Social Security Online Code of Federal Regulations

CFR 20 Title Page

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

The body system listings in parts A and B of the Listing of Impairments will no longer be effective on the following dates unless extended by the Commissioner or revised and promulgated again.

1. Growth Impairment (100.00): July 2, 2007.

2. Musculoskeletal System (1.00 and 101.00): February 19, 2009.

3. Special Senses and Speech (2.00 and 102.00): February 20, 2015.

4. Respiratory System (3.00 and 103.00): July 2, 2007.

5. Cardiovascular System (4.00 and 104.00): January 13, 2011.

6. Digestive System (5.00 and 105.00): July 2, 2007.

7. Genitourinary Impairments (6.00 and 106.00): September 6, 2013.

8. Hematological Disorders (7.00 and 107.00): July 2, 2007.

9. Skin Disorders (8.00 and 108.00): July 9, 2012.

10. Endocrine System (9.00 and 109.00): July 2, 2007.

11. Impairments That Affect Multiple Body Systems (10.00 and 110.00): October 31, 2013

12. Neurological (11.00 and 111.00): July 2, 2007.

13. Mental Disorders (12.00 and 112.00): July 2, 2007.

14. Malignant Neoplastic Diseases (13.00 and 113.00): December 15, 2009.

15. Immune System (14.00 and 114.00): July 2, 2007.

Part A

Criteria applicable to individuals age 18 and over and to children under age 18 where criteria

are appropriate.

Sec.

1.00 Musculoskeletal System

A. *Disorders of the musculoskeletal system* may result from hereditary, congenital, or acquired pathologic processes. Impairments may result from infectious, inflammatory, or degenerative processes, traumatic or developmental events, or neoplastic, vascular, or toxic/metabolic diseases.

B. *Loss of function.*

1. *General.* Under this section, loss of function may be due to bone or joint deformity or destruction from any cause; miscellaneous disorders of the spine with or without radiculopathy or other neurological deficits; amputation; or fractures or soft tissue injuries, including burns, requiring prolonged periods of immobility or convalescence. For inflammatory arthritides that may result in loss of function because of inflammatory peripheral joint or axial arthritis or sequelae, or because of extra-articular features, see 14.00B6. Impairments with neurological causes are to be evaluated under 11.00ff.

2. How We Define Loss of Function in These Listings

a. *General.* Regardless of the cause(s) of a musculoskeletal impairment, functional loss for purposes of these listings is defined as the inability to ambulate effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment, or the inability to perform fine and gross movements effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment. The inability to ambulate effectively or the inability to perform fine and gross movements effectively must have lasted, or be expected to last, for at least 12 months. For the purposes of these criteria, consideration of the ability to perform these activities must be from a physical standpoint alone. When there is an inability to perform these activities due to a mental impairment, the criteria in 12.00ff are to be used. We will determine whether an individual can ambulate effectively or can perform fine and gross movements effectively based on the medical and other evidence in the case record, generally without developing additional evidence about the individual's ability to perform the specific activities listed as examples in 1.00B2b(2) and 1.00B2c.

b. What We Mean by Inability To Ambulate Effectively

(1) *Definition.* Inability to ambulate effectively means an extreme limitation of the ability to walk; *i.e.*, an impairment(s) that interferes very seriously with the individual's ability to independently initiate, sustain, or complete activities. Ineffective ambulation is defined generally as having insufficient lower extremity functioning (see 1.00J) to permit independent ambulation without the use of a hand-held assistive device(s) that limits the functioning of both upper extremities. (Listing 1.05C is an exception to this general definition because the individual has the use of only one upper extremity due to amputation of a hand.)

(2) *To ambulate effectively,* individuals must be capable of sustaining a reasonable walking pace over a sufficient distance to be able to carry out activities of daily living. They must have the ability to travel without companion assistance to and from a place of employment or school. Therefore, examples of ineffective ambulation include, but are not limited to, the inability to walk without the use of a walker, two crutches or two canes, the inability to walk a block at a reasonable pace on rough or uneven surfaces, the inability to use standard public transportation, the inability to carry out routine ambulatory activities, such as shopping and banking, and the inability to climb a few steps at a reasonable pace with the use of a single hand rail. The ability to walk independently about one's home without the use of assistive devices does not, in and of itself, constitute effective ambulation.

c. *What we mean by inability to perform fine and gross movements effectively.* Inability to perform fine and gross movements effectively means an extreme loss of function of both upper extremities; *i.e.*, an impairment(s) that interferes very seriously with the individual's ability to independently initiate, sustain, or complete activities. To use their upper extremities effectively, individuals must be capable of sustaining such functions as reaching, pushing, pulling, grasping, and fingering to be able to carry out activities of daily living. Therefore, examples of inability to perform fine and gross movements effectively include, but are not limited to, the inability to prepare a simple meal and feed oneself, the inability to take care of personal hygiene, the inability to sort and handle papers or files, and the inability to place files in a file cabinet at or above waist level.

d. *Pain or other symptoms.* Pain or other symptoms may be an important factor contributing to

functional loss. In order for pain or other symptoms to be found to affect an individual's ability to perform basic work activities, medical signs or laboratory findings must show the existence of a medically determinable impairment(s) that could reasonably be expected to produce the pain or other symptoms. The musculoskeletal listings that include pain or other symptoms among their criteria also include criteria for limitations in functioning as a result of the listed impairment, including limitations caused by pain. It is, therefore, important to evaluate the intensity and persistence of such pain or other symptoms carefully in order to determine their impact on the individual's functioning under these listings. See also §§404.1525(f) and 404.1529 of this part, and §§416.925(f) and 416.929 of part 416 of this chapter.

C. Diagnosis and Evaluation

1. *General.* Diagnosis and evaluation of musculoskeletal impairments should be supported, as applicable, by detailed descriptions of the joints, including ranges of motion, condition of the musculature (e.g., weakness, atrophy), sensory or reflex changes, circulatory deficits, and laboratory findings, including findings on x-ray or other appropriate medically acceptable imaging. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Purchase of certain medically acceptable imaging.* While any appropriate medically acceptable imaging is useful in establishing the diagnosis of musculoskeletal impairments, some tests, such as CAT scans and MRIs, are quite expensive, and we will not routinely purchase them. Some, such as myelograms, are invasive and may involve significant risk. We will not order such tests. However, when the results of any of these tests are part of the existing evidence in the case record we will consider them together with the other relevant evidence.

3. *Consideration of electrodiagnostic procedures.* Electrodiagnostic procedures may be useful in establishing the clinical diagnosis, but do not constitute alternative criteria to the requirements of 1.04.

D. *The physical examination* must include a detailed description of the rheumatological, orthopedic, neurological, and other findings appropriate to the specific impairment being evaluated. These physical findings must be determined on the basis of objective observation during the examination and not simply a report of the individual's allegation; e.g., "He says his leg is weak, numb." Alternative testing methods should be used to verify the abnormal findings; e.g., a seated straight-leg raising test in addition to a supine straight-leg raising test. Because abnormal physical findings may be intermittent, their presence over a period of time must be established by a record of ongoing management and evaluation. Care must be taken to ascertain that the reported examination findings are consistent with the individual's daily activities.

E. Examination of the Spine

1. *General.* Examination of the spine should include a detailed description of gait, range of motion of the spine given quantitatively in degrees from the vertical position (zero degrees) or, for straight-leg raising from the sitting and supine position (zero degrees), any other

appropriate tension signs, motor and sensory abnormalities, muscle spasm, when present, and deep tendon reflexes. Observations of the individual during the examination should be reported; e.g., how he or she gets on and off the examination table. Inability to walk on the heels or toes, to squat, or to arise from a squatting position, when appropriate, may be considered evidence of significant motor loss. However, a report of atrophy is not acceptable as evidence of significant motor loss without circumferential measurements of both thighs and lower legs, or both upper and lower arms, as appropriate, at a stated point above and below the knee or elbow given in inches or centimeters. Additionally, a report of atrophy should be accompanied by measurement of the strength of the muscle(s) in question generally based on a grading system of 0 to 5, with 0 being complete loss of strength and 5 being maximum strength. A specific description of atrophy of hand muscles is acceptable without measurements of atrophy but should include measurements of grip and pinch strength.

2. *When neurological abnormalities persist.* Neurological abnormalities may not completely subside after treatment or with the passage of time. Therefore, residual neurological abnormalities that persist after it has been determined clinically or by direct surgical or other observation that the ongoing or progressive condition is no longer present will not satisfy the required findings in 1.04. More serious neurological deficits (paraparesis, paraplegia) are to be evaluated under the criteria in 11.00ff.

F. *Major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

G. *Measurements of joint motion* are based on the techniques described in the chapter on the extremities, spine, and pelvis in the current edition of the "Guides to the Evaluation of Permanent Impairment" published by the American Medical Association.

H. Documentation

1. *General.* Musculoskeletal impairments frequently improve with time or respond to treatment. Therefore, a longitudinal clinical record is generally important for the assessment of severity and expected duration of an impairment unless the claim can be decided favorably on the basis of the current evidence.

2. *Documentation of medically prescribed treatment and response.* Many individuals, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever evidence of such treatment is available it must be considered.

3. *When there is no record of ongoing treatment.* Some individuals will not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In such cases, evaluation will be made on the basis of the current objective medical evidence and other available evidence, taking into consideration the individual's medical history, symptoms, and medical source opinions. Even though an individual who does not receive treatment may not be able to show an impairment that meets the criteria of one of the musculoskeletal listings, the individual may have an impairment(s)

equivalent in severity to one of the listed impairments or be disabled based on consideration of his or her residual functional capacity (RFC) and age, education and work experience.

4. *Evaluation when the criteria of a musculoskeletal listing are not met.* These listings are only examples of common musculoskeletal disorders that are severe enough to prevent a person from engaging in gainful activity. Therefore, in any case in which an individual has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will consider medical equivalence. (See §§404.1526 and 416.926.) Individuals who have an impairment(s) with a level of severity that does not meet or equal the criteria of the musculoskeletal listings may or may not have the RFC that would enable them to engage in substantial gainful activity. Evaluation of the impairment(s) of these individuals should proceed through the final steps of the sequential evaluation process in §§404.1520 and 416.920 (or, as appropriate, the steps in the medical improvement review standard in §§404.1594 and 416.994).

I. Effects of Treatment

1. *General.* Treatments for musculoskeletal disorders may have beneficial effects or adverse side effects. Therefore, medical treatment (including surgical treatment) must be considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the disorder, and in terms of any side effects that may further limit the individual.

2. *Response to treatment.* Response to treatment and adverse consequences of treatment may vary widely. For example, a pain medication may relieve an individual's pain completely, partially, or not at all. It may also result in adverse effects, e.g., drowsiness, dizziness, or disorientation, that compromise the individual's ability to function. Therefore, each case must be considered on an individual basis, and include consideration of the effects of treatment on the individual's ability to function.

3. *Documentation.* 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 finding regarding the impact of treatment must be based on a sufficient period of treatment to permit proper consideration or judgment about future functioning.

J. Orthotic, Prosthetic, or Assistive Devices

1. *General.* Consistent with clinical practice, individuals with musculoskeletal impairments may be examined with and without the use of any orthotic, prosthetic, or assistive devices as explained in this section.

2. *Orthotic devices.* Examination should be with the orthotic device in place and should include an evaluation of the individual's maximum ability to function effectively with the orthosis. It is unnecessary to routinely evaluate the individual's ability to function without the orthosis in place. If the individual has difficulty with, or is unable to use, the orthotic device, the medical basis for the difficulty should be documented. In such cases, if the impairment involves a lower extremity or extremities, the examination should include information on the individual's ability to ambulate effectively without the device in place unless contraindicated by the medical judgment of a physician who has treated or examined the individual.

3. *Prosthetic devices.* Examination should be with the prosthetic device in place. In amputations involving a lower extremity or extremities, it is unnecessary to evaluate the individual's ability to walk without the prosthesis in place. However, the individual's medical ability to use a prosthesis to ambulate effectively, as defined in 1.00B2b, should be evaluated. The condition of the stump should be evaluated without the prosthesis in place.

4. *Hand-held assistive devices.* When an individual with an impairment involving a lower extremity or extremities uses a hand-held assistive device, such as a cane, crutch or walker, examination should be with and without the use of the assistive device unless contraindicated by the medical judgment of a physician who has treated or examined the individual. The individual's ability to ambulate with and without the device provides information as to whether, or the extent to which, the individual is able to ambulate without assistance. The medical basis for the use of any assistive device (e.g., instability, weakness) should be documented. The requirement to use a hand-held assistive device may also impact on the individual's functional capacity by virtue of the fact that one or both upper extremities are not available for such activities as lifting, carrying, pushing, and pulling.

K. *Disorders of the spine,* listed in 1.04, result in limitations because of distortion of the bony and ligamentous architecture of the spine and associated impingement on nerve roots (including the cauda equina) or spinal cord. Such impingement on nerve tissue may result from a herniated nucleus pulposus, spinal stenosis, arachnoiditis, or other miscellaneous conditions. Neurological abnormalities resulting from these disorders are to be evaluated by referral to the neurological listings in 11.00ff, as appropriate. (See also 1.00B and E.)

1. *Herniated nucleus pulposus* is a disorder frequently associated with the impingement of a nerve root. Nerve root compression results in a specific neuro-anatomic distribution of symptoms and signs depending upon the nerve root(s) compromised.

2. Spinal Arachnoiditis

a. *General.* Spinal arachnoiditis is a condition characterized by adhesive thickening of the arachnoid which may cause intermittent ill-defined burning pain and sensory dysesthesia, and may cause neurogenic bladder or bowel incontinence when the cauda equina is involved.

b. *Documentation.* Although the cause of spinal arachnoiditis is not always clear, it may be associated with chronic compression or irritation of nerve roots (including the cauda equina) or the spinal cord. For example, there may be evidence of spinal stenosis, or a history of spinal trauma or meningitis. Diagnosis must be confirmed at the time of surgery by gross description, microscopic examination of biopsied tissue, or by findings on appropriate medically acceptable imaging. Arachnoiditis is sometimes used as a diagnosis when such a diagnosis is unsupported by clinical or laboratory findings. Therefore, care must be taken to ensure that the diagnosis is documented as described in 1.04B. Individuals with arachnoiditis, particularly when it involves the lumbosacral spine, are generally unable to sustain any given position or posture for more than a short period of time due to pain.

3. *Lumbar spinal stenosis* is a condition that may occur in association with degenerative processes, or as a result of a congenital anomaly or trauma, or in association with Paget's disease of the bone. *Pseudoclaudication,* which may result from lumbar spinal stenosis, is manifested as pain and weakness, and may impair ambulation. Symptoms are usually bilateral, in the low back, buttocks, or thighs, although some individuals may experience only

leg pain and, in a few cases, the leg pain may be unilateral. The pain generally does not follow a particular neuro-anatomical distribution, *i.e.*, it is distinctly different from the radicular type of pain seen with a herniated intervertebral disc, is often of a dull, aching quality, which may be described as "discomfort" or an "unpleasant sensation," or may be of even greater severity, usually in the low back and radiating into the buttocks region bilaterally. The pain is provoked by extension of the spine, as in walking or merely standing, but is reduced by leaning forward. The distance the individual has to walk before the pain comes on may vary. Pseudoclaudication differs from peripheral vascular claudication in several ways. Pedal pulses and Doppler examinations are unaffected by pseudoclaudication. Leg pain resulting from peripheral vascular claudication involves the calves, and the leg pain in vascular claudication is ordinarily more severe than any back pain that may also be present. An individual with vascular claudication will experience pain after walking the same distance time after time, and the pain will be relieved quickly when walking stops.

4. *Other miscellaneous conditions* that may cause weakness of the lower extremities, sensory changes, areflexia, trophic ulceration, bladder or bowel incontinence, and that should be evaluated under 1.04 include, but are not limited to, osteoarthritis, degenerative disc disease, facet arthritis, and vertebral fracture. Disorders such as spinal dysrhaphism (e.g., spina bifida), diastematomyelia, and tethered cord syndrome may also cause such abnormalities. In these cases, there may be gait difficulty and deformity of the lower extremities based on neurological abnormalities, and the neurological effects are to be evaluated under the criteria in 11.00ff.

L. *Abnormal curvatures of the spine.* Abnormal curvatures of the spine (specifically, scoliosis, kyphosis and kyphoscoliosis) can result in impaired ambulation, but may also adversely affect functioning in body systems other than the musculoskeletal system. For example, an individual's ability to breathe may be affected; there may be cardiac difficulties (e.g., impaired myocardial function); or there may be disfigurement resulting in withdrawal or isolation. When there is impaired ambulation, evaluation of equivalence may be made by reference to 14.09A. When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 14.09B. When there is respiratory or cardiac involvement or an associated mental disorder, evaluation may be made under 3.00ff, 4.00ff, or 12.00ff, as appropriate. Other consequences should be evaluated according to the listing for the affected body system.

M. *Under continuing surgical management,* as used in 1.07 and 1.08, refers to surgical procedures and any other associated treatments related to the efforts directed toward the salvage or restoration of functional use of the affected part. It may include such factors as post-surgical procedures, surgical complications, infections, or other medical complications, related illnesses, or related treatments that delay the individual's attainment of maximum benefit from therapy. When burns are not under continuing surgical management, see 8.00F.

N. *After maximum benefit from therapy has been achieved* in situations involving fractures of an upper extremity (1.07), or soft tissue injuries (1.08), *i.e.*, there have been no significant changes in physical findings or on appropriate medically acceptable imaging for any 6-month period after the last definitive surgical procedure or other medical intervention, evaluation must be made on the basis of the demonstrable residuals, if any. A finding that 1.07 or 1.08 is met must be based on a consideration of the symptoms, signs, and laboratory findings associated with recent or anticipated surgical procedures and the resulting recuperative periods, including any related medical complications, such as infections, illnesses, and therapies which impede or delay the efforts toward restoration of function. Generally, when there has been no surgical

or medical intervention for 6 months after the last definitive surgical procedure, it can be concluded that maximum therapeutic benefit has been reached. Evaluation at this point must be made on the basis of the demonstrable residual limitations, if any, considering the individual's impairment-related symptoms, signs, and laboratory findings, any residual symptoms, signs, and laboratory findings associated with such surgeries, complications, and recuperative periods, and other relevant evidence.

O. *Major function of the face and head,* for purposes of listing 1.08, relates to impact on any or all of the activities involving vision, hearing, speech, mastication, and the initiation of the digestive process.

P. *When surgical procedures have been performed,* documentation should include a copy of the operative notes and available pathology reports.

Q. *Effects of obesity.* Obesity is a medically determinable impairment that is often associated with disturbance of the musculoskeletal system, and disturbance of this system can be a major cause of disability in individuals with obesity. The combined effects of obesity with musculoskeletal impairments can be greater than the effects of each of the impairments considered separately. Therefore, when determining whether an individual with obesity has a listing-level impairment or combination of impairments, and when assessing a claim at other steps of the sequential evaluation process, including when assessing an individual's residual functional capacity, adjudicators must consider any additional and cumulative effects of obesity.

1.01 Category of Impairments, Musculoskeletal

1.02 *Major dysfunction of a joint(s) (due to any cause):* Characterized by gross anatomical deformity (e.g., subluxation, contracture, bony or fibrous ankylosis, instability) and chronic joint pain and stiffness with signs of limitation of motion or other abnormal motion of the affected joint(s), and findings on appropriate medically acceptable imaging of joint space narrowing, bony destruction, or ankylosis of the affected joint(s). With:

A. Involvement of one major peripheral weight-bearing joint (*i.e.*, hip, knee, or ankle), resulting in inability to ambulate effectively, as defined in 1.00B2b;

or

B. Involvement of one major peripheral joint in each upper extremity (*i.e.*, shoulder, elbow, or wrist-hand), resulting in inability to perform fine and gross movements effectively, as defined in 1.00B2c.

1.03 *Reconstructive surgery or surgical arthrodesis of a major weight-bearing joint,* with inability to ambulate effectively, as defined in 1.00B2b, and return to effective ambulation did not occur, or is not expected to occur, within 12 months of onset.

1.04 *Disorders of the spine* (e.g., herniated nucleus pulposus, spinal arachnoiditis, spinal stenosis, osteoarthritis, degenerative disc disease, facet arthritis, vertebral fracture), resulting in compromise of a nerve root (including the cauda equina) or the spinal cord. With:

A. Evidence of nerve root compression characterized by neuro-anatomic distribution of pain,

limitation of motion of the spine, motor loss (atrophy with associated muscle weakness or muscle weakness) accompanied by sensory or reflex loss and, if there is involvement of the lower back, positive straight-leg raising test (sitting and supine);

or

B. Spinal arachnoiditis, confirmed by an operative note or pathology report of tissue biopsy, or by appropriate medically acceptable imaging, manifested by severe burning or painful dysesthesia, resulting in the need for changes in position or posture more than once every 2 hours;

or

C. Lumbar spinal stenosis resulting in pseudoclaudication, established by findings on appropriate medically acceptable imaging, manifested by chronic nonradicular pain and weakness, and resulting in inability to ambulate effectively, as defined in 1.00B2b.

1.05 *Amputation (due to any cause).*

A. Both hands; or

or

B. One or both lower extremities at or above the tarsal region, with stump complications resulting in medical inability to use a prosthetic device to ambulate effectively, as defined in 1.00B2b, which have lasted or are expected to last for at least 12 months;

or

C. One hand and one lower extremity at or above the tarsal region, with inability to ambulate effectively, as defined in 1.00B2b; OR

D. Hemipelvectomy or hip disarticulation.

1.06 *Fracture of the femur, tibia, pelvis, or one or more of the tarsal bones.* With:

A. Solid union not evident on appropriate medically acceptable imaging and not clinically solid;

and

B. Inability to ambulate effectively, as defined in 1.00B2b, and return to effective ambulation did not occur or is not expected to occur within 12 months of onset.

1.07 *Fracture of an upper extremity* with nonunion of a fracture of the shaft of the humerus, radius, or ulna, under continuing surgical management, as defined in 1.00M, directed toward restoration of functional use of the extremity, and such function was not restored or expected to be restored within 12 months of onset.

1.08 *Soft tissue injury (e.g., burns)* of an upper or lower extremity, trunk, or face and head,

under continuing surgical management, as defined in 1.00M, directed toward the salvage or restoration of major function, and such major function was not restored or expected to be restored within 12 months of onset. Major function of the face and head is described in 1.00O.

2.00 Special Senses and Speech

A. *How do we evaluate visual disorders?*

1. *What are visual disorders?* Visual disorders are abnormalities of the eye, the optic nerve, the optic tracts, or the brain that may cause a loss of visual acuity or visual fields. A loss of visual acuity limits your ability to distinguish detail, read, or do fine work. A loss of visual fields limits your ability to perceive visual stimuli in the peripheral extent of vision.

2. *How do we define statutory blindness?* Statutory blindness is blindness as defined in sections 216(i)(1) and 1614(a)(2) of the Social Security Act (the Act). The Act defines blindness as visual acuity of 20/200 or less in the better eye with the use of a correcting lens. We use your best-corrected visual acuity for distance in the better eye when we determine if this definition is met. The Act also provides that an eye that has a visual field limitation such that the widest diameter of the visual field subtends an angle no greater than 20 degrees is considered as having visual acuity of 20/200 or less. You have statutory blindness only if your visual disorder meets the criteria of 2.02 or 2.03A. You do not have statutory blindness if your visual disorder medically equals the criteria of 2.02 or 2.03A, or if it meets or medically equals 2.03B, 2.03C, or 2.04. If your visual disorder medically equals the criteria of 2.02 or 2.03A, or if it meets or medically equals 2.03B, 2.03C, or 2.04, we will find that you have a disability if your visual disorder also meets the duration requirement.

3. *What evidence do we need to establish statutory blindness under title XVI?* For title XVI, the only evidence we need to establish statutory blindness is evidence showing that your visual acuity in your better eye or your visual field in your better eye meets the criteria in 2.00A2, provided that those measurements are consistent with the other evidence in your case record. We do not need to document the cause of your blindness. Also, there is no duration requirement for statutory blindness under title XVI (see §§416.981 and 416.983).

4. *What evidence do we need to evaluate visual disorders, including those that result in statutory blindness under title II?*

a. To evaluate your visual disorder, we usually need a report of an eye examination that includes measurements of the best-corrected visual acuity or the extent of the visual fields, as appropriate. If there is a loss of visual acuity or visual fields, the cause of the loss must be documented. A standard eye examination will usually reveal the cause of any visual acuity loss. An eye examination can also reveal the cause of some types of visual field deficits. If the eye examination does not reveal the cause of the visual loss, we will request the information that was used to establish the presence of the visual disorder.

b. A cortical visual disorder is a disturbance of the posterior visual pathways or occipital lobes of the brain in which the visual system does not interpret what the eyes are seeing. It may result from such causes as traumatic brain injury, stroke, cardiac arrest, near drowning, a central nervous system infection such as meningitis or encephalitis, a tumor, or surgery. It can be temporary or permanent, and the amount of visual loss can vary. It is possible to have a cortical visual disorder and not have any abnormalities observed in a standard eye

examination. Therefore, a diagnosis of a cortical visual disorder must be confirmed by documentation of the cause of the brain lesion. If neuroimaging or visual evoked response (VER) testing was performed, we will request a copy of the report or other medical evidence that describes the findings in the report.

c. If your visual disorder does not satisfy the criteria in 2.02, 2.03, or 2.04, we will also request a description of how your visual disorder impacts your ability to function.

5. *How do we measure best-corrected visual acuity?*

a. *Testing for visual acuity.* When we need to measure your best-corrected visual acuity, we will use visual acuity testing that was carried out using Snellen methodology or any other testing methodology that is comparable to Snellen methodology.

b. *Determining best-corrected visual acuity.* (i) Best-corrected visual acuity is the optimal visual acuity attainable with the use of a corrective lens. In some instances, this assessment may be performed using a specialized lens; for example, a contact lens. We will use the visual acuity measurements obtained with a specialized lens only if you have demonstrated the ability to use the specialized lens on a sustained basis. However, we will not use visual acuity measurements obtained with telescopic lenses because they significantly reduce the visual field. If you have an absent response to VER testing in an eye, we can determine that your best-corrected visual acuity is 20/200 or less in that eye. However, if you have a positive response to VER testing in an eye, we will not use that result to determine your best-corrected visual acuity in that eye. Additionally, we will not use the results of pinhole testing or automated refraction acuity to determine your best-corrected visual acuity.

(ii) We will use the best-corrected visual acuity for distance in your better eye when we determine whether your loss of visual acuity satisfies the criteria in 2.02. The best-corrected visual acuity for distance is usually measured by determining what you can see from 20 feet. If your visual acuity is measured for a distance other than 20 feet, we will convert it to a 20-foot measurement. For example, if your visual acuity is measured at 10 feet and is reported as 10/40, we will convert this to 20/80.

6. *How do we measure visual fields?*

a. *Testing for visual fields.*

(i) We generally need visual field testing when you have a visual disorder that could result in visual field loss, such as glaucoma, retinitis pigmentosa, or optic neuropathy, or when you display behaviors that suggest a visual field loss.

(ii) When we need to measure the extent of your visual field loss, we will use visual field measurements obtained with an automated static threshold perimetry test performed on a perimeter, like the Humphrey Field Analyzer, that satisfies all of the following requirements:

A. The perimeter must use optical projection to generate the test stimuli.

B. The perimeter must have an internal normative database for automatically comparing your performance with that of the general population.

C. The perimeter must have a statistical analysis package that is able to calculate visual field indices, particularly mean deviation.

D. The perimeter must demonstrate the ability to correctly detect visual field loss and correctly identify normal visual fields.

E. The perimeter must demonstrate good test-retest reliability.

F. The perimeter must have undergone clinical validation studies by three or more independent laboratories with results published in peer-reviewed ophthalmic journals.

(iii) The test must use a white size III Goldmann stimulus and a 31.5 apostilb (10 cd/m$^{[2]}$) white background. The stimuli locations must be no more than 6 degrees apart horizontally or vertically. Measurements must be reported on standard charts and include a description of the size and intensity of the test stimulus.

(iv) To determine statutory blindness based on visual field loss (2.03A), we need a test that measures the central 24 to 30 degrees of the visual field; that is, the area measuring 24 to 30 degrees from the point of fixation. Acceptable tests include the Humphrey 30-2 or 24-2 tests.

(v) The criterion in 2.03B is based on the use of a test performed on a Humphrey Field Analyzer that measures the central 30 degrees of the visual field. We can also use comparable results from other acceptable perimeters, for example, a mean defect of 22 on an acceptable Octopus test, to determine that the criterion in 2.03B is met. We cannot use tests that do not measure the central 30 degrees of the visual field, such as the Humphrey 24-2 test, to determine if your impairment meets or medically equals 2.03B.

(vi) We measure the extent of visual field loss by determining the portion of the visual field in which you can see a white III4e stimulus. The "III" refers to the standard Goldmann test stimulus size III, and the "4e" refers to the standard Goldmann intensity filters used to determine the intensity of the stimulus.

(vii) In automated static threshold perimetry, the intensity of the stimulus varies. The intensity of the stimulus is expressed in decibels (dB). We need to determine the dB level that corresponds to a 4e intensity for the particular perimeter being used. We will then use the dB printout to determine which points would be seen at a 4e intensity level. For example, in Humphrey Field Analyzers, a 10 dB stimulus is equivalent to a 4e stimulus. A dB level that is higher than 10 represents a dimmer stimulus, while a dB level that is lower than 10 represents a brighter stimulus. Therefore, for tests performed on Humphrey Field Analyzers, any point seen at 10 dB or higher is a point that would be seen with a 4e stimulus.

(viii) We can also use visual field measurements obtained using kinetic perimetry, such as the Humphrey "SSA Test Kinetic" or Goldmann perimetry, instead of automated static threshold perimetry. The kinetic test must use a white III4e stimulus projected on a white 31.5 apostilb (10 cd/m$^{[2]}$) background. In automated kinetic tests, such as the Humphrey "SSA Test Kinetic," testing along a meridian stops when you see the stimulus. Because of this, automated kinetic testing does not detect limitations in the central visual field. If your visual disorder has progressed to the point at which it is likely to result in a significant limitation in the central visual field, such as a scotoma (see 2.00A8c), we will not use automated kinetic perimetry to evaluate your visual field loss. Instead, we will assess your visual field loss using automated

static threshold perimetry or manual kinetic perimetry.

(ix) We will not use the results of visual field screening tests, such as confrontation tests, tangent screen tests, or automated static screening tests, to determine that your impairment meets or medically equals a listing or to evaluate your residual functional capacity. However, we can consider normal results from visual field screening tests to determine whether your visual disorder is severe when these test results are consistent with the other evidence in your case record. (See §§404.1520(c), 404.1521, 416.920(c), and 416.921.) We will not consider normal test results to be consistent with the other evidence if either of the following applies:

A. The clinical findings indicate that your visual disorder has progressed to the point that it is likely to cause visual field loss, or

B. You have a history of an operative procedure for retinal detachment.

b. *Use of corrective lenses.* You must not wear eyeglasses during the visual field examination because they limit your field of vision. Contact lenses or perimetric lenses may be used to correct visual acuity during the visual field examination in order to obtain the most accurate visual field measurements. For this single purpose, you do not need to demonstrate that you have the ability to use the contact or perimetric lenses on a sustained basis.

7. *How do we calculate visual efficiency?*

a. *Visual acuity efficiency.* We use the percentage shown in Table 1 that corresponds to the best-corrected visual acuity for distance in your better eye.

b. *Visual field efficiency.* We use kinetic perimetry to calculate visual field efficiency by adding the number of degrees seen along the eight principal meridians in your better eye and dividing by 500. (See Table 2.)

c. *Visual efficiency.* We calculate the percent of visual efficiency by multiplying the visual acuity efficiency by the visual field efficiency and converting the decimal to a percentage. For example, if your visual acuity efficiency is 75 percent and your visual field efficiency is 64 percent, we will multiply 0.75 × 0.64 to determine that your visual efficiency is 0.48, or 48 percent.

8. *How do we evaluate specific visual problems?*

a. *Statutory blindness.* Most test charts that use Snellen methodology do not have lines that measure visual acuity between 20/100 and 20/200. Newer test charts, such as the Bailey-Lovie or the Early Treatment Diabetic Retinopathy Study (ETDRS), do have lines that measure visual acuity between 20/100 and 20/200. If your visual acuity is measured with one of these newer charts, and you cannot read any of the letters on the 20/100 line, we will determine that you have statutory blindness based on a visual acuity of 20/200 or less. For example, if your best-corrected visual acuity for distance in the better eye was determined to be 20/160 using an ETDRS chart, we will find that you have statutory blindness. Regardless of the type of test chart used, you do not have statutory blindness if you can read at least one letter on the 20/100 line. For example, if your best-corrected visual acuity for distance in the better eye was determined to be 20/125+1 using an ETDRS chart, we will find that you do not have statutory blindness as you are able to read one letter on the 20/100 line.

b. *Blepharospasm.* This movement disorder is characterized by repetitive, bilateral, involuntary closure of the eyelids. If you have this disorder, you may have measurable visual acuities and visual fields that do not satisfy the criteria of 2.02 or 2.03. Blepharospasm generally responds to therapy. However, if therapy is not effective, we will consider how the involuntary closure of your eyelids affects your ability to maintain visual functioning over time.

c. *Scotoma.* A scotoma is a non-seeing area in the visual field surrounded by a seeing area. When we measure the visual field, we subtract the length of any scotoma, other than the normal blind spot, from the overall length of any diameter on which it falls.

B. *Otolaryngology*

1. *Hearing impairment.* Hearing ability should be evaluated in terms of the person's ability to hear and distinguish speech.

Loss of hearing can be quantitatively determined by an audiometer which meets the standards of the American National Standards Institute (ANSI) for air and bone conducted stimuli (*i.e.*, ANSI S 3.6-1969 and ANSI S 3.13-1972, or subsequent comparable revisions) and performing all hearing measurements in an environment which meets the ANSI standard for maximal permissible background sound (ANSI S 3.1-1977).

Speech discrimination should be determined using a standardized measure of speech discrimination ability in quiet at a test presentation level sufficient to ascertain maximum discrimination ability. The speech discrimination measure (test) used, and the level at which testing was done, must be reported.

Hearing tests should be preceded by an otolaryngologic examination and should be performed by or under the supervision of an otolaryngologist or audiologist qualified to perform such tests.

In order to establish an independent medical judgment as to the level of impairment in a claimant alleging deafness, the following examinations should be reported: Otolaryngologic examination, pure tone air and bone audiometry, speech reception threshold (SRT), and speech discrimination testing. A copy of reports of medical examination and audiologic evaluations must be submitted.

Cases of alleged "deaf mutism" should be documented by a hearing evaluation. Records obtained from a speech and hearing rehabilitation center or a special school for the deaf may be acceptable, but if these reports are not available, or are found to be inadequate, a current hearing evaluation should be submitted as outlined in the preceding paragraph.

2. *Vertigo associated with disturbances of labyrinthine-vestibular function, including Meniere's disease.* These disturbances of balance are characterized by an hallucination of motion or loss of position sense and a sensation of dizziness which may be constant or may occur in paroxysmal attacks. Nausea, vomiting, ataxia, and incapacitation are frequently observed, particularly during the acute attack. It is important to differentiate the report of rotary vertigo from that of "dizziness" which is described as lightheadedness, unsteadiness, confusion, or syncope.

Meniere's disease is characterized by paroxysmal attacks of vertigo, tinnitus, and fluctuating hearing loss. Remissions are unpredictable and irregular, but may be longlasting; hence, the

severity of impairment is best determined after prolonged observation and serial reexaminations.

The diagnosis of a vestibular disorder requires a comprehensive neuro-otolaryngologic examination with a detailed description of the vertiginous episodes, including notation of frequency, severity, and duration of the attacks. Pure tone and speech audiometry with the appropriate special examinations, such as Bekesy audiometry, are necessary. Vestibular functions is assessed by positional and caloric testing, preferably by electronystagmography. When polytomograms, contrast radiography, or other special tests have been performed, copies of the reports of these tests should be obtained in addition to appropriate medically acceptable imaging reports of the skull and temporal bone. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

3. *Loss of speech.* In evaluating the loss of speech, the ability to produce speech by any means includes the use of mechanical or electronic devices that improve voice or articulation. Impairments of speech may also be evaluated under the body system for the underlying disorder, such as neurological disorders, 11.00ff.

C. *How do we evaluate impairments that do not meet one of the special senses and speech listings?*

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

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

2.01 Category of Impairments, Special Senses and Speech

2.02 *Loss of visual acuity.* Remaining vision in the better eye after best correction is 20/200 or less.

2.03 *Contraction of the visual field in the better eye,* with:

A. The widest diameter subtending an angle around the point of fixation no greater than 20 degrees;

OR

B. A mean deviation of -22 or worse, determined by automated static threshold perimetry as described in 2.00A6a(v);

OR

C. A visual field efficiency of 20 percent or less as determined by kinetic perimetry (see 2.00A7b).

2.04 *Loss of visual efficiency.* Visual efficiency of the better eye of 20 percent or less after best correction (see 2.00A7c).

2.07 *Disturbance of labyrinthine-vestibular function (including Meniere's disease),* characterized by a history of frequent attacks of balance disturbance, tinnitus, and progressive loss of hearing. With both A and B:

A. Disturbed function of vestibular labyrinth demonstrated by caloric or other vestibular tests; and

B. Hearing loss established by audiometry.

2.08 *Hearing impairments* (hearing not restorable by a hearing aid) manifested by:

A. Average hearing threshold sensitivity for air conduction of 90 decibels or greater and for bone conduction to corresponding maximal levels, in the better ear, determined by the simple average of hearing threshold levels at 500, 1000 and 2000 hz. (see 2.00B1); or

B. Speech discrimination scores of 40 percent or less in the better ear;

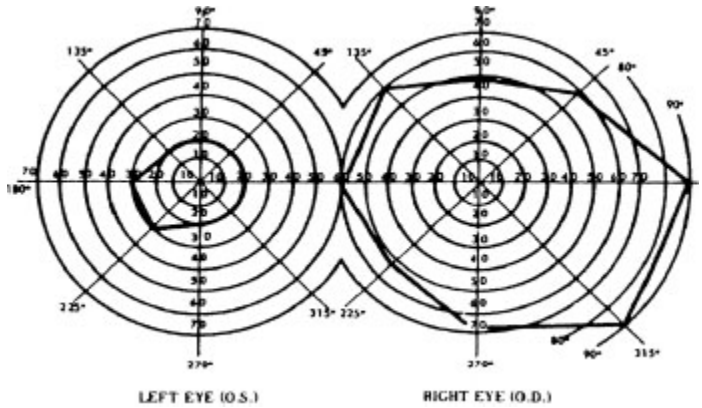
2.09 *Loss of speech* due to any cause, with inability to produce by any means speech that can be heard, understood, or sustained.

Table 1.—Percentage of Visual Acuity Efficiency Corresponding to the Best-Corrected Visual Acuity Measurement for Distance in the Better Eye

Snellen		Percent visual acuity efficiency
English	Metric	
20/16	6/5	100
20/20	6/6	100
20/25	6/7.5	95
20/30	6/9	90
20/40	6/12	85
20/50	6/15	75
20/60	6/18	70
20/70	6/21	65
20/80	6/24	60

20/100	6/30	50

Table 2.—Chart of Visual Fields



1. The diagram of the right eye illustrates the extent of a normal visual field as measured with a III4e stimulus. The sum of the eight principal meridians of this field is 500 degrees.

2. The diagram of the left eye illustrates a visual field contracted to 30 degrees in two meridians and to 20 degrees in the remaining six meridians. The percent of visual field efficiency of this field is: (2 × 30) + (6 × 20) = 180÷500 = 0.36 or 36 percent visual field efficiency.

3.00 Respiratory System

A. *Introduction.* The listings in this section describe impairments resulting from respiratory disorders based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of treatment prescribed by a treating source. Respiratory disorders along with any associated impairment(s) must be established by medical evidence. Evidence must be provided in sufficient detail to permit an independent reviewer to evaluate the severity of the impairment.

Many individuals, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is evidence of such treatment, the longitudinal clinical record must include a description of the treatment prescribed by the treating source and response in addition to information about the nature and severity of the impairment. It is important to document any prescribed treatment and response, because this medical management may have improved the individual's functional status. The longitudinal record should provide information regarding functional recovery, if any.

Some individuals will not have received ongoing treatment or have an ongoing relationship with the medical community, despite the existence of a severe impairment(s). An individual who does not receive treatment may or may not be able to show the existence of an

impairment that meets the criteria of these listings. Even if an individual does not show that his or her impairment meets the criteria of these listings, the individual may have an impairment(s) equivalent in severity to one of the listed impairments or be disabled because of a limited residual functional capacity. Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the individual's functioning, and the frequency, severity, and duration of symptoms. Also, the asthma listing specifically includes a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.

Impairments caused by chronic disorders of the respiratory system generally produce irreversible loss of pulmonary function due to ventilatory impairments, gas exchange abnormalities, or a combination of both. The most common symptoms attributable to these disorders are dyspnea on exertion, cough, wheezing, sputum production, hemoptysis, and chest pain. Because these symptoms are common to many other diseases, a thorough medical history, physical examination, and chest x-ray or other appropriate imaging technique are required to establish chronic pulmonary disease. Pulmonary function testing is required to assess the severity of the respiratory impairment once a disease process is established by appropriate clinical and laboratory findings.

Alterations of pulmonary function can be due to obstructive airway disease (e.g., emphysema, chronic bronchitis, asthma), restrictive pulmonary disorders with primary loss of lung volume (e.g., pulmonary resection, thoracoplasty, chest cage deformity as in kyphoscoliosis or obesity), or infiltrative interstitial disorders (e.g., diffuse pulmonary fibrosis). Gas exchange abnormalities without significant airway obstruction can be produced by interstitial disorders. Disorders involving the pulmonary circulation (e.g., primary pulmonary hypertension, recurrent thromboembolic disease, primary or secondary pulmonary vasculitis) can produce pulmonary vascular hypertension and, eventually, pulmonary heart disease (cor pulmonale) and right heart failure. Persistent hypoxemia produced by any chronic pulmonary disorder also can result in chronic pulmonary hypertension and right heart failure. Chronic infection, caused most frequently by mycobacterial or mycotic organisms, can produce extensive and progressive lung destruction resulting in marked loss of pulmonary function. Some disorders, such as bronchiectasis, cystic fibrosis, and asthma, can be associated with intermittent exacerbations of such frequency and intensity that they produce a disabling impairment, even when pulmonary function during periods of relative clinical stability is relatively well-maintained.

Respiratory impairments usually can be evaluated under these listings on the basis of a complete medical history, physical examination, a chest x-ray or other appropriate imaging techniques, and spirometric pulmonary function tests. In some situations, most typically with a diagnosis of diffuse interstitial fibrosis or clinical findings suggesting cor pulmonale, such as cyanosis or secondary polycythemia, an impairment may be underestimated on the basis of spirometry alone. More sophisticated pulmonary function testing may then be necessary to determine if gas exchange abnormalities contribute to the severity of a respiratory impairment. Additional testing might include measurement of diffusing capacity of the lungs for carbon monoxide or resting arterial blood gases. Measurement of arterial blood gases during exercise is required infrequently. In disorders of the pulmonary circulation, right heart catheterization with angiography and/or direct measurement of pulmonary artery pressure may have been done to establish a diagnosis and evaluate severity. When performed, the results of the procedure should be obtained. Cardiac catheterization will not be purchased.

These listings are examples of common respiratory disorders that are severe enough to prevent a person from engaging in any gainful activity. When an individual has a medically determinable impairment that is not listed, an impairment which does not meet a listing, or a combination of impairments no one of which meets a listing, we will consider whether the individual's impairment or combination of impairments is medically equivalent in severity to a listed impairment. Individuals who have an impairment(s) with a level of severity which does not meet or equal the criteria of the listings may or may not have the residual functional capacity (RFC) which would enable them to engage in substantial gainful activity. Evaluation of the impairment(s) of these individuals will proceed through the final steps of the sequential evaluation process.

B. *Mycobacterial, mycotic, and other chronic persistent infections of the lung.* These disorders are evaluated on the basis of the resulting limitations in pulmonary function. Evidence of chronic infections, such as active mycobacterial diseases or mycoses with positive cultures, drug resistance, enlarging parenchymal lesions, or cavitation, is not, by itself, a basis for determining that an individual has a disabling impairment expected to last 12 months. In those unusual cases of pulmonary infection that persist for a period approaching 12 consecutive months, the clinical findings, complications, therapeutic considerations, and prognosis must be carefully assessed to determine whether, despite relatively well-maintained pulmonary function, the individual nevertheless has an impairment that is expected to last for at least 12 consecutive months and prevent gainful activity.

C. *Episodic respiratory disease.* When a respiratory impairment is episodic in nature, as can occur with exacerbations of asthma, cystic fibrosis, bronchiectasis, or chronic asthmatic bronchitis, the frequency and intensity of episodes that occur despite prescribed treatment are often the major criteria for determining the level of impairment. Documentation for these exacerbations should include available hospital, emergency facility and/or physician records indicating the dates of treatment; clinical and laboratory findings on presentation, such as the results of spirometry and arterial blood gas studies (ABGS); the treatment administered; the time period required for treatment; and the clinical response. Attacks of asthma, episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum), or respiratory failure as referred to in paragraph B of 3.03, 3.04, and 3.07, are defined as prolonged symptomatic episodes lasting one or more days and requiring intensive treatment, such as intravenous bronchodilator or antibiotic administration or prolonged inhalational bronchodilator therapy in a hospital, emergency room or equivalent setting. Hospital admissions are defined as inpatient hospitalizations for longer than 24 hours. The medical evidence must also include information documenting adherence to a prescribed regimen of treatment as well as a description of physical signs. For asthma, the medical evidence should include spirometric results obtained between attacks that document the presence of baseline airflow obstruction.

D. *Cystic fibrosis* is a disorder that affects either the respiratory or digestive body systems or both and is responsible for a wide and variable spectrum of clinical manifestations and complications. Confirmation of the diagnosis is based upon an elevated sweat sodium concentration or chloride concentration accompanied by one or more of the following: the presence of chronic obstructive pulmonary disease, insufficiency of exocrine pancreatic function, meconium ileus, or a positive family history. The quantitative pilocarpine iontophoresis procedure for collection of sweat content must be utilized. Two methods are acceptable: the "Procedure for the Quantitative Iontophoretic Sweat Test for Cystic Fibrosis" published by the Cystic Fibrosis Foundation and contained in, "A Test for Concentration of Electrolytes in Sweat in Cystic Fibrosis of the Pancreas Utilizing Pilocarpine Iontophoresis,"

Gibson, I.E., and Cooke, R.E., *Pediatrics,* Vol. 23: 545, 1959; or the "Wescor Macroduct System." To establish the diagnosis of cystic fibrosis, the sweat sodium or chloride content must be analyzed quantitatively using an acceptable laboratory technique. Another diagnostic test is the "CF gene mutation analysis" for homozygosity of the cystic fibrosis gene. The pulmonary manifestations of this disorder should be evaluated under 3.04. The nonpulmonary aspects of cystic fibrosis should be evaluated under the digestive body system (5.00). Because cystic fibrosis may involve the respiratory and digestive body systems, the combined effects of the involvement of these body systems must be considered in case adjudication.

E. *Documentation of pulmonary function testing.* The results of spirometry that are used for adjudication under paragraphs A and B of 3.02 and paragraph A of 3.04 should be expressed in liters (L), body temperature and pressure saturated with water vapor (BTPS). The reported one-second forced expiratory volume (FEV_1) and forced vital capacity (FVC) should represent the largest of at least three satisfactory forced expiratory maneuvers. Two of the satisfactory spirograms should be reproducible for both pre-bronchodilator tests and, if indicated, post-bronchodilator tests. A value is considered reproducible if it does not differ from the largest value by more than 5 percent or 0.1 L, whichever is greater. The highest values of the FEV_1 and FVC, whether from the same or different tracings, should be used to assess the severity of the respiratory impairment. Peak flow should be achieved early in expiration, and the spirogram should have a smooth contour with gradually decreasing flow throughout expiration. The zero time for measurement of the FEV_1 and FVC, if not distinct, should be derived by linear back-extrapolation of peak flow to zero volume. A spirogram is satisfactory for measurement of the FEV_1 if the expiratory volume at the back-extrapolated zero time is less than 5 percent of the FVC or 0.1 L, whichever is greater. The spirogram is satisfactory for measurement of the FVC if maximal expiratory effort continues for at least 6 seconds, or if there is a plateau in the volume-time curve with no detectable change in expired volume (VE) during the last 2 seconds of maximal expiratory effort.

Spirometry should be repeated after administration of an aerosolized bronchodilator under supervision of the testing personnel if the pre-bronchodilator FEV_1 value is less than 70 percent of the predicted normal value. Pulmonary function studies should not be performed unless the clinical status is stable (e.g., the individual is not having an asthmatic attack or suffering from an acute respiratory infection or other chronic illness). Wheezing is common in asthma, chronic bronchitis, or chronic obstructive pulmonary disease and does not preclude testing. The effect of the administered bronchodilator in relieving bronchospasm and improving ventilatory function is assessed by spirometry. If a bronchodilator is not administered, the reason should be clearly stated in the report. Pulmonary function studies performed to assess airflow obstruction without testing after bronchodilators cannot be used to assess levels of impairment in the range that prevents any gainful work activity, unless the use of bronchodilators is contraindicated. Post-bronchodilator testing should be performed 10 minutes after bronchodilator administration. The dose and name of the bronchodilator administered should be specified. The values in paragraphs A and B of 3.02 must only be used as criteria for the level of ventilatory impairment that exists during the individual's most stable state of health (*i.e.*, any period in time except during or shortly after an exacerbation).

The appropriately labeled spirometric tracing, showing the claimant's name, date of testing, distance per second on the abscissa and distance per liter (L) on the ordinate, must be incorporated into the file. The manufacturer and model number of the device used to measure and record the spirogram should be stated. The testing device must accurately measure both

time and volume, the latter to within 1 percent of a 3 L calibrating volume. If the spirogram was generated by any means other than direct pen linkage to a mechanical displacement-type spirometer, the testing device must have had a recorded calibration performed previously on the day of the spirometric measurement.

If the spirometer directly measures flow, and volume is derived by electronic integration, the linearity of the device must be documented by recording volume calibrations at three different flow rates of approximately 30 L/min (3 L/6 sec), 60 L/min (3 L/3 sec), and 180 L/min (3 L/sec). The volume calibrations should agree to within 1 percent of a 3 L calibrating volume. The proximity of the flow sensor to the individual should be noted, and it should be stated whether or not a BTPS correction factor was used for the calibration recordings and for the individual's actual spirograms.

The spirogram must be recorded at a speed of at least 20 mm/sec, and the recording device must provide a volume excursion of at least 10 mm/L. If reproductions of the original spirometric tracings are submitted, they must be legible and have a time scale of at least 20 mm/sec and a volume scale of at least 10 mm/L to permit independent measurements. Calculation of FEV_1 from a flow-volume tracing is not acceptable, *i.e.*, the spirogram and calibrations must be presented in a volume-time format at a speed of at least 20 mm/sec and a volume excursion of at least 10 mm/L to permit independent evaluation.

A statement should be made in the pulmonary function test report of the individual's ability to understand directions as well as his or her effort and cooperation in performing the pulmonary function tests.

The pulmonary function tables in 3.02 and 3.04 are based on measurement of standing height without shoes. If an individual has marked spinal deformities (e.g., kyphoscoliosis), the measured span between the fingertips with the upper extremities abducted 90 degrees should be substituted for height when this measurement is greater than the standing height without shoes.

F. *Documentation of chronic impairment of gas exchange.*

1. *Diffusing capacity of the lungs for carbon monoxide (DLCO).* A diffusing capacity of the lungs for carbon monoxide study should be purchased in cases in which there is documentation of chronic pulmonary disease, but the existing evidence, including properly performed spirometry, is not adequate to establish the level of functional impairment. Before purchasing DLCO measurements, the medical history, physical examination, reports of chest x-ray or other appropriate imaging techniques, and spirometric test results must be obtained and reviewed because favorable decisions can often be made based on available evidence without the need for DLCO studies. Purchase of a DLCO study may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

The DLCO should be measured by the single breath technique with the individual relaxed and seated. At sea level, the inspired gas mixture should contain approximately 0.3 percent carbon monoxide (CO), 10 percent helium (He), 21 percent oxygen (O_2), and the balance nitrogen. At altitudes above sea level, the inspired O_2 concentration may be raised to provide an inspired O_2 tension of approximately 150 mm Hg. Alternatively, the sea level mixture may be employed

at altitude and the measured DLCO corrected for ambient barometric pressure. Helium may be replaced by another inert gas at an appropriate concentration. The inspired volume (VI) during the DLCO maneuver should be at least 90 percent of the previously determined vital capacity (VC). The inspiratory time for the VI should be less than 2 seconds, and the breath-hold time should be between 9 and 11 seconds. The washout volume should be between 0.75 and 1.00 L, unless the VC is less than 2 L. In this case, the washout volume may be reduced to 0.50 L; any such change should be noted in the report. The alveolar sample volume should be between 0.5 and 1.0 L and be collected in less than 3 seconds. At least 4 minutes should be allowed for gas washout between repeat studies.

