

Medicaid Disability Manual

114.00 Immune System Disorders

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

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

a. The dysfunction may be due to problems in antibody production, impaired cell-mediated immunity, a combined type of antibody/cellular deficiency, impaired phagocytosis, or complement deficiency.

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

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

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

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

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

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

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

C. Definitions

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. “Appropriate” means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.
2. *Constitutional symptoms or signs*, as used in these listings, means severe fatigue, fever, malaise, or involuntary weight loss. *Severe fatigue* means a frequent sense of exhaustion that results in significantly reduced physical activity or mental function. *Malaise* means frequent feelings of illness, bodily discomfort, or lack of well-being that result in significantly reduced physical activity or mental function.
3. *Disseminated* means that a condition is spread over a considerable area. The type and extent of the spread will depend on your specific disease.
4. *Dysfunction* means that one or more of the body regulatory mechanisms are impaired, causing either an excess or deficiency of immunocompetent cells or their products.
5. *Extra-articular* means “other than the joints”; for example, an organ(s) such as the heart, lungs, kidneys, or skin.
6. *Inability to ambulate effectively* has the same meaning as in 101.00B2b.
7. *Inability to perform fine and gross movements effectively* has the same meaning as in 101.00B2c.
8. *Major peripheral joints* has the same meaning as in 101.00F.
9. *Persistent* means that a sign(s) or symptom(s) has continued over time. The precise meaning will depend on the specific immune system disorder, the usual course of the disorder, and the other circumstances of your clinical course.
10. *Recurrent* means that a condition that previously responded adequately to an appropriate course of treatment returns after a period of remission or regression. The precise meaning, such as the extent of response or remission and the time periods involved, will depend on the specific disease or condition you have, the body system affected, the usual course of the disorder and its treatment, and the other facts of your particular case.

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

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

D. How do we document and evaluate the listed autoimmune disorders?

1. *Systemic lupus erythematosus (114.02).*

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

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

2. *Systemic vasculitis (114.03).*

a. *General.*

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

(ii) Children can develop the vasculitis of Kawasaki disease, of which the most serious manifestation is formation of coronary artery aneurysms and related complications. We evaluate heart problems related to Kawasaki disease under the criteria in the cardiovascular listings (104.00). Children can also develop the vasculitis of anaphylactoid purpura (Henoch-Schoenlein purpura), which may cause intestinal and renal disorders. We evaluate intestinal and renal disorders related to vasculitis of anaphylactoid purpura under the criteria in the digestive

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(105.00) or genitourinary (106.00) listings. Other clinical patterns include, but are not limited to, polyarteritis nodosa, Takayasu's arteritis (aortic arch arteritis), and Wegener's granulomatosis.

b. *Documentation of systemic vasculitis.* Angiography or tissue biopsy confirms a diagnosis of systemic vasculitis when the disease is suspected clinically. When you have had angiography or tissue biopsy for systemic vasculitis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase angiography or tissue biopsy.

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

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

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

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

(i) Localized scleroderma (linear scleroderma and morphea) is more common in children than systemic scleroderma. To assess the severity of the impairment, we need a description of the extent of involvement of linear scleroderma and the location of the lesions. For example, linear scleroderma involving the arm but not crossing any joints is not as functionally limiting as sclerodactyly (scleroderma localized to the fingers). Linear scleroderma of a lower extremity involving skin thickening and atrophy of underlying muscle or bone can result in contractures and leg length discrepancy. In such cases, we may evaluate your impairment under the musculoskeletal listings (101.00).

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

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

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

4. *Polymyositis and dermatomyositis (114.05)*.

a. *General*.

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

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

b. *Documentation of polymyositis and dermatomyositis*. Generally, but not always, polymyositis is associated with elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, and aldolase), and characteristic abnormalities on electromyography and muscle biopsy. In children, the diagnosis of dermatomyositis is supported largely by medical history, findings on physical examination that include the characteristic skin findings, and elevated serum muscle enzymes. Muscle inflammation or vasculitis depicted on MRI is additional evidence supporting the diagnosis of childhood dermatomyositis. When you have had electromyography, muscle biopsy, or MRI for polymyositis or dermatomyositis, we will make every reasonable effort to obtain reports of the results of that procedure. However, we will not purchase electromyography, muscle biopsy, or MRI.

