CD Part 14

Immune System Disorders

A. Definitions

The following definitions are for words used in this chapter and during the SSA disability process. If you need additional definitions, consult a good medical dictionary, available in most bookstores and libraries. You can also look at online medical dictionaries like the one at www.medlineplus.gov.

Abscess. An infection occurring as a localized collection of pus.

Anemia. Low red blood cell count usually determined by a decrease in the hematocrit.

Angiography. Any technique to produce images of arteries, such as by x-ray or MRI scan. Usually involves injection of contrast material into the artery to make it visible. Also known as *arteriography.*

Ankylosing spondylitis. Inflammatory disease of the spine and its supporting ligaments, as well as of the sacroiliac joints and sometimes other parts of the body.

Ankylosis. When a joint or spine is fixed so that it can't move. Ankylosis may be caused by arthritis that fuses joint bones together. For example, an arthritic bone spur may grow across the space of the knee joint so that the joint cannot move. Ankylosis can also be caused by soft tissue damage around a joint, such as scarring of skin and inflamed fibrosis of ligaments and tendons.

Anorexia. Loss of appetite. Anorexia is a frequent problem in chronic diseases, including cancer.

Anterior and lateral ligaments. Ligaments that run up and down the outside of the spine, helping hold the vertebrae in place.

Anterior neck flexors. Muscles that move the head forward in a standing position or upward in a lying (supine) position.

Antibodies. See *immunoglobulins.*

Antigens. Substances that trigger the body's immune system. Antigens are usually foreign substances such as allergens, bacteria, or viruses, but there are also *autoimmune disorders* in which the immune system mistakenly reacts to its own tissues as if they were antigens.

Antinuclear antibodies (ANA). Abnormal antibodies are found in the bodily fluids of many patients with autoimmune diseases, such as systemic lupus. ANA results are reported by laboratories in degrees of abnormality called titers. A higher titer reading suggests a more severe disease. Although it is possible for positive ANA results to occur in normal people, they would be in the low titer ranges of 1:20 to 1:40, which are generally interpreted as negative results.

Anus. Terminal opening of the gastrointestinal tract, through which feces exits the body.

Apophyseal articulations. Parts of the upper and lower surfaces of vertebrae that attach to the intervertebral discs and anterior and lateral ligaments and run up and down the outside of the spine.

Arteritis. Inflammation of an artery.

Arthralgia. Joint pain. Not the same as arthritis, which is disease affecting a joint. Arthralgia usually accompanies arthritis.

Arthropathy. Joint disease.

Aspiration. Getting food or other foreign materials into the trachea or lungs, as caused by defects in the swallowing mechanism.

Atrophy. To get smaller.

Autoimmune disorders. Disorders in which a person's immune system forms antibodies against their own tissues.

Bacteremia. The presence of bacteria in the blood.

Bacteria. Microscopic plants consisting of a single cell.

Behcet's syndrome. A potentially life-threatening immune disorder that causes painful ulcerations of mucous membranes, such as the mouth and genitalia. Multiple other body systems can also be involved with the inflammation, such as the eye (uveitis), joints (arthritis), colon (colitis), and central nervous system (encephalopathy).

Biopsy. The process of taking a sample of tissue for detailed analysis of various kinds. Biopsy specimens are observed grossly with the eyes, microscopically with a variety of possible tissue stains, and in some cases may undergo specific chemical and DNA analysis.

Bronchi. Larger airways branching from the trachea to the lungs.

Bronchial washings. Samples of fluid created by putting small amounts of salt solution (saline) into a bronchus and then sucking it back out. The saline washes over a suspected abnormal area that may contain cancerous cells, bacteria, fungi, or other evidence of abnormality. The saline contains salt

(NaCl) at normal body concentration so that it does not irritate tissues or damage any material it picks up.

Bronchitis. Inflammation of bronchial airways; may be associated with infection or other sources of irritation such as allergy, smoke, or chemical fumes.

Calcinosis. Abnormal calcium deposits in tissues.

Candidiasis. A common type of fungal infection, frequently referred to as a "yeast infection."

Cardiac arrhythmia. Abnormality in the rate or rhythm of the heart beat.

Cardiomyopathy. Any disease of heart muscle. Many things can cause cardiomyopathies including ischemia, viruses, drugs, alcohol, and autoimmune disorders like systemic lupus erythematosus (SLE).

Cell-mediated immunity. Immune system functions carried out by cells, particularly white cells known as T lymphocytes.

Cervical lymph nodes. Lymph nodes in the neck.

Cervical spine. Spine in the neck.

Cervix. The lower part of the uterus, extending into the vagina. PAP smears are done on the cervix.

Chorioretinitis. Inflammation of the choroid and retina of the eye. The choroid is a vascular layer in the wall of the eye that supplies blood to the retina, and carries nerves and blood to the front parts of the eye.

Chronic. Persistent.

Clinical abnormalities. Physical or mental abnormalities that can be directly observed in a patient, in contrast to the study of laboratory abnormalities such as blood tests, x-rays, etc.

Complement. Any of a group of about 30 substances, usually protein enzymes, that can be found in the blood or on the surfaces of cells. Complement reactions are complex. Basically, complement assists in producing an inflammatory response to infections and in destroying viruses and bacteria. Complement can also play a role in immune system disorders.

Computerized axial tomography scan (CAT scan, CT scan). X-rays taken under computer guidance, consisting of many picture slices of high resolution. They show much greater detail than plain x-rays.

Congenital. Dating from the time of birth. Congenital disorders may result from abnormal genes or some abnormality in the intrauterine environment before birth.

Constitutional. Affecting the whole body; not local.

Contracture. When a limb strongly resists movement from a fixed abnormal position as a result of fibrosis or scarring of ligaments, tendons, muscles, or other soft tissues around joints. Contractures of limbs in a bent position are the most common and are known as "flexion contractures."

Corticosteroids. Drugs that have the same action as the natural steroid hormone cortisol. Corticosteroids help combat many diseases, but their side effects can be serious if taken for a long time. Chronic use of steroids also indicates the severity of the disease being treated.

Cricopharyngeal muscles. Muscles deep in the neck and just above the beginning of the esophagus; weakness of these muscles could result in dysphagia.

Cryoglobulinemia. Cryoglobulins in the blood. Cryoglobulins are abnormal immunoglobulins (antibodies) that become insoluble in blood when exposed to slightly lower than normal temperatures. Certain areas of the body are particularly susceptible to cold—the fingertips, earlobes, tip of the nose, the toes, and the cheeks. With exposure to cold, cryoglobulins may gel in the blood stream, so that blood flow slows with resultant symptoms and physical abnormalities in the area affected. Symptoms disappear with warming of the body part so that the abnormal immunoglobulin dissolves again in the blood.

Culture. Growing of bacteria, viruses, fungi, or cells on an appropriate nutrient medium. Cultures are important in diagnosing the cause of an infection, and in determining the drug sensitivity of various microscopic organisms so that the correct drug can be administered.

Cytology. The study of cells. Cytologic tests may involve various kinds of staining of cells to reveal abnormalities such as cancer, or the extraction of DNA or other cellular materials for study.

Diaphragm. The right and left dome-shaped sheets of muscle that separate the chest and abdominal cavities. Movement of the diaphragm changes the air pressure inside the chest, thereby helping air movement in breathing. When the diaphragms move down, the space inside the chest gets larger and the resulting drop in pressure inside the chest causes air to move into the lungs through the mouth or nose. When the diaphragms move back up, the pressure inside the chest increases and air moves out of the lungs through the mouth or nose.

Digital. Reference to the fingers.

Dilation. Widening.

Dorsolumbar spine. Spine area in the lower part of the chest and upper part of the lower back.

Dysmotility. Abnormal movement, such as abnormal contractions in the muscles of the esophagus.

Dysphagia. Difficulty swallowing.

Eczema. General word for a type of itchy skin inflammation. Erythema develops in the area of skin involved as a result of inflammation, followed by oozing of clear fluid that tends to produce crusting. Small blisters are present (vesiculation), and there may also be scaling and thickening of the skin in advanced cases.

Effusion (of joint). Abnormal collection of fluid in a joint space.

Electromyogram (EMG). Recording of a muscle's electrical response to electrical stimulation. The size (amplitude), number (frequency), and shape of the electrical outputs from stimulated muscles provide important information that can be related to both nerve diseases and muscle diseases.

Encephalopathy. Any physical abnormality of the brain that produces mental abnormalities, especially when the brain as a whole is affected. Encephalopathy may be reversible or irreversible, depending on the cause.

Erosion of bone. Areas of bone loss due to a disease process.

Erythema. Redness of the skin caused by increased blood flow in the small capillary blood vessels. Erythema often accompanies inflammation, because inflammation is associated with the release of substances that dilate blood vessels and increase blood flow.

Erythrocyte sedimentation rate (ESR). Test that measures how quickly red blood cells settle; the faster the settling, the more abnormal the result. An elevated ESR indicates some type of inflammation somewhere in the body; it does not diagnose one particular disease. A normal ESR is about 10 mm/hr or less in men and 20 mm/hr or less in women, depending on the method used by the reporting laboratory.

Esophageal dysmotility. Abnormal contractions of muscles in the esophagus. Esophageal dysmotility is associated with fibrosis of the esophagus, which can lead to narrowing of the esophagus with difficulty eating and maintaining adequate nutrition. Pain may also accompany esophageal contractions.

Esophagitis. Inflammation of the esophagus. The most common cause of esophagitis is gastroesophageal reflux.

Extra-articular. Any location outside of the joints, as in the lung, eye or heart.

Facet joints. Small joints between vertebrae.

Failure to thrive. A condition in which infants fail to gain weight at a normally expected rate, or they lose weight. Failure to thrive is often caused by nonphysical factors associated with parental care of the infant. However, it may also result from any type of severe physical disease.

Fibrosis. Degenerative process involving the replacement of normal tissue with fiber-like tissue.

FIGO. International Federation of Gynecology and Obstetrics.

Fine movements. Coordinated manipulation with the fingers, such as picking up coins, buttoning a shirt, typing, playing the piano, or handling anything with the fingertips.

Fungating. Lesions that appear as fungus-like growths. Fungating is a descriptive term about the appearance of an abnormality—bulky and fungus-like—and does not necessarily imply the presence of a fungal infection.

Fusion. See *ankylosis*.

Gangrene. The death of soft tissues, associated with a loss of blood supply and possibly followed by bacterial infection. If there is no bacterial infection, the gangrene is said to be dry gangrene.

Giant cell arteritis. A type of vasculitis that can affect any artery. For example, inflammation of head arteries (such as the temporal artery) can result in headache. Decreased blood flow to the optic nerve can impair vision.

Gross movements. Grasping and holding onto fairly large objects with the hand as a whole, such as turning a door knob, lifting a pan, or handling a wrench.

Helminth. A type of parasitic worm.

Helper/suppressor ratio. The number of helper lymphocytes (CD4 lymphocytes) divided by the number of suppressor lymphocytes (CD8 lymphocytes). An abnormally low CD4/CD8 ratio

suggests that CD4 lymphocytes are being destroyed, such as would be expected with aggressive human immunodeficiency virus (HIV) infection.

Hepatitis. Inflammation of the liver. A common cause of hepatitis is alcohol abuse. A number of viruses (known by letters; for example, A, B, C, D, and E) can cause hepatitis, as can toxins and drugs.

Hilar lymph nodes. Nodes situated in the hilum of each lung, a location where arteries, veins, and lymphatic vessels enter the right or left lung.

Histological tests. Tests performed on pieces of biopsied tissue.

Histology. The study of tissues. Tissues are groups of cells with a specialized function.

Humoral immunity. Immune system functions carried out by antibodies. Antibodies are produced by B lymphocytes.

Hypoplasia. Underdevelopment of an organ or other bodily structure.

Immunoglobulins (Ig). Chemicals produced by plasma cells that are part of the body's immune response to antigens. Immunoglobulins perform many specialized functions. The various types of immunoglobulins are G, M, A, D, and E. These are abbreviated as IgG, IgM, IgA, IgD, and IgE. Also known as *antibodies*.

Immunologic. Reference to the immune system.

Inflammation of joints or other tissues. Redness, swelling, pain, warmth, and tenderness. Because skin tones vary, a lack of redness doesn't rule out inflammation if the other findings are present.

Intercostal muscles. The muscles between the ribs, important in controlling the size of the chest in breathing movements.

Iridocyclitis. Inflammation of the eye's iris and ciliary body.

Ischemic ulcers. Areas of dead tissue resulting from poor blood supply. In autoimmune disorders, ischemic ulcers may develop in the fingertips and toes as a result of vasospasm.

Lesion. Abnormality.

Leukopenia. Decreased white cell count in blood.

Leukoplakia. White patches in the mouth that cannot be rubbed off. The patches are caused by abnormal skin cells and represent a precancerous condition. A common cause of leukoplakia is chewing tobacco and is a danger signal requiring medical evaluation. A particular type of leukoplakia, oral hairy leukoplakia, may indicate impending AIDS. Leukoplakia should not be confused with white patches caused by candida fungal infection that may occur on the tongue or elsewhere in the mouth.

Ligaments. Flat, flexible, tough connective tissue that extends between bones and across joints to hold bones in position.

Lordosis. Curvature of the spine normally present to a moderate degree in the lumbar spine and to a mild degree in the cervical spine. The spine looks as if a flexible straight rod had been pushed forward from behind while the bottom remained in place. The lay term is swayback.

Loss of motion (LOM). See *range of motion*.

Lumbar spine. Spine area in the lower back.

Lymph nodes. Specialized collections of cells found in various locations along the system of lymph vessels. Lymph nodes function for the immune system and contain lymphocytes. For example, lymph nodes can trap and destroy bacteria. Lymph nodes also may contain cancerous cells that are being spread through the lymphatic system. Therefore, biopsy of lymph nodes is important in determining whether cancer has metastasized from the original tumor.

