2).

f. The interactive and cumulative effects of your treatments. For example, many individuals with immune system disorders receive treatment both for their immune system disorders and for the manifestations of the disorders or co-occurring impairments, such as treatment for HIV infection and hepatitis C. The interactive and cumulative effects of these treatments may be greater than the effects of each treatment considered separately.

g. The duration of your treatment.

h. Any other aspects of treatment that may interfere with your ability to function.

2. *Variability of your response to treatment.* Your response to treatment and the adverse or beneficial consequences of your treatment may vary widely. The effects of your treatment may be temporary or long term. For example, some individuals may show an initial positive response to a drug or combination of drugs followed by a decrease in effectiveness. When we evaluate your response to treatment and how your treatment may affect you, we consider such factors as disease activity before treatment, requirements for changes in therapeutic regimes, the time required for therapeutic effectiveness of a particular drug or drugs, the limited number of drug combinations that may be available for your impairment(s), and the time-limited efficacy of some drugs. For example, an individual with HIV infection or another immune deficiency disorder who develops pneumonia or tuberculosis may not respond to the same antibiotic regimen used in treating individuals without these disorders or may not respond to an antibiotic that he or she responded to before. Therefore, we must consider the effects of your treatment on an individual basis, including the effects of your treatment on your ability to function.

3. *How we evaluate the effects of treatment for autoimmune disorders on your ability to function.* Some medications may have acute or long-term side effects. When we consider the effects of corticosteroids or other treatments for autoimmune disorders on your ability to function, we consider the factors in 14.00G1 and 14.00G2. Long-term corticosteroid treatment can cause ischemic necrosis of bone, posterior subcapsular cataract, weight gain, glucose intolerance, increased susceptibility to infection, and osteoporosis that may result in a loss of function. In addition, medications used in the treatment of autoimmune disorders may also have effects on mental function including cognition (for example, memory), concentration, and mood.

4. *How we evaluate the effects of treatment for immune deficiency disorders, excluding HIV infection, on your ability to function.* When we consider the effects of your treatment for your immune deficiency disorder on your ability to function, we consider the factors in 14.00G1 and 14.00G2. A frequent need for treatment such as intravenous immunoglobulin and gamma interferon therapy can be intrusive and interfere with your ability to work on a sustained basis. We will also consider whether you have chronic side effects from these or other medications, including fatigue, fever, headaches, high blood pressure, joint swelling, muscle aches, nausea, shortness of breath, or limitations in mental function including cognition (for example, memory), concentration, and mood.

5. *How we evaluate the effects of treatment for HIV infection on your ability to function.*

a. *General.* When we consider the effects of antiretroviral drugs (including the effects of highly active antiretroviral therapy (HAART)) and the effects of treatments for the manifestations of HIV infection on your ability to function, we consider the factors in 14.00G1 and 14.00G2. Side effects of antiretroviral drugs include, but are not limited to: Bone marrow suppression, pancreatitis, gastrointestinal intolerance (nausea, vomiting, diarrhea), neuropathy, rash, hepatotoxicity, lipodystrophy, glucose intolerance, and lactic acidosis. In addition, medications used in the treatment of HIV infection may also have effects on mental function, including cognition (for example, memory), concentration, and mood, and may result in malaise, fatigue, joint and muscle pain, and insomnia. The symptoms of HIV infection and the side effects of medication may be indistinguishable from each other. We will consider all of your functional limitations, whether they result from your symptoms of HIV infection or the side effects of your treatment.

b. *Structured treatment interruptions.* A structured treatment interruption (STI, also called a ''drug holiday'') is a treatment practice during which your treating source advises you to stop taking your medications temporarily. An STI in itself does not imply that your medical condition has improved or that you are noncompliant with your treatment because you are following your treating source's advice. Therefore, if you have stopped taking medication because your treating source prescribed or recommended an STI, we will not find that you are failing to follow treatment or draw inferences about the severity of your impairment on this fact alone. We will consider why your treating source has prescribed or recommended an STI and all the other information in your case record when we determine the severity of your impairment.

6. *When there is no record of ongoing treatment.* If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the medical severity and duration of your immune system impairment on the basis of the current objective medical evidence and other evidence in your case record, taking into consideration your medical history, symptoms, clinical and laboratory findings, and medical source opinions. If you have just begun treatment and we cannot determine whether you are disabled based on the evidence we have, we may need to wait to determine the effect of the treatment on your ability to function. The amount of time we need to wait will depend on the facts of your case. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the immune system listings, but your immune system impairment may medically equal a listing or be disabling based on a consideration of your residual functional capacity, age, education, and work experience.

H. *How do we consider your symptoms, including your constitutional symptoms or pain?*

Your symptoms, including pain, fatigue, and malaise, may be important factors in our determination whether your immune system disorder(s) meets or medically equals a listing or in our determination whether you are otherwise able to work. In order for us to consider your symptoms, you must have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 14.00 and in our other regulations. See §§ 404.1528, 404.1529, 416.928, and 416.929.

I. *How do we use the functional criteria in these listings?*

1. The following listings in this body system include standards for evaluating the

limitations resulting from repeated manifestations of immune system disorders that do not meet the criteria of the other sections of their respective listings: 14.02B, for systemic lupus erythematosus; 14.03B, for systemic vasculitis; 14.04D, for systemic sclerosis (scleroderma); 14.05E, for polymyositis and dermatomyositis; 14.06B, for undifferentiated and mixed connective tissue disease; 14.07C, for immune deficiency disorders, excluding HIV infection; 14.08K, for HIV infection; 14.09D, for inflammatory arthritides; and 14.10B, for Sjöogren's syndrome.

2. When we use one of the listings cited in 14.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder(s) on your ability to function on a sustained basis. Important factors we will consider when we evaluate your functioning under these listings include, but are not limited to: Your symptoms, the frequency and duration of manifestations of your immune system disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

3. As used in these listings, ''repeated'' means that the manifestations occur on an average of three times a year, or once every 4 months, each lasting 2 weeks or more; or the manifestations do not last for 2 weeks but occur substantially more frequently than three times in a year or once every 4 months; or they occur less frequently than an average of three times a year or once every 4 months but last substantially longer than 2 weeks.

4. To satisfy the functional criterion in a listing, your immune system disorder must result in a marked level of limitation in one of three general areas of functioning: Activities of daily living, social functioning, or difficulties in completing tasks due to deficiencies in concentration, persistence, or pace. Functional limitation may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This could result from persistent or intermittent symptoms, such as depression, fatigue, or pain, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. You may also have limitations because of your treatment and its side effects (see 14.00G).

5. When ''marked'' is used as a standard for measuring the degree of functional limitation, it means more than moderate but less than extreme. We do not define ''marked'' by a specific number of different activities of daily living in which your functioning is impaired, different behaviors in which your social functioning is impaired, or tasks that you are able to complete, but by the nature and overall degree of interference with your functioning. You may have a marked limitation when several activities or functions are impaired, or even when only one is impaired. Also, you need not be totally precluded from performing an activity to have a marked limitation, as long as the degree of limitation seriously interferes with your ability to function independently, appropriately, and effectively. The term ''marked'' does not imply that you must be confined to bed, hospitalized, or in a nursing home.

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

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

8. *Completing tasks in a timely manner* involves the ability to sustain concentration, persistence, or pace to permit timely completion of tasks commonly found in work settings. We will find that you have ''marked'' difficulty completing tasks if you have serious limitation in your ability to sustain concentration or pace adequate to complete work-related tasks because of symptoms, such as pain, fatigue, anxiety, or difficulty concentrating, caused by your immune system disorder (including manifestations of the disorder) or its treatment even if you are able to do some routine activities of daily living.