A DLCO should be reported in units of ml CO, standard temperature, pressure, dry (STPD)/min/mm Hg uncorrected for hemoglobin concentration and be based on a single-breath alveolar volume determination. Abnormal hemoglobin or hematocrit values, and/or carboxyhemoglobin levels should be reported along with diffusing capacity.

The DLCO value used for adjudication should represent the mean of at least two acceptable measurements, as defined above. In addition, two acceptable tests should be within 10 percent of each other or 3 ml CO(STPD)/min/mm Hg, whichever is larger. The percent difference should be calculated as 100×(test 1-test 2)/average DLCO.

The ability of the individual to follow directions and perform the test properly should be described in the written report. The report should include tracings of the VI, breath-hold maneuver, and VE appropriately labeled with the name of the individual and the date of the test. The time axis should be at least 20 mm/sec and the volume axis at least 10 mm/L. The percentage concentrations of inspired O_2 and inspired and expired CO and He for each of the maneuvers should be provided. Sufficient data must be provided, including documentation of the source of the predicted equation, to permit verification that the test was performed adequately, and that, if necessary, corrections for anemia or carboxyhemoglobin were made appropriately.

2. *Arterial blood gas studies (ABGS).* An ABGS performed at rest (while breathing room air, awake and sitting or standing) or during exercise should be analyzed in a laboratory certified by a State or Federal agency. If the laboratory is not certified, it must submit evidence of participation in a national proficiency testing program as well as acceptable quality control at the time of testing. The report should include the altitude of the facility and the barometric pressure on the date of analysis.

Purchase of resting ABGS may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided. If the results of a DLCO study are greater than 40 percent of predicted normal but less than 60 percent of predicted normal, purchase of resting ABGS should be considered. Before purchasing resting ABGS, a program physician, preferably one experienced in the care of patients with pulmonary disease, must review all clinical and laboratory data short of this procedure, including spirometry, to determine whether obtaining the test would present a significant risk to the individual.

3. *Exercise testing.* Exercise testing with measurement of arterial blood gases during exercise may be appropriate in cases in which there is documentation of chronic pulmonary disease, but full development, short of exercise testing, is not adequate to establish if the impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably

decided. In this context, "full development" means that results from spirometry and measurement of DLCO and resting ABGS have been obtained from treating sources or through purchase. Exercise arterial blood gas measurements will be required infrequently and should be purchased only after careful review of the medical history, physical examination, chest x-ray or other appropriate imaging techniques, spirometry, DLCO, electrocardiogram (ECG), hematocrit or hemoglobin, and resting blood gas results by a program physician, preferably one experienced in the care of patients with pulmonary disease, to determine whether obtaining the test would presents a significant risk to the individual. Oximetry and capillary blood gas analysis are not acceptable substitutes for the measurement of arterial blood gases. Arterial blood gas samples obtained after the completion of exercise are not acceptable for establishing an individual's functional capacity.

Generally, individuals with a DLCO greater than 60 percent of predicted normal would not be considered for exercise testing with measurement of blood gas studies. The exercise test facility must be provided with the claimant's clinical records, reports of chest x-ray or other appropriate imaging techniques, and any spirometry, DLCO, and resting blood gas results obtained as evidence of record. The testing facility must determine whether exercise testing present a significant risk to the individual; if it does, the reason for not performing the test must be reported in writing.

4. *Methodology.* Individuals considered for exercise testing first should have resting arterial blood partial pressure of oxygen (PO_2), resting arterial blood partial pressure of carbon dioxide (PCO_2) and negative log of hydrogen ion concentration (pH) determinations by the testing facility. The sample should be obtained in either the sitting or standing position. The individual should then perform exercise under steady state conditions, preferably on a treadmill, breathing room air, for a period of 4 to 6 minutes at a speed and grade providing an oxygen consumption of approximately 17.5 ml/kg/min (5 METs). If a bicycle ergometer is used, an exercise equivalent of 5 METs (e.g., 450 kpm/min, or 75 watts, for a 176 pound (80 kilogram) person) should be used. If the individual is able to complete this level of exercise without achieving listing-level hypoxemia, then he or she should be exercised at higher workloads to determine exercise capacity. A warm-up period of treadmill walking or cycling may be performed to acquaint the individual with the exercise procedure. If during the warm-up period the individual cannot achieve an exercise level of 5 METs, a lower workload may be selected in keeping with the estimate of exercise capacity. The individual should be monitored by ECG throughout the exercise and in the immediate post-exercise period. Blood pressure and an ECG should be recorded during each minute of exercise. During the final 2 minutes of a specific level of steady state exercise, an arterial blood sample should be drawn and analyzed for oxygen pressure (or tension) (PO_2), carbon dioxide pressure (or tension) (PCO_2), and pH. At the discretion of the testing facility, the sample may be obtained either from an indwelling arterial catheter or by direct arterial puncture. If possible, in order to evaluate exercise capacity more accurately, a test site should be selected that has the capability to measure minute ventilation, O_2 consumption, and carbon dioxide (CO_2) production. If the claimant fails to complete 4 to 6 minutes of steady state exercise, the testing laboratory should comment on the reason and report the actual duration and levels of exercise performed. This comment is necessary to determine if the individual's test performance was limited by lack of effort or other impairment (e.g., cardiac, peripheral vascular, musculoskeletal, neurological).

The exercise test report should contain representative ECG strips taken before, during and after exercise; resting and exercise arterial blood gas values; treadmill speed and grade

settings, or, if a bicycle ergometer was used, exercise levels expressed in watts or kpm/min; and the duration of exercise. Body weight also should be recorded. If measured, O_2 consumption (STPD), minute ventilation (BTPS), and CO_2 production (STPD) also should be reported. The altitude of the test site, its normal range of blood gas values, and the barometric pressure on the test date must be noted.

G. *Chronic cor pulmonale and pulmonary vascular disease.* The establishment of an impairment attributable to irreversible cor pulmonale secondary to chronic pulmonary hypertension requires documentation by signs and laboratory findings of right ventricular overload or failure (e.g., an early diastolic right-sided gallop on auscultation, neck vein distension, hepatomegaly, peripheral edema, right ventricular outflow tract enlargement on x-ray or other appropriate imaging techniques, right ventricular hypertrophy on ECG, and increased pulmonary artery pressure measured by right heart catheterization available from treating sources). Cardiac catheterization will not be purchased. Because hypoxemia may accompany heart failure and is also a cause of pulmonary hypertension, and may be associated with hypoventilation and respiratory acidosis, arterial blood gases may demonstrate hypoxemia (decreased PO_2), CO_2 retention (increased PCO_2), and acidosis (decreased pH). Polycythemia with an elevated red blood cell count and hematocrit may be found in the presence of chronic hypoxemia.

P-pulmonale on the ECG does not establish chronic pulmonary hypertension or chronic cor pulmonale. Evidence of florid right heart failure need not be present at the time of adjudication for a listing (e.g., 3.09) to be satisfied, but the medical evidence of record should establish that cor pulmonale is chronic and irreversible.

H. *Sleep-related breathing disorders.* Sleep-related breathing disorders (sleep apneas) are caused by periodic cessation of respiration associated with hypoxemia and frequent arousals from sleep. Although many individuals with one of these disorders will respond to prescribed treatment, in some, the disturbed sleep pattern and associated chronic nocturnal hypoxemia cause daytime sleepiness with chronic pulmonary hypertension and/or disturbances in cognitive function. Because daytime sleepiness can affect memory, orientation, and personality, a longitudinal treatment record may be needed to evaluate mental functioning. Not all individuals with sleep apnea develop a functional impairment that affects work activity. When any gainful work is precluded, the physiologic basis for the impairment may be chronic cor pulmonale. Chronic hypoxemia due to episodic apnea may cause pulmonary hypertension (see 3.00G and 3.09). Daytime somnolence may be associated with disturbance in cognitive vigilance. Impairment of cognitive function may be evaluated under organic mental disorders (12.02).

I. *Effects of obesity.* Obesity is a medically determinable impairment that is often associated with disturbance of the respiratory system, and disturbance of this system can be a major cause of disability in individuals with obesity. The combined effects of obesity with respiratory impairments can be greater than the effects of each of the impairments considered separately. Therefore, when determining whether an individual with obesity has a listing-level impairment or combination of impairments, and when assessing a claim at other steps of the sequential evaluation process, including when assessing an individual's residual functional capacity, adjudicators must consider any additional and cumulative effects of obesity.

3.01 Category of Impairments, Respiratory System.

3.02 *Chronic pulmonary insufficiency.*

A. Chronic obstructive pulmonary disease, due to any cause, with the FEV_1 equal to or less than the values specified in table I corresponding to the person's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

Table I

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
154 or less	60 or less	1.05
155-160	61-63	1.15
161-165	64-65	1.25
166-170	66-67	1.35
171-175	68-69	1.45
176-180	70-71	1.55
181 or more	72 or more	1.65

Or

B. Chronic restrictive ventilatory disease, due to any cause, with the FVC equal to or less than the values specified in table II corresponding to the person's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

Table II

Height without shoes (centimeters)	Height without shoes (inches)	FVC equal to or less than (L, BTPS)
154 or less	60 or less	1.25
155-160	61-63	1.35
161-165	64-65	1.45
166-170	66-67	1.55
171-175	68-69	1.65
176-180	70-71	1.75
181 or more	72 or more	1.85

Or

C. Chronic impairment of gas exchange due to clinically documented pulmonary disease. With:

1. Single breath DLCO (see 3.00F1) less than 10.5 ml/min/mm Hg or less than 40 percent of the predicted normal value. (Predicted values must either be based on data obtained at the test site or published values from a laboratory using the same technique as the test site. The source of the predicted values should be reported. If they are not published, they should be submitted in the form of a table or nomogram); or

2. Arterial blood gas values of PO_2 and simultaneously determined PCO_2 measured while at rest (breathing room air, awake and sitting or standing) in a clinically stable condition on at least two occasions, three or more weeks apart within a 6-month period, equal to or less than

the values specified in the applicable table III-A or III-B or III-C:

Table III—A

[Applicable at test sites less than 3,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 equal to or less than (mm. Hg)
30 or below	65
31	64
32	63
33	62
34	61
35	60
36	59
37	58
38	57
39	56
40 or above	55

Table III—B

[Applicable at test sites 3,000 through 6,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 equal to or less than (mm. Hg)
30 or below	60
31	59
32	58
33	57
34	56
35	55
36	54
37	53
38	52
39	51
40 or above	50

Table III—C

[Applicable at test sites over 6,000 feet above sea level]

Arterial PCO_2 (mm. Hg) and	Arterial PO_2 or equal to or less than (mm. Hg)
30 or below	55
31	54
32	53
33	52
34	51
35	50
36	49
37	48
38	47

39	46
40 or above	45

Or

3. Arterial blood gas values of PO_2 and simultaneously determined PCO_2 during steady state exercise breathing room air (level of exercise equivalent to or less than 17.5 ml O_2 consumption/kg/min or 5 METs) equal to or less than the values specified in the applicable table III-A or III-B or III-C in 3.02C2.

3.03 *Asthma.* With:

A. Chronic asthmatic bronchitis. Evaluate under the criteria for chronic obstructive pulmonary disease in 3.02A;

Or

B. Attacks (as defined in 3.00C), in spite of prescribed treatment and requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each in-patient hospitalization for longer than 24 hours for control of asthma counts as two attacks, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of attacks.

3.04 *Cystic fibrosis.* With:

A. An FEV_1 equal to or less than the appropriate value specified in table IV corresponding to the individual's height without shoes. (In cases of marked spinal deformity, see 3.00E.);

Or

B. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;

Or

C. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased bacterial infection occurring at least once every 6 months and requiring intravenous or nebulization antimicrobial therapy.

Table IV

[Applicable only for evaluation under 3.04A—cystic fibrosis]

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
154 or less	60 or less	1.45
155-159	61-62	1.55

160-164	63-64	1.65
165-169	65-66	1.75
170-174	67-68	1.85
175-179	69-70	1.95
180 or more	71 or more	2.05

3.05 [Reserved]

3.06 *Pneumoconiosis* (demonstrated by appropriate imaging techniques). Evaluate under the appropriate criteria in 3.02.

3.07 *Bronchiectasis* (demonstrated by appropriate imaging techniques). With:

A. Impairment of pulmonary function due to extensive disease. Evaluate under the appropriate criteria in 3.02;

Or

B. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each in-patient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation of at least 12 consecutive months must be used to determine the frequency of episodes.

3.08 *Mycobacterial, mycotic, and other chronic persistent infections of the lung* (see 3.00B). Evaluate under the appropriate criteria in 3.02.

3.09 *Cor pulmonale secondary to chronic pulmonary vascular hypertension.* Clinical evidence of cor pulmonale (documented according to 3.00G) with:

A. Mean pulmonary artery pressure greater than 40 mm Hg;

Or

B. Arterial hypoxemia. Evaluate under the criteria in 3.02C2.

3.10 *Sleep-related breathing disorders.* Evaluate under 3.09 (chronic cor pulmonale) or 12.02 (organic mental disorders).

3.11 *Lung transplant.* Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment.

4.00 Cardiovascular System

A. General

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

a. We mean any disorder that affects the proper functioning of the heart or the circulatory system (that is, arteries, veins, capillaries, and the lymphatic drainage). The disorder can be congenital or acquired.

b. Cardiovascular impairment results from one or more of four consequences of heart disease:

(i) Chronic heart failure or ventricular dysfunction.

(ii) Discomfort or pain due to myocardial ischemia, with or without necrosis of heart muscle.

(iii) Syncope, or near syncope, due to inadequate cerebral perfusion from any cardiac cause, such as obstruction of flow or disturbance in rhythm or conduction resulting in inadequate cardiac output.

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

c. Disorders of the veins or arteries (for example, obstruction, rupture, or aneurysm) may cause impairments of the lower extremities (peripheral vascular disease), the central nervous system, the eyes, the kidneys, and other organs. We will evaluate peripheral vascular disease under 4.11 or 4.12 and impairments of another body system(s) under the listings for that body system(s).

2. *What do we consider in evaluating cardiovascular impairments?* The listings in this section describe cardiovascular impairments based on symptoms, signs, laboratory findings, response to a regimen of prescribed treatment, and functional limitations.

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

a. *Medical consultant* is an individual defined in §§404.1616(a) and 416.1016(a). This term does not include medical sources who provide consultative examinations for us. We use the abbreviation "MC" throughout this section to designate a medical consultant.

b. *Persistent* means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been present, or is expected to be present, for a continuous period of at least 12 months, such that a pattern of continuing severity is established.

c. *Recurrent* means that the longitudinal clinical record shows that, within a consecutive 12-month period, the finding(s) occurs at least three times, with intervening periods of improvement of sufficient duration that it is clear that separate events are involved.

d. *Appropriate medically acceptable imaging* means that the technique used is the proper one to evaluate and diagnose the impairment and is commonly recognized as accurate for assessing the cited finding.

e. *A consecutive 12-month period* means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with an application or continuing disability review.

f. *Uncontrolled* means the impairment does not adequately respond to standard prescribed

medical treatment.

B. Documenting Cardiovascular Impairment

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular impairment. A longitudinal clinical record covering a period of not less than 3 months of observations and treatment is usually necessary, unless we can make a determination or decision based on the current evidence.

2. *Why is a longitudinal clinical record important?* We will usually need a longitudinal clinical record to assess the severity and expected duration of your impairment(s). If you have a listing-level impairment, you probably will have received medically prescribed treatment. Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your treating or other medical source. It should also include your response to this medical management, as well as information about the nature and severity of your impairment. The record will provide us with information on your functional status over an extended period of time and show whether your ability to function is improving, worsening, or unchanging.

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

a. You may not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In this situation, we will base our evaluation on the current objective medical evidence and the other evidence we have. If you do not receive treatment, you cannot show an impairment that meets the criteria of most of these listings. However, we may find you disabled because you have another impairment(s) that in combination with your cardiovascular impairment medically equals the severity of a listed impairment or based on consideration of your residual functional capacity and age, education, and work experience.

b. Unless we can decide your claim favorably on the basis of the current evidence, a longitudinal record is still important. In rare instances where there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the severity and duration of your impairment.

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

a. We will wait when we have information showing that your impairment is not yet stable and the expected change in your impairment might affect our determination or decision. In these situations, we need to wait to properly evaluate the severity and duration of your impairment during a stable period. Examples of when we might wait are:

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

(ii) If you have recently had a corrective cardiac procedure; for example, coronary artery bypass grafting.

(iii) If you have started new drug therapy and your response to this treatment has not yet been established; for example, beta-blocker therapy for dilated congestive cardiomyopathy.

b. In these situations, we will obtain more evidence 3 months following the event before we evaluate your impairment. However, we will not wait if we have enough information to make a determination or decision based on all of the relevant evidence in your case.

5. *Will we purchase any studies?* In appropriate situations, we will purchase studies necessary to substantiate the diagnosis or to document the severity of your impairment, generally after we have evaluated the medical and other evidence we already have. We will not purchase studies involving exercise testing if there is significant risk involved or if there is another medical reason not to perform the test. We will follow sections 4.00C6, 4.00C7, and 4.00C8 when we decide whether to purchase exercise testing.

6. *What studies will we not purchase?* We will not purchase any studies involving cardiac catheterization, such as coronary angiography, arteriograms, or electrophysiological studies. However, if the results of catheterization are part of the existing evidence we have, we will consider them together with the other relevant evidence. See 4.00C15a.

C. Using Cardiovascular Test Results

1. *What is an ECG?*

a. *ECG* stands for *electrocardiograph* or *electrocardiogram.* An electrocardiograph is a machine that records electrical impulses of your heart on a strip of paper called an electrocardiogram or a *tracing.* To record the ECG, a technician positions a number of small contacts (or *leads*) on your arms, legs, and across your chest to connect them to the ECG machine. An ECG may be done while you are resting or exercising.

b. The ECG tracing may indicate that you have a heart abnormality. It may indicate that your heart muscle is not getting as much oxygen as it needs (ischemia), that your heart rhythm is abnormal (arrhythmia), or that there are other abnormalities of your heart, such as left ventricular enlargement.

2. *How do we evaluate ECG evidence?* We consider a number of factors when we evaluate ECG evidence:

a. An original or legible copy of the 12-lead ECG obtained at rest must be appropriately dated and labeled, with the standardization inscribed on the tracing. Alteration in standardization of specific leads (such as to accommodate large QRS amplitudes) must be identified on those leads.

(i) Detailed descriptions or computer-averaged signals without original or legible copies of the ECG as described in listing 4.00C2a are not acceptable.

(ii) The effects of drugs or electrolyte abnormalities must be considered as possible noncardiac causes of ECG abnormalities of ventricular repolarization; that is, those involving the ST segment and T wave. If available, the predrug (especially digitalis glycosides) ECG should be submitted.

b. ECGs obtained in conjunction with treadmill, bicycle, or arm exercise tests should meet the following specifications:

(i) ECG reports must include the original calibrated ECG tracings or a legible copy.

(ii) A 12-lead baseline ECG must be recorded in the upright position before exercise.

(iii) A 12-lead ECG should be recorded at the end of each minute of exercise.

(iv) If ECG documentation of the effects of hyperventilation is obtained, the exercise test should be deferred for at least 10 minutes because metabolic changes of hyperventilation may alter the physiologic and ECG-recorded response to exercise.

(v) Post-exercise ECGs should be recorded using a generally accepted protocol consistent with the prevailing state of medical knowledge and clinical practice.

(vi) All resting, exercise, and recovery ECG strips must have the standardization inscribed on the tracing. The ECG strips should be labeled to indicate the date, the times recorded and the relationship to the stage of the exercise protocol. The speed and grade (treadmill test) or work rate (bicycle or arm ergometric test) should be recorded. The highest level of exercise achieved, heart rate and blood pressure levels during testing, and the reason(s) for terminating the test (including limiting signs or symptoms) must be recorded.

3. *What are exercise tests and what are they used for?*

a. Exercise tests have you perform physical activity and record how your cardiovascular system responds. Exercise tests usually involve walking on a treadmill, but other forms of exercise, such as an exercise bicycle or an arm exercise machine, may be used. Exercise testing may be done for various reasons; such as to evaluate the severity of your coronary artery disease or peripheral vascular disease or to evaluate your progress after a cardiac procedure or an acute event, like a myocardial infarction (heart attack). Exercise testing is the most widely used testing for identifying the presence of myocardial ischemia and for estimating maximal aerobic capacity (usually expressed in METs—metabolic equivalents) if you have heart disease.

b. We include exercise tolerance test (ETT) criteria in 4.02B3 (chronic heart failure) and 4.04A (ischemic heart disease). To meet the ETT criteria in these listings, the ETT must be a sign-or symptom-limited test in which you exercise while connected to an ECG until you develop a sign or symptom that indicates that you have exercised as much as is considered safe for you.

c. In 4.12B, we also refer to exercise testing for peripheral vascular disease. In this test, you walk on a treadmill, usually for a specified period of time, and the individual who administers the test measures the effect of exercise on the flow of blood in your legs, usually by using ultrasound. The test is also called an exercise Doppler test. Even though this test is intended to evaluate peripheral vascular disease, it will be stopped for your safety if you develop abnormal signs or symptoms because of heart disease.

d. Each type of test is done in a certain way following specific criteria, called a *protocol.* For our program, we also specify certain aspects of how any exercise test we purchase is to be done. See 4.00C10 and 4.00C17.

4. *Do ETTs have limitations?* An ETT provides an estimate of aerobic capacity for walking on a grade, bicycling, or moving one's arms in an environmentally controlled setting. Therefore, ETT

results do not correlate with the ability to perform other types of exertional activities, such as lifting and carrying heavy loads, and do not provide an estimate of the ability to perform activities required for work in all possible work environments or throughout a workday. Also, certain medications (such as beta blockers) and conduction disorders (such as left or right bundle branch blocks) can cause false-negative or false-positive results. Therefore, we must consider the results of an ETT together with all the other relevant evidence in your case record.

5. *How does an ETT with measurement of maximal or peak oxygen uptake VO_2) differ from other ETTs?* Occasionally, medical evidence will include the results of an ETT with VO_2. While ETTs without measurement of VO_2 provide only an estimate of aerobic capacity, measured maximal or peak oxygen uptake provides an accurate measurement of aerobic capacity, which is often expressed in METs (metabolic equivalents). The MET level may not be indicated in the report of attained maximal or peak VO_2 testing, but can be calculated as follows: 1 MET = 3.5 milliliters (ml) of oxygen uptake per kilogram (kg) of body weight per minute. For example, a 70 kg (154 lb.) individual who achieves a maximal or peak VO_2 of 1225 ml in 1 minute has attained 5 METs (1225 ml/70 kg/1 min = 17.5 ml/kg/min. 17.5/3.5 = 5 METs).

6. *When will we consider whether to purchase an exercise test?*

a. We will consider whether to purchase an exercise test when:

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

(ii) We need to assess your residual functional capacity and there is insufficient evidence in the record to make a determination or decision.

b. We will not purchase an exercise test when we can make our determination or decision based on the evidence we already have.

7. *What must we do before purchasing an exercise test?*

a. Before we purchase an exercise test, an MC, preferably one with experience in the care of patients with cardiovascular disease, must review the pertinent history, physical examinations, and laboratory tests that we have to determine whether the test would present a significant risk to you or if there is some other medical reason not to purchase the test (see 4.00C8).

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

c. The MC, in accordance with the regulations and other instructions on consultative examinations, will generally give great weight to the treating source's opinion about the risk of exercise testing to you and will generally not override it. In the rare situation in which the MC does override the treating source's opinion, the MC must prepare a written rationale

documenting the reasons for overriding the opinion.

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

e. We must also provide your records to the medical source who performs the exercise test for review prior to conducting the test if the source does not already have them. The medical source who performs the exercise test has the ultimate responsibility for deciding whether you would be at risk.

8. *When will we not purchase an exercise test or wait before we purchase an exercise test?*

a. We will not purchase an exercise test when an MC finds that you have one of the following significant risk factors:

(i) Unstable angina not previously stabilized by medical treatment.

(ii) Uncontrolled cardiac arrhythmias causing symptoms or hemodynamic compromise.

(iii) An implanted cardiac defibrillator.

(iv) Symptomatic severe aortic stenosis.

(v) Uncontrolled symptomatic heart failure.

(vi) Aortic dissection.

(vii) Severe pulmonary hypertension (pulmonary artery systolic pressure greater than 60 mm Hg).

(viii) Left main coronary stenosis of 50 percent or greater that has not been bypassed.

(ix) Moderate stenotic valvular disease with a systolic gradient across the aortic valve of 50 mm Hg or greater.

(x) Severe arterial hypertension (systolic greater than 200 mm Hg or diastolic greater than 110 mm Hg).

(xi) Hypertrophic cardiomyopathy with a systolic gradient of 50 mm Hg or greater.

b. We also will not purchase an exercise test when you are prevented from performing exercise testing due to another impairment affecting your ability to use your arms and legs.

c. We will not purchase an ETT to document the presence of a cardiac arrhythmia.

d. We will wait to purchase an exercise test until 3 months after you have had one of the following events. This will allow for maximal, attainable restoration of functional capacity.

(i) Acute myocardial infarction.

(ii) Surgical myocardial revascularization (bypass surgery).

(iii) Other open-heart surgical procedures.

(iv) Percutaneous transluminal coronary angioplasty with or without stenting.

e. If you are deconditioned after an extended period of bedrest or inactivity and could improve with activity, or if you are in acute heart failure and are expected to improve with treatment, we will wait an appropriate period of time for you to recuperate before we purchase an exercise test.

9. *What do we mean by a "timely" test?*

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

b. However, an exercise test that is older than 12 months, especially an abnormal one, can still provide information important to our adjudication. For example, a test that is more than 12 months old can provide evidence of ischemic heart disease or peripheral vascular disease, information on decreased aerobic capacity, or information about the duration or onset of your impairment. Such tests can be an important component of the longitudinal record.

c. When we evaluate a test that is more than 12 months old, we must consider the results in the context of all the relevant evidence, including why the test was performed and whether there has been an intervening event or improvement or worsening of your impairment.

d. We will purchase a new exercise test only if we cannot make a determination or decision based on the evidence we have.

10. *How must ETTs we purchase be performed?*

a. The ETT must be a sign- or symptom-limited test characterized by a progressive multistage regimen. It must be performed using a generally accepted protocol consistent with the prevailing state of medical knowledge and clinical practice. A description of the protocol that was followed must be provided, and the test must meet the requirements of 4.00C2b and this section. A radionuclide perfusion scan may be useful for detecting or confirming ischemia when resting ECG abnormalities, medications, or other factors may decrease the accuracy of ECG interpretation of ischemia. (The perfusion imaging is done at the termination of exercise, which may be at a higher MET level than that at which ischemia first occurs. If the imaging confirms the presence of reversible ischemia, the exercise ECG may be useful for detecting the MET level at which ischemia initially appeared.) Exercise tests may also be performed using echocardiography to detect stress-induced ischemia and left ventricular dysfunction (see 4.00C12 and 4.00C13).

b. The exercise test must be paced to your capabilities and be performed following the generally accepted standards for adult exercise test laboratories. With a treadmill test, the speed, grade (incline), and duration of exercise must be recorded for each exercise test stage

performed. Other exercise test protocols or techniques should use similar workloads. The exercise protocol may need to be modified in individual cases to allow for a lower initial workload with more slowly graded increments than the standard Bruce protocol.

c. Levels of exercise must be described in terms of workload and duration of each stage; for example, treadmill speed and grade, or bicycle ergometer work rate in kpm/min or watts.

d. The exercise laboratory's physical environment, staffing, and equipment must meet the generally accepted standards for adult exercise test laboratories.

11. *How do we evaluate ETT results?* We evaluate ETT results on the basis of the work level at which the test becomes abnormal, as documented by onset of signs or symptoms and any ECG or imaging abnormalities. The absence of an ischemic response on an ETT alone does not exclude the diagnosis of ischemic heart disease. We must consider the results of an ETT in the context of all of the other evidence in your case record.

12. *When are ETTs done with imaging?* When resting ECG abnormalities preclude interpretation of ETT tracings relative to ischemia, a radionuclide (for example, thallium-201 or technetium-99m) perfusion scan or echocardiography in conjunction with an ETT provides better results. You may have resting ECG abnormalities when you have a conduction defect—for example, Wolff-Parkinson-White syndrome, left bundle branch block, left ventricular hypertrophy—or when you are taking digitalis or other antiarrhythmic drugs, or when resting ST changes are present. Also, these techniques can provide a reliable estimate of ejection fraction.

13. *Will we purchase ETTs with imaging?* We may purchase an ETT with imaging in your case after an MC, preferably one with experience in the care of patients with cardiovascular disease, has reviewed your medical history and physical examination, any report(s) of appropriate medically acceptable imaging, ECGs, and other appropriate tests. We will consider purchasing an ETT with imaging when other information we have is not adequate for us to assess whether you have severe ventricular dysfunction or myocardial ischemia, there is no significant risk involved (see 4.00C8a), and we cannot make our determination or decision based on the evidence we already have.

14. *What are drug-induced stress tests?* These tests are designed primarily to provide evidence about myocardial ischemia or prior myocardial infarction, but do not require you to exercise. These tests are used when you cannot exercise or cannot exercise enough to achieve the desired cardiac stress. Drug-induced stress tests can also provide evidence about heart chamber dimensions and function; however, these tests do not provide information about your aerobic capacity and cannot be used to help us assess your ability to function. Some of these tests use agents, such as Persantine or adenosine, that dilate the coronary arteries and are used in combination with nuclear agents, such as thallium or technetium (for example, Cardiolyte or Myoview), and a myocardial scan. Other tests use agents, such as dobutamine, that stimulate the heart to contract more forcefully and faster to simulate exercise and are used in combination with a 2-dimensional echocardiogram. We may, when appropriate, purchase a drug-induced stress test to confirm the presence of myocardial ischemia after a review of the evidence in your file by an MC, preferably one with experience in the care of patients with cardiovascular disease.

15. *How do we evaluate cardiac catheterization evidence?*

a. We will not purchase cardiac catheterization; however, if you have had catheterization, we will make every reasonable effort to obtain the report and any ancillary studies. We will consider the quality and type of data provided and its relevance to the evaluation of your impairment. For adults, we generally see two types of catheterization reports: Coronary arteriography and left ventriculography.

b. For coronary arteriography, the report should provide information citing the method of assessing coronary arterial lumen diameter and the nature and location of obstructive lesions. Drug treatment at baseline and during the procedure should be reported. Some individuals with significant coronary atherosclerotic obstruction have collateral vessels that supply the myocardium distal to the arterial obstruction so that there is no evidence of myocardial damage or ischemia, even with exercise. When the results of quantitative computer measurements and analyses are included in your case record, we will consider them in interpreting the severity of stenotic lesions.

c. For left ventriculography, the report should describe the wall motion of the myocardium with regard to any areas of hypokinesis (abnormally decreased motion), akinesis (lack of motion), or dyskinesis (distortion of motion), and the overall contraction of the ventricle as measured by the ejection fraction. Measurement of chamber volumes and pressures may be useful. Quantitative computer analysis provides precise measurement of segmental left ventricular wall thickness and motion. There is often a poor correlation between left ventricular function at rest and functional capacity for physical activity.

16. *What details should exercise Doppler test reports contain?* The reports of exercise Doppler tests must describe the level of exercise; for example, the speed and grade of the treadmill settings, the duration of exercise, symptoms during exercise, and the reasons for stopping exercise if the expected level of exercise was not attained. They must also include the blood pressures at the ankle and other pertinent sites measured after exercise and the time required for the systolic blood pressure to return toward or to the pre-exercise level. The graphic tracings, if available, should also be included with the report. All tracings must be annotated with the standardization used by the testing facility.

17. *How must exercise Doppler tests we purchase be performed?* When we purchase an exercise Doppler test, you must exercise on a treadmill at 2 mph on a 12 percent grade for up to 5 minutes. The reports must include the information specified in 4.00C16. Because this is an exercise test, we must evaluate whether such testing would put you at significant risk, in accordance with the guidance found in 4.00C6, 4.00C7, and 4.00C8.

D. Evaluating Chronic Heart Failure

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

a. *CHF* is the inability of the heart to pump enough oxygenated blood to body tissues. This syndrome is characterized by symptoms and signs of pulmonary or systemic congestion (fluid retention) or limited cardiac output. Certain laboratory findings of cardiac functional and structural abnormality support the diagnosis of CHF. There are two main types of CHF:

(i) *Predominant systolic dysfunction* (the inability of the heart to contract normally and expel sufficient blood), which is characterized by a dilated, poorly contracting left ventricle and reduced ejection fraction (abbreviated EF, it represents the percentage of the blood in the

ventricle actually pumped out with each contraction), and

(ii) *Predominant diastolic dysfunction* (the inability of the heart to relax and fill normally), which is characterized by a thickened ventricular muscle, poor ability of the left ventricle to distend, increased ventricular filling pressure, and a normal or increased EF.

b. CHF is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, or rheumatic, congenital, or other heart disease. However, if the CHF is the result of primary pulmonary hypertension secondary to disease of the lung (cor pulmonale), we will evaluate your impairment using 3.09, in the respiratory system listings.

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

a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization.

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

(ii) An LVEDD greater than 6.0 cm or an EF of 30 percent or less measured during a period of stability (that is, not during an episode of acute heart failure) may be associated clinically with systolic failure.

(iii) Left ventricular posterior wall thickness added to septal thickness totaling 2.5 cm or greater with left atrium enlarged to 4.5 cm or greater may be associated clinically with diastolic failure.

(iv) However, these measurements alone do not reflect your functional capacity, which we evaluate by considering all of the relevant evidence. In some situations, we may need to purchase an ETT to help us assess your functional capacity.

(v) Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema. These findings need not be present on each report, since CHF may be controlled by prescribed treatment.

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

(i) Symptoms of congestion or of limited cardiac output include easy fatigue, weakness, shortness of breath (dyspnea), cough, or chest discomfort at rest or with activity. Individuals with CHF may also experience shortness of breath on lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). They may

also experience cardiac arrhythmias resulting in palpitations, lightheadedness, or fainting.

(ii) Signs of congestion may include hepatomegaly, ascites, increased jugular venous distention or pressure, rales, peripheral edema, or rapid weight gain. However, these signs need not be found on all examinations because fluid retention may be controlled by prescribed treatment.

3. *Is it safe for you to have an ETT, if you have CHF?* The presence of CHF is not necessarily a contraindication to an ETT, unless you are having an acute episode of heart failure. Measures of cardiac performance are valuable in helping us evaluate your ability to do work-related activities. Exercise testing has been safely used in individuals with CHF; therefore, we may purchase an ETT for evaluation under 4.02B3 if an MC, preferably one experienced in the care of patients with cardiovascular disease, determines that there is no significant risk to you. (See 4.00C6 for when we will consider the purchase of an ETT. See 4.00C7-4.00C8 for what we must do before we purchase an ETT and when we will not purchase one.) ST segment changes from digitalis use in the treatment of CHF do not preclude the purchase of an ETT.

4. *How do we evaluate CHF using 4.02?*

a. We must have objective evidence, as described in 4.00D2, that you have chronic heart failure.

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

c. In 4.02B2, the phrase *periods of stabilization* means that, for at least 2 weeks between episodes of acute heart failure, there must be objective evidence of clearing of the pulmonary edema or pleural effusions and evidence that you returned to, or you were medically considered able to return to, your prior level of activity.

d. Listing 4.02B3c requires a decrease in systolic blood pressure below the baseline level (taken in the standing position immediately prior to exercise) or below any systolic pressure reading recorded during exercise. This is because, normally, systolic blood pressure and heart rate increase gradually with exercise. Decreases in systolic blood pressure below the baseline level that occur during exercise are often associated with ischemia-induced left ventricular dysfunction resulting in decreased cardiac output. However, a blunted response (that is, failure of the systolic blood pressure to rise 10 mm Hg or more), particularly in the first 3 minutes of exercise, may be drug-related and is not necessarily associated with left ventricular dysfunction. Also, some individuals with increased sympathetic responses because of deconditioning or apprehension may increase their systolic blood pressure and heart rate above their baseline level just before and early into exercise. This can be associated with a drop in systolic pressure in early exercise that is not due to left ventricular dysfunction. Therefore, an early decrease in systolic blood pressure must be interpreted within the total context of the test; that is, the presence or absence of symptoms such as lightheadedness, ischemic changes, or arrhythmias on the ECG.

E. Evaluating Ischemic Heart Disease

1. *What is ischemic heart disease (IHD)? IHD* results when one or more of your coronary arteries is narrowed or obstructed or, in rare situations, constricted due to vasospasm,

interfering with the normal flow of blood to your heart muscle (ischemia). The obstruction may be the result of an embolus, a thrombus, or plaque. When heart muscle tissue dies as a result of the reduced blood supply, it is called a myocardial infarction (heart attack).

2. *What causes chest discomfort of myocardial origin?*

a. Chest discomfort of myocardial ischemic origin, commonly known as angina pectoris, is usually caused by coronary artery disease (often abbreviated CAD). However, ischemic discomfort may be caused by a noncoronary artery impairment, such as aortic stenosis, hypertrophic cardiomyopathy, pulmonary hypertension, or anemia.

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

3. *What are the characteristics of typical angina pectoris?* Discomfort of myocardial ischemic origin (angina pectoris) is discomfort that is precipitated by effort or emotion and promptly relieved by rest, sublingual nitroglycerin (that is, nitroglycerin tablets that are placed under the tongue), or other rapidly acting nitrates. Typically, the discomfort is located in the chest (usually substernal) and described as pressing, crushing, squeezing, burning, aching, or oppressive. Sharp, sticking, or cramping discomfort is less common. Discomfort occurring with activity or emotion should be described specifically as to timing and usual inciting factors (type and intensity), character, location, radiation, duration, and response to nitrate treatment or rest.

4. *What is atypical angina? Atypical angina* describes discomfort or pain from myocardial ischemia that is felt in places other than the chest. The common sites of cardiac pain are the inner aspect of the left arm, neck, jaw(s), upper abdomen, and back, but the discomfort or pain can be elsewhere. When pain of cardiac ischemic origin presents in an atypical site in the absence of chest discomfort, the source of the pain may be difficult to diagnose. To represent atypical angina, your discomfort or pain should have precipitating and relieving factors similar to those of typical chest discomfort, and we must have objective medical evidence of myocardial ischemia; for example, ECG or ETT evidence or appropriate medically acceptable imaging.

5. *What is anginal equivalent?* Often, individuals with IHD will complain of shortness of breath (dyspnea) on exertion without chest pain or discomfort. In a minority of such situations, the shortness of breath is due to myocardial ischemia; this is called *anginal equivalent*. To represent anginal equivalent, your shortness of breath should have precipitating and relieving factors similar to those of typical chest discomfort, and we must have objective medical evidence of myocardial ischemia; for example, ECG or ETT evidence or appropriate medically acceptable imaging. In these situations, it is essential to establish objective evidence of myocardial ischemia to ensure that you do not have effort dyspnea due to non-ischemic or non-cardiac causes.

6. *What is variant angina?*

a. *Variant angina* (Prinzmetal's angina, vasospastic angina) refers to the occurrence of anginal episodes at rest, especially at night, accompanied by transitory ST segment elevation (or, at times, ST depression) on an ECG. It is due to severe spasm of a coronary artery, causing ischemia of the heart wall, and is often accompanied by major ventricular arrhythmias, such as

ventricular tachycardia. We will consider variant angina under 4.04 only if you have spasm of a coronary artery in relation to an obstructive lesion of the vessel. If you have an arrhythmia as a result of variant angina, we may consider your impairment under 4.05.

b. Variant angina may also occur in the absence of obstructive coronary disease. In this situation, an ETT will not demonstrate ischemia. The diagnosis will be established by showing the typical transitory ST segment changes during attacks of pain, and the absence of obstructive lesions shown by catheterization. Treatment in cases where there is no obstructive coronary disease is limited to medications that reduce coronary vasospasm, such as calcium channel blockers and nitrates. In such situations, we will consider the frequency of anginal episodes despite prescribed treatment when evaluating your residual functional capacity.

c. Vasospasm that is catheter-induced during coronary angiography is not variant angina.

7. *What is silent ischemia?*

a. Myocardial ischemia, and even myocardial infarction, can occur without perception of pain or any other symptoms; when this happens, we call it *silent ischemia*. Pain sensitivity may be altered by a variety of diseases, most notably diabetes mellitus and other neuropathic disorders. Individuals also vary in their threshold for pain.

b. Silent ischemia occurs most often in:

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

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

c. ST depression can result from a variety of factors, such as postural changes and variations in cardiac sympathetic tone. In addition, there are differences in how different Holter monitors record the electrical responses. Therefore, we do not consider the Holter monitor reliable for the diagnosis of silent ischemia except in the situation described in 4.00E7b(ii).

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

9. *How do we evaluate IHD using 4.04?*

a. We must have objective evidence, as described under 4.00C, that your symptoms are due to myocardial ischemia.

b. Listing-level changes on the ECG in 4.04A1 are the classically accepted changes of horizontal or downsloping ST depression occurring both during exercise and recovery. Although we recognize that ischemic changes may at times occur only during exercise or recovery, and may at times be upsloping with only junctional ST depression, such changes can be false positive; that is, occur in the absence of ischemia. Diagnosis of ischemia in this situation requires radionuclide or echocardiogram confirmation. See 4.00C12 and 4.00C13.

c. Also in 4.04A1, we require that the depression of the ST segment last for at least 1 minute of recovery because ST depression that occurs during exercise but that rapidly normalizes in recovery is a common false-positive response.

d. In 4.04A2, we specify that the ST elevation must be in non-infarct leads during both exercise and recovery. This is because, in the absence of ECG signs of prior infarction, ST elevation during exercise denotes ischemia, usually severe, requiring immediate termination of exercise. However, if there is baseline ST elevation in association with a prior infarction or ventricular aneurysm, further ST elevation during exercise does not necessarily denote ischemia and could be a false-positive ECG response. Diagnosis of ischemia in this situation requires radionuclide or echocardiogram confirmation. See 4.00C12 and 4.00C13.

e. Listing 4.04A3 requires a decrease in systolic blood pressure below the baseline level (taken in the standing position immediately prior to exercise) or below any systolic pressure reading recorded during exercise. This is the same finding required in 4.02B3c. See 4.00D4d for full details.

f. In 4.04B, each of the three ischemic episodes must require revascularization or be not amenable to treatment. *Revascularization* means angioplasty (with or without stent placement) or bypass surgery. However, reocclusion that occurs after a revascularization procedure but during the same hospitalization and that requires a second procedure during the same hospitalization will not be counted as another ischemic episode. Not amenable means that the revascularization procedure could not be done because of another medical impairment or because the vessel was not suitable for revascularization.

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

h. In 4.04C1 the term *nonbypassed* means that the blockage is in a vessel that is potentially bypassable; that is, large enough to be bypassed and considered to be a cause of your ischemia. These vessels are usually major arteries or one of a major artery's major branches. A vessel that has become obstructed again after angioplasty or stent placement and has remained obstructed or is not amenable to another revascularization is considered a nonbypassed vessel for purposes of this listing. When you have had revascularization, we will not use the pre-operative findings to assess the current severity of your coronary artery disease under 4.04C, although we will consider the severity and duration of your impairment prior to your surgery in making our determination or decision.

F. Evaluating Arrhythmias

1. *What is an arrhythmia?* An *arrhythmia* is a change in the regular beat of the heart. Your heart may seem to skip a beat or beat irregularly, very quickly (tachycardia), or very slowly (bradycardia).

2. *What are the different types of arrhythmias?*

a. There are many types of arrhythmias. Arrhythmias are identified by where they occur in the heart (atria or ventricles) and by what happens to the heart's rhythm when they occur.

b. Arrhythmias arising in the cardiac atria (upper chambers of the heart) are called atrial or supraventricular arrhythmias. Ventricular arrhythmias begin in the ventricles (lower chambers). In general, ventricular arrhythmias caused by heart disease are the most serious.

3. *How do we evaluate arrhythmias using 4.05?*

a. We will use 4.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator and you have uncontrolled recurrent episodes of syncope or near syncope. If your arrhythmias are controlled, we will evaluate your underlying heart disease using the appropriate listing. For other considerations when we evaluate arrhythmias in the presence of an implanted cardiac defibrillator, see 4.00F4.

b. We consider *near syncope* to be a period of altered consciousness, since syncope is a loss of consciousness or a faint. It is not merely a feeling of light-headedness, momentary weakness, or dizziness.

c. For purposes of 4.05, there must be a documented association between the syncope or near syncope and the recurrent arrhythmia. The recurrent arrhythmia, not some other cardiac or non-cardiac disorder, must be established as the cause of the associated symptom. This documentation of the association between the symptoms and the arrhythmia may come from the usual diagnostic methods, including Holter monitoring (also called ambulatory electrocardiography) and tilt-table testing with a concurrent ECG. Although an arrhythmia may be a coincidental finding on an ETT, we will not purchase an ETT to document the presence of a cardiac arrhythmia.

4. *What will we consider when you have an implanted cardiac defibrillator and you do not have arrhythmias that meet the requirements of 4.05?*

a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in individuals who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group at risk for sudden cardiac death consists of individuals with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in individuals with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator is a unique form of treatment; it rescues an individual from what may have been cardiac arrest. However, as a consequence of the shock (s), individuals may experience psychological distress, which we may evaluate under the mental disorders listings in 12.00ff.

b. Most implantable cardiac defibrillators have rhythm-correcting and pacemaker capabilities.

In some individuals, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. (The shock is like being kicked in the chest.) Implanted cardiac defibrillators may deliver inappropriate shocks, often repeatedly, in response to benign arrhythmias or electrical malfunction. Also, exposure to strong electrical or magnetic fields, such as from MRI (magnetic resonance imaging), can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. We must consider the frequency of, and the reason(s) for, the shocks when evaluating the severity and duration of your impairment.

c. In general, the exercise limitations imposed on individuals with an implanted cardiac defibrillator are those dictated by the underlying heart impairment. However, the exercise limitations may be greater when the implanted cardiac defibrillator delivers an inappropriate shock in response to the increase in heart rate with exercise, or when there is exercise-induced ventricular arrhythmia.

G. Evaluating Peripheral Vascular Disease

1. *What is peripheral vascular disease (PVD)?* Generally, *PVD* is any impairment that affects either the arteries (peripheral arterial disease) or the veins (venous insufficiency) in the extremities, particularly the lower extremities. The usual effect is blockage of the flow of blood either from the heart (arterial) or back to the heart (venous). If you have peripheral arterial disease, you may have pain in your calf after walking a distance that goes away when you rest (intermittent claudication); at more advanced stages, you may have pain in your calf at rest or you may develop ulceration or gangrene. If you have venous insufficiency, you may have swelling, varicose veins, skin pigmentation changes, or skin ulceration.

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

3. *What is brawny edema? Brawny edema* (4.11A) is swelling that is usually dense and feels firm due to the presence of increased connective tissue; it is also associated with characteristic skin pigmentation changes. It is not the same thing as pitting edema. Brawny edema generally does not pit (indent on pressure), and the terms are not interchangeable. Pitting edema does not satisfy the requirements of 4.11A.

4. *What is lymphedema and how will we evaluate it?*

a. *Lymphedema* is edema of the extremities due to a disorder of the lymphatic circulation; at its worst, it is called elephantiasis. Primary lymphedema is caused by abnormal development of lymph vessels and may be present at birth (congenital lymphedema), but more often develops during the teens (lymphedema praecox). It may also appear later, usually after age 35 (lymphedema tarda). Secondary lymphedema is due to obstruction or destruction of normal lymphatic channels due to tumor, surgery, repeated infections, or parasitic infection such as filariasis. Lymphedema most commonly affects one extremity.

b. Lymphedema does not meet the requirements of 4.11, although it may medically equal the severity of that listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular listing, such as 4.11, or a musculoskeletal listing, such as 1.02A or 1.03. If no

listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we assess your residual functional capacity.

5. *When will we purchase exercise Doppler studies for evaluating peripheral arterial disease (PAD)?* If we need additional evidence of your PAD, we will generally purchase exercise Doppler studies (see 4.00C16 and 4.00C17) when your resting ankle/brachial systolic blood pressure ratio is at least 0.50 but less than 0.80, and only rarely when it is 0.80 or above. We will not purchase exercise Doppler testing if you have a disease that results in abnormal arterial calcification or small vessel disease, but will use your resting toe systolic blood pressure or resting toe/brachial systolic blood pressure ratio. (See 4.00G7c and 4.00G8.) There are no current medical standards for evaluating exercise toe pressures. Because any exercise test stresses your entire cardiovascular system, we will purchase exercise Doppler studies only after an MC, preferably one with experience in the care of patients with cardiovascular disease, has determined that the test would not present a significant risk to you and that there is no other medical reason not to purchase the test (see 4.00C6, 4.00C7, and 4.00C8).

6. *Are there any other studies that are helpful in evaluating PAD?* Doppler studies done using a recording ultrasonic Doppler unit and strain-gauge plethysmography are other useful tools for evaluating PAD. A recording Doppler, which prints a tracing of the arterial pulse wave in the femoral, popliteal, dorsalis pedis, and posterior tibial arteries, is an excellent evaluation tool to compare wave forms in normal and compromised peripheral blood flow. Qualitative analysis of the pulse wave is very helpful in the overall assessment of the severity of the occlusive disease. Tracings are especially helpful in assessing severity if you have small vessel disease related to diabetes mellitus or other diseases with similar vascular changes, or diseases causing medial calcifications when ankle pressure is either normal or falsely high.

7. *How do we evaluate PAD under 4.12?*

a. The ankle blood pressure referred to in 4.12A and B is the higher of the pressures recorded from the posterior tibial and dorsalis pedis arteries in the affected leg. The higher pressure recorded from the two sites is the more significant measurement in assessing the extent of arterial insufficiency. Techniques for obtaining ankle systolic blood pressures include Doppler (See 4.00C16 and 4.00C17), plethysmographic studies, or other techniques. We will request any available tracings generated by these studies so that we can review them.

b. In 4.12A, the ankle/brachial systolic blood pressure ratio is the ratio of the systolic blood pressure at the ankle to the systolic blood pressure at the brachial artery; both taken at the same time while you are lying on your back. We do not require that the ankle and brachial pressures be taken on the same side of your body. This is because, as with the ankle pressure, we will use the higher brachial systolic pressure measured. Listing 4.12A is met when your resting ankle/brachial systolic blood pressure ratio is less than 0.50. If your resting ankle/brachial systolic blood pressure ratio is 0.50 or above, we will use 4.12B to evaluate the severity of your PAD, unless you also have a disease causing abnormal arterial calcification or small vessel disease, such as diabetes mellitus. See 4.00G7c and 4.00G8.

c. We will use resting toe systolic blood pressures or resting toe/brachial systolic blood pressure ratios (determined the same way as ankle/brachial ratios, see 4.00G7b) when you have intermittent claudication and a disease that results in abnormal arterial calcification (for example, Monckeberg's sclerosis or diabetes mellitus) or small vessel disease (for example,

diabetes mellitus). These diseases may result in misleadingly high blood pressure readings at the ankle. However, high blood pressures due to vascular changes related to these diseases seldom occur at the toe level. While the criteria in 4.12C and 4.12D are intended primarily for individuals who have a disease causing abnormal arterial calcification or small vessel disease, we may also use them for evaluating anyone with PAD.

8. *How are toe pressures measured?* Toe pressures are measured routinely in most vascular laboratories through one of three methods: most frequently, photoplethysmography; less frequently, plethysmography using strain gauge cuffs; and Doppler ultrasound. Toe pressure can also be measured by using any blood pressure cuff that fits snugly around the big toe and is neither too tight nor too loose. A neonatal cuff or a cuff designed for use on fingers or toes can be used in the measurement of toe pressure.

9. *How do we use listing 4.12 if you have had a peripheral graft?* Peripheral grafting serves the same purpose as coronary grafting; that is, to bypass a narrow or obstructed arterial segment. If intermittent claudication recurs or persists after peripheral grafting, we may purchase Doppler studies to assess the flow of blood through the bypassed vessel and to establish the current severity of the peripheral arterial impairment. However, if you have had peripheral grafting done for your PAD, we will not use the findings from before the surgery to assess the current severity of your impairment, although we will consider the severity and duration of your impairment prior to your surgery in making our determination or decision.

H. Evaluating Other Cardiovascular Impairments

1. *How will we evaluate hypertension?* Because *hypertension* (high blood pressure) generally causes disability through its effects on other body systems, we will evaluate it by reference to the specific body system(s) affected (heart, brain, kidneys, or eyes) when we consider its effects under the listings. We will also consider any limitations imposed by your hypertension when we assess your residual functional capacity.