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

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

(ii) If you are of preschool age through adolescence (age 3 to attainment of age 18), weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to ambulate effectively. Weakness of your shoulder girdle muscles may result in your inability to perform

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lifting, carrying, and reaching overhead, and also may seriously affect your ability to perform activities requiring fine movements. We evaluate these limitations under 114.05A.

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

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

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

6. *Inflammatory arthritis (114.09).*

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

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

- (i) Reactive arthropathies;
- (ii) Juvenile ankylosing spondylitis;
- (iii) Psoriatic arthritis;
- (iv) SEA syndrome (seronegative enthesopathy arthropathy syndrome);
- (v) Behçet's disease; and

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(vi) Inflammatory bowel disease

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

(i) Juvenile rheumatoid arthritis;

(ii) Sjögren's syndrome;

(iii) Psoriatic arthritis;

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

(v) Lyme disease; and

(vi) Inflammatory bowel disease.

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

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

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

(ii) Listing-level severity is shown in 114.09B, 114.09C2, and 114.09D by inflammatory arthritis that involves various combinations of complications of one or more major peripheral joints or involves other joints, such as inflammation or deformity, extra-articular features, repeated manifestations, and constitutional symptoms and signs. Extra-articular impairments may also meet listings in other body systems.

(iii) Extra-articular features of inflammatory arthritis may involve any body system; for example: Musculoskeletal (heel enthesopathy), ophthalmologic (iridocyclitis, keratoconjunctivitis sicca, uveitis), pulmonary (pleuritis, pulmonary fibrosis or nodules, restrictive lung disease), cardiovascular (aortic valve insufficiency, arrhythmias, coronary arteritis, myocarditis, pericarditis, Raynaud's phenomenon, systemic vasculitis), renal (amyloidosis of the kidney), hematologic (chronic anemia, thrombocytopenia), neurologic (peripheral neuropathy,

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radiculopathy, spinal cord or cauda equina compression with sensory and motor loss), mental (cognitive dysfunction, poor memory), and immune system (Felty's syndrome (hypersplenism with compromised immune competence)).

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

7. *Sjögren's syndrome (114.10).*

a. *General.*

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

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

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

E. How do we document and evaluate immune deficiency disorders, excluding HIV infection?

1. *General.*

a. Immune deficiency disorders can be classified as:

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

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

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

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

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

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

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

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

(ii) Immunosuppressant therapy, such as frequent infections.

(iii) Significant deterioration of other organ systems.

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

a. The frequency and severity of infections.

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

c. Significant deterioration of other organ systems.

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

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

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reports of the results of that testing. However, we will not purchase laboratory testing to establish whether you have HIV infection.

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

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

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

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

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

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

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

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

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

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

(ii) For newborn and younger infants (birth to attainment of age 1), a CD4 (T4) count of 1500/mm³ or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iii) For older infants and toddlers from 12 to 18 months of age, a CD4 (T4) count of 750/mm³ or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

(iv) An abnormal CD4/CD8 ratio.

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

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

2. *CD4 tests.* Children who have HIV infection or other disorders of the immune system may have tests showing a reduction of either the absolute count or the percentage of their T-helper lymphocytes (CD4 cells). The extent of immune suppression correlates with the level or rate of decline of the CD4 count (relative to the age of the young child). By age 6, children have CD4 counts comparable to those levels found in adults. Generally, in these children when the CD4 count is below 200/mm³ (or below 14 percent of the total lymphocyte count) the susceptibility to opportunistic infection is greatly increased. Although a reduced CD4 count alone does not establish a definitive diagnosis of HIV infection, a CD4 count below 200 does offer supportive evidence when there are clinical findings, but not a definitive diagnosis of an opportunistic infection(s). However, a reduced CD4 count *alone* does not document the severity or functional consequences of HIV infection.