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

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

2. Individuals with immune system disorders, including HIV infection, may manifest signs or symptoms of a mental impairment or of another physical impairment. We may evaluate these impairments under any affected body system. For example, we will evaluate:

a. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 1.00ff.

b. Ocular involvement, such as dry eye, under 2.00ff.

c. Respiratory impairments, such as pleuritis, under 3.00ff.

d. Cardiovascular impairments, such as cardiomyopathy, under 4.00ff.

e. Digestive impairments, such as hepatitis (including hepatitis C), under 5.00ff.

f. Genitourinary impairments, such as nephropathy, under 6.00ff.

g. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 7.00ff.

h. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 8.00ff.

i. Neurologic impairments, such as neuropathy or seizures, under 11.00ff.

j. Mental disorders, such as depression, anxiety, or cognitive deficits, under 12.00ff.

k. Allergic disorders, such as asthma or atopic dermatitis, under 3.00ff or 8.00ff or under the criteria in another affected body system.

l. Syphilis or neurosyphilis under the criteria for the affected body system; for example, 2.00 Special senses and speech, 4.00 Cardiovascular system, or 11.00 Neurological.

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

14.01 *Category of Impairments, Immune System Disorders*

14.02 *Systemic lupus erythematosus.* As described in 14.00D1. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Repeated manifestations of SLE but without the requisite findings in A, resulting in at least two of the constitutional symptoms or signs in A2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.03 *Systemic vasculitis.* As described in 14.00D2. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Repeated manifestations of systemic vasculitis but without the requisite findings

in A, resulting in at least two of the constitutional symptoms or signs in A2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.04 *Systemic sclerosis (scleroderma).* As described in 14.00D3. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. With one of the following:

1. Toe contractures or fixed deformity of one or both feet, resulting in the inability to ambulate effectively as defined in 14.00C6; or

2. Finger contractures or fixed deformity in both hands, resulting in the inability to perform fine and gross movements effectively as defined in 14.00C7; or

3. Atrophy with irreversible damage in one or both lower extremities, resulting in the inability to ambulate effectively as defined in 14.00C6; or

4. Atrophy with irreversible damage in both upper extremities, resulting in the inability to perform fine and gross movements effectively as defined in 14.00C7.

OR

C. Raynaud's phenomenon, characterized by:

1. Gangrene of a toe or finger in at least two extremities, or of a toe and finger; or

2. Ischemia with ulcerations of toes or fingers, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively as defined in 14.00C6 and 14.00C7; or

D. Repeated manifestations of systemic sclerosis (scleroderma) but without the requisite findings in A, B, or C, resulting in at least two of the constitutional symptoms or signs in A2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.05 *Polymyositis and dermatomyositis.* As described in 14.00D4. With:

A. Proximal limb-girdle (pelvic or shoulder) muscle weakness, resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively as defined in 14.00C6 and 14.00C7.

OR

B. Impaired swallowing (dysphagia) with aspiration due to muscle weakness.

OR

C. Impaired respiration due to intercostal and diaphragmatic muscle weakness.

OR

D. Diffuse calcinosis with limitation of joint mobility or intestinal motility.

OR

E. Repeated manifestations of polymyositis or dermatomyositis but without the requisite findings in A, B, or C, resulting in at least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.06 *Undifferentiated and mixed connective tissue disease.* As described in 14.00D5. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Repeated manifestations of undifferentiated or mixed connective tissue disease but without the requisite findings in A, resulting in at least two of the constitutional symptoms or signs in A2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.07 *Immune deficiency disorders, excluding HIV infection.* As described in 14.00E. With:

A. One or more of the following infections. The infection(s) must either be resistant to treatment, or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

OR

B. Stem cell transplantation as described under 14.00E3. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

C. Repeated manifestations of an immune deficiency disorder but without the requisite findings in A or B, resulting in at least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social function.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.08 *Human immunodeficiency virus (HIV) infection.* With documentation as described in 14.00F and one of the following:

A. Bacterial infections:

1. Mycobacterial infection (for example, caused by M. avium-intracellulare, M. kansasii, or M. tuberculosis) at a site other than the lungs, skin, or cervical or hilar lymph nodes, or pulmonary tuberculosis resistant to treatment; or

2. Nocardiosis; or

3. Salmonella bacteremia, recurrent non-typhoid; or

4. Multiple or recurrent bacterial infection(s), including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment three or more times in a 12-month period.

OR

B. Fungal Infections:

1. Aspergillosis; or

2. Candidiasis involving the esophagus, trachea, bronchi, or lungs, or at another site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes; or

3. Coccidioidomycosis, at a site other than the lungs or lymph nodes; or

4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis); or

5. Histoplasmosis, at a site other than the lungs or lymph nodes; or

6. Mucormycosis; or

7. Pneumocystis carinii (jiroveci) pneumonia or extrapulmonary pneumocystis carinii (jiroveci) infection.

OR

C. Protozoan or helminthic infections:

1. Cryptosporidiosis, isosporiasis, or microsporidiosis, with diarrhea lasting for 1 month or longer; or

2. Strongyloidiasis, extra-intestinal; or

3. Toxoplasmosis of an organ other than the liver, spleen, or lymph nodes.

OR

D. Viral infections:

1. Cytomegalovirus disease (documented as described in 14.00F3b(ii)) at a site other than the liver, spleen or lymph nodes; or

2. Herpes simplex virus causing:

a. Mucocutaneous infection (for example, oral, genital, perianal) lasting for 1 month or longer; or

b. Infection at a site other than the skin or mucous membranes (for example, bronchitis, pneumonitis, esophagitis, or encephalitis); or

c. Disseminated infection; or

3. Herpes zoster:

a. Disseminated; or

b. With multidermatomal eruptions that are resistant to treatment; or

4. Progressive multifocal leukoencephalopathy.

OR

E. Malignant neoplasms:

1. Carcinoma of the cervix, invasive, FIGO stage II and beyond; or

2. Kaposi's sarcoma with:

a. Extensive oral lesions; or

b. Involvement of the gastrointestinal tract, lungs, or other visceral organs; or

3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease); or

4. Squamous cell carcinoma of the anus.

OR

F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above), with extensive fungating or ulcerating lesions not responding to treatment (for example, dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal candida, condyloma caused by human papillomavirus, genital ulcerative disease).

OR

G. HIV encephalopathy, characterized by cognitive or motor dysfunction that limits function and progresses.

OR

H. HIV wasting syndrome, characterized by involuntary weight loss of 10 percent or more of baseline (or other significant involuntary weight loss, as described in 14.00F5) and, in the absence of a concurrent illness that could explain the findings, either:

1. Chronic diarrhea with two or more loose stools daily lasting for 1 month or longer; or

2. Chronic weakness and documented fever greater than 38 °C (100.4 °F) for the majority of 1 month or longer.

OR

I. Diarrhea, lasting for 1 month or longer, resistant to treatment, and requiring intravenous hydration, intravenous alimentation, or tube feeding.

OR

J. One or more of the following infections (other than described in A–I, above). The infection(s) must either be resistant to treatment, or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

OR

K. Repeated (as defined in 14.00I3) manifestations of HIV infection, including those listed in 14.08A–J, but without the requisite findings for those listings (for example, carcinoma of the cervix not meeting the criteria in 14.08E, diarrhea not meeting the criteria in 14.08I), or other manifestations (for example, oral hairy leukoplakia, myositis, pancreatitis, hepatitis, peripheral neuropathy, glucose intolerance, muscle weakness, cognitive or other mental impairment) resulting in significant, documented symptoms or signs (for example, fatigue, fever, malaise, involuntary weight loss, pain, night sweats, nausea, vomiting, headaches, or insomnia) and one of the following at the marked level (as defined in 14.00I5):

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.09 *Inflammatory arthritis.* As described in 14.00D6. With:

A. Persistent inflammation or deformity in two or more major peripheral joints resulting in the inability to ambulate effectively or inability to perform fine and gross movements effectively as defined in 14.00C6 and 14.00C7.

