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

4. *Polymyositis and dermatomyositis (114.05).*

a. *General.*

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

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

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

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

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

(ii) If you are of preschool age through adolescence (age 3 to attainment of age 18), weakness of your pelvic girdle muscles that results in your inability to rise independently from a squatting or sitting position or to climb stairs may be an indication that you are unable to walk without assistance. Weakness of your shoulder girdle muscles may result in your inability to

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

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

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

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

6. *Inflammatory arthritis (114.09).*

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

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

(i) Reactive arthropathies;

(ii) Juvenile ankylosing spondylitis;

(iii) Psoriatic arthritis;

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

(v) Behçet's disease; and

(vi) Inflammatory bowel disease