

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 common infections as well as opportunistic infections, cancers, or other conditions listed in 114.11.

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

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laboratory tests. We explain the information we need in more detail in the sections below.

C. Definitions

1. *Appropriate medically acceptable imaging* includes, but is not limited to, angiography, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. “Appropriate” means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

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

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

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

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

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

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

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

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

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

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

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

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

1. *Systemic lupus erythematosus (114.02).*

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

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

2. *Systemic vasculitis (114.03).*

a. *General.*

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

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

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

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

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

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

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

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

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

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

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

4. *Polymyositis and dermatomyositis (114.05).*

a. *General.*

(i) Polymyositis and dermatomyositis are related disorders that are characterized by an inflammatory process in striated muscle, occurring alone or in association with other autoimmune disorders. The most common manifestations are symmetric weakness, and less frequently, pain and tenderness of the proximal

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limb-girdle (shoulder or pelvic) musculature. There may also be involvement of the cervical, cricopharyngeal, esophageal, intercostal, and diaphragmatic muscles.

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

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

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

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

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

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

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

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

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findings of extremely high antibody titers to extractable nuclear antigen (ENA) or ribonucleoprotein (RNP) without high titers of anti-dsDNA or anti-SM antibodies. There are often clinical findings suggestive of SLE or childhood dermatomyositis. Many children later develop features of scleroderma.

6. *Inflammatory arthritis (114.09).*

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

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

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

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

- (i) Juvenile rheumatoid arthritis;
- (ii) Sjögren's syndrome;
- (iii) Psoriatic arthritis;
- (iv) Crystal deposition disorders (gout and pseudogout);
- (v) Lyme disease; and
- (vi) Inflammatory bowel disease.

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

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

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

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

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

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

7. *Sjögren's syndrome (114.10).*

a. *General.*

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

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

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

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

1. *General.*

a. Immune deficiency disorders can be classified as:

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

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

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

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

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

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

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

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- (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 HIV infection?* Any child with HIV infection, including one with a diagnosis of acquired immune deficiency syndrome (AIDS), may be found disabled under 114.11 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.*

a. *Definitive documentation of HIV infection.* We may document a diagnosis of HIV infection by positive findings on one or more of the following definitive laboratory tests:

- (i) HIV antibody screening test (for example, enzyme immunoassay, or EIA), confirmed by a supplemental HIV antibody test such as the Western blot (immunoblot) or immunofluorescence assay, for any child age 18 months or older.
- (ii) HIV nucleic acid (DNA or RNA) detection test (for example, polymerase chain reaction, or PCR).
- (iii) HIV p24 antigen (p24Ag) test, for any child age 1 month or older.
- (iv) Isolation of HIV in viral culture.
- (v) Other tests that are highly specific for detection of HIV and that are consistent with the prevailing state of medical knowledge.

b. We will make every reasonable effort to obtain the results of your laboratory testing. Pursuant to § 416.919f, we will purchase examinations or tests necessary to make a determination in your claim if no other acceptable documentation exists.

c. *Other acceptable documentation of HIV infection.* We may also document HIV infection without definitive laboratory evidence.

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(i) We will accept a persuasive report from a physician that a positive diagnosis of your HIV infection was confirmed by an appropriate laboratory test(s), such as those described in 114.00F1a. To be persuasive, this report must state that you had the appropriate definitive laboratory test(s) for diagnosing your HIV infection and provide the results. The report must also be consistent with the remaining evidence of record.

(ii) We may also document HIV infection by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence, 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, we will accept a diagnosis of HIV infection without definitive laboratory evidence of the HIV infection if you have an opportunistic disease that is predictive of a defect in cell-mediated immunity (for example, toxoplasmosis of the brain or Pneumocystis pneumonia (PCP)), and there is no other known cause of diminished resistance to that disease (for example, long-term steroid treatment or lymphoma). In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

2. Documentation of the manifestations of HIV infection.

a. *Definitive documentation of manifestations of HIV infection.* We may document manifestations of HIV infection by positive findings on definitive laboratory tests, such as culture, microscopic examination of biopsied tissue or other material (for example, bronchial washings), serologic tests, or on other generally acceptable definitive tests consistent with the prevailing state of medical knowledge and clinical practice.

b. We will make every reasonable effort to obtain the results of your laboratory testing. Pursuant to § 416.919f, we will purchase examinations or tests necessary to make a determination of your claim if no other acceptable documentation exists.

c. *Other acceptable documentation of manifestations of HIV infection.* We may also document manifestations of HIV infection without definitive laboratory evidence.