3. *Documentation of the manifestations of HIV infection.* The medical evidence must also include documentation of the manifestations of HIV infection. Documentation may be by laboratory evidence or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

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

b. *Other acceptable documentation of the manifestations of HIV infection.* We may also document manifestations of HIV infection without the definitive laboratory evidence described in 114.00F3a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. For example, many conditions are now commonly diagnosed based on some or all of the following:

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Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing. The following are examples of how we may document manifestations of HIV infection with other appropriate evidence.

(i) Although a definitive diagnosis of PCP requires identifying the organism in bronchial washings, induced sputum, or lung biopsy, these tests are frequently bypassed if PCP can be diagnosed presumptively. Supportive evidence may include: Fever, dyspnea, hypoxia, CD4 count below 200 in children 6 years of age or older, and no evidence of bacterial pneumonia. Also supportive are bilateral lung interstitial infiltrates on x-ray, a typical pattern on CAT scan, or a gallium scan positive for pulmonary uptake. Response to anti-PCP therapy usually requires 5-7 days, and such a response can be supportive of the diagnosis.

(ii) Documentation of *Cytomegalovirus* (CMV) disease (114.08D) may present special problems because definitive diagnosis (except for chorioretinitis, which may be diagnosed by an ophthalmologist or optometrist on funduscopic examination) requires identification of viral inclusion bodies or a positive culture from the affected organ and the absence of any other infectious agent likely to be causing the disease. A positive serology test does not establish a definitive diagnosis of CMV disease, but does offer supportive evidence of a presumptive diagnosis of CMV disease. Other clinical findings that support a presumptive diagnosis of CMV may include: Fever, urinary culture positive for CMV, and CD4 count below 200 in children 6 years of age or older. A clear response to anti-CMV therapy also supports a diagnosis.

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

(iv) Candidiasis of the esophagus (also known as *Candida* esophagitis) may be presumptively diagnosed based on symptoms of retrosternal pain on swallowing (odynophagia) and either oropharyngeal thrush (white patches or plaques) diagnosed on physical examination or by microscopic documentation of *Candida* fungal elements from a noncultured specimen scraped from the oral mucosa. Treatment with oral (systemic) antifungal agents usually produces improvement after 5 or more days of therapy, and such a response can be supportive of the diagnosis.

4. *HIV infection manifestations specific to children.*

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

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developmental abnormalities. Adolescents may also exhibit neurological abnormalities, such as HIV encephalopathy, or have growth problems. HIV infection that affects the digestive system and results in malnutrition also may be evaluated under 105.08.

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

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

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

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

- a. The effects of medications you take.
- b. Adverse side effects (acute and chronic).
- c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).
- d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).
- e. Variability of your response to treatment (see 114.00G2).

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

g. The duration of your treatment.

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

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

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

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

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

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

b. *Structured treatment interruptions.* A structured treatment interruption (STI, also called a “drug holiday”) is a treatment practice during which your treating source advises you to stop taking your medications temporarily. An STI in itself does not imply that your medical condition has improved; nor does it imply that you are noncompliant with your treatment because you are following your treating source’s advice. Therefore, if you have stopped taking medication because your treating source prescribed or recommended an STI, we will not find that you are failing to follow treatment or draw inferences about the severity of your impairment on this fact alone. We will consider why your treating source has prescribed or recommended an STI and all the other information in your case record when we determine the severity of your impairment.

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

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

Your symptoms, including pain, severe fatigue, and malaise, may be important factors in our determination whether your immune system disorder(s) meets or medically equals a listing or in our determination whether you otherwise have marked and severe functional limitations. In order for us to consider your symptoms, you must have medical signs or laboratory findings showing the existence of a medically determinable impairment(s) that could reasonably be expected to produce the symptoms. If you have such an impairment(s), we will evaluate the intensity, persistence, and functional effects of your symptoms using the rules throughout 114.00 and in our other regulations. See §§416.928, and 416.929. Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any

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inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

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

1. The following listings in this body system include standards for evaluating the functional limitations resulting from immune system disorders: 114.02B, for systemic lupus erythematosus; 114.03B, for systemic vasculitis; 114.04D, for systemic sclerosis (scleroderma); 114.05E, for polymyositis and dermatomyositis; 114.06B, for undifferentiated and mixed connective tissue disease; 114.07C, for immune deficiency disorders, excluding HIV infection; 114.08L, for HIV infection; 114.09D, for inflammatory arthritis; and 114.10B, for Sjögren's syndrome.

