Polymyositis — Comprehensive overview covers symptoms, causes, treatment of this inflammatory muscle disease. Polymyositis (pol-e-mi-o-SI-tis) is an uncommon connective tissue disease. It's a type of inflammatory myopathy, which is characterized by muscle inflammation and weakness. The most noticeable characteristic of polymyositis is weakness of the skeletal muscles, which control movement. Polymyositis can occur at any age, but it mostly affects adults sometime between their 30s and 50s. It's more common in blacks than in whites, and women are affected more often than men are. Polymyositis signs and symptoms usually develop gradually, over weeks or months. Periods of remission in polymyositis, during which symptoms improve spontaneously, rarely occur. However, treatment can improve your muscle strength and function.

Symptoms

Signs and symptoms of polymyositis usually appear gradually, so it may be difficult to pinpoint when they first started. They may also fluctuate from week to week or month to month. Progressive muscle weakness is the most common polymyositis symptom. It typically affects the muscles closest to the trunk, such as those in your hips, thighs, shoulders, upper arms and neck. The weakness is symmetrical, affecting both the left and right sides of your body, and tends to gradually worsen. Although the muscle weakness starts subtly, after it progresses over the course of the disease it can make it difficult for you to climb stairs, rise from a seated position, lift objects or reach overhead.

Other polymyositis signs and symptoms include:

- Difficulty swallowing (dysphagia)
- Difficulty speaking
- Mild joint or muscle tenderness
- Fatigue
- Shortness of breath

Causes

Your immune system is responsible for helping to eliminate invaders (antigens) such as infectious organisms. The key cells in your immune system are lymphocytes known as B cells and T cells, which originate in your bone marrow. After T cells further develop in your thymus, all of your immune system cells gather in your lymph nodes and spleen. Antigens (triangular shapes above) are ingested (1), partially digested (2) and then presented to helper T cells by special cells called macrophages (3). This process activates the helper T cell to release hormones (lymphokines) that help B cells develop (4). These hormones, along with recognition of further antigens (5), change the B cell into an antibody-producing plasma cell (6). The antibodies (Y shapes above) produced can be one of several types (IgG, IgM, IgA, IgE and IgD) (7). The antibody "fits" the antigen much like a lock fits a key. The antigen is thus rendered harmless. The helper T cells also aid in development of cytotoxic T cells (8), which can kill antigens directly; memory T cells are produced (9) so that re-exposure to the same antigen will provide a more rapid and effective response (10).

Polymyositis is in a group of diseases or disorders of the muscles called inflammatory myopathies. The cause of most inflammatory myopathies is unknown.

Infections caused by bacteria, parasites or viruses can cause inflammatory myopathies, but in most cases of polymyositis, doctors aren't able to identify a preceding infection. Some doctors think certain people may have a genetic susceptibility to the disease.

Inflammatory myopathies share many characteristics with autoimmune disorders, in which your immune system attacks normal body components. Normally, your immune system works to protect your healthy cells from attacks by foreign substances, such as bacteria and viruses. If you

have polymyositis, an unknown cause may act as a trigger for your immune system to begin producing autoimmune antibodies (autoantibodies) that attack your body's own tissues.

Many people with polymyositis show a detectable level of autoantibodies in their blood.

Complications

Possible complications of polymyositis include:

- **Difficulty swallowing.** If the muscles in your esophagus are affected, you may have problems swallowing (dysphagia), which in turn may cause weight loss and malnutrition.
- **Aspiration and pneumonia.** Difficulty swallowing may also lead to entrance of food or liquids, including saliva, into your lungs (aspiration), which can lead to pneumonia.
- **Breathing problems.** If your chest muscles are affected by the disease, you may experience breathing problems, such as shortness of breath or in severe cases, respiratory failure.
- **Calcium deposits.** Late in the disease, particularly if you've had the disease for a long time, deposits of calcium can occur in your muscles, skin and connective tissues (calcinosis).

Associated conditions
Although these are not complications, polymyositis is often associated with other conditions that may cause further complications of their own, or in combination with polymyositis symptoms. Associated conditions include:

- **Raynaud's