(i) We will accept a persuasive report from a physician that a positive diagnosis of your manifestation of HIV infection was confirmed by an appropriate laboratory test(s). To be persuasive, this report must state that you had the appropriate definitive laboratory test(s) for diagnosing your manifestation of HIV infection and provide the results. The report must also be consistent with the remaining evidence of record.

(ii) We may also document manifestations of HIV infection without the definitive laboratory evidence described in 114.00F2a, provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence in your case record. For example, many conditions are now commonly diagnosed based on some or all of the following: Medical history, clinical manifestations, laboratory findings (including appropriate medically acceptable imaging), and treatment responses. In such cases, we will make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

3. Disorders associated with HIV infection (114.11A-E).

a. *Multicentric Castleman disease (MCD, 114.11A)* affects multiple groups of lymph nodes and

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organs containing lymphoid tissue. This widespread involvement distinguishes MCD from localized (or unicentric) Castleman disease, which affects only a single set of lymph nodes. While not a cancer, MCD is known as a lymphoproliferative disorder. Its clinical presentation and progression is similar to that of lymphoma, and its treatment may include radiation or chemotherapy. We require characteristic findings on microscopic examination of the biopsied lymph nodes or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice to establish the diagnosis. Localized (or unicentric) Castleman disease does not meet or medically equal the criterion in 114.11A, but we may evaluate it under the criteria in 114.11G or 14.11I in part A.

b. *Primary central nervous system lymphoma* (PCNSL, 114.11B) originates in the brain, spinal cord, meninges, or eye. Imaging tests (for example, MRI) of the brain, while not diagnostic, may show a single lesion or multiple lesions in the white matter of the brain. We require characteristic findings on microscopic examination of the cerebral spinal fluid or of the biopsied brain tissue, or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice to establish the diagnosis.

c. *Primary effusion lymphoma* (PEL, 114.11C) is also known as body cavity lymphoma. We require characteristic findings on microscopic examination of the effusion fluid or of the biopsied tissue from the affected internal organ, or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice to establish the diagnosis.

d. *Progressive multifocal leukoencephalopathy* (PML, 114.11D) is a progressive neurological degenerative syndrome caused by the John Cunningham (JC) virus in immunosuppressed children. Clinical findings of PML include clumsiness, progressive weakness, and visual and speech changes. Personality and cognitive changes may also occur. We require appropriate clinical findings, characteristic white matter lesions on MRI, and a positive PCR test for the JC virus in the cerebrospinal fluid to establish the diagnosis. We also accept a positive brain biopsy for JC virus or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice to establish the diagnosis.

e. *Pulmonary Kaposi sarcoma* (Kaposi sarcoma in the lung, 114.11E) is the most serious form of Kaposi sarcoma (KS). Other internal KS tumors (for example, tumors of the gastrointestinal tract) have a more variable prognosis. We require characteristic findings on microscopic examination of the induced sputum, bronchoalveolar lavage washings, or of the biopsied transbronchial tissue, or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice to establish the diagnosis.

4. *CD4 measurement (114.11F)*. To evaluate your HIV infection under 114.11F, we require one measurement of your absolute CD4 count (also known as CD4 count or CD4+ T-helper lymphocyte count) or CD4 percentage for children from birth to attainment of age 5, or one measurement of your absolute CD4 count for children from age 5 to attainment of age 18. These measurements (absolute CD4 count or CD4 percentage) must occur within the period we are considering in connection with your application or continuing disability review. If you have more than one CD4 measurement within this period, we will use your lowest absolute CD4 count or your lowest CD4 percentage.

5. *Complications of HIV infection requiring hospitalization (114.11G)*.

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a. Complications of HIV infection may include infections (common or opportunistic), cancers, and other conditions. Examples of complications that may result in hospitalization include: Depression; diarrhea; immune reconstitution inflammatory syndrome; malnutrition; and PCP and other severe infections.

b. Under 114.11G, we require three hospitalizations within a 12-month period that are at least 30 days apart and that result from a complication(s) of HIV infection. The hospitalizations may be for the same complication or different complications of HIV infection and are not limited to the examples of complications that may result in hospitalization listed in 114.00F5a. All three hospitalizations must occur within the period we are considering in connection with your application or continuing disability review. Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

c. We will use the rules on medical equivalence in § 416.926 to evaluate your HIV infection if you have fewer, but longer, hospitalizations, or more frequent, but shorter, hospitalizations, or if you receive nursing, rehabilitation, or other care in alternative settings.

6. *Neurological manifestations specific to children (114.11H)*. The methods of identifying and evaluating neurological manifestations 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. Neurological manifestations may present in the loss of acquired developmental milestones (developmental regression) in infants and young children or, in the loss of acquired intellectual abilities in school-age children and adolescents. A child may demonstrate loss of intellectual abilities by a decrease in IQ scores, by forgetting information previously learned, by inability to learn new information, or by a sudden onset of a new learning disability. When infants and young children present with serious developmental delays (without regression), we evaluate the child's impairment(s) under 112.00.