2. *How will we evaluate symptomatic congenital heart disease? Congenital heart disease* is any abnormality of the heart or the major blood vessels that is present at birth. Because of improved treatment methods, more children with congenital heart disease are living to adulthood. Although some types of congenital heart disease may be corrected by surgery, many individuals with treated congenital heart disease continue to have problems throughout their lives (symptomatic congenital heart disease). If you have congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias, we will evaluate your impairment under 4.02 or 4.05. Otherwise, we will evaluate your impairment under 4.06.

3. *What is cardiomyopathy and how will we evaluate it? Cardiomyopathy* is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy). There are various types of cardiomyopathy, which fall into two major categories: *Ischemic* and *nonischemic* cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes several types: Dilated, hypertrophic, and restrictive. We will evaluate cardiomyopathy under 4.02, 4.04, 4.05, or 11.04, depending on its effects on you.

4. *How will we evaluate valvular heart disease?* We will evaluate valvular heart disease under the listing appropriate for its effect on you. Thus, we may use 4.02, 4.04, 4.05, 4.06, or an appropriate neurological listing in 11.00ff.

5. *What do we consider when we evaluate heart transplant recipients?*

a. After your heart transplant, we will consider you disabled for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year.

b. However, heart transplant patients generally meet our definition of disability before they undergo transplantation. We will determine the onset of your disability based on the facts in your case.

c. We will not assume that you became disabled when your name was placed on a transplant waiting list. This is because you may be placed on a waiting list soon after diagnosis of the cardiac disorder that may eventually require a transplant. Physicians recognize that candidates for transplantation often have to wait months or even years before a suitable donor heart is found, so they place their patients on the list as soon as permitted.

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

6. *When does an aneurysm have "dissection not controlled by prescribed treatment," as required under 4.10?* An aneurysm (or bulge in the aorta or one of its major branches) is *dissecting* when the inner lining of the artery begins to separate from the arterial wall. We consider the dissection not controlled when you have persistence of chest pain due to progression of the dissection, an increase in the size of the aneurysm, or compression of one or more branches of the aorta supplying the heart, kidneys, brain, or other organs. An aneurysm with dissection can cause heart failure, renal (kidney) failure, or neurological complications. If you have an aneurysm that does not meet the requirements of 4.10 and you have one or more of these associated conditions, we will evaluate the condition(s) using the appropriate listing.

7. *What is hyperlipidemia and how will we evaluate it? Hyperlipidemia* is the general term for an elevation of any or all of the lipids (fats or cholesterol) in the blood; for example, hypertriglyceridemia, hypercholesterolemia, and hyperlipoproteinemia. These disorders of lipoprotein metabolism and transport can cause defects throughout the body. The effects most likely to interfere with function are those produced by atherosclerosis (narrowing of the arteries) and coronary artery disease. We will evaluate your lipoprotein disorder by considering its effects on you.

8. *What is Marfan syndrome and how will we evaluate it?*

a. Marfan syndrome is a genetic connective tissue disorder that affects multiple body systems, including the skeleton, eyes, heart, blood vessels, nervous system, skin, and lungs. There is no specific laboratory test to diagnose Marfan syndrome. The diagnosis is generally made by medical history, including family history, physical examination, including an evaluation of the

ratio of arm/leg size to trunk size, a slit lamp eye examination, and a heart test(s), such as an echocardiogram. In some cases, a genetic analysis may be useful, but such analyses may not provide any additional helpful information.

b. The effects of Marfan syndrome can range from mild to severe. In most cases, the disorder progresses as you age. Most individuals with Marfan syndrome have abnormalities associated with the heart and blood vessels. Your heart's mitral valve may leak, causing a heart murmur. Small leaks may not cause symptoms, but larger ones may cause shortness of breath, fatigue, and palpitations. Another effect is that the wall of the aorta may be weakened and abnormally stretch (aortic dilation). This aortic dilation may tear, dissect, or rupture, causing serious heart problems or sometimes sudden death. We will evaluate the manifestations of your Marfan syndrome under the appropriate body system criteria, such as 4.10, or if necessary, consider the functional limitations imposed by your impairment.

I. Other Evaluation Issues

1. *What effect does obesity have on the cardiovascular system and how will we evaluate it?* Obesity is a medically determinable impairment that is often associated with disorders of the cardiovascular system. Disturbance of this system can be a major cause of disability if you have obesity. Obesity may affect the cardiovascular system because of the increased workload the additional body mass places on the heart. Obesity may make it harder for the chest and lungs to expand. This can mean that the respiratory system must work harder to provide needed oxygen. This in turn would make the heart work harder to pump blood to carry oxygen to the body. Because the body would be working harder at rest, its ability to perform additional work would be less than would otherwise be expected. Thus, the combined effects of obesity with cardiovascular impairments can be greater than the effects of each of the impairments considered separately. We must consider any additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular impairment or a listing-level cardiovascular impairment (or a combination of impairments that medically equals the severity of a listed impairment), and when we assess your residual functional capacity.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, we may need to defer evaluation of the impairment for a period of up to 3 months from the date treatment began to permit consideration of treatment effects, unless we can make a determination or decision using the evidence we have. See 4.00B4.

3. *How do we evaluate impairments that do not meet one of the cardiovascular listings?*

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

b. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairments(s) medically equals a listing. (See §§404.1526 and

416.926.) If you have a severe impairment(s) that does not meet or medically equal the criteria of a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. Therefore, we proceed to the fourth and, if necessary, the fifth steps of the sequential evaluation process in §§404.1520 and 416.920. If you are an adult, we use the rules in §§404.1594 or 416.994, as appropriate, when we decide whether you continue to be disabled.

4.01 Category of Impairments, Cardiovascular System

4.02 *Chronic heart failure* while on a regimen of prescribed treatment, with symptoms and signs described in 4.00D2. The required level of severity for this impairment is met when the requirements in *both A and B* are satisfied.

A. Medically documented presence of one of the following:

1. Systolic failure (see 4.00D1a(i)), with left ventricular end diastolic dimensions greater than 6.0 cm or ejection fraction of 30 percent or less during a period of stability (not during an episode of acute heart failure); or

2. Diastolic failure (see 4.00D1a(ii)), with left ventricular posterior wall plus septal thickness totaling 2.5 cm or greater on imaging, with an enlarged left atrium greater than or equal to 4.5 cm, with normal or elevated ejection fraction during a period of stability (not during an episode of acute heart failure);

AND

B. Resulting in one of the following:

1. Persistent symptoms of heart failure which very seriously limit the ability to independently initiate, sustain, or complete activities of daily living in an individual for whom an MC, preferably one experienced in the care of patients with cardiovascular disease, has concluded that the performance of an exercise test would present a significant risk to the individual; or

2. Three or more separate episodes of acute congestive heart failure within a consecutive 12-month period (see 4.00A3e), with evidence of fluid retention (see 4.00D2b(ii)) from clinical and imaging assessments at the time of the episodes, requiring acute extended physician intervention such as hospitalization or emergency room treatment for 12 hours or more, separated by periods of stabilization (see 4.00D4c); or

3. Inability to perform on an exercise tolerance test at a workload equivalent to 5 METs or less due to:

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

b. Three or more consecutive premature ventricular contractions (ventricular tachycardia), or increasing frequency of ventricular ectopy with at least 6 premature ventricular contractions per minute; or

c. Decrease of 10 mm Hg or more in systolic pressure below the baseline systolic blood pressure or the preceding systolic pressure measured during exercise (see 4.00D4d) due to

left ventricular dysfunction, despite an increase in workload; or

d. Signs attributable to inadequate cerebral perfusion, such as ataxic gait or mental confusion.

4.04 *Ischemic heart disease*, with symptoms due to myocardial ischemia, as described in 4.00E3-4.00E7, while on a regimen of prescribed treatment (see 4.00B3 if there is no regimen of prescribed treatment), with one of the following:

A. Sign-or symptom-limited exercise tolerance test demonstrating at least one of the following manifestations at a workload equivalent to 5 METs or less:

1. Horizontal or downsloping depression, in the absence of digitalis glycoside treatment or hypokalemia, of the ST segment of at least -0.10 millivolts (-1.0 mm) in at least 3 consecutive complexes that are on a level baseline in any lead other than aVR, and depression of at least -0.10 millivolts lasting for at least 1 minute of recovery; or

2. At least 0.1 millivolt (1 mm) ST elevation above resting baseline in non-infarct leads during both exercise and 1 or more minutes of recovery; or

3. Decrease of 10 mm Hg or more in systolic pressure below the baseline blood pressure or the preceding systolic pressure measured during exercise (see 4.00E9e) due to left ventricular dysfunction, despite an increase in workload; or

4. Documented ischemia at an exercise level equivalent to 5 METs or less on appropriate medically acceptable imaging, such as radionuclide perfusion scans or stress echocardiography.

OR

B. Three separate ischemic episodes, each requiring revascularization or not amenable to revascularization (see 4.00E9f), within a consecutive 12-month period (see 4.00A3e).

OR

C. Coronary artery disease, demonstrated by angiography (obtained independent of Social Security disability evaluation) or other appropriate medically acceptable imaging, and in the absence of a timely exercise tolerance test or a timely normal drug-induced stress test, an MC, preferably one experienced in the care of patients with cardiovascular disease, has concluded that performance of exercise tolerance testing would present a significant risk to the individual, with both 1 and 2:

1. Angiographic evidence showing:

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

b. 70 percent or more narrowing of another nonbypassed coronary artery; or

c. 50 percent or more narrowing involving a long (greater than 1 cm) segment of a nonbypassed coronary artery; or

d. 50 percent or more narrowing of at least two nonbypassed coronary arteries; or

e. 70 percent or more narrowing of a bypass graft vessel; and

2. Resulting in very serious limitations in the ability to independently initiate, sustain, or complete activities of daily living.

4.05 *Recurrent arrhythmias*, not related to reversible causes, such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled (see 4.00A3f), recurrent (see 4.00A3c) episodes of cardiac syncope or near syncope (see 4.00F3b), despite prescribed treatment (see 4.00B3 if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography, or by other appropriate medically acceptable testing, coincident with the occurrence of syncope or near syncope (see 4.00F3c).

4.06 *Symptomatic congenital heart disease* (cyanotic or acyanotic), documented by appropriate medically acceptable imaging (see 4.00A3d) or cardiac catheterization, with one of the following:

A. Cyanosis at rest, and:

1. Hematocrit of 55 percent or greater; or

2. Arterial O_2 saturation of less than 90 percent in room air, or resting arterial PO_2 of 60 Torr or less.

OR

B. Intermittent right-to-left shunting resulting in cyanosis on exertion (e.g., Eisenmenger's physiology) and with arterial PO_2 of 60 Torr or less at a workload equivalent to 5 METs or less.

OR

C. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure.

4.09 *Heart transplant*. Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under the appropriate listing.

4.10 *Aneurysm of aorta or major branches*, due to any cause (e.g., atherosclerosis, cystic medial necrosis, Marfan syndrome, trauma), demonstrated by appropriate medically acceptable imaging, with dissection not controlled by prescribed treatment (see 4.00H6).

4.11 *Chronic venous insufficiency* of a lower extremity with incompetency or obstruction of the deep venous system and one of the following:

A. Extensive brawny edema (see 4.00G3) involving at least two-thirds of the leg between the ankle and knee or the distal one-third of the lower extremity between the ankle and hip.

OR

B. Superficial varicosities, stasis dermatitis, and either recurrent ulceration or persistent ulceration that has not healed following at least 3 months of prescribed treatment.

4.12 *Peripheral arterial disease*, as determined by appropriate medically acceptable imaging (see 4.00A3d, 4.00G2, 4.00G5, and 4.00G6), causing intermittent claudication (see 4.00G1) and one of the following:

A. Resting ankle/brachial systolic blood pressure ratio of less than 0.50.

OR

B. Decrease in systolic blood pressure at the ankle on exercise (see 4.00G7a and 4.00C16-4.00C17) of 50 percent or more of pre-exercise level and requiring 10 minutes or more to return to pre-exercise level.

OR

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

OR

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

5.00 Digestive System

A. *Disorders of the digestive system* which result in a marked impairment usually do so because of interference with nutrition, multiple recurrent inflammatory lesions, or complications of disease, such as fistulae, abscesses, or recurrent obstruction. Such complications usually respond to treatment. These complications must be shown to persist on repeated examinations despite therapy for a reasonable presumption to be made that a marked impairment will last for a continuous period of at least 12 months.

B. *Malnutrition or weight loss from gastrointestinal disorders.* When the primary disorder of the digestive tract has been established (e.g. enterocolitis, chronic pancreatitis, postgastrointestinal resection, or esophageal stricture, stenosis, or obstruction), the resultant interference with nutrition will be considered under the criteria in 5.08. This will apply whether the weight loss is due to primary or secondary disorders of malabsorption, malassimilation or obstruction.

C. *Surgical diversion of the intestinal tract,* including colostomy or ileostomy, are not listed since they do not represent impairments which preclude all work activity if the individual is able to maintain adequate nutrition and function of the stoma. Dumping syndrome which may follow gastric resection rarely represents a marked impairment which would continue for 12 months. Peptic ulcer disease with recurrent ulceration after definitive surgery ordinarily responds to treatment. To be considered a severe impairment which will last for at least 12 months, a recurrent ulcer after definitive surgery must be demonstrated, despite therapy, by repeated appropriate medically acceptable imaging of the upper gastrointestinal tract or by gastroscopic examinations. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means

that the technique used is the proper one to support the evaluation and diagnosis of the impairment. Definitive surgical procedures are those designed to control the ulcer disease process (*i.e.*, vagotomy and pyloroplasty, subtotal gastrectomy, etc.). Simple closure of a perforated ulcer does not constitute definitive surgical therapy for peptic ulcer disease.

5.01 Category of Impairments, Digestive System

5.02 *Recurrent upper gastrointestinal hemorrhage from undetermined cause* with anemia manifested by hematocrit of 30 percent or less on repeated examinations.

5.03 *Stricture, stenosis, or obstruction of the esophagus (demonstrated by endoscopy or other appropriate medically acceptable imaging)* with weight loss as described under listing 5.08.

5.04 *Peptic ulcer disease (demonstrated by endoscopy or other appropriate medically acceptable imaging).* With:

A. Recurrent ulceration after definitive surgery persistent despite therapy; or

B. Inoperable fistula formation; or

C. Recurrent obstruction demonstrated by endoscopy or other appropriate medically acceptable imaging; or,

D. Weight loss as described under §5.08.

5.05 *Chronic liver disease (e.g., portal, postnecrotic, or biliary cirrhosis; chronic active hepatitis; Wilson's disease).* With:

A. Esophageal varices (demonstrated by endoscopy or other appropriate medically acceptable imaging) with a documented history of massive hemorrhage attributable to these varices. Consider under a disability for 3 years following the last massive hemorrhage; thereafter, evaluate the residual impairment; or

B. Performance of a shunt operation for esophageal varices. Consider under a disability for 3 years following surgery; thereafter, evaluate the residual impairment; or

C. Serum bilirubin of 2.5 mg. per deciliter (100 ml.) or greater persisting on repeated examinations for at least 5 months; or

D. Ascites, not attributable to other causes, recurrent or persisting for at least 5 months, demonstrated by abdominal paracentesis or associated with persistent hypoalbuminemia of 3.0 gm. per deciliter (100 ml.) or less; or

E. Hepatic encephalopathy. Evaluate under the criteria in listing 12.02; or

F. Confirmation of chronic liver disease by liver biopsy (obtained independent of Social Security disability evaluation) and one of the following:

1. Ascites not attributable to other causes, recurrent or persisting for at least 3 months, demonstrated by abdominal paracentesis or associated with persistent hypoalbuminemia of

3.0 gm. per deciliter (100 ml.) or less; or

2. Serum bilirubin of 2.5 mg. per deciliter (100 ml) or greater on repeated examinations for at least 3 months; or

3. Hepatic cell necrosis or inflammation, persisting for at least 3 months, documented by repeated abnormalities of prothrombin time and enzymes indicative of hepatic dysfunction.

5.06 *Chronic ulcerative or granulomatous colitis (demonstrated by endoscopy, barium enema, biopsy, or operative findings).* With:

A. Recurrent bloody stools documented on repeated examinations and anemia manifested by hematocrit of 30 percent or less on repeated examinations; or

B. Persistent or recurrent systemic manifestations, such as arthritis, iritis, fever, or liver dysfunction, not attributable to other causes; or

C. Intermittent obstruction due to intractable abscess, fistula formation, or stenosis; or

D. Recurrence of findings of A, B, or C above after total colectomy; or

E. Weight loss as described under §5.08.

5.07 *Regional enteritis (demonstrated by operative findings, barium studies, biopsy, or endoscopy).* With:

A. Persistent or recurrent intestinal obstruction evidenced by abdominal pain, distention, nausea, and vomiting and accompanied by stenotic areas of small bowel with proximal intestinal dilation; or

B. Persistent or recurrent systemic manifestations such as arthritis, iritis, fever, or liver dysfunction, not attributable to other causes; or

C. Intermittent obstruction due to intractable abscess or fistula formation; or

D. Weight loss as described under §5.08.

5.08 *Weight loss due to any persisting gastrointestinal disorder:* (The following weights are to be demonstrated to have persisted for at least 3 months despite prescribed therapy and expected to persist at this level for at least 12 months.) With:

A. Weight equal to or less than the values specified in table I or II; or

B. Weight equal to or less than the values specified in table III or IV and one of the following abnormal findings on repeated examinations:

1. Serum albumin of 3.0 gm. per deciliter (100 ml.) or less; or

2. Hematocrit of 30 percent or less; or

3. Serum calcium of 8.0 mg. per deciliter (100 ml.) (4.0 mEq./L) or less; or

4. Uncontrolled diabetes mellitus due to pancreatic dysfunction with repeated hyperglycemia, hypoglycemia, or ketosis; or

5. Fat in stool of 7 gm. or greater per 24-hour stool specimen; or

6. Nitrogen in stool of 3 gm, or greater per 24-hour specimen; or

7. Persistent or recurrent ascites or edema not attributable to other causes.

Tables of weight reflecting malnutrition scaled according to height and sex—To be used only in connection with 5.08.

Table I—Men

Height (inches)[1]	Weight (pounds)
61	90
62	92
63	94
64	97
65	99
66	102
67	106
68	109
69	112
70	115
71	118
72	122
73	125
74	128
75	131
76	134

[1]Height measured without shoes.

Table II—Women

Height (inches)[1]	Weight (pounds)
58	77
59	79
60	82
61	84
62	86
63	89
64	91
65	94

66	98
67	101
68	104
69	107
70	110
71	114
72	117
73	120

[1]Height measured without shoes.

Table III—Men

Height (inches)[1]	Weight (pounds)
61	95
62	98
63	100
64	103
65	106
66	109
67	112
68	116
69	119
70	122
71	126
72	129
73	133
74	136
75	139
76	143

[1]Height measured without shoes.

Table IV—Women

Height (inches)[1]	Weight (pounds)
58	82
59	84
60	87
61	89
62	92
63	94
64	97
65	100
66	104

67	107
68	111
69	114
70	117
71	121
72	124
73	128

[1]Height measured without shoes.

5.09 *Liver transplant.* Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment(s).

6.00 Genitourinary Impairments

A. What impairments do these listings cover?

1. We use these listings to evaluate genitourinary impairments resulting from chronic renal disease.

2. We use the criteria in 6.02 to evaluate renal dysfunction due to any chronic renal disease, such as chronic glomerulonephritis, hypertensive renal vascular disease, diabetic nephropathy, chronic obstructive uropathy, and hereditary nephropathies.

3. We use the criteria in 6.06 to evaluate nephrotic syndrome due to glomerular disease.

B. What do we mean by the following terms in these listings?

1. *Anasarca* is generalized massive edema (swelling).

2. *Creatinine* is a normal product of muscle metabolism.

3. *Creatinine clearance test* is a test for renal function based on the rate at which creatinine is excreted by the kidney.

4. *Diastolic hypertension* is elevated diastolic blood pressure.

5. *Fluid overload syndrome* associated with renal disease occurs when there is excessive sodium and water retention in the body that cannot be adequately removed by the diseased kidneys. Symptoms and signs of vascular congestion may include fatigue, shortness of breath, hypertension, congestive heart failure, accumulation of fluid in the abdomen (ascites) or chest (pleural effusions), and peripheral edema.

6. *Glomerular disease* can be classified into two broad categories, nephrotic and nephritic. Nephrotic conditions are associated with increased urinary protein excretion and nephritic conditions are associated with inflammation of the internal structures of the kidneys.

7. *Hemodialysis,* or *dialysis,* is the removal of toxic metabolic byproducts from the blood by

diffusion in an artificial kidney machine.

8. *Motor neuropathy* is neuropathy or polyneuropathy involving only the motor nerves.

9. *Nephrotic syndrome* is a general name for a group of diseases involving defective kidney glomeruli, characterized by heavy proteinuria, hypoalbuminemia, hyperlipidemia, and varying degrees of edema.

10. *Neuropathy* is a problem in peripheral nerve function (that is, in any part of the nervous system except the brain and spinal cord) that causes pain, numbness, tingling, and muscle weakness in various parts of the body.

11. *Osteitis fibrosa* is fibrous degeneration with weakening and deformity of bones.

12. *Osteomalacia* is a softening of the bones.

13. *Osteoporosis* is a thinning of the bones with reduction in bone mass resulting from the depletion of calcium and bone protein.

14. *Pathologic fractures* are fractures resulting from weakening of the bone structure by pathologic processes, such as osteomalacia and osteoporosis.

15. *Peritoneal dialysis* is a method of hemodialysis in which the dialyzing solution is introduced into and removed from the peritoneal cavity either continuously or intermittently.

16. *Proteinuria* is excess protein in the urine.

17. *Renal* means pertaining to the kidney.

18. *Renal osteodystrophy* refers to a variety of bone disorders usually caused by chronic kidney failure.

19. *Sensory neuropathy* is neuropathy or polyneuropathy that involves only the sensory nerves.

20. *Serum albumin* is a major plasma protein that is responsible for much of the plasma colloidal osmotic pressure and serves as a transport protein.

21. *Serum creatinine* is the amount of creatinine in the blood and is measured to evaluate kidney function.

C. What evidence do we need?

1. We need a longitudinal record of your medical history that includes records of treatment, response to treatment, hospitalizations, and laboratory evidence of renal disease that indicates its progressive nature. The laboratory or clinical evidence will indicate deterioration of renal function, such as elevation of serum creatinine.

2. We generally need a longitudinal clinical record covering a period of at least 3 months of

observations and treatment, unless we can make a fully favorable determination or decision without it. The record should include laboratory findings, such as serum creatinine or serum albumin values, obtained on more than one examination over the 3-month period.

3. When you are undergoing dialysis, we should have laboratory findings showing your renal function before you started dialysis.

4. The medical evidence establishing the clinical diagnosis of nephrotic syndrome must include a description of the extent of edema, including pretibial, periorbital, or presacral edema. The medical evidence should describe any ascites, pleural effusion, or pericardial effusion. Levels of serum albumin and proteinuria must be included.

5. If a renal biopsy has been performed, the evidence should include a copy of the report of the microscopic examination of the specimen. However, if we do not have a copy of the microscopic examination in the evidence, we can accept a statement from an acceptable medical source that a biopsy was performed, with a description of the results.

D. How do we consider the effects of treatment?

We consider factors such as the:

1. Type of therapy.

2. Response to therapy.

3. Side effects of therapy.

4. Effects of any post-therapeutic residuals.

5. Expected duration of treatment.

E. What other things do we consider when we evaluate your chronic renal disease under specific listings?

1. *Chronic hemodialysis or peritoneal dialysis* (6.02A). A report from an acceptable medical source describing the chronic renal disease and the need for ongoing dialysis is sufficient to satisfy the requirements in 6.02A.

2. *Kidney transplantation* (6.02B). If you have undergone kidney transplantation, we will consider you to be disabled for 12 months following the surgery because, during the first year, there is a greater likelihood of rejection of the organ and recurrent infection. After the first year posttransplantation, we will base our continuing disability evaluation on your residual impairment(s). We will include absence of symptoms, signs, and laboratory findings indicative of kidney dysfunction in our consideration of whether medical improvement (as defined in §§404.1579(b)(1) and (c)(1), 404.1594(b)(1) and (c)(1), 416.994(b)(1)(i) and (b)(2)(i), or 416.994a, as appropriate) has occurred. We will consider the:

a. Occurrence of rejection episodes.

b. Side effects of immunosuppressants, including corticosteroids.

c. Frequency of any renal infections.

d. Presence of systemic complications such as other infections, neuropathy, or deterioration of other organ systems.

3. *Renal osteodystrophy* (6.02C1). This condition is bone deterioration resulting from chronic renal disease. The resultant bone disease includes the impairments described in 6.02C1.

4. *Persistent motor or sensory neuropathy* (6.02C2). The longitudinal clinical record must show that the neuropathy is a "severe" impairment as defined in §§404.1520(c) and 416.920(c) that has lasted or can be expected to last for a continuous period of at least 12 months.

5. *Nephrotic syndrome* (6.06). The longitudinal clinical record should include a description of prescribed therapy, response to therapy, and any side effects of therapy. In order for your nephrotic syndrome to meet 6.06A or B, the medical evidence must document that you have the appropriate laboratory findings required by these listings and that your anasarca has persisted for at least 3 months despite prescribed therapy. However, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in your case record. We may also evaluate complications of your nephrotic syndrome, such as orthostatic hypotension, recurrent infections, or venous thromboses, under the appropriate listing for the resultant impairment.

F. What does the term "persistent" mean in these listings?

Persistent means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been at, or is expected to be at, the level specified in the listing for a continuous period of at least 12 months.

G. How do we evaluate impairments that do not meet one of the genitourinary listings?

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

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (See §§404.1526 and 416.926.) If you have a severe impairment(s) that does not meet or medically equal the criteria of a listing, you may or may not have the residual functional capacity to engage in substantial gainful activity. Therefore, we proceed to the fourth and, if necessary, the fifth steps of the sequential evaluation process in §§404.1520 and 416.920. When we decide whether you continue to be disabled, we use the rules in §§404.1579(b)(1) and (c)(1), 404.1594(b)(1) and (c)(1), 416.994(b)(1)(i) and (b)(2)(i), or 416.994a, as appropriate.

6.01 Category of Impairments, Genitourinary Impairments

6.02 *Impairment of renal function*, due to any chronic renal disease that has lasted or can be expected to last for a continuous period of at least 12 months. With:

A. *Chronic hemodialysis or peritoneal dialysis* (see 6.00E1).

or

B. *Kidney transplantation.* Consider under a disability for 12 months following surgery; thereafter, evaluate the residual impairment (see 6.00E2).

or

C. *Persistent elevation of serum creatinine* to 4 mg per deciliter (dL) (100 ml) or greater or *reduction of creatinine clearance* to 20 ml per minute or less, over at least 3 months, with one of the following:

1. Renal osteodystrophy (see 6.00E3) manifested by severe bone pain and appropriate medically acceptable imaging demonstrating abnormalities such as osteitis fibrosa, significant osteoporosis, osteomalacia, or pathologic fractures; or

2. Persistent motor or sensory neuropathy (see 6.00E4); or

3. Persistent fluid overload syndrome with:

a. Diastolic hypertension greater than or equal to diastolic blood pressure of 110 mm Hg; or

b. Persistent signs of vascular congestion despite prescribed therapy (see 6.00B5); or

4. Persistent anorexia with recent weight loss and current weight meeting the values in 5.08, table III or IV.

6.06 *Nephrotic syndrome,* with anasarca, persisting for at least 3 months despite prescribed therapy (see 6.00E5). With:

A. Serum albumin of 3.0 g per dL (100 ml) or less and proteinuria of 3.5 g or greater per 24 hours.

or

B. Proteinuria of 10.0 g or greater per 24 hours.

7.00 Hematological Disorders

A. *Impairment caused by anemia* should be evaluated according to the ability of the individual to adjust to the reduced oxygen carrying capacity of the blood. A gradual reduction in red cell mass, even to very low values, is often well tolerated in individuals with a healthy cardiovascular system.

B. *Chronicity is indicated by* persistence of the condition for at least 3 months. The laboratory findings cited must reflect the values reported on more than one examination over that 3-month period. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

C. *Sickle cell disease* refers to a chronic hemolytic anemia associated with sickle cell hemoglobin, either homozygous or in combination with thalassemia or with another abnormal hemoglobin (such as C or F).

Appropriate hematologic evidence for sickle cell disease, such as hemoglobin electrophoresis, must be included. Vasoocclusive or aplastic episodes should be documented by description of severity, frequency, and duration.

Major visceral episodes include meningitis, osteomyelitis, pulmonary infections or infarctions, cerebrovascular accidents, congestive heart failure, genito-urinary involvement, etc.

D. *Coagulation defects.* Chronic inherited coagulation disorders must be documented by appropriate laboratory evidence. Prophylactic therapy such as with antihemophilic globulin (AHG) concentrate does not in itself imply severity.

7.01 Category of Impairments, Hemic and Lymphatic System

7.02 *Chronic anemia (hematocrit persisting at 30 percent or less due to any cause).* With:

A. Requirement of one or more blood transfusions on an average of at least once every 2 months; or

B. Evaluation of the resulting impairment under criteria for the affected body system.

7.05 *Sickle cell disease, or one of its variants.* With:

A. Documented painful (thrombotic) crises occurring at least three times during the 5 months prior to adjudication; or

B. Requiring extended hospitalization (beyond emergency care) at least three times during the 12 months prior to adjudication; or

C. Chronic, severe anemia with persistence of hematocrit of 26 percent or less; or

D. Evaluate the resulting impairment under the criteria for the affected body system.

7.06 *Chronic thrombocytopenia (due to any cause)* with platelet counts repeatedly below 40,000/cubic millimeter. With:

A. At least one spontaneous hemorrhage, requiring transfusion, within 5 months prior to adjudication; or

B. Intracranial bleeding within 12 months prior to adjudication.

7.07 *Hereditary telangiectasia* with hemorrhage requiring transfusion at least three times during the 5 months prior to adjudication.

7.08 *Coagulation defects (hemophilia or a similar disorder)* with spontaneous hemorrhage requiring transfusion at least three times during the 5 months prior to adjudication.

7.09 *Polycythemia vera (with erythrocytosis, splenomegaly, and leukocytosis or thrombocytosis).* Evaluate the resulting impairment under the criteria for the affected body system.

7.10 *Myelofibrosis (myeloproliferative syndrome).* With:

A. Chronic anemia. Evaluate according to the criteria of §7.02; or

B. Documented recurrent systemic bacterial infections occurring at least 3 times during the 5 months prior to adjudication; or

C. Intractable bone pain with radiologic evidence of osteosclerosis.

7.11-7.14 [Reserved]

7.15 *Chronic granulocytopenia (due to any cause).* With both A and B:

A. Absolute neutrophil counts repeatedly below 1,000 cells/cubic millimeter; and

B. Documented recurrent systemic bacterial infections occurring at least 3 times during the 5 months prior to adjudication.

7.16 [Reserved]

7.17 *Aplastic anemias* with bone marrow or stem cell transplantation. Consider under a disability for 12 months following transplantation; thereafter, evaluate according to the primary characteristics of the residual impairment.

8.00 Skin Disorders

A. *What skin disorders do we evaluate with these listings?* We use these listings to evaluate skin disorders that may result from hereditary, congenital, or acquired pathological processes. The kinds of impairments covered by these listings are: Ichthyosis, bullous diseases, chronic infections of the skin or mucous membranes, dermatitis, hidradenitis suppurativa, genetic photosensitivity disorders, and burns.

B. *What documentation do we need?* When we evaluate the existence and severity of your skin disorder, we generally need information about the onset, duration, frequency of flareups, and prognosis of your skin disorder; the location, size, and appearance of lesions; and, when applicable, history of exposure to toxins, allergens, or irritants, familial incidence, seasonal variation, stress factors, and your ability to function outside of a highly protective environment. To confirm the diagnosis, we may need laboratory findings (for example, results of a biopsy obtained independently of Social Security disability evaluation or blood tests) or evidence from other medically acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

C. *How do we assess the severity of your skin disorder(s)?* We generally base our assessment of severity on the extent of your skin lesions, the frequency of flareups of your skin lesions, how your symptoms (including pain) limit you, the extent of your treatment, and how your treatment affects you.

1. *Extensive skin lesions.* Extensive skin lesions are those that involve multiple body sites or critical body areas, and result in a very serious limitation. Examples of extensive skin lesions that result in a very serious limitation include but are not limited to:

a. Skin lesions that interfere with the motion of your joints and that very seriously limit your use of more than one extremity; that is, two upper extremities, two lower extremities, or one upper and one lower extremity.

b. Skin lesions on the palms of both hands that very seriously limit your ability to do fine and gross motor movements.

c. Skin lesions on the soles of both feet, the perineum, or both inguinal areas that very seriously limit your ability to ambulate.

2. *Frequency of flareups.* If you have skin lesions, but they do not meet the requirements of any of the listings in this body system, you may still have an impairment that prevents you from doing any gainful activity when we consider your condition over time, especially if your flareups result in extensive skin lesions, as defined in C1 of this section. Therefore, if you have frequent flareups, we may find that your impairment(s) is medically equal to one of these listings even though you have some periods during which your condition is in remission. We will consider how frequent and serious your flareups are, how quickly they resolve, and how you function between flareups to determine whether you have been unable to do any gainful activity for a continuous period of at least 12 months or can be expected to be unable to do any gainful activity for a continuous period of at least 12 months. We will also consider the frequency of your flareups when we determine whether you have a severe impairment and when we need to assess your residual functional capacity.

3. *Symptoms (including pain).* Symptoms (including pain) may be important factors contributing to the severity of your skin disorder(s). We assess the impact of symptoms as explained in §§404.1528, 404.1529, 416.928, and 416.929 of this chapter.

4. *Treatment.* We assess the effects of medication, therapy, surgery, and any other form of treatment you receive when we determine the severity and duration of your impairment(s). Skin disorders frequently respond to treatment; however, response to treatment can vary widely, with some impairments becoming resistant to treatment. Some treatments can have side effects that can in themselves result in limitations.

a. We assess the effects of continuing treatment as prescribed by determining if there is improvement in the symptoms, signs, and laboratory findings of your disorder, and if you experience side effects that result in functional limitations. To assess the effects of your treatment, we may need information about:

i. The treatment you have been prescribed (for example, the type, dosage, method, and frequency of administration of medication or therapy);

ii. Your response to the treatment;

iii. Any adverse effects of the treatment; and

iv. The expected duration of the treatment.

b. Because treatment itself or the effects of treatment may be temporary, in most cases sufficient time must elapse to allow us to evaluate the impact and expected duration of treatment and its side effects. Except under 8.07 and 8.08, you must follow continuing treatment as prescribed for at least 3 months before your impairment can be determined to meet the requirements of a skin disorder listing. (See 8.00H if you are not undergoing treatment or did not have treatment for 3 months.) We consider your specific response to treatment when we evaluate the overall severity of your impairment.

D. *How do we assess impairments that may affect the skin and other body systems?* When your impairment affects your skin and has effects in other body systems, we first evaluate the predominant feature of your impairment under the appropriate body system. Examples include, but are not limited to the following.

1. *Tuberous sclerosis* primarily affects the brain. The predominant features are seizures, which we evaluate under the neurological listings in 11.00, and developmental delays or other mental disorders, which we evaluate under the mental disorders listings in 12.00.

2. *Malignant tumors of the skin* (for example, malignant melanomas) are cancers, or neoplastic diseases, which we evaluate under the listings in 13.00.

3. *Connective tissue disorders and other immune system disorders* (for example, systemic lupus erythematosus, scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than one body system. We first evaluate these disorders under the immune system listings in 14.00. We evaluate lupus erythematosus under 14.02, scleroderma under 14.04, symptomatic HIV infection under 14.08, and Sjögren's syndrome under 14.03, 14.09, or any other appropriate listing in section 14.00.

4. *Disfigurement or deformity* resulting from skin lesions may result in loss of sight, hearing, speech, and the ability to chew (mastication). We evaluate these impairments and their effects under the special senses and speech listings in 2.00 and the digestive system listings in 5.00. Facial disfigurement or other physical deformities may also have effects we evaluate under the mental disorders listings in 12.00, such as when they affect mood or social functioning.

E. *How do we evaluate genetic photosensitivity disorders?*

1. *Xeroderma pigmentosum (XP).* When you have XP, your impairment meets the requirements of 8.07A if you have clinical and laboratory findings showing that you have the disorder. (See 8.00E3.) People who have XP have a lifelong hypersensitivity to all forms of ultraviolet light and generally lead extremely restricted lives in highly protective environments in order to prevent skin cancers from developing. Some people with XP also experience problems with their eyes, neurological problems, mental disorders, and problems in other body systems.

2. *Other genetic photosensitivity disorders.* Other genetic photosensitivity disorders may vary in their effects on different people, and may not result in an inability to engage in any gainful activity for a continuous period of at least 12 months. Therefore, if you have a genetic photosensitivity disorder other than XP (established by clinical and laboratory findings as described in 8.00E3), you must show that you have either extensive skin lesions or an inability to function outside of a highly protective environment to meet the requirements of 8.07B. You must also show that your impairment meets the duration requirement. By *inability to function*

outside of a highly protective environment we mean that you must avoid exposure to ultraviolet light (including sunlight passing through windows and light from unshielded fluorescent bulbs), wear protective clothing and eyeglasses, and use opaque broad-spectrum sunscreens in order to avoid skin cancer or other serious effects. Some genetic photosensitivity disorders can have very serious effects in other body systems, especially special senses and speech (2.00), neurological (11.00), mental (12.00), and neoplastic (13.00). We will evaluate the predominant feature of your impairment under the appropriate body system, as explained in 8.00D.

3. *Clinical and laboratory findings.*

a. *General.* We need documentation from an acceptable medical source, as defined in §§404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment. In general, we must have evidence of appropriate laboratory testing showing that you have XP or another genetic photosensitivity disorder. We will find that you have XP or another genetic photosensitivity disorder based on a report from an acceptable medical source indicating that you have the impairment, supported by definitive genetic laboratory studies documenting appropriate chromosomal changes, including abnormal DNA repair or another DNA or genetic abnormality specific to your type of photosensitivity disorder.

b. *What we will accept as medical evidence instead of the actual laboratory report.* When we do not have the actual laboratory report, we need evidence from an acceptable medical source that includes appropriate clinical findings for your impairment and that is persuasive that a positive diagnosis has been confirmed by appropriate laboratory testing at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive genetic laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

F. *How do we evaluate burns?* Electrical, chemical, or thermal burns frequently affect other body systems; for example, musculoskeletal, special senses and speech, respiratory, cardiovascular, renal, neurological, or mental. Consequently, we evaluate burns the way we evaluate other disorders that can affect the skin and other body systems, using the listing for the predominant feature of your impairment. For example, if your soft tissue injuries are under continuing surgical management (as defined in 1.00M), we will evaluate your impairment under 1.08. However, if your burns do not meet the requirements of 1.08 and you have extensive skin lesions that result in a very serious limitation (as defined in 8.00C1) that has lasted or can be expected to last for a continuous period of at least 12 months, we will evaluate them under 8.08.

G. *How do we determine if your skin disorder(s) will continue at a disabling level of severity in order to meet the duration requirement?* For all of these skin disorder listings except 8.07 and 8.08, we will find that your impairment meets the duration requirement if your skin disorder results in extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. By *persist,* we mean that the longitudinal clinical record shows that, with few exceptions, your lesions have been at the level of severity specified in the listing. For 8.07A, we will presume that you meet the duration requirement. For 8.07B and 8.08, we will consider all of the relevant medical and other information in your case record to determine whether your skin disorder meets the duration requirement.

H. *How do we assess your skin disorder(s) if your impairment does not meet the requirements of one of these listings?*

1. These listings are only examples of common skin disorders that we consider severe enough to prevent you from engaging in any gainful activity. For most of these listings, if you do not have continuing treatment as prescribed, if your treatment has not lasted for at least 3 months, or if you do not have extensive skin lesions that have persisted for at least 3 months, your impairment cannot meet the requirements of these skin disorder listings. (This provision does not apply to 8.07 and 8.08.) However, we may still find that you are disabled because your impairment(s) meets the requirements of a listing in another body system or medically equals the severity of a listing. (See §§404.1526 and 416.926 of this chapter.) We may also find you disabled at the last step of the sequential evaluation process.

2. If you have not received ongoing treatment or do not have an ongoing relationship with the medical community despite the existence of a severe impairment(s), or if your skin lesions have not persisted for at least 3 months but you are undergoing continuing treatment as prescribed, you may still have an impairment(s) that meets a listing in another body system or that medically equals a listing. If you do not have an impairment(s) that meets or medically equals a listing, we will assess your residual functional capacity and proceed to the fourth and, if necessary, the fifth step of the sequential evaluation process in §§404.1520 and 416.920 of this chapter. When we decide whether you continue to be disabled, we use the rules in §§404.1594 and 416.994 of this chapter.

8.01 Category of Impairments, Skin Disorders

8.02 *Ichthyosis,* with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.03 *Bullous disease* (for example, pemphigus, erythema multiforme bullosum, epidermolysis bullosa, bullous pemphigoid, dermatitis herpetiformis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.04 *Chronic infections of the skin or mucous membranes,* with extensive fungating or extensive ulcerating skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.05 *Dermatitis* (for example, psoriasis, dyshidrosis, atopic dermatitis, exfoliative dermatitis, allergic contact dermatitis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.06 *Hidradenitis suppurativa,* with extensive skin lesions involving both axillae, both inguinal areas or the perineum that persist for at least 3 months despite continuing treatment as prescribed.

8.07 *Genetic photosensitivity disorders*, established as described in 8.00E.

A. Xeroderma pigmentosum. Consider the individual disabled from birth.

B. Other genetic photosensitivity disorders, with:

1. Extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months, or

2. Inability to function outside of a highly protective environment for a continuous period of at least 12 months (see 8.00E2).

8.08 *Burns,* with extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months (see 8.00F).

9.00 Endocrine System

Cause of impairment. Impairment is caused by overproduction or underproduction of hormones, resulting in structural or functional changes in the body. Where involvement of other organ systems has occurred as a result of a primary endocrine disorder, these impairments should be evaluated according to the criteria under the appropriate sections. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

9.01 Category of Impairments, Endocrine System

9.02 *Thyroid Disorders.*

Evaluate the resulting impairment under the criteria for the affected body system.

9.03 *Hyperparathyroidism.* With:

A. Generalized decalcification of bone on appropriate medically acceptable imaging study and elevation of plasma calcium to 11 mg. per deciliter (100 ml.) or greater; or

B. A resulting impairment. Evaluate according to the criteria in the affected body system.

9.04 *Hypoparathyroidism.* With:

A. Severe recurrent tetany; or

B. Recurrent generalized convulsions; or

C. Lenticular cataracts. Evaluate under the criteria in 2.00ff.

9.05 *Neurohypophyseal insufficiency (diabetes insipidus).* With urine specific gravity of 1.005 or below, persistent for at least 3 months and recurrent dehydration.

9.06 *Hyperfunction of the adrenal cortex.* Evaluate the resulting impairment under the criteria for the affected body system.

9.08 *Diabetes mellitus.* With:

A. Neuropathy demonstrated by significant and persistent disorganization of motor function in two extremities resulting in sustained disturbance of gross and dexterous movements, or gait and station (see 11.00C); or

B. Acidosis occurring at least on the average of once every 2 months documented by appropriate blood chemical tests (pH or PCO_2 or bicarbonate levels); or

C. Retinitis proliferans; evaluate the visual impairment under the criteria in 2.02, 2.03, or 2.04.

10.00 Impairments That Affect Multiple Body Systems

A. What Impairment Do We Evaluate Under This Body System?

1. *General.* We evaluate non-mosaic Down syndrome under this body system.

2. *What is Down syndrome?* Down syndrome is a condition in which there are three copies of chromosome 21 within the cells of the body instead of the normal two copies per cell. The three copies may be separate (trisomy), or one chromosome 21 copy may be attached to a different chromosome (translocation). This extra chromosomal material changes the orderly development of the body and brain. Down syndrome is characterized by a complex of physical characteristics, delayed physical development, and mental retardation. Down syndrome exists in non-mosaic and mosaic forms.

3. *What is non-mosaic Down syndrome?*

a. Non-mosaic Down syndrome occurs when you have an extra copy of chromosome 21 in every cell of your body. At least 98 percent of people with Down syndrome have this form (which includes either trisomy or translocation type chromosomal abnormalities). Virtually all cases of non-mosaic Down syndrome affect the mental, neurological, and skeletal systems, and they are often accompanied by heart disease, impaired vision, hearing problems, and other conditions.

b. We evaluate adults with confirmed non-mosaic Down syndrome under 10.06. If you have confirmed non-mosaic Down syndrome, we consider you disabled from birth.

4. *What is mosaic Down syndrome?*

a. Mosaic Down syndrome occurs when you have some cells with the normal two copies of chromosome 21 and some cells with an extra copy of chromosome 21. When this occurs, there is a mixture of two types of cells. Mosaic Down syndrome occurs in only 1-2 percent of people with Down syndrome, and there is a wide range in the level of severity of the impairment. Mosaic Down syndrome can be profound and disabling, but it can also be so slight as to be undetected clinically.

b. We evaluate adults with confirmed mosaic Down syndrome under the listing criteria in any affected body system(s) on an individual case basis, as described in 10.00C.

B. What Documentation Do We Need To Establish That You Have Non-Mosaic Down Syndrome?

1. *General.* We need documentation from an acceptable medical source, as defined in §§404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment.

2. *Definitive chromosomal analysis*. We will find that you have non-mosaic Down syndrome based on a report from an acceptable medical source that indicates that you have the impairment and that includes the actual laboratory report of definitive chromosomal analysis showing that you have the impairment. *Definitive chromosomal analysis* means karyotype analysis. In this case, we do not additionally require a clinical description of the diagnostic physical features of your impairment.

3. *What if we do not have the results of definitive chromosomal analysis?* When we do not have the actual laboratory report of definitive chromosomal analysis, we need evidence from an acceptable medical source that includes a clinical description of the diagnostic physical features of Down syndrome, and that is persuasive that a positive diagnosis has been confirmed by definitive chromosomal analysis at some time prior to our evaluation. To be persuasive, the report must state that definitive chromosomal analysis was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record; for example, evidence showing your limitations in adaptive functioning or signs of a mental disorder that can be associated with non-mosaic Down syndrome, your educational history, or the results of psychological testing.

C. How Do We Evaluate Other Impairments That Affect Multiple Body Systems?

1. Non-mosaic Down syndrome (10.06) is an example of an impairment that commonly affects multiple body systems and that we consider significant enough to prevent you from doing any gainful activity. If you have a different severe impairment(s) that affects multiple body systems, we must also consider whether your impairment(s) meets the criteria of a listing in another body system.

2. There are many other impairments that can cause deviation from, or interruption of, the normal function of the body or interfere with development; for example, congenital anomalies, chromosomal disorders, dysmorphic syndromes, metabolic disorders, and perinatal infectious diseases. In these impairments, the degree of deviation or interruption may vary widely from individual to individual. Therefore, the resulting functional limitations and the progression of those limitations also vary widely. For this reason, we evaluate the specific effects of these impairments on you under the listing criteria in any affected body system(s) on an individual case basis. Examples of such impairments include triple X syndrome (XXX syndrome), fragile X syndrome, phenylketonuria (PKU), caudal regression syndrome, and fetal alcohol syndrome.

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

10.01 Category of Impairments, Impairments That Affect Multiple Body Systems

10.06 *Non-mosaic Down syndrome,* established as described in 10.00B.

11.00 Neurological

A. *Epilepsy.* In epilepsy, regardless of etiology, degree of impairment will be determined according to type, frequency, duration, and sequelae of seizures. At least one detailed description of a typical seizure is required. Such description includes the presence or absence of aura, tongue bites, sphincter control, injuries associated with the attack, and postictal phenomena. The reporting physician should indicate the extent to which description of seizures reflects his own observations and the source of ancillary information. Testimony of persons other than the claimant is essential for description of type and frequency of seizures if professional observation is not available.

Under 11.02 and 11.03, the criteria can be applied only if the impairment persists despite the fact that the individual is following prescribed antiepileptic treatment. Adherence to prescribed antiepileptic therapy can ordinarily be determined from objective clinical findings in the report of the physician currently providing treatment for epilepsy. Determination of blood levels of phenytoin sodium or other antiepileptic drugs may serve to indicate whether the prescribed medication is being taken. When seizures are occurrring at the frequency stated in 11.02 or 11.03, evalution of the severity of the impairment must include consideration of the serum drug levels. Should serum drug levels appear therapeutically inadequate, consideration should be given as to whether this is caused by individual idiosyncrasy in absorption of metabolism of the drug. Blood drug levels should be evaluated in conjunction with all the other evidence to determine the extent of compliance. When the reported blood drug levels are low, therefore, the information obtained from the treating source should include the physician's statement as to why the levels are low and the results of any relevant diagnostic studies concerning the blood levels. Where adequate seizure control is obtained only with unusually large doses, the possibility of impairment resulting from the side effects of this medication must be also assessed. Where documentation shows that use of alcohol or drugs affects adherence to prescribed therapy or may play a part in the precipitation of seizures, this must also be considered in the overall assessment of impairment level.

B. *Brain tumors.* We evaluate malignant brain tumors under the criteria in 13.13. For benign brain tumors, we determine the severity and duration of the impairment on the basis of symptoms, signs, and laboratory findings (11.05).

C. *Persistent disorganization of motor function* in the form of paresis or paralysis, tremor or other involuntary movements, ataxia and sensory disturbances (any or all of which may be due to cerebral, cerebellar, brain stem, spinal cord, or peripheral nerve dysfunction) which occur singly or in various combinations, frequently provides the sole or partial basis for decision in cases of neurological impairment. The assessment of impairment depends on the degree of interference with locomotion and/or interference with the use of fingers, hands, and arms.

D. *In conditions which are episodic in character,* such as multiple sclerosis or myasthenia gravis, consideration should be given to frequency and duration of exacerbations, length of remissions, and permanent residuals.

E. *Multiple sclerosis.* The major criteria for evaluating impairment caused by multiple sclerosis are discussed in listing 11.09. Paragraph A provides criteria for evaluating disorganization of motor function and gives reference to 11.04B (11.04B then refers to 11.00C). Paragraph B provides references to other listings for evaluating visual or mental impairments caused by multiple sclerosis. Paragraph C provides criteria for evaluating the impairment of individuals who do not have muscle weakness or other significant disorganization of motor function at rest, but who do develop muscle weakness on activity as a result of fatigue.

Use of the criteria in 11.09C is dependent upon (1) documenting a diagnosis of multiple sclerosis, (2) obtaining a description of fatigue considered to be characteristic of multiple sclerosis, and (3) obtaining evidence that the system has actually become fatigued. The evaluation of the magnitude of the impairment must consider the degree of exercise and the severity of the resulting muscle weakness.

The criteria in 11.09C deals with motor abnormalities which occur on activity. If the disorganization of motor function is present at rest, paragraph A must be used, taking into account any further increase in muscle weakness resulting from activity.

Sensory abnormalities may occur, particularly involving central visual acuity. The decrease in visual acuity may occur after brief attempts at activity involving near vision, such as reading. This decrease in visual acuity may not persist when the specific activity is terminated, as with rest, but is predictably reproduced with resumption of the activity. The impairment of central visual acuity in these cases should be evaluated under the criteria in listing 2.02, taking into account the fact that the decrease in visual acuity will wax and wane.

Clarification of the evidence regarding central nervous system dysfunction responsible for the symptoms may require supporting technical evidence of functional impairment such as evoked response tests during exercise.

F. *Traumatic brain injury (TBI).* The guidelines for evaluating impairments caused by cerebral trauma are contained in 11.18. Listing 11.18 states that cerebral trauma is to be evaluated under 11.02, 11.03, 11.04, and 12.02, as applicable.

TBI may result in neurological and mental impairments with a wide variety of posttraumatic symptoms and signs. The rate and extent of recovery can be highly variable and the long-term outcome may be difficult to predict in the first few months post-injury. Generally, the neurological impairment(s) will stabilize more rapidly than any mental impairment(s). Sometimes a mental impairment may appear to improve immediately following TBI and then worsen, or, conversely, it may appear much worse initially but improve after a few months. Therefore, the mental findings immediately following TBI may not reflect the actual severity of your mental impairment(s). The actual severity of a mental impairment may not become apparent until 6 months post-injury.