Lymphomas. Cancers of the lymph nodes and spleen that result in abnormal lymphocytes. Lymphoma can invade any organ of the body. *Hodgkin's lymphoma* and *non-Hodgkin's lymphoma* are two important classifications.

Magnetic Resonance Imaging (MRI). A method of producing pictures of internal body structures using magnetic fields and radiofrequency fields. MRIs do not utilize x-rays or other radiation.

Malaise. A general feeling of body discomfort and tiredness.

Meninges. Membranes covering the brain and spinal cord. The thickest, outer meningeal membrane is called the dura mater which surgical or other medical reports usually just call the "dura."

Meningitis. Inflammation of the meninges. The word is also used in association with a particular kind of infection of the meninges. For example, cryptococcal meningitis is infection of the meninges with the cryptococcus fungus, viral meningitis means infection with a virus.

Microcephaly. An abnormally small head.

Morphea. A localized form of scleroderma. At first the morphea skin patches may be soft with a violet tint, but over time become harder and attain a yellowish or ivory color. The local lesions of morphea may spread out over the body to produce a more generalized involvement of the skin.

Motility. Motility means movement and is especially used in reference to the esophagus, stomach, and intestines. These organs have specific normal patterns of motility based on contractions of the muscles within them.

Mucocutaneous. Reference to the skin and mucous membranes.

Mucous membranes. The moist membranes covering the inner surfaces of the lips, the inside of the mouth, and the inner surfaces of the female genitals (vulva).

Mycobacterial infections. Mycobacteria are a group of bacteria, including the species causing tuberculosis (TB). Atypical mycobacteria (for example, M. kansasii, M. intracellulare) are nontuberculosis-type bacteria that may also cause infection.

Mycotic infections. Infections caused by fungi.

Myositis. Myositis means inflammation of muscle. *Polymyositis* is a disorder characterized by widespread inflammation of muscle.

Nephropathy. Kidney disease.

Neuropathy. Any disease of peripheral nerves. Peripheral nerves are those connecting the spinal cord to the various organs and tissues of the body. Kidney failure is one possible cause of neuropathy. Neuropathy is best demonstrated by weakness, decreased reflexes, loss of sensation, and decreased nerve conduction velocity (NCV).

Neurosyphilis. Syphilis of the nervous system, particularly infection of the spinal cord and brain.

Opportunistic diseases. Diseases that arise as a result of a weakened immune system, caused by diseases such as AIDS.

Oral hairy leukoplakia. Leukoplakia refers to white patches on one or both sides of the tongue. The leukoplakia may also have a hairy appearance because old skin cells fail to fall off and instead become fine filaments that look like hairs. About 75% of HIV patients with oral hairy leukoplakia later develop AIDS. However, any person with severe suppression of their immune system—such as transplant patients receiving potent immune-suppressing drugs—can develop oral hairy leukoplakia. The leukoplakia itself often produces no symptoms.

Otitis media. Middle ear infection.

Parasite. A plant or animal that lives off another plant or animal. Parasites can be microscopic or as large as a worm.

Pelvic inflammatory disease (PID). Infection involving the female reproductive organs: the ovaries, fallopian tubes, or uterus.

Percentile. Method of comparing something, like height or weight, to normal expected values, in order to decide the probability that it is normal or abnormal. For example, a person with a weight in the 60th percentile is heavier than 60% of other people and lighter than 40% of other people.

Perianal. Near the anus.

Pericardium. The thin, moist, membrane that surrounds the heart.

Peripheral joints. Refers to joints of the limbs, in contrast to those of the spine.

Pneumonitis. Inflammation of lung tissue, whether from infection or inhalation of irritating substances like smoke, dust, or chemical fumes.

Polyarteritis nodosa. Immune disorder of unknown cause producing widespread arterial inflammation. Any organ in the body can potentially be damaged, depending on the location of the arteritis: Most commonly involved are arteries to the kidney, liver, gastrointestinal tract, and heart. If arteries supplying the brain are involved, headaches or even seizures can occur. This disorder progresses slowly in some cases, while in others it is fatal within months. It is most common in middle-aged men. The most frequent symptoms are arthralgia, weakness, and abdominal pain. High blood pressure may be present. Most patients die within a year without treatment from causes like heart failure, bleeding, ruptured aneurysms, kidney failure, or gastrointestinal bleeding. Even with treatment the five-year survival rate is generally poor. Also known as *polyarteritis, periarteritis,* and *periarteritis nodosa*.

Prognosis. The likelihood of recovery from a disorder.

Protozoans. Microscopic, single-celled animals. Some cause disease in humans.

Psoriasis. A chronic hereditary skin disorder usually characterized by white—sometimes pinkish—scaly, raised, and flat lesions. Any part of the skin surface may be involved, but lesions are most common on the elbows and knees.

Range of motion (ROM). How well a joint moves. ROM is extremely important in determining how limiting arthritis is likely to be. For example, a knee joint that has only a small degree of motion will limit the ability to walk and otherwise use the legs much more than a knee joint with a normal or mildly restricted range of motion. A person's ROM may be limited not only by arthritis, but also by loss of flexibility in the soft tissues around the joints. ROM is usually reported in degrees of flexion (bending of a limb or the spine) and extension (straightening a limb), abduction (movement of a limb away from the body in a right or left direction), adduction (movement of a limb toward the body from a right or left position), rotation, etc., depending on the joint involved.

For Social Security disability purposes, all ROM measurements must be passive—meaning measured when you relax and let a doctor move the joint for you. The only exception is the spine, for which you must actively participate in movements. Active range of motion is where you voluntarily move a joint. Measurements of active ROM are considered unreliable because they depend on the applicants to honestly move their joints to the maximum degree when asked. Active ROM tests can lead to serious disagreements between you and the SSA. If you state that you cannot bend, but physical tests or x-rays do not verify your claim, the SSA does not have to believe you. SSA evaluations frequently reveal (through physical examinations, x-rays, and other laboratory tests) that applicants alleging incapacitating arthritis and inability to move joints actually have a good ROM and minimal abnormalities.

Raynaud's phenomenon (disease). Raynaud's phenomenon is a disorder characterized by episodes of arterial vasospasm, especially involving the fingers or toes. Raynaud's phenomenon may exist alone and be fairly harmless. However, it is frequently associated with some immune system disorders. When it is, it is referred to as Raynaud's *disorder,* although some doctors do not make this distinction. Raynaud's phenomenon is usually triggered by cold or emotion. There is a pattern of pale color to the area affected, because blood flow decreases. Then a bluish or purplish coloration appears. As the episode ends and blood flow is restored, the part affected becomes redder.

Reiter's syndrome. An immune system disease classically involving inflammation of the urethra (urethritis), eye (conjunctivitis), and joints (arthritis). However, this classic triad of findings is present in only a minority of people with the disorder. Multiple other organ systems, such as the heart and nervous system, can also be involved. This syndrome is most common in men in the 20 to 40 age range.

Retrovirus. Any virus of the family Retroviridae, such as the human immunodeficiency virus (HIV) that causes AIDS. Retroviruses have a single-stranded ribonucleic acid (RNA) core, but use an enzyme called reverse transcriptase to produce a DNA (deoxyribonucleic acid) copy of their RNA. This viral DNA then integrates itself into the DNA of the host cell that the virus has infected. In the case of the AIDS virus, invasion of host T4 (CD4) lymphocytes leads to cell death with resulting immune deficiency.

Rheumatoid arthritis (RA). A disease of the immune system, particularly associated with swelling, redness, and tenderness of the joints. The small joints of the hands are most susceptible, but any joint in the body can be affected. If not treated adequately, RA can result in extensive bone destruction and deformity. Although best known as a cause of severe arthritis, RA has numerous other potentially harmful effects on the body that can involve the lungs, spleen, heart, pericardium, blood vessels, nerves, or eyes, and it can even produce anemia.

Rheumatoid factor (RF). Certain abnormal antibodies that the body has produced and that are especially associated with rheumatoid arthritis. However, it is possible to have rheumatoid arthritis without testing positive for RF. At the same time, you may test positive even if you don't have arthritis. RF is reported by laboratories as "positive" or "negative," and also as degrees of abnormality called titers. For example, higher titers, such as 1:500 versus 1:50, suggest more severe disease activity.

Sacroiliac joints. Joints between the pelvic bones and the sacrum of the spine.

Sacroiliitis. Inflammation of a sacroiliac joint.

Sclerodactyly. Scleroderma affecting the fingers. Hard, thickened skin may make use of the fingers difficult.

Scleroderma. The fibrotic hardening and thickening of skin caused by the autoimmune disorder that causes progressive systemic sclerosis (PSS). The presence of scleroderma doesn't necessarily mean there is full-blown PSS. However, some doctors use the word "scleroderma" to mean "progressive systemic sclerosis," and this fact should be taken into account when an examiner is considering a claimant's diagnosis.

Sepsis. Severe illness resulting from an infection in tissues or the blood with disease-causing microorganisms, particularly bacteria or fungi. Sepsis can result from the microorganisms themselves or from toxins they produce. *Septicemia* specifically means sepsis involving the blood. However, note that infection of a joint known as *septic arthritis* is not usually associated with sepsis; the effect of the infection may be confined to the joint space.

Serological tests. Blood tests related to immune processes, such as levels of antibodies and antigens in the blood. Serological tests are important in establishing prior infection as well as monitoring ongoing infection in some diseases. Serological tests can apply to bacterial, viral, fungal, or protozoan infections.

Sjogren's syndrome. A disorder characterized by decreased tear and/or saliva production. Decreased tear production results in a drying of the eyes, a condition known as keratoconjunctivitis sicca. The whole complex of abnormalities (such as tooth decay and damage to the tongue) associated with decreased saliva production is called xerostomia. Ninety percent of cases are women. Sjogren's syndrome can be caused by connective tissue diseases, cancers, liver disease, inflammation of the spine (spondylitis), and others.

Spine. Bony vertebrae stacked on top of each other and separated by intervertebral discs that permit some degree of cushioning and flexibility. The seven vertebrae of the neck (C1-C7) are called the cervical spine. The 12 vertebrae in the chest are the thoracic spine (T1-T12), while the five vertebrae in the lower back are known as the lumbar spine (L1-L5). Beneath the lumbar spine is the sacrum, which consists of a triangular piece of bone of sacral vertebrae fused together (S1-S4). At the end of the spinal column is the tailbone (coccyx). The vertebrae forming the spine are overlaid and connected by many spinal muscles and ligaments. They also form small joints between each other called facet joints.

Spondylitis. Inflammation of the spine. The term does not necessarily signify arthropathy.

Spondyloarthropathy. A group of disorders involving the joints of the spine. If the arthropathy also involves an inflammatory process, the resulting disorder is a type of inflammatory spondyloarthropathy.

Subcutaneous nodules. Lumpy abnormalities of tissues beneath the skin that are sometimes associated with rheumatoid arthritis.

Syndrome. A set of signs and symptoms that occur together.

Synovial membranes. Membranes that surround and help lubricate joints; they become inflamed and tender in active rheumatoid arthritis.

Systemic. Affecting the body as a whole.

Takayasu's arteritis (aortic arch syndrome). Vasculitis of unknown cause involving the aorta and some of its major branches. Joint inflammation and pain early in the disease are eventually followed by decreased blood flow to organs and limbs. For example, decreased blood flow to the brain can impair thinking, while poor blood flow to the limbs can cause tissue damage such as ulcerations of the fingers or toes.

Telangiectasia. Lesions characterized by an area of permanent dilation of blood vessels in the skin or mucous membranes. These lesions can be seen with the unaided eye.

Thrombocytopenia. Decreased platelets in blood.

Ulna. Small bone in the forearm between the elbow and wrist, on the same side of the forearm as the little finger.

Ulnar deviation. Deformities of the fingers that severely limit use of the hands, usually found in rheumatoid arthritis. This condition typically results in a sideways pointing of the fingers.

Undifferentiated. Lacking specialization, as when a connective tissue disorder has features of multiple specific disorders.

Valgus. Reference to a deformity in which a body part is bent outward from the midline of the body.

Varus. Reference to a deformity in which a body part is bent inward toward the midline of the body.

Vascular. Reference to blood vessels.

Vasculitis. Vasculitis means inflammation of a blood vessel, which could be an artery or vein. However, the word vasculitis is usually used in reference to arterial inflammation. *Systemic vasculitis* is more serious, since many arteries are involved. *Arteritis* specifically refers to arterial inflammation.

Vasospasm. An abnormal contraction of muscles in the walls of arteries affected, causing those vessels to narrow. Such vasospasm decreases the ability of the artery to provide blood to the tissues it serves.

Viral inclusion bodies. Microscopic clumps of material in cells associated with some types of viral infection, such as cytomegalovirus.

Vulva. The external female genitalia.

Vulvovaginal. Reference the external female genitalia and vagina.

Wegener's granulomatosis. A form of arteritis causing damage to the respiratory system (nose, sinuses, and lungs), as well as the kidneys and heart. However, any organ system can be involved. This is a serious disorder with numerous possible serious effects, including neurological involvement. Arthralgias may be present. The mortality untreated is extremely high. Although about 90% of cases will improve with initial treatment, a third of cases will relapse and about 20% will ultimately die.

Whipple's disease. An immune disorder that predominantly affects males and may involve numerous body systems. The effects of this disorder may include abdominal pain, arthritis, malabsorption of nutrients from the intestine, nervous system abnormalities, anemia, and skin, lung, or heart disease.

Zygoma. Cheekbone.

B. General Information

While most systems in the human body consist of bones, muscles, veins, arteries, and organs, the immune system is made of different types of cells that protect the body from disease.

1. Parts of the Immune System

The disorders listed by the SSA as making a person eligible for disability benefits include deficiencies of one or more parts of the immune system, including:

- antibody-producing B lymphocyte cells
- various types of cells associated with cell-mediated immunity including T lymphocyte cells
- white cells known as macrophages and monocytes, and
- components of the complement system.