OR

B. Inflammation or deformity in one or more major peripheral joints, but with less joint involvement than in A and extra-articular features that do not satisfy the criteria of a listing, with:

1. Involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

C. Ankylosing spondylitis or other spondyloarthropathies, with:

1. Ankylosis (fixation) of the dorsolumbar or cervical spines as shown by appropriate medically acceptable imaging and measured on physical examination at 45° or more of flexion from the vertical position (zero degrees); or

2. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 30° or more of flexion (but less than 45°) measured from the vertical position (zero degrees), and involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity.

OR

D. Repeated manifestations of inflammatory arthritis but without the requisite findings in A, B, or C, resulting in at least two of the constitutional symptoms or signs in B2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

14.10 *Sjögren's syndrome.* As described in 14.00D7. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Repeated manifestations of Sjögren's syndrome but without the requisite findings in A, resulting in at least two of the constitutional symptoms or signs in A2, and one of the following at the marked level:

1. Limitation of activities of daily living.

2. Limitation in maintaining social functioning.

3. Limitation in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

* * * * *

Part B

* * * * *

101.00 Musculoskeletal System

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B. *Loss of function.*

1. *General.* * * * For inflammatory arthritides that result in loss of function because of inflammatory peripheral joint or axial arthritis or sequelae, or because of extra-articular features, see 114.00D6. * * *

* * * * *

L. *Abnormal curvatures of the spine.* * * * When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 114.09C. * * *

* * * * *

108.00 Skin Disorders

* * * * *

D. *How do we assess impairments that may affect the skin and other body systems?*

* * * * *

3. *Autoimmune disorders and other immune system disorders* (for example, systemic lupus erythematosus, scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than one body system. We first evaluate these disorders under the immune system listings in 114.00. We evaluate lupus erythematosus under 114.02, scleroderma under 114.04, symptomatic HIV infection under 114.08, and Sjögren's syndrome under 114.10.

* * * * *

114.00 Immune System Disorders

A. *What disorders do we evaluate under the immune system listings?*

1. *We evaluate immune system disorders that cause dysfunction in one or more components of your immune system.*

a. These listings are examples of immune system disorders that are severe enough to result in marked and severe functional limitations. The dysfunction may be due to problems in antibody production, impaired cell-mediated immunity, a combined type of antibody/cellular deficiency, impaired phagocytosis, or complement deficiency.

b. Immune system disorders may result in recurrent and unusual infections, or inflammation and dysfunction of the body's own tissues. Immune system disorders can cause a deficit in a single organ or body system that results in extreme (that is, very serious) loss of function. They can also cause lesser degrees of limitations in two or more organs or body systems, and when associated with symptoms or signs such as fatigue, fever, malaise, diffuse musculoskeletal pain, or involuntary weight loss, can also result in extreme limitation. In children, immune system disorders or their treatment may also affect growth, development, and performance of age-appropriate activities.

c. In this preface, we organize the discussions of immune system disorders in three categories: Autoimmune disorders; Immune deficiency disorders, excluding human immunodeficiency virus (HIV) infection; and HIV infection.

2. *Autoimmune disorders (114.00D).* Autoimmune disorders are caused by dysfunctional immune responses directed against the body's own tissues, resulting in chronic, multisystem impairments that differ in clinical manifestations, course, and outcome. They are sometimes referred to as rheumatic diseases, connective tissue disorders, or collagen vascular disorders. Some of the features of autoimmune disorders in children differ from the features of the same disorders in adults. The impact of the disorders or their treatment on physical, psychological, and developmental growth of pre-pubertal children may be considerable, and often differs from that of post-pubertal adolescents or adults.

3. *Immune deficiency disorders, excluding HIV infection (114.00E).* Immune deficiency disorders are characterized by recurrent or unusual infections that respond poorly to treatment, and are often associated with complications affecting other parts of the body. Immune deficiency disorders are classified as either *primary* (congenital) or *acquired*. Children with immune deficiency disorders also have an increased risk of malignancies and of having autoimmune disorders.

4. *Human immunodeficiency virus (HIV) infection (114.00F).* HIV infection is caused by a specific retrovirus and may be characterized by increased susceptibility to opportunistic infections, cancers, or other conditions as described in 114.08.

B. *What information do we need to show that you have an immune system disorder?* Generally, we need your medical history, report(s) of physical examination, report(s) of laboratory findings, and in some instances, appropriate medically acceptable imaging or tissue biopsy reports to show that you have an immune system disorder. Therefore, we will make every reasonable effort to obtain your medical history, medical findings, and results of laboratory tests. We explain the information we need in more detail in the sections below.

C. *Definitions*

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. ''Appropriate'' means that the technique used is one that is generally accepted and consistent with the prevailing state of medical knowledge and clinical practice to support the evaluation and diagnosis of the impairment.

2. *Constitutional symptoms or signs* means fatigue, fever, malaise, or involuntary weight loss. *Severe fatigue* means a frequent sense of exhaustion that results in significantly reduced physical activity or mental function. *Malaise* means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.

3. *Disseminated* means that a condition is spread over a considerable area. The type and extent of the spread will depend on your specific disease.

4. *Dysfunction* means that one or more of the body regulatory mechanisms are impaired, causing either an excess or deficiency of immunocompetent cells or their products.

5. *Extra-articular* means ''other than the joints''; for example, the effect is in an organ(s) such as the heart, lungs, kidneys, or skin.

6. *Inability to ambulate effectively* has the same meaning as in 101.00B2b.

7. *Inability to perform fine and gross movements effectively* has the same meaning as in 101.00B2c.

8. *Major peripheral joints* has the same meaning as in 101.00F.

9. *Persistent* means that a sign(s) or symptom(s) has continued over time. The precise meaning will depend on the specific immune system disorder, the usual course of the disorder, and the other circumstances of your clinical course.

10. *Recurrent* means that a condition that previously responded adequately to an appropriate course of treatment returns after a period of remission or regression. The precise meaning, such as the extent of response or remission and the time periods involved, will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

11. *Resistant to treatment* means that a condition did not respond adequately to an appropriate course of treatment. Whether a response is adequate or a course of treatment is appropriate will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

12. *Severe* describes medical severity as used by the medical community. The term does not have the same meaning as it does when we use it in connection with a finding at the second step of the sequential evaluation process in § 416.924.

D. *What are the listed autoimmune disorders in these listings?*

1. *Systemic lupus erythematosus (114.02).*

a. *General.* Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that can affect any organ or body system. It is frequently, but not always, accompanied by constitutional symptoms or signs (fatigue, fever, malaise, involuntary weight loss). Major organ or body system involvement can include: Respiratory (pleuritis, pneumonitis), cardiovascular (endocarditis, myocarditis, pericarditis, vasculitis), renal (glomerulonephritis), hematologic (anemia, leukopenia, thrombocytopenia), skin (photosensitivity), neurologic (seizures), mental (anxiety, fluctuating cognition (''lupus fog''), mood disorders, organic brain syndrome, psychosis), or immune system (inflammatory arthritis) disorders. Immunologically, there is an array of circulating serum auto-antibodies and pro- and anti-coagulant proteins that may occur in a highly variable pattern.

b. *Documentation of SLE.* Generally, but not always, the medical evidence will show that your SLE satisfies the criteria in the current ''Criteria for the Classification of Systemic Lupus Erythematosus'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

2. *Systemic vasculitis (114.03).*

a. *General.* (i) Vasculitis is an inflammation of blood vessels. It may occur acutely in association with adverse drug reactions, certain chronic infections, and occasionally, malignancies. More often, it is chronic and the cause is unknown. Symptoms vary depending on which blood vessels are involved. Systemic vasculitis may also be associated with other autoimmune disorders; for example, SLE or dermatomyositis.