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

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

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

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

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

a. Growth impairment under 100.00.

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- b. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 101.00.
 - c. Ocular involvement, such as dry eye, under 102.00.
 - d. Respiratory impairments, such as pleuritis, under 103.00.
 - e. Cardiovascular impairments, such as cardiomyopathy, under 104.00.
 - f. Digestive impairments, such as hepatitis (including hepatitis C) or weight loss as a result of HIV infection that affects the digestive system, under 105.00.
 - g. Genitourinary impairments, such as nephropathy, under 106.00.
 - h. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 107.00.
 - i. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 108.00.
 - j. Neurologic impairments, such as neuropathy or seizures, under 111.00.
 - k. Mental disorders, such as depression, anxiety, or cognitive deficits, under 112.00.
 - l. Allergic disorders, such as asthma or atopic dermatitis, under 103.00 or 108.00 or under the criteria in another affected body system.
 - m. Syphilis or neurosyphilis under the criteria for the affected body system, for example, 102.00 Special senses and speech, 104.00 Cardiovascular system, or 111.00 Neurological.
3. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §416.926.) If it does not, we will also consider whether you have an impairment(s) that functionally equals the listings. (See §416.926a.) We use the rules in §416.994a when we decide whether you continue to be disabled.

114.01 Category of Impairments, Immune System Disorders

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

- A. Involvement of two or more organs/body systems, with:
 - 1. One of the organs/body systems involved to at least a moderate level of severity; and
 - 2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

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OR

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

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or
2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or
3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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

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

1. One of the organs/body systems involved to at least a moderate level of severity; and
2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

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

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or
2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or
3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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

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

1. One of the organs/body systems involved to at least a moderate level of severity; and
2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

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B. With one of the following:

1. Toe contractures or fixed deformity of one or both feet, resulting in the inability to ambulate effectively as defined in 114.00C6; or
2. Finger contractures or fixed deformity in both hands, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7; or
3. Atrophy with irreversible damage in one or both lower extremities, resulting in the inability to ambulate effectively as defined in 114.00C6; or
4. Atrophy with irreversible damage in both upper extremities, resulting in the inability to perform fine and gross movements effectively as defined in 114.00C7.

OR

C. Raynaud's phenomenon, characterized by:

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

OR

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

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or
2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or
3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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

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

OR

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

OR

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

OR

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

OR

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

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

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

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

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

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

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

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

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

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

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

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

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

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

1. Sepsis; or
2. Meningitis; or
3. Pneumonia; or
4. Septic arthritis; or
5. Endocarditis; or
6. Sinusitis documented by appropriate medically acceptable imaging.

OR

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

OR

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

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or
2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or
3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

114.08 Human immunodeficiency virus (HIV) infection. With documentation as described in 114.00F and one of the following:

A. Bacterial infections:

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1. Mycobacterial infection (for example, caused by *M. avium-intracellulare*, *M. kansasii*, or *M. tuberculosis*) at a site other than the lungs, skin, or cervical or hilar lymph nodes, or pulmonary tuberculosis resistant to treatment; or
2. Nocardiosis; or
3. *Salmonella* bacteremia, recurrent non-typhoid; or
4. In a child less than 13 years of age, multiple or recurrent pyogenic bacterial infections (sepsis, pneumonia, meningitis, bone or joint infection, or abscess of an internal organ or body cavity, but not otitis media or superficial skin or mucosal abscesses) occurring two or more times in 2 years (for children age 13 and older, see 114.00F4c); or
5. Multiple or recurrent bacterial infections, including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment three or more times in a 12-month period.