2. Dysfunction of the Immune System

Two types of immune system dysfunction that are important to understand are connective tissue disorder (Subsection a, below) and HIV (Subsection b, below).

a. Connective Tissue Disorder

Abnormal function of the immune system may result in the development of a connective tissue disorder. Connective tissue disorders include several chronic disorders affecting multiple organs. These various connective disorders differ in their clinical manifestation, course of illness, and outcome. They generally evolve and persist for months or years, may result in loss of functional abilities, and may require long-term and repeated evaluation and treatment.

The documentation needed to establish the existence of a connective tissue disorder is medical history, physical examination, selected laboratory studies, medically acceptable imaging techniques, and in some instances, tissue biopsy. However, the Social Security Administration will not purchase diagnostic tests or procedures that involve significant risk, such as biopsies or angiograms. Generally, the existing medical evidence will contain this information.

A medical record covering a continuous period of at least three months is necessary for assessment of the severity and duration of an impairment. This medical record must be "clinical," meaning involving actual visits to a treating doctor for examination. The records should also show active disease despite prescribed treatment during this three-month period with the expectation that the disease will remain active for 12 months.

To permit the accurate application of a listing, specific diagnostic findings should be documented in the clinical record for each of the connective tissue disorders. These diagnostic findings must be present for systemic lupus erythematosus (SLE), systemic vasculitis, systemic sclerosis and scleroderma, polymyositis or dermatomyositis, and undifferentiated connective tissue disorders. See the comments under each listing for a discussion of the required diagnostic findings.

In addition to the limitations caused by a connective tissue disorder itself, any harmful side effects of treatment must be considered. For example, long-term use of corticosteroids such as prednisone may result in the deterioration of bones, which, in turn, can result in functional loss.

Connective tissue disorders may prevent performance of any gainful work activity because of severe loss of function in a single organ or body system or lesser degrees of functional loss in two or more organs or body systems. Such functional loss may also result from fatigue, fever, malaise, and weight loss.

The SSA uses the term "severe" in connective disorder listings to describe medical severity in the common meaning of the word. The word "severe" does not have the same meaning as it does in some other federal regulations the SSA uses, where it simply means more than slight or mild.

Allergic disorders, such as asthma or allergic skin disorders like atopic dermatitis, are discussed and evaluated under the appropriate listing of the affected body system, such as those listings dealing with skin disorders (CD Part 8).

b. Human Immunodeficiency Virus (HIV) Infection

Infections with the human immunodeficiency virus (HIV) continue as a worldwide problem. HIV started in Africa, and was acquired by humans in the form of a mutated simian immunodeficiency virus (SIV) from monkeys or apes. In other words, there was somehow a cross-species transmission of the virus. Although in this country the original infection was concentrated in the male homosexual population, HIV infection is now a serious risk of unprotected sexual intercourse in all groups of people regardless of their sexual orientation. Transmission occurs through microscopic abrasions on contact surfaces during sexual intercourse.

HIV infection can also take place through blood transfusions or any other open place where infected blood can enter the body. Medical personnel have been infected by exposure to HIV-infected blood through small cuts or damaged skin where the virus could enter. An accidental needle stick with a syringe used to draw HIV-infected blood is very dangerous because it injects virus into the tissues where it can enter the bloodstream. People receiving blood transfusions are at small risk, because the blood supplies in the United States are carefully checked for HIV infection. However, it is important that authorities keep up with detection of new HIV strains that may appear (see below). Other viruses, like hepatitis, can also be transmitted by accidental needle sticks. In general, it is a good idea to treat all exposure to blood as very dangerous, even from people who are thought to be healthy. Babies can be infected with HIV from their mothers by breast-feeding.

Although HIV infection may lead to AIDS, it is not, by itself, the same as having the acquired immunodeficiency syndrome (AIDS). Infections with HIV may produce no symptoms, and there are thought to be several million people in the United States who do not know they are infected. They are, however, capable of infecting others. Claimants with no significant symptoms or other abnormalities other than HIV infection are not considered disabled by the SSA, since they have no functional restrictions that would prevent work. This is an important point, because claimants with HIV infection alone frequently apply for disability benefits and erroneously allege that they have AIDS.

The SSA should allow all cases of accurately diagnosed AIDS; some cases of HIV infection without AIDS are allowable if their condition is severe enough, even though they don't officially qualify for a diagnosis of AIDS. This is because it is the claimant's medical condition, rather than a diagnosis, that the SSA evaluates. See Listing 14.08 and accompanying comments.

The human immunodeficiency virus damages the immune system by destroying CD4 (T4) lymphocytes. When HIV infection damages the immune system enough, there is risk of developing various cancers, as well as parasitic, viral, bacterial, fungal, and protozoan infections. Additional infections resulting

from HIV infection are known as opportunistic infections, because it is the "opportunity" provided by a weakened immune system that allows them to do their damage. When these severe additional infections or cancer appear in people with HIV infection, AIDS is then the proper diagnosis and marked functional limitations may be present. Weight loss and malnutrition that may also be a problem in AIDS, is known as wasting syndrome. Sexually transmitted diseases (STDs), such as syphilis and gonorrhea, are also much more common in people with HIV infection and can cause additional difficulties that must be taken into account during disability determinations.

HIV-1 is the technical name for the type of HIV that is causing AIDS in most of the world. Unless otherwise stated, reference to HIV in the listings and discussion means HIV-1. Another form of HIV—HIV-2—can also cause AIDS, but it is confined mostly to West Africa. HIV-2 progresses more slowly than HIV-1, but could potentially qualify under the listing if such a case were encountered.

HIV is a type of retrovirus, which multiplies by taking over the genetic material of the host cells it infects. The host cells, such as CD4 lymphocytes, are killed in the process. HIV can also lie dormant in various kinds of host cells they have not killed, so that even if the virus is completely wiped out of the bloodstream, a recurrence of infection can occur if medication is stopped. Once a person is infected with HIV, it will probably not be possible to ever completely eliminate it from the body.

HIV-1 has been thought to consist of two major groups of viruses that can infect humans and cause AIDS: the M ("Majority") group and the O ("Outlier") group. There are nine subtypes of Group M (A through I). However, a newer virus, found in 1995 in Africa, called YBF30, has been designated as a new group N. Conventional HIV tests, including those done on blood supplies, do not detect YBF30. The spread of YBF30 has been limited, but that may not remain the case.

HIV is the most serious infectious agent facing the world today. About 80% of those who are infected with HIV are known as typical progressors with a median survival of ten years. About 10–15% of people with HIV infection are "rapid progressors," with a median survival of only two or three years. About 5–10% of those infected are "nonprogressors," who remain without symptoms for as long as ten years.

Specific requirements for disability based on HIV infection can be found in Listing 14.08 for adults, or Listing 114.08 for children. Additional comments may also be found there.

c. Inflammatory Arthritis

Inflammatory arthritis isn't simply one disease—it is associated with a number of disorders, whose cause, nature, and severity of flare-ups and outcome may differ dramatically. Depending on the type of inflammatory arthritis, there may be involvement of the peripheral joints, the spine, or both. Spinal involvement is found especially in cases caused by psoriasis, ankylosing spondylitis, Reiter's syndrome, Behcet's disease, Whipple's disease, or by unknown factors (undifferentiated spondylitis). Inflammation of the soft tissues around joints is usually evaluated under SSA Listing 14.09 (adults) or 114.09 (children). To the extent bony joint damage with deformity is present as a result of inflammation, SSA evaluation would be done under Listing 1.02 or 1.03 (adults). Similarly, the SSA would evaluate deformity under Listing 101.02 or 101.03 (children).

The abnormalities that sometimes accompany the various disorders causing inflammatory arthritis may be found in numerous body systems other than the joints. For example, these disorders may include:

- inflammation of the eye
- inflammation of the membranes lining the chest and covering the lungs (pleuritis)
- fibrosis of the lungs
- inflammation of the heart muscle (myocarditis) or pericardium (pericarditis)
- abnormal heart rhythms (cardiac arrhythmias)
- damage to heart valves
- inflammation of arteries supplying the heart (coronary arteritis)
- Raynaud's phenomena
- inflammation of arteries generally (systemic vasculitis)
- amyloidosis of the kidney
- chronic anemia
- decreased blood platelets (thrombocytopenia)

- neuropathy, or other neurological abnormalities, or
- inflammation of heel ligaments (heel enthesopathy).

In children, inflammation of joints and other organs may affect growth, development, attainment of age-appropriate skills, and performance of age-appropriate activities. If the child has growth impairments, the SSA would evaluate these under Listings 100.01 and 100.02.

The SSA cannot find you disabled simply because treatment of your arthritis requires the chronic use of steroid drugs. Each case must be evaluated on its own merits, taking into consideration the severity of the underlying impairment and any other adverse consequences of treatment, such as steroid drug side effects.

C. Specific Listings and Residual Functional Capacity

The listings that follow are from the federal regulations. They have been interpreted and commented on for greater ease of understanding while explaining their requirements. It is impossible to discuss here all of the medical possibilities related to every kind of disorder, and you may need to seek help from your treating doctor to more fully understand how your particular impairment relates to these listings. The discussions of residual functional capacity do not apply to children.

1. Listing 14.02: Systemic Lupus Erythematosus (Adults)

Systemic lupus erythematosus (SLE) is an autoimmune disorder, which means the body's immune system reacts against healthy tissues. Almost any part of the body can be involved, including the nervous system, kidneys, intestine, skin, muscles, joints, eyes, lungs, bone marrow, or heart. That is why SLE is referred to as a multisystem disorder. SLE may show periods of increased severity (exacerbations) involving a particular organ, followed by some degree of improvement and involvement of some other organ. Or, multiple organs may be involved in different degrees of severity at the same time. SLE is caused by genetic abnormalities that are more common in females and people of Asian heritage.

The damage and symptoms caused by SLE vary greatly. Some people have mild disease controllable with medication and few symptoms, while others have rapidly progressive disease despite treatment. A particularly ominous problem is decreased kidney function related to SLE damage.

SLE may produce constitutional symptoms and signs such as fever, fatigability, malaise, and weight loss. Other possible problems include anemia, a decreased white cell count (leukopenia), or a decreased platelet count (thrombocytopenia). A variety of circulating serum autoantibodies directed against the patient's own tissues can occur, but are highly variable in pattern.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must be diagnosed as suffering from systemic lupus erythematosus. Additionally, you must satisfy part Ⓐ or Ⓑ.

Ⓐ One of the following:

- Joint involvement, as described under the criteria in listings with 1 as the prefix (CD Part 1)
- Muscle involvement, as described under the criteria in Listing 14.05, below
- Eye involvement, as described under the criteria in listings with 2 as the prefix (CD Part 2)
- Respiratory involvement, as described under the criteria in listings with 3 as the prefix (CD Part 3)
- Heart or blood vessel involvement, as described under the criteria in listings with 4 as the prefix (CD Part 4 or 14.04Ⓓ, below)
- Digestive involvement, as described under the criteria in listings with 5 as the prefix (CD Part 5)
- Kidney involvement, as described under the criteria in listings with 6 as the prefix (CD Part 6)
- Blood involvement, as described under the criteria in listings with 7 as the prefix (CD Part 7)
- Skin involvement, as described under the criteria in listings with 8 as the prefix (CD Part 8)

- Nervous system involvement, as described under the criteria in listings with 11 as the prefix (CD Part 11), or
- Mental involvement, as described under the criteria in listings with 12 as the prefix (CD Part 12).

Ⓑ Significant (more than slight) damage to at least one of the organs or body systems listed in part Ⓐ, and at least slight damage to another. For example, you could have significantly painful hand joints and a minor inflammation of the eye. Additionally, you must have significant constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss documented in your medical records. Significant constitutional symptoms and signs would be those sufficiently severe to have more than a slight effect in your ability to function in your daily activities or a work environment. Medical judgment on a case-by-case basis is required to make the above determinations.

b. Residual Functional Capacity

Since SLE can affect so many different areas of the body, each case must be evaluated on an individual basis. SLE can cause feelings of weakness and fatigue that are not obvious on physical examination. Also, lupus often causes the skin to be hypersensitive to sunlight. A claimant with this should not be given an outdoor job. There is a disorder affecting only the skin, called discoid lupus. This is not as serious as systemic lupus, but should still receive restrictions on exposure to excessive sunlight. Reference should also be made to comments about RFC under whatever body system and specific listings are most relevant to the claimant's problems. For example, eye involvement would be evaluated under the listings described in CD Part 2 and whatever RFC discussions are appropriate to the specific impairment involved. As another example, SLE can result in abnormalities of the blood, including anemia, thrombocytopenia, and leukopenia. Anemia can lead to decreased exercise tolerance and weakness; thrombocytopenia can lead to an excessive tendency to bleed; and leukopenia can produce susceptibility to infection. Various RFC possibilities related to these blood disorders can be found in CD Part 7.

2. Listing 114.02: Systemic Lupus Erythematosus (Children)

See the comments under adult Listing 14.02.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of systemic lupus erythematosus. Additionally, the child must satisfy part Ⓐ or part Ⓑ, below.

Ⓐ One of the following:

- Growth impairment, as described under the criteria in listings with 100 as the prefix (CD Part 1)
- Musculoskeletal involvement, as described under the criteria in listings with 101 as the prefix (CD Part 1)
- Muscle involvement, as described under the criteria in Listing 14.05, below
- Eye involvement, as described under the criteria in listings with 102 as the prefix (CD Part 2)
- Respiratory involvement, as described under the criteria in listings with 103 as the prefix (CD Part 3)
- Heart or blood vessel (cardiovascular) involvement, as described under the criteria in listings with 104 as the prefix (CD Part 4) or 14.04Ⓓ, below
- Digestive involvement, as described under the criteria in listings with 105 as the prefix (CD Part 5)
- Kidney involvement, as described under the criteria in listings with 106 as the prefix (CD Part 6)
- Blood involvement, as described under the criteria in listings with 107 as the prefix (CD Part 7)
- Skin involvement, as described under the criteria in listings with 8 as the prefix (CD Part 8)
- Hormone (endocrine) involvement, as described under the criteria in listings with 109 as the prefix (CD Part 9)
- Nervous system involvement, as described under the criteria in listings with 111 as the prefix (CD Part 11), or
- Mental involvement, as described under the criteria in listings with 112 as the prefix (CD Part 12).