7. *Growth failure due to HIV immune suppression (114.11I)*.

a. To evaluate growth failure due to HIV immune suppression, we require documentation of the laboratory values described in 114.11I1 and the growth measurements in 114.11I2 or 114.11I3 within the same consecutive 12-month period. The dates of laboratory findings may be different from the dates of growth measurements.

b. Under 114.11I2 and 114.11I3, we use the appropriate table under 105.08B in the digestive system to determine whether a child's growth is less than the third percentile.

(i) For children from birth to attainment of age 2, we use the weight-for-length table corresponding to the child's gender (Table I or Table II).

(ii) For children from age 2 to attainment of age 18, we use the body mass index (BMI)-for-age corresponding to the child's gender (Table III or Table IV).

(iii) BMI is the ratio of a child's weight to the square of his or her height. We calculate BMI using the formulas in 105.00G2c.

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

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

- a. The effects of medications you take.
- b. Adverse side effects (acute and chronic).
- c. The intrusiveness and complexity of your treatment (for example, the dosing schedule, need for injections).
- d. The effect of treatment on your mental functioning (for example, cognitive changes, mood disturbance).
- e. Variability of your response to treatment (see 114.00G2).
- f. The interactive and cumulative effects of your treatments. For example, many children with immune system disorders receive treatment both for their immune system disorders and for the manifestations of the disorders or co-occurring impairments, such as treatment for HIV infection and hepatitis C. The interactive and cumulative effects of these treatments may be greater than the effects of each treatment considered separately.
- g. The duration of your treatment.
- h. Any other aspects of treatment that may interfere with your ability to function.

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

3. *How we evaluate the effects of treatment for autoimmune disorders on your ability to function.* Some medications may have acute or long-term side effects. When we consider the effects of corticosteroids or other treatments for autoimmune disorders on your ability to function, we

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consider the factors in 114.00G1 and 114.00G2. Long-term corticosteroid treatment can cause ischemic necrosis of bone, posterior subcapsular cataract, impaired growth, weight gain, glucose intolerance, increased susceptibility to infection, and osteopenia that may result in a loss of function. In addition, medications used in the treatment of autoimmune disorders may also have effects on mental functioning, including cognition (for example, memory), concentration, and mood.

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

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

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

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

6. When there is no record of ongoing treatment. If you have not received ongoing treatment or have not had an ongoing relationship with the medical community despite the existence of a severe impairment(s), we will evaluate the medical severity and duration of your immune system disorder on the basis of the current objective medical evidence and other evidence in your case record, taking into consideration your medical history, symptoms, clinical and laboratory findings, and

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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 §§416.921, and 416.929. Additionally, when we assess the credibility of your complaints about your symptoms and their functional effects, we will not draw any inferences from the fact that you do not receive treatment or that you are not following treatment without considering all of the relevant evidence in your case record, including any explanations you provide that may explain why you are not receiving or following treatment.

I. *How do we consider the impact of your immune system disorder on your functioning?*

1. We will consider all relevant information in your case record to determine the full impact of your immune system disorder, including HIV infection, on your ability to function. 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, diarrhea, severe fatigue, or pain, resulting in a limitation of your ability to acquire information, to concentrate, to persevere at a task, to interact with others, to move about, or to cope with stress. You may also have limitations because of your treatment and its side effects (see 114.00G).

2. Important factors we will consider when we evaluate your functioning include, but are not limited to: Your symptoms (see 114.00H), 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 (see 114.00G). See §§416.924a and 416.926a for additional guidance on the factors we consider when we evaluate your functioning.

3. We will use the rules in §§ 416.924a and 416.926a to evaluate your functional limitations and determine whether your impairment functionally equals the listings.

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

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2. Children with immune system disorders, including HIV infection, may manifest signs or symptoms of a mental impairment or of another physical impairment. For example, HIV infection may accelerate the onset of conditions such as diabetes or affect the course of or treatment options for diseases such as cardiovascular disease or hepatitis. We may evaluate these impairments under the affected body system. For example, we will evaluate:

- a. Growth impairment under 100.00.
- b. Musculoskeletal involvement, such as surgical reconstruction of a joint, under 101.00.
- c. Ocular involvement, such as dry eye, under 102.00.
- d. Respiratory impairments, such as pleuritis, under 103.00.
- e. Cardiovascular impairments, such as cardiomyopathy, under 104.00.
- f. Digestive impairments, such as hepatitis (including hepatitis C) or weight loss as a result of HIV infection that affects the digestive system, under 105.00.
- g. Genitourinary impairments, such as nephropathy, under 106.00.
- h. Hematologic abnormalities, such as anemia, granulocytopenia, and thrombocytopenia, under 107.00.
- i. Skin impairments, such as persistent fungal and other infectious skin eruptions, and photosensitivity, under 108.00.
- j. Neurologic impairments, such as neuropathy or seizures, under 111.00.
- k. Mental disorders, such as depression, anxiety, or cognitive deficits, under 112.00.
- l. Allergic disorders, such as asthma or atopic dermatitis, under 103.00 or 108.00 or under the criteria in another affected body system.
- m. Syphilis or neurosyphilis under the criteria for the affected body system, for example, 102.00 Special senses and speech, 104.00 Cardiovascular system, or 111.00 Neurological.

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

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114.01 Category of Impairments, Immune System Disorders

114.02 Systemic lupus erythematosus. As described in 114.00D1. With involvement of two or more organs/body systems, and with:

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

114.03 Systemic vasculitis. As described in 114.00D2.

With involvement of two or more organs/body systems, with:

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

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 and signs (severe fatigue, fever, malaise, or involuntary weight loss).

OR

B. With one of the following:

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

C. Raynaud's phenomenon, characterized by:

1. Gangrene involving at least two extremities;

or

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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.

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

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

OR

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

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

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

114.06 *Undifferentiated and mixed connective tissue disease* as described in 114.00D5. With involvement of two or more organs/body systems, and with:

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

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

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

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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.

114.08 (Reserved)

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 or gross movements effectively (as defined in 114.00C7).

OR

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

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

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

OR

C. Ankylosing spondylitis or other spondyloarthropathies, with:

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

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

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114.10 Sjögren's syndrome. As described in 114.00D7. With involvement of two or more organs/body systems, and with:

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

114.11 Human immunodeficiency virus (HIV) infection. With documentation as described in 114.00F1 and one of the following:

A. Multicentric (not localized or unicentric) Castleman disease affecting multiple groups of lymph nodes or organs containing lymphoid tissue (see 114.00F3a).

OR

B. Primary central nervous system lymphoma (see 114.00F3b).

OR

C. Primary effusion lymphoma (see 114.00F3c).

OR

D. Progressive multifocal leukoencephalopathy (see 114.00F3d).

OR

E. Pulmonary Kaposi sarcoma (see 114.00F3e).

OR

F. Absolute CD4 count or CD4 percentage (see 114.00F4):

1. For children from birth to attainment of age 1, absolute CD4 count of 500 cells/mm³ or less, or CD4 percentage of less than 15 percent; or
2. For children from age 1 to attainment of age 5, absolute CD4 count of 200 cells/mm³ or less, or CD4 percentage of less than 15 percent; or
3. For children from age 5 to attainment of age 18, absolute CD4 count of 50 cells/mm³ or less.

OR

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G. Complication(s) of HIV infection requiring at least three hospitalizations within a 12-month period and at least 30 days apart (see 114.00F5). Each hospitalization must last at least 48 hours, including hours in a hospital emergency department immediately before the hospitalization.

OR

H. A neurological manifestation of HIV infection (for example, HIV encephalopathy or peripheral neuropathy) (see 114.00F6) resulting in one of the following:

1. Loss of previously acquired developmental milestones or intellectual ability (including the sudden onset of a new learning disability), documented on two examinations at least 60 days apart; or
2. Progressive motor dysfunction affecting gait and station or fine and gross motor skills, documented on two examinations at least 60 days apart; or
3. Microcephaly with head circumference that is less than the third percentile for age, documented on two examinations at least 60 days apart; or
4. Brain atrophy, documented by appropriate medically acceptable imaging.

OR

I. Immune suppression and growth failure (see 114.00F7) documented by 1 and 2, or by 1 and 3:

1. CD4 measurement:
 - a. For children from birth to attainment of age 5, CD4 percentage of less than 20 percent; or
 - b. For children from age 5 to attainment of age 18, absolute CD4 count of less than 200 cells/mm³ or CD4 percentage of less than 14 percent; and
2. For children from birth to attainment of age 2, three weight-for-length measurements that are:
 - a. Within a consecutive 12-month period; and
 - b. At least 60 days apart; and
 - c. Less than the third percentile on the appropriate weight-for-length table under 105.08B1; or
3. For children from age 2 to attainment of age 18, three BMI-for-age measurements that are:
 - a. Within a consecutive 12-month period; and
 - b. At least 60 days apart; and
 - c. Less than the third percentile on the appropriate BMI-for-age table under 105.08B2.