In some cases, evidence of a profound neurological impairment is sufficient to permit a finding of disability within 3 months post-injury. If a finding of disability within 3 months post-injury is not possible based on any neurological impairment(s), we will defer adjudication of the claim until we obtain evidence of your neurological or mental impairments at least 3 months post-injury. If a finding of disability still is not possible at that time, we will again defer adjudication of the claim until we obtain evidence at least 6 months post-injury. At that time, we will fully evaluate any neurological and mental impairments and adjudicate the claim.

G. *Amyotrophic Lateral Sclerosis (ALS).* 1. Amyotrophic lateral sclerosis (ALS), sometimes called Lou Gehrig's disease, is a progressive, invariably fatal neurological disease that attacks the nerve cells (motor neurons) responsible for controlling voluntary muscles. Eventually, all muscles under voluntary control are affected, and individuals with ALS ultimately lose their ability to move their arms and legs, and their capacity to swallow, speak, and breath. Most people with ALS die from respiratory failure. There is currently no cure for ALS, and most treatments are designed only to relieve symptoms and improve the quality of life.

2. Diagnosis of ALS is based on history, neurological findings consistent with the diagnosis of ALS, and electrophysiological and neuroimaging testing to rule out other impairments that may cause similar signs and symptoms. The diagnosis may also be supported by electrophysiological studies (electromyography or nerve conduction studies), but these tests may be negative or only suggestive of the diagnosis. There is no single test that establishes the existence of ALS.

3. For purposes of 11.10, documentation of the diagnosis must be by generally accepted methods consistent with the prevailing state of medical knowledge and clinical practice. The evidence should include documentation of a clinically appropriate medical history, neurological findings consistent with the diagnosis of ALS, and the results of any electrophysiological and neuroimaging testing.

11.01 Category of Impairments, Neurological

11.02 *Epilepsy—convulsive epilepsy, (grand mal or psychomotor), documented by detailed description of a typical seizure pattern, including all associated phenomena; occurring more frequently than once a month in spite of at least 3 months of prescribed treatment.* With:

A. Daytime episodes (loss of consciousness and convulsive seizures) or

B. Nocturnal episodes manifesting residuals which interfere significantly with activity during the day.

11.03 *Epilepsy—nonconvulsive epilepsy (petit mal, psychomotor, or focal), documented by detailed description of a typical seizure pattern, including all associated phenomena; occurring more frequently than once weekly in spite of at least 3 months of prescribed treatment.* With alteration of awareness or loss of consciousness and transient postictal manifestations of unconventional behavior or significant interference with activity during the day.

11.04 *Central nervous system vascular accident.* With one of the following more than 3 months post-vascular accident:

A. Sensory or motor aphasia resulting in ineffective speech or communication; or

B. Significant and persistent disorganization of motor function in two extremities, resulting in sustained disturbance of gross and dexterous movements, or gait and station (see 11.00C).

11.05 *Benign brain tumors.* Evaluate under 11.02, 11.03, 11.04, or the criteria of the affected body system.

11.06 *Parkinsonian syndrome* with the following signs: Significant rigidity, brady kinesia, or tremor in two extremities, which, singly or in combination, result in sustained disturbance of gross and dexterous movements, or gait and station.

11.07 *Cerebral palsy.* With:

A. IQ of 70 or less; or

B. Abnormal behavior patterns, such as destructiveness or emotional instability: or

C. Significant interference in communication due to speech, hearing, or visual defect; or

D. Disorganization of motor function as described in 11.04B.

11.08 *Spinal cord or nerve root lesions, due to any cause* with disorganization of motor function as described in 11.04B.

11.09 *Multiple sclerosis.* With:

A. Disorganization of motor function as described in 11.04B; or

B. Visual or mental impairment as described under the criteria in 2.02, 2.03, 2.04, or 12.02; or

C. Significant, reproducible fatigue of motor function with substantial muscle weakness on repetitive activity, demonstrated on physical examination, resulting from neurological dysfunction in areas of the central nervous system known to be pathologically involved by the multiple sclerosis process.

11.10 *Amyotrophic lateral sclerosis* established by clinical and laboratory findings, as described in 11.00G.

11.11 *Anterior poliomyelitis.* With:

A. Persistent difficulty with swallowing or breathing; or

B. Unintelligible speech; or

C. Disorganization of motor function as described in 11.04B.

11.12 *Myasthenia gravis.* With:

A. Significant difficulty with speaking, swallowing, or breathing while on prescribed therapy; or

B. Significant motor weakness of muscles of extremities on repetitive activity against resistance while on prescribed therapy.

11.13 *Muscular dystrophy* with disorganization of motor function as described in 11.04B.

11.14 *Peripheral neuropathies.*

With disorganization of motor function as described in 11.04B, in spite of prescribed treatment.

11.15 [Reserved]

11.16 *Subacute combined cord degeneration (pernicious anemia) with disorganization of motor function as decribed in 11.04B or 11.15B, not significantly improved by prescribed treatment.*

11.17 *Degenerative disease not listed elsewhere, such as Huntington's chorea, Friedreich's*

ataxia, and spino-cerebellar degeneration. With:

A. Disorganization of motor function as described in 11.04B; or

B. Chronic brain syndrome. Evaluate under 12.02.

11.18 *Cerebral trauma:*

Evaluate under the provisions of 11.02, 11.03, 11.04 and 12.02, as applicable.

11.19 *Syringomyelia.*

With:

A. Significant bulbar signs; or

B. Disorganization of motor function as described in 11.04B.

12.00 Mental Disorders

A. *Introduction.* The evaluation of disability on the basis of mental disorders requires documentation of a medically determinable impairment(s), consideration of the degree of limitation such impairment(s) may impose on your ability to work, and consideration of whether these limitations have lasted or are expected to last for a continuous period of at least 12 months. The listings for mental disorders are arranged in nine diagnostic categories: Organic mental disorders (12.02); schizophrenic, paranoid and other psychotic disorders (12.03); affective disorders (12.04); mental retardation (12.05); anxiety-related disorders (12.06); somatoform disorders (12.07); personality disorders (12.08); substance addiction disorders (12.09); and autistic disorder and other pervasive developmental disorders (12.10). Each listing, except 12.05 and 12.09, consists of a statement describing the disorder(s) addressed by the listing, paragraph A criteria (a set of medical findings), and paragraph B criteria (a set of impairment-related functional limitations). There are additional functional criteria (paragraph C criteria) in 12.02, 12.03, 12.04, and 12.06, discussed herein. We will assess the paragraph B criteria before we apply the paragraph C criteria. We will assess the paragraph C criteria only if we find that the paragraph B criteria are not satisfied. We will find that you have a listed impairment if the diagnostic description in the introductory paragraph and the criteria of both paragraphs A and B (or A and C, when appropriate) of the listed impairment are satisfied.

The criteria in paragraph A substantiate medically the presence of a particular mental disorder. Specific symptoms, signs, and laboratory findings in the paragraph A criteria of any of the listings in this section cannot be considered in isolation from the description of the mental disorder contained at the beginning of each listing category. Impairments should be analyzed or reviewed under the mental category(ies) indicated by the medical findings. However, we may also consider mental impairments under physical body system listings, using the concept of medical equivalence, when the mental disorder results in physical dysfunction. (See, for instance, 12.00D12 regarding the evaluation of anorexia nervosa and other eating disorders.)

The criteria in paragraphs B and C describe impairment-related functional limitations that are incompatible with the ability to do any gainful activity. The functional limitations in paragraphs B and C must be the result of the mental disorder described in the diagnostic description, that

is manifested by the medical findings in paragraph A.

The structure of the listing for mental retardation (12.05) is different from that of the other mental disorders listings. Listing 12.05 contains an introductory paragraph with the diagnostic description for mental retardation. It also contains four sets of criteria (paragraphs A through D). If your impairment satisfies the diagnostic description in the introductory paragraph and any one of the four sets of criteria, we will find that your impairment meets the listing. Paragraphs A and B contain criteria that describe disorders we consider severe enough to prevent your doing any gainful activity without any additional assessment of functional limitations. For paragraph C, we will assess the degree of functional limitation the additional impairment(s) imposes to determine if it significantly limits your physical or mental ability to do basic work activities, *i.e.,* is a "severe" impairment(s), as defined in §§404.1520(c) and 416.920(c). If the additional impairment(s) does not cause limitations that are "severe" as defined in §§404.1520(c) and 416.920(c), we will not find that the additional impairment(s) imposes "an additional and significant work-related limitation of function," even if you are unable to do your past work because of the unique features of that work. Paragraph D contains the same functional criteria that are required under paragraph B of the other mental disorders listings.

The structure of the listing for substance addiction disorders, 12.09, is also different from that for the other mental disorder listings. Listing 12.09 is structured as a reference listing; that is, it will only serve to indicate which of the other listed mental or physical impairments must be used to evaluate the behavioral or physical changes resulting from regular use of addictive substances.

The listings are so constructed that an individual with an impairment(s) that meets or is equivalent in severity to the criteria of a listing could not reasonably be expected to do any gainful activity. These listings are only examples of common mental disorders that are considered severe enough to prevent an individual from doing any gainful activity. When you have a medically determinable severe mental impairment that does not satisfy the diagnostic description or the requirements of the paragraph A criteria of the relevant listing, the assessment of the paragraph B and C criteria is critical to a determination of equivalence.

If your impairment(s) does not meet or is not equivalent in severity to the criteria of any listing, you may or may not have the residual functional capacity (RFC) to do substantial gainful activity (SGA). The determination of mental RFC is crucial to the evaluation of your capacity to do SGA when your impairment(s) does not meet or equal the criteria of the listings, but is nevertheless severe.

RFC is a multidimensional description of the work-related abilities you retain in spite of your medical impairments. An assessment of your RFC complements the functional evaluation necessary for paragraphs B and C of the listings by requiring consideration of an expanded list of work-related capacities that may be affected by mental disorders when your impairment(s) is severe but neither meets nor is equivalent in severity to a listed mental disorder.

B. *Need for medical evidence.* We must establish the existence of a medically determinable impairment(s) of the required duration by medical evidence consisting of symptoms, signs, and laboratory findings (including psychological test findings). Symptoms are your own description of your physical or mental impairment(s). Psychiatric signs are medically demonstrable phenomena that indicate specific psychological abnormalities, e.g., abnormalities of behavior, mood, thought, memory, orientation, development, or perception, as described by an

appropriate medical source. Symptoms and signs generally cluster together to constitute recognizable mental disorders described in the listings. The symptoms and signs may be intermittent or continuous depending on the nature of the disorder.

C. *Assessment of severity.* We measure severity according to the functional limitations imposed by your medically determinable mental impairment(s). We assess functional limitations using the four criteria in paragraph B of the listings: Activities of daily living; social functioning; concentration, persistence, or pace; and episodes of decompensation. Where we use "marked" as a standard for measuring the degree of limitation, it means more than moderate but less than extreme. A marked limitation may arise when several activities or functions are impaired, or even when only one is impaired, as long as the degree of limitation is such as to interfere seriously with your ability to function independently, appropriately, effectively, and on a sustained basis. See §§404.1520a and 416.920a.

1. *Activities of daily living* include adaptive activities such as cleaning, shopping, cooking, taking public transportation, paying bills, maintaining a residence, caring appropriately for your grooming and hygiene, using telephones and directories, and using a post office. In the context of your overall situation, we assess the quality of these activities by their independence, appropriateness, effectiveness, and sustainability. We will determine the extent to which you are capable of initiating and participating in activities independent of supervision or direction.

We do not define "marked" by a specific number of different activities of daily living in which functioning is impaired, but by the nature and overall degree of interference with function. For example, if you do a wide range of activities of daily living, we may still find that you have a marked limitation in your daily activities if you have serious difficulty performing them without direct supervision, or in a suitable manner, or on a consistent, useful, routine basis, or without undue interruptions or distractions.

2. *Social functioning* refers to your capacity to interact independently, appropriately, effectively, and on a sustained basis with other individuals. Social functioning includes the ability to get along with others, such as family members, friends, neighbors, grocery clerks, landlords, or bus drivers. You may demonstrate impaired social functioning by, for example, a history of altercations, evictions, firings, fear of strangers, avoidance of interpersonal relationships, or social isolation. You may exhibit strength in social functioning by such things as your ability to initiate social contacts with others, communicate clearly with others, or interact and actively participate in group activities. We also need to consider cooperative behaviors, consideration for others, awareness of others' feelings, and social maturity. Social functioning in work situations may involve interactions with the public, responding appropriately to persons in authority (e.g., supervisors), or cooperative behaviors involving coworkers.

We do not define "marked" by a specific number of different behaviors in which social functioning is impaired, but by the nature and overall degree of interference with function. For example, if you are highly antagonistic, uncooperative, or hostile but are tolerated by local storekeepers, we may nevertheless find that you have a marked limitation in social functioning because that behavior is not acceptable in other social contexts.

3. *Concentration, persistence, or pace* refers to the ability to sustain focused attention and concentration sufficiently long to permit the timely and appropriate completion of tasks commonly found in work settings. Limitations in concentration, persistence, or pace are best observed in work settings, but may also be reflected by limitations in other settings. In addition,

major limitations in this area can often be assessed through clinical examination or psychological testing. Wherever possible, however, a mental status examination or psychological test data should be supplemented by other available evidence.

On mental status examinations, concentration is assessed by tasks such as having you subtract serial sevens or serial threes from 100. In psychological tests of intelligence or memory, concentration is assessed through tasks requiring short-term memory or through tasks that must be completed within established time limits.

In work evaluations, concentration, persistence, or pace is assessed by testing your ability to sustain work using appropriate production standards, in either real or simulated work tasks (e.g., filing index cards, locating telephone numbers, or disassembling and reassembling objects). Strengths and weaknesses in areas of concentration and attention can be discussed in terms of your ability to work at a consistent pace for acceptable periods of time and until a task is completed, and your ability to repeat sequences of action to achieve a goal or an objective.

We must exercise great care in reaching conclusions about your ability or inability to complete tasks under the stresses of employment during a normal workday or work week based on a time-limited mental status examination or psychological testing by a clinician, or based on your ability to complete tasks in other settings that are less demanding, highly structured, or more supportive. We must assess your ability to complete tasks by evaluating all the evidence, with an emphasis on how independently, appropriately, and effectively you are able to complete tasks on a sustained basis.

We do not define "marked" by a specific number of tasks that you are unable to complete, but by the nature and overall degree of interference with function. You may be able to sustain attention and persist at simple tasks but may still have difficulty with complicated tasks. Deficiencies that are apparent only in performing complex procedures or tasks would not satisfy the intent of this paragraph B criterion. However, if you can complete many simple tasks, we may nevertheless find that you have a marked limitation in concentration, persistence, or pace if you cannot complete these tasks without extra supervision or assistance, or in accordance with quality and accuracy standards, or at a consistent pace without an unreasonable number and length of rest periods, or without undue interruptions or distractions.

4. *Episodes of decompensation* are exacerbations or temporary increases in symptoms or signs accompanied by a loss of adaptive functioning, as manifested by difficulties in performing activities of daily living, maintaining social relationships, or maintaining concentration, persistence, or pace. Episodes of decompensation may be demonstrated by an exacerbation in symptoms or signs that would ordinarily require increased treatment or a less stressful situation (or a combination of the two). Episodes of decompensation may be inferred from medical records showing significant alteration in medication; or documentation of the need for a more structured psychological support system (e.g., hospitalizations, placement in a halfway house, or a highly structured and directing household); or other relevant information in the record about the existence, severity, and duration of the episode.

The term *repeated episodes of decompensation, each of extended duration* in these listings means three episodes within 1 year, or an average of once every 4 months, each lasting for at least 2 weeks. If you have experienced more frequent episodes of shorter duration or less

frequent episodes of longer duration, we must use judgment to determine if the duration and functional effects of the episodes are of equal severity and may be used to substitute for the listed finding in a determination of equivalence.

D. *Documentation.* The evaluation of disability on the basis of a mental disorder requires sufficient evidence to (1) establish the presence of a medically determinable mental impairment(s), (2) assess the degree of functional limitation the impairment(s) imposes, and (3) project the probable duration of the impairment(s). See §§404.1512 and 416.912 for a discussion of what we mean by "evidence" and how we will assist you in developing your claim. Medical evidence must be sufficiently complete and detailed as to symptoms, signs, and laboratory findings to permit an independent determination. In addition, we will consider information you provide from other sources when we determine how the established impairment(s) affects your ability to function. We will consider all relevant evidence in your case record.

1. *Sources of evidence.*

a. *Medical evidence.* There must be evidence from an acceptable medical source showing that you have a medically determinable mental impairment. See §§404.1508, 404.1513, 416.908, and 416.913. We will make every reasonable effort to obtain all relevant and available medical evidence about your mental impairment(s), including its history, and any records of mental status examinations, psychological testing, and hospitalizations and treatment. Whenever possible, and appropriate, medical source evidence should reflect the medical source's considerations of information from you and other concerned persons who are aware of your activities of daily living; social functioning; concentration, persistence, or pace; or episodes of decompensation. Also, in accordance with standard clinical practice, any medical source assessment of your mental functioning should take into account any sensory, motor, or communication abnormalities, as well as your cultural and ethnic background.

b. *Information from the individual.* Individuals with mental impairments can often provide accurate descriptions of their limitations. The presence of a mental impairment does not automatically rule you out as a reliable source of information about your own functional limitations. When you have a mental impairment and are willing and able to describe your limitations, we will try to obtain such information from you. However, you may not be willing or able to fully or accurately describe the limitations resulting from your impairment(s). Thus, we will carefully examine the statements you provide to determine if they are consistent with the information about, or general pattern of, the impairment as described by the medical and other evidence, and to determine whether additional information about your functioning is needed from you or other sources.

c. *Other information.* Other professional health care providers (e.g., psychiatric nurse, psychiatric social worker) can normally provide valuable functional information, which should be obtained when available and needed. If necessary, information should also be obtained from nonmedical sources, such as family members and others who know you, to supplement the record of your functioning in order to establish the consistency of the medical evidence and longitudinality of impairment severity, as discussed in 12.00D2. Other sources of information about functioning include, but are not limited to, records from work evaluations and rehabilitation progress notes.

2. *Need for longitudinal evidence.* Your level of functioning may vary considerably over time.

The level of your functioning at a specific time may seem relatively adequate or, conversely, rather poor. Proper evaluation of your impairment(s) must take into account any variations in the level of your functioning in arriving at a determination of severity over time. Thus, it is vital to obtain evidence from relevant sources over a sufficiently long period prior to the date of adjudication to establish your impairment severity.

3. *Work attempts.* You may have attempted to work or may actually have worked during the period of time pertinent to the determination of disability. This may have been an independent attempt at work or it may have been in conjunction with a community mental health or sheltered program, and it may have been of either short or long duration. Information concerning your behavior during any attempt to work and the circumstances surrounding termination of your work effort are particularly useful in determining your ability or inability to function in a work setting. In addition, we should also examine the degree to which you require special supports (such as those provided through supported employment or transitional employment programs) in order to work.

4. *Mental status examination.* The mental status examination is performed in the course of a clinical interview and is often partly assessed while the history is being obtained. A comprehensive mental status examination generally includes a narrative description of your appearance, behavior, and speech; thought process (e.g., loosening of associations); thought content (e.g., delusions); perceptual abnormalities (e.g., hallucinations); mood and affect (e.g., depression, mania); sensorium and cognition (e.g., orientation, recall, memory, concentration, fund of information, and intelligence); and judgment and insight. The individual case facts determine the specific areas of mental status that need to be emphasized during the examination.

5. *Psychological testing.*

a. Reference to a "standardized psychological test" indicates the use of a psychological test measure that has appropriate validity, reliability, and norms, and is individually administered by a qualified specialist. By "qualified," we mean the specialist must be currently licensed or certified in the State to administer, score, and interpret psychological tests and have the training and experience to perform the test.

b. Psychological tests are best considered as standardized sets of tasks or questions designed to elicit a range of responses. Psychological testing can also provide other useful data, such as the specialist's observations regarding your ability to sustain attention and concentration, relate appropriately to the specialist, and perform tasks independently (without prompts or reminders). Therefore, a report of test results should include both the objective data and any clinical observations.

c. The salient characteristics of a good test are: (1) Validity, *i.e.,* the test measures what it is supposed to measure; (2) reliability, *i.e.,* the consistency of results obtained over time with the same test and the same individual; (3) appropriate normative data, *i.e.,* individual test scores can be compared to test data from other individuals or groups of a similar nature, representative of that population; and (4) wide scope of measurement, *i.e.,* the test should measure a broad range of facets/aspects of the domain being assessed. In considering the validity of a test result, we should note and resolve any discrepancies between formal test results and the individual's customary behavior and daily activities.

6. *Intelligence tests.*

a. The results of standardized intelligence tests may provide data that help verify the presence of mental retardation or organic mental disorder, as well as the extent of any compromise in cognitive functioning. However, since the results of intelligence tests are only part of the overall assessment, the narrative report that accompanies the test results should comment on whether the IQ scores are considered valid and consistent with the developmental history and the degree of functional limitation.

b. Standardized intelligence test results are essential to the adjudication of all cases of mental retardation that are not covered under the provisions of 12.05A. Listing 12.05A may be the basis for adjudicating cases where the results of standardized intelligence tests are unavailable, e.g., where your condition precludes formal standardized testing.

c. Due to such factors as differing means and standard deviations, identical IQ scores obtained from different tests do not always reflect a similar degree of intellectual functioning. The IQ scores in 12.05 reflect values from tests of general intelligence that have a mean of 100 and a standard deviation of 15; e.g., the Wechsler series. IQs obtained from standardized tests that deviate from a mean of 100 and a standard deviation of 15 require conversion to a percentile rank so that we can determine the actual degree of limitation reflected by the IQ scores. In cases where more than one IQ is customarily derived from the test administered, e.g., where verbal, performance, and full scale IQs are provided in the Wechsler series, we use the lowest of these in conjunction with 12.05.

d. Generally, it is preferable to use IQ measures that are wide in scope and include items that test both verbal and performance abilities. However, in special circumstances, such as the assessment of individuals with sensory, motor, or communication abnormalities, or those whose culture and background are not principally English-speaking, measures such as the Test of Nonverbal Intelligence, Third Edition (TONI-3), Leiter International Performance Scale-Revised (Leiter-R), or Peabody Picture Vocabulary Test—Third Edition (PPVT-III) may be used.

e. We may consider exceptions to formal standardized psychological testing when an individual qualified by training and experience to perform such an evaluation is not available, or in cases where appropriate standardized measures for your social, linguistic, and cultural background are not available. In these cases, the best indicator of severity is often the level of adaptive functioning and how you perform activities of daily living and social functioning.

7. *Personality measures and projective testing techniques.* Results from standardized personality measures, such as the Minnesota Multiphasic Personality Inventory-Revised (MMPI-II), or from projective types of techniques, such as the Rorschach and the Thematic Apperception Test (TAT), may provide useful data for evaluating several types of mental disorders. Such test results may be useful for disability evaluation when corroborated by other evidence, including results from other psychological tests and information obtained in the course of the clinical evaluation, from treating and other medical sources, other professional health care providers, and nonmedical sources. Any inconsistency between test results and clinical history and observation should be explained in the narrative description.

8. *Neuropsychological assessments.* Comprehensive neuropsychological examinations may be used to establish the existence and extent of compromise of brain function, particularly in

cases involving organic mental disorders. Normally, these examinations include assessment of cerebral dominance, basic sensation and perception, motor speed and coordination, attention and concentration, visual-motor function, memory across verbal and visual modalities, receptive and expressive speech, higher-order linguistic operations, problem-solving, abstraction ability, and general intelligence. In addition, there should be a clinical interview geared toward evaluating pathological features known to occur frequently in neurological disease and trauma, e.g., emotional lability, abnormality of mood, impaired impulse control, passivity and apathy, or inappropriate social behavior. The specialist performing the examination may administer one of the commercially available comprehensive neuropsychological batteries, such as the Luria-Nebraska or the Halstead-Reitan, or a battery of tests selected as relevant to the suspected brain dysfunction. The specialist performing the examination must be properly trained in this area of neuroscience.

9. *Screening tests.* In conjunction with clinical examinations, sources may report the results of screening tests; *i.e.,* tests used for gross determination of level of functioning. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

10. *Traumatic brain injury (TBI).* In cases involving TBI, follow the documentation and evaluation guidelines in 11.00F.

11. *Anxiety disorders.* In cases involving agoraphobia and other phobic disorders, panic disorders, and posttraumatic stress disorders, documentation of the anxiety reaction is essential. At least one detailed description of your typical reaction is required. The description should include the nature, frequency, and duration of any panic attacks or other reactions, the precipitating and exacerbating factors, and the functional effects. If the description is provided by a medical source, the reporting physician or psychologist should indicate the extent to which the description reflects his or her own observations and the source of any ancillary information. Statements of other persons who have observed you may be used for this description if professional observation is not available.

12. *Eating disorders.* In cases involving anorexia nervosa and other eating disorders, the primary manifestations may be mental or physical, depending upon the nature and extent of the disorder. When the primary functional limitation is physical, e.g., when severe weight loss and associated clinical findings are the chief cause of inability to work, we may evaluate the impairment under the appropriate physical body system listing. Of course, we must also consider any mental aspects of the impairment, unless we can make a fully favorable determination or decision based on the physical impairment(s) alone.

E. *Chronic mental impairments.* Particular problems are often involved in evaluating mental impairments in individuals who have long histories of repeated hospitalizations or prolonged outpatient care with supportive therapy and medication. For instance, if you have chronic organic, psychotic, and affective disorders, you may commonly have your life structured in such a way as to minimize your stress and reduce your symptoms and signs. In such a case, you may be much more impaired for work than your symptoms and signs would indicate. The results of a single examination may not adequately describe your sustained ability to function. It is, therefore, vital that we review all pertinent information relative to your condition, especially at times of increased stress. We will attempt to obtain adequate descriptive information from all sources that have treated you in the time period relevant to the determination or decision.

F. *Effects of structured settings.* Particularly in cases involving chronic mental disorders, overt symptomatology may be controlled or attenuated by psychosocial factors such as placement in a hospital, halfway house, board and care facility, or other environment that provides similar structure. Highly structured and supportive settings may also be found in your home. Such settings may greatly reduce the mental demands placed on you. With lowered mental demands, overt symptoms and signs of the underlying mental disorder may be minimized. At the same time, however, your ability to function outside of such a structured or supportive setting may not have changed. If your symptomatology is controlled or attenuated by psychosocial factors, we must consider your ability to function outside of such highly structured settings. For these reasons, identical paragraph C criteria are included in 12.02, 12.03, and 12.04. The paragraph C criterion of 12.06 reflects the uniqueness of agoraphobia, an anxiety disorder manifested by an overwhelming fear of leaving the home.

G. *Effects of medication.* We must give attention to the effects of medication on your symptoms, signs, and ability to function. While drugs used to modify psychological functions and mental states may control certain primary manifestations of a mental disorder, e.g., hallucinations, impaired attention, restlessness, or hyperactivity, such treatment may not affect all functional limitations imposed by the mental disorder. In cases where overt symptomatology is attenuated by the use of such drugs, particular attention must be focused on the functional limitations that may persist. We will consider these functional limitations in assessing the severity of your impairment. See the paragraph C criteria in 12.02, 12.03, 12.04, and 12.06.

Drugs used in the treatment of some mental illnesses may cause drowsiness, blunted effect, or other side effects involving other body systems. We will consider such side effects when we evaluate the overall severity of your impairment. Where adverse effects of medications contribute to the impairment severity and the impairment(s) neither meets nor is equivalent in severity to any listing but is nonetheless severe, we will consider such adverse effects in the RFC assessment.

H. *Effects of treatment.* With adequate treatment some individuals with chronic mental disorders not only have their symptoms and signs ameliorated, but they also return to a level of function close to the level of function they had before they developed symptoms or signs of their mental disorders. Treatment may or may not assist in the achievement of a level of adaptation adequate to perform sustained SGA. See the paragraph C criteria in 12.02, 12.03, 12.04, and 12.06.

I. *Technique for reviewing evidence in mental disorders claims to determine the level of impairment severity.* We have developed a special technique to ensure that we obtain, consider, and properly evaluate all the evidence we need to evaluate impairment severity in claims involving mental impairment(s). We explain this technique in §§404.1520a and 416.920a.

12.01 Category of Impairments—Mental

12.02 *Organic Mental Disorders:* Psychological or behaviorial abnormalities associated with a dysfunction of the brain. History and physical examination or laboratory tests demonstrate the presence of a specific organic factor judged to be etiologically related to the abnormal mental state and loss of previously acquired functional abilities.

The required level of severity for these disorders is met when the requirements in both A and B

are satisfied, or when the requirements in C are satisfied.

A. Demonstration of a loss of specific cognitive abilities or affective changes and the medically documented persistence of at least one of the following:

1. Disorientation to time and place; or

2. Memory impairment, either short-term (inability to learn new information), intermediate, or long-term (inability to remember information that was known sometime in the past); or

3. Perceptual or thinking disturbances (e.g., hallucinations, delusions); or

4. Change in personality; or

5. Disturbance in mood; or

6. Emotional lability (e.g., explosive temper outbursts, sudden crying, etc.) and impairment in impulse control; or

7. Loss of measured intellectual ability of at least 15 I.Q. points from premorbid levels or overall impairment index clearly within the severely impaired range on neuropsychological testing, e.g., the Luria-Nebraska, Halstead-Reitan, etc.;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic organic mental disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:

1. Repeated episodes of decompensation, each of extended duration; or

2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or

3. Current history of 1 or more years' inability to function outside a highly supportive living

arrangement, with an indication of continued need for such an arrangement.

12.03 *Schizophrenic, Paranoid and Other Psychotic Disorders:* Characterized by the onset of psychotic features with deterioration from a previous level of functioning.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in C are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one or more of the following:

1. Delusions or hallucinations; or

2. Catatonic or other grossly disorganized behavior; or

3. Incoherence, loosening of associations, illogical thinking, or poverty of content of speech if associated with one of the following:

a. Blunt affect; or

b. Flat affect; or

c. Inappropriate affect;

or

4. Emotional withdrawal and/or isolation;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic schizophrenic, paranoid, or other psychotic disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:

1. Repeated episodes of decompensation, each of extended duration; or

2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or

3. Current history of 1 or more years' inability to function outside a highly supportive living arrangement, with an indication of continued need for such an arrangement.

12.04 *Affective Disorders:* Characterized by a disturbance of mood, accompanied by a full or partial manic or depressive syndrome. Mood refers to a prolonged emotion that colors the whole psychic life; it generally involves either depression or elation.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in C are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one of the following:

1. Depressive syndrome characterized by at least four of the following:

a. Anhedonia or pervasive loss of interest in almost all activites; or

b. Appetite disturbance with change in weight; or

c. Sleep disturbance; or

d. Psychomotor agitation or retardation; or

e. Decreased energy; or

f. Feelings of guilt or worthlessness; or

g. Difficulty concentrating or thinking; or

h. Thoughts of suicide; or

i. Hallucinations, delusions, or paranoid thinking; or

2. Manic syndrome characterized by at least three of the following:

a. Hyperactivity; or

b. Pressure of speech; or

c. Flight of ideas; or

d. Inflated self-esteem; or

e. Decreased need for sleep; or

f. Easy distractability; or

g. Involvement in activities that have a high probability of painful consequences which are not recognized; or

h. Hallucinations, delusions or paranoid thinking;

or

3. Bipolar syndrome with a history of episodic periods manifested by the full symptomatic picture of both manic and depressive syndromes (and currently characterized by either or both syndromes);

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration;

OR

C. Medically documented history of a chronic affective disorder of at least 2 years' duration that has caused more than a minimal limitation of ability to do basic work activities, with symptoms or signs currently attenuated by medication or psychosocial support, and one of the following:

1. Repeated episodes of decompensation, each of extended duration; or

2. A residual disease process that has resulted in such marginal adjustment that even a minimal increase in mental demands or change in the environment would be predicted to cause the individual to decompensate; or

3. Current history of 1 or more years' inability to function outside a highly supportive living arrangement, with an indication of continued need for such an arrangement.

12.05 *Mental retardation:* Mental retardation refers to significantly subaverage general intellectual functioning with deficits in adaptive functioning initially manifested during the developmental period; *i.e.*, the evidence demonstrates or supports onset of the impairment before age 22.

The required level of severity for this disorder is met when the requirements in A, B, C, or D are satisfied.

A. Mental incapacity evidenced by dependence upon others for personal needs (e.g., toileting, eating, dressing, or bathing) and inability to follow directions, such that the use of standardized

measures of intellectual functioning is precluded;

OR

B. A valid verbal, performance, or full scale IQ of 59 or less;

OR

C. A valid verbal, performance, or full scale IQ of 60 through 70 and a physical or other mental impairment imposing an additional and significant work-related limitation of function;

OR

D. A valid verbal, performance, or full scale IQ of 60 through 70, resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

12.06 *Anxiety Related Disorders:* In these disorders anxiety is either the predominant disturbance or it is experienced if the individual attempts to master symptoms; for example, confronting the dreaded object or situation in a phobic disorder or resisting the obsessions or compulsions in obsessive compulsive disorders.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied, or when the requirements in both A and C are satisfied.

A. Medically documented findings of at least one of the following:

1. Generalized persistent anxiety accompanied by three out of four of the following signs or symptoms:

a. Motor tension; or

b. Autonomic hyperactivity; or

c. Apprehensive expectation; or

d. Vigilance and scanning;

or

2. A persistent irrational fear of a specific object, activity, or situation which results in a compelling desire to avoid the dreaded object, activity, or situation; or

3. Recurrent severe panic attacks manifested by a sudden unpredictable onset of intense apprehension, fear, terror and sense of impending doom occurring on the average of at least once a week; or

4. Recurrent obsessions or compulsions which are a source of marked distress; or

5. Recurrent and intrusive recollections of a traumatic experience, which are a source of marked distress;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

OR

C. Resulting in complete inability to function independently outside the area of one's home.

12.07 *Somatoform Disorders:* Physical symptoms for which there are no demonstrable organic findings or known physiological mechanisms.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented by evidence of one of the following:

1. A history of multiple physical symptoms of several years duration, beginning before age 30, that have caused the individual to take medicine frequently, see a physician often and alter life patterns significantly; or

2. Persistent nonorganic disturbance of one of the following:

a. Vision; or

b. Speech; or

c. Hearing; or

d. Use of a limb; or

e. Movement and its control (e.g., coordination disturbance, psychogenic seizures, akinesia, dyskinesia; or

f. Sensation (e.g., diminished or heightened).

3. Unrealistic interpretation of physical signs or sensations associated with the preoccupation or belief that one has a serious disease or injury;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

12.08 *Personality Disorders:* A personality disorder exists when personality traits are inflexible and maladaptive and cause either significant impairment in social or occupational functioning or subjective distress. Characteristic features are typical of the individual's long-term functioning and are not limited to discrete episodes of illness.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Deeply ingrained, maladaptive patterns of behavior associated with one of the following:

1. Seclusiveness or autistic thinking; or

2. Pathologically inappropriate suspiciousness or hostility; or

3. Oddities of thought, perception, speech and behavior; or

4. Persistent disturbances of mood or affect; or

5. Pathological dependence, passivity, or aggressivity; or

6. Intense and unstable interpersonal relationships and impulsive and damaging behavior;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

12.09 *Substance Addiction Disorders:* Behavioral changes or physical changes associated with the regular use of substances that affect the central nervous system.

The required level of severity for these disorders is met when the requirements in any of the following (A through I) are satisfied.

A. Organic mental disorders. Evaluate under 12.02.

B. Depressive syndrome. Evaluate under 12.04.

C. Anxiety disorders. Evaluate under 12.06.

D. Personality disorders. Evaluate under 12.08.

E. Peripheral neuropathies. Evaluate under 11.14.

F. Liver damage. Evaluate under 5.05.

G. Gastritis. Evaluate under 5.04.

H. Pancreatitis. Evaluate under 5.08.

I. Seizures. Evaluate under 11.02 or 11.03.

12.10 *Autistic disorder and other pervasive developmental disorders:* Characterized by qualitative deficits in the development of reciprocal social interaction, in the development of verbal and nonverbal communication skills, and in imaginative activity. Often, there is a markedly restricted repertoire of activities and interests, which frequently are stereotyped and repetitive.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of the following:

1. For autistic disorder, all of the following:

a. Qualitative deficits in reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity; and

c. Markedly restricted repertoire of activities and interests;

OR

2. For other pervasive developmental disorders, both of the following:

a. Qualitative deficits in reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity;

AND

B. Resulting in at least two of the following:

1. Marked restriction of activities of daily living; or

2. Marked difficulties in maintaining social functioning; or

3. Marked difficulties in maintaining concentration, persistence, or pace; or

4. Repeated episodes of decompensation, each of extended duration.

13.00 Malignant Neoplastic Diseases

A. *What impairments do these listings cover?* We use these listings to evaluate all malignant neoplasms except certain neoplasms associated with human immunodeficiency virus (HIV) infection. We use the criteria in 14.08E to evaluate carcinoma of the cervix, Kaposi's sarcoma, lymphoma, and squamous cell carcinoma of the anus if you also have HIV infection.

B. *What do we consider when we evaluate malignant neoplastic diseases under these listings?* We consider factors such as the:

1. Origin of the malignancy.

2. Extent of involvement.

3. Duration, frequency, and response to antineoplastic therapy. Antineoplastic therapy means surgery, irradiation, chemotherapy, hormones, immunotherapy, or bone marrow or stem cell transplantation. When we refer to surgery as an antineoplastic treatment, we mean surgical excision for treatment, not for diagnostic purposes.

4. Effects of any post-therapeutic residuals.

C. *How do we apply these listings?* We apply the criteria in a specific listing to a malignancy originating from that specific site.

D. *What evidence do we need?*

1. We need medical evidence that specifies the type, extent, and site of the primary, recurrent, or metastatic lesion. When the primary site cannot be identified, we will use evidence documenting the site(s) of metastasis to evaluate the impairment under 13.27.

2. For operative procedures, including a biopsy or a needle aspiration, we generally need a copy of both the:

a. Operative note.

b. Pathology report.

3. When we cannot get these documents, we will accept the summary of hospitalization(s) or other medical reports. This evidence should include details of the findings at surgery and, whenever appropriate, the pathological findings.

4. In some situations we may also need evidence about recurrence, persistence, or progression of the malignancy, the response to therapy, and any significant residuals. (*See* 13.00G.)

E. *When do we need longitudinal evidence?*

1. *Tumors with distant metastases.* We generally do not need longitudinal evidence for tumors that have metastasized beyond the regional lymph nodes because these tumors usually meet the requirements of a listing. Exceptions are for tumors with distant metastases that are expected to respond to antineoplastic therapy. For these exceptions, we usually need a longitudinal record of 3 months after therapy starts to determine whether the intended effect of therapy has been achieved and is likely to persist.

2. *Other malignancies.* When there are no distant metastases, many of the listings require that we consider your response to initial antineoplastic therapy; that is, the initial planned treatment regimen. This therapy may consist of a single modality or a combination of modalities (multimodal) given in close proximity as a unified whole, and is usually planned before any treatment(s) is initiated. Examples of multimodal therapy include:

a. Surgery followed by chemotherapy or radiation.

b. Chemotherapy followed by surgery.

c. Chemotherapy and concurrent radiation.

3. *Types of treatment.* Whenever the initial planned therapy is a single modality, enough time must pass to allow a determination about whether the therapy will achieve its intended effect. If the treatment fails, the failure will often happen within 6 months after the treatment starts, and there will often be a change in the treatment regimen. Whenever the initial planned therapy is multimodal, a determination about the effectiveness of the therapy usually cannot be made until the effects of all the planned modalities can be determined. In some cases, we may need to defer adjudication until the effectiveness of therapy can be assessed. However, we do not need to defer adjudication to determine whether the therapy will achieve its intended effect if we can make a fully favorable determination or decision based on the length and effects of therapy, or the residuals of the malignancy or therapy (*see* 13.00G).

F. *How do we evaluate impairments that do not meet one of the malignant neoplastic diseases listings?*

1. These listings are only examples of malignant neoplastic diseases that we consider severe enough to prevent you from doing any gainful activity. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether you have an

impairment(s) that meets the criteria of a listing in another body system.

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

G. *How do we consider the effects of therapy?*

1. *How we consider the effects of therapy under the listings.* In many cases, malignancies meet listing criteria only if the therapy does not achieve the intended effect: the malignancy persists, progresses, or recurs despite treatment. However, as explained in the following paragraphs, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in the case record.

2. *Effects can vary widely.*

a. Because the therapy and its toxicity may vary widely, we consider each case on an individual basis. We will request a specific description of the therapy, including these items:

i. Drugs given.

ii. Dosage.

iii. Frequency of drug administration.

iv. Plans for continued drug administration.

v. Extent of surgery.

vi. Schedule and fields of radiation therapy.

b. We will also request a description of the complications or adverse effects of therapy, such as the following:

i. Continuing gastrointestinal symptoms.

ii. Persistent weakness.

iii. Neurological complications.

iv. Cardiovascular complications.

v. Reactive mental disorders.

3. *Effects of therapy may change.* Because the severity of the adverse effects of antineoplastic

therapy may change during treatment, enough time must pass to allow us to evaluate the therapy's effect. The residual effects of treatment are temporary in most instances. But on occasion, the effects may be disabling for a consecutive period of at least 12 months.

4. *When the initial antineoplastic therapy is effective.* We evaluate any post-therapeutic residual impairment(s) not included in these listings under the criteria for the affected body system. We must consider any complications of therapy. When the residual impairment(s) does not meet or medically equal a listing, we must consider its effect on your ability to do substantial gainful activity.

H. *How long do we consider your impairment to be disabling?*

1. In some listings, we specify that we will consider your impairment to be disabling until a particular point in time (for example, at least 18 months from the date of diagnosis). We may consider your impairment to be disabling beyond this point when the medical and other evidence justifies it.

2. When a listing does not contain such a specification, we will consider an impairment(s) that meets or medically equals a listing in this body system to be disabling until at least 3 years after onset of complete remission. When the impairment(s) has been in complete remission for at least 3 years, that is, the original tumor and any metastases have not been evident for at least 3 years, the impairment(s) will no longer meet or medically equal the criteria of a listing in this body system.

3. Following the appropriate period, we will consider any residuals, including residuals of the malignancy or therapy (see 13.00G), in determining whether you are disabled.

I. *What do these terms in the listings mean?*

1. *Inoperable:* Surgery is thought to be of no therapeutic value or the surgery cannot be performed. Examples of when surgery cannot be performed include a tumor that is too large or that invades crucial structures, or an intolerance of anesthesia or surgery due to other medical conditions. This term does not include situations in which the tumor could have been surgically removed but another method of treatment was chosen; for example, an attempt at organ preservation. The determination whether a tumor is inoperable usually occurs before attempts to shrink the tumor with chemotherapy or radiation.

2. *Unresectable:* The operation was performed, but the malignant tumor was not removed. This term includes situations in which a tumor is incompletely resected or the surgical margins are positive.

3. *Persistent:* Failure to achieve a complete remission.

4. *Progressive:* The malignancy became more extensive after treatment.

5. *Recurrent, relapse:* A malignancy that had been in complete remission or entirely removed by surgery has returned.

J. *Can we establish the existence of a disabling impairment prior to the date of the evidence that shows the malignancy satisfies the criteria of a listing?* Yes. We will consider factors such

as:

1. The type of malignancy and its location.

2. The extent of involvement when the malignancy was first demonstrated.

3. Your symptoms.

K. *How do we evaluate specific malignant neoplastic diseases?*

1. *Lymphoma.*

a. Many low grade or indolent (non-aggressive) lymphomas are controlled by well-tolerated treatment modalities, although they may produce intermittent symptoms and signs. Therefore, we may defer adjudication of these cases for an appropriate period after initiation of therapy to determine whether the therapy will achieve its intended effect. (*See* 13.00E3.) For a low grade or indolent lymphoma, the intended effect of therapy is usually stability of the disease process. When stability has been achieved, we will assess severity on the basis of the extent of involvement of other organ systems and residuals from therapy.

b. A change in therapy for low grade or indolent lymphomas is usually an indicator that the therapy is not achieving its intended effect. However, it does not indicate this if the change is based on your (or your physician's) choice rather than a failure to achieve stability. If the therapy is changed due solely to choice, the requirements of listing 13.05A2 are not met.

c. We consider Hodgkin's disease that recurs more than 12 months after completing initial antineoplastic therapy to be a new disease rather than a recurrence.

2. *Leukemia.*

a. *Acute leukemia.* The initial diagnosis of acute leukemia, including the accelerated or blast phase of chronic myelogenous (granulocytic) leukemia, is based upon definitive bone marrow examination. Additional diagnostic information is based on chromosomal analysis, cytochemical and surface marker studies on the abnormal cells, or other methods consistent with the prevailing state of medical knowledge and clinical practice. Recurrent disease must be documented by peripheral blood, bone marrow, or cerebrospinal fluid examination. The initial and follow-up pathology reports should be included.

b. *Chronic myelogenous leukemia (CML).* The diagnosis of CML should be based upon documented granulocytosis, including immature forms such as differentiated or undifferentiated myelocytes and myeloblasts, and a chromosomal analysis that demonstrates the Philadelphia chromosome. In the absence of a chromosomal analysis, or if the Philadelphia chromosome is not present, the diagnosis may be made by other methods consistent with the prevailing state of medical knowledge and clinical practice.

c. *Chronic lymphocytic leukemia.*

i. The diagnosis of chronic lymphocytic leukemia (CLL) must be documented by evidence of a chronic lymphocytosis of at least 10,000/mm[3] for 3 months or longer, or other acceptable

diagnostic techniques consistent with the prevailing state of medical knowledge and clinical practice.

ii. We evaluate the complications and residual impairment(s) from CLL under the appropriate listings, such as 13.05A2, 7.02, and 7.15.

d. *Elevated white cell count.* In cases of chronic leukemia (either myelogenous or lymphocytic), an elevated white cell count, in itself, is not ordinarily a factor in determining the severity of the impairment.

3. *Macroglobulinemia or heavy chain disease.* The diagnosis of these diseases must be confirmed by protein electrophoresis or immunoelectrophoresis. We evaluate the resulting impairment(s) under the criteria of 7.02, 7.06, 7.08, or any other affected body system.

4. *Bilateral primary breast cancer.* We evaluate bilateral primary breast cancer (synchronous or metachronous) under 13.10A, which covers local primary disease, and not as a primary disease that has metastasized.

5. *Carcinoma-in-situ.* Carcinoma-in-situ, or preinvasive carcinoma, usually responds to treatment. When we use the term "carcinoma" in these listings, it does not include carcinoma-in-situ.

6. *Brain tumors.* We use the criteria in 13.13 to evaluate malignant brain tumors. We will evaluate any complications of malignant brain tumors, such as resultant neurological or psychological impairments, under the criteria for the affected body system. We evaluate benign brain tumors under 11.05.

L. *How do we evaluate malignant neoplastic diseases treated by bone marrow or stem cell transplantation?* Bone marrow or stem cell transplantation is performed for a variety of malignant neoplastic diseases.

1. *Acute leukemia (including T-cell lymphoblastic lymphoma) or accelerated or blast phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of transplantation, whichever is later.

2. *Lymphoma, multiple myeloma, or chronic phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 12 months from the date of transplantation.

3. *Other malignancies.* We will evaluate any other malignant neoplastic disease treated with bone marrow or stem cell transplantation under 13.28, regardless of whether there is another listing that addresses that impairment. The length of time we will consider you to be disabled depends on whether you undergo allogeneic or autologous transplantation.

a. *Allogeneic bone marrow or stem cell transplantation.* If you undergo allogeneic transplantation (transplantation from an unrelated donor or a related donor other than an identical twin), we will consider you to be disabled until at least 12 months from the date of transplantation.

b. *Autologous bone marrow or stem cell transplantation.* If you undergo autologous transplantation (transplantation of your own cells or cells from your identical twin (syngeneic transplantation)), we will consider you to be disabled until at least 12 months from the date of the first treatment under the treatment plan that includes transplantation. The first treatment usually refers to the initial therapy given to prepare you for transplantation.

4. *Evaluating disability after the appropriate time period has elapsed.* We consider any residual impairment(s), such as complications arising from:

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

b. Immunosuppressant therapy, such as frequent infections.

c. Significant deterioration of other organ systems.

13.01 Category of Impairments, Malignant Neoplastic Diseases

13.02 *Soft tissue tumors of the head and neck (except salivary glands—13.08—and thyroid gland—13.09).*

A. Inoperable or unresectable.

OR

B. Persistent disease following initial multimodal antineoplastic therapy.

OR

C. Recurrent disease following initial antineoplastic therapy, except local vocal cord recurrence.

OR

D. With metastases beyond the regional lymph nodes.

OR

E. Soft tissue tumors of the head and neck not addressed in A-D, with multimodal antineoplastic therapy. Consider under a disability until at least 18 months from the date of diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.03 *Skin.*

A. Sarcoma or carcinoma with metastases to or beyond the regional lymph nodes.

OR

B. Melanoma, with either 1 or 2:

1. Recurrent after wide excision (except an additional primary melanoma at a different site, which is not considered to be recurrent disease).

2. Palpable nodal metastases or metastases to adjacent skin (satellite lesions) or elsewhere.

13.04 *Soft tissue sarcoma.*

A. With regional or distant metastases.

OR

B. Persistent or recurrent following initial antineoplastic therapy.

13.05 *Lymphoma (including mycosis fungoides, but excluding T-cell lymphoblastic lymphoma—13.06).* (*See* 13.00K1 and 13.00K2c.)

A. Non-Hodgkin's lymphoma, as described in 1 or 2:

1. Intermediate or high-grade lymphoma persistent or recurrent following initial antineoplastic therapy.

2. Low-grade or indolent lymphoma requiring initiation of more than one antineoplastic treatment regimen within a consecutive 12-month period. Consider under a disability from at least the date of initiation of the treatment regimen that failed within 12 months.

OR

B. Hodgkin's disease with failure to achieve clinically complete remission, or recurrent disease within 12 months of completing initial antineoplastic therapy.

OR

C. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.06 *Leukemia.* (*See* 13.00K2.)

A. Acute leukemia (including T-cell lymphoblastic lymphoma). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Chronic myelogenous leukemia, as described in 1 or 2:

1. Accelerated or blast phase. Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell

transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

2. Chronic phase, as described in a or b:

a. Consider under a disability until at least 12 months from the date of bone marrow or stem cell transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

b. Progressive disease following initial antineoplastic therapy.

13.07 *Multiple myeloma (confirmed by appropriate serum or urine protein electrophoresis and bone marrow findings).*

A. Failure to respond or progressive disease following initial antineoplastic therapy.

OR

B. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.08 *Salivary glands*—carcinoma or sarcoma with metastases beyond the regional lymph nodes.

13.09 *Thyroid gland.*

A. Anaplastic (undifferentiated) carcinoma.

OR

B. Carcinoma with metastases beyond the regional lymph nodes progressive despite radioactive iodine therapy.

13.10 *Breast (except sarcoma—13.04).* (*See* 13.00K4.)

A. Locally advanced carcinoma (inflammatory carcinoma, tumor of any size with direct extension to the chest wall or skin, tumor of any size with metastases to the ipsilateral internal mammary nodes).

OR

B. Carcinoma with distant metastases.

OR

C. Recurrent carcinoma, except local recurrence that remits with antineoplastic therapy.

13.11 *Skeletal system*—carcinoma or sarcoma.

A. Inoperable or unresectable.

OR

B. Recurrent tumor (except local recurrence) after initial antineoplastic therapy.

OR

C. With distant metastases.

OR

D. All other tumors originating in bone with multimodal antineoplastic therapy. Consider under a disability for 12 months from the date of diagnosis. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

13.12 *Maxilla, orbit, or temporal fossa.*

A. Sarcoma or carcinoma of any type with regional or distant metastases.

OR

B. Carcinoma of the antrum with extension into the orbit or ethmoid or sphenoid sinus.

OR

C. Tumors with extension to the base of the skull, orbit, meninges, or sinuses.

13.13 *Nervous system.* (*See* 13.00K6.)

A. Central nervous system neoplasms (brain and spinal cord), as described in 1 or 2:

1. Highly malignant tumors, such as Grades III and IV astrocytomas, glioblastoma multiforme, ependymoblastoma, medulloblastoma or other primitive neuroectodermal tumors (PNETs) with documented metastases, diffuse intrinsic brain stem gliomas, or primary sarcomas.

2. Any central nervous system neoplasm progressive or recurrent following initial antineoplastic therapy.

OR

B. Peripheral nerve or spinal root neoplasm, as described in 1 or 2:

1. Metastatic.

2. Progressive or recurrent following initial antineoplastic therapy.

13.14 *Lungs*.

A. Non-small-cell carcinoma—inoperable, unresectable, recurrent, or metastatic disease to or beyond the hilar nodes.

OR

B. Small-cell (oat cell) carcinoma.

13.15 *Pleura or mediastinum*.