Ⓑ Significant (more than slight) damage to at least one of the organs or body systems listed in part Ⓐ, and at least slight damage to another. For example, the child could have significant anemia and a minor growth impairment. Additionally, the child must have significant constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss documented in his or her medical records. Significant constitutional symptoms and signs would be those sufficiently severe to have more than a slight effect in the child's ability to function in age-appropriate daily activities. Medical judgment on a case-by-case basis is required to make the above determinations.

3. Listing 14.03: Systemic Vasculitis (Adults)

Systemic vasculitis means generalized inflammation of the arterial system. It occurs acutely in association with adverse drug reactions, certain chronic infections, and, occasionally, cancers. More often it is chronic and of unknown origin. There are several clinical patterns, including (but not limited to) classical polyarteritis nodosa, aortic arch arteritis, giant cell arteritis, Wegener's granulomatosis, and vasculitis associated with other disorders (for example, rheumatoid arthritis, systemic lupus erythematosus (SLE), Sjogren's syndrome, cryoglobulinemia).

The diagnosis is confirmed by angiography or tissue biopsy when the disease is suspected from a clinical evaluation. These studies must be done to satisfy the listing. However, the SSA does not request or perform angiography or biopsies, since these are invasive procedures. Most patients with systemic vasculitis will have the results of the angiogram or biopsy in their medical records.

Vasculitis of the skin may or may not be associated with systemic involvement, and the patterns of vascular involvement are highly variable. Such skin involvement may include decreased blood flow to areas of skin (ischemia).

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must be diagnosed with systemic vasculitis. Additionally, you must satisfy part Ⓐ or Ⓑ, below.

Ⓐ Involvement of a single organ or body system, as described under the criteria in Listing 14.02Ⓐ, above.

Ⓑ Significant (more than slight) damage to at least one of the organs or body systems listed in part Ⓐ, and at least slight damage to another. For example, you could have significant cardiac arrhythmias and minor joint inflammation. Additionally, you must have significant constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss documented in your medical records. Significant constitutional symptoms and signs would be those sufficiently severe to have more than a slight effect on your ability to function in your daily activities or a work environment. Medical judgment on a case-by-case basis is required to make the above determinations.

b. Residual Functional Capacity

You should refer to the comments about RFC under whatever body system and specific listings are most relevant to your physical problems. For example, kidney involvement would be evaluated under the listings described in CD Part 6, including the accompanying RFC discussions.

4. Listing 114.03: Systemic Vasculitis (Children)

This listing has the same requirements as 14.03, above, as well as the additional consideration of growth impairments (CD Part 1).

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must be diagnosed with systemic vasculitis as described in Listing 14.03 or, if there is growth impairment, as described under the criteria in listings with the prefix 100 (CD Part 1).

5. Listing 14.04: Systemic Sclerosis and Scleroderma (Adults)

Systemic sclerosis is an autoimmune disease also known as progressive systemic sclerosis (PSS). The word "progressive" in the acronym PSS does not mean that the disorder must be worsening to qualify

under the listing. In fact, the SSA may have removed the word progressive from its listing to avoid such confusion. Systemic sclerosis is the generalized form of this connective tissue disorder, while scleroderma refers to the hardening and thickening of skin. It is possible to have scleroderma without having systemic sclerosis, but systemic sclerosis frequently includes scleroderma.

There are no diagnostic laboratory tests for scleroderma or systemic sclerosis. This is an incurable disease with no very effective treatment other than supportive care and treatment for secondary disorders such as high blood pressure or heart failure.

The clinical hallmark of systemic sclerosis and scleroderma is thickening of the skin. In addition to skin and blood vessels, the major organs and body systems involvement may include the gastrointestinal tract, lungs, heart, kidneys, and muscle. Although arthritis can occur, abnormal joint function results primarily from thickening of the skin and other soft tissue, as well as fibrosis and contractures. Scleroderma itself, even without systemic sclerosis, can cause degeneration not only of skin, but of underlying soft tissues such as muscle, ligaments, and tendons.

There is a disorder known as the CREST syndrome that may slowly progress to systemic sclerosis over a period of years. CREST stands for **C**alcinosis, **R**aynaud's phenomena, **E**sophageal dysmotility, **S**clerodactyly, and **T**elangiectasia. These disorders are defined at the beginning of this chapter. Refer to these definitions if you see the term CREST in your medical file or other SSA documents.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have a diagnosis of systemic sclerosis and scleroderma. Additionally, you must satisfy part Ⓐ, Ⓑ, Ⓒ, or Ⓓ, below.

Ⓐ One of the following:

- Muscle involvement, as described under the criteria in Listing 14.05.
- Respiratory involvement, as described under the criteria in listings with 3 as the prefix (CD Part 3)
- Heart or blood vessel (cardiovascular) involvement, as described under the criteria in listings with 4 as the prefix (CD Part 4)
- Digestive involvement, as described under the criteria in listings with 5 as the prefix (CD Part 5), or
- Kidney involvement, as described under the criteria in listings with 6 as the prefix (CD Part 6).

Ⓑ Significant (more than slight) damage to at least one of the organs or body systems listed in part Ⓐ, and at least slight damage to another. For example, you could have a a significant problem with scleroderma and a minor degree of lung fibrosis. Additionally, you must have significant constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss documented in your medical records. Significant constitutional symptoms and signs would be those sufficiently severe to have more than a slight effect in your ability to function in your daily activities or a work environment. Medical judgment on a case-by-case basis is required to make the above determinations.

Ⓒ Generalized scleroderma with digital contractures. This means there is a generalized hardening of the skin, as well as inability to move the fingers normally as a result of hardening of the soft tissues around them. There is no requirement that the fingers have to be fixed to such a degree that no movement is possible.

Ⓓ Severe Raynaud's phenomena, characterized by digital ulcerations, ischemia, or gangrene. This part of the listing concerns the complication of decreased blood flow (ischemia) to the fingers as a result of Raynaud's phenomenon. Gangrene may appear as black spots, especially on the fingertips, which is evidence of severe ischemia. If the gangrenous tissue sloughs off, it leaves ulcers.

b. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account residual impairment from systemic sclerosis or scleroderma and the treatment given. Particular attention should be given to the ability to use the hands. Even if gross function is intact, fine (small) movements with the fingers may be affected and limit the kinds of work that you can perform. Reference should also be made to comments about RFC under whatever body system and specific listings

are most relevant to your problems. For example, heart involvement would be evaluated under the listings described in CD Part 4 and the accompanying RFC discussions.

6. Listing 114.04: Systemic Sclerosis and Scleroderma (Children)

See comments under adult Listing 14.04, above, with the additional consideration of possible growth impairments, which would be evaluated under the 100-prefix listings (CD Part 1).

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of systemic sclerosis and scleroderma. Additionally, the child must satisfy part Ⓐ or Ⓑ, below.

Ⓐ See the criteria in Listing 14.04 or, if there is growth impairment, see the criteria in listings with 100 as the prefix (CD Part 1).

Ⓑ Linear scleroderma. In linear scleroderma, areas of skin abnormality appear as lines or bands rather than the patchy shape characteristic of most scleroderma. In addition, 1, 2, 3, or 4 must be satisfied.

1. Fixed valgus or varus deformities of both hands or both feet. These are deformities in which the hands or feet are bent in a fixed position away from or toward the body.
2. Marked destruction or marked atrophy of an extremity. This requirement is satisfied by damage to an arm or leg so severe that it has no useful function.
3. Facial disfigurement from hypoplasia of the mandible, maxilla, or zygoma resulting in a mental disorder, as described under the criteria in listings with 112 as the prefix (CD Part 12).
4. Seizure disorder, as described under the criteria in listings with 111 as the prefix (CD Part 11).

7. Listing 14.05: Polymyositis or Dermatomyositis (Adults)

Polymyositis or dermatomyositis are autoimmune diseases. They are primarily inflammatory processes found in striated muscles, which control bone movement. Both diseases can occur alone or in association with other connective tissue disorders or malignancy. Polymyositis and dermatomyositis are thought to be part of the same disorder, but with dermatomyositis the skin also becomes inflamed. While the muscle inflammation caused by these diseases can often be effectively treated, the cause of the disease is unknown and there is no cure. The majority of cases (65–75%) will respond to corticosteroid drugs, such as prednisone, which suppress the abnormal immune system process inflaming the muscles (myositis). Other drugs (for example, methotrexate, azathioprine) that suppress the immune system may also be tried. Any muscles can be affected.

There are no tests that clearly diagnose polymyositis or dermatomyositis. Diagnosis is partly based on the exclusion of other disorders like muscular dystrophy, metabolic diseases, endocrine diseases, drug side effects, and infections.

The diagnosis is supported by elevated serum muscle enzymes, characteristic abnormalities on electromyography, and myositis seen through a muscle biopsy. CPK and aldolase enzymes can be measured by a simple blood test to determine if the disease is worsening or responding to therapy, as higher levels indicate greater muscle inflammation. The SSA should consider elevations in these enzymes as strong evidence that weakness and fatigability are real and limiting. By looking at the laboratory reports in your own medical records, you can determine whether the enzymes are elevated, because the test reports will include your results and the expected values in a normal person. You can also find out whether the results of enzyme tests are abnormal from your treating doctor or medical books available in most bookstores or libraries. It is quite possible that the SSA could send you for a blood test to measure enzyme levels. Such tests can help the SSA determine the severity of your disease. That is relevant to the listing and any possible RFC, even though enzyme tests are not specifically mentioned by the listing. For example, the SSA is much more likely to believe that you have severe muscle weakness under part Ⓐ of the listing if your muscle enzymes are elevated than if the results are normal.

On the other hand, informed medical judgment is needed to evaluate enzyme tests results, because you could be truly weak and still have normal test results. How is this possible? Steroid drugs—such as prednisone—taken over a period of months can cause muscle weakness, a condition known as steroid myopathy. In this case, the SSA would be in error to dismiss your complaints of muscle weakness based on normal enzyme reports. It is important that the SSA obtain information over as long a period of time as possible, so that an accurate picture can be obtained of your health, rather than a snapshot of your condition at the time you apply for disability.

Electromyography (EMG) should show abnormally decreased electrical activity from significantly inflamed muscles. Evaluation of biopsy specimens under a microscope should show the characteristic degenerating and regenerating muscle fibers and presence of inflammatory cells. If you see an EMG report, look for mention of these abnormalities showing active muscle inflammation.

Skin lesions associated with dermatomyositis may appear in various forms, including characteristic violaceus patches—lesions with a purplish coloration. Itching may be intense and require treatment with antihistamines or other drugs. The skin should be protected from excessive exposure to sunlight by the use of sunscreens.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have a diagnosis of polymyositis or dermatomyositis. Additionally, you must satisfy part Ⓐ, Ⓑ, Ⓒ, or Ⓓ, below.

Ⓐ Severe proximal limb-girdle (shoulder or pelvic) muscle weakness. These are the muscles most likely to be weak. Pain and muscle tenderness may also be present and occasionally are the main symptom. Weakness of the pelvic girdle satisfying this part of the listing would result in significant difficulty climbing stairs or rising from a chair without use of the arms. Proximal limb weakness in the upper extremities may result in inability to lift objects, and interference with dressing and combing the hair.

Ⓑ Less severe limb-girdle muscle weakness than in part Ⓐ, associated with neck (cervical) muscle weakness. The anterior neck flexors are muscles that move the head forward in a standing position or upward in a lying (supine) position. Weakness of these flexors satisfies the requirement for muscle weakness in the neck, if the head cannot even be lifted from a pillow. Additionally, 1 or 2 must be present to at least a moderate level of severity, as determined by medical judgment.

1. Difficulty swallowing (dysphagia) and episodes of aspiration due to cricopharyngeal muscle weakness. The cricopharyngeal muscles are deep in the neck and just above the beginning of the esophagus; weakness of these muscles could result in dysphagia; episodes of aspiration can occur by getting food into the trachea and the lungs (aspiration). The muscles in the upper part of the esophagus may also be weakened, increasing the chances of aspiration. Note that while aspiration of food contents into the trachea can easily result in pneumonia, it is not a requirement of the listing.
2. Impaired breathing due to intercostal and diaphragmatic muscle weakness. The intercostal muscles, located between the ribs, are important for proper breathing. The diaphragm consists of two sheets of muscle between the chest and abdomen and is also important in respiration.

Ⓒ Association with a cancerous tumor, as described under the criteria in listings with 13 as a prefix (CD Part 13).

Ⓓ Association with generalized connective tissue disease, described under the criteria in Listings 14.02, 14.03, 14.04, or 14.06. If you examine those listings, they further refer to additional listings, such as those dealing with musculoskeletal impairments (CD Part 1), breathing disorders (CD Part 3), or heart disease (CD Part 4). The following facts are relevant to part Ⓓ:

- It is possible to have isolated dermatomyositis with skin involvement which may precede the development of polymyositis by several years in about 10% of cases.
- Polymyositis or dermatomyositis are known to be associated with a much higher risk than normal of developing some form of cancer, especially in older adults.

- Breathing difficulties can result from fibrosis of lung tissue itself, in addition to weakness of the muscles of respiration.
- Some patients also have painful, inflamed joints.
- The heart muscle may be inflamed (myocarditis), but only in a few patients.

b. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account residual impairment from both the polymyositis or dermatomyositis and treatment. As stated above, chronic use of steroid drugs can weaken muscles and should be considered a source of possible weakness even if muscle inflammation is under control. There should be avoidance of prolonged, intense sunlight exposure. Reference should also be made to comments about RFC under whatever body system and specific listings are most relevant to your problems.