(ii) Children can develop the vasculitis of Kawasaki disease, of which the most serious manifestation is formation of coronary artery aneurysms and related complications. We evaluate heart problems related to Kawasaki disease under the criteria in the cardiovascular listings (104.00ff). Children can also develop the vasculitis of anaphylactoid purpura (Henoch-Schoenlein purpura), which may cause intestinal and renal disorders. We evaluate intestinal and renal disorders related to vasculitis of anaphylactoid purpura under the criteria in the digestive (105.00ff) or genitourinary (106.00ff) listings. Other clinical patterns include, but are not limited to, polyarteritis nodosa, Takayasu's arteritis (aortic arch arteritis), and Wegener's granulomatosis.

b. *Documentation of systemic vasculitis.* Angiography or tissue biopsy confirms a diagnosis of systemic vasculitis when the disease is suspected clinically. Usually the results will be in your medical records.

3. *Systemic sclerosis (scleroderma) (114.04).*

a. *General.* Systemic sclerosis (scleroderma) constitutes a spectrum of disease in which thickening of the skin is the clinical hallmark. Raynaud's phenomenon, often medically severe and progressive, is present frequently and may be the peripheral manifestation of a vasospastic abnormality in the heart, lungs, and kidneys. The CREST syndrome (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) is a variant that may slowly progress over years to the generalized process, systemic sclerosis.

b. *Diffuse cutaneous systemic sclerosis.* In diffuse cutaneous systemic sclerosis (also known as diffuse scleroderma), major organ or systemic involvement can include the gastrointestinal tract, lungs, heart, kidneys, and muscle in addition to skin or blood vessels. Although arthritis can occur, joint dysfunction results primarily from soft tissue/cutaneous thickening, fibrosis, and contractures.

c. *Localized scleroderma (linear scleroderma and morphea).*

(i) Localized scleroderma (linear scleroderma and morphea) is more common in children than systemic scleroderma. The extent of involvement of linear scleroderma and a description of the lesions are important in assessing the severity of the impairment.

For example, linear scleroderma involving the arm but not crossing any joints is not as functionally limiting as sclerodactyly (scleroderma localized to the fingers). Linear scleroderma of a lower extremity involving skin thickening and atrophy of underlying muscle or bone can result in contracture(s) and leg length discrepancies. In such cases, evaluation under the musculoskeletal (101.00ff) listings may be appropriate.

(ii) When there is isolated morphea of the face causing facial disfigurement from unilateral hypoplasia of the mandible, maxilla, zygoma, or orbit, adjudication may be more appropriate under the criteria in the special senses listings (102.00ff) or mental disorders listings (112.00ff).

(iii) Chronic variants of these syndromes include disseminated morphea, Shulman's disease (diffuse fasciitis with eosinophilia), and eosinophilia-myalgia syndrome (often associated with toxins such as toxic oil or contaminated tryptophan), all of which can impose medically severe musculoskeletal impairment and may also lead to restrictive pulmonary disease. We evaluate these variants of the disease under the criteria in the musculoskeletal listings (101.00ff) or respiratory system listings (103.00ff).

d. *Documentation of systemic sclerosis (scleroderma).* Documentation involves differentiating the clinical features of systemic sclerosis (scleroderma) from other autoimmune disorders; however, there may be an overlap.

4. *Polymyositis and dermatomyositis (114.05).*

a. *General.*

(i) Polymyositis and dermatomyositis are related disorders that are characterized by an inflammatory process in striated muscle, occurring alone or in association with other autoimmune disorders. Symmetric weakness, and less frequently pain and tenderness of the proximal limb-girdle (shoulder or pelvic) musculature, are the most common manifestations. There may also be involvement of the cervical, cricopharyngeal, esophageal, intercostal, and diaphragmatic muscles.

(ii) Polymyositis occurs rarely in children; the more common presentation in children is dermatomyositis with symmetric proximal muscle weakness and characteristic skin rash. The clinical course of dermatomyositis can be more severe when it is accompanied by systemic vasculitis rather than just localized to striated muscle. Late in the disease, some children with dermatomyositis develop calcinosis of the skin and subcutaneous tissues, muscles and joints. We evaluate the involvement of other organs/body systems under the criteria for the listings in the affected body system.

b. *Documentation of polymyositis and dermatomyositis.* Generally, but not always, polymyositis is associated with elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, aldolase), and characteristic abnormalities on electromyography and muscle biopsy. In children, the diagnosis of dermatomyositis is supported largely by medical history, findings on physical examination that include the characteristic skin rash, and elevated serum muscle enzymes. Additional evidence of the diagnosis of childhood dermatomyositis is depiction on MRI of muscle inflammation or vasculitis.

c. *Additional information about how we evaluate polymyositis and dermatomyositis under the listings.*

(i) In newborn and younger infants (birth to attainment of age 1), we consider muscle weakness that affects motor skills, such as head control, reaching, grasping, taking solids, or self-feeding under 114.05A. In older infants and toddlers (age 1 to attainment of age 3), we also consider muscle weakness affecting the child's ability to roll over, sit, crawl, or walk under 114.05A.

(ii) If you are of preschool age through adolescence (age 3 to attainment of age 18), weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to ambulate effectively. Weakness of your shoulder girdle muscles may result in your inability to perform lifting, carrying, and reaching overhead, and also may seriously affect your ability to perform activities requiring fine movements. We evaluate these limitations under 114.05A.

5. *Undifferentiated and mixed connective tissue disease (114.06).*

a. *General.* This listing includes syndromes with clinical and immunologic features of several autoimmune disorders, but which do not satisfy the criteria for any of the specific disorders described. For example, you may have clinical features of systemic lupus erythematosus and systemic vasculitis, and the serologic (blood test) findings of rheumatoid arthritis. The most common pattern of undifferentiated autoimmune disorders in children is mixed connective tissue disease (MCTD).

b. *Documentation of undifferentiated and mixed connective tissue disease.* Undifferentiated connective tissue disease is diagnosed when clinical features and serologic (blood test) findings, such as rheumatoid factor or antinuclear antibody (consistent with an autoimmune disorder) are present but do not satisfy the criteria for a specific disease. Children with MCTD have laboratory findings of extremely high antibody titers to extractable nuclear antigen (ENA) or ribonucleoprotein (RNP) without high titers of anti-dsDNA or anti-SM antibodies. There are often clinical findings suggestive of SLE or childhood dermatomyositis. Many children later develop features of scleroderma.