A. Malignant mesothelioma of pleura.

OR

B. Tumors of the mediastinum, as described in 1 or 2:

1. With metastases to or beyond the regional lymph nodes.

2. Persistent or recurrent following initial antineoplastic therapy.

13.16 *Esophagus or stomach*.

A. Carcinoma or sarcoma of the esophagus.

OR

B. Carcinoma or sarcoma of the stomach, as described in 1 or 2:

1. Inoperable, unresectable, extending to surrounding structures, or recurrent.

2. With metastases to or beyond the regional lymph nodes.

13.17 *Small intestine*—carcinoma, sarcoma, or carcinoid.

A. Inoperable, unresectable, or recurrent.

OR

B. With metastases beyond the regional lymph nodes.

13.18 *Large intestine (from ileocecal valve to and including anal canal)*.

A. Adenocarcinoma that is inoperable, unresectable, or recurrent.

OR

B. Squamous cell carcinoma of the anus, recurrent after surgery.

OR

C. With metastases beyond the regional lymph nodes.

13.19 *Liver or gallbladder*—tumors of the liver, gallbladder, or bile ducts.

13.20 *Pancreas.*

A. Carcinoma (except islet cell carcinoma).

OR

B. Islet cell carcinoma that is inoperable or unresectable and physiologically active.

13.21 *Kidneys, adrenal glands, or ureters*—carcinoma.

A. Inoperable, unresectable, or recurrent.

OR

B. With metastases to or beyond the regional lymph nodes.

13.22 *Urinary bladder*—carcinoma.

A. With infiltration beyond the bladder wall.

OR

B. Recurrent after total cystectomy.

OR

C. Inoperable or unresectable.

OR

D. With metastases to or beyond the regional lymph nodes.

13.23 *Cancers of the female genital tract*—carcinoma or sarcoma.

A. Uterus (corpus), as described in 1, 2, or 3:

1. Invading adjoining organs.

2. With metastases to or beyond the regional lymph nodes.

3. Persistent or recurrent following initial antineoplastic therapy.

OR

B. Uterine cervix, as described in 1 or 2:

1. Extending to the pelvic wall, lower portion of the vagina, or adjacent or distant organs.

2. Persistent or recurrent following initial antineoplastic therapy.

OR

C. Vulva, as described in 1, 2, or 3:

1. Invading adjoining organs.

2. With metastases to or beyond the regional lymph nodes.

3. Persistent or recurrent following initial antineoplastic therapy.

OR

D. Fallopian tubes, as described in 1 or 2:

1. Extending to the serosa or beyond.

2. Persistent or recurrent following initial antineoplastic therapy.

OR

E. Ovaries, as described in 1 or 2:

1. All tumors except germ-cell tumors, with at least one of the following:

a. Tumor extension beyond the pelvis; for example, tumor implants on peritoneal, omental, or bowel surfaces.

b. Metastases to or beyond the regional lymph nodes.

c. Ruptured ovarian capsule, tumor on the serosal surface of the ovary, ascites with malignant cells, or positive peritoneal washings.

d. Recurrent following initial antineoplastic therapy.

2. Germ-cell tumors—progressive or recurrent following initial antineoplastic therapy.

13.24 *Prostate gland*—carcinoma.

A. Progressive or recurrent despite initial hormonal intervention.

OR

B. With visceral metastases.

13.25 *Testicles*—tumor with metastatic disease progressive or recurrent following initial

chemotherapy.

13.26 *Penis*—carcinoma with metastases to or beyond the regional lymph nodes.

13.27 *Primary site unknown after appropriate search for primary*—metastatic carcinoma or sarcoma, except for solitary squamous cell carcinoma in the neck.

13.28 *Malignant neoplastic diseases treated by bone marrow or stem cell transplantation.* (*See* 13.00L.)

A. Allogeneic transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Autologous transplantation. Consider under a disability until at least 12 months from the date of the first treatment under the treatment plan that includes transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

14.00 Immune System

A. Listed disorders include impairments involving deficiency of one or more components of the immune system (*i.e.*, antibody-producing B cells; a number of different types of cells associated with cell-mediated immunity including T-lymphocytes, macrophages and monocytes; and components of the complement system).

B. Dysregulation of the immune system may result in the development of a connective tissue disorder. Connective tissue disorders include several chronic multisystem disorders that differ in their clinical manifestation, course, and outcome. They generally evolve and persist for months or years, may result in loss of functional abilities, and may require long-term, repeated evaluation and management.

The documentation needed to establish the existence of a connective tissue disorder is medical history, physical examination, selected laboratory studies, appropriate medically acceptable imaging, and, in some instances, tissue biopsy. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment. However, the Social Security Administration will not purchase diagnostic tests or procedures that may involve significant risk, such as biopsies or angiograms. Generally, the existing medical evidence will contain this information.

A longitudinal clinical record of at least 3 months demonstrating active disease despite prescribed treatment during this period with the expectation that the disease will remain active for 12 months is necessary for assessment of severity and duration of impairment.

To permit appropriate application of a listing, the specific diagnostic features that should be documented in the clinical record for each of the disorders are summarized for systemic lupus

erythematosus (SLE), systemic vasculitis, systemic sclerosis and scleroderma, polymyositis or dermatomyositis, undifferentiated connective tissue disorders, and the inflammatory arthritides.

In addition to the limitations caused by the connective tissue disorder *per se,* the chronic adverse effects of treatment (e.g., corticosteroid-related ischemic necrosis of bone) may result in functional loss.

These disorders may preclude performance of any gainful activity by reason of serious loss of function because of disease affecting a single organ or body system, or lesser degrees of functional loss because of disease affecting two or more organs/body systems associated with significant constitutional symptoms and signs of severe fatigue, fever, malaise, weight loss, and joint pain and stiffness. We use the term "severe" in these listings to describe medical severity; the term does not have the same meaning as it does when we use it in connection with a finding at the second step of the sequential evaluation processes in §§404.1520, 416.920, and 416.924.

1. Systemic lupus erythematosus (14.02)—This disease is characterized clinically by constitutional symptoms and signs (e.g., fever, fatigability, malaise, weight loss), multisystem involvement and, frequently, anemia, leukopenia, or thrombocytopenia. Immunologically, an array of circulating serum auto-antibodies can occur, but are highly variable in pattern. Generally the medical evidence will show that patients with this disease will fulfill The 1982 Revised Criteria for the Classification of Systemic Lupus Erythematosus of the American College of Rheumatology. (Tan, E.M., et al., Arthritis Rheum. 25: 11271-1277, 1982).

2. Systemic vasculitis (14.03)—This disease occurs acutely in association with adverse drug reactions, certain chronic infections and, occasionally, malignancies. More often it is idiopathic and chronic. There are several clinical patterns, including classical polyarteritis nodosa, aortic arch arteritis, giant cell arteritis, Wegener's granulomatosis, and vasculitis associated with other connective tissue disorders (e.g., rheumatoid arthritis, SLE, Sjögren's syndrome, cryoglobulinemia). Cutaneous vasculitis may or may not be associated with systemic involvement and the patterns of vascular and ischemic involvement are highly variable. The diagnosis is confirmed by angiography or tissue biopsy when the disease is suspected clinically. Most patients who are stated to have this disease will have the results of the confirmatory angiogram or biopsy in their medical records.

3. Systemic sclerosis and scleroderma (14.04)—These disorders constitute a spectrum of disease in which thickening of the skin is the clinical hallmark. Raynaud's phenomena, often severe and progressive, are especially frequent and may be the peripheral manifestation of a generalized vasospastic abnormality in the heart, lungs, and kidneys. The CREST syndrome (calcinosis, Raynaud's phenomena, esophageal dysmotility, sclerodactyly, telangiectasia) is a variant that may slowly progress to the generalized process, systemic sclerosis, over years. In addition to skin and blood vessels, the major organ/body system involvement includes the gastrointestinal tract, lungs, heart, kidneys, and muscle. Although arthritis can occur, joint dysfunction results primarily from soft tissue/cutaneous thickening, fibrosis, and contractures.

4. Polymyositis or dermatomyositis (14.05)—This disorder is primarily an inflammatory process in striated muscle, which can occur alone or in association with other connective tissue disorders or malignancy. Weakness and, less frequently, pain and tenderness of the proximal limb-girdle musculature are the cardinal manifestations. Involvement of the cervical muscles, the cricopharyngeals, the intercostals, and diaphragm may occur in those with listing-level

disease. Weakness of the pelvic girdle, as contemplated in Listing 14.05A, may 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 hair. Weakness of anterior neck flexors may impair the ability to lift the head from the pillow in bed. The diagnosis is supported by elevated serum muscle enzymes (creatine phosphokinase (CPK), aminotransferases, aldolase), characteristic abnormalities on electromyography, and myositis on muscle biopsy.

5. Undifferentiated connective tissue disorder (14.06)—This listing includes syndromes with clinical and immunologic features of several connective tissue disorders, but that do not satisfy the criteria for any of the disorders described; for instance, the individual may have clinical features of systemic lupus erythematosus and systemic vasculitis and the serologic findings of rheumatoid arthritis. It also includes overlap syndromes with clinical features of more than one established connective tissue disorder. For example, the individual may have features of both rheumatoid arthritis and scleroderma. The correct designation of this disorder is important for assessment of prognosis.

6. *Inflammatory arthritis (14.09)* includes a vast array of disorders that differ in cause, course, and outcome. For example, inflammatory spondyloarthropathies include ankylosing spondylitis, Reiter's syndrome and other reactive arthropathies, psoriatic arthropathy, Behçet's disease, and Whipple's disease, as well as undifferentiated spondylitis. Inflammatory arthritis of peripheral joints likewise comprises many disorders, including rheumatoid arthritis, Sjögren's syndrome, psoriatic arthritis, crystal deposition disorders, and Lyme disease. Clinically, inflammation of major joints may be the dominant problem causing difficulties with ambulation or fine and gross movements, or the arthritis may involve other joints or cause less restriction of ambulation or other movements but be complicated by extra-articular features that cumulatively result in serious functional deficit. When persistent deformity without ongoing inflammation is the dominant feature of the impairment, it should be evaluated under 1.02, or, if there has been surgical reconstruction, 1.03.

a. In 14.09A, the term *major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

b. The terms *inability to ambulate effectively and inability to perform fine and gross movements effectively* in 14.09A have the same meaning as in 1.00B2b and 1.00B2c and must have lasted, or be expected to last, for at least 12 months.

c. Inability to ambulate effectively is implicit in 14.09B. Even though individuals who demonstrate the findings of 14.09B will not ordinarily require bilateral upper limb assistance, the required ankylosis of the cervical or dorsolumbar spine will result in an extreme loss of the ability to see ahead, above, and to the side.

d. As in 14.02 through 14.06, extra-articular features of an inflammatory arthritis may satisfy the criteria for a listing in an involved extra-articular body system. Such impairments may be found to meet a criterion of 14.09C. Extra-articular impairments of lesser severity should be

evaluated under 14.09D and 14.09E. Commonly occurring extra-articular impairments include keratoconjunctivitis sicca, uveitis, iridocyclitis, pleuritis, pulmonary fibrosis or nodules, restrictive lung disease, pericarditis, myocarditis, cardiac arrhythmias, aortic valve insufficiency, coronary arteritis, Raynaud's phenomena, systemic vasculitis, amyloidosis of the kidney, chronic anemia, thrombocytopenia, hypersplenism with compromised immune competence (Felty's syndrome), peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss, and heel enthesopathy with functionally limiting pain.

e. The fact that an individual is dependent on steroids, or any other drug, for the control of inflammatory arthritis is, in and of itself, insufficient to find disability. Advances in the treatment of inflammatory connective tissue disease and in the administration of steroids for its treatment have corrected some of the previously disabling consequences of continuous steroid use. Therefore, each case must be evaluated on its own merits, taking into consideration the severity of the underlying impairment and any adverse effects of treatment.

C. Allergic disorders (e.g., asthma or atopic dermatitis) are discussed and evaluated under the appropriate listing of the affected body system.

D. Human immunodeficiency virus (HIV) infection.

1. HIV infection is caused by a specific retrovirus and may be characterized by susceptibility to one or more opportunistic diseases, cancers, or other conditions, as described in 14.08. Any individual 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 14.08 or is of equivalent severity to any impairment in 14.08.

2. Definitions. In 14.08, the terms "resistant to treatment," "recurrent," and "disseminated" 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 14.08I, "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 percent of baseline is always considered significant, loss of less than 10 percent may or may not be significant, depending on the individual's baseline weight and body habitus. (For example, a 7-pound weight loss in a 100-pound 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.)

3. 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.

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

i. 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 (e.g., Western blot, immunofluorescence assay).

ii. A specimen that contains HIV antigen (e.g., serum specimen, lymphocyte culture, or cerebrospinal fluid (CSF) specimen).

iii. Other test(s) that are highly specific for detection of HIV (e.g., 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, every reasonable effort must be made to obtain reports of the results of that testing.

Individuals who have HIV infection or other disorders of the immune system may undergo tests to determine T-helper lymphocyte (CD4) counts. 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 percent or less), the susceptibility to opportunistic disease is considerably increased. However, a reduced CD4 count alone does not establish a definitive diagnosis of HIV infection, or document the severity or functional effects of HIV infection.

b. Other acceptable documentation of HIV infection.

HIV infection may also be documented without the definitive laboratory evidence described in paragraph a, 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, 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 if the individual has an opportunistic disease (e.g., toxoplasmosis of the brain, pneumocystis carinii pneumonia (PCP)) predictive of a defect in cell-mediated immunity, and there is no other known cause of diminished resistance to that disease (e.g., long-term steroid treatment, lymphoma). In such cases, every reasonable effort must be made to obtain full details of the history, medical findings, and results of testing.

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

a. Documentation of the manifestations of HIV infection by definitive diagnosis.

The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection is by culture, serological test, or microscopic examination of biopsied tissue or other material (e.g., bronchial washings). Therefore, every reasonable effort must be made to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histological 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 radiographic studies) 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 (see 14.00D3a, above), that alone does not establish the presence, severity, or functional effects of a manifestation of HIV infection.

b. Other acceptable documentation of the manifestations of HIV infection.

Manifestations of HIV infection may also be documented without the definitive laboratory evidence described in paragraph a, 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 diagnosis(es) 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 (14.08D) 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.

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

Many of these manifestations (e.g. vulvovaginal candidiasis, pelvic inflammatory disease) occur in women with or without HIV infection, but can be more severe or resistant to treatment, or occur more frequently in a woman whose immune system is suppressed. Therefore, when evaluating the claim of a woman with HIV infection, it is important to consider gynecologic and other problems specific to women, including any associated symptoms (e.g., pelvic pain), in assessing the severity of the impairment and resulting functional limitations. Manifestations of HIV infection in women may be evaluated under the specific criteria (e.g., cervical cancer under 14.08E), under an applicable general category (e.g., pelvic inflammatory disease under

14.08A5) or, in appropriate cases, under 14.08N.

6. Evaluation. The criteria in 14.08 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 an individual's impairment(s) meets or equals in severity any other listing in appendix 1 of subpart P (e.g., a neoplastic disorder listed in 13.00ff). Although 14.08 includes cross-references to other listings for the more common manifestations of HIV infection, other listings may apply.

In addition, the impact of all impairments, 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 (e.g., anxiety, depression), or of another physical impairment. Medical evidence should include documentation of all physical and mental impairments, and the impairment(s) should be evaluated not only under the relevant listing(s) in 14.08, but under any other appropriate listing(s).

It is also important to remember that individuals with HIV infection, like all other individuals, are evaluated under the full five-step sequential evaluation process described in §404.1520 and §416.920. If an individual with HIV infection is working and engaging in substantial gainful activity (SGA), or does not have a severe impairment, the case will be decided at the first or second step of the sequential evaluation process, and does not require evaluation under these listings. For an individual with HIV infection who is not engaging in SGA and has a severe impairment, but whose impairment(s) does not meet or equal in severity the criteria of a listing, evaluation must proceed through the final steps of the sequential evaluation process (or, as appropriate, the steps in the medical improvement review standard) before any conclusion can be reached on the issue of disability.

7. Effect of treatment. Medical treatment must be considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the specific disorder, or of the HIV infection itself (e.g., antiretroviral agents) and in terms of any side effects of treatment that may further impair the individual.

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 individuals without HIV infection, but another individual 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 individual'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.

8. Functional criteria. Paragraph N of 14.08 establishes standards for evaluating manifestations of HIV infection that do not meet the requirements listed in 14.08A-M. Paragraph N is applicable for manifestations that are not listed in 14.08A-M, as well as those listed in 14.08A-M that do not meet the criteria of any of the rules in 14.08A-M.

For individuals with HIV infection evaluated under 14.08N, 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 individuals 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 exacerbation and remission in the disease course; and the functional impact of treatment for the disease, including the side effects of medication.

As used in 14.08N, "repeated" means that the conditions occur on an average of 3 times a year, or once every 4 months, each lasting 2 weeks or more; or the conditions do not last for 2 weeks but occur substantially more frequently than 3 times in a year or once every 4 months; or they occur less often than an average of 3 times a year or once every 4 months but last substantially longer than 2 weeks.

To meet the criteria in 14.08N, an individual with HIV infection must demonstrate a marked level of restriction in one of three general areas of functioning: activities of daily living; social functioning; and difficulties in completing tasks due to deficiencies in concentration, persistence, or pace. Functional restrictions may result from the impact of the disease process itself on mental or physical functioning, or both. This 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 individual'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, an individual need not be totally precluded from performing 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 individual 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. An individual 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 (even though he or she is able to perform some self-care activities) would have marked limitation of activities of daily living.

Social functioning includes the capacity to interact appropriately and communicate effectively with others. An individual with HIV infection who, because of symptoms or a pattern of exacerbation and remission caused by the illness or its treatment, cannot engage in social interaction on a sustained basis (even though he or she is able to communicate with close friends or relatives) would have marked difficulty maintaining social functioning.

Completing tasks in a timely manner involves the ability to sustain concentration, persistence, or pace to permit timely completion of tasks commonly found in work settings. 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.

14.01 Category of Impairments, Immune System

14.02 Systemic lupus erythematosus. Documented as described in 14.00B1, with:

A. One of the following:

1. Joint involvement, as described under the criteria in 1.00; or

2. Muscle involvement, as described under the criteria in 14.05; or

3. Ocular involvement, as described under the criteria in 2.00ff; or

4. Respiratory involvement, as described under the criteria in 3.00ff; or

5. Cardiovascular involvement, as described under the criteria in 4.00ff or 14.04D; or

6. Digestive involvement, as described under the criteria in 5.00ff; or

7. Renal involvement, as described under the criteria in 6.00ff; or

8. Hematologic involvement, as described under the criteria in 7.00ff; or

9. Skin involvement, as described under the criteria in 8.00ff; or

10. Neurological involvement, as described under the criteria in 11.00ff; or

11. Mental involvement, as described under the criteria in 12.00ff.

or

B. Lesser involvement of two or more organs/body systems listed in paragraph A, with significant, documented, constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss. At least one of the organs/body systems must be involved to at least a moderate level of severity.

14.03 Systemic vasculitis. Documented as described in 14.00B2, including documentation by angiography or tissue biopsy, with:

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

or

B. Lesser involvement of two or more organs/body systems listed in 14.02A, with significant, documented, constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss. At least one of the organs/body systems must be involved to at least a moderate level of severity.

14.04 Systemic sclerosis and scleroderma. Documented as described in 14.00B3, with:

A. One of the following:

1. Muscle involvement, as described under the criteria in 14.05; or

2. Respiratory involvement, as described under the criteria in 3.00ff; or

3. Cardiovascular involvement, as described under the criteria in 4.00ff; or

4. Digestive involvement, as described under the criteria in 5.00ff; or

5. Renal involvement, as described under the criteria in 6.00ff.

or

B. Lesser involvement of two or more organs/body systems listed in paragraph A, with significant, documented, constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss. At least one of the organs/body systems must be involved to at least a moderate level of severity.

or

C. Generalized scleroderma with digital contractures.

or

D. Severe Raynaud's phenomena, characterized by digital ulcerations, ischemia, or gangrene.

14.05 Polymyositis or dermatomyositis. Documented as described in 14.00B4, with:

A. Severe proximal limb-girdle (shoulder and/or pelvic) muscle weakness, as described in 14.00B4.

or

B. Less severe limb-girdle muscle weakness than in 14.05A, associated with cervical muscle weakness and one of the following to at least a moderate level of severity:

1. Impaired swallowing with dysphagia and episodes of aspiration due to cricopharyngeal weakness, or

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

or

C. If associated with malignant tumor, as described under the criteria in 13.00ff.

or

D. If associated with generalized connective tissue disease, described under the criteria in 14.02, 14.03, 14.04, or 14.06.

14.06 Undifferentiated connective tissue disorder. Documented as described in 14.00B5, and with impairment as described under the criteria in 14.02A, 14.02B, or 14.04.

14.07 Immunoglobulin deficiency syndromes or deficiencies of cell-mediated immunity, excepting HIV infection. Associated with documented, recurrent severe infection occurring 3 or more times within a 5-month period.

14.08 Human immunodeficiency virus (HIV) infection. With documentation as described in 14.00D3 and one of the following:

A. Bacterial infections:

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

2. Nocardiosis; or

3. Salmonella bacteremia, recurrent non-typhoid; or

4. Syphilis or neurosyphilis—evaluate sequelae under the criteria for the affected body system (e.g., 2.00 Special Senses and Speech, 4.00 Cardiovascular System, 11.00 Neurological); or

5. Multiple or recurrent bacterial infection(s), including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment 3 or more times in 1 year.

or

B. Fungal infections:

1. Aspergillosis; or

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

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

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

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

6. Mucormycosis.

or

C. Protozoan or helminthic infections:

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

2. Pneumocystis carinii pneumonia or extrapulmonary pneumocystis carinii infection; or

3. Strongyloidiasis, extra-intestinal; or

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

or

D. Viral infections:

1. Cytomegalovirus disease (documented as described in 14.00D4b) at a site other than the liver, spleen, or lymph nodes; or

2. Herpes simplex virus causing:

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

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

c. Disseminated infection; or

3. Herpes zoster, either disseminated or with multidermatomal eruptions that are resistant to treatment; or

4. Progressive multifocal leukoencephalopathy; or

5. Hepatitis, as described under the criteria in 5.05.

or

E. Malignant neoplasms:

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

2. Kaposi's sarcoma with:

a. Extensive oral lesions; or

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

c. Involvement of the skin or mucous membranes, as described under the criteria in 14.08F; or

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

4. Squamous cell carcinoma of the anus.

or

F. Conditions of the skin or mucous membranes (other than described in B2, D2, or D3, above) with extensive fungating or ulcerating lesions not responding to treatment (e.g., dermatological conditions such as eczema or psoriasis, vulvovaginal or other mucosal candida, condyloma caused by human papillomavirus, genital ulcerative disease), or evaluate under the criteria in 8.00ff.

or

G. Hematologic abnormalities:

1. Anemia, as described under the criteria in 7.02; or

2. Granulocytopenia, as described under the criteria in 7.15; or

3. Thrombocytopenia, as described under the criteria in 7.06.

or

H. Neurological abnormalities:

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

2. Other neurological manifestations of HIV infection (e.g., peripheral neuropathy) as described under the criteria in 11.00ff.

or

I. HIV wasting syndrome, characterized by involuntary weight loss of 10 percent or more of baseline (or other significant involuntary weight loss, as described in 14.00D2) and, in the absence of a concurrent illness that could explain the findings, either:

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

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

or

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

or

K. Cardiomyopathy, as described under the criteria in 4.00ff or 11.04.

or

L. Nephropathy, as described under the criteria in 6.00ff.

or

M. One or more of the following infections (other than described in A-L, above), resistant to treatment or requiring hospitalization or intravenous treatment 3 or more times in 1 year (or evaluate sequelae under the criteria for the affected body system).

1. Sepsis; or

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

or

N. Repeated (as defined in 14.00D8) manifestations of HIV infection (including those listed in 14.08A-M, but without the requisite findings, e.g., carcinoma of the cervix not meeting the criteria in 14.08E, diarrhea not meeting the criteria in 14.08J, or other manifestations, e.g., oral hairy leukoplakia, myositis) resulting in significant, documented symptoms or signs (e.g., fatigue, fever, malaise, weight loss, pain, night sweats) and one of the following at the marked level (as defined in 14.00D8):

1. Restriction of activities of daily living; or

2. Difficulties in maintaining social functioning; or

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

14.09 *Inflammatory arthritis.* Documented as described in 14.00B6, with one of the following:

A. History of joint pain, swelling, and tenderness, and signs on current physical examination of joint inflammation or deformity in two or more major joints resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 14.00B6b and 1.00B2b and B2c;

or

B. Ankylosing spondylitis or other spondyloarthropathy, with diagnosis established by findings of unilateral or bilateral sacroiliitis (e.g., erosions or fusions), shown by appropriate medically

acceptable imaging, with both:

1. History of back pain, tenderness, and stiffness, and

2. Findings on physical examination of ankylosis (fixation) of the dorsolumbar or cervical spine at 45° or more of flexion measured from the vertical position (zero degrees);

or

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

or

D. Inflammatory arthritis, with signs of peripheral joint inflammation on current examination, but with lesser joint involvement than in A and lesser extra-articular features than in C, and:

1. Significant, documented constitutional symptoms and signs (e.g., fatigue, fever, malaise, weight loss), and

2. Involvement of two or more organs/body systems (see 14.00B6d). At least one of the organs/body systems must be involved to at least a moderate level of severity.

or

E. Inflammatory spondylitis or other inflammatory spondyloarthropathies, with lesser deformity than in B and lesser extra-articular features than in C, with signs of unilateral or bilateral sacroiliitis on appropriate medically acceptable imaging; and with the extra-articular features described in 14.09D.

Part B

Medical criteria for the evaluation of impairments of children under age 18 (where criteria in part A do not give appropriate consideration to the particular disease process in childhood).

Sec.

100.00 Growth Impairment.

101.00 Musculoskeletal System.

102.00 Special Senses and Speech.

103.00 Respiratory System.

104.00 Cardiovascular System.

105.00 Digestive System.

106.00 Genitourinary Impairments.

107.00 Hematological Disorders.

108.00 Skin Disorders

109.00 Endocrine System.

110.00 Impairments That Affect Multiple Body Systems.

111.00 Neurological.

112.00 Mental Disorders.

113.00 Malignant Neoplastic Diseases.

114.00 Immune System.

100.00 Growth Impairment

A. *Impairment of growth* may be disabling in itself or it may be an indicator of the severity of the impairment due to a specific disease process.

Determinations of growth impairment should be based upon the comparison of current height with at least three previous determinations, including length at birth, if available. Heights (or lengths) should be plotted on a standard growth chart, such as derived from the National Center for Health Statistics: NCHS Growth Charts. Height should be measured without shoes. Body weight corresponding to the ages represented by the heights should be furnished. The adult heights of the child's natural parents and the heights and ages of siblings should also be furnished. This will provide a basis upon which to identify those children whose short stature represents a familial characteristic rather than a result of disease. This is particularly true for adjudication under 100.02B.

B. *Bone age determinations* should include a full descriptive report of medically acceptable imaging specifically obtained to determine bone age and must cite the standardization method used. Where appropriate medically acceptable imaging must be obtained currently as a basis for adjudication under 100.03, views or scans of the left hand and wrist should be ordered. In addition appropriate medically acceptable imaging of the knee and ankle should be obtained when cessation of growth is being evaluated in an older child at, or past, puberty. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myeolgraphy, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

C. The criteria in this section are applicable until closure of the major epiphyses. The cessation of significant increase in height at that point would prevent the application of these criteria.

100.01 Category of Impairments, Growth

100.02 *Growth impairment,* considered to be related to an additional specific medically determinable impairment, and one of the following:

A. Fall of greater than 15 percentiles in height which is sustained; or

B. Fall to, or persistence of, height below the third percentile.

100.03 *Growth impairment,* not identified as being related to a

101.00 Musculoskeletal System

A. *Disorders of the musculoskeletal system* may result from hereditary, congenital, or acquired pathologic processes. Impairments may result from infectious, inflammatory, or degenerative processes, traumatic or developmental events, or neoplastic, vascular, or toxic/metabolic diseases.

B. Loss of Function

1. *General.* Under this section, loss of function may be due to bone or joint deformity or destruction from any cause; miscellaneous disorders of the spine with or without radiculopathy or other neurological deficits; amputation; or fractures or soft tissue injuries, including burns, requiring prolonged periods of immobility or convalescence. For inflammatory arthritides that result in loss of function because of inflammatory peripheral joint or axial arthritis or sequelae, or because of extra-articular features, see 114.00E. Impairments with neurological causes are to be evaluated under 111.00ff.

2. How We Define Loss of Function in These Listings

a. *General.* Regardless of the cause(s) of a musculoskeletal impairment, functional loss for purposes of these listings is defined as the inability to ambulate effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment, or the inability to perform fine and gross movements effectively on a sustained basis for any reason, including pain associated with the underlying musculoskeletal impairment. The inability to ambulate effectively or the inability to perform fine and gross movements effectively must have lasted, or be expected to last, for at least 12 months. For the purposes of these criteria, consideration of the ability to perform these activities must be from a physical standpoint alone. When there is an inability to perform these activities due to a mental impairment, the criteria in 112.00ff are to be used. We will determine whether a child can ambulate effectively or can perform fine and gross movements effectively based on the medical and other evidence in the case record, generally without developing additional evidence about the child's ability to perform the specific activities listed as examples in 101.00B2b(2) and (3) and 101.00B2c(2) and (3).

b. What We Mean by Inability To Ambulate Effectively

(1) *Definition.* Inability to ambulate effectively means an extreme limitation of the ability to walk; *i.e.*, an impairment that interferes very seriously with the child's ability to independently initiate, sustain, or complete activities. Ineffective ambulation is defined generally as having insufficient lower extremity functioning (see 101.00J) to permit independent ambulation without the use of a hand-held assistive device(s) that limits the functioning of both upper extremities. (Listing 101.05C is an exception to this general definition because the child has the use of only one upper extremity due to amputation of a hand.)

(2) *How we assess inability to ambulate effectively for children too young to be expected to walk independently.* For children who are too young to be expected to walk independently, consideration of function must be based on assessment of limitations in the ability to perform comparable age-appropriate activities with the lower extremities, given normal developmental expectations. 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.

(3) *How we assess inability to ambulate effectively for older children.* Older children, who would be expected to be able to walk when compared to other children the same age who do not have impairments, must be capable of sustaining a reasonable walking pace over a sufficient distance to be able to carry out age-appropriate activities. They must have the ability to travel age-appropriately without extraordinary assistance to and from school or a place of employment. Therefore, examples of ineffective ambulation for older children include, but are not limited to, the inability to walk without the use of a walker, two crutches or two canes, the inability to walk a block at a reasonable pace on rough or uneven surfaces, the inability to use standard public transportation, the inability to carry out age-appropriate school activities independently, and the inability to climb a few steps at a reasonable pace with the use of a single hand rail. The ability to walk independently about the child's home or a short distance at school without the use of assistive devices does not, in and of itself, constitute effective ambulation.

c. What We Mean by Inability To Perform Fine and Gross Movements Effectively

(1) *Definition.* Inability to perform fine and gross movements effectively means an extreme loss of function of both upper extremities; *i.e.*, an impairment that interferes very seriously with the child's ability to independently initiate, sustain, or complete activities. To use their upper extremities effectively, a child must be capable of sustaining such functions as reaching, pushing, pulling, grasping, and fingering in an age-appropriate manner to be able to carry out age-appropriate activities.

(2) *How we assess inability to perform fine and gross movements in very young children*. For very young children, we consider limitations in the ability to perform comparable age-appropriate activities involving the upper extremities compared to the ability of children the same age who do not have impairments. 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.

(3) *How we assess inability to perform fine and gross movements in older children.* For older children, examples of inability to perform fine and gross movements effectively include, but are not limited to, the inability to prepare a simple meal and feed oneself, the inability to take care of personal hygiene, or the inability to sort and handle papers or files, depending upon which activities are age-appropriate.

d. *Pain or other symptoms.* Pain or other symptoms may be an important factor contributing to functional loss. In order for pain or other symptoms to be found to affect a child's ability to function in an age-appropriate manner or to perform basic work activities, medical signs or laboratory findings must show the existence of a medically determinable impairment(s) that could reasonably be expected to produce the pain or other symptoms. The musculoskeletal listings that include pain or other symptoms among their criteria also include criteria for

limitations in functioning as a result of the listed impairment, including limitations caused by pain. It is, therefore, important to evaluate the intensity and persistence of such pain or other symptoms carefully in order to determine their impact on the child's functioning under these listings. See also §§404.1525(f) and 404.1529 of this part, and §§416.925(f) and 416.929 of part 416 of this chapter.

C. Diagnosis and Evaluation

1. *General.* Diagnosis and evaluation of musculoskeletal impairments should be supported, as applicable, by detailed descriptions of the joints, including ranges of motion, condition of the musculature (e.g., weakness, atrophy), sensory or reflex changes, circulatory deficits, and laboratory findings, including findings on x-ray or other appropriate medically acceptable imaging. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

2. *Purchase of certain medically acceptable imaging.* While any appropriate medically acceptable imaging is useful in establishing the diagnosis of musculoskeletal impairments, some tests, such as CAT scans and MRIs, are quite expensive, and we will not routinely purchase them. Some, such as myelograms, are invasive and may involve significant risk. We will not order such tests. However, when the results of any of these tests are part of the existing evidence in the case record we will consider them together with the other relevant evidence.

3. *Consideration of electrodiagnostic procedures.* Electrodiagnostic procedures may be useful in establishing the clinical diagnosis, but do not constitute alternative criteria to the requirements of 101.04.

D. *The physical examination* must include a detailed description of the rheumatological, orthopedic, neurological, and other findings appropriate to the specific impairment being evaluated. These physical findings must be determined on the basis of objective observation during the examination and not simply a report of the child's allegation; e.g., "He says his leg is weak, numb." Alternative testing methods should be used to verify the abnormal findings; e.g., a seated straight-leg raising test in addition to a supine straight-leg raising test. Because abnormal physical findings may be intermittent, their presence over a period of time must be established by a record of ongoing management and evaluation. Care must be taken to ascertain that the reported examination findings are consistent with the child's age and activities.

E. Examination of the Spine

1. *General.* Examination of the spine should include a detailed description of gait, range of motion of the spine given quantitatively in degrees from the vertical position (zero degrees) or, for straight-leg raising from the sitting and supine position (zero degrees), any other appropriate tension signs, motor and sensory abnormalities, muscle spasm, when present, and deep tendon reflexes. Observations of the child during the examination should be reported; e.g., how he or she gets on and off the examination table. Inability to walk on the heels or toes, to squat, or to arise from a squatting position, when appropriate, may be considered evidence

of significant motor loss. However, a report of atrophy is not acceptable as evidence of significant motor loss without circumferential measurements of both thighs and lower legs, or both upper and lower arms, as appropriate, at a stated point above and below the knee or elbow given in inches or centimeters. Additionally, a report of atrophy should be accompanied by measurement of the strength of the muscle(s) in question generally based on a grading system of 0 to 5, with 0 being complete loss of strength and 5 being maximum strength. A specific description of atrophy of hand muscles is acceptable without measurements of atrophy but should include measurements of grip and pinch strength. However, because of the unreliability of such measurement in younger children, these data are not applicable to children under 5 years of age.

2. *When neurological abnormalities persist.* Neurological abnormalities may not completely subside after treatment or with the passage of time. Therefore, residual neurological abnormalities that persist after it has been determined clinically or by direct surgical or other observation that the ongoing or progressive condition is no longer present will not satisfy the required findings in 101.04. More serious neurological deficits (paraparesis, paraplegia) are to be evaluated under the criteria in 111.00ff.

F. *Major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

G. *Measurements of joint motion* are based on the techniques described in the chapter on the extremities, spine, and pelvis in the current edition of the "Guides to the Evaluation of Permanent Impairment" published by the American Medical Association.

H. Documentation.

1. *General.* Musculoskeletal impairments frequently improve with time or respond to treatment. Therefore, a longitudinal clinical record is generally important for the assessment of severity and expected duration of an impairment unless the child is a newborn or the claim can be decided favorably on the basis of the current evidence.

2. *Documentation of medically prescribed treatment and response.* Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever evidence of such treatment is available it must be considered.

3. *When there is no record of ongoing treatment.* Some children will not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In such cases, evaluation will be made on the basis of the current objective medical evidence and other available evidence, taking into consideration the child's medical history, symptoms, and medical source opinions. Even though a child who does not receive treatment may not be able to show an impairment that meets the criteria of one of the musculoskeletal listings, the child may have an impairment(s) that is either medically or, in the case of a claim for benefits under part 416 of this chapter, functionally equivalent in severity to one of the listed impairments.

4. *Evaluation when the criteria of a musculoskeletal listing are not met.* These listings are only examples of common musculoskeletal disorders that are severe enough to find a child disabled. Therefore, in any case in which a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will consider whether the child's impairment(s) is medically or, in the case of a claim for benefits under part 416 of this chapter, functionally equivalent in severity to the criteria of a listing. (See §§404.1526, 416.926, and 416.926a.) Individuals with claims for benefits under part 404, who have an impairment(s) with a level of severity that does not meet or equal the criteria of the musculoskeletal listings may or may not have the RFC that would enable them to engage in substantial gainful activity. Evaluation of the impairment(s) of these individuals should proceed through the final steps of the sequential evaluation process in §404.1520 (or, as appropriate, the steps in the medical improvement review standard in §404.1594).

I. Effects of Treatment

1. *General.* Treatments for musculoskeletal disorders may have beneficial effects or adverse side effects. Therefore, medical treatment (including surgical treatment) must be considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the disorder, and in terms of any side effects that may further limit the child.

2. *Response to treatment.* Response to treatment and adverse consequences of treatment may vary widely. For example, a pain medication may relieve a child's pain completely, partially, or not at all. It may also result in adverse effects, e.g., drowsiness, dizziness, or disorientation, that compromise the child's ability to function. Therefore, each case must be considered on an individual basis, and include consideration of the effects of treatment on the child's ability to function.

3. *Documentation.* 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 finding regarding the impact of treatment must be based on a sufficient period of treatment to permit proper consideration or judgment about future functioning.

J. Orthotic, Prosthetic, or Assistive Devices

1. *General.* Consistent with clinical practice, children with musculoskeletal impairments may be examined with and without the use of any orthotic, prosthetic, or assistive devices as explained in this section.

2. *Orthotic devices.* Examination should be with the orthotic device in place and should include an evaluation of the child's maximum ability to function effectively with the orthosis. It is unnecessary to routinely evaluate the child's ability to function without the orthosis in place. If the child has difficulty with, or is unable to use, the orthotic device, the medical basis for the difficulty should be documented. In such cases, if the impairment involves a lower extremity or extremities, the examination should include information on the child's ability to ambulate effectively without the device in place unless contraindicated by the medical judgment of a physician who has treated or examined the child.

3. *Prosthetic devices.* Examination should be with the prosthetic device in place. In

amputations involving a lower extremity or extremities, it is unnecessary to evaluate the child's ability to walk without the prosthesis in place. However, the child's medical ability to use a prosthesis to ambulate effectively, as defined in 101.00B2b, should be evaluated. The condition of the stump should be evaluated without the prosthesis in place.

4. *Hand-held assistive devices.* When a child with an impairment involving a lower extremity or extremities uses a hand-held assistive device, such as a cane, crutch or walker, examination should be with and without the use of the assistive device unless contraindicated by the medical judgment of a physician who has treated or examined the child. The child's ability to ambulate with and without the device provides information as to whether, or the extent to which, the child is able to ambulate without assistance. The medical basis for the use of any assistive device (e.g., instability, weakness) should be documented. The requirement to use a hand-held assistive device may also impact on the child's functional capacity by virtue of the fact that one or both upper extremities are not available for such activities as lifting, carrying, pushing, and pulling.

K. *Disorders of the spine,* listed in 101.04, result in limitations because of distortion of the bony and ligamentous architecture of the spine and associated impingement on nerve roots (including the cauda equina) or spinal cord. Such impingement on nerve tissue may result from a herniated nucleus pulposus or other miscellaneous conditions. Neurological abnormalities resulting from these disorders are to be evaluated by referral to the neurological listings in 111.00ff, as appropriate. (See also 101.00B and E.)

1. *Herniated nucleus pulposus* is a disorder frequently associated with the impingement of a nerve root, but occurs infrequently in children. Nerve root compression results in a specific neuro-anatomic distribution of symptoms and signs depending upon the nerve root(s) compromised.

2. *Other miscellaneous conditions* that may cause weakness of the lower extremities, sensory changes, areflexia, trophic ulceration, bladder or bowel incontinence, and that should be evaluated under 101.04 include, but are not limited to, lysosomal disorders, metabolic disorders, vertebral osteomyelitis, vertebral fractures and achondroplasia. Disorders such as spinal dysrhaphism, (e.g., spina bifida) diastematomyelia, and tethered cord syndrome may also cause such abnormalities. In these cases, there may be gait difficulty and deformity of the lower extremities based on neurological abnormalities, and the neurological effects are to be evaluated under the criteria in 111.00ff.

L. *Abnormal curvatures of the spine.* Abnormal curvatures of the spine (specifically, scoliosis, kyphosis and kyphoscoliosis) can result in impaired ambulation, but may also adversely affect functioning in body systems other than the musculoskeletal system. For example, a child's ability to breathe may be affected; there may be cardiac difficulties (e.g., impaired myocardial function); or there may be disfigurement resulting in withdrawal or isolation. When there is impaired ambulation, evaluation of equivalence may be made by reference to 114.09A. When the abnormal curvature of the spine results in symptoms related to fixation of the dorsolumbar or cervical spine, evaluation of equivalence may be made by reference to 114.09B. When there is respiratory or cardiac involvement or an associated mental disorder, evaluation may be made under 103.00ff, 104.00ff, or 112.00ff, as appropriate. Other consequences should be evaluated according to the listing for the affected body system.

M. *Under continuing surgical management,* as used in 101.07 and 101.08, refers to surgical

procedures and any other associated treatments related to the efforts directed toward the salvage or restoration of functional use of the affected part. It may include such factors as post-surgical procedures, surgical complications, infections, or other medical complications, related illnesses, or related treatments that delay the child's attainment of maximum benefit from therapy. When burns are not under continuing surgical management, see 108.00F.

N. *After maximum benefit from therapy has been achieved* in situations involving fractures of an upper extremity (101.07), or soft tissue injuries (101.08), *i.e.*, there have been no significant changes in physical findings or on appropriate medically acceptable imaging for any 6-month period after the last definitive surgical procedure or other medical intervention, evaluation must be made on the basis of the demonstrable residuals, if any. A finding that 101.07 or 101.08 is met must be based on a consideration of the symptoms, signs, and laboratory findings associated with recent or anticipated surgical procedures and the resulting recuperative periods, including any related medical complications, such as infections, illnesses, and therapies which impede or delay the efforts toward restoration of function. Generally, when there has been no surgical or medical intervention for 6 months after the last definitive surgical procedure, it can be concluded that maximum therapeutic benefit has been reached. Evaluation at this point must be made on the basis of the demonstrable residual limitations, if any, considering the child's impairment-related symptoms, signs, and laboratory findings, any residual symptoms, signs, and laboratory findings associated with such surgeries, complications, and recuperative periods, and other relevant evidence.

O. *Major function of the face and head,* for purposes of listing 101.08, relates to impact on any or all of the activities involving vision, hearing, speech, mastication, and the initiation of the digestive process.

P. *When surgical procedures have been performed,* documentation should include a copy of the operative notes and available pathology reports.

101.01 Category of Impairments, Musculoskeletal

101.02 *Major dysfunction of a joint(s) (due to any cause):* Characterized by gross anatomical deformity (e.g., subluxation, contracture, bony or fibrous ankylosis, instability) and chronic joint pain and stiffness with signs of limitation of motion or other abnormal motion of the affected joint(s), and findings on appropriate medically acceptable imaging of joint space narrowing, bony destruction, or ankylosis of the affected joint(s). With:

A. Involvement of one major peripheral weight-bearing joint (*i.e.*, hip, knee, or ankle), resulting in inability to ambulate effectively, as defined in 101.00B2b;

or

B. Involvement of one major peripheral joint in each upper extremity (*i.e.*, shoulder, elbow, or wrist-hand), resulting in inability to perform fine and gross movements effectively, as defined in 101.00B2c.

101.03 *Reconstructive surgery or surgical arthrodesis of a major weight-bearing joint,* with inability to ambulate effectively, as defined in 101.00B2b, and return to effective ambulation did not occur, or is not expected to occur, within 12 months of onset.

101.04 *Disorders of the spine* (e.g., lysosomal disorders, metabolic disorders, vertebral osteomyelitis, vertebral fracture, achondroplasia) resulting in compromise of a nerve root (including the cauda equina) or the spinal cord, with evidence of nerve root compression characterized by neuro-anatomic distribution of pain, limitation of motion of the spine, motor loss (atrophy with associated muscle weakness or muscle weakness) accompanied by sensory or reflex loss and, if there is involvement of the lower back, positive straight-leg raising test (sitting and supine).

101.05 *Amputation (due to any cause).*

A. Both hands;

or

B. One or both lower extremities at or above the tarsal region, with stump complications resulting in medical inability to use a prosthetic device to ambulate effectively, as defined in 101.00B2b, which have lasted or are expected to last for at least 12 months;

or

C. One hand and one lower extremity at or above the tarsal region, with inability to ambulate effectively, as defined in 101.00B2b;

or

D. Hemipelvectomy or hip disarticulation.

101.06 *Fracture of the femur, tibia, pelvis, or one or more of the tarsal bones.* With:

A. Solid union not evident on appropriate medically acceptable imaging, and not clinically solid;

and

B. Inability to ambulate effectively, as defined in 101.00B2b, and return to effective ambulation did not occur or is not expected to occur within 12 months of onset.

101.07 *Fracture of an upper extremity* with nonunion of a fracture of the shaft of the humerus, radius, or ulna, under continuing surgical management, as defined in 101.00M, directed toward restoration of functional use of the extremity, and such function was not restored or expected to be restored within 12 months of onset.

101.08 *Soft tissue injury (e.g., burns)* of an upper or lower extremity, trunk, or face and head, under continuing surgical management, as defined in 101.00M, directed toward the salvage or restoration of major function, and such major function was not restored or expected to be restored within 12 months of onset. Major function of the face and head is described in 101.00O.

102.00 Special Senses and Speech

A. *How do we evaluate visual disorders?*

1. *What are visual disorders?* Visual disorders are abnormalities of the eye, the optic nerve, the optic tracts, or the brain that may cause a loss of visual acuity or visual fields. A loss of visual acuity limits your ability to distinguish detail, read, do fine work, or perform other age-appropriate activities. A loss of visual fields limits your ability to perceive visual stimuli in the peripheral extent of vision.

2. *How do we define statutory blindness?* Statutory blindness is blindness as defined in sections 216(i)(1) and 1614(a)(2) of the Social Security Act (the Act). The Act defines blindness as visual acuity of 20/200 or less in the better eye with the use of a correcting lens. We use your best-corrected visual acuity for distance in the better eye when we determine if this definition is met. The Act also provides that an eye that has a visual field limitation such that the widest diameter of the visual field subtends an angle no greater than 20 degrees is considered as having visual acuity of 20/200 or less. You have statutory blindness only if your visual disorder meets the criteria of 102.02 or 102.03A. You do not have statutory blindness if your visual disorder medically equals the criteria of 102.02 or 102.03A, or if it meets or medically equals 102.03B, 102.03C, or 102.04. If your visual disorder medically equals the criteria of 102.02 or 102.03A, or if it meets or medically equals 102.03B, 102.03C, or 102.04, we will find that you have a disability if your visual disorder also meets the duration requirement.

3. *What evidence do we need to establish statutory blindness under title XVI?* For title XVI, the only evidence we need to establish statutory blindness is evidence showing that your visual acuity in your better eye or your visual field in your better eye meets the criteria in 102.00A2, provided that those measurements are consistent with the other evidence in your case record. We do not need to document the cause of your blindness. Also, there is no duration requirement for statutory blindness under title XVI (see §§416.981 and 416.983).

4. W*hat evidence do we need to evaluate visual disorders, including those that result in statutory blindness under title II?*

a. To evaluate your visual disorder, we usually need a report of an eye examination that includes measurements of the best-corrected visual acuity or the extent of the visual fields, as appropriate. If there is a loss of visual acuity or visual fields, the cause of the loss must be documented. A standard eye examination will usually reveal the cause of any visual acuity loss. An eye examination can also reveal the cause of some types of visual field deficits. If the eye examination does not reveal the cause of the visual loss, we will request the information that was used to establish the presence of the visual disorder.

b. A cortical visual disorder is a disturbance of the posterior visual pathways or occipital lobes of the brain in which the visual system does not interpret what the eyes are seeing. It may result from such causes as traumatic brain injury, stroke, cardiac arrest, near drowning, a central nervous system infection such as meningitis or encephalitis, a tumor, or surgery. It can be temporary or permanent, and the amount of visual loss can vary. It is possible to have a cortical visual disorder and not have any abnormalities observed in a standard eye examination. Therefore, a diagnosis of a cortical visual disorder must be confirmed by documentation of the cause of the brain lesion. If neuroimaging or visual evoked response (VER) testing was performed, we will request a copy of the report or other medical evidence that describes the findings in the report.

c. If your visual disorder does not satisfy the criteria in 102.02, 102.03, or 102.04, we will also

request a description of how your visual disorder impacts your ability to function.

5. *How do we measure best-corrected visual acuity?*

a. *Testing for visual acuity.*

(i) When we need to measure your best-corrected visual acuity, we will use visual acuity testing that was carried out using Snellen methodology or any other testing methodology that is comparable to Snellen methodology.

(ii) We consider tests such as the Landolt C test or the tumbling-E test, which are used to evaluate young children who are unable to participate in testing using Snellen methodology, to be comparable to testing using Snellen methodology. These alternate methods for measuring visual acuity should be performed by specialists with expertise in assessment of childhood vision.

(iii) If you are unable to participate in testing using Snellen methodology or other comparable testing, we will consider your fixation and visual-following behavior. If both these behaviors are absent, we will consider the anatomical findings or the results of neuroimaging, electroretinogram, or VER testing when this testing has been performed.

b. Determining best-corrected visual acuity. (i) Best-corrected visual acuity is the optimal visual acuity attainable with the use of a corrective lens. In some instances, this assessment may be performed using a specialized lens; for example, a contact lens. We will use the visual acuity measurements obtained with a specialized lens only if you have demonstrated the ability to use the specialized lens on a sustained basis. However, we will not use visual acuity measurements obtained with telescopic lenses because they significantly reduce the visual field. If you have an absent response to VER testing in an eye, we can determine that your best-corrected visual acuity is 20/200 or less in that eye. However, if you have a positive response to VER testing in an eye, we will not use that result to determine your best-corrected visual acuity in that eye. Additionally, we will not use the results of pinhole testing or automated refraction acuity to determine your best-corrected visual acuity.

(ii) We will use the best-corrected visual acuity for distance in your better eye when we determine whether your loss of visual acuity satisfies the criteria in 102.02A. The best-corrected visual acuity for distance is usually measured by determining what you can see from 20 feet. If your visual acuity is measured for a distance other than 20 feet, we will convert it to a 20-foot measurement. For example, if your visual acuity is measured at 10 feet and is reported as 10/40, we will convert this to 20/80.

(iii) If you cannot participate in visual acuity testing, we will determine that your best-corrected visual acuity is 20/200 or less in your better eye if your visual disorder meets the criteria in 102.02B. To meet 102.02B1, your impairment must result in the absence of fixation and visual-following behavior and abnormal anatomical findings indicating a visual acuity of 20/200 or less in your better eye. Such abnormal anatomical findings include, but are not limited to, the presence of Stage III or worse retinopathy of prematurity despite surgery, hypoplasia of the optic nerve, albinism with macular aplasia, and bilateral optic atrophy. To meet 102.02B2, your impairment must result in the absence of fixation and visual-following behavior and abnormal neuroimaging documenting damage to the cerebral cortex which would be expected to prevent the development of a visual acuity better than 20/200 in your better eye. Such abnormal

neuroimaging includes, but is not limited to, neuroimaging showing bilateral encephalomyelitis or bilateral encephalomalacia.

6. *How do we measure visual fields?*

a. *Testing for visual fields.*

(i) We generally need visual field testing when you have a visual disorder that could result in visual field loss, such as glaucoma, retinitis pigmentosa, or optic neuropathy, or when you display behaviors that suggest a visual field loss.

(ii) When we need to measure the extent of your visual field loss, we will use visual field measurements obtained with an automated static threshold perimetry test performed on a perimeter, like the Humphrey Field Analyzer, that satisfies all of the following requirements:

A. The perimeter must use optical projection to generate the test stimuli.

B. The perimeter must have an internal normative database for automatically comparing your performance with that of the general population.

C. The perimeter must have a statistical analysis package that is able to calculate visual field indices, particularly mean deviation.