8. Listing 114.05: Polymyositis or Dermatomyositis (Children)

See comments under adult Listing 14.05. Part Ⓑ provides additional possible criteria for allowance.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of polymyositis or dermatomyositis. Additionally, the child must satisfy part Ⓐ or Ⓑ, below.

Ⓐ See the criteria in adult Listing 14.05.

Ⓑ With one of the following:

1. Multiple fixed joints (joint contractures). Multiple only means more than one joint must be involved. One hand and wrist together count as one joint; individual finger joints cannot be counted separately.
2. Generalized skin calcification with formation of a hard crust outside the skin (exoskeleton). The child's entire skin surface does not have to be covered with calcium deposits. The skin must be involved generally, rather than at one or two locations.
3. Generalized arterial inflammation (systemic vasculitis) as described in Listing 14.03.

9. Listing 14.06: Undifferentiated Connective Tissue Disorder (Adults)

This listing deals with tissue disorder syndromes that have clinical (physical) and immunologic (laboratory) features of several connective tissue disorders, but that do not satisfy the criteria for any of the disorders described in other listings. For instance, there may be overlap syndromes with the signs and symptoms of rheumatoid arthritis and scleroderma. Or there may be clinical findings of systemic lupus and systemic vasculitis, but blood tests that confirm findings of rheumatoid arthritis. The correct diagnosis of undifferentiated connective tissue disorder is important for determining prognosis.

No actual evaluation is done under this listing. Claimants are evaluated under other listings or a combination of listings. The important point is that the SSA not disregard abnormalities that don't fit into a neat diagnostic category.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have a diagnosis of undifferentiated connective tissue disorder with impairment as described under the criteria in Listings 14.02Ⓐ, 14.02Ⓑ, or 14.04.

b. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account residual impairment from both the connective tissue disorder and treatment given. See the RFC discussion under whatever listing is used for evaluation.

10. Listing 114.06: Undifferentiated Connective Tissue Disorder (Children)

See comments under adult Listing 14.06.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of undifferentiated connective tissue disorder with impairment as described under the criteria in Listings 114.02 or 114.04.

11. Listing 14.07: Immune Deficiency Disorders Other Than Those Caused by HIV (Adults)

This listing deals with disorders associated with decreased immunity other than those caused by human immunodeficiency virus (HIV) infection, which is considered under another listing. Immune deficiency disorders are those in which there is either a deficiency of some type of antibody (immunoglobulin) or white cells that play an important role in the immune system. Immunity involving antibodies is known as humoral immunity, while immunity involving white cells is known as cell-mediated immunity. Antibodies are produced by white cells known as B lymphocytes, while in cell-mediated immunity T lymphocytes (for example, CD4 (T4) lymphocytes) play a direct role in controlling infection. This listing can be used for either type of immunity.

Various types of immune deficiency may be present at birth or arise later in life. Adult deficiencies not associated with HIV infection are more often an antibody deficiency than a white cell deficiency. There may be a deficiency of one or more types of antibody. For example, combined variable immunodeficiency disorder can occur in adults, especially in association with autoimmune diseases like systemic lupus erythematosus (SLE), and involves the deficiency of multiple types of antibodies. Antibody deficiencies can be treated with immunoglobulin injections to bring them back to normal levels.

Infection is the risk for those with antibody deficiencies, especially IgG or IgM. Therefore, the listing deals with repeated medically severe infections. Most likely, such infections would result in hospitalization and treatment with intravenous antibiotics, but hospitalization is not a requirement of the listing.

Various kinds of bodily damage done by recurrent infection might require evaluation under another listing. For example, repeated episodes of pneumonia could result in chronic lung damage. If this listing is not satisfied, evaluation would then also be done under the breathing disorder listings (CD Part 3).

a. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have a diagnosis of immune deficiency disorders other than those caused by HIV associated with documented, recurrent severe infection occurring three or more times within a five-month period.

b. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account residual impairment from both the immune deficiency disorder and treatment given. Recurrent severe infections can result in so much time being sick and receiving treatment that physical strength and stamina can be affected. Reference should also be made to the discussions of RFC for particular disorders that might result as a complication of bacterial infections, such as the RFCs discussed in regard to lung disease caused by repeated pulmonary infections (CD Part 3). Damage to the kidneys would be evaluated under the listing for kidney disorders (CD Part 6).

12. Listing 114.07: Congenital Immune Deficiency Disease (Children)

See the comments under adult Listing 14.07. You may encounter listings using the words "gamma globulin." That is an old way of referring to immunoglobulins, but remains part of the name of some medical disorders. In congenital hypogammaglobulinemia there is a decrease in all types of antibodies. In congenital dysgammaglobulinemia there is a decrease in some types of antibodies. The risk of severe infection accompanies extremely low antibody levels. Other abnormalities, such as described in part Ⓐ2, below, may also be present.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of congenital immune deficiency disease and either part Ⓐ or Ⓑ, below.

Ⓐ Hypogammaglobulinemia or dysgammaglobulinemia, with:

1. Documented, recurrent severe infections occurring three or more times within a five-month period, or
2. An associated disorder such as growth retardation, chronic lung disease, connective tissue disorder, or tumor. Evaluate according to the appropriate body system listing.

Ⓑ Thymic dysplastic syndromes (such as Swiss or diGeorge). The thymus gland in the neck plays an important role in the immune system, especially in developing cell-mediated immunity. The Swiss and diGeorge syndromes are rare congenital disorders involving a poorly developed or abnormal thymus. The risk of severe infection is so high that these diagnoses alone are sufficient for allowance.

13. Listing 14.08: Human Immunodeficiency Virus (HIV) Infection (Adults)

a. General Information

Human immunodeficiency virus (HIV) infection is caused by a type of retrovirus. HIV weakens the immune system by destroying CD4 (T4) lymphocytes. By suppressing the immune system, HIV infection makes an infected person susceptible to one or more opportunistic diseases, cancers, or other conditions described in this listing. Any person with HIV infection, including one with a diagnosis of acquired immunodeficiency syndrome (AIDS), may be found disabled under this listing if his or her impairment meets any of the criteria in this listing or is of equivalent severity to any impairment in this listing.

b. Definitions

In this listing, the meanings of the terms "resistant to treatment," "recurrent," and "disseminated" used by the SSA have the same general meaning as used by the medical community. The precise meaning of any of these terms will depend upon the specific disease or condition in question, the body system affected, the usual course of the disorder and its treatment, and the other circumstances of the case.

- "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 appropriate, will depend on the facts of the particular case.
- "Recurrent" means that a condition that responded adequately to an appropriate course of treatment has returned after a period of remission or regression. The extent of response (or remission) and the time periods involved will depend on the facts of the particular case.
- "Disseminated" means that a condition is spread widely over a considerable area or body system(s). The type and extent of the spread will depend on the specific disease.
- As used in Part I of the listing, "significant involuntary weight loss" does not correspond to a specific minimum amount or percentage of weight loss, although, for purposes of this listing, an involuntary weight loss of at least 10% of baseline is always considered significant. Loss of less than 10% may or may not be significant, depending on the individual's baseline weight and body build (also called habitus). For example, a seven-pound weight loss in a 100-pound female who is 63 inches tall might be considered significant; but a 14-pound weight loss in a 200-pound female who is the same height might not be significant.

c. Manifestations Specific to Women

Most women with severe suppression of their immune system caused by HIV infection show the typical opportunistic infections and other conditions, such as pneumocystis carinii pneumonia (PCP), yeast fungal infection of the esophagus (candida esophagitis), extreme malnutrition and weight loss (wasting syndrome), cryptococcosis (cryptococcal fungal infection), and toxoplasmosis. However, HIV infection may have different manifestations in women than in men. Doctors working for the SSA must carefully evaluate the medical evidence and be alert to the variety of medical conditions specific to or common in women with HIV infection that may affect their ability to function in the workplace.

Many of these manifestations (for example, vulvo-vaginal candidiasis, pelvic inflammatory disease) occur in women with or without HIV infection, but

can be more severe or resistant to treatment, or occur more frequently in a woman whose immune system is suppressed. Vulvovaginal candidiasis is a yeast fungal infection of the female genitalia; pelvic inflammatory disease (PID) refers to infection involving the female reproductive organs: the ovaries, fallopian tubes, or uterus. Therefore, when evaluating the claim of a woman with HIV infection, it is important to consider gynecologic and other problems specific to women, including any associated symptoms (for example, pelvic pain), in determining the severity of the impairment and resulting functional limitations. Manifestations of HIV infection in women may be evaluated under the specific criteria (for example, cervical cancer under part Ⓔ), under an applicable general category (for example, pelvic inflammatory disease under part Ⓐ5), or in appropriate cases, under part Ⓝ.

d. Evaluation

The criteria in this listing do not describe the full spectrum of diseases or conditions manifested by people with HIV infection. As in any case, consideration must be given to whether an individual's impairment meets or equals in severity any other listing (for example, the cancer listings with prefixes of 13 in CD Part 13). Although this listing includes cross-references to other listings for the more common manifestations of HIV infection, additional other listings may also apply.

In addition, the effect of all impairments on the claimant, whether or not related to HIV infection, must be considered. For example, individuals with HIV infection may manifest signs and symptoms of a mental impairment (for example, anxiety and depression), or of another physical impairment. Medical evidence should include documentation of all physical and mental impairments, and the impairments should be evaluated not only under other listings mentioned in Listing 14.08, but under any other appropriate listings.

e. 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.

i. Documentation of HIV Infection by Definitive Diagnosis

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

- A serum specimen that contains HIV antibodies. HIV antibodies are usually detected by a screening test. The most commonly used screening test is the ELISA. Although this test is highly sensitive, it may yield false positive results. Therefore, positive results from an ELISA must be confirmed by a more definitive test, such as the Western blot or immunofluorescence assay (IFA).
- A specimen that contains immune-reactive parts of the human immunodeficiency virus (HIV antigen), such as serum specimens, lymphocyte cultures, or cerebrospinal fluid (CSF) specimens.
- Other tests that are highly specific for detection of HIV, such as polymerase chain reaction (PCR), or that are acceptable methods of detection consistent with the prevailing state of medical knowledge.

When laboratory testing for HIV infection has been performed, the SSA must make every reasonable effort to obtain the results of that testing.

Individuals who have HIV infection or other disorders of the immune system may undergo tests to determine the T-helper lymphocyte (CD4) count in the blood, because CD4 cells are destroyed by HIV. The extent of immune depression correlates with the level or rate of decline of the CD4 count. In general, when the CD4 count is 200/mm^3 or less (14% or less), the susceptibility to opportunistic disease is considerably increased. CD4 lymphocytes are necessary for effective cell-mediated immunity, and such immunity declines as the number of CD4 cells in the blood fall. However, a low CD4 count alone does not establish a definite diagnosis of HIV infection. Nor will it document the severity or functional effects of HIV infection.

ii. Other Acceptable Documentation of HIV Infection

HIV infection may also be documented without the definitive laboratory tests described above, if such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is

consistent with the other evidence. If no definitive laboratory evidence is available, HIV infection may be documented by the medical history, clinical and laboratory findings, and diagnoses indicated in the medical evidence.

For example, a diagnosis of HIV infection will be accepted without definitive laboratory evidence if the claimant has an opportunistic disease, such as toxoplasmosis of the brain or pneumocystis carinii pneumonia (PCP). Toxoplasmosis and PCP are caused by protozoan organisms that only seriously infect people with weak immune systems. Their presence strongly suggests that the person has a defect in cell-mediated immunity caused by a low CD4 lymphocyte count. However, to reach this conclusion without definitive laboratory evidence of HIV infection there must be no other known cause of diminished resistance to the opportunistic disease concerned. For instance, long-term steroid treatment and lymphoma-type cancer can also suppress the immune system. In all cases without definitive laboratory evidence of HIV infection, the SSA must make every reasonable effort to obtain full details of the history, medical findings, and results of testing. Problems can arise if the treating doctor, hospital, or other medical facility has lost the records documenting HIV infection, or simply refuses to give them to the SSA.

f. Documentation of Manifestations of HIV Infection

A person infected with HIV may not have any symptoms, much less any opportunistic diseases or other functional limitations. Therefore, the medical evidence must also include documentation of any possibly disabling manifestations of HIV infection. Such manifestations could include, for example, opportunistic infections such as pneumocystis carinii pneumonia, or cancer. Such manifestations would justify a diagnosis of AIDS. While claimants with impairments that satisfy the listing would generally be considered to have AIDS, the listing itself does not mention AIDS, because it is the severity of the impairments that matters, not whether there is a diagnosis of AIDS. A treating doctor cannot establish severity on the basis of diagnosis. Documentation of manifestations of HIV infection may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

i. Documentation of Manifestations of HIV Infection by Definitive Diagnosis

The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serological test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). Therefore, the SSA must make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a test of tissue (histological test) or other test has been performed, the evidence should include a copy of the appropriate report. If the report is not obtainable, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including x-ray reports) or microscopic examination of the appropriate tissues or body fluids. Various test reports can be unobtainable for a variety of reasons, such as loss or simply refusal by a treating doctor, hospital, or other facility to give the information to the SSA.