6. *Inflammatory arthritis (114.09).*

a. *General.* The inflammatory arthritides include a vast array of disorders that differ in cause, course, and outcome. Clinically, inflammation of major peripheral joints may be the dominant manifestation causing difficulties with ambulation or fine and gross movements; there may be joint pain, swelling, and tenderness. The arthritis may affect other joints, or cause less functional limitations in ambulation or performance of fine and gross movements. However, in combination with extra-articular features, including constitutional symptoms or signs (fatigue, fever, malaise, involuntary weight loss), inflammatory arthritis may result in an extreme limitation. You may also have impaired growth as a result of the inflammatory arthritides because of its effects on the immature skeleton, open epiphyses, and young cartilage and bone. We evaluate any associated growth impairment under the criteria in 100.00ff.

b. *Inflammatory arthritides involving the axial spine (spondyloarthropathies).* In children, inflammatory arthritides involving the axial spine may be associated with heterogeneous disorders such as:

(i) Reactive arthropathies;

(ii) Juvenile ankylosing spondylitis;

(iii) Psoriatic arthritis;

(iv) SEA syndrome (seronegative enthesopathy arthropathy syndrome);

(v) Behçet's disease; and

(vi) Inflammatory bowel disease.

c. *Inflammatory arthritides involving the peripheral joints.* In children, the inflammatory arthropathies involving peripheral joints may be associated with disorders such as:

(i) Juvenile rheumatoid arthritis;

(ii) Sjögren's syndrome;

(iii) Psoriatic arthritis;

(iv) Crystal deposition disorders (gout and pseudogout);

(v) Lyme disease; and

(vi) Inflammatory bowel disease.

d. *Documentation of inflammatory arthritides.* Generally, but not always, the diagnosis of inflammatory arthritis is made by the clinical features and serologic findings described in the most recent edition of the *Primer on Rheumatic Diseases* published by the Arthritis Foundation.

e. *How we evaluate the inflammatory arthritides under the listings.*

(i) Listing-level severity in 114.09A and 114.09C1 is shown by an impairment that results in an ''extreme'' (very serious) limitation. In 114.09A, the criterion is satisfied with persistent inflammation or deformity in two or more major peripheral joints resulting in the inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 114.00C6 and 114.00C7. In 114.09C1, if you have the required ankylosis (fixation) of your cervical or dorsolumbar spine, we will find that you have an extreme limitation in your ability to see in front of you, above you, and to the side. Therefore, inability to ambulate effectively is implicit in 114.09C1, even though you might not require bilateral upper limb assistance.

(ii) Listing-level severity is shown in 114.09B, 114.09C2, and 114.09D when the arthritis does not result in the extreme limitation in 114.09A or 114.09C1, involves one or more major peripheral joints, or involves other joints, but is complicated by extra-articular features that cumulatively result in an ''extreme'' (very serious) limitation or ''marked'' (serious) limitations in at least two areas of functioning. Extra-articular impairments may also meet listings in other body systems.

(iii) Extra-articular features of inflammatory arthritis may involve any body system. Commonly occurring extra-articular impairments include: Musculoskeletal (heel enthesopathy), ophthalmologic (iridocyclitis, keratoconjunctivitis sicca, uveitis), pulmonary (pleuritis, pulmonary fibrosis or

nodules, restrictive lung disease), cardiovascular (aortic valve insufficiency, arrhythmias, coronary arteritis, myocarditis, pericarditis, Raynaud's phenomenon, systemic vasculitis), renal (amyloidosis of the kidney), hematologic (chronic anemia, thrombocytopenia), neurologic (peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss), and immune system (Felty's syndrome (hypersplenism with compromised immune competence)) disorders.

(iv) If permanent deformity of a major peripheral joint is the dominant feature of your impairment, we evaluate your impairment under 101.02.

(v) If there has been surgical reconstruction of a major weight-bearing joint, we evaluate your impairment under 101.03.

(vi) If both inflammation and chronic deformities are present, we evaluate your impairment under the criteria of any appropriate listing.

7. *Sjögren's syndrome (114.10).*

a. *General.* (i) Sjögren's syndrome is an immune-mediated disorder of the exocrine glands. Involvement of the lacrimal and salivary glands is the hallmark feature, resulting in symptoms of dry eyes and dry mouth, and possible complications such as corneal damage, blepharitis (eyelid inflammation), dysphagia (difficulty in swallowing), dental caries, and the inability to speak for extended periods of time. Involvement of the exocrine glands of the upper airways may result in persistent dry cough.

(ii) Many other organ systems may be involved, including musculoskeletal (arthritis, myositis), respiratory (interstitial fibrosis), gastrointestinal (dysmotility, dysphagia, involuntary weight loss), genitourinary (interstitial cystitis, renal tubular acidosis), skin (purpura, vasculitis,), neurologic (central nervous system disorders, cranial and peripheral neuropathies), mental (cognitive dysfunction, poor memory), and neoplastic (lymphoma). Fatigue and malaise are frequently reported. Sjögren's syndrome may be associated with other autoimmune disorders (for example, rheumatoid arthritis or SLE); usually the clinical features of the associated disorder predominate.

b. *Documentation of Sjögren's syndrome.* If you have Sjögren's syndrome, the medical evidence will generally, but not always, show that your disease satisfies the criteria in the current ''Criteria for the Classification of Sjögren's Syndrome'' by the American College of Rheumatology found in the most recent edition of the *Primer on the Rheumatic Diseases* published by the Arthritis Foundation.

E. *How do we evaluate immune deficiency disorders, excluding HIV infection (114.07)?*

1. *General.*

a. Immune deficiency disorders can be classified as:

(i) *Primary* (congenital); for example, X-linked agammaglobulinemia, thymic hypoplasia (DiGeorge syndrome), severe combined immunodeficiency (SCID), chronic granulomatous disease (CGD), C1 esterase inhibitor deficiency.

(ii) *Acquired;* for example, medication-related.

b. Primary immune deficiency disorders are seen mainly in children. However, recent advances in the treatment of these disorders have allowed many affected children to survive well into adulthood. Occasionally, these disorders are first diagnosed in adolescence or adulthood.

2. *Documentation of immune deficiency disorders.* The medical evidence must include documentation of the specific type of immune deficiency. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

3. *Immune deficiency disorders treated by stem cell transplantation.*

a. *Evaluation in the first 12 months.* If you undergo stem cell transplantation for your immune deficiency disorder, we will consider you disabled until at least 12 months from the date of the transplant.

b. *Evaluation after the 12-month period has elapsed.* After the 12-month period has elapsed, we will consider any residuals of your immune deficiency disorder as well as any residual impairment(s) resulting from treatment, such as complications arising from:

(i) Graft-versus-host (GVH) disease.

(ii) Immunosuppressant therapy, such as frequent infections.

(iii) Significant deterioration of other organ systems.

4. *Medication-induced immune suppression.* Medication effects can result in varying degrees of immune suppression, but most resolve when the medication is ceased. However, if you are prescribed medication for long-term immune suppression, such as after an organ transplant, we will evaluate:

a. The frequency and severity of infections.

b. Residuals from the organ transplant itself, after the 12-month period has elapsed.

c. Significant deterioration of other organ systems.

F. *How do we evaluate human immunodeficiency virus (HIV) infection?* Any child with HIV infection, including one with a diagnosis of acquired immune deficiency syndrome (AIDS), may be found disabled under 114.08 if his or her impairment meets the criteria in that listing or is medically equivalent to the criteria in that listing.

1. *Documentation of HIV infection.* The medical evidence must include documentation of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for HIV infection, we will make every reasonable effort to obtain reports of the results of that testing.

a. *Documentation of HIV infection by definitive diagnosis.* A definitive diagnosis of HIV infection is documented by one or more of the following laboratory tests:

(i) HIV antibody tests. HIV antibodies are usually first detected by an ELISA screening test performed on serum. Because the ELISA can yield false positive results, confirmation is required using a more definitive test, such as a Western blot or an immunofluorescence assay. Positive results on these tests are considered to be diagnostic of HIV infection in a child age 18 months or older. (See b. below, for information about HIV antibody testing in children younger than 18 months of age.)

(ii) Positive ''viral load'' (VL) tests. These tests are normally used to quantitate the amount of the virus present but also document HIV infection. Such tests include the quantitative plasma HIV RNA, quantitative plasma HIV branched DNA, and reverse transcriptase-polymerase chain reaction (RT–PCR).

(iii) HIV DNA detection by polymerase chain reaction (PCR).