D. The perimeter must demonstrate the ability to correctly detect visual field loss and correctly identify normal visual fields.

E. The perimeter must demonstrate good test-retest reliability.

F. The perimeter must have undergone clinical validation studies by three or more independent laboratories with results published in peer-reviewed ophthalmic journals.

(iii) The test must use a white size III Goldmann stimulus and a 31.5 apostilb (10 cd/m$^{[2]}$) white background. The stimuli locations must be no more than 6 degrees apart horizontally or vertically. Measurements must be reported on standard charts and include a description of the size and intensity of the test stimulus.

(iv) To determine statutory blindness based on visual field loss (102.03A), we need a test that measures the central 24 to 30 degrees of the visual field; that is, the area measuring 24 to 30 degrees from the point of fixation. Acceptable tests include the Humphrey 30-2 or 24-2 tests.

(v) The criterion in 102.03B is based on the use of a test performed on a Humphrey Field Analyzer that measures the central 30 degrees of the visual field. We can also use comparable results from other acceptable perimeters; for example, a mean defect of 22 on an acceptable Octopus test, to determine that the criterion in 102.03B is met. We cannot use tests that do not measure the central 30 degrees of the visual field, such as the Humphrey 24-2 test, to determine if your impairment meets or medically equals 102.03B.

(vi) We measure the extent of visual field loss by determining the portion of the visual field in which you can see a white III4e stimulus. The "III" refers to the standard Goldmann test

stimulus size III, and the "4e" refers to the standard Goldmann intensity filters used to determine the intensity of the stimulus.

(vii) In automated static threshold perimetry, the intensity of the stimulus varies. The intensity of the stimulus is expressed in decibels (dB). We need to determine the dB level that corresponds to a 4e intensity for the particular perimeter being used. We will then use the dB printout to determine which points would be seen at a 4e intensity level. For example, in Humphrey Field Analyzers, a 10 dB stimulus is equivalent to a 4e stimulus. A dB level that is higher than 10 represents a dimmer stimulus, while a dB level that is lower than 10 represents a brighter stimulus. Therefore, for tests performed on Humphrey Field Analyzers, any point seen at 10 dB or higher is a point that would be seen with a 4e stimulus.

(viii) We can also use visual field measurements obtained using kinetic perimetry, such as the Humphrey "SSA Test Kinetic" or Goldmann perimetry, instead of automated static threshold perimetry. The kinetic test must use a white III4e stimulus projected on a white 31.5 apostilb (10 cd/m$^{[2]}$) background. In automated kinetic tests, such as the Humphrey "SSA Test Kinetic," testing along a meridian stops when you see the stimulus. Because of this, automated kinetic testing does not detect limitations in the central visual field. If your visual disorder has progressed to the point at which it is likely to result in a significant limitation in the central visual field, such as a scotoma (see 102.00A8c), we will not use automated kinetic perimetry to evaluate your visual field loss. Instead, we will assess your visual field loss using automated static threshold perimetry or manual kinetic perimetry.

(ix) We will not use the results of visual field screening tests, such as confrontation tests, tangent screen tests, or automated static screening tests, to determine that your impairment meets or medically equals a listing, or functionally equals the listings. However, we can consider normal results from visual field screening tests to determine whether your visual disorder is severe when these test results are consistent with the other evidence in your case record. (See §416.924(c).) We will not consider normal test results to be consistent with the other evidence if either of the following applies:

A. The clinical findings indicate that your visual disorder has progressed to the point that it is likely to cause visual field loss; or

B. You have a history of an operative procedure for retinal detachment.

b. *Use of corrective lenses.* You must not wear eyeglasses during the visual field examination because they limit your field of vision. Contact lenses or perimetric lenses may be used to correct visual acuity during the visual field examination in order to obtain the most accurate visual field measurements. For this single purpose, you do not need to demonstrate that you have the ability to use the contact or perimetric lenses on a sustained basis.

7. *How do we calculate visual efficiency?*

a. *Visual acuity efficiency.* We use the percentage shown in Table 1 that corresponds to the best-corrected visual acuity for distance in your better eye.

b. *Visual field efficiency.* We use kinetic perimetry to calculate visual field efficiency by adding the number of degrees seen along the eight principal meridians in your better eye and dividing by 500. (See Table 2.)

c. *Visual efficiency.* We calculate the percent of visual efficiency by multiplying the visual acuity efficiency by the visual field efficiency and converting the decimal to a percentage. For example, if your visual acuity efficiency is 75 percent and your visual field efficiency is 64 percent, we will multiply 0.75 × 0.64 to determine that your visual efficiency is 0.48, or 48 percent.

8. *How do we evaluate specific visual problems?*

a. *Statutory blindness.* Most test charts that use Snellen methodology do not have lines that measure visual acuity between 20/100 and 20/200. Newer test charts, such as the Bailey-Lovie or the Early Treatment Diabetic Retinopathy Study (ETDRS), do have lines that measure visual acuity between 20/100 and 20/200. If your visual acuity is measured with one of these newer charts, and you cannot read any of the letters on the 20/100 line, we will determine that you have statutory blindness based on a visual acuity of 20/200 or less. For example, if your best-corrected visual acuity for distance in the better eye was determined to be 20/160 using an ETDRS chart, we will find that you have statutory blindness. Regardless of the type of test chart used, you do not have statutory blindness if you can read at least one letter on the 20/100 line. For example, if your best-corrected visual acuity for distance in the better eye was determined to be 20/125+1 using an ETDRS chart, we will find that you do not have statutory blindness as you are able to read one letter on the 20/100 line.

b. *Blepharospasm.* This movement disorder is characterized by repetitive, bilateral, involuntary closure of the eyelids. If you have this disorder, you may have measurable visual acuities and visual fields that do not satisfy the criteria of 102.02 or 102.03. Blepharospasm generally responds to therapy. However, if therapy is not effective, we will consider how the involuntary closure of your eyelids affects your ability to maintain visual functioning over time.

c. *Scotoma.* A scotoma is a non-seeing area in the visual field surrounded by a seeing area. When we measure the visual field, we subtract the length of any scotoma, other than the normal blind spot, from the overall length of any diameter on which it falls.

B. *Hearing impairments in children.* The criteria for hearing impairments in children take into account that a lesser impairment in hearing which occurs at an early age may result in a severe speech and language disorder.

Improvement by a hearing aid, as predicted by the testing procedure, must be demonstrated to be feasible in that child, since younger children may be unable to use a hearing aid effectively.

The type of audiometric testing performed must be described and a copy of the results must be included. The pure tone air conduction hearing levels in 102.08 are based on American National Standard Institute Specifications for Audiometers, S3.6-1969 (ANSI-1969). The report should indicate the specifications used to calibrate the audiometer.

The finding of a severe impairment will be based on the average hearing levels at 500, 1000, 2000, and 3000 Hertz (Hz) in the better ear, and on speech discrimination, as specified in §102.08.

C. *How do we evaluate impairments that do not meet one of the special senses and speech listings?*

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

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

102.01 *Category of Impairments, Special Senses and Speech*

102.02 *Loss of visual acuity.*

A. Remaining vision in the better eye after best correction is 20/200 or less;

OR

B. An inability to participate in testing using Snellen methodology or other comparable visual acuity testing and clinical findings that fixation and visual-following behavior are absent in the better eye, and:

1. Abnormal anatomical findings indicating a visual acuity of 20/200 or less in the better eye; or

2. Abnormal neuroimaging documenting damage to the cerebral cortex which would be expected to prevent the development of a visual acuity better than 20/200 in the better eye; or

3. Abnormal electroretinogram documenting the presence of Leber's congenital amaurosis or achromatopsia; or

4. An absent response to VER testing in the better eye.

102.03 *Contraction of the visual field in the better eye,* with:

A. The widest diameter subtending an angle around the point of fixation no greater than 20 degrees;

OR

B. A mean deviation of -22 or worse, determined by automated static threshold perimetry as described in 102.00A6a(v);

OR

C. A visual field efficiency of 20 percent or less as determined by kinetic perimetry (see 102.00A7b).

102.04 *Loss of visual efficiency.* Visual efficiency of the better eye of 20 percent or less after

best correction (see 102.00A7c).

102.08 *Hearing impairments.*

A. For children below 5 years of age at time of adjudication, inability to hear air conduction thresholds at an average of 40 decibels (db) hearing level or greater in the better ear; or

B. For children 5 years of age and above at time of adjudication:

1. Inability to hear air conduction thresholds at an average of 70 decibels (db) or greater in the better ear; or

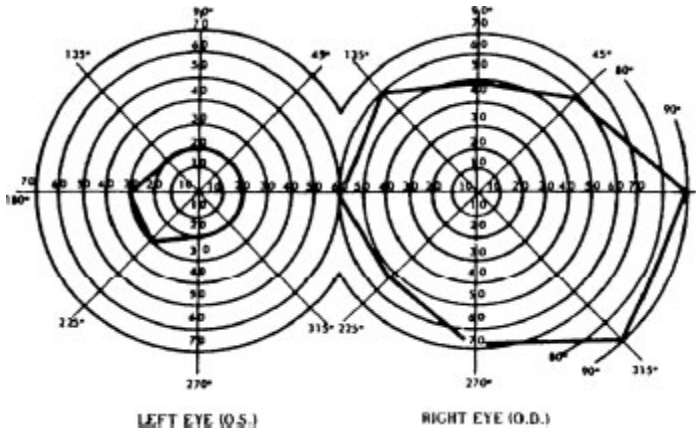
2. Speech discrimination scores at 40 percent or less in the better ear; or

3. Inability to hear air conduction thresholds at an average of 40 decibels (db) or greater in the better ear, and a speech and language disorder which significantly affects the clarity and content of the speech and is attributable to the hearing impairment.

Table 1.—Percentage of Visual Acuity Efficiency Corresponding to the Best-Corrected Visual Acuity Measurement for Distance in the Better Eye

Snellen		Percent visual acuity efficiency
English	Metric	
20/16	6/5	100
20/20	6/6	100
20/25	6/7.5	95
20/30	6/9	90
20/40	6/12	85
20/50	6/15	75
20/60	6/18	70
20/70	6/21	65
20/80	6/24	60
20/100	6/30	50

Table 2.—Chart of Visual Fields



1. The diagram of the right eye illustrates the extent of a normal visual field as measured with a III4e stimulus. The sum of the eight principal meridians of this field is 500 degrees.

2. The diagram of the left eye illustrates a visual field contracted to 30 degrees in two meridians and to 20 degrees in the remaining six meridians. The percent of visual field efficiency of this field is: (2 × 30) + (6 × 20) = 180 ÷ 500 = 0.36 or 36 percent visual field efficiency.

103.00 Respiratory System

A. *Introduction.* The listings in this section describe impairments resulting from respiratory disorder based on symptoms, physical signs, laboratory test abnormalities, and response to a regimen of treatment prescribed by a treating source. Respiratory disorders, along with any associated impairment(s) must be established by medical evidence. Evidence must be provided in sufficient detail to permit an independent reviewer to evaluate the severity of the impairment. Reasonable efforts should be made to ensure evaluation by a program physician specializing in childhood respiratory impairments or a qualified pediatrician.

Many children, especially those who have listing-level impairments, will have received the benefit of medically prescribed treatment. Whenever there is such evidence, the longitudinal clinical record must include a description of the treatment prescribed by the treating source and response, in addition to information about the nature and severity of the impairment. It is important to document any prescribed treatment and response because this medical management may have improved the child's functional status. The longitudinal record should provide information regarding functional recovery, if any.

Some children will not have received ongoing treatment or have an ongoing relationship with the medical community, despite the existence of a severe impairment(s). A child who does not receive treatment may or may not be able to show an impairment that meets the criteria of these listings. Even if a child does not show that his or her impairment meets the criteria of these listings, the child may have an impairment(s) that medically or functionally equals the listings. Unless the claim can be decided favorably on the basis of the current evidence, a longitudinal record is still important because it will provide information about such things as the ongoing medical severity of the impairment, the level of the child's functioning, and the frequency, severity, and duration of symptoms. Also, the asthma listing specifically includes a requirement for continuing signs and symptoms despite a regimen of prescribed treatment.

Evaluation should include consideration of adverse effects of respiratory impairment in all relevant body systems, and especially on the child's growth and development or mental functioning, as described under the growth impairment (100.00), neurological (111.00), and mental disorders (112.00) listings.

It must be remembered that these listings are only examples of common respiratory disorders that are severe enough to find a child disabled. When a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will make a determination whether the child's impairment(s) medically or functionally equals the listings. (See §§404.1526, 416.926, and 416.926a.)

B. *Documentation of Pulmonary Function Testing.* The results of spirometry that are used for adjudication, under the 103.02 A and B, 103.03, and 103.04 of these listings should be expressed in liters (L), body temperature and pressure saturated with water vapor (BTPS). The reported one-second forced expiratory volume (FEV_1) and forced vital capacity (FVC) should represent the largest of at least three satisfactory forced expiratory maneuvers. Two of the satisfactory spirograms should be reproducible for both pre-bronchodilator tests and, if indicated, post-bronchodilator tests. A value is considered reproducible if it does not differ from the largest value by more than 5 percent or 0.1 L, whichever is greater. The highest values of the FEV_1 and FVC, whether from the same or different tracings, should be used to assess the severity of the respiratory impairment. Peak flow should be achieved early in expiration, and the spirogram should have a smooth contour with gradually decreasing flow throughout expiration. The zero time for measurement of the FEV_1 and FVC, if not distinct, should be derived by linear back-extrapolation of peak flow to zero volume. A spirogram is satisfactory for measurement of the FEV_1 if the expiratory volume at the back-extrapolated zero time is less than 5 percent of the FVC or 0.1 L, whichever is greater. The spirogram is satisfactory for measurement of the FVC if maximal expiratory effort continues for at least 6 seconds, or if there is a plateau in the volume-time curve with no detectable change in expired volume (VE) during the last 2 seconds of maximal expiratory effort.

Spirometry should be repeated after administration of an aerosolized bronchodilator under supervision of the testing personnel if the pre-bronchodilator FEV_1 value is less than the appropriate reference value in table I or III, as appropriate. If a bronchodilator is not administered, the reason should be clearly stated in the report. Pulmonary function studies should not be performed unless the clinical status is stable (e.g., the child is not having an asthmatic attack or suffering from an acute respiratory infection or other chronic illness). Wheezing is common in asthma, chronic bronchitis, or chronic obstructive pulmonary disease and does not preclude testing. Pulmonary function studies performed to assess airflow obstruction without testing after bronchodilators cannot be used to assess levels of impairment in the range that prevents a child from performing age-appropriate activities, unless the use of bronchodilators is contraindicated. Post-bronchodilator testing should be performed 10 minutes after bronchodilator administration. The dose and name of the bronchodilator administered should be specified. The values in 103.02 and 103.04 must only be used as criteria for the level of ventilatory impairment that exists during the child's most stable state of health (*i.e.*, any period in time except during or shortly after an exacerbation).

The appropriately labeled spirometric tracing, showing the child's name, date of testing, distance per second on the abscissa and distance per liter (L) on the ordinate, must be

incorporated into the file. The manufacturer and model number of the device used to measure and record the spirogram should be stated. The testing device must accurately measure both time and volume, the latter to within 1 percent of a 3 L calibrating volume. If the spirogram was generated by any means other than direct pen linkage to a mechanical displacement-type spirometer, the testing device must have had a recorded calibration performed previously on the day of the spirometric measurement.

If the spirometer directly measures flow, and volume is derived by electronic integration, the linearity of the device must be documented by recording volume calibrations at three different flow rates of approximately 30 L/min (3 L/6 sec), 60 L/min (3 L/3 sec), and 180 L/min (3 L/sec). The volume calibrations should agree to within 1 percent of a 3 L calibrating volume. The proximity of the flow sensor to the child should be noted, and it should be stated whether or not a BTPS correction factor was used for the calibration recordings and for the child's actual spirograms.

The spirogram must be recorded at a speed of at least 20 mm/sec and the recording device must provide a volume excursion of at least 10 mm/L. If reproductions of the original spirometric tracings are submitted, they must be legible and have a time scale of at least 20 mm/sec and a volume scale of at least 10 mm/L to permit independent measurements. Calculation of FEV_1 from a flow volume tracing is not acceptable, *i.e.*, the spirogram and calibrations must be presented in a volume-time format at a speed of at least 20 mm/sec and a volume excursion of at least 10 mm/L to permit independent evaluation.

A statement should be made in the pulmonary function test report of the child's ability to understand directions, as well as his or her efforts and cooperation in performing the pulmonary function tests.

Purchase of a pulmonary function test is appropriate only when the child is capable of performing reproducible forced expiratory maneuvers. This capability usually occurs around age 6. Purchase of a pulmonary function test may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

The pulmonary function tables in 103.02 and 103.04 are based on measurement of standing height without shoes. If a child has marked spinal deformities (e.g., kyphoscoliosis), the measured span between the fingertips with the upper extremities abducted 90 degrees should be substituted for height when this measurement is greater than the standing height without shoes.

C. *Documentation of chronic impairment of gas exchange.*

1. *Arterial blood gas studies (ABGS).* An ABGS performed at rest (while breathing room air, awake and sitting or standing) should be analyzed in a laboratory certified by a State or Federal agency. If the laboratory is not certified, it must submit evidence of participation in a national proficiency testing program as well as acceptable quality control at the time of testing. The report should include the altitude of the facility and the barometric pressure on the date of analysis.

Purchase of resting ABGS may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be

favorably decided. Before purchasing resting ABGS, a program physician, preferably one experienced in the care of children with pulmonary disease, must review the clinical and laboratory data short of this procedure, including spirometry, to determine whether obtaining the test would present a significant risk to the child.

2. *Oximetry.* Pulse oximetry may be substituted for arterial blood gases in children under 12 years of age. The oximetry unit should employ the basic technology of spectrophotometric plethysmography as described in Taylor, M.B., and Whitwain, J.G., "Current Status of Pulse Oximetry," "Anesthesia," Vol. 41, No. 9, pp. 943-949, 1986. The unit should provide a visual display of the pulse signal and the corresponding oxygen saturation. A hard copy of the readings (heart rate and saturation) should be provided. Readings should be obtained for a minimum of 5 minutes. The written report should describe patient activity during the recording, *i.e.*, sleep rate, feeding, or exercise. Correlation between the actual heart rate determined by a trained observer and that displayed by the oximeter should be provided. A statement should be made in the report of the child's effort and cooperation during the test.

Purchase of oximetry may be appropriate when there is a question of whether an impairment meets or is equivalent in severity to a listing, and the claim cannot otherwise be favorably decided.

D. *Cystic fibrosis* is a disorder that affects either the respiratory or digestive body systems or both and may impact on a child's growth and development. It is responsible for a wide and variable spectrum of clinical manifestations and complications. Confirmation of the diagnosis is based upon an elevated sweat sodium concentration or chloride concentration accompanied by one or more of the following: the presence of chronic obstructive pulmonary disease, insufficiency of exocrine pancreatic function, meconium ileus, or a positive family history. The quantitative pilocarpine iontophoresis procedure for collection of sweat content must be utilized. Two methods are acceptable: the "Procedure for the Quantitative Iontophoretic Sweat Test for Cystic Fibrosis," published by the Cystic Fibrosis Foundation and contained in, "A Test for Concentration of Electrolytes in Sweat in Cystic Fibrosis of the Pancreas Utilizing Pilocarpine Iontophoresis," Gibson, I.E., and Cooke, R.E., "Pediatrics," Vol 23: 545, 1959; or the "Wescor Macroduct System." To establish the diagnosis of cystic fibrosis, the sweat sodium or chloride content must be analyzed quantitatively using an acceptable laboratory technique. Another diagnostic test is the "CF gene mutation analysis" for homozygosity of the cystic fibrosis gene. The pulmonary manifestations of this disorder should be evaluated under 103.04. The nonpulmonary aspects of cystic fibrosis should be evaluated under the listings for the digestive system (105.00) or growth impairments (100.00). Because cystic fibrosis may involve the respiratory and digestive body systems, as well as impact on a child's growth and development, the combined effects of this involvement must be considered in case adjudication.

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

E. *Bronchopulmonary dysplasia (BPD).* Bronchopulmonary dysplasia is a form of chronic obstructive pulmonary disease that arises as a consequence of acute lung injury in the newborn period and treatment of hyaline membrane disease, meconium aspiration, neonatal pneumonia and apnea of prematurity. The diagnosis is established by the requirement for

continuous or nocturnal supplemental oxygen for more than 30 days, in association with characteristic changes on medically acceptable imaging and clinical signs of respiratory dysfunction, including retractions, rales, wheezing, and tachypnea.

103.01 Category of Impairments, Respiratory System

103.02 *Chronic pulmonary insufficiency.* With:

A. Chronic obstructive pulmonary disease, due to any cause, with the FEV_1 equal to or less than the value specified in table I corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Table I

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
119 or less	46 or less	0.65
120-129	47-50	0.75
130-139	51-54	0.95
140-149	55-58	1.15
150-159	59-62	1.35
160-164	63-64	1.45
165-169	65-66	1.55
170 or more	67 or more	1.65

Or

B. Chronic restrictive ventilatory disease, due to any cause, with the FVC equal to or less than the value specified in table II corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Table II

Height without shoes (centimeters)	Height without shoes (inches)	FVC equal to or less than (L, BTPS)
119 or less	46 or less	0.65
120-129	47-50	0.85
130-139	51-54	1.05
140-149	55-58	1.25
150-159	59-62	1.45
160-164	63-64	1.65
165-169	65-66	1.75
170 or more	67 or more	2.05

Or

C. Frequent need for:

1. Mechanical ventilation; or

2. Nocturnal supplemental oxygen as required by persistent or recurrent episodes of hypoxemia;

Or

D. The presence of a tracheostomy in a child under 3 years of age;

Or

E. Bronchopulmonary dysplasia characterized by two of the following:

1. Prolonged expirations; or

2. Intermittent wheezing or increased respiratory effort as evidenced by retractions, flaring and tachypnea; or

3. Hyperinflation and scarring on a chest radiograph or other appropriate imaging techniques; or

4. Bronchodilator or diuretic dependency; or

5. A frequent requirement for nocturnal supplemental oxygen; or

6. Weight disturbance with:

a. 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) which persists for 2 months or longer; or

b. 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) which persists for 2 months or longer;

Or

F. Two required hospital admissions (each longer than 24 hours) within a 6-month period for recurrent lower respiratory tract infections or acute respiratory distress associated with:

1. Chronic wheezing or chronic respiratory distress; or

2. Weight disturbance with:

a. 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) which persists for 2 months or longer; or

b. 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) which persists for 2 months or longer;

Or

G. Chronic hypoventilation ($PaCO_2$ greater than 45 mm Hg) or chronic cor pulmonale as described under the appropriate criteria in 104.02;

Or

H. Growth impairment as described under the criteria in 100.00.

103.03 *Asthma.* With:

A. FEV_1 equal to or less than the value specified in table I of 103.02A;

Or

B. Attacks (as defined in 3.00C), in spite of prescribed treatment and requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for control of asthma counts as two attacks, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of attacks;

Or

C. Persistent low-grade wheezing between acute attacks or absence of extended symptom-free periods requiring daytime and nocturnal use of sympathomimetic bronchodilators with one of the following:

1. Persistent prolonged expiration with radiographic or other appropriate imaging techniques evidence of pulmonary hyperinflation or peribronchial disease; or

2. Short courses of corticosteroids that average more than 5 days per month for at least 3 months during a 12-month period;

Or

D. Growth impairment as described under the criteria in 100.00.

103.04 *Cystic fibrosis.* With:

A. An FEV_1 equal to or less than the appropriate value specified in table III corresponding to the child's height without shoes. (In cases of marked spinal deformity, see 103.00B.);

Or

B. For children in whom pulmonary function testing cannot be performed, the presence of two of the following:

1. History of dyspnea on exertion or accumulation of secretions as manifested by repetitive

coughing or cyanosis; or

2. Persistent bilateral rales and rhonchi or substantial reduction of breath sounds related to mucous plugging of the trachea or bronchi; or

3. Appropriate medically acceptable imaging evidence of extensive disease, such as thickening of the proximal bronchial airways or persistence of bilateral peribronchial infiltrates;

Or

C. Persistent pulmonary infection accompanied by superimposed, recurrent, symptomatic episodes of increased bacterial infection occurring at least once every 6 months and requiring intravenous or nebulization antimicrobial treatment;

Or

D. Episodes of bronchitis or pneumonia or hemoptysis (more than blood-streaked sputum) or respiratory failure (documented according to 3.00C), requiring physician intervention, occurring at least once every 2 months or at least six times a year. Each inpatient hospitalization for longer than 24 hours for treatment counts as two episodes, and an evaluation period of at least 12 consecutive months must be used to determine the frequency of episodes;

Or

E. Growth impairment as described under the criteria in 100.00.

Table III

[Applicable only for evaluation under 103.04A—cystic fibrosis]

Height without shoes (centimeters)	Height without shoes (inches)	FEV_1 equal to or less than (L, BTPS)
119 or less	46 or less	0.75
120-129	47-50	0.85
130-139	51-54	1.05
140-149	55-58	1.35
150-159	59-62	1.55
160-164	63-64	1.85
165-169	65-66	2.05
170 or more	67 or more	2.25

103.05 *Lung transplant.* Consider under a disability for 12 months following the date of surgery; thereafter, evaluate the residual impairment(s).

104.00 Cardiovascular System

A. General

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

a. We mean any disorder that affects the proper functioning of the heart or the circulatory system (that is, arteries, veins, capillaries, and the lymphatic drainage). The disorder can be congenital or acquired.

b. Cardiovascular impairment results from one or more of four consequences of heart disease:

(i) Chronic heart failure or ventricular dysfunction.

(ii) Discomfort or pain due to myocardial ischemia, with or without necrosis of heart muscle.

(iii) Syncope, or near syncope, due to inadequate cerebral perfusion from any cardiac cause, such as obstruction of flow or disturbance in rhythm or conduction resulting in inadequate cardiac output.

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

c. Disorders of the veins or arteries (for example, obstruction, rupture, or aneurysm) may cause impairments of the lower extremities (peripheral vascular disease), the central nervous system, the eyes, the kidneys, and other organs. We will evaluate peripheral vascular disease under 4.11 or 4.12 in part A, and impairments of another body system(s) under the listings for that body system(s).

2. *What do we consider in evaluating cardiovascular impairments?* The listings in this section describe cardiovascular impairments based on symptoms, signs, laboratory findings, response to a regimen of prescribed treatment, and functional limitations.

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

a. *Medical consultant* is an individual defined in §§404.1616(a) and 416.1016(a). This term does not include medical sources who provide consultative examinations for us. We use the abbreviation "MC" throughout this section to designate a medical consultant.

b. *Persistent* means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been present, or is expected to be present, for a continuous period of at least 12 months, such that a pattern of continuing severity is established.

c. *Recurrent* means that the longitudinal clinical record shows that, within a consecutive 12-month period, the finding(s) occurs at least three times, with intervening periods of improvement of sufficient duration that it is clear that separate events are involved.

d. *Appropriate medically acceptable imaging* means that the technique used is the proper one to evaluate and diagnose the impairment and is commonly recognized as accurate for assessing the cited finding.

e. *A consecutive 12-month period* means a period of 12 consecutive months, all or part of which must occur within the period we are considering in connection with an application or continuing disability review.

f. *Currently present* means that the finding is present at the time of adjudication.

g. *Uncontrolled* means the impairment does not respond adequately to standard prescribed medical treatment.

B. Documenting Cardiovascular Impairment

1. *What basic documentation do we need?* We need sufficiently detailed reports of history, physical examinations, laboratory studies, and any prescribed treatment and response to allow us to assess the severity and duration of your cardiovascular impairment. A longitudinal clinical record covering a period of not less than 3 months of observations and treatment is usually necessary, unless we can make a determination or decision based on the current evidence.

2. *Why is a longitudinal clinical record important?* We will usually need a longitudinal clinical record to assess the severity and expected duration of your impairment(s). If you have a listing-level impairment, you probably will have received medically prescribed treatment. Whenever there is evidence of such treatment, your longitudinal clinical record should include a description of the ongoing management and evaluation provided by your treating or other medical source. It should also include your response to this medical management, as well as information about the nature and severity of your impairment. The record will provide us with information on your functional status over an extended period of time and show whether your ability to function is improving, worsening, or unchanging.

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

a. You may not have received ongoing treatment or have an ongoing relationship with the medical community despite the existence of a severe impairment(s). In this situation, we will base our evaluation on the current objective medical evidence and the other evidence we have. If you do not receive treatment, you cannot show an impairment that meets the criteria of these listings. However, we may find you disabled because you have another impairment(s) that in combination with your cardiovascular impairment medically equals the severity of a listed impairment or that functionally equals the listings.

b. Unless we can decide your claim favorably on the basis of the current evidence, a longitudinal record is still important. In rare instances where there is no or insufficient longitudinal evidence, we may purchase a consultative examination(s) to help us establish the severity and duration of your impairment.

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

a. We will wait when we have information showing that your impairment is not yet stable and the expected change in your impairment might affect our determination or decision. In these situations, we need to wait to properly evaluate the severity and duration of your impairment during a stable period. Examples of when we might wait are:

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

(ii) If you have recently had a corrective cardiac procedure; for example, open-heart surgery.

(iii) If you have started new drug therapy and your response to this treatment has not yet been established; for example, beta-blocker therapy for dilated congestive cardiomyopathy.

b. In these situations, we will obtain more evidence 3 months following the event before we evaluate your impairment. However, we will not wait if we have enough information to make a determination or decision based on all of the relevant evidence in your case.

5. *Will we purchase any studies?* In appropriate situations, we will purchase studies necessary to substantiate the diagnosis or to document the severity of your impairment, generally after we have evaluated the medical and other evidence we already have. We will not purchase studies involving exercise testing if there is significant risk involved or if there is another medical reason not to perform the test. We will follow sections 4.00C6, 4.00C7, 4.00C8, and 104.00B7 when we decide whether to purchase exercise testing. We will make a reasonable effort to obtain any additional studies from a qualified medical source in an office or center experienced in pediatric cardiac assessment. (See §416.919g.)

6. *What studies will we not purchase?* We will not purchase any studies involving cardiac catheterization, such as coronary angiography, arteriograms, or electrophysiological studies. However, if the results of catheterization are part of the existing evidence we have, we will consider them together with the other relevant evidence. See 4.00C15a in part A.

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

a. ETTs, though increasingly used, are still less frequently indicated in children than in adults, and can rarely be performed successfully by children under 6 years of age. An ETT may be of value in the assessment of some arrhythmias, in the assessment of the severity of chronic heart failure, and in the assessment of recovery of function following cardiac surgery or other treatment.

b. We will purchase an ETT in a childhood claim only if we cannot make a determination or decision based on the evidence we have and an MC, preferably one with experience in the care of children with cardiovascular impairments, has determined that an ETT is needed to evaluate your impairment. We will not purchase an ETT if you are less than 6 years of age. If we do purchase an ETT for a child age 12 or younger, it must be performed by a qualified medical source in a specialty center for pediatric cardiology or other facility qualified to perform exercise tests of children.

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

C. Evaluating Chronic Heart Failure

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

a. *CHF* is the inability of the heart to pump enough oxygenated blood to body tissues. This syndrome is characterized by symptoms and signs of pulmonary or systemic congestion (fluid retention) or limited cardiac output. Certain laboratory findings of cardiac functional and structural abnormality support the diagnosis of CHF.

b. CHF is considered in these listings as a single category whether due to atherosclerosis (narrowing of the arteries), cardiomyopathy, hypertension, or rheumatic, congenital, or other heart disease. However, if the CHF is the result of primary pulmonary hypertension secondary to disease of the lung (cor pulmonale), we will evaluate your impairment using 3.09 in the

respiratory system listings in part A.

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

a. Cardiomegaly or ventricular dysfunction must be present and demonstrated by appropriate medically acceptable imaging, such as chest x-ray, echocardiography (M-Mode, 2-dimensional, and Doppler), radionuclide studies, or cardiac catheterization.

(i) Cardiomegaly is present when:

(*A*) Left ventricular diastolic dimension or systolic dimension is greater than 2 standard deviations above the mean for the child's body surface area;

(*B*) Left ventricular mass is greater than 2 standard deviations above the mean for the child's body surface area; or

(*C*) Chest x-ray (6 foot PA film) is indicative of cardiomegaly if the cardiothoracic ratio is over 60 percent at 1 year of age or less, or 55 percent or greater at more than 1 year of age.

(ii) Ventricular dysfunction is present when indices of left ventricular function, such as fractional shortening or ejection fraction (the percentage of the blood in the ventricle actually pumped out with each contraction), are greater than 2 standard deviations below the mean for the child's age. (Fractional shortening, also called shortening fraction, reflects the left ventricular systolic function in the absence of segmental wall motion abnormalities and has a linear correlation with ejection fraction. In children, fractional shortening is more commonly used than ejection fraction.)

(iii) However, these measurements alone do not reflect your functional capacity, which we evaluate by considering all of the relevant evidence.

(iv) Other findings on appropriate medically acceptable imaging may include increased pulmonary vascular markings, pleural effusion, and pulmonary edema. These findings need not be present on each report, since CHF may be controlled by prescribed treatment.

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

(i) Symptoms of congestion or of limited cardiac output include easy fatigue, weakness, shortness of breath (dyspnea), cough, or chest discomfort at rest or with activity. Children with CHF may also experience shortness of breath on lying flat (orthopnea) or episodes of shortness of breath that wake them from sleep (paroxysmal nocturnal dyspnea). They may also experience cardiac arrhythmias resulting in palpitations, lightheadedness, or fainting. Fatigue or exercise intolerance in an infant may be manifested by prolonged feeding time, often associated with excessive respiratory effort and sweating.

(ii) During infancy, other manifestations of chronic heart failure may include failure to gain

weight or involuntary loss of weight and repeated lower respiratory tract infections.

(iii) Signs of congestion may include hepatomegaly, ascites, increased jugular venous distention or pressure, rales, peripheral edema, rapid shallow breathing (tachypnea), or rapid weight gain. However, these signs need not be found on all examinations because fluid retention may be controlled by prescribed treatment.

D. Evaluating Congenital Heart Disease

1. *What is congenital heart disease?* Congenital heart disease is any abnormality of the heart or the major blood vessels that is present at birth. Examples include:

a. *Abnormalities of cardiac septation,* including ventricular septal defect or atrioventricular canal;

b. *Abnormalities resulting in cyanotic heart disease,* including tetralogy of Fallot or transposition of the great arteries;

c. *Valvular defects or obstructions to ventricular outflow,* including pulmonary or aortic stenosis or coarctation of the aorta; and

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

2. *How will we evaluate symptomatic congenital heart disease?*

a. Because of improved treatment methods, more children with congenital heart disease are living longer. Although some types of congenital heart disease may be corrected by surgery, many children with treated congenital heart disease continue to have problems throughout their lives (symptomatic congenital heart disease). If you have congenital heart disease that results in chronic heart failure with evidence of ventricular dysfunction or in recurrent arrhythmias, we will evaluate your impairment under 104.02 or 104.05. Otherwise, we will evaluate your impairment under 104.06.

b. For 104.06A2, we will accept pulse oximetry measurements instead of arterial O_2, but the arterial O_2 values are preferred, if available.

c. For 104.06D, examples of impairments that in most instances will require life-saving surgery or a combination of surgery and other major interventional procedures (for example, multiple "balloon" catheter procedures) before age 1 include, but are not limited to, the following:

(i) Hypoplastic left heart syndrome,

(ii) Critical aortic stenosis with neonatal heart failure,

(iii) Critical coarctation of the aorta, with or without associated anomalies,

(iv) Complete atrioventricular canal defects,

(v) Transposition of the great arteries,

(vi) Tetralogy of Fallot,

(vii) Pulmonary atresia with intact ventricular septum,

(viii) Single ventricle,

(ix) Tricuspid atresia, and

(x) Multiple ventricular septal defects.

E. Evaluating Arrhythmias

1. *What is an arrhythmia?* An *arrhythmia* is a change in the regular beat of the heart. Your heart may seem to skip a beat or beat irregularly, very quickly (tachycardia), or very slowly (bradycardia).

2. *What are the different types of arrhythmias?*

a. There are many types of arrhythmias. Arrhythmias are identified by where they occur in the heart (atria or ventricles) and by what happens to the heart's rhythm when they occur.

b. Arrhythmias arising in the cardiac atria (upper chambers of the heart) are called atrial or supraventricular arrhythmias. Ventricular arrhythmias begin in the ventricles (lower chambers). In general, ventricular arrhythmias caused by heart disease are the most serious.

3. *How do we evaluate arrhythmias using 104.05?*

a. We will use 104.05 when you have arrhythmias that are not fully controlled by medication, an implanted pacemaker, or an implanted cardiac defibrillator and you have uncontrolled recurrent episodes of syncope or near syncope. If your arrhythmias are controlled, we will evaluate your underlying heart disease using the appropriate listing. For other considerations when we evaluate arrhythmias in the presence of an implanted cardiac defibrillator, see 104.00E4.

b. We consider *near syncope* to be a period of altered consciousness, since syncope is a loss of consciousness or a faint. It is not merely a feeling of light-headedness, momentary weakness, or dizziness.

c. For purposes of 104.05, there must be a documented association between the syncope or near syncope and the recurrent arrhythmia. The recurrent arrhythmia, not some other cardiac or non-cardiac disorder, must be established as the cause of the associated symptom. This documentation of the association between the symptoms and the arrhythmia may come from the usual diagnostic methods, including Holter monitoring (also called ambulatory electrocardiography) and tilt-table testing with a concurrent ECG. Although an arrhythmia may be a coincidental finding on an ETT, we will not purchase an ETT to document the presence of a cardiac arrhythmia.

4. *What will we consider when you have an implanted cardiac defibrillator and you do not have*

arrhythmias that meet the requirements of 104.05?

a. Implanted cardiac defibrillators are used to prevent sudden cardiac death in children who have had, or are at high risk for, cardiac arrest from life-threatening ventricular arrhythmias. The largest group of children at risk for sudden cardiac death consists of children with cardiomyopathy (ischemic or non-ischemic) and reduced ventricular function. However, life-threatening ventricular arrhythmias can also occur in children with little or no ventricular dysfunction. The shock from the implanted cardiac defibrillator is a unique form of treatment; it rescues a child from what may have been cardiac arrest. However, as a consequence of the shock(s), children may experience psychological distress, which we may evaluate under the mental disorders listings in 112.00ff.

b. Most implantable cardiac defibrillators have rhythm-correcting and pacemaker capabilities. In some children, these functions may result in the termination of ventricular arrhythmias without an otherwise painful shock. (The shock is like being kicked in the chest.) Implanted cardiac defibrillators may deliver inappropriate shocks, often repeatedly, in response to benign arrhythmias or electrical malfunction. Also, exposure to strong electrical or magnetic fields, such as from MRI (magnetic resonance imaging), can trigger or reprogram an implanted cardiac defibrillator, resulting in inappropriate shocks. We must consider the frequency of, and the reason(s) for, the shocks when evaluating the severity and duration of your impairment.

c. In general, the exercise limitations imposed on children with an implanted cardiac defibrillator are those dictated by the underlying heart impairment. However, the exercise limitations may be greater when the implanted cardiac defibrillator delivers an inappropriate shock in response to the increase in heart rate with exercise, or when there is exercise-induced ventricular arrhythmia.

F. Evaluating Other Cardiovascular Impairments

1. *What is ischemic heart disease (IHD) and how will we evaluate it in children? IHD* results when one or more of your coronary arteries is narrowed or obstructed or, in rare situations, constricted due to vasospasm, interfering with the normal flow of blood to your heart muscle (ischemia). The obstruction may be the result of an embolus, a thrombus, or plaque. When heart muscle tissue dies as a result of the reduced blood supply, it is called a myocardial infarction (heart attack). Ischemia is rare in children, but when it occurs, its effects on children are the same as on adults. If you have IHD, we will evaluate it under 4.00E and 4.04 in part A.

2. *How will we evaluate hypertension?* Because *hypertension* (high blood pressure) generally causes disability through its effects on other body systems, we will evaluate it by reference to the specific body system(s) affected (heart, brain, kidneys, or eyes) when we consider its effects under the listings. We will also consider any limitations imposed by your hypertension when we consider whether you have an impairment that functionally equals the listings.

3. *What is cardiomyopathy and how will we evaluate it? Cardiomyopathy* is a disease of the heart muscle. The heart loses its ability to pump blood (heart failure), and in some instances, heart rhythm is disturbed, leading to irregular heartbeats (arrhythmias). Usually, the exact cause of the muscle damage is never found (idiopathic cardiomyopathy). There are various types of cardiomyopathy, which fall into two major categories: *Ischemic* and *nonischemic* cardiomyopathy. Ischemic cardiomyopathy typically refers to heart muscle damage that results from coronary artery disease, including heart attacks. Nonischemic cardiomyopathy includes

several types: Dilated, hypertrophic, and restrictive. We will evaluate cardiomyopathy under 4.04 in part A, 104.02, 104.05, or 111.06, depending on its effects on you.

4. *How will we evaluate valvular heart disease?* We will evaluate valvular heart disease under the listing appropriate for its effect on you. Thus, we may use 4.04 in part A, 104.02, 104.05, 104.06, or an appropriate neurological listing in 111.00ff.

5. *What do we consider when we evaluate heart transplant recipients?*

a. After your heart transplant, we will consider you disabled for 1 year following the surgery because there is a greater likelihood of rejection of the organ and infection during the first year.

b. However, heart transplant patients generally meet our definition of disability before they undergo transplantation. We will determine the onset of your disability based on the facts in your case.

c. We will not assume that you became disabled when your name was placed on a transplant waiting list. This is because you may be placed on a waiting list soon after diagnosis of the cardiac disorder that may eventually require a transplant. Physicians recognize that candidates for transplantation often have to wait months or even years before a suitable donor heart is found, so they place their patients on the list as soon as permitted.

d. When we do a continuing disability review to determine whether you are still disabled, we will evaluate your residual impairment(s), as shown by symptoms, signs, and laboratory findings, including any side effects of medication. We will consider any remaining symptoms, signs, and laboratory findings indicative of cardiac dysfunction in deciding whether medical improvement (as defined in §416.994a) has occurred.

6. *How will we evaluate chronic rheumatic fever or rheumatic heart disease?* The diagnosis should be made in accordance with the current revised Jones criteria for guidance in the diagnosis of rheumatic fever. We will evaluate persistence of rheumatic fever activity under 104.13. If you have evidence of chronic heart failure or recurrent arrhythmias associated with rheumatic heart disease, we will use 104.02 or 104.05.

7. *What is hyperlipidemia and how will we evaluate it? Hyperlipidemia* is the general term for an elevation of any or all of the lipids (fats or cholesterol) in the blood; for example, hypertriglyceridemia, hypercholesterolemia, and hyperlipoproteinemia. These disorders of lipoprotein metabolism and transport can cause defects throughout the body. The effects most likely to interfere with function are those produced by atherosclerosis (narrowing of the arteries) and coronary artery disease. We will evaluate your lipoprotein disorder by considering its effects on you.

8. *How will we evaluate Kawasaki disease?* We will evaluate Kawasaki disease under the listing appropriate to its effects on you, which may include major coronary artery aneurysm or heart failure. A major coronary artery aneurysm may cause ischemia or arrhythmia, which we will evaluate under 4.04 in part A or 104.05. We will evaluate chronic heart failure under 104.02.

9. *What is lymphedema and how will we evaluate it?*

a. *Lymphedema* is edema of the extremities due to a disorder of the lymphatic circulation; at its worst, it is called elephantiasis. Primary lymphedema is caused by abnormal development of lymph vessels and may be present at birth (congenital lymphedema), but more often develops during the teens (lymphedema praecox). Secondary lymphedema is due to obstruction or destruction of normal lymphatic channels due to tumor, surgery, repeated infections, or parasitic infection such as filariasis. Lymphedema most commonly affects one extremity.

b. Lymphedema does not meet the requirements of 4.11 in part A, although it may medically equal the severity of that listing. We will evaluate lymphedema by considering whether the underlying cause meets or medically equals any listing or whether the lymphedema medically equals a cardiovascular listing, such as 4.11, or a musculoskeletal listing, such as 101.02A or 101.03. If no listing is met or medically equaled, we will evaluate any functional limitations imposed by your lymphedema when we consider whether you have an impairment that functionally equals the listings.

10. *What is Marfan syndrome and how will we evaluate it?*

a. Marfan syndrome is a genetic connective tissue disorder that affects multiple body systems, including the skeleton, eyes, heart, blood vessels, nervous system, skin, and lungs. There is no specific laboratory test to diagnose Marfan syndrome. The diagnosis is generally made by medical history, including family history, physical examination, including an evaluation of the ratio of arm/leg size to trunk size, a slit lamp eye examination, and a heart test(s), such as an echocardiogram. In some cases, a genetic analysis may be useful, but such analyses may not provide any additional helpful information.

b. The effects of Marfan syndrome can range from mild to severe. In most cases, the disorder progresses as you age. Most individuals with Marfan syndrome have abnormalities associated with the heart and blood vessels. Your heart's mitral valve may leak, causing a heart murmur. Small leaks may not cause symptoms, but larger ones may cause shortness of breath, fatigue, and palpitations. Another effect is that the wall of the aorta may be weakened and stretch (aortic dilation). This aortic dilation may tear, dissect, or rupture, causing serious heart problems or sometimes sudden death. We will evaluate the manifestations of your Marfan syndrome under the appropriate body system criteria, such as 4.10 in part A, or if necessary consider the functional limitations imposed by your impairment.

G. Other Evaluation Issues

1. *What effect does obesity have on the cardiovascular system and how will we evaluate it?* Obesity is a medically determinable impairment that is often associated with disorders of the cardiovascular system. Disturbance of this system can be a major cause of disability in children with obesity. Obesity may affect the cardiovascular system because of the increased workload the additional body mass places on the heart. Obesity may make it harder for the chest and lungs to expand. This can mean that the respiratory system must work harder to provide needed oxygen. This in turn would make the heart work harder to pump blood to carry oxygen to the body. Because the body would be working harder at rest, its ability to perform additional work would be less than would otherwise be expected. Thus, the combined effects of obesity with cardiovascular impairments can be greater than the effects of each of the impairments considered separately. We must consider any additional and cumulative effects of obesity when we determine whether you have a severe cardiovascular impairment or a listing-level cardiovascular impairment (or a combination of impairments that medically equals a

listing), and when we determine whether your impairment(s) functionally equals the listings.

2. *How do we relate treatment to functional status?* In general, conclusions about the severity of a cardiovascular impairment cannot be made on the basis of type of treatment rendered or anticipated. The amount of function restored and the time required for improvement after treatment (medical, surgical, or a prescribed program of progressive physical activity) vary with the nature and extent of the disorder, the type of treatment, and other factors. Depending upon the timing of this treatment in relation to the alleged onset date of disability, we may need to defer evaluation of the impairment for a period of up to 3 months from the date treatment began to permit consideration of treatment effects, unless we can make a determination or decision using the evidence we have. See 104.00B4.

3. *How do we evaluate impairments that do not meet one of the cardiovascular listings?*

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

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

104.01 Category of Impairments, Cardiovascular System

104.02. *Chronic heart failure* while on a regimen of prescribed treatment, with symptoms and signs described in 104.00C2, and with one of the following:

A. Persistent tachycardia at rest (see Table I);

OR

B. Persistent tachypnea at rest (see Table II) or markedly decreased exercise tolerance (see 104.00C2b);

OR

C. Growth disturbance with:

1. An involuntary weight loss or failure to gain weight at an appropriate rate for age, resulting in a fall of 15 percentiles from an established growth curve (on current NCHS/CDC growth chart) which is currently present (see 104.00A3f) and has persisted for 2 months or longer; or

2. An involuntary weight loss or failure to gain weight at an appropriate rate for age, resulting in a fall to below the third percentile from an established growth curve (on current NCHS/CDC growth chart) which is currently present (see 104.00A3f) and has persisted for 2 months or longer.

Table I—Tachycardia at rest

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

Table II—Tachypnea at rest

Age	Respiratory rate over (per minute)
Under 1 yr	40
1 through 5 yrs	35
6 through 9 yrs	30
Over 9 yrs	25

104.05 *Recurrent arrhythmias,* not related to reversible causes such as electrolyte abnormalities or digitalis glycoside or antiarrhythmic drug toxicity, resulting in uncontrolled (see 104.00A3g), recurrent (see 104.00A3c) episodes of cardiac syncope or near syncope (see 104.00E3b), despite prescribed treatment (see 104.00B3 if there is no prescribed treatment), and documented by resting or ambulatory (Holter) electrocardiography, or by other appropriate medically acceptable testing, coincident with the occurrence of syncope or near syncope (see 104.00E3c).

104.06 *Congenital heart disease,* documented by appropriate medically acceptable imaging (see 104.00A3d) or cardiac catheterization, with one of the following:

A. Cyanotic heart disease, with persistent, chronic hypoxemia as manifested by:

1. Hematocrit of 55 percent or greater on two evaluations 3 months or more apart within a consecutive 12-month period (see 104.00A3e); or

2. Arterial O_2 saturation of less than 90 percent in room air, or resting arterial PO_2 of 60 Torr or less; or

3. Hypercyanotic spells, syncope, characteristic squatting, or other incapacitating symptoms directly related to documented cyanotic heart disease; or

4. Exercise intolerance with increased hypoxemia on exertion.

OR

B. Secondary pulmonary vascular obstructive disease with pulmonary arterial systolic pressure elevated to at least 70 percent of the systemic arterial systolic pressure.

OR

C. Symptomatic acyanotic heart disease, with ventricular dysfunction interfering very seriously with the ability to independently initiate, sustain, or complete activities.

OR

D. For infants under 12 months of age at the time of filing, with life-threatening congenital heart impairment that will require or already has required surgical treatment in the first year of life, and the impairment is expected to be disabling (because of residual impairment following surgery, or the recovery time required, or both) until the attainment of at least 1 year of age, consider the infant to be under disability until the attainment of at least age 1; thereafter, evaluate impairment severity with reference to the appropriate listing.

104.09 *Heart transplant.* Consider under a disability for 1 year following surgery; thereafter, evaluate residual impairment under the appropriate listing.

104.13 *Rheumatic heart disease,* with persistence of rheumatic fever activity manifested by significant murmurs(s), cardiac enlargement or ventricular dysfunction (see 104.00C2a), and other associated abnormal laboratory findings; for example, an elevated sedimentation rate or ECG findings, for 6 months or more in a consecutive 12-month period (see 104.00A3e). Consider under a disability for 18 months from the established onset of impairment, then evaluate any residual impairment(s).

105.00 Digestive System

A. *Disorders of the digestive system* which result in disability usually do so because of interference with nutrition and growth, multiple recurrent inflammatory lesions, or other complications of the disease. Such lesions or complications usually respond to treatment. To constitute a listed impairment, these must be shown to have persisted or be expected to persist despite prescribed therapy for a continuous period of at least 12 months.

B. *Documentation of gastrointestinal impairments* should include pertinent operative findings, appropriate medically acceptable imaging studies, endoscopy, and biopsy reports. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment.

C. *Growth retardation and malnutrition.* When the primary disorder of the digestive tract has been documented, evaluate resultant malnutrition under the criteria described in 105.08. Evaluate resultant growth impairment under the criteria described in 100.03. Intestinal disorders, including surgical diversions and potentially correctable congenital lesions, do not represent a severe impairment if the individual is able to maintain adequate nutrition growth and development.

D. *Multiple congenital anomalies.* See related criteria, and consider as a combination of impairments.

105.01 Category of Impairments, Digestive.

105.03 *Esophageal obstruction, caused by atresia, stricture, or stenosis* with malnutrition as

described under the criteria in 105.08.

105.05 *Chronic liver disease.* With one of the following:

A. Inoperable biliary atresia demonstrated by appropriate medically acceptable imaging or surgery; or

B. Intractable ascites not attributable to other causes, with serum albumin of 3.0 gm./100 ml. or less; or

C. Esophageal varices (demonstrated by endoscopy or other appropriate medically acceptable imaging); or

D. Hepatic coma, documentated by findings from hospital records; or

E. Hepatic encephalopathy. Evaluate under the criteria in 112.02; or

F. Chronic active inflammation or necrosis documented by SGOT persistently more than 100 units or serum bilirubin of 2.5 mg. percent or greater.