Although a reduced CD4 lymphocyte count may show that there is an increased susceptibility to opportunistic infections and diseases, that alone does not establish the presence, severity, or functional effects of a manifestation of HIV infection.

ii. Other Acceptable Documentation of Manifestations of HIV Infection

Manifestations of HIV infection may also be documented without the definitive laboratory evidence described above provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and other evidence. If no definitive laboratory evidence is available, manifestations of HIV infection may be documented by medical history, clinical and laboratory findings, and diagnoses indicated in the medical evidence. In such cases, the SSA must make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

Documentation of cytomegalovirus (CMV) disease (part Ⓓ) presents special problems, because diagnosis

requires identification of viral inclusion bodies or a positive culture from the affected organ, and the absence of any other infectious agent. A positive serology test identifies infection with the virus, but does not confirm a disease process. With the exception of chorioretinitis (which may be diagnosed by an ophthalmologist), documentation of CMV disease requires confirmation by biopsy or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

g. Effect of Treatment

Medical treatment must be considered in terms of its effectiveness in decreasing the signs, symptoms, and laboratory abnormalities of the specific disorder, or of the HIV infection itself. For example, the antiretroviral drugs used to treat HIV and AIDS may have side effects of treatment that may further impair a claimant.

Response to treatment and adverse or beneficial consequences of treatment may vary widely. For example, an individual with HIV infection who develops pneumonia or tuberculosis may respond to the same antibiotic regimen used in treating people without HIV infection, but another person with HIV infection may not respond to the same regimen. Therefore, each case must be considered on an individual basis, along with the effects of treatment on the person's ability to function.

A specific description of the drugs or treatment given (including surgery), dosage, frequency of administration, and a description of the complications or response to treatment should be obtained. The effects of treatment may be temporary or long-term. As such, the decision regarding the impact of treatment should be based on a sufficient period of treatment to permit proper consideration.

h. Functional Criteria

Part Ⓝ of this listing establishes standards for evaluating manifestations of HIV infection that do not meet the requirements listed in parts Ⓐ through Ⓜ.

For individuals with HIV infection evaluated under part Ⓝ, listing-level severity will be assessed in terms of the functional limitations imposed by the impairment. The full impact of signs, symptoms, and laboratory findings on the claimant's ability to function must be considered. Important factors to be considered in evaluating the functioning of people with HIV infection include, but are not limited to:

- symptoms, such as fatigue and pain
- characteristics of the illness, such as the frequency and duration of manifestations or periods of worsening (exacerbation) and improvement (remission), and
- the effect of the treatment for the disease on the claimant's ability to function, including the side effects of medication.

As used in part Ⓝ, "repeated" means that:

- The conditions occur on an average of three times a year, or once every four months, each lasting two weeks or more, or
- The conditions do not last for two weeks but occur substantially more frequently than three times in a year or once every four months, or
- The conditions occur less often than an average of three times a year or once every four months, but last substantially longer than two weeks at a time.

To meet the criteria in part Ⓝ, an individual with HIV infection must demonstrate a marked level of restriction in one of three general areas of functioning:

- activities of daily living (ADLs)
- social functioning, and
- difficulties in completing tasks due to deficiencies in concentration, persistence, or speed of work (pace).

Functional restrictions may be caused by the effect of the disease process on mental or physical functioning, or both. For example, these effects could result from extended or intermittent symptoms, such as depression, fatigue, or pain, resulting in a limitation of the ability to concentrate, to persevere at a task, or to perform the task at an acceptable rate of speed. Limitations may also result from the side effects of medication.

When "marked" is used as a standard for measuring the degree of functional limitation, it means more than moderate, but less than extreme. A marked limitation does not represent a quantitative measure of the person's ability to do an activity for a certain percentage of the time. A marked limitation may be present when several activities or functions

are impaired or even when only one is impaired. However, a claimant need not be totally unable to perform an activity to have a marked limitation, as long as the degree of limitation is such as to seriously interfere with the ability to function independently, appropriately, and effectively. The term "marked" does not imply that the impaired person is confined to bed, hospitalized, or in a nursing home.

Activities of daily living include, but are not limited to, such activities as doing household chores, grooming and hygiene, using a post office, taking public transportation, and paying bills. A person with HIV infection who, because of symptoms such as pain imposed by the illness or its treatment, is not able to maintain a household or take public transportation on a sustained basis or without assistance, would have marked limitation of activities of daily living. This would be true even though he or she is able to perform some self-care activities.

Social functioning includes the capacity to interact appropriately and communicate effectively with others. A person with HIV infection who, because of symptoms or a pattern of worsening and improvement caused by the illness or its treatment, cannot engage in social interaction on a sustained basis (even though they are able to communicate with close friends or relatives) would have marked difficulty maintaining social functioning.

Completing workplace tasks in a timely manner involves the ability to sustain concentration, persistence, or pace. An individual with HIV infection who, because of HIV-related fatigue or other symptoms, is unable to sustain concentration or pace adequate to complete simple work-related tasks (even though he or she is able to do routine activities of daily living) would have marked difficulty completing tasks.

i. Listing Level Severity

For your condition to be severe enough to meet this listing, you must have a diagnosis of human immunodeficiency virus (HIV) infection and your condition must match Ⓐ, Ⓑ, Ⓒ, Ⓓ, Ⓔ, Ⓕ, Ⓖ, Ⓗ, Ⓘ, Ⓙ, Ⓚ, Ⓛ, Ⓜ, or Ⓝ, below.

Ⓐ Bacterial infections:

1. Mycobacterial infection such as tuberculosis (TB) or related infections (for example, M. avium-intracellulare, M. kansasii) at a site other than the lungs, skin, or cervical or hilar lymph nodes. Also, pulmonary tuberculosis resistant to treatment qualifies under this part of the listing.
2. Nocardiosis.
3. Salmonella bacteria in the blood (bacteremia), recurrent nontyphoid type.
4. Syphilis. Evaluate under the criteria for the affected body system, such as the listings dealing with vision (listings with a 2 prefix, CD Part 2), blood vessels (listings with a 4 prefix, CD Part 4), or the nervous system (listings with an 11 prefix, CD Part 11).
5. Multiple or recurrent bacterial infections, including infections of the female reproductive organs (pelvic inflammatory disease), requiring hospitalization or intravenous antibiotic treatment three or more times in one year.

Ⓑ Fungal infections:

1. Aspergillosis.
2. Candidiasis, at a site other than the skin, urinary tract, or intestinal tract. Also, candidiasis at a site other than the mucous membranes of the mouth, vulva, or vagina. Candidiasis involving the esophagus, trachea, bronchi, or lungs qualifies under this part of the listing.
3. Coccidioidomycosis, at a site other than the lungs or lymph nodes.
4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis).
5. Histoplasmosis, at a site other than the lungs or lymph nodes.
6. Mucormycosis.

Ⓒ Protozoan or parasitic worm (helminthic) infections:

1. Cryptosporidiosis, isosporiasis, or microsporidiosis protozoan infections, with diarrhea lasting for one month or longer.
2. Pneumocystis carinii pneumonia or pneumocystis carinii infection occurring outside of the lungs.
3. Strongyloidiasis parasitic worm infection occurring outside of the intestines.
4. Toxoplasmosis protozoan infection of an organ other than the liver, spleen, or lymph nodes.

Ⓓ Viral infections:

1. Cytomegalovirus disease at a site other than the liver, spleen, or lymph nodes.

2. Herpes simplex virus causing:
   a. Skin or mucous membrane infection affecting the mouth, genitals, or perianal region, lasting for one 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. Infection that has spread widely throughout the body.
3. Herpes zoster, either disseminated or with eruptions along multiple nerves (multidermatomal) that are resistant to treatment.
4. Progressive viral brain destruction known as multifocal leukoencephalopathy.
5. Hepatitis, as described under the criteria in Listing 5.05 (CD Part 5).

Ⓔ Cancer:
1. Carcinoma of the cervix, invasive, FIGO stage II, and beyond.
2. Kaposi's sarcoma with:
   a. Extensive oral lesions, or
   b. Involvement of the gastrointestinal tract, lungs, or other large internal (visceral) organs, or
   c. Involvement of the skin or mucous membranes, as described under the criteria in 14.08Ⓕ.
3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, Hodgkin's disease).
4. Squamous cell carcinoma of the anus.

Ⓕ Conditions of the skin or mucous membranes (other than described in parts Ⓑ2, Ⓓ2, or Ⓓ3, above) with extensive fungating or ulcerating lesions not responding to treatment. Examples of qualifying conditions are eczema or psoriasis, vulvovaginal candidiasis or other mucosal candida, venereal warts on the genitalia (condyloma) caused by human papillomavirus, and ulcerative diseases of the genitalia. Or, if more appropriate, the lesions can be evaluated under the criteria of the listings dealing with skin disorders (listings with an 8 prefix, CD Part 8).

Ⓖ Blood (hematologic) abnormalities:
1. Decreased red blood cells (anemia), as described under the criteria in Listing 7.02 (CD Part 7).
2. Decreased white blood cells (granulocytopenia), as described under the criteria in Listing 7.15 (CD Part 7).
3. Decreased platelets (thrombocytopenia), as described under the criteria in Listing 7.06 (CD Part 7).

Ⓗ Neurological abnormalities:
1. Encephalopathy caused by HIV, characterized by thinking (cognitive) or movement abnormalities (motor dysfunction) that limit function and progressively worsen.
2. Other neurological manifestations of HIV infection such as nerve damage (peripheral neuropathy) as described under the criteria in the listings dealing with nervous system disorders (listings with an 11 prefix, CD Part 11).

Ⓘ HIV wasting syndrome, characterized by involuntary weight loss of 10% or more of baseline (or other significant involuntary weight loss) and, in the absence of a concurrent illness that could explain the findings, either:
1. Chronic diarrhea with two or more loose stools daily, lasting for one month or longer, or
2. Chronic weakness and documented fever greater than 38° C (100.4° F) for the majority of one month or longer.

Ⓙ Diarrhea, lasting for one month or longer, resistant to treatment, and requiring intravenous fluids (hydration), intravenous feeding (alimentation), or tube feeding.

Ⓚ Cardiomyopathy, as described under the criteria in the listings dealing with heart disease (listings with a 4 prefix, CD Part 4) or the neurological listing dealing with strokes (11.04, CD Part 11).

Ⓛ Nephropathy, as described under the criteria in the listings dealing with kidney disease (listings with a 6 prefix, CD Part 6).

Ⓜ One or more of the following infections (other than described in parts Ⓐ through Ⓛ, above), resistant to treatment or requiring hospitalization or intravenous treatment three or more times in one year. Or evaluate abnormalities caused by the infection (sequelae) under the criteria for the affected body system, including:
- infection affecting the whole body (sepsis)
- infection of the meninges covering the brain or spinal cord (meningitis)

- infection of the lungs (pneumonia)
- infection of a joint (septic arthritis)
- infection of the heart (endocarditis), and
- infection of a sinus (sinusitis) documented by x-rays.

Ⓝ Repeated manifestations of HIV infection. These manifestations can include those listed in parts Ⓐ through Ⓜ that don't have findings required by those parts of the listing—for example, carcinoma of the cervix not meeting the criteria in part Ⓔ, or diarrhea not meeting the criteria in part Ⓙ. Other manifestations of HIV infection can also be considered, such as white lesions on the sides of the tongue (oral hairy leukoplakia), or muscle inflammation (myositis). Whatever the manifestation, it must result in significant, documented symptoms or signs (for example, fatigue, fever, malaise, weight loss, pain, night sweats) and one of the following at the marked level of severity:

- restriction of activities of daily living
- difficulties in maintaining social functioning, or
- difficulties in completing tasks in a timely manner due to deficiencies in concentration, persistence, or pace.

j. Residual Functional Capacity

Medical judgment must be applied to individual cases, taking into account residual impairment from both the HIV-associated disorders and treatment given. Additionally, reference should be made to comments about RFCs under other listings as appropriate to the types of problems related to the HIV infection. Particular attention should be paid to whether the person has the strength to stand and walk six to eight hours a day. If not, the RFC cannot be higher than sedentary work. Especially in people over 50 years of age, allowance on a medical-vocational basis becomes much more likely with a sedentary RFC. (See Chapter 9 for a detailed discussion of medical-vocational rules.)

14. Listing 114.08: Human Immunodeficiency Virus (HIV) Infection (Children)

SSA policies on HIV infections in children and adults have many similarities. But there are enough differences that a separate discussion is necessary.

a. General

See "General" in Listing 14.08, above.

b. Definitions

See "Definitions" in Listing 14.08, above.

c. HIV Infection in Children

The clinical manifestation and course of disease in children who become infected with HIV at or near the time of birth (perinatally) or in the first six years of life may differ from that in older children and adults. In addition, survival times are shorter for children infected in the first year of life compared to those who become infected as older children or as adults. Infants may fail to gain weight (failure to thrive) or develop pneumocystis carinii pneumonia (PCP). Young children may have recurrent infections, neurological problems, or developmental abnormalities. Older children may also exhibit neurological abnormalities, such as HIV brain damage (encephalopathy), or failure to thrive.

The methods of identifying and evaluating neurological 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 older children, impaired brain growth can be documented by brain atrophy on a CAT scan. Neurological abnormalities can also be observed in a younger child by the loss of previously acquired, or marked delays in achieving, developmental milestones. In an older child, this type of neurological abnormality would generally be demonstrated by the loss of previously acquired intellectual abilities. Although loss of previously acquired intellectual abilities can be documented by a decrease in intelligence quotient (IQ) scores or demonstrated if a child forgets information he or she previously learned, it can also be shown if the child is unable to learn new information. This could include the sudden acquisition of a new learning disability.

Children with HIV infection may contract any of a broad range of bacterial infections. Certain major infections caused by pyogenic (pus-forming) bacteria—for example, some pneumonias—can be severely limiting, especially in preadolescent children. These major bacterial infections should be evaluated under part Ⓐ5, which requires two or more such

infections within a two-year period. Although part Ⓐ5 applies only to children younger than 13 years of age, an older child may be found to have an impairment of equivalent severity if the circumstances of the case warrant (for example, delayed puberty).