OR

B. Fungal infections:

1. Aspergillosis; or
2. Candidiasis involving the esophagus, trachea, bronchi, or lungs, or at a site other than the skin, urinary tract, intestinal tract, or oral or vulvovaginal mucous membranes; or
3. Coccidioidomycosis, at a site other than the lungs or lymph nodes; or
4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis); or
5. Histoplasmosis, at a site other than the lungs or lymph nodes; or
6. Mucormycosis; or
7. *Pneumocystis* pneumonia or extrapulmonary *Pneumocystis* infection.

OR

C. Protozoan or helminthic infections:

1. Cryptosporidiosis, isosporiasis, or microsporidiosis, with diarrhea lasting for 1 month or longer; or
2. Strongyloidiasis, extra-intestinal; or
3. Toxoplasmosis of an organ other than the liver, spleen, or lymph nodes.

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OR

D. Viral infections:

1. *Cytomegalovirus* disease (documented as described in 114.00F3b(ii)) at a site other than the liver, spleen, or lymph nodes; or
2. Herpes simplex virus causing:
 - a. Mucocutaneous infection (for example, oral, genital, perianal) lasting for 1 month or longer; or
 - b. Infection at a site other than the skin or mucous membranes (for example, bronchitis, pneumonitis, esophagitis, or encephalitis); or
 - c. Disseminated infection; or
3. Herpes zoster:
 - a. Disseminated; or
 - b. With multidermatomal eruptions that are resistant to treatment; or
4. Progressive multifocal leukoencephalopathy.

OR

E. Malignant neoplasms:

1. Carcinoma of the cervix, invasive, FIGO stage II and beyond; or
2. Kaposi's sarcoma with:
 - a. Extensive oral lesions; or
 - b. Involvement of the gastrointestinal tract, lungs, or other visceral organs; or
3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease); or
4. Squamous cell carcinoma of the anal canal or anal margin.

OR

- F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above), with extensive fungating or ulcerating lesions not responding to treatment (for example,

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dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal *Candida*, condyloma caused by human *Papillomavirus*, genital ulcerative disease).

OR

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

1. Loss of previously acquired, or marked delay in achieving, developmental milestones or intellectual ability (including the sudden onset of a new learning disability); or
2. Impaired brain growth (acquired microcephaly or brain atrophy--see 114.00F4b); or
3. Progressive motor dysfunction affecting gait and station or fine and gross motor skills.

OR

H. Growth disturbance, with:

1. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall of 15 percentiles from an established growth curve (on standard growth charts) that persists for 2 months or longer; or
2. An involuntary weight loss (or failure to gain weight at an appropriate rate for age) resulting in a fall to below the third percentile from an established growth curve (on standard growth charts) that persists for 2 months or longer; or
3. Involuntary weight loss of 10 percent or more of baseline (computed based on pounds, kilograms, or body mass index (BMI)) that persists for 2 months or longer.

OR

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

OR

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

OR

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

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1. Sepsis; or
2. Meningitis; or
3. Pneumonia; or
4. Septic arthritis; or
5. Endocarditis; or
6. Sinusitis documented by appropriate medically acceptable imaging.

OR

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

1. For children from birth to attainment of age 1, at least one of the criteria in paragraphs A-E of 112.12; or
2. For children age 1 to attainment of age 3, at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or
3. For children age 3 to attainment of age 18, at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

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

A. Persistent inflammation or persistent deformity of:

1. One or more major peripheral weight-bearing joints resulting in the inability to ambulate effectively (as defined in 114.00C6); or
2. One or more major peripheral joints in each upper extremity resulting in the inability to perform fine and gross movements effectively (as defined in 114.00C7).

OR

B. Inflammation or deformity in one or more major peripheral joints with:

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

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2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

C. Ankylosing spondylitis or other spondyloarthropathies, with:

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

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

OR

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

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

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

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

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

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

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

2. At least two of the constitutional symptoms or signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

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

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

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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.