105.07 *Chronic inflammatory bowel disease (such as ulcerative colitis, regional enteritis), as documented in 105.00.* With one of the following:

A. Intestinal manifestations or complications, such as obstruction, abscess, or fistula formation which has lasted or is expected to last 12 months; or

B. Malnutrition as described under the criteria in 105.08; or

C. Growth impairment as described under the criteria in 100.03.

105.08 *Malnutrition, due to demonstrable gastrointestinal disease causing either a fall of 15 percentiles of weight which persists or the persistence of weight which is less than the third percentile (on standard growth charts).* And one of the following:

A. Stool fat excretion per 24 hours:

1. More than 15 percent in infants less than 6 months.

2. More than 10 percent in infants 6-18 months.

3. More than 6 percent in children more than 18 months; or

B. Persistent hematocrit of 30 percent or less despite prescribed therapy; or

C. Serum carotene of 40 mcg./100 ml. or less; or

D. Serum albumin of 3.0 gm./100 ml. or less.

105.09 *Liver transplant.* Consider under a disability for 12 months following the date of surgery;

thereafter, evaluate the residual impairment.

106.00 Genitourinary Impairments

A. What impairments do these listings cover?

1. We use these listings to evaluate genitourinary impairments resulting from chronic renal disease and congenital genitourinary disorders.

2. We use the criteria in 106.02 to evaluate renal dysfunction due to any chronic renal disease, such as chronic glomerulonephritis, hypertensive renal vascular disease, diabetic nephropathy, chronic obstructive uropathy, and hereditary nephropathies.

3. We use the criteria in 106.06 to evaluate nephrotic syndrome due to glomerular disease.

4. We use the criteria in 106.07 to evaluate congenital genitourinary impairments such as ectopic ureter, extrophic urinary bladder, urethral valves, and neurogenic bladder.

B. What do we mean by the following terms in these listings?

1. *Anasarca* is generalized massive edema (swelling).

2. *Creatinine* is a normal product of muscle metabolism.

3. *Creatinine clearance test* is a test for renal function based on the rate at which creatinine is excreted by the kidney.

4. *Glomerular disease* can be classified into two broad categories, nephrotic and nephritic. Nephrotic conditions are associated with increased urinary protein excretion and nephritic conditions are associated with inflammation of the internal structures of the kidneys.

5. *Hemodialysis,* or *dialysis,* is the removal of toxic metabolic byproducts from the blood by diffusion in an artificial kidney machine.

6. *Nephrotic syndrome* is a general name for a group of diseases involving defective kidney glomeruli, characterized by heavy proteinuria, hypoalbuminemia, hyperlipidemia, and varying degrees of edema.

7. *Neuropathy* is a problem in peripheral nerve function (that is, in any part of the nervous system except the brain and spinal cord) that causes pain, numbness, tingling, and muscle weakness in various parts of the body.

8. *Parenteral antibiotics* refer to the administration of antibiotics by intravenous, intramuscular, or subcutaneous injection.

9. *Peritoneal dialysis* is a method of hemodialysis in which the dialyzing solution is introduced into and removed from the peritoneal cavity either continuously or intermittently.

10. *Proteinuria* is excess protein in the urine.

11. *Renal* means pertaining to the kidney.

12. *Serum albumin* is a major plasma protein that is responsible for much of the plasma colloidal osmotic pressure and serves as a transport protein.

13. *Serum creatinine* is the amount of creatinine in the blood and is measured to evaluate kidney function.

C. What evidence do we need?

1. We need a longitudinal record of your medical history that includes records of treatment, response to treatment, hospitalizations, and laboratory evidence of renal disease that indicates its progressive nature or of congenital genitourinary impairments that documents their recurrent or episodic nature. The laboratory or clinical evidence will indicate deterioration of renal function, such as elevation of serum creatinine, or changes in genitourinary function, such as episodes of electrolyte disturbance.

2. We generally need a longitudinal clinical record covering a period of at least 3 months of observations and treatment, unless we can make a fully favorable determination or decision without it. The record should include laboratory findings, such as serum creatinine or serum albumin values, obtained on more than one examination over the 3-month period.

3. When you are undergoing dialysis, we should have laboratory findings showing your renal function before you started dialysis.

4. The medical evidence establishing the clinical diagnosis of nephrotic syndrome must include a description of the extent of edema, including pretibial, periorbital, or presacral edema. The medical evidence should describe any ascites, pleural effusion, or pericardial effusion. Levels of serum albumin and proteinuria must be included.

5. If a renal biopsy has been performed, the evidence should include a copy of the report of the microscopic examination of the specimen. However, if we do not have a copy of the microscopic examination in the evidence, we can accept a statement from an acceptable medical source that a biopsy was performed, with a description of the results.

6. The medical evidence documenting congenital genitourinary impairments should include treating physician records, operative reports, and hospital records. It should describe the frequency of your episodes, prescribed treatment, laboratory findings, and any surgical procedures performed.

D. How do we consider the effects of treatment?

We consider factors such as the:

1. Type of therapy.

2. Response to therapy.

3. Side effects of therapy.

4. Effects of any post-therapeutic residuals.

5. Expected duration of treatment.

E. What other things do we consider when we evaluate your genitourinary impairment under specific listings?

1. *Chronic hemodialysis or peritoneal dialysis* (106.02A). A report from an acceptable medical source describing the chronic renal disease and the need for ongoing dialysis is sufficient to satisfy the requirements in 106.02A.

2. *Kidney transplantation* (106.02B). If you have undergone kidney transplantation, we will consider you to be disabled for 12 months following the surgery because, during the first year, there is a greater likelihood of rejection of the organ and recurrent infection. After the first year posttransplantation, we will base our continuing disability evaluation on your residual impairment(s). We will include absence of symptoms, signs, and laboratory findings indicative of kidney dysfunction in our consideration of whether medical improvement (as defined in §§404.1594(b)(1) and (c)(1) and 416.994a, as appropriate) has occurred. We will consider the:

a. Occurrence of rejection episodes.

b. Side effects of immunosuppressants, including corticosteroids.

c. Frequency of any renal infections.

d. Presence of systemic complications such as other infections, neuropathy, or deterioration of other organ systems.

3. *Nephrotic syndrome* (106.06). The longitudinal clinical record should include a description of prescribed therapy, response to therapy, and any side effects of therapy. In order for your nephrotic syndrome to meet 106.06A or B, the medical evidence must document that you have the appropriate laboratory findings required by these listings and that your anasarca has persisted for at least 3 months despite prescribed therapy. However, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in your case record. We may also evaluate complications of your nephrotic syndrome, such as orthostatic hypotension, recurrent infections, or venous thromboses, under the appropriate listing for the resultant impairment.

4. *Congenital genitourinary impairments* (106.07).

a. Each of the listings in 106.07 requires a longitudinal clinical record showing that at least three events have occurred within a consecutive 12-month period with intervening periods of improvement. *Events* include urologic surgical procedures, hospitalizations, and treatment with parenteral antibiotics. To meet the requirements of these listings, there must be at least 1 month (that is, 30 days) between the events in order to ensure that we are evaluating separate episodes.

b. Diagnostic cystoscopy does not satisfy the requirement for repeated urologic surgical procedures in 106.07A.

c. In 106.07B, *systemic infection* means an infection requiring an initial course of parenterally administered antibiotics occurring at least once every 4 months or at least 3 times a year.

d. In 106.07C, appropriate laboratory and clinical evidence document electrolyte disturbance. Hospitalizations are inpatient hospitalizations for 24 hours or more.

F. What does the term "persistent" mean in these listings?

Persistent means that the longitudinal clinical record shows that, with few exceptions, the required finding(s) has been at, or is expected to be at, the level specified in the listing for a continuous period of at least 12 months.

G. How do we evaluate impairments that do not meet one of the genitourinary listings?

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

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing, or, in the case of a claim for SSI payments, functionally equals the listings. (See §§404.1526, 416.926, and 416.926a.) When we decide whether a child receiving SSI payments continues to be disabled, we use the rules in §416.994a.

106.01 Category of Impairments, Genitourinary Impairments

106.02 *Impairment of renal function,* due to any chronic renal disease that has lasted or can be expected to last for a continuous period of at least 12 months. With:

A. *Chronic hemodialysis or peritoneal dialysis* (see 106.00E1).

or

B. *Kidney transplantation.* Consider under a disability for 12 months following surgery; thereafter, evaluate the residual impairment (see 106.00E2).

or

C. *Persistent elevation of serum creatinine* to 3 mg per deciliter (dL) (100 ml) or greater, over at least 3 months.

or

D. *Reduction of creatinine clearance* to 30 ml per minute (43 liters/24 hours) per 1.73 m2 of body surface area over at least 3 months.

106.06 *Nephrotic syndrome,* with anasarca, persisting for at least 3 months despite prescribed therapy. (See 106.00E3.) With:

A. Serum albumin of 2.0 g/dL (100 ml) or less.

or

B. Proteinuria of 40 mg/m2/hr or greater.

106.07 *Congenital genitourinary impairments* (see 106.00E4) resulting in one of the following:

A. Repeated urologic surgical procedures, occurring at least 3 times in a consecutive 12-month period.

or

B. Documented episodes of systemic infection requiring an initial course of parenteral antibiotics, occurring at least 3 times in a consecutive 12-month period (see 106.00E4).

or

C. Hospitalization (see 106.00E4d) for episodes of electrolyte disturbance, occurring at least 3 times in a consecutive 12-month period.

107.00 Hematological Disorders

A. *Sickle cell disease.* Refers to a chronic hemolytic anemia associated with sickle cell hemoglobin, either homozygous or in combination with thalassemia or with another abnormal hemoglobin (such as C or F).

Appropriate hematologic evidence for sickle cell disease, such as hemoglobin electrophoresis must be included. Vaso-occlusive, hemolytic, or aplastic episodes should be documented by description of severity, frequency, and duration.

Disability due to sickle cell disease may be solely the result of a severe, persistent anemia or may be due to the combination of chronic progressive or episodic manifestations in the presence of a less severe anemia.

Major visceral episodes causing disability include meningitis, osteomyelitis, pulmonary infections or infarctions, cerebrovascular accidents, congestive heart failure, genitourinary involvement, etc.

B. *Coagulation defects.* Chronic inherited coagulation disorders must be documented by appropriate laboratory evidence such as abnormal thromboplastin generation, coagulation time, or factor assay.

107.01 Category of Impairments, Hemic and Lymphatic.

107.03 *Hemolytic anemia (due to any cause).* Manifested by persistence of hematocrit of 26 percent or less despite prescribed therapy, and reticulocyte count of 4 percent or greater.

107.05 *Sickle cell disease.* With:

A. Recent, recurrent, severe vaso-occlusive crises (musculoskeletal, vertebral, abdominal); or

B. A major visceral complication in the 12 months prior to application; or

C. A hyperhemolytic or aplastic crisis within 12 months prior to application; or

D. Chronic, severe anemia with persistence of hematocrit of 26 percent or less; or

E. Congestive heart failure, cerebrovascular damage, or emotional disorder as described under the criteria in 104.02, 111.00ff, or 112.00ff.

107.06 *Chronic idiopathic thrombocytopenic purpura of childhood* with purpura and thrombocytopenia of 40,000 platelets/cu. mm. or less despite prescribed therapy or recurrent upon withdrawal of treatment.

107.08 *Inherited coagulation disorder.* With:

A. Repeated spontaneous or inappropriate bleeding; or

B. Hemarthrosis with joint deformity.

108.00 Skin Disorders

A. *What skin disorders do we evaluate with these listings?* We use these listings to evaluate skin disorders that may result from hereditary, congenital, or acquired pathological processes. The kinds of impairments covered by these listings are: Ichthyosis, bullous diseases, chronic infections of the skin or mucous membranes, dermatitis, hidradenitis suppurativa, genetic photosensitivity disorders, and burns.

B. *What documentation do we need?* When we evaluate the existence and severity of your skin disorder, we generally need information about the onset, duration, frequency of flareups, and prognosis of your skin disorder; the location, size, and appearance of lesions; and, when applicable, history of exposure to toxins, allergens, or irritants, familial incidence, seasonal variation, stress factors, and your ability to function outside of a highly protective environment. To confirm the diagnosis, we may need laboratory findings (for example, results of a biopsy obtained independently of Social Security disability evaluation or blood tests) or evidence from other medically acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

C. *How do we assess the severity of your skin disorders(s)?* We generally base our assessment of severity on the extent of your skin lesions, the frequency of flareups of your skin lesions, how your symptoms (including pain) limit you, the extent of your treatment, and how your treatment affects you.

1. *Extensive skin lesions.* Extensive skin lesions are those that involve multiple body sites or critical body areas, and result in a very serious limitation. Examples of extensive skin lesions that result in a very serious limitation include but are not limited to:

a. Skin lesions that interfere with the motion of your joints and that very seriously limit your use of more than one extremity; that is, two upper extremities, two lower extremities, or one upper

and one lower extremity.

b. Skin lesions on the palms of both hands that very seriously limit your ability to do fine and gross motor movements.

c. Skin lesions on the soles of both feet, the perineum, or both inguinal areas that very seriously limit your ability to ambulate.

2. *Frequency of flareups.* If you have skin lesions, but they do not meet the requirements of any of the listings in this body system, you may still have an impairment that results in marked and severe functional limitations when we consider your condition over time, especially if your flareups result in extensive skin lesions, as defined in C1 of this section. Therefore, if you have frequent flareups, we may find that your impairment(s) is medically equal to one of these listings even though you have some periods during which your condition is in remission. We will consider how frequent and serious your flareups are, how quickly they resolve, and how you function between flareups to determine whether you have marked and severe functional limitations that have lasted for a continuous period of at least 12 months or that can be expected to last for a continuous period of at least 12 months. We will also consider the frequency of your flareups when we determine whether you have a severe impairment and when we need to assess functional equivalence.

3. *Symptoms (including pain).* Symptoms (including pain) may be important factors contributing to the severity of your skin disorder(s). We assess the impact of symptoms as explained in §§404.1528, 404.1529, 416.928, and 416.929 of this chapter.

4. *Treatment.* We assess the effects of medication, therapy, surgery, and any other form of treatment you receive when we determine the severity and duration of your impairment(s). Skin disorders frequently respond to treatment; however, response to treatment can vary widely, with some impairments becoming resistant to treatment. Some treatments can have side effects that can in themselves result in limitations.

a. We assess the effects of continuing treatment as prescribed by determining if there is improvement in the symptoms, signs, and laboratory findings of your disorder, and if you experience side effects that result in functional limitations. To assess the effects of your treatment, we may need information about:

i. The treatment you have been prescribed (for example, the type, dosage, method and frequency of administration of medication or therapy);

ii. Your response to the treatment;

iii. Any adverse effects of the treatment; and

iv. The expected duration of the treatment.

b. Because treatment itself or the effects of treatment may be temporary, in most cases sufficient time must elapse to allow us to evaluate the impact and expected duration of treatment and its side effects. Except under 108.07 and 108.08, you must follow continuing treatment as prescribed for at least 3 months before your impairment can be determined to meet the requirements of a skin disorder listing. (See 108.00H if you are not undergoing

treatment or did not have treatment for 3 months.) We consider your specific response to treatment when we evaluate the overall severity of your impairment.

D. *How do we assess impairments that may affect the skin and other body systems?* When your impairment affects your skin and has effects in other body systems, we first evaluate the predominant feature of your impairment under the appropriate body system. Examples include, but are not limited to the following.

1. *Tuberous sclerosis* primarily affects the brain. The predominant features are seizures, which we evaluate under the neurological listings in 111.00, and developmental delays or other mental disorders, which we evaluate under the mental disorders listings in 112.00.

2. *Malignant tumors of the skin* (for example, malignant melanoma) are cancers, or neoplastic diseases, which we evaluate under the listings in 113.00.

3. *Connective tissue disorders and other immune system disorders* (for example, systemic lupus erythematosus, scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than one body system. We first evaluate these disorders under the immune system listings in 114.00. We evaluate lupus erythematosus under 114.02, scleroderma under 114.04, symptomatic HIV infection under 114.08, and Sjögren's syndrome under 114.03, 114.09, or any other appropriate listing in section 114.00.

4. *Disfigurement or deformity* resulting from skin lesions may result in loss of sight, hearing, speech, and the ability to chew (mastication). We evaluate these impairments and their effects under the special senses and speech listings in 102.00 and the digestive system listings in 105.00. Facial disfigurement or other physical deformities may also have effects we evaluate under the mental disorders listings in 112.00, such as when they affect mood or social functioning.

5. We evaluate *erythropoietic porphyrias* under the hemic and lymphatic listings in 107.00.

6. We evaluate *hemangiomas associated with thrombocytopenia and hemorrhage* (for example, Kasabach-Merritt syndrome) involving coagulation defects, under the hemic and lymphatic listings in 107.00. But, when hemangiomas impinge on vital structures or interfere with function, we evaluate their primary effects under the appropriate body system.

E. *How do we evaluate genetic photosensitivity disorders?*

1. *Xeroderma pigmentosum (XP).* When you have XP, your impairment meets the requirements of 108.07A if you have clinical and laboratory findings showing that you have the disorder. (See 108.00E3.) People who have XP have a lifelong hypersensitivity to all forms of ultraviolet light and generally lead extremely restricted lives in highly protective environments in order to prevent skin cancers from developing. Some people with XP also experience problems with their eyes, neurological problems, mental disorders, and problems in other body systems.

2. *Other genetic photosensitivity disorders.* Other genetic photosensitivity disorders may vary in their effects on different people, and may not result in marked and severe functional limitations for a continuous period of at least 12 months. Therefore, if you have a genetic photosensitivity disorder other than XP (established by clinical and laboratory findings as

described in 108.00E3), you must show that you have either extensive skin lesions or an inability to function outside of a highly protective environment to meet the requirements of 108.07B. You must also show that your impairment meets the duration requirement. By *inability to function outside of a highly protective environment* we mean that you must avoid exposure to ultraviolet light (including sunlight passing through windows and light from unshielded fluorescent bulbs), wear protective clothing and eyeglasses, and use opaque broad-spectrum sunscreens in order to avoid skin cancer or other serious effects. Some genetic photosensitivity disorders can have very serious effects in other body systems, especially special senses and speech (102.00), neurological (111.00), mental (112.00), and neoplastic (113.00). We will evaluate the predominant feature of your impairment under the appropriate body system, as explained in 108.00D.

3. *Clinical and laboratory findings*.

a. *General*. We need documentation from an acceptable medical source, as defined in §§404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment. In general, we must have evidence of appropriate laboratory testing showing that you have XP or another genetic photosensitivity disorder. We will find that you have XP or another genetic photosensitivity disorder based on a report from an acceptable medical source indicating that you have the impairment, supported by definitive genetic laboratory studies documenting appropriate chromosomal changes, including abnormal DNA repair or another DNA or genetic abnormality specific to your type of photosensitivity disorder.

b. *What we will accept as medical evidence instead of the actual laboratory report*. When we do not have the actual laboratory report, we need evidence from an acceptable medical source that includes appropriate clinical findings for your impairment and that is persuasive that a positive diagnosis has been confirmed by appropriate laboratory testing at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive genetic laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

F. *How do we evaluate burns?* Electrical, chemical, or thermal burns frequently affect other body systems; for example, musculoskeletal, special senses and speech, respiratory, cardiovascular, renal, neurological, or mental. Consequently, we evaluate burns the way we evaluate other disorders that can affect the skin and other body systems, using the listing for the predominant feature of your impairment. For example, if your soft tissue injuries are under continuing surgical management (as defined in 101.00M), we will evaluate your impairment under 101.08. However, if your burns do not meet the requirements of 101.08 and you have extensive skin lesions that result in a very serious limitation (as defined in 108.00C1) that has lasted or can be expected to last for a continuous period of at least 12 months, we will evaluate them under 108.08.

G. *How do we determine if your skin disorder(s) will continue at a disabling level of severity in order to meet the duration requirement?* For all of these skin disorder listings except 108.07 and 108.08, we will find that your impairment meets the duration requirement if your skin disorder results in extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. By *persist*, we mean that the longitudinal clinical record shows that, with few exceptions, your lesions have been at the level of severity specified in the listing. For 108.07A, we will presume that you meet the duration requirement. For 108.07B and 108.08, we will consider all of the relevant medical and other information in your case record to

determine whether your skin disorder meets the duration requirement.

H. *How do we assess your skin disorder(s) if your impairment does not meet the requirements of one of these listings?*

1. These listings are only examples of common skin disorders that we consider severe enough to result in marked and severe functional limitations. For most of these listings, if you do not have continuing treatment as prescribed, if your treatment has not lasted for at least 3 months, or if you do not have extensive skin lesions that have persisted for at least 3 months, your impairment cannot meet the requirements of these skin disorder listings. (This provision does not apply to 108.07 and 108.08.) However, we may still find that you are disabled because your impairment(s) meets the requirements of a listing in another body system, medically equals (see §§404.1526 and 416.926 of this chapter) the severity of a listing, or functionally equals the severity of the listings.

2. If you have not received ongoing treatment or do not have an ongoing relationship with the medical community despite the existence of a severe impairment(s), or if your skin lesions have not persisted for at least 3 months but you are undergoing continuing treatment as prescribed, you may still have an impairment(s) that meets a listing in another body system or that medically equals a listing. If you do not have an impairment(s) that meets or medically equals a listing, we will consider whether your impairment(s) functionally equals the listings. (See §416.924 of this chapter.) When we decide whether you continue to be disabled, we use the rules in §416.994a of this chapter.

108.01 Category of Impairments, Skin Disorders

108.02 *Ichthyosis*, with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.03 *Bullous disease* (for example, pemphigus, erythema multiforme bullosum, epidermolysis bullosa, bullous pemphigoid, dermatitis herpetiformis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.04 *Chronic infections of the skin or mucous membranes*, with extensive fungating or extensive ulcerating skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.05 *Dermatitis* (for example, psoriasis, dyshidrosis, atopic dermatitis, exfoliative dermatitis, allergic contact dermatitis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

108.06 *Hidradenitis suppurativa*, with extensive skin lesions involving both axillae, both inguinal areas, or the perineum that persist for at least 3 months despite continuing treatment as prescribed.

108.07 *Genetic photosensitivity disorders,* established as described in 108.00E.

A. Xeroderma pigmentosum. Consider the individual disabled from birth.

B. Other genetic photosensitivity disorders, with:

1. Extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months, or

2. Inability to function outside of a highly protective environment for a continuous period of at least 12 months (see 108.00E2).

108.08 *Burns*, with extensive skin lesions that have lasted or can be expected to last for a continuous period of at least 12 months. (See 108.00F).

109.00 Endocrine System

A. *Cause of disability.* Disability is caused by a disturbance in the regulation of the secretion or metabolism of one or more hormones which are not adequately controlled by therapy. Such disturbances or abnormalities usually respond to treatment. To constitute a listed impairment these must be shown to have persisted or be expected to persist despite prescribed therapy for a continuous period of at least 12 months.

B. *Growth.* Normal growth is usually a sensitive indicator of health as well as of adequate therapy in children. Impairment of growth may be disabling in itself or may be an indicator of a severe disorder involving the endocrine system or other body systems. Where involvement of other organ systems has occurred as a result of a primary endocrine disorder, these impairments should be evaluated according to the criteria under the appropriate sections.

C. *Documentation.* Description of characteristic history, physical findings, and diagnostic laboratory data must be included. Results of laboratory tests will be considered abnormal if outside the normal range or greater than two standard deviations from the mean of the testing laboratory. Reports in the file should contain the information provided by the testing laboratory as to their normal values for that test.

D. *Hyperfunction of the adrenal cortex.* Evidence of growth retardation must be documented as described in 100.00. Elevated blood or urinary free cortisol levels are not acceptable in lieu of urinary 17-hydroxycorticosteroid excretion for the diagnosis of adrenal cortical hyperfunction.

E. *Adrenal cortical insufficiency.* Documentation must include persistent low plasma cortisol or low urinary 17-hydroxycorticosteroids or 17-ketogenic steroids and evidence of unresponsiveness to ACTH stimulation.

109.01 Category of Impairments, Endrocrine

109.02 *Thyroid Disorders.*

A. Hyperthyroidism (as documented in 109.00C). With clinical manifestations despite prescribed therapy, and one of the following:

1. Elevated serum thyroxine (T_4) and either elevated free T_4 or resin T_3 uptake; or

2. Elevated thyroid uptake of radioiodine; or

3. Elevated serum triiodothyronine (T_3).

B. *Hypothyroidism.* With one of the following, despite prescribed therapy:

1. IQ of 70 or less; or

2. Growth impairment as described under the criteria in 100.02 A and B; or

3. Precocious puberty.

109.03 *Hyperparathyroidism (as documented in 109.00C).* With:

A. Repeated elevated total or ionized serum calcium; or

B. Elevated serum parathyroid hormone.

109.04 *Hypoparathyroidism or Pseudohypoparathyroidism.* With:

A. Severe recurrent tetany or convulsions which are unresponsive to prescribed therapy; or

B. Growth retardation as described under criteria in 100.02 A and B.

109.05 *Diabetes insipidus, documented by pathologic hypertonic saline or water deprivation test.* And one of the following:

A. Intracranial space-occupying lesion, before or after surgery; or

B. Unresponsiveness to Pitressin; or

C. Growth retardation as described under the criteria in 100.02 A and B; or

D. Unresponsive hypothalmic thirst center, with chronic or recurrent hypernatremia; or

E. Decreased visual fields attributable to a pituitary lesion.

109.06 *Hyperfunction of the adrenal cortex (Primary or secondary).* With:

A. Elevated urinary 17-hyroxycortico-steroids (or 17-ketogenic steroids) as documented in 109.00 C and D; and

B. Unresponsiveness to low-dose dexamethasone suppression.

109.07 *Adrenal cortical insufficiency (as documented in 109.00 C and E)* with recent, recurrent episodes of circulatory collapse.

109.08 *Juvenile diabetes mellitus (as documented in 109.00C) requiring parenteral insulin.* And one of the following, despite prescribed therapy:

A. Recent, recurrent hospitalizations with acidosis; or

B. Recent, recurrent episodes of hypoglycemia; or

C. Growth retardation as described under the criteria in 100.02 A or B; or

D. Impaired renal function as described under the criteria in 106.00ff.

109.09 *Iatrogenic hypercorticoid state.*

With chronic glucocorticoid therapy resulting in one of the following:

A. Osteoporosis; or

B. Growth retardation as described under the criteria in 100.02 A or B; or

C. Diabetes mellitus as described under the criteria in 109.08; or

D. Myopathy as described under the criteria in 111.06; or

E. Emotional disorder as described under the criteria in 112.00ff.

109.10 *Pituitary dwarfism (with documented growth hormone deficiency).* And growth impairment as described under the criteria in 100.02B.

109.11 *Adrenogenital syndrome.* With:

A. Recent, recurrent self-losing episodes despite prescribed therapy; or

B. Inadequate replacement therapy manifested by accelerated bone age and virilization, or

C. Growth impairment as described under the criteria in 100.02 A or B.

109.12 *Hypoglycemia (as documented in 109.00C).* With recent, recurrent hypoglycemic episodes producing convulsion or coma.

109.13 *Gonadal Dysgenesis (Turner's Syndrome), chromosomally proven.* Evaluate the resulting impairment under the criteria for the appropriate body system.

110.00 Impairments That Affect Multiple Body Systems

A. What Kinds of Impairments Do We Evaluate Under This Body System?

1. *General*. We use these listings when you have a single impairment that affects two or more body systems. Under these listings, we evaluate impairments that affect multiple body systems due to non-mosaic Down syndrome or a catastrophic congenital abnormality or disease. These kinds of impairments generally produce long-term, if not lifelong, interference with age-appropriate activities. Some of them result in early death or interfere very seriously with development. We use the term "very seriously" in these listings to describe an "extreme" limitation of functioning as defined in §416.926a(e)(3).

2. *What is Down syndrome?* Down syndrome is a condition in which there are three copies of chromosome 21 within the cells of the body instead of the normal two copies per cell. The

three copies may be separate (trisomy), or one chromosome 21 copy may be attached to a different chromosome (translocation). This extra chromosomal material changes the orderly development of the body and brain. Down syndrome is characterized by a complex of physical characteristics, delayed physical development, and mental retardation. Down syndrome exists in non-mosaic and mosaic forms.

3. *What is non-mosaic Down syndrome?*

a. Non-mosaic Down syndrome occurs when you have an extra copy of chromosome 21 in every cell of your body. At least 98 percent of people with Down syndrome have this form (which includes either trisomy or translocation type chromosomal abnormalities). Virtually all cases of non-mosaic Down syndrome affect the mental, neurological, and skeletal systems, and they are often accompanied by heart disease, impaired vision, hearing problems, and other conditions.

b. We evaluate children with confirmed non-mosaic Down syndrome under 110.06. If you have confirmed non-mosaic Down syndrome, we consider you disabled from birth.

4. *What is mosaic Down syndrome?*

a. Mosaic Down syndrome occurs when you have some cells with the normal two copies of chromosome 21 and some cells with an extra copy of chromosome 21. When this occurs, there is a mixture of two types of cells. Mosaic Down syndrome occurs in only 1-2 percent of people with Down syndrome, and there is a wide range in the level of severity of the impairment. Mosaic Down syndrome can be profound and disabling, but it can also be so slight as to be undetected clinically.

b. We evaluate children with confirmed mosaic Down syndrome under the listing criteria in any affected body system(s) on an individual case basis, as described in 110.00C.

5. *What are catastrophic congenital abnormalities or diseases?*

a. Catastrophic congenital abnormalities or diseases are present at birth, although they may not be apparent immediately. They cause deviation from, or interruption of, the normal function of the body and are reasonably certain to result in early death or to interfere very seriously with development.

b. We evaluate catastrophic congenital abnormalities or diseases under 110.08.

B. What Documentation Do We Need To Establish That You Have an Impairment That Affects Multiple Body Systems?

1. *General*. We need documentation from an acceptable medical source, as defined in §§404.1513(a) and 416.913(a), to establish that you have a medically determinable impairment. In general, the documentation should include a clinical description of the diagnostic physical features associated with your multiple body system impairment, and any appropriate laboratory tests.

2. *Non-mosaic Down syndrome (110.06)*.

a. *Definitive chromosomal analysis*. We will find that you have non-mosaic Down syndrome based on a report from an acceptable medical source that indicates that you have the impairment and that includes the actual laboratory report of definitive chromosomal analysis showing that you have the impairment. *Definitive chromosomal analysis* for Down syndrome means karyotype analysis. When we have the laboratory report of the actual karyotype analysis, we do not additionally require a clinical description of the physical features of Down syndrome.

b. *What if you have Down syndrome and we do not have the results of definitive chromosomal analysis?* When you have Down syndrome and we do not have the actual laboratory report of definitive chromosomal analysis, we need evidence from an acceptable medical source that includes a clinical description of the diagnostic physical features of your impairment, and that is persuasive that a positive diagnosis has been confirmed by definitive chromosomal analysis at some time prior to our evaluation. To be persuasive, the report must state that definitive chromosomal analysis was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record; for example, evidence showing your limitations in adaptive functioning or signs of a mental disorder that can be associated with non-mosaic Down syndrome, your educational history, or the results of psychological testing.

3. *Catastrophic congenital abnormalities or diseases (110.08)*.

a. *Genetic disorders*. For genetic multiple body system impairments (other than non-mosaic Down syndrome), such as Trisomy 13 (Patau Syndrome or Trisomy D), Trisomy 18 (Edwards' Syndrome or Trisomy E), chromosomal deletion syndromes (for example, deletion 5p syndrome, also called cri du chat syndrome), or inborn metabolic disorders (for example, Tay-Sachs disease), we need evidence from an acceptable medical source that includes a clinical description of the diagnostic physical features of your impairment, and the report of the definitive laboratory study (for example, genetic analysis or evidence of biochemical abnormalities) that is diagnostic of your impairment. When we do not have the actual laboratory report, we need evidence from an acceptable medical source that is persuasive that a positive diagnosis was confirmed by appropriate laboratory analysis at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

b. *Other disorders*. For infants born with other kinds of catastrophic congenital abnormalities (for example, anencephaly, cyclopia), we need evidence from an acceptable medical source that includes a clinical description of the diagnostic physical features of the impairment.

C. How Do We Evaluate Impairments That Affect Multiple Body Systems and That Do Not Meet the Criteria of the Listings in This Body System?

1. These listings are examples of impairments that commonly affect multiple body systems and that we consider significant enough to result in marked and severe functional limitations. If your severe impairment(s) does not meet the criteria of any of these listings, we must also consider whether your impairment(s) meets the criteria of a listing in another body system.

2. There are many other impairments that can cause deviation from, or interruption of, the normal function of the body or interfere with development; for example, congenital anomalies,

chromosomal disorders, dysmorphic syndromes, metabolic disorders, and perinatal infectious diseases. In these impairments, the degree of deviation or interruption may vary widely from child to child. Therefore, the resulting functional limitations and the progression of those limitations are more variable than with the catastrophic congenital abnormalities and diseases we include in these listings. For this reason, we evaluate the specific effects of these impairments on you under the listing criteria in any affected body system(s) on an individual case basis. Examples of such impairments include, but are not limited to, triple X syndrome (XXX syndrome), fragile X syndrome, phenylketonuria (PKU), caudal regression syndrome, and fetal alcohol syndrome.

3. If you have a severe medically determinable impairment(s) that does not meet a listing, we will consider whether your impairment(s) medically equals a listing. If your impairment(s) does not meet or medically equal a listing, we will consider whether it functionally equals the listings. (See §§404.1526, 416.926, and 416.926a.) When we decide whether you continue to be disabled, we use the rules in §416.994a.

110.01 Category of Impairments, Impairments That Affect Multiple Body Systems

110.06 *Non-mosaic Down syndrome,* established as described in 110.00B.

110.08 *A catastrophic congenital abnormality or disease*, established as described in 110.00B, and:

A. Death usually is expected within the first months of life, and the rare individuals who survive longer are profoundly impaired (for example, anencephaly, trisomy 13 or 18, cyclopia);

or

B. That interferes very seriously with development; for example, cri du chat syndrome (deletion 5p syndrome) or Tay-Sachs disease (acute infantile form).

111.00 Neurological

A. *Convulsive epilepsy* must be substantiated by at least one detailed description of a typical seizure. Report of recent documentation should include a neurological examination with frequency of episodes and any associated phenomena substantiated.

Young children may have convulsions in association with febrile illnesses. Proper use of 111.02 and 111.03 requires that epilepsy be established. Although this does not exclude consideration of seizures occurring during febrile illnesses, it does require documentation of seizures during nonfebrile periods.

There is an expected delay in control of epilepsy when treatment is started, particularly when changes in the treatment regimen are necessary. Therefore, an epileptic disorder should not be considered to meet the requirements of 111.02 or 111.03 unless it is shown that convulsive episodes have persisted more than three months after prescribed therapy began.

B. *Nonconvulsive epilepsy.* Classical petit mal seizures must be documented by characteristic EEG pattern, plus information as to age at onset and frequency of clinical seizures.

C. *Motor dysfunction.* As described in 111.06, motor dysfunction may be due to any neurological disorder. It may be due to static or progressive conditions involving any area of the nervous system and producing any type of neurological impairment. This may include weakness, spasticity, lack of coordination, ataxia, tremor, athetosis, or sensory loss. Documentation of motor dysfunction must include neurologic findings and description of type of neurologic abnormality (e.g., spasticity, weakness), as well as a description of the child's functional impairment (*i.e.*, what the child is unable to do because of the abnormality). Where a diagnosis has been made, evidence should be included for substantiation of the diagnosis (e.g., blood chemistries and muscle biopsy reports), wherever applicable.

D. *Impairment of communication.* The documentation should include a description of a recent comprehensive evaluation, including all areas of affective and effective communication, performed by a qualified professional.

E. *Brain tumors.* We evaluate malignant brain tumors under the criteria in 113.13. For benign brain tumors, we determine the severity and duration of the impairment on the basis of symptoms, signs, and laboratory findings (111.05).

111.01 Category of Impairment, Neurological

111.02 *Major motor seizure disorder.*

A. *Convulsive epilepsy.* In a child with an established diagnosis of epilepsy, the occurrence of more than one major motor seizure per month despite at least three months of prescribed treatment. With:

1. Daytime episodes (loss of consciousness and convulsive seizures); or

2. Nocturnal episodes manifesting residuals which interfere with activity during the day.

B. *Convulsive epilepsy syndrome.* In a child with an established diagnosis of epilepsy, the occurrence of at least one major motor seizure in the year prior to application despite at least three months of prescribed treatment. And one of the following:

1. IQ of 70 or less; or

2. Significant interference with communication due to speech, hearing, or visual defect; or

3. Significant mental disorder; or

4. Where significant adverse effects of medication interfere with major daily activities.

111.03 *Nonconvulsive epilepsy.* In a child with an established seizure disorder, the occurrence of more than one minor motor seizure per week, with alteration of awareness or loss of consciousness, despite at least three months of prescribed treatment.

111.05 *Benign brain tumors.* Evaluate under 111.02, 111.03, 111.06, 111.09 or the criteria of the affected body system.

111.06 *Motor dysfunction (due to any neurological disorder).* Persistent disorganization or

deficit of motor function for age involving two extremities, which (despite prescribed therapy) interferes with age-appropriate major daily activities and results in disruption of:

A. Fine and gross movements; or

B. Gait and station.

111.07 *Cerebral Palsy.* With:

A. Motor dysfunction meeting the requirements of 101.02 or 111.06; or

B. Less severe motor dysfunction (but more than slight) and one of the following:

1. IQ of 70 or less; or

2. Seizure disorder, with at least one major motor seizure in the year prior to application; or

3. Significant interference with communication due to speech, hearing or visual defect; or

4. Significant emotional disorder.

111.08 *Meningomyelocele (and related disorders).* With one of the following despite prescribed treatment:

A. Motor dysfunction meeting the requirements of 101.02 or 111.06; or

B. Less severe motor dysfunction (but more than slight), and:

1. Urinary or fecal incontinence when inappropriate for age; or

2. IQ of 70 or less; or

C. Four extremity involvement; or

D. Noncompensated hydrocephalus producing interference with mental or motor developmental progression.

111.09 *Communication impairment, associated with documented neurological disorder.* And one of the following:

A. Documented speech deficit which significantly affects the clarity and content of the speech; or

B. Documented comprehension deficit resulting in ineffective verbal communication for age; or

C. Impairment of hearing as described under the criteria in 102.08.

112.00 Mental Disorders

A. *Introduction:* The structure of the mental disorders listings for children under age 18 parallels the structure for the mental disorders listings for adults but is modified to reflect the presentation of mental disorders in children. The listings for mental disorders in children are arranged in 11 diagnostic categories: Organic mental disorders (112.02); schizophrenic, delusional (paranoid), schizoaffective, and other psychotic disorders (112.03); mood disorders (112.04); mental retardation (112.05); anxiety disorders (112.06); somatoform, eating, and tic disorders (112.07); personality disorders (112.08); psychoactive substance dependence disorders (112.09); autistic disorder and other pervasive developmental disorders (112.10); attention deficit hyperactivity disorder (112.11); and developmental and emotional disorders of newborn and younger infants (112.12).

There are significant differences between the listings for adults and the listings for children. There are disorders found in children that have no real analogy in adults; hence, the differences in the diagnostic categories for children. The presentation of mental disorders in children, particularly the very young child, may be subtle and of a character different from the signs and symptoms found in adults. For example, findings such as separation anxiety, failure to mold or bond with the parents, or withdrawal may serve as findings comparable to findings that mark mental disorders in adults. The activities appropriate to children, such as learning, growing, playing, maturing, and school adjustment, are also different from the activities appropriate to the adult and vary widely in the different childhood stages.

Each listing begins with an introductory statement that describes the disorder or disorders addressed by the listing. This is followed (except in listings 112.05 and 112.12) by paragraph A criteria (a set of medical findings) and paragraph B criteria (a set of impairment-related functional limitations). An individual will be found to have a listed impairment when the criteria of both paragraphs A and B of the listed impairment are satisfied.

The purpose of the criteria in paragraph A is to substantiate medically the presence of a particular mental disorder. Specific symptoms and signs under any of the listings 112.02 through 112.12 cannot be considered in isolation from the description of the mental disorder contained at the beginning of each listing category. Impairments should be analyzed or reviewed under the mental category(ies) indicated by the medical findings.

Paragraph A of the listings is a composite of medical findings which are used to substantiate the existence of a disorder and may or may not be appropriate for children at specific developmental stages. However, a range of medical findings is included in the listings so that no age group is excluded. For example, in listing 112.02A7, emotional lability and crying would be inappropriate criteria to apply to older infants and toddlers, age 1 to attainment of age 3; whereas in 112.02A1, developmental arrest, delay, or regression are appropriate criteria for older infants and toddlers. Whenever the adjudicator decides that the requirements of paragraph A of a particular mental listing are satisfied, then that listing should be applied regardless of the age of the child to be evaluated.

The purpose of the paragraph B criteria is to describe impairment-related functional limitations which are applicable to children. Standardized tests of social or cognitive function and adaptive behavior are frequently available and appropriate for the evaluation of children and, thus, such tests are included in the paragraph B functional parameters. The functional restrictions in paragraph B must be the result of the mental disorder which is manifested by the medical findings in paragraph A.

We did not include separate C criteria for listings 112.02, 112.03, 112.04, and 112.06, as are found in the adult listings, because for the most part we do not believe that the residual disease processes described by these listings are commonly found in children. However, in unusual cases where these disorders are found in children and are comparable to the severity and duration found in adults, we may use the adult listings 12.02C, 12.03C, 12.04C, and 12.06C criteria to evaluate such cases.

The structure of the listings for Mental Retardation (112.05) and Developmental and Emotional Disorders of Newborn and Younger Infants (112.12) is different from that of the other mental disorders. Listing 112.05 (Mental Retardation) contains six sets of criteria. If an impairment satisfies the diagnostic description in the introductory paragraph and any one of the six sets of criteria, we will find that the child's impairment meets the listing. For listings 112.05D and 112.05F, we will assess the degree of functional limitation the additional impairment(s) imposes to determine if it causes more than minimal functional limitations, *i.e.,* is a "severe" impairment(s), as defined in §416.924(c). If the additional impairment(s) does not cause limitations that are "severe" as defined in §416.924(c), we will not find that the additional impairment(s) imposes an additional and significant limitation of function. Listing 112.12 (Developmental and Emotional Disorders of Newborn and Younger Infants) contains five criteria, any one of which, if satisfied, will result in a finding that the infant's impairment meets the listing.

It must be remembered that these listings are only examples of common mental disorders that are severe enough to find a child disabled. When a child has a medically determinable impairment that is not listed, an impairment that does not meet the requirements of a listing, or a combination of impairments no one of which meets the requirements of a listing, we will make a determination whether the child's impairment(s) medically or functionally equals the listings. (See §§404.1526, 416.926, and 416.926a.) This determination can be especially important in older infants and toddlers (age 1 to attainment of age 3), who may be too young for identification of a specific diagnosis, yet demonstrate serious functional limitations. Therefore, the determination of equivalency is necessary to the evaluation of any child's case when the child does not have an impairment that meets a listing.

B. *Need for Medical Evidence:* The existence of a medically determinable impairment of the required duration must be established by medical evidence consisting of symptoms, signs, and laboratory findings (including psychological or developmental test findings). Symptoms are complaints presented by the child. Psychiatric signs are medically demonstrable phenomena that indicate specific psychological abnormalities, e.g., abnormalities of behavior, mood, thought, memory, orientation, development, or perception, as described by an appropriate medical source. Symptoms and signs generally cluster together to constitute recognizable mental disorders described in paragraph A of the listings. These findings may be intermittent or continuous depending on the nature of the disorder.

C. *Assessment of Severity:* In childhood cases, as with adults, severity is measured according to the functional limitations imposed by the medically determinable mental impairment. However, the range of functions used to assess impairment severity for children varies at different stages of maturation. The functional areas that we consider are: Motor function; cognitive/communicative function; social function; personal function; and concentration, persistence, or pace. In most functional areas, there are two alternative methods of documenting the required level of severity: (1) Use of standardized tests alone, where appropriate test instruments are available, and (2) use of other medical findings. (See 112.00D

for explanation of these documentation requirements.) The use of standardized tests is the preferred method of documentation if such tests are available.

Newborn and younger infants (birth to attainment of age 1) have not developed sufficient personality differentiation to permit formulation of appropriate diagnoses. We have, therefore, assigned listing 112.12 for Developmental and Emotional Disorders of Newborn and Younger Infants for the evaluation of mental disorders of such children. Severity of these disorders is based on measures of development in motor, cognitive/communicative, and social functions. When older infants and toddlers (age 1 to attainment of age 3) do not clearly satisfy the paragraph A criteria of any listing because of insufficient developmental differentiation, they must be evaluated under the rules for equivalency. The principles for assessing the severity of impairment in such children, described in the following paragraphs, must be employed.

Generally, when we assess the degree of developmental delay imposed by a mental impairment, we will use an infant's or toddler's chronological age; *i.e.,* the child's age based on birth date. If the infant or toddler was born prematurely, however, we will follow the rules in §416.924b(b) to determine whether we should use the infant's or toddler's corrected chronological age; *i.e.*, the chronological age adjusted by the period of gestational prematurity.

In defining the severity of functional limitations, two different sets of paragraph B criteria corresponding to two separate age groupings have been established, in addition to listing 112.12, which is for children who have not attained age 1. These age groups are: older infants and toddlers (age 1 to attainment of age 3) and children (age 3 to attainment of age 18). However, the discussion below in 112.00C1, 2, 3, and 4, on the age-appropriate areas of function, is broken down into four age groupings: older infants and toddlers (age 1 to attainment of age 3), preschool children (age 3 to attainment of age 6), primary school children (age 6 to attainment of age 12), and adolescents (age 12 to attainment of age 18). This was done to provide specific guidance on the age group variances in disease manifestations and methods of evaluation.

Where "marked" is used as a standard for measuring the degree of limitation it means more than moderate but less than extreme. A marked limitation may arise when several activities or functions are impaired, or even when only one is impaired, as long as the degree of limitation is such as to interfere seriously with the ability to function (based upon age-appropriate expectations) independently, appropriately, effectively, and on a sustained basis. When standardized tests are used as the measure of functional parameters, a valid score that is two standard deviations below the norm for the test will be considered a marked restriction.

1. *Older infants and toddlers (age 1 to attainment of age 3).* In this age group, impairment severity is assessed in three areas: (a) Motor development, (b) cognitive/communicative function, and (c) social function.

a. *Motor development.* Much of what we can discern about mental function in these children frequently comes from observation of the degree of development of fine and gross motor function. Developmental delay, as measured by a good developmental milestone history confirmed by medical examination, is critical. This information will ordinarily be available in the existing medical evidence from the claimant's treating sources and other medical sources, supplemented by information from nonmedical sources, such as parents, who have observed the child and can provide pertinent historical information. It may also be available from standardized testing. If the delay is such that the older infant or toddler has not achieved motor

development generally acquired by children no more than one-half the child's chronological age, the criteria are satisfied.

b. *Cognitive/communicative function.* Cognitive/communicative function is measured using one of several standardized infant scales. Appropriate tests for the measure of such function are discussed in 112.00D. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases, the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

For older infants and toddlers, alternative criteria covering disruption in communication as measured by their capacity to use simple verbal and nonverbal structures to communicate basic needs are provided.

c. *Social function.* Social function in older infants and toddlers is measured in terms of the development of relatedness to people (e.g., bonding and stranger anxiety) and attachment to animate or inanimate objects. Criteria are provided that use standard social maturity scales or alternative criteria that describe marked impairment in socialization.

2. *Preschool children (age 3 to attainment of age 6).* For the age groups including preschool children through adolescence, the functional areas used to measure severity are: (a) Cognitive/communicative function, (b) social function, (c) personal function, and (d) deficiencies of concentration, persistence, or pace resulting in frequent failure to complete tasks in a timely manner. After 36 months, motor function is no longer felt to be a primary determinant of mental function, although, of course, any motor abnormalities should be documented and evaluated.

a. *Cognitive/communicative function.* In the preschool years and beyond, cognitive function can be measured by standardized tests of intelligence, although the appropriate instrument may vary with age. A primary criterion for limited cognitive function is a valid verbal, performance, or full scale IQ of 70 or less. The listings also provide alternative criteria, consisting of tests of language development or bizarre speech patterns.

b. *Social function.* Social functioning refers to a child's capacity to form and maintain relationships with parents, other adults, and peers. Social functioning includes the ability to get along with others (e.g., family members, neighborhood friends, classmates, teachers). Impaired social functioning may be caused by inappropriate externalized actions (e.g., running away, physical aggression—but not self-injurious actions, which are evaluated in the personal area of functioning), or inappropriate internalized actions (e.g., social isolation, avoidance of interpersonal activities, mutism). Its severity must be documented in terms of intensity, frequency, and duration, and shown to be beyond what might be reasonably expected for age. Strength in social functioning may be documented by such things as the child's ability to respond to and initiate social interaction with others, to sustain relationships, and to participate in group activities. Cooperative behaviors, consideration for others, awareness of others' feelings, and social maturity, appropriate to a child's age, also need to be considered. Social functioning in play and school may involve interactions with adults, including responding appropriately to persons in authority (e.g., teachers, coaches) or cooperative behaviors involving other children. Social functioning is observed not only at home but also in preschool programs.

c. *Personal function.* Personal functioning in preschool children pertains to self-care; *i.e.*, personal needs, health, and safety (feeding, dressing, toileting, bathing; maintaining personal hygiene, proper nutrition, sleep, health habits; adhering to medication or therapy regimens; following safety precautions). Development of self-care skills is measured in terms of the child's increasing ability to help himself/herself and to cooperate with others in taking care of these needs. Impaired ability in this area is manifested by failure to develop such skills, failure to use them, or self-injurious actions. This function may be documented by a standardized test of adaptive behavior or by a careful description of the full range of self-care activities. These activities are often observed not only at home but also in preschool programs.

d. *Concentration, persistence, or pace.* This function may be measured through observations of the child in the course of standardized testing and in the course of play.

3. *Primary school children (age 6 to attainment of age 12).* The measures of function here are similar to those for preschool-age children except that the test instruments may change and the capacity to function in the school setting is supplemental information. Standardized measures of academic achievement, e.g., Wide Range Achievement Test-Revised, Peabody Individual Achievement Test, etc., may be helpful in assessing cognitive impairment. Problems in social functioning, especially in the area of peer relationships, are often observed firsthand by teachers and school nurses. As described in 112.00D, *Documentation,* school records are an excellent source of information concerning function and standardized testing and should always be sought for school-age children.

As it applies to primary school children, the intent of the functional criterion described in paragraph B2d, *i.e.*, deficiencies of concentration, persistence, or pace resulting in failure to complete tasks in a timely manner, is to identify the child who cannot adequately function in primary school because of a mental impairment. Although grades and the need for special education placement are relevant factors which must be considered in reaching a decision under paragraph B2d, they are not conclusive. There is too much variability from school district to school district in the expected level of grading and in the criteria for special education placement to justify reliance solely on these factors.

4. *Adolescents (age 12 to attainment of age 18).* Functional criteria parallel to those for primary school children (cognitive/communicative; social; personal; and concentration, persistence, or pace) are the measures of severity for this age group. Testing instruments appropriate to adolescents should be used where indicated. Comparable findings of disruption of social function must consider the capacity to form appropriate, stable, and lasting relationships. If information is available about cooperative working relationships in school or at part-time or full-time work, or about the ability to work as a member of a group, it should be considered when assessing the child's social functioning. Markedly impoverished social contact, isolation, withdrawal, and inappropriate or bizarre behavior under the stress of socializing with others also constitute comparable findings. (Note that self-injurious actions are evaluated in the personal area of functioning.)

a. Personal functioning in adolescents pertains to self-care. It is measured in the same terms as for younger children, the focus, however, being on the adolescent's ability to take care of his or her own personal needs, health, and safety without assistance. Impaired ability in this area is manifested by failure to take care of these needs or by self-injurious actions. This function may be documented by a standardized test of adaptive behavior or by careful descriptions of the full range of self-care activities.

b. In adolescents, the intent of the functional criterion described in paragraph B2d is the same as in primary school children, However, other evidence of this functional impairment may also be available, such as from evidence of the child's performance in work or work-like settings.

D. *Documentation:* 1. The presence of a mental disorder in a child must be documented on the basis of reports from acceptable sources of medical evidence. See §§404.1513 and 416.913. Descriptions of functional limitations may be available from these sources, either in the form of standardized test results or in other medical findings supplied by the sources, or both. (Medical findings consist of symptoms, signs, and laboratory findings.) Whenever possible, a medical source's findings should reflect the medical source's consideration of information from parents or other concerned individuals who are aware of the child's activities of daily living, social functioning, and ability to adapt to different settings and expectations, as well as the medical source's findings and observations on examination, consistent with standard clinical practice. As necessary, information from nonmedical sources, such as parents, should also be used to supplement the record of the child's functioning to establish the consistency of the medical evidence and longitudinality of impairment severity.

2. For some newborn and younger infants, it may be very difficult to document the presence or severity of a mental disorder. Therefore, with the exception of some genetic diseases and catastrophic congenital anomalies, it may be necessary to defer making a disability decision until the child attains age 3 months of age in order to obtain adequate observation of behavior or affect. See, also, 110.00 of this part. This period could be extended in cases of premature infants depending on the degree of prematurity and the adequacy of documentation of their developmental and emotional status.