Otherwise, bacterial infections are evaluated under part Ⓐ6. The criteria of the listing are met if one or more bacterial infections occurs and requires hospitalization or intravenous antibiotic treatment three or more times in one year. Pelvic inflammatory disease in older female children should be evaluated under multiple or recurrent bacterial infections in part Ⓐ6.

d. Evaluation of HIV Infection in Children

The criteria in this listing do not describe the full spectrum of diseases or conditions manifested by individuals with HIV infection. As in any case, consideration must be given to whether a child's impairment meets or equals in severity any other listing (for example, the cancer listings with the prefix 113, CD Part 13). Although this listing includes cross-references to other listings for the more common manifestations of HIV infection, other additional listings may also apply.

In addition, the effect of all impairments on a claimant, whether or not related to HIV infection, must be considered. Individuals with HIV infection may manifest signs and symptoms of a mental impairment (for example, anxiety or depression), or of another physical impairment. Medical evidence should include documentation of all physical and mental impairments, and the impairments should be evaluated not only under other listings mentioned in Listing 14.08, but under any other appropriate listings.

e. Documentation of HIV Infection in Children

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.

i. Documentation of HIV Infection in Children by Definitive Diagnosis

A definitive diagnosis of HIV infection in children is documented by one or more of the following laboratory tests:

1. For a child 24 months of age or older, a serum specimen that contains HIV antibodies. HIV antibodies are usually detected by a screening test. The most commonly used screening test is the ELISA. Although this test is highly sensitive, it may yield false positive results. Therefore, positive results from an ELISA must be confirmed by a more definitive test, such as the Western blot or immunofluorescence assay (IFA).
2. A specimen that contains immune-reactive parts of the human immunodeficiency virus (HIV antigen), such as serum specimens, lymphocyte cultures, or cerebrospinal fluid (CSF) specimens.
3. Other tests that are highly specific for detection of HIV, such as polymerase chain reaction (PCR), or that are acceptable methods of detection consistent with the prevailing state of medical knowledge.

ii. Other Acceptable Documentation of HIV Infection in Children

HIV infection cannot be documented in children younger than 24 months of age by a serum specimen containing HIV antibodies. This is because women with HIV infection often transfer HIV antibodies to their newborns. The mother's antibodies can persist in the infant for up to 24 months, even if the infant is not HIV-infected. Only 20% to 30% of such infants with HIV antibodies are actually infected. However, the presence of HIV antibodies accompanied by evidence of significantly depressed T-helper lymphocytes (CD4), an abnormal CD4/CD8 ratio, or abnormal immunoglobulin G (IgG) may be used to document HIV infection in a child under 24 months of age, even though such testing is not a basis for a definitive diagnosis. Immunoglobulin G is an antibody produced in response to HIV infection. The helper/suppressor ratio is the number of helper lymphocytes (CD4 lymphocytes) divided by the number of suppressor lymphocytes (CD8 lymphocytes). An abnormally low CD4/CD8 ratio suggests that CD4 lymphocytes are being destroyed.

For children from birth to 24 months of age who have tested positive for HIV antibodies, HIV infection may be documented by one or more of the following:

1. For an infant 12 months of age or less, a CD4 (T4) count of 1,500/mm³ or less, or a CD4 count less than or equal to 20% of total lymphocytes.
2. For an infant from 12 to 24 months of age, a CD4 (T4) count of 750/mm³ or less, or a CD4 count less than or equal to 20% of total lymphocytes.
3. An abnormal CD4/CD8 ratio.
4. An IgG significantly greater than or less than the normal range for age.

HIV infection may also be documented without the definitive laboratory tests described above, if such documentation is consistent with the prevailing state of medical knowledge and clinical practice and with the other evidence. If no definitive laboratory evidence is available, HIV infection may be documented by the medical history, clinical and laboratory findings, and diagnosis(es) indicated in the medical evidence.

For example, a diagnosis of HIV infection will be accepted without definitive laboratory evidence of HIV infection if the claimant has an opportunistic disease, such as toxoplasmosis of the brain or pneumocystis carinii pneumonia (PCP). Toxoplasmosis and PCP are caused by protozoan organisms that only seriously infect people with weak immune systems. Their presence strongly suggests that the child has a defect in cell-mediated immunity caused by a low CD4 lymphocyte count. However, to reach this conclusion without definitive laboratory evidence of HIV infection, there must be no other known cause of diminished resistance to the opportunistic disease concerned. For instance, long-term steroid treatment and lymphoma-type cancer can also suppress the immune system. In all cases without definitive laboratory evidence of HIV infection, the SSA must make every reasonable effort to obtain full details of the history, medical findings, and results of testing.

f. Documentation of Manifestations of HIV Infection

A person with HIV infection does not necessarily have symptoms, much less opportunistic diseases or other functional limitations. Therefore, the medical evidence must also include documentation of any possibly disabling manifestations of HIV infection. Such manifestations could include, for example, opportunistic infections such as pneumocystis carinii pneumonia, or cancer. Such manifestations would justify a diagnosis of AIDS. However, the listing itself does not mention AIDS because it is the severity of the impairments that matters and not whether there is a diagnosis of AIDS. A treating doctor cannot establish severity on the basis of an AIDS diagnosis. Documentation of manifestations of HIV infection may be by laboratory evidence or by other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

i. Documentation of Manifestations of HIV Infection in Children by Definitive Diagnosis

The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serological test, or microscopic examination of biopsied tissue or other material (for example, bronchial washings). Therefore, the SSA must make every reasonable effort to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a test of tissue (histological test) or other test has been performed, the evidence should include a copy of the appropriate report. If the report is not obtainable, the summary of hospitalization or a report from the treating source should include details of the findings and results of the diagnostic studies (including x-ray reports) or microscopic examination of the appropriate tissues or body fluids.

Although a reduced CD4 lymphocyte count may show that there is an increased susceptibility to opportunistic infections and diseases, that alone does not establish the presence, severity, or functional effects of a manifestation of HIV infection in a child.

ii. Other Acceptable Documentation of the Manifestations of HIV Infection in Children

Manifestations of HIV infection may also be documented without the definitive laboratory evidence described above provided that such documentation is consistent with the prevailing state of medical knowledge and clinical practice and is consistent with the other evidence. If no definitive laboratory evidence is available, manifestations of HIV infection may be documented by medical history, clinical and laboratory findings, and diagnoses

indicated in the medical evidence. In such cases, every reasonable effort must be made to obtain full details of the history, medical findings, and results of testing.

Documentation of cytomegalovirus (CMV) disease (part Ⓓ) presents special problems because diagnosis requires identification of viral inclusion bodies or a positive culture from the affected organ, and the absence of any other infectious agent. A positive serology test identifies infection with the virus, but does not confirm a disease process. With the exception of chorioretinitis (which may be diagnosed by an ophthalmologist), documentation of CMV disease requires confirmation by biopsy or other generally acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

g. Effect of Treatment

Medical treatment must be considered in terms of its effectiveness in decreasing the signs, symptoms, and laboratory abnormalities of the specific disorder, or of the HIV infection itself. For example, the antiretroviral drugs used to treat HIV and AIDS may have side effects of treatment that may further impair a claimant.

Response to treatment and adverse or beneficial consequences of treatment may vary widely. For example, a child with HIV infection who develops a middle ear infection (otitis media) may respond to the same antibiotic regimen used in treating children without HIV infection, but another child with HIV infection may not respond to the same regimen. Therefore, each case must be considered on an individual basis, along with the effects of treatment on the child's ability to function.

A specific description of the drugs or treatment given (including surgery), their dosage, frequency of administration, and a description of the complications or response to treatment should be obtained. The effects of treatment may be temporary or long-term. As such, the decision regarding the impact of treatment should be based on a sufficient period of treatment to permit proper consideration.

h. Functional Criteria

Part Ⓞ of this listing establishes standards for evaluating manifestations of HIV infection that do not meet the requirements listed in parts Ⓐ through Ⓝ. Part Ⓞ is applicable for manifestations that are not listed in parts Ⓐ through Ⓝ, as well as those that are listed in parts Ⓐ through Ⓝ but do not meet the criteria of any of the rules in parts Ⓐ through Ⓝ.

For children with HIV infection evaluated under part Ⓞ, listing-level severity will be assessed in terms of the functional limitations imposed by the impairment. The full impact of signs, symptoms and laboratory findings on the child's ability to function must be considered. Important factors to be considered in evaluating the functioning of children with HIV infection include—but are not limited to:

- symptoms, such as fatigue and pain
- characteristics of the illness, such as the frequency and duration of manifestations or periods of worsening and improvement (exacerbation and remission) in the disease course, and
- the functional impact of treatment for the disease, including the side effects of medication.

To meet the criteria in part Ⓞ, a child with HIV infection must demonstrate a level of restriction in either one or two (depending on the child's age) of the general areas of functioning applicable to the child's age group.

i. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child must have a diagnosis of human immunodeficiency virus (HIV) infection and part Ⓐ, Ⓑ, Ⓒ, Ⓓ, Ⓔ, Ⓕ, Ⓖ, Ⓗ, Ⓘ, Ⓙ, Ⓚ, Ⓛ, Ⓜ, Ⓝ, or Ⓞ, below.

Ⓐ Bacterial infections:

1. Mycobacterial infection such as tuberculosis (TB) or related infections (for example, caused by M. avium-intracellulare, M. kansasii) at a site other than the lungs, skin, or cervical or hilar lymph nodes. Also, pulmonary tuberculosis resistant to treatment qualifies under this part of the listing.
2. Nocardiosis.
3. Salmonella bacteria in the blood (bacteremia), recurrent nontyphoid type.
4. Syphilis (evaluate under the criteria for the affected body system, such as the listings dealing with vision (listings with a 2 prefix, CD Part 2), heart or blood vessels (listings with a 4 prefix, CD Part 4), or the nervous system (listings with an 11 prefix, CD Part 11).

5. In a child younger than 13 years of age, multiple or recurrent pus-forming bacterial infections (pyogenic bacterial infections) of the following types: sepsis, pneumonia, meningitis, bone or joint infection, or abscess of an internal organ or body cavity (excluding otitis media or superficial skin or mucosal abscesses) occurring two or more times in two years.
6. Multiple or recurrent bacterial infections, including infections of the female reproductive organs (pelvic inflammatory disease), requiring hospitalization or intravenous antibiotic treatment three or more times in one year.

Ⓑ Fungal infections:
1. Aspergillosis.
2. Candidiasis, at a site other than the skin, urinary tract, or intestinal tract. Also, candidiasis at a site other than the mucous membranes of the mouth, vulva, or vagina. Candidiasis involving the esophagus, trachea, bronchi, or lungs qualifies under this part of the listing.
3. Coccidioidomycosis, at a site other than the lungs or lymph nodes.
4. Cryptococcosis, at a site other than the lungs (for example, cryptococcal meningitis).
5. Histoplasmosis, at a site other than the lungs or lymph nodes.
6. Mucormycosis.

Ⓒ Protozoan or parasitic worm (helminthic) infections:
1. Cryptosporidiosis, isosporiasis, or microsporidiosis protozoan infections, with diarrhea lasting for one month or longer.
2. Pneumocystis carinii pneumonia or pneumocystis carinii infection occurring outside of the lungs.
3. Strongyloidiasis parasitic worm infection occurring outside of the intestines.
4. Toxoplasmosis protozoan infection of an organ other than the liver, spleen, or lymph nodes.

Ⓓ Viral infections:
1. Cytomegalovirus disease at a site other than the liver, spleen, or lymph nodes.
2. Herpes simplex virus causing:
   a. Skin or mucous membrane infection affecting the mouth, genitals, or perianal region, lasting for one 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. Infection that has spread widely throughout the body (disseminated infection).
3. Herpes zoster, either disseminated or with eruptions along multiple nerves (multi-dermatomal) that are resistant to treatment.
4. Progressive viral brain destruction known as multifocal leukoencephalopathy.
5. Liver inflammation (hepatitis), as described under the criteria in Listing 105.05 (CD Part 5).

Ⓔ Cancer:
1. Carcinoma of the cervix, invasive, FIGO stage II and beyond.
2. Kaposi's sarcoma with:
   a. Extensive oral lesions, or
   b. Involvement of the gastrointestinal tract, lungs, or other large internal (visceral) organs, or
   c. Involvement of the skin or mucous membranes, as described under the criteria in part Ⓕ.
3. Lymphoma (for example, primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other non-Hodgkin's lymphoma, or Hodgkin's disease).
4. Squamous cell carcinoma of the anus.

Ⓕ Conditions of the skin or mucous membranes (other than described in parts Ⓑ2, Ⓓ2, or Ⓓ3, above) with extensive fungating or ulcerating lesions not responding to treatment. Examples of qualifying conditions are eczema or psoriasis, vulvovaginal candidiasis or other mucosal candida, venereal warts on the genitalia (condyloma) caused by human papillomavirus, and ulcerative diseases of the genitalia. Or, if more appropriate, the lesions can be evaluated under the criteria of the listings dealing with skin disorders (listings with an 8 prefix, CD Part 8).

Ⓖ Blood (hematologic) abnormalities:
1. Decreased red blood cells (anemia), as described under the criteria in Listing 7.02 (CD Part 7).
2. Decreased white blood cells (granulocytopenia), as described under the criteria in Listing 7.15 (CD Part 7).

3. Decreased platelets (thrombocytopenia), as described under the criteria in Listing 107.06 or 7.06 (CD Part 7).

Ⓗ Neurological manifestations of HIV infection such as brain damage (HIV encephalopathy) or nerve damage (peripheral neuropathy), as described under the criteria in the nervous system listings (listings with a 111 prefix, CD Part 11), or resulting in one or more of the following:

1. Loss of previously acquired, or marked delay in achieving, developmental milestones or intellectual ability (including the sudden acquisition of a new learning disability).
2. Decreased brain growth, as demonstrated by a cessation of head growth (acquired microcephaly) or by brain atrophy.
3. Progressive movement abnormalities (motor dysfunction) resulting in:
   - significantly decreased ability to stand (station) and walk (gait), or
   - significantly decreased ability to make large (gross) and small (dexterous) movements with the fingers.