(iv) A specimen that contains HIV antigen (for example, serum specimen, lymphocyte culture, or cerebrospinal fluid), in a child age 1 month or older.

(v) A positive viral culture for HIV from peripheral blood mononuclear cells (PBMC).

(vi) An immunoglobulin A (IgA) serological assay that is specific for HIV.

(vii) Other tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge.

b. *Documentation of HIV infection in children from birth to the attainment of 18 months.* For children from birth to the attainment of 18 months of age, and who have tested positive for HIV antibodies, HIV infection is documented by:

(i) One or more of the tests listed in F1a(ii)–F1a(vii).

(ii) For newborn and younger infants (birth to attainment of age 1), a CD4 (T4) count of 1500/mm^3 or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iii) For older infants and toddlers from 12 to 18 months of age, a CD4 (T4) count of 750/mm^3 or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iv) An abnormal CD4/CD8 ratio.

(v) A severely diminished immunoglobulin G (IgG) level (<4 g/l or 400 mg/dl), or significantly greater than normal range for age.

c. *Other acceptable documentation of HIV infection.* We may also document HIV infection without the definitive laboratory evidence described in 114.00F1a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice, and is consistent with the other evidence in your case record. If no definitive laboratory evidence is available, we may document HIV infection by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence. For example, we will accept a diagnosis of HIV infection without definitive laboratory evidence if you have an opportunistic disease that is predictive of a defect in cell-mediated immunity (for example, Pneumocystis carinii pneumonia (PCP)), and there is no other known cause of diminished resistance to that disease (for example, long-term steroid treatment, lymphoma). In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

2. *CD4 tests.* Children who have HIV infection or other disorders of the immune system may have tests showing a reduction of either the absolute count or the percentage

of their T-helper lymphocytes (CD4 cells). The extent of immune suppression correlates with the level or rate of decline of the CD4 count. At age 6, children begin to have CD4 counts comparable to the levels found in adults. Generally, in these children when the CD4 count is 200/mm^3 or less (14 percent or less) the susceptibility to opportunistic infection is greatly increased. Although a reduced CD4 count alone does not establish a definitive diagnosis of HIV infection, a CD4 count below 200 does offer supportive evidence when there are clinical findings, but not a definitive diagnosis of an opportunistic infection(s). However, a reduced CD4 count *alone* does not document the severity or functional consequences of HIV infection.

3. *Documentation of the manifestations of HIV infection.* The medical evidence must also include documentation of the manifestations of HIV infection. Documentation may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice. When you have had laboratory testing for a manifestation of HIV infection, we will make every reasonable effort to obtain reports of the results of that testing.

a. *Documentation of the manifestations of HIV infection by definitive diagnosis.* The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serologic test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). We will make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histologic or other test has been performed, the evidence should include a copy of the appropriate report. If we cannot obtain the report, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including appropriate medically acceptable imaging studies) or microscopic examination of the appropriate tissues or body fluids.

b. *Other acceptable documentation of the manifestations of HIV infection.* We may also document manifestations of HIV infection without the definitive laboratory evidence described in 114.00F3a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice, and is consistent with the other evidence in your case record. If no definitive evidence is available, we may document the manifestations of HIV infection with other appropriate evidence. For example, many conditions are now commonly diagnosed based on some or all of the following: Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

(i) Although a definitive diagnosis of PCP requires identifying the organism in bronchial washings, induced sputum, or lung biopsy, these tests are frequently bypassed if PCP can be diagnosed presumptively. (**Note:** *Pneumocystis carinii* is now known as *Pneumocystis jiroveci;* however, ''PCP'' remains in common usage for the pneumonia caused by this organism.) Supportive evidence includes: Fever, dyspnea, hypoxia, and CD4 count below 200 in children 6 years of age or older. Also supportive are bilateral lung interstitial infiltrates on x-ray, or a typical pattern on CT scan, or a gallium scan positive for pulmonary uptake. Response to anti-PCP therapy usually requires 5–7 days.

(ii) Documentation of cytomegalovirus (CMV) disease (114.08D) may present special problems because definitive diagnosis (except for chorioretinitis, which may be diagnosed by an ophthalmologist on funduscopic exam) requires identification of viral inclusion bodies or a positive culture from the affected organ and the absence of any other infectious agent likely to be causing the disease. A positive serology test identifies a history of infection with CMV, but it does not confirm an active disease process. Therefore, a presumptive diagnosis of CMV disease requires corroborating evidence that CMV is causing the disease. Supportive evidence includes: Fever, positive CMV serology test, urinary culture positive for CMV, and CD4 count below 200 in children 6 years of age or older. A clear response to anti-CMV therapy also supports a diagnosis.

(iii) A definitive diagnosis of toxoplasmosis of the brain is made by brain biopsy, but this procedure carries significant risk and is not commonly performed. This condition is usually diagnosed presumptively based on symptoms or signs of fever, headache, focal neurologic deficits, seizures, typical lesions on brain imaging, and a positive serology test.

4. *HIV infection manifestations specific to children.*

a. *General.* The clinical manifestation and course of disease in children who become infected with HIV perinatally or in the first 6 years of life may differ from that in adolescents (age 12 to attainment of age 18) and adults. Newborn and younger infants (birth to attainment of age 1) and older infants and toddlers (age 1 to attainment of age 3) may present with failure to thrive or PCP; preschool children (age 3 to attainment of age 6) and primary school children (age 6 to attainment of age 12) may present with recurrent infections, neurological problems, or developmental abnormalities. Adolescents may also exhibit neurological abnormalities such as HIV encephalopathy, or have growth problems.

b. *Neurologic abnormalities.* The methods of identifying and evaluating neurologic abnormalities may vary depending on a child's age. For example, in an infant impaired brain growth can be documented by a decrease in the growth rate of the head. In an older child, impaired brain growth may be documented by brain atrophy on a CT scan or MRI. Neurologic abnormalities in infants and young children may present as serious developmental delays or in the loss of previously acquired developmental milestones. In school-age children and adolescents, this type of neurologic abnormality would generally present as the loss of previously acquired intellectual abilities. This may be evidenced by a decrease in intelligence quotient (IQ) scores, by a child forgetting information he or she previously learned, by being unable to learn new information, or by a sudden onset of a new learning disability.

c. *Bacterial infections.* Children with HIV infection may contract any of a broad range of bacterial infections. Certain major infections caused by pyogenic bacteria (for example, some pneumonias) can be severely limiting, especially in pre-adolescent children. We evaluate these major bacterial infections under 114.08A4. Although 114.08A4 applies only to children under 13 years of age, children age 13 and older may have an impairment that medically equals this listing if the circumstances of the case warrant; for example, if there is delayed puberty. We will evaluate pelvic inflammatory disease in older girls under 114.08A5.

G. *How will we consider the effect of treatment in evaluating your autoimmune disorder, immune deficiency disorder, or HIV infection?*

1. *General.* If your impairment does not otherwise meet the requirements of a listing we will consider your medical treatment both in terms of its effectiveness in improving the signs, symptoms, and laboratory abnormalities of your specific immune system disorder or its manifestations, and in terms of any side effects that limit your functioning. We will make every reasonable effort to obtain a specific description of the treatment you receive (including surgery) for your immune system disorder. We consider:

a. The effects of medications you take.

b. Adverse side effects (acute and chronic).

c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).

d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).

e. Variability of your response to treatment (see 114.00G2).

f. The interactive and cumulative effects of your treatments. For example, many individuals with immune system disorders receive treatment both for their immune system disorders and for the manifestations of the disorders or co-occuring impairments, such as treatment for HIV infection and hepatitis C. The interactive and cumulative effects of these treatments may be greater than the effects of each treatment considered separately.

g. The duration of your treatment.

h. Any other aspects of treatment that may interfere with your ability to function.