3. For infants and toddlers, programs of early intervention involving occupational, physical, and speech therapists, nurses, social workers, and special educators, are a rich source of data. They can provide the developmental milestone evaluations and records on the fine and gross motor functioning of these children. This information is valuable and can complement the medical examination by a physician or psychologist. A report of an interdisciplinary team that contains the evaluation and signature of an acceptable medical source is considered acceptable medical evidence rather than supplemental data.

4. In children with mental disorders, particularly those requiring special placement, school records are a rich source of data, and the required reevaluations at specified time periods can provide the longitudinal data needed to trace impairment progression over time.

5. In some cases where the treating sources lack expertise in dealing with mental disorders of children, it may be necessary to obtain evidence from a psychiatrist, psychologist, or pediatrician with experience and skill in the diagnosis and treatment of mental disorders as they appear in children. In these cases, however, every reasonable effort must be made to obtain the records of the treating sources, since these records will help establish a longitudinal picture that cannot be established through a single purchased examination.

6. Reference to a "standardized psychological test" indicates the use of a psychological test measure that has appropriate validity, reliability, and norms, and is individually administered by a qualified specialist. By "qualified," we mean the specialist must be currently licensed or certified in the State to administer, score, and interpret psychological tests and have the training and experience to perform the test.

7. Psychological tests are best considered as standardized sets of tasks or questions designed to elicit a range of responses. Psychological testing can also provide other useful data, such as the specialist's observations regarding the child's ability to sustain attention and concentration, relate appropriately to the specialist, and perform tasks independently (without prompts or reminders). Therefore, a report of test results should include both the objective data and any clinical observations.

8. The salient characteristics of a good test are: (1) Validity, *i.e.,* the test measures what it is supposed to measure; (2) reliability, *i.e.,* the consistency of results obtained over time with the same test and the same individual; (3) appropriate normative data, *i.e.,* individual test scores can be compared to test data from other individuals or groups of a similar nature, representative of that population; and (4) wide scope of measurement, *i.e.,* the test should measure a broad range of facets/aspects of the domain being assessed. In considering the validity of a test result, we should note and resolve any discrepancies between formal test results and the child's customary behavior and daily activities.

9. Identical IQ scores obtained from different tests do not always reflect a similar degree of intellectual functioning. The IQ scores in listing 112.05 reflect values from tests of general intelligence that have a mean of 100 and a standard deviation of 15, e.g., the Wechsler series. IQs obtained from standardized tests that deviate significantly from a mean of 100 and standard deviation of 15 require conversion to a percentile rank so that the actual degree of limitation reflected by the IQ scores can be determined. In cases where more than one IQ is customarily derived from the test administered, e.g., where verbal, performance, and full scale IQs are provided in the Wechsler series, the lowest of these is used in conjunction with listing 112.05.

10. IQ test results must also be sufficiently current for accurate assessment under 112.05. Generally, the results of IQ tests tend to stabilize by the age of 16. Therefore, IQ test results obtained at age 16 or older should be viewed as a valid indication of the child's current status, provided they are compatible with the child's current behavior. IQ test results obtained between ages 7 and 16 should be considered current for 4 years when the tested IQ is less than 40, and for 2 years when the IQ is 40 or above. IQ test results obtained before age 7 are current for 2 years if the tested IQ is less than 40 and 1 year if at 40 or above.

11. Standardized intelligence test results are essential to the adjudication of all cases of mental retardation that are not covered under the provisions of listings 112.05A, 112.05B, and 112.05F. Listings 112.05A, 112.05B, and 112.05F may be the bases for adjudicating cases where the results of standardized intelligence tests are unavailable, e.g., where the child's young age or condition precludes formal standardized testing.

12. In conjunction with clinical examinations, sources may report the results of screening tests, *i.e.,* tests used for gross determination of level of functioning. Screening instruments may be useful in uncovering potentially serious impairments, but often must be supplemented by other data. However, in some cases the results of screening tests may show such obvious abnormalities that further testing will clearly be unnecessary.

13. Where reference is made to developmental milestones, this is defined as the attainment of particular mental or motor skills at an age-appropriate level, *i.e.,* the skills achieved by an infant or toddler sequentially and within a given time period in the motor and manipulative areas, in general understanding and social behavior, in self-feeding, dressing, and toilet

training, and in language. This is sometimes expressed as a developmental quotient (DQ), the relation between developmental age and chronological age as determined by specific standardized measurements and observations. Such tests include, but are not limited to, the Cattell Infant Intelligence Scale, the Bayley Scales of Infant Development, and the Revised Stanford-Binet. Formal tests of the attainment of developmental milestones are generally used in the clinical setting for determination of the developmental status of infants and toddlers.

14. Formal psychological tests of cognitive functioning are generally in use for preschool children, for primary school children, and for adolescents except for those instances noted below.

15. Generally, it is preferable to use IQ measures that are wide in scope and include items that test both verbal and performance abilities. However, in special circumstances, such as the assessment of children with sensory, motor, or communication abnormalities, or those whose culture and background are not principally English-speaking, measures such as the Test of Nonverbal Intelligence, Third Edition (TONI-3), Leiter International Performance Scale-Revised (Leiter-R), or Peabody Picture Vocabulary Test—Third Edition (PPVT-III) may be used.

16. We may consider exceptions for formal standardized psychological testing when an individual qualified by training and experience to perform such an evaluation is not available, or in cases where appropriate standardized measures for the child's social, linguistic, and cultural background are not available. In these cases, the best indicator of severity is often the level of adaptive functioning and how the child performs activities of daily living and social functioning.

17. Comprehensive neuropsychological examinations may be used to establish the existence and extent of compromise of brain function, particularly in cases involving organic mental disorders. Normally these examinations include assessment of cerebral dominance, basic sensation and perception, motor speed and coordination, attention and concentration, visual-motor function, memory across verbal and visual modalities, receptive and expressive speech, higher-order linguistic operations, problem-solving, abstraction ability, and general intelligence. In addition, there should be a clinical interview geared toward evaluating pathological features known to occur frequently in neurological disease and trauma, e.g., emotional lability, abnormality of mood, impaired impulse control, passivity and apathy, or inappropriate social behavior. The specialist performing the examination may administer one of the commercially available comprehensive neuropsychological batteries, such as the Luria-Nebraska or Halstead-Reitan, or a battery of tests selected as relevant to the suspected brain dysfunction. The specialist performing the examination must be properly trained in this area of neuroscience.

E. *Effect of Hospitalization or Residential Placement:* As with adults, children with mental disorders may be placed in a variety of structured settings outside the home as part of their treatment. Such settings include, but are not limited to, psychiatric hospitals, developmental disabilities facilities, residential treatment centers and schools, community-based group homes, and workshop facilities. The reduced mental demands of such structured settings may attenuate overt symptomatology and superficially make the child's level of adaptive functioning appear better than it is. Therefore, the capacity of the child to function outside highly structured settings must be considered in evaluating impairment severity. This is done by determining the degree to which the child can function (based upon age-appropriate expectations) independently, appropriately, effectively, and on a sustained basis outside the highly structured setting.

On the other hand, there may be a variety of causes for placement of a child in a structured setting which may or may not be directly related to impairment severity and functional ability. Placement in a structured setting in and of itself does not equate with a finding of disability. The severity of the impairment must be compared with the requirements of the appropriate listing.

F. *Effects of Medication:* Attention must be given to the effect of medication on the child's signs, symptoms, and ability to function. While drugs used to modify psychological functions and mental states may control certain primary manifestations of a mental disorder, e.g., hallucinations, impaired attention, restlessness, or hyperactivity, such treatment may not affect all functional limitations imposed by the mental disorder. In cases where overt symptomatology is attenuated by the use of such drugs, particular attention must be focused on the functional limitations that may persist. These functional limitations must be considered in assessing impairment severity.

Psychotropic medicines used in the treatment of some mental illnesses may cause drowsiness, blunted affect, or other side effects involving other body systems. Such side effects must be considered in evaluating overall impairment severity.

112.01 Category of Impairments, Mental

112.02 *Organic Mental Disorders:* Abnormalities in perception, cognition, affect, or behavior associated with dysfunction of the brain. The history and physical examination or laboratory tests, including psychological or neuropsychological tests, demonstrate or support the presence of an organic factor judged to be etiologically related to the abnormal mental state and associated deficit or loss of specific cognitive abilities, or affective changes, or loss of previously acquired functional abilities.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence of at least one of the following:

1. Developmental arrest, delay or regression; or

2. Disorientation to time and place; or

3. Memory impairment, either short-term (inability to learn new information), intermediate, or long-term (inability to remember information that was known sometime in the past); or

4. Perceptual or thinking disturbance (e.g., hallucinations, delusions, illusions, or paranoid thinking); or

5. Disturbance in personality (e.g., apathy, hostility); or

6. Disturbance in mood (e.g., mania, depression); or

7. Emotional lability (e.g., sudden crying); or

8. Impairment of impulse control (e.g., disinhibited social behavior, explosive temper

outbursts); or

9. Impairment of cognitive function, as measured by clinically timely standardized psychological testing; or

10. Disturbance of concentration, attention, or judgment;

AND

B. Select the appropriate age group to evaluate the severity of the impairment:

1. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the following:

a. Gross or fine motor development at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings (see 112.00C); or

b. Cognitive/communicative function at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings of equivalent cognitive/communicative abnormality, such as the inability to use simple verbal or nonverbal behavior to communicate basic needs or concepts; or

c. Social function at a level generally acquired by children no more than one-half the child's chronological age, documented by:

(1) An appropriate standardized test; or

(2) Other medical findings of an equivalent abnormality of social functioning, exemplified by serious inability to achieve age-appropriate autonomy as manifested by excessive clinging or extreme separation anxiety; or

d. Attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in two or more areas covered by a., b., or c., as measured by an appropriate standardized test or other appropriate medical findings.

2. For children (age 3 to attainment of age 18), resulting in at least two of the following:

a. Marked impairment in age-appropriate cognitive/communicative function, documented by medical findings (including consideration of historical and other information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, the results of appropriate standardized psychological

tests, or for children under age 6, by appropriate tests of language and communication; or

b. Marked impairment in age-appropriate social functioning, documented by history and medical findings (including consideration of information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, the results of appropriate standardized tests; or

c. Marked impairment in age-appropriate personal functioning, documented by history and medical findings (including consideration of information from parents or other individuals who have knowledge of the child, when such information is needed and available) and including, if necessary, appropriate standardized tests; or

d. Marked difficulties in maintaining concentration, persistence, or pace.

112.03 *Schizophrenic, Delusional (Paranoid), Schizoaffective, and Other Psychotic Disorders:* Onset of psychotic features, characterized by a marked disturbance of thinking, feeling, and behavior, with deterioration from a previous level of functioning or failure to achieve the expected level of social functioning.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence, for at least 6 months, either continuous or intermittent, of one or more of the following:

1. Delusions or hallucinations; or

2. Catatonic, bizarre, or other grossly disorganized behavior; or

3. Incoherence, loosening of associations, illogical thinking, or poverty of content of speech; or

4. Flat, blunt, or inappropriate affect; or

5. Emotional withdrawal, apathy, or isolation;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.04 *Mood Disorders:* Characterized by a disturbance of mood (referring to a prolonged emotion that colors the whole psychic life, generally involving either depression or elation), accompanied by a full or partial manic or depressive syndrome.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented persistence, either continuous or intermittent, of one of the following:

1. Major depressive syndrome, characterized by at least five of the following, which must include either depressed or irritable mood or markedly diminished interest or pleasure:

a. Depressed or irritable mood; or

b. Markedly diminished interest or pleasure in almost all activities; or

c. Appetite or weight increase or decrease, or failure to make expected weight gains; or

d. Sleep disturbance; or

e. Psychomotor agitation or retardation; or

f. Fatigue or loss of energy; or

g. Feelings of worthlessness or guilt; or

h. Difficulty thinking or concentrating; or

i. Suicidal thoughts or acts; or

j. Hallucinations, delusions, or paranoid thinking;

OR

2. Manic syndrome, characterized by elevated, expansive, or irritable mood, and at least three of the following:

a. Increased activity or psychomotor agitation; or

b. Increased talkativeness or pressure of speech; or

c. Flight of ideas or subjectively experienced racing thoughts; or

d. Inflated self-esteem or grandiosity; or

e. Decreased need for sleep; or

f. Easy distractibility; or

g. Involvement in activities that have a high potential of painful consequences which are not recognized; or

h. Hallucinations, delusions, or paranoid thinking;

OR

3. Bipolar or cyclothymic syndrome with a history of episodic periods manifested by the full symptomatic picture of both manic and depressive syndromes (and currently or most recently

characterized by the full or partial symptomatic picture of either or both syndromes);

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.05 *Mental Retardation:* Characterized by significantly subaverage general intellectual functioning with deficits in adaptive functioning.

The required level of severity for this disorder is met when the requirements in A, B, C, D, E, or F are satisfied.

A. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02;

OR

B. Mental incapacity evidenced by dependence upon others for personal needs (grossly in excess of age-appropriate dependence) and inability to follow directions such that the use of standardized measures of intellectual functioning is precluded;

OR

C. A valid verbal, performance, or full scale IQ of 59 or less;

OR

D. A valid verbal, performance, or full scale IQ of 60 through 70 and a physical or other mental impairment imposing an additional and significant limitation of function;

OR

E. A valid verbal, performance, or full scale IQ of 60 through 70 and:

1. For older infants and toddlers (age 1 to attainment of age 3), resulting in attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in either paragraphs B1a or B1c of 112.02; or

2. For children (age 3 to attainment of age 18), resulting in at least one of paragraphs B2b or B2c or B2d of 112.02;

OR

F. Select the appropriate age group:

1. For older infants and toddlers (age 1 to attainment of age 3), resulting in attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in paragraph B1b of 112.02, and a physical or other mental impairment imposing an additional and significant limitation of function;

OR

2. For children (age 3 to attainment of age 18), resulting in the satisfaction of 112.02B2a, and a physical or other mental impairment imposing an additional and significant limitation of function.

112.06 *Anxiety Disorders:* In these disorders, anxiety is either the predominant disturbance or is experienced if the individual attempts to master symptoms, e.g., confronting the dreaded object or situation in a phobic disorder, attempting to go to school in a separation anxiety disorder, resisting the obsessions or compulsions in an obsessive compulsive disorder, or confronting strangers or peers in avoidant disorders.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of at least one of the following:

1. Excessive anxiety manifested when the child is separated, or separation is threatened, from a parent or parent surrogate; or

2. Excessive and persistent avoidance of strangers; or

3. Persistent unrealistic or excessive anxiety and worry (apprehensive expectation), accompanied by motor tension, autonomic hyperactivity, or vigilance and scanning; or

4. A persistent irrational fear of a specific object, activity, or situation which results in a compelling desire to avoid the dreaded object, activity, or situation; or

5. Recurrent severe panic attacks, manifested by a sudden unpredictable onset of intense apprehension, fear, or terror, often with a sense of impending doom, occurring on the average of at least once a week; or

6. Recurrent obsessions or compulsions which are a source of marked distress; or

7. Recurrent and intrusive recollections of a traumatic experience, including dreams, which are a source of marked distress;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.07 *Somatoform, Eating, and Tic Disorders:* Manifested by physical symptoms for which

there are no demonstrable organic findings or known physiologic mechanisms; or eating or tic disorders with physical manifestations.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of one of the following:

1. An unrealistic fear and perception of fatness despite being underweight, and persistent refusal to maintain a body weight which is greater than 85 percent of the average weight for height and age, as shown in the most recent edition of the *Nelson Textbook of Pediatrics,* Richard E. Behrman and Victor C. Vaughan, III, editors, Philadelphia: W. B. Saunders Company; or

2. Persistent and recurrent involuntary, repetitive, rapid, purposeless motor movements affecting multiple muscle groups with multiple vocal tics; or

3. Persistent nonorganic disturbance of one of the following:

a. Vision; or

b. Speech; or

c. Hearing; or

d. Use of a limb; or

e. Movement and its control (e.g., coordination disturbance, psychogenic seizures); or

f. Sensation (diminished or heightened); or

g. Digestion or elimination; or

4. Preoccupation with a belief that one has a serious disease or injury;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.08 *Personality Disorders:* Manifested by pervasive, inflexible, and maladaptive personality traits, which are typical of the child's long-term functioning and not limited to discrete episodes of illness.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Deeply ingrained, maladaptive patterns of behavior, associated with one of the following:

1. Seclusiveness or autistic thinking; or

2. Pathologically inappropriate suspiciousness or hostility; or

3. Oddities of thought, perception, speech, and behavior; or

4. Persistent disturbances of mood or affect; or

5. Pathological dependence, passivity, or aggressiveness; or

6. Intense and unstable interpersonal relationships and impulsive and exploitative behavior; or

7. Pathological perfectionism and inflexibility;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.09 *Psychoactive Substance Dependence Disorders:* Manifested by a cluster of cognitive, behavioral, and physiologic symptoms that indicate impaired control of psychoactive substance use with continued use of the substance despite adverse consequences.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of at least four of the following:

1. Substance taken in larger amounts or over a longer period than intended and a great deal of time is spent in recovering from its effects; or

2. Two or more unsuccessful efforts to cut down or control use; or

3. Frequent intoxication or withdrawal symptoms interfering with major role obligations; or

4. Continued use despite persistent or recurring social, psychological, or physical problems; or

5. Tolerance, as characterized by the requirement for markedly increased amounts of substance in order to achieve intoxication; or

6. Substance taken to relieve or avoid withdrawal symptoms;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the

appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.10 *Autistic Disorder and Other Pervasive Developmental Disorders:* Characterized by qualitative deficits in the development of reciprocal social interaction, in the development of verbal and nonverbal communication skills, and in imaginative activity. Often, there is a markedly restricted repertoire of activities and interests, which frequently are stereotyped and repetitive.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of the following:

1. For autistic disorder, all of the following:

a. Qualitative deficits in the development of reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity; and

c. Markedly restricted repertoire of activities and interests;

OR

2. For other pervasive developmental disorders, both of the following:

a. Qualitative deficits in the development of reciprocal social interaction; and

b. Qualitative deficits in verbal and nonverbal communication and in imaginative activity;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraphs B2 of 112.02.

112.11 *Attention Deficit Hyperactivity Disorder:* Manifested by developmentally inappropriate degrees of inattention, impulsiveness, and hyperactivity.

The required level of severity for these disorders is met when the requirements in both A and B are satisfied.

A. Medically documented findings of all three of the following:

1. Marked inattention; and

2. Marked impulsiveness; and

3. Marked hyperactivity;

AND

B. For older infants and toddlers (age 1 to attainment of age 3), resulting in at least one of the appropriate age-group criteria in paragraph B1 of 112.02; or, for children (age 3 to attainment of age 18), resulting in at least two of the appropriate age-group criteria in paragraph B2 of 112.02.

112.12 *Developmental and Emotional Disorders of Newborn and Younger Infants (Birth to attainment of age 1):* Developmental or emotional disorders of infancy are evidenced by a deficit or lag in the areas of motor, cognitive/communicative, or social functioning. These disorders may be related either to organic or to functional factors or to a combination of these factors.

The required level of severity for these disorders is met when the requirements of A, B, C, D, or E are satisfied.

A. Cognitive/communicative functioning generally acquired by children no more than one-half the child's chronological age, as documented by appropriate medical findings (e.g., in infants 0-6 months, markedly diminished variation in the production or imitation of sounds and severe feeding abnormality, such as problems with sucking swallowing, or chewing) including, if necessary, a standardized test;

OR

B. Motor development generally acquired by children no more than one-half the child's chronological age, documented by appropriate medical findings, including if necessary, a standardized test;

OR

C. Apathy, over-excitability, or fearfulness, demonstrated by an absent or grossly excessive response to one of the following:

1. Visual stimulation; or

2. Auditory stimulation; or

3. Tactile stimulation;

OR

D. Failure to sustain social interaction on an ongoing, reciprocal basis as evidenced by:

1. Inability by 6 months to participate in vocal, visual, and motoric exchanges (including facial expressions); or

2. Failure by 9 months to communicate basic emotional responses, such as cuddling or exhibiting protest or anger; or

3. Failure to attend to the caregiver's voice or face or to explore an inanimate object for a period of time appropriate to the infant's age;

OR

E. Attainment of development or function generally acquired by children no more than two-thirds of the child's chronological age in two or more areas (*i.e.*, cognitive/communicative, motor, and social), documented by appropriate medical findings, including if necessary, standardized testing.

113.00 Malignant Neoplastic Diseases

A. *What impairments do these listings cover?* We use these listings to evaluate all malignant neoplasms except certain neoplasms associated with human immunodeficiency virus (HIV) infection. We use the criteria in 114.08E to evaluate carcinoma of the cervix, Kaposi's sarcoma, lymphoma, and squamous cell carcinoma of the anus if you also have HIV infection.

B. *What do we consider when we evaluate malignant neoplastic diseases under these listings?* We consider factors such as the:

1. Origin of the malignancy.

2. Extent of involvement.

3. Duration, frequency, and response to antineoplastic therapy. Antineoplastic therapy means surgery, irradiation, chemotherapy, hormones, immunotherapy, or bone marrow or stem cell transplantation. When we refer to surgery as an antineoplastic treatment, we mean surgical excision for treatment, not for diagnostic purposes.

4. Effects of any post-therapeutic residuals.

C. *How do we apply these listings?* We apply the criteria in a specific listing to a malignancy originating from that specific site.

D. *What evidence do we need?*

1. We need medical evidence that specifies the type, extent, and site of the primary, recurrent, or metastatic lesion. In the rare situation in which the primary site cannot be identified, we will use evidence documenting the site(s) of metastasis to evaluate the impairment under 13.27 in part A.

2. For operative procedures, including a biopsy or a needle aspiration, we generally need a copy of both the:

a. Operative note.

b. Pathology report.

3. When we cannot get these documents, we will accept the summary of hospitalization(s) or other medical reports. This evidence should include details of the findings at surgery and,

whenever appropriate, the pathological findings.

4. In some situations we may also need evidence about recurrence, persistence, or progression of the malignancy, the response to therapy, and any significant residuals. (*See* 113.00G.)

E. *When do we need longitudinal evidence?*

1. *Tumors with distant metastases.* Most malignant tumors of childhood consist of a local lesion with metastases to regional lymph nodes and, less often, distant metastases. We generally do not need longitudinal evidence for tumors that have metastasized beyond the regional lymph nodes because these tumors usually meet the requirements of a listing. Exceptions are for tumors with distant metastases that are expected to respond to antineoplastic therapy. For these exceptions, we usually need a longitudinal record of 3 months after therapy starts to determine whether the intended effect of therapy has been achieved and is likely to persist.

2. *Other malignancies.* When there are no distant metastases, many of the listings require that we consider your response to initial antineoplastic therapy; that is, the initial planned treatment regimen. This therapy may consist of a single modality or a combination of modalities (multimodal) given in close proximity as a unified whole, and is usually planned before any treatment(s) is initiated. Examples of multimodal therapy include:

a. Surgery followed by chemotherapy or radiation.

b. Chemotherapy followed by surgery.

c. Chemotherapy and concurrent radiation.

3. *Types of treatment.* Whenever the initial planned therapy is a single modality, enough time must pass to allow a determination about whether the therapy will achieve its intended effect. If the treatment fails, the failure will often happen within 6 months after treatment starts, and there will often be a change in the treatment regimen. Whenever the initial planned therapy is multimodal, a determination about the effectiveness of the therapy usually cannot be made until the effects of all the planned modalities can be determined. In some cases, we may need to defer adjudication until the effectiveness of therapy can be assessed. However, we do not need to defer adjudication to determine whether the therapy will achieve its intended effect if we can make a fully favorable determination or decision based on the length and effects of therapy, or the residuals of the malignancy or therapy (*see* 113.00G).

F. *How do we evaluate impairments that do not meet one of the malignant neoplastic diseases listings?*

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

2. If you have a severe medically determinable impairment(s) that does not meet a listing, we will determine whether your impairment(s) medically equals a listing. (*See* §§404.1526 and

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

G. *How do we consider the effects of therapy?*

1. *How we consider the effects of therapy under the listings.* In many cases, malignancies meet listing criteria only if the therapy does not achieve the intended effect: the malignancy persists, progresses, or recurs despite treatment. However, as explained in the following paragraphs, we will not delay adjudication if we can make a fully favorable determination or decision based on the evidence in the case record.

2. *Effects can vary widely.*

a. Because the therapy and its toxicity may vary widely, we consider each case on an individual basis. We will request a specific description of the therapy, including these items:

i. Drugs given.

ii. Dosage.

iii. Frequency of drug administration.

iv. Plans for continued drug administration.

v. Extent of surgery.

vi. Schedule and fields of radiation therapy.

b. We will also request a description of the complications or adverse effects of therapy, such as the following:

i. Continuing gastrointestinal symptoms.

ii. Persistent weakness.

iii. Neurological complications.

iv. Cardiovascular complications.

v. Reactive mental disorders.

3. *Effects of therapy may change.* Because the severity of the adverse effects of antineoplastic therapy may change during treatment, enough time must pass to allow us to evaluate the therapy's effect. The residual effects of treatment are temporary in most instances. But on occasion, the effects may be disabling for a consecutive period of at least 12 months.

4. *When the initial antineoplastic therapy is effective.* We evaluate any post-therapeutic residual impairment(s) not included in these listings under the criteria for the affected body

system. We must consider any complications of therapy. When the residual impairment(s) does not meet a listed impairment, we must consider whether it medically equals a listing, or, as appropriate, functionally equals the listings.

H. *How long do we consider your impairment to be disabling?*

1. In some listings, we specify that we will consider your impairment to be disabling until a particular point in time (for example, at least 12 months from the date of diagnosis). We may consider your impairment to be disabling beyond this point when the medical and other evidence justifies it.

2. When a listing does not contain such a specification, we will consider an impairment(s) that meets or medically equals a listing in this body system to be disabling until at least 3 years after onset of complete remission. When the impairment(s) has been in complete remission for at least 3 years, that is, the original tumor and any metastases have not been evident for at least 3 years, the impairment(s) will no longer meet or equal the criteria of a listing in this body system.

3. Following the appropriate period, we will consider any residuals, including residuals of the malignancy or therapy (*see* 113.00G), in determining whether you are disabled.

I. *What do these terms in the listings mean?*

1. *Persistent:* Failure to achieve a complete remission.

2. *Progressive:* The malignancy became more extensive after treatment.

3. *Recurrent, relapse:* A malignancy that had been in complete remission or entirely removed by surgery has returned.

J. *Can we establish the existence of a disabling impairment prior to the date of the evidence that shows the malignancy satisfies the criteria of a listing?* Yes. We will consider factors such as:

1. The type of malignancy and its location.

2. The extent of involvement when the malignancy was first demonstrated.

3. Your symptoms.

K. *How do we evaluate specific malignant neoplastic diseases?*

1. *Lymphoma.*

a. Listing 113.05 provides criteria for evaluating intermediate or high grade lymphomas that have not responded to antineoplastic therapy. Low grade or indolent lymphomas are rare in children. We will evaluate low grade or indolent lymphomas under 13.05 in part A.

b. We consider Hodgkin's disease that recurs more than 12 months after completing initial antineoplastic therapy to be a new disease rather than a recurrence.

c. Many children with lymphoma are treated according to a long-term protocol that can result in significant adverse medical, social, and emotional consequences. (*See* 113.00G.)

2. *Leukemia.*

a. *Acute leukemia.* The initial diagnosis of acute leukemia, including the accelerated or blast phase of chronic myelogenous (granulocytic) leukemia, is based upon definitive bone marrow examination. Additional diagnostic information is based on chromosomal analysis, cytochemical and surface marker studies on the abnormal cells, or other methods consistent with the prevailing state of medical knowledge and clinical practice. Recurrent disease must be documented by peripheral blood, bone marrow, or cerebrospinal fluid examination. The initial and follow-up pathology reports should be included.

b. *Chronic myelogenous leukemia (CML).* The diagnosis of CML should be based upon documented granulocytosis, including immature forms such as differentiated or undifferentiated myelocytes and myeloblasts, and a chromosomal analysis that demonstrates the Philadelphia chromosome. In the absence of a chromosomal analysis, or if the Philadelphia chromosome is not present, the diagnosis may be made by other methods consistent with the prevailing state of medical knowledge and clinical practice.

c. *Juvenile chronic myelogenous leukemia (JCML).* JCML is a rare, Philadelphia-chromosome-negative childhood leukemia that is aggressive and clinically similar to acute myelogenous leukemia. We evaluate JCML under 113.06A.

d. *Elevated white cell count.* In cases of chronic leukemia, an elevated white cell count, in itself, is not ordinarily a factor in determining the severity of the impairment.

3. *Malignant solid tumors.* The tumors we consider under 113.03 include the histiocytosis syndromes except for solitary eosinophilic granuloma. Therefore, we will not evaluate brain tumors (*see* 113.13) or thyroid tumors (*see* 113.09) under this listing.

4. *Brain tumors.* We use the criteria in 113.13 to evaluate malignant brain tumors. We will evaluate any complications of malignant brain tumors, such as resultant neurological or psychological impairments, under the criteria for the affected body system. We evaluate benign brain tumors under 111.05.

5. *Retinoblastoma.* The treatment for bilateral retinoblastoma usually results in a visual impairment. We will evaluate any resulting visual impairment under 102.02.

L. *How do we evaluate malignant neoplastic diseases treated by bone marrow or stem cell transplantation?* Bone marrow or stem cell transplantation is performed for a variety of malignant neoplastic diseases.

1. *Acute leukemia (including T-cell lymphoblastic lymphoma and JCML) or accelerated or blast phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of transplantation, whichever is later.

2. *Lymphoma or chronic phase of CML.* If you undergo bone marrow or stem cell transplantation for any of these disorders, we will consider you to be disabled until at least 12

months from the date of transplantation.

3. *Evaluating disability after the appropriate time period has elapsed.* We consider any residual impairment(s), such as complications arising from:

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

b. Immunosuppressant therapy, such as frequent infections.

c. Significant deterioration of other organ systems.

113.01 Category of Impairments, Malignant Neoplastic Diseases

113.03 *Malignant solid tumors.* Consider under a disability:

A. For 2 years from the date of initial diagnosis. Thereafter, evaluate any residual impairment (s) under the criteria for the affected body system.

OR

B. For 2 years from the date of recurrence of active disease. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

113.05 *Lymphoma (excluding T-cell lymphoblastic lymphoma—113.06).* (*See* 113.00K1.)

A. Non-Hodgkins lymphoma, including Burkitt's and anaplastic large cell. Persistent or recurrent following initial antineoplastic therapy.

OR

B. Hodgkin's disease with failure to achieve clinically complete remission, or recurrent disease within 12 months of completing initial antineoplastic therapy.

OR

C. With bone marrow or stem cell transplantation. Consider under a disability until at least 12 months from the date of transplantation. Thereafter, evaluate any residual impairment(s) under the criteria of the affected body system.

113.06 *Leukemia.* (*See* 113.00K2.)

A. Acute leukemia (including T-cell lymphoblastic lymphoma and juvenile chronic myelogenous leukemia (JCML)). Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

OR

B. Chronic myelogenous leukemia (except JCML), as described in 1 or 2:

1. Accelerated or blast phase. Consider under a disability until at least 24 months from the date of diagnosis or relapse, or at least 12 months from the date of bone marrow or stem cell transplantation, whichever is later. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

2. Chronic phase, as described in a or b:

a. Consider under a disability until at least 12 months from the date of bone marrow or stem cell transplantation. Thereafter, evaluate any residual impairment(s) under the criteria for the affected body system.

b. Progressive disease following initial antineoplastic therapy.

113.09 *Thyroid gland.*

A. Anaplastic (undifferentiated) carcinoma.

OR

B. Carcinoma with metastases beyond the regional lymph nodes progressive despite radioactive iodine therapy.

113.12 *Retinoblastoma.*

A. With extension beyond the orbit.

OR

B. Persistent or recurrent following initial antineoplastic therapy.

OR

C. With regional or distant metastases.

113.13 *Brain tumors.* (*See* 113.00K4.) Highly malignant tumors, such as Grades III and IV astrocytomas, glioblastoma multiforme, ependymoblastoma, medulloblastoma or other primitive neuroectodermal tumors (PNETs) with documented metastases, diffuse intrinsic brain stem gliomas, or primary sarcomas.

113.21 *Neuroblastoma.*

A. With extension across the midline.

OR

B. With distant metastases.

OR

C. Recurrent.

OR

D. With onset at age 1 year or older.

114.00 Immune System

A. Listed disorders include impairments involving deficiency of one or more components of the immune system (*i.e.*, antibody-producing B cells; a number of different types of cells associated with cell-mediated immunity including T-lymphocytes, macrophages and monocytes; and components of the complement system).

B. *Dysregulation of the immune system* may result in the development of a connective tissue disorder. Connective tissue disorders include several chronic multisystem disorders that differ in their clinical manifestation, course, and outcome. These disorders are described in part A, 14.00B; inflammatory arthritis is also described in 114.00E.

Some of the features of connective tissue disorders in children may differ from the features in adults. When the clinical features are the same as that seen in adults, the principles and concepts in part A, 14.00B apply.

The documentation needed to establish the existence of a connective tissue disorder is medical history, physical examination, selected laboratory studies, appropriate medically acceptable imaging, and, in some instances, tissue biopsy. Medically acceptable imaging includes, but is not limited to, x-ray imaging, computerized axial tomography (CAT scan) or magnetic resonance imaging (MRI), with or without contrast material, myelography, and radionuclear bone scans. "Appropriate" means that the technique used is the proper one to support the evaluation and diagnosis of the impairment. However, the Social Security Administration will not purchase diagnostic tests or procedures that may involve significant risk, such as biopsies or angiograms. Generally, the existing medical evidence will contain this information.

In addition to the limitations caused by the connective tissue disorder *per se,* the chronic adverse effects of treatment (e.g., corticosteroid-related ischemic necrosis of bone) may result in functional loss.

A longitudinal clinical record of at least 3 months demonstrating active disease despite prescribed treatment during this period with the expectation that the disease will remain active for 12 months is necessary for assessment of severity and duration of impairment.

In children the impairment may affect growth, development, attainment of age-appropriate skills, and performance of age-appropriate activities. The limitations may be the result of serious loss of function because of disease affecting a single organ or body system, or lesser degrees of functional loss because of disease affecting two or more organs/body systems associated with significant constitutional symptoms and signs of severe fatigue, fever, malaise, weight loss, and joint pain and stiffness. We use the term "severe" in these listings to describe medical severity; the term does not have the same meaning as it does when we use it in

connection with a finding at the second step of the sequential evaluation processes in §§404.1520, 416.920, and 416.924.

C. Allergies, growth impairments and Kawasaki disease.

1. Allergic disorders (e.g., asthma or atopic dermatitis) are discussed and evaluated under the appropriate listing of the affected body system.

2. If growth is affected by the disorder or its treatment by immunosuppressive drugs, 100.00, Growth impairment, may apply. Children may have growth impairment as a result of the inflammatory arthritides because of the diseases' potential effects on the immature skeleton, open epiphyses, and young cartilage and bone. In such situations, the growth impairment should be evaluated under 100.00ff.

3. Kawasaki disease, also known as mucocutaneous lymph node syndrome, is characterized by multisystem manifestations, but significant functional impairment is usually due to disease of the coronary arteries, which should be evaluated under 104.00.

D. Human immunodeficiency virus (HIV) infection.

1. HIV infection is caused by a specific retrovirus and may be characterized by susceptibility to one or more opportunistic diseases, cancers, or other conditions, as described in 114.08. Any child 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 114.08 or is of equivalent severity to an impairment in 114.08.

2. Definitions. In 114.08, the terms "resistant to treatment," "recurrent," and "disseminated" 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.

3. 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.

a. 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:

i. 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 (e.g., Western blot, immunofluorescence assay). (See paragraph b, below, for information about HIV antibody testing in children younger than 24 months of age).

ii. A specimen that contains HIV antigen (e.g., serum specimen, lymphocyte culture, or cerebrospinal fluid (CSF) specimen).

iii. An immunoglobulin A (IgA) serological assay specific for HIV.

iv. Other test(s) that are highly specific for detection of HIV in children (e.g., 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, every reasonable effort must be made to obtain reports of the results of that testing.

b. Other acceptable documentation of HIV infection in children.

As noted in paragraph a, above, HIV infection is not documented in children under 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 percent of such infants are actually infected. Therefore, the presence of serum HIV antibodies alone does not establish the presence of HIV infection in a child under 24 months of age. 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.

For children from birth to the attainment of 24 months of age who have tested positive for HIV antibodies (see D3a above), HIV infection may be documented by one or more of the following:

i. For an infant 12 months of age or less, a CD4 (T4) count of 1500/mm[3] or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

ii. For an infant from 12 to 24 months of age, a CD4 (T4) count of 750/mm[3] or less, or a CD4 count less than or equal to 20 percent of total lymphocytes.

iii. An abnormal CD4/CD8 ratio.

iv. An IgG significantly greater than or less than the normal range for age.

HIV infection in children may also be documented without the definitive laboratory evidence described in paragraph a, or the other laboratory evidence discussed 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 such laboratory evidence is not 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 in children will be accepted without definitive laboratory evidence if the child has an opportunistic disease (e.g., Pneumocystis carinii pneumonia (PCP)) predictive of a defect in cell-mediated immunity, and there is no other known cause of diminished resistance to that disease (e.g., long-term steroid treatment, lymphoma). In such cases, every reasonable effort must be made to obtain full details of the history, medical findings, and results of testing.

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

a. Documentation of the manifestations of HIV infection in children by definitive diagnosis.

The definitive method of diagnosing opportunistic diseases or conditions that are manifestations of HIV infection in children is by culture, serological test, or microscopic examination of biopsied tissue or other material (e.g., bronchial washings). Therefore, every reasonable effort must be made to obtain specific laboratory evidence of an opportunistic disease or other condition whenever this information is available. If a histological 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 radiographic studies) or microscopic examination of the appropriate tissues or body fluids.

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

b. Other acceptable documentation of the manifestations of HIV infection in children.

Manifestations of HIV infection in children may also be documented without the definitive laboratory evidence described in paragraph a, 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 diagnosis(es) 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 (114.08D) 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.

5. HIV infection in children. The clinical manifestation and course of disease in children who become infected with HIV perinatally or in the first 6 years of life may differ from that in 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 present with failure to thrive or pneumocystis carinii pneumonia (PCP); young children may present with recurrent infections, neurological problems, or developmental abnormalities. Older children may also exhibit neurological abnormalities, such as HIV 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 in 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 bacteria, e.g., some pneumonias, can be severely limiting, especially in pre-adolescent children. These major bacterial infections should be evaluated under 114.08A5, which requires two or more such infections within a 2-year period. Although 114.08A5 applies only to children less 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 (e.g., delayed puberty).

Otherwise, bacterial infections are evaluated under 114.08A6. The criteria of the listing are met if one or more bacterial infection(s) occurs and requires hospitalization or intravenous antibiotic treatment 3 or more times in 1 year. Pelvic inflammatory disease in older female children should be evaluated under multiple or recurrent bacterial infections (114.08A6).

6. Evaluation of HIV infection in children. The criteria in 114.08 do not describe the full spectrum of diseases or conditions manifested by children with HIV infection. As in any case, consideration must be given to whether a child's impairment(s) meets, medically equals, or functionally equals the severity of any other listing in appendix 1 of subpart P; e.g., a neoplastic disorder listed in 113.00ff. (See §§404.1526, 416.926, and 416.926a.) Although 114.08 includes cross-references to other listings for the more common manifestations of HIV infection, additional listings may also apply.

In addition, the impact of all impairments, whether or not related to the HIV infection, must be considered. Children with HIV infection may manifest signs and symptoms of a mental impairment (e.g., anxiety, depression), or of another physical impairment. Medical evidence should include documentation of all physical and mental impairments and the impairment(s) should be evaluated not only under the relevant listing(s) in 114.08, but under any other appropriate listing(s).

It is also important to remember that children with HIV infection, like all others, are evaluated under the full sequential evaluation process described in §416.924. If a child with HIV infection is working and engaging in substantial gainful activity (SGA), or does not have a severe

impairment, the case will be decided at the first or second step of the sequential evaluation process, and does not require evaluation under these listings. For a child with HIV infection who is not engaging in SGA and has a severe impairment, but whose impairment(s) does not meet the criteria of a listing, consideration will be given to whether the child's impairment or combination of impairments is either medically or functionally equivalent in severity to any listed impairment.

7. Effect of treatment. Medical treatment must be considered in terms of its effectiveness in ameliorating the signs, symptoms, and laboratory abnormalities of the specific disorder, or of the HIV infection itself (e.g. antiretroviral agents) and in terms of any side effects of treatment that may further impair the child.

Response to treatment and adverse or beneficial consequences of treatment may vary widely. For example, a child with HIV infection who develops 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), 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.

8. Functional criteria. Paragraph O of 114.08 establishes standards for evaluating manifestations of HIV infection that do not meet the requirements listed in 114.08A-N. Paragraph O is applicable for manifestations that are not listed in 114.08A-N, as well as those listed in 114.08A-N that do not meet the criteria of any of the rules in 114.08A-N.

For children with HIV infection evaluated under 114.08O, 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 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 114.08O, 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. (See 112.00C for additional discussion of these areas of functioning).

E. *Inflammatory arthritis (114.09)* includes a vast array of disorders that differ in cause, course, and outcome. For example, in children inflammatory spondyloarthropathies include juvenile ankylosing spondylitis, reactive arthropathies, psoriatic arthropathy, and Behçet's disease, as well as undifferentiated spondylitis. Inflammatory arthritis of peripheral joints likewise comprises many disorders, including juvenile rheumatoid arthritis, Sjögren's syndrome, psoriatic arthritis, crystal deposition disorders, and Lyme disease. Clinically, inflammation of major joints may be the dominant problem causing difficulties with ambulation or fine and gross

movements, or the arthritis may involve other joints or cause less restriction of age-appropriate ambulation or other movements but be complicated by extra-articular features that cumulatively result in serious functional deficit. When persistent deformity without ongoing inflammation is the dominant feature of the impairment, it should be evaluated under 101.02, or, if there has been surgical reconstruction, 101.03.

1. Because the features of inflammatory connective tissue diseases in children are modified by such factors as the child's limited antigenic exposure and immune reactivity, the acute inflammatory connective tissue diseases must be differentiated from each other in order to evaluate duration factors and responses to specific treatments. Chronic conditions must be differentiated from short-term reversible disorders, and also from other connective tissue diseases.

2. In 114.09A, the term *major joints* refers to the major peripheral joints, which are the hip, knee, shoulder, elbow, wrist-hand, and ankle-foot, as opposed to other peripheral joints (e.g., the joints of the hand or forefoot) or axial joints (*i.e.*, the joints of the spine.) The wrist and hand are considered together as one major joint, as are the ankle and foot. Since only the ankle joint, which consists of the juncture of the bones of the lower leg (tibia and fibula) with the hindfoot (tarsal bones), but not the forefoot, is crucial to weight bearing, the ankle and foot are considered separately in evaluating weight bearing.

3. The terms *inability to ambulate effectively* and *inability to perform fine and gross movements effectively* in 114.09A have the same meaning as in 101.00B2b and 101.00B2c and must have lasted, or be expected to last, for at least 12 months.

4. Inability to ambulate effectively is implicit in 114.09B. Even though children who demonstrate the findings of 114.09B will not ordinarily require bilateral upper limb assistance, the required ankylosis of the cervical or dorsolumbar spine will result in an extreme loss of the ability to see ahead, above, and to the side.

5. As in 114.02 through 114.06, extra-articular features of an inflammatory arthritis may satisfy the criteria for a listing in an involved extra-articular body system. Such impairments may be found to meet a criterion of 114.09C. Extra-articular impairments of lesser severity should be evaluated under 114.09D and 114.09E. Commonly occurring extra-articular impairments include keratoconjunctivitis sicca, uveitis, iridocyclitis, pleuritis, pulmonary fibrosis or nodules, restrictive lung disease, pericarditis, myocarditis, cardiac arrhythmias, aortic valve insufficiency, coronary arteritis, Raynaud's phenomena, systemic vasculitis, amyloidosis of the kidney, chronic anemia, thrombocytopenia, hypersplenism with compromised immune competence (Felty's syndrome), peripheral neuropathy, radiculopathy, spinal cord or cauda equina compression with sensory and motor loss, and heel enthesopathy with functionally limiting pain.

6. The fact that a child is dependent on steroids, or any other drug, for the control of inflammatory arthritis is, in and of itself, insufficient to find disability. Advances in the treatment of inflammatory connective tissue disease and in the administration of steroids for its treatment have corrected some of the previously disabling consequences of continuous steroid use. Therefore, each case must be evaluated on its own merits, taking into consideration the severity of the underlying impairment and any adverse effects of treatment.

114.01 Category of Impairments, Immune System

114.02 *Systemic lupus erythematosus.* Documented as described in 14.00B1 and 114.00B, with:

A. One of the following:

1. Growth impairment, as described under the criteria in 100.00ff; or

2. Musculoskeletal involvement, as described under the criteria in 101.00ff; or

3. Muscle involvement, as described under the criteria in 14.05; or

4. Ocular involvement, as described under the criteria in 102.00ff; or

5. Respiratory involvement, as described under the criteria in 103.00ff; or

6. Cardiovascular involvement, as described under the criteria in 104.00ff or 14.04D; or

7. Digestive involvement, as described under the criteria in 105.00ff; or

8. Renal involvement, as described under the criteria in 106.00ff; or

9. Hematologic involvement, as described under the criteria in 107.00ff; or

10. Skin involvement, as described under the criteria in 8.00ff; or

11. Endocrine involvement, as described under the criteria in 109.00ff; or

12. Neurological involvement, as described under the criteria in 111.00ff; or

13. Mental involvement, as described under the criteria in 112.00ff.

or

B. Lesser involvement of two or more organs/body systems listed in paragraph A, with significant, documented, constitutional symptoms and signs of severe fatigue, fever, malaise, and weight loss. At least one of the organs/body systems must be involved to at least a moderate level of severity.

114.03 *Systemic vasculitis.* As described under the criteria in 14.03 or, if growth impairment, as described under the criteria in 100.00ff.

114.04 *Systemic sclerosis and scleroderma.* Documented as described in 14.00B3 and 114.00B, and:

A. As described under the criteria in 14.04 or, if growth impairment, as described under the criteria in 100.00ff.

or

B. Linear scleroderma, with one of the following:

1. Fixed valgus or varus deformities of both hands or both feet; or

2. Marked destruction or marked atrophy of an extremity; or

3. Facial disfigurement from hypoplasia of the mandible, maxilla, or zygoma resulting in an impairment as described under the criteria in 112.00ff; or

4. Seizure disorder, as described under the criteria in 111.00ff.

114.05 *Polymyositis or dermatomyositis.* Documented as described in 14.00B4 and 114.00B, and:

A. As described under the criteria in 14.05.

or

B. With one of the following:

1. Multiple joint contractures; or

2. Diffuse cutaneous calcification with formation of an exoskeleton; or

3. Systemic vasculitis as described under the criteria in 14.03.

114.06 *Undifferentiated connective tissue disorder.* As described under the criteria in 114.02 or 114.04.

114.07 *Congenital immune deficiency disease.*

A. Hypogammaglobulinemia or dysgammaglobulinemia, with:

1. Documented, recurrent severe infections occurring 3 or more times within a 5-month period; or

2. An associated disorder such as growth retardation, chronic lung disease, collagen disorder or tumor. Evaluate according to the appropriate body system listing.

or

B. Thymic dysplastic syndromes (such as Swiss, diGeorge).

114.08 *Human immunodeficiency virus (HIV) infection.* With documentation as described in 114.00D3 and one of the following:

A. Bacterial infections:

1. Mycobacterial infection (e.g., caused by M. avium-intracellulare, M. kansasii, or M.

tuberculosis) at a site other than the lungs, skin, or cervical or hilar lymph nodes; or pulmonary tuberculosis resistant to treatment; or

2. Nocardiosis; or

3. Salmonella bacteremia, recurrent non-typhoid.

4. Syphilis or neurosyphilis—evaluate sequelae under the criteria for the affected body system (e.g., 102.00 Special Senses and Speech, 104.00 Cardiovascular System, 111.00 Neurological); or

5. In a child less than 13 years of age, multiple or recurrent pyogenic bacterial infection(s) 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 2 or more times in 2 years; or

6. Other multiple or recurrent bacterial infection(s), including pelvic inflammatory disease, requiring hospitalization or intravenous antibiotic treatment 3 or more times in 1 year.

or

B. Fungal infections:

1. Aspergillosis; or

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

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

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

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

6. Mucormycosis.

or

C. Protozoan or helminthic infections:

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

2. Pneumocystis carinii pneumonia or extrapulmonary pneumocystis carinii infection; or

3. Strongyloidiasis, extra-intestinal; or

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

or

D. Viral infections:

1. Cytomegalovirus disease (documented as described in 114.00D4b) at a site other than the liver, spleen, or lymph nodes; or

2. Herpes simplex virus causing:

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

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

c. Disseminated infection; or

3. Herpes zoster, either disseminated or with multidermatomal eruptions that are resistant to treatment; or

4. Progressive multifocal leukoencephalopathy; or

5. Hepatitis, as described under the criteria of 105.05.

or

E. Malignant neoplasms:

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

2. Karposi's sarcoma with:

a. Extensive oral lesions; or

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

c. Involvement of the skin or mucous membranes as described under the criteria of 114.08F; or

3. Lymphoma (e.g., primary lymphoma of the brain, Burkitt's lymphoma, immunoblastic sarcoma, other Non-Hodgkin's lymphoma, Hodgkin's disease); or

4. Squamous cell carcinoma of the anus.

or

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

8.00ff.

or

G. Hematologic abnormalities:

1. Anemia, as described under the criteria in 7.02; or

2. Granulocytopenia, as described under the criteria in 7.15; or

3. Thrombocytopenia, as described under the criteria of 107.06 or 7.06.

or

H. Neurological manifestations of HIV infection (e.g., HIV encephalopathy, peripheral neuropathy), as described under the criteria in 111.00ff, 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); or

2. Impaired brain growth (acquired microcephaly or brain atrophy—see 114.00D5); or

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

or

I. 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 2 months or longer; or

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

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

4. Growth impairment as described under the criteria in 100.00ff.

or

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

or

K. Cardiomyopathy, as described under the criteria in 104.00ff or 11.04.

or

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

or

M. Nephropathy, as described under the criteria in 106.00.

or

N. One or more of the following infections (other than described in A-M, above), resistant to treatment or requiring hospitalization or intravenous treatment 3 or more times in 1 year (or evaluate sequelae under the criteria for the affected body system):

1. Sepsis;

2. Meningitis; or

3. Pneumonia; or

4. Septic arthritis; or

5. Endocarditis; or

6. Sinusitis documented by appropriate medically acceptable imaging.

or

O. Any other manifestation(s) of HIV infection (including any listed in 114.08A-N, but without the requisite findings, e.g., oral candidiasis not meeting the criteria in 114.08F, diarrhea not meeting the criteria in 114.08J, or any other manifestation(s), e.g., oral hairy leukoplakia, hepatomegaly), resulting in one of the following:

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

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

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

114.09 *Inflammatory arthritis.* Documented as described in 114.00E, with one of the following:

A. History of joint pain, swelling, and tenderness, and signs on current physical examination of

joint inflammation or deformity in two or more major joints resulting in inability to ambulate effectively or inability to perform fine and gross movements effectively, as defined in 114.00E3 and 101.00B2b and B2c;

or

B. Ankylosing spondylitis or other spondyloarthropathy, with diagnosis established by findings of unilateral or bilateral sacroiliitis (e.g., erosions or fusions), shown by appropriate medically acceptable imaging, with both:

1. History of back pain, tenderness, and stiffness, and

2. Findings on physical examination of ankylosis (fixation) of the dorsolumbar or cervical spine at 45° or more of flexion measured from the vertical position (zero degrees);

or

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

or

D. Inflammatory arthritis, with signs of peripheral joint inflammation on current examination, but with lesser joint involvement than in A and lesser extra-articular features than in C, and:

1. Significant, documented constitutional symptoms and signs (e.g., fatigue, fever, malaise, weight loss), and

2. Involvement of two or more organs/body systems (see 114.00E5). At least one of the organs/body systems must be involved to at least a moderate level of severity.

or

E. Inflammatory spondylitis or other inflammatory spondyloarthropathies, with lesser deformity than in B and lesser extra-articular features than in C, with signs of unilateral or bilateral sacroiliitis on appropriate medically acceptable imaging; and with the extra-articular features described in 114.09D.

[50 FR 35066, Aug. 28, 1985]

EDITORIAL NOTE: For FEDERAL REGISTER citations affecting appendix 1 to subpart P of part 404, see the List of CFR Sections Affected, which appears in the Finding Aids section of the printed volume and on GPO Access.