Ⓘ 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 established growth curve (on standard growth charts) that persists for two months or longer.
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 established growth curve (on standard growth charts) that persists for two months or longer.
3. Involuntary weight loss greater than 10% of baseline that persists for two months or longer.
4. Decreased growth as described under the criteria in growth impairment listings (listings with a 100 prefix, CD Part 1).

Ⓙ Diarrhea, lasting for one month or longer, resistant to treatment and requiring intravenous fluids (hydration), intravenous feeding (alimentation), or tube feeding.

Ⓚ Cardiomyopathy, as described under the criteria in the listings dealing with heart disease (listings with a 104 prefix, CD Part 4) or the neurological listing dealing with strokes in adults (11.04, CD Part 11).

Ⓛ 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.

Ⓜ Nephropathy as described under the criteria in the listings for kidney disease (listings with a 106 prefix, CD Part 6).

Ⓝ One or more of the following infections (other than described in parts Ⓐ through Ⓛ, above), resistant to treatment or requiring hospitalization or intravenous treatment three or more times in one year. Or evaluate abnormalities caused by the infection (sequelae) under the criteria for the affected body system, including:

1. Infection affecting the whole body (sepsis).
2. Infection of the meninges covering the brain or spinal cord (meningitis).
3. Infection of the lungs (pneumonia).
4. Infection of a joint (septic arthritis).
5. Infection of the heart (endocarditis).
6. Infection of a sinus (sinusitis) documented by x-rays.

Ⓞ Any other manifestations of HIV infection. Such manifestations can include those listed in parts Ⓐ through Ⓝ that don't have findings required by those parts of the listing—for example, oral candidiasis not meeting the criteria in part Ⓕ, or diarrhea not meeting the criteria in part Ⓙ. Also, other manifestations of HIV infection can be considered, such as white lesions on the sides of the tongue (oral hairy leukoplakia) or an enlarged liver (hepatomegaly). Whatever the manifestation, it must result in one of the following:

1. For children from birth to attainment of age one, at least one of the criteria in parts Ⓐ through Ⓔ of Listing 112.12.
2. For children age one to attainment of age three, at least one of the appropriate age-group criteria in part Ⓑ1 of Listing 112.02.
3. For children age three to attainment of age 18, at least two of the appropriate age-group criteria in part Ⓑ2 of Listing 112.02.

15. Listing 14.09: Inflammatory Arthritis (Adults)

Rheumatoid or inflammatory arthritis is a disease of the immune system that causes inflammation: tenderness, swelling, and pain in the tissues surrounding the joints (see general discussion in Section B, above). The cause of the inflammation does not matter, and inflammation caused by other conditions, for example psoriasis, can also qualify a person under this listing. Both rheumatoid and psoriatic arthritis most often affect the small joints of the hands. Rheumatoid arthritis (RA) is the most common inflammatory joint disorder the SSA sees.

Your joints may be red, warm, and swollen as a result of inflammation. You may suffer stiffness of joints, joint effusions, weight loss, or fever, but none of these are specifically required by the listing.

For purposes of the following discussion, the term "major joints" refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist/hand, and ankle/foot.

a. Listing Level Severity

For your condition to be severe enough to meet this listing, your condition must satisfy Ⓐ, Ⓑ, Ⓒ, Ⓓ, or Ⓔ, below.

Ⓐ You must have a history of joint pain, swelling, and tenderness. Also, on current physical examination, you must show signs of joint inflammation or deformity in two or more major joints, making you either unable to walk effectively or unable to perform fine and gross movements effectively.

Part Ⓐ can be satisfied by ineffective use of either the upper or lower extremities as follows.

Walking. Extreme limitation in your ability to walk means you must be unable to sustain a reasonable walking pace over a sufficient distance to be able to carry out your activities of daily living. You would not have the ability to travel without a companion's assistance to and from a place of employment or school. More specifically, some examples of ineffective ambulation given by the SSA include:

- inability to walk without the use of a walker
- inability to walk without the use of two crutches or two canes
- inability to walk a block at a reasonable pace on rough or uneven surfaces
- inability to use standard public transportation
- inability to carry out ordinary activities involving walking, such as shopping and banking, or
- inability to climb a few steps at a reasonable pace with the use of a single handrail.

The listing does not require you to be completely unable to walk in all circumstances. For example, your ability to walk about your home without someone else's help or the use of assistive devices does not, in and of itself, mean you cannot qualify under the listing. To qualify, you must have serious difficulty in starting, sustaining, or completing activities. Also understand that the use of only one crutch or cane would not necessarily restrict you from qualifying under the listing, provided that your functional limitations are severe enough. In addition, the SSA recognizes that people who cannot walk effectively might be able to stand without assistive devices. Therefore, your ability to stand without assistance would not disqualify you under the listing.

Performance of fine and gross movements. To use your upper extremities effectively in carrying out your activities of daily living, you must be able to perform such functions as reaching, pushing, pulling, grasping, and fingering. Examples of being unable to effectively perform fine and gross movements include, but are not limited to, being unable to prepare a simple meal and feed yourself, to take care of personal hygiene, to sort and handle papers or files, or to place files in a file cabinet at or above waist level.

To qualify under part Ⓐ, it is not necessary that you have a *total* inability to use your upper extremities. You must, however, have serious difficulty in starting, sustaining, or completing activities.

Ⓑ Ankylosing spondylitis or other spondyloarthropathy. This diagnosis must be established by findings of unilateral or bilateral sacroiliitis, as demonstrated by the presence of erosion or fusion (ankylosis) of your sacroiliac joints. These abnormalities must show up on imaging tests such as plain x-rays, an MRI scan, or a CT scan.

Additionally, part Ⓑ requires you to have both 1 and 2, below.

1. A history of back pain, tenderness, and stiffness.
2. Findings on physical examination of ankylosis (fixation) of your dorsolumbar or cervical spine at 45 degrees or more of flexion measured from the vertical position (zero degrees).

Note that to satisfy part Ⓑ, you don't have to be relying on an assistive device like a cane in order to walk. A person with the symptoms and other findings in part Ⓑ has their gaze fixed downward at a sharp angle, which produces extreme functional limitation by restricting vision ahead, above, and to the side. Walking will be extremely limited, so it is not necessary for the SSA to spell out that fact under part Ⓑ.

Ⓒ An impairment as described under the criteria in Listing 14.02Ⓐ.

Ⓓ Inflammatory arthritis, with signs of peripheral joint inflammation on current examination. However, you can have less joint involvement than in part Ⓐ and fewer extra-articular features than in part Ⓒ.

Additionally, part D requires you to have both 1 and 2, below:

1. Significant, documented constitutional symptoms and signs (e.g., fatigue, fever, malaise, weight loss).
2. Involvement of two or more organs/body systems. At least one of the organs/body systems must be involved to at least a moderate level of severity. Moderate means more than slight or mild, as a matter of medical judgment.

Ⓔ Inflammatory spondylitis of any cause. Your deformity can be less than in part Ⓑ, and you don't have to have extra-articular abnormalities so severe that they satisfy some other listing as required in part Ⓒ. However, you must have signs of sacroiliitis in at least one sacroiliac joint and the extra-articular features described in 14.09Ⓓ 1 and 2.

b. Residual Functional Capacity

Upper Extremity Dysfunction: The SSA needs information regarding how well you can use your upper extremities—specifically, the extent of your ability to push, pull, lift, carry, and grasp objects and do small movements with your fingers (fine manipulations). Think of all the things you cannot do because of pain, deformity, or fatigue. Can you pick up coins? Easily grasp and turn doorknobs? Open jars? If you were unable to perform prior work because of arthritis, exactly how did the arthritis interfere? Specific examples of why you can no longer perform the job are much better than vague generalizations such as, "I was in pain" or "My arthritis bothered me." For instance, how much weight can you lift and carry? Did your pain limit your use of hand controls that were necessary for you to work? Exactly how? Include environmental factors; for example, arthritis that is tolerable working in normal temperatures might be limiting in the cold. If you have significant arthritis in your shoulder, pain will probably limit the amount of overhead work you can do. Shoulder, elbow, or hand arthritis will limit how much pushing and pulling you can do.

Note that the use of an assistive device such as a cane ties up the use of an arm and hand. So if you require a cane to walk, the SSA cannot refer you to jobs requiring you to lift and carry with both arms while walking.

Lower Extremity Dysfunction: In evaluating your RFC, the SSA must determine how long you can stand and walk on arthritic joints. Let the SSA know if the arthritis is severe enough that you can't stand or walk for most of a workday; have your treating doctor provide supportive statements. For the SSA to claim that you can perform light, medium, or heavy work, you must be able to walk or stand six to eight hours a day. Significant arthritis in a major joint of a lower extremity would prevent such standing or walking. Even if your hands and arms are unaffected by the arthritis, you'll be restricted to sedentary work. If you are older and have a limited education, these restrictions may mean that you'll be awarded benefits on the basis of your RFC.

If you had an arthritic hip, knee, or ankle joint replaced with an artificial one, see the RFC comments under Listing 1.03.

Other Functional Limitations: The limitations you face due to the involvement of organs other than your joints must be evaluated on a case-by-case basis. If your back is involved, it is extremely important to document your limitations in bending, sitting, standing, and walking. Also, back pain and back stiffness can be pointed out as separate limitations.

16. Listing 114.09: Inflammatory Arthritis (Children)

See comments under adult Listing 14.09.

a. Listing Level Severity

For the child's condition to be severe enough to meet this listing, the child's condition must satisfy Ⓐ, Ⓑ, Ⓒ, Ⓓ, or Ⓔ, below.

Ⓐ The child must have a history of joint pain, swelling, and tenderness. A current physical examination must reveal signs of joint inflammation or deformity in two or more major joints. This inflammation or deformity must result in the child's being unable to walk effectively or perform fine and gross movements.

Walking: For children who are too young to be expected to walk independently, the SSA considers their function in terms of how well they can perform age-appropriate activities with their lower extremities. For such children, an extreme level of limitation means skills or performance at no greater than one-half of age-appropriate expectations based on an overall developmental assessment rather than on one or two isolated skills.

Older children who qualify would not have the ability to travel without a companion's assistance to and from a place of employment or school. More specifically, some examples of ineffective ambulation given by the SSA include the child's:

- inability to walk without the use of a walker
- inability to walk without the use of two crutches or two canes
- inability to walk a block at a reasonable, age-appropriate pace on rough or uneven surfaces
- inability to use standard public transportation
- inability to carry out ordinary age-appropriate activities involving walking, such as shopping and school activities, or
- inability to climb a few steps at a reasonable pace with the use of a single handrail.

The listing does not require that the child be completely unable to walk in all circumstances. For example, the child's ability to walk around the house (or short distances at school) without someone else's help or the use of assistive devices does not, in and of itself, mean the child cannot qualify under the listing. The listing requires that the child have serious difficulty in starting, sustaining, or completing activities. Also understand that the use of only one crutch or cane would not necessarily restrict the child from qualifying under the listing, provided that the child's functional limitations are severe enough. In addition, the SSA recognizes that people who cannot walk effectively might be able to stand without assistive devices. Therefore, the child's ability to stand without assistance would not disqualify him or her under the listing.

Performance of fine and gross movements: In cases of very young children, the SSA looks at the limitations in the child's ability to perform age-appropriate activities involving the upper extremities. To determine whether such children have an extreme limitation, see the limitations for persistent motor dysfunction for infants and young children described in Listing 110.07Ⓐ.

For an older child to use his or her upper extremities effectively in carrying out age-appropriate activities of daily living, she must be able to perform age-appropriate functions like reaching, pushing, pulling, grasping, and fingering. Therefore, in older children, examples of inability to effectively perform fine and gross movements include being to prepare a simple meal and feed themselves, to take care of personal hygiene or to sort and handle papers or files (depending on which activities are age-appropriate). To qualify under part Ⓐ, it is not necessary that the child be *totally* unable to use her upper extremities. The requirement is that he or she have serious difficulty in starting, sustaining, or completing age-appropriate activities.

Ⓑ Ankylosing spondylitis or other spondyloarthropathy. This diagnosis must be established by findings of unilateral or bilateral sacroiliitis, as shown by erosion or fusion (ankylosis) of the sacroiliac joints. These abnormalities must appear on imaging tests such as plain x-rays, an MRI scan, or a CT scan.

Additionally, part Ⓑ requires the child to have both 1 and 2, below.

1. A history of back pain, tenderness, and stiffness.
2 Findings on physical examination of ankylosis (fixation) of the dorsolumbar or cervical spine at

45 degrees or more of flexion measured from the vertical position (zero degrees).

Note that to satisfy part Ⓑ, the child need not require an assistive device like a cane in order to walk. A person with the symptoms and other findings in part Ⓑ has their gaze fixed downward at a sharp angle, which produces extreme functional limitation by restricting vision ahead, above, and to the side. Walking will be extremely limited, so it is not necessary for the SSA to spell out that fact under part Ⓑ.

Ⓒ An impairment as described under the criteria in Listing 114.02Ⓐ.

Ⓓ Inflammatory arthritis, with signs of peripheral joint inflammation on current examination. However, the child can have less joint involvement than in part Ⓐ and fewer extra-articular features than in part Ⓒ.

Additionally, part Ⓓ requires that the child have both 1 and 2, below:

1. Significant, documented constitutional symptoms and signs (such as fatigue, fever, malaise, or weight loss).
2. Involvement of two or more organs/body systems. At least one of the organs/body systems must be involved to at least a moderate level of severity. Moderate means more than slight or mild, as a matter of medical judgment.

Ⓔ Inflammatory spondylitis of any cause. The child can have lesser deformity than in part Ⓑ, and doesn't have to have extra-articular abnormalities so severe that the child satisfies some other listing as required in part Ⓒ. However, the child must show signs of sacroiliitis in at least one sacroiliac joint as well as the extra-articular features described in 14.09Ⓓ 1 and 2. ■