2. *Variability of your response to treatment.* Your response to treatment and the adverse or beneficial consequences of your treatment may vary widely. The effects of your treatment may be temporary or long term. For example, some individuals may show an initial positive response to a drug or combination of drugs followed by a decrease in effectiveness. When we evaluate your response to treatment and how your treatment may affect you, we consider such factors as disease activity before treatment,

requirements for changes in therapeutic regimes, the time required for therapeutic effectiveness of a particular drug or drugs, the limited number of drug combinations that may be available for your impairment(s), and the time-limited efficacy of some drugs. For example, a child with HIV infection or another immune deficiency disorder who develops otitis media may not respond to the same antibiotic regimen used in treating children without these disorders or may not respond to an antibiotic that he or she responded to before. Therefore, we must consider the effects of your treatment on an individual basis, including the effects of your treatment on your ability to function.

3. *How we evaluate the effects of treatment for autoimmune disorders on your ability to function.* Some medications may have acute or long-term side effects. When we consider the effects of corticosteroids or other treatments for autoimmune disorders on your ability to function, we consider the factors in 114.00G1 and 114.00G2. Long-term corticosteroid treatment can cause ischemic necrosis of bone, posterior subcapsular cataract, impaired growth, weight gain, glucose intolerance, increased susceptibility to infection, and osteopenia that may result in a loss of function. In addition, medications used in the treatment of autoimmune disorders may also have effects on mental function including cognition (for example, memory), concentration, and mood.

4. *How we evaluate the effects of treatment for immune deficiency disorders, excluding HIV infection, on your ability to function.* When we consider the effects of your treatment for your immune deficiency disorder on your ability to function, we consider the factors in 114.00G1 and 114.00G2. A frequent need for treatment such as intravenous immunoglobulin and gamma interferon therapy can be intrusive and interfere with your ability to function. We will also consider whether you have chronic side effects from these or other medications, including fatigue, fever, headaches, high blood pressure, joint swelling, muscle aches, nausea, shortness of breath, or limitations in mental function including cognition (for example, memory) concentration, and mood.

5. *How we evaluate the effects of treatment for HIV infection on your ability to function.*

a. *General.* When we consider the effects of antiretroviral drugs (including the effects of highly active antiretroviral therapy (HAART)) and the effects of treatments for the manifestations of HIV infection on your ability to function, we consider the factors in 114.00G1 and 114.00G2. Side effects of antiretroviral drugs include, but are not limited to: Bone marrow suppression, pancreatitis, gastrointestinal intolerance (nausea, vomiting, diarrhea), neuropathy, rash, hepatotoxicity, lipodystrophy, glucose intolerance, and lactic acidosis. In addition, medications used in the treatment of HIV infection may also have effects on mental function, including cognition (for example, memory), concentration, and mood, and may result in malaise, fatigue, joint and muscle pain, and insomnia. The symptoms of HIV infection and the side effects of medication may be indistinguishable from each other. We will consider all of your functional limitations, whether they result from your symptoms of HIV infection or the side effects of your treatment.

b. *Structured treatment interruptions.* A structured treatment interruption (STI, also called a ''drug holiday'') is a treatment practice during which your treating source advises you to stop taking your medications temporarily. An STI in itself does not imply that your medical condition has improved or that you are noncompliant with your treatment because you are following your treating source's advice. Therefore, if you have stopped taking medication because your treating source prescribed or recommended an STI, we will not find that you are failing to follow treatment or draw inferences about the severity of your impairment on this fact alone. We will consider why your treating source has prescribed or recommended an STI and all the other information in your case record when we determine the severity of your impairment.

6. *When there is no record of ongoing treatment.* If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the medical severity and duration of your immune system impairment on the basis of the current objective medical evidence and other evidence in your case record, taking into consideration your medical history, symptoms, clinical and laboratory findings, and medical source opinions. If you have just begun treatment and we cannot determine whether you are disabled based on the evidence we have, we may need to wait to determine the effect of the treatment on your ability to function. The amount of time we need to wait will depend on the facts of your case. If you have not received treatment, you may not be able to show an impairment that meets the criteria of one of the immune system listings, but your immune system impairment may medically equal a listing or functionally equal the listings.

H. *How do we consider your symptoms, including your constitutional symptoms or pain?*

Your symptoms, including pain, fatigue, and malaise, may be important factors in our determination whether your immune system disorder(s) meets or medically equals a listing or in our determination whether you otherwise have marked and severe functional limitations. In order for us to consider your symptoms, you must have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 114.00 and in our other regulations. See §§ 416.928, and 416.929.

I. *How do we use the functional criteria in these listings?*

1. The following listings in this body system include standards for evaluating the limitations resulting from manifestations of immune system disorders that do not meet the criteria of the other sections of their respective listings: 114.02B, for systemic lupus erythematosus; 114.03B, for systemic vasculitis; 114.04D, for systemic sclerosis (scleroderma); 114.05E, for polymyositis and dermatomyositis; 114.06B, for undifferentiated and mixed connective tissue disease; 114.07C, for immune deficiency disorders, excluding HIV infection; 114.08L, for HIV infection; 114.09D, for inflammatory arthritides; and 114.10B, for Sjögren's syndrome.

2. When we use one of the listings cited in 114.00I1, we will consider all relevant information in your case record to determine the full impact of your immune system disorder(s) on your ability to function on a sustained basis. Important factors we will consider when we evaluate your functioning under these listings include, but are not limited to: Your symptoms, the frequency and duration of manifestations of your immune system disorder, periods of exacerbation and remission, and the functional impact of your treatment, including the side effects of your medication.

3. To satisfy the functional criterion in a listing, your immune system disorder must result in an ''extreme'' limitation in one domain of functioning or ''marked'' limitations in two domains of functioning depending on your age. (See 112.00C for additional discussion of these areas of functioning and §§ 416.924a and 416.926a for additional guidance on the evaluation of functioning in children.) Functional limitation may result from the impact of the disease process itself on your mental functioning, physical functioning, or both your mental and physical functioning. This could result from persistent or intermittent symptoms, such as depression, fatigue, or pain, resulting in a limitation of your ability to do a task, to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. You may also have limitations because of your treatment and its side effects (see 114.00G).

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

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

2. Individuals with immune system disorders, including HIV infection, may manifest signs or symptoms of a mental impairment or of another physical impairment. We may evaluate these impairments under any affected body system. For example, we will evaluate:

a. Growth impairment under 100.00ff.

b. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 101.00ff.

c. Ocular involvement, such as dry eye, under 102.00ff

d. Respiratory impairments, such as pleuritis, under 103.00ff.

e. Cardiovascular impairments, such as cardiomyopathy, under 104.00ff.

f. Digestive impairments, such as hepatitis (including hepatitis C), under 105.00ff.

g. Genitourinary impairments, such as nephropathy, under 106.00ff.

h. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 107.00ff.

i. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 108.00ff.

j. Neurologic impairments, such as neuropathy or seizures, under 111.00ff.

k. Mental disorders, such as depression, anxiety, or cognitive deficits, under 112.00ff.

l. Allergic disorders, such as asthma or atopic dermatitis, under 103.00ff or 108.00ff or under the criteria in another affected body system.

m. Syphilis or neurosyphilis under the criteria for the affected body system; for example, 102.00 Special senses and speech, 104.00 Cardiovascular system, or 111.00 Neurological.

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

114.01 *Category of Impairments, Immune System Disorders*

114.02 *Systemic lupus erythematosus.* As described in 114.00D1. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Any other manifestation(s) of SLE resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.03 *Systemic vasculitis.* As described in 114.00D2. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Any other manifestation(s) of systemic vasculitis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.04 *Systemic sclerosis (scleroderma).* As described in 114.00D3. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. With one of the following:

1. Toe contractures or fixed deformity of one or both feet, resulting in the inability to ambulate effectively as defined in 114.00C6; or

2. Finger contractures or fixed deformity in both hands, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7; or

3. Atrophy with irreversible damage in one or both lower extremities, resulting in the inability to ambulate effectively as defined in 114.00C6; or

4. Atrophy with irreversible damage in both upper extremities, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7.

OR

C. Raynaud's phenomenon, characterized by:

1. Gangrene of a toe or finger in at least two extremities, or of a toe and finger; or

2. Ischemia with ulcerations of toes or fingers, resulting in the inability to ambulate effectively or to perform fine and gross movements effectively as defined in 114.00C6 and 114.00C7; or

D. Any other manifestation(s) of systemic sclerosis (scleroderma) resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.05 *Polymyositis and dermatomyositis.* As described in 114.00D4. With:

A. Proximal limb-girdle (pelvic or shoulder) muscle weakness, resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively as defined in 114.00C6 and 114.00C7.

OR

B. Impaired swallowing (dysphagia) and aspiration due to muscle weakness.

OR

C. Impaired respiration due to intercostal and diaphragmatic muscle weakness.

OR

D. Diffuse calcinosis with limitation of joint mobility or intestinal motility.

OR

E. Any other manifestation(s) of polymyositis or dermatomyositis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.06 *Undifferentiated and mixed connective tissue disease.* As described in 114.00D5. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Any other manifestation(s) of undifferentiated or mixed connective tissue disease resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.07 *Immune deficiency disorders, excluding HIV infection.* As described in 114.00E. With:

A. One or more of the following infections. The infection(s) must either be resistant to treatment, or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

OR

B. Stem cell transplantation as described under 114.00E3. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

C. Any other manifestations(s) of an immune deficiency disorder resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A–E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.08 *Human immunodeficiency virus (HIV) infection.* With documentation as

described in 114.00F and one of the following:

A. Bacterial infections:

1. Mycobacterial infection (for example, caused by M. avium-intracellulare, M. kansasii, or M. tuberculosis) at a site other than the lungs, skin, or cervical or hilar lymph nodes, or pulmonary tuberculosis resistant to treatment; or

2. Nocardiosis; or

3. Salmonella bacteremia, recurrent non-typhoid; or

4. In a child less than 13 years of age, multiple or recurrent pyogenic bacterial infection(s) (sepsis, pneumonia, meningitis, bone or joint infection, or abscess of an internal organ or body cavity, but not otitis media or superficial skin or mucosal abscesses) occurring two or more times in 2 years; or

5. Multiple or recurrent bacterial infection(s), including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment three or more times in a 12-month period.

OR

B. Fungal infections:

1. Aspergillosis; or

2. Candidiasis involving the esophagus, trachea, bronchi, or lungs, or at another site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes; or

3. Coccidioidomycosis, at a site other than the lungs or lymph nodes; or

4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis); or

5. Histoplasmosis, at a site other than the lungs or lymph nodes; or

6. Mucormycosis; or

7. Pneumocystis carinii (jiroveci) pneumonia or extrapulmonary pneumocystis carinii (jiroveci) infection.

OR

C. Protozoan or helminthic infections:

1. Cryptosporidiosis, isosporiasis, or microsporidiosis, with diarrhea lasting for 1 month or longer; or

2. Strongyloidiasis, extra-intestinal; or

3. Toxoplasmosis of an organ other than the liver, spleen, or lymph nodes.

OR

D. Viral infections:

1. Cytomegalovirus disease (documented as described in 114.00F3b(ii)) at a site other than the liver, spleen, or lymph nodes; or

2. Herpes simplex virus causing:

a. Mucocutaneous infection (for example, oral, genital, perianal) lasting for 1 month or longer; or

b. Infection at a site other than the skin or mucous membranes (for example, bronchitis, pneumonitis, esophagitis, or encephalitis); or

c. Disseminated infection; or

3. Herpes zoster:

a. Disseminated; or

b. With multidermatomal eruptions that are resistant to treatment; or

4. Progressive multifocal leukoencephalopathy.

OR

E. Malignant neoplasms:

1. Carcinoma of the cervix, invasive, FIGO stage II and beyond; or

2. Kaposi's sarcoma with:

a. Extensive oral lesions; or

b. Involvement of the gastrointestinal tract, lungs, or other visceral organs; or

3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease); or

4. Squamous cell carcinoma of the anus.

OR

F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above), with extensive fungating or ulcerating lesions not responding to treatment (for example, dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal candida, condyloma caused by human papillomavirus, genital ulcerative disease).

OR

G. Neurological manifestations of HIV infection (for example, HIV encephalopathy, peripheral neuropathy) resulting in one of the following:

1. Loss of previously acquired, or marked delay in achieving, developmental milestones or intellectual ability (including the sudden onset of a new learning disability); or

2. Impaired brain growth (acquired microcephaly or brain atrophy—see 114.00F4b); or

3. Progressive motor dysfunction affecting gait and station or fine and gross motor skills.

OR

H. Growth disturbance, with:

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

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

3. Involuntary weight loss of 10 percent or more of baseline that persists for 2 months or longer.

OR

I. Diarrhea, lasting for 1 month or longer, resistant to treatment, and requiring intravenous hydration, intravenous alimentation, or tube feeding.

OR

J. Lymphoid interstitial pneumonia/pulmonary lymphoid hyperplasia (LIP/PLH complex), with respiratory symptoms that significantly interfere with age-appropriate activities, and that cannot be controlled by prescribed treatment.

OR

K. One or more of the following infections (other than described in A-J, above). The infection(s) must either be resistant to treatment, or require hospitalization or intravenous treatment three or more times in a 12-month period.

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

OR

L. Any other manifestation(s) of HIV infection, including those listed in 114.08A–K, but without the requisite findings for those listings (for example, oral candidiasis not meeting the criteria in 114.08F, diarrhea not meeting the criteria in 114.08I), or other manifestation(s) (for example, oral hairy leukoplakia, hepatomegaly), resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.09 *Inflammatory arthritis. As described in 114.00D6.* With:

A. Persistent inflammation or deformity in two or more major peripheral joints resulting in the inability to ambulate effectively or the inability to perform fine and gross movements effectively as defined in 114.00C6 and 114.00C7.

OR

B. Inflammation or deformity in one or more major peripheral joints, but with less joint involvement than in A and extra-articular features that do not satisfy the criteria of a listing, with:

1. Involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

C. Ankylosing spondylitis or other spondyloarthropathies, with:

1. Ankylosis (fixation) of the dorsolumbar or cervical spines as shown by appropriate medically acceptable imaging and measured on physical examination at 45° or more of flexion from the vertical position (zero degrees); or

2. Ankylosis (fixation) of the dorsolumbar or cervical spine as shown by appropriate medically acceptable imaging and measured on physical examination at 30° or more of flexion (but less than 45°) measured from the vertical position (zero degrees), and involvement of two or more organs/body systems with one of the organs/body systems involved to at least a moderate level of severity.

OR

D. Any other manifestation(s) of inflammatory arthritis resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.10 *Sjögren's syndrome.* As described n 114.00D7. With:

A. Involvement of two or more organs/body systems, with:

1. One of the organs/body systems involved to at least a moderate level of severity, and

2. At least two of the following constitutional symptoms or signs: Severe fatigue, fever, malaise, or involuntary weight loss.

OR

B. Any other manifestation(s) of Sjögren's syndrome resulting in one of the following:

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or

2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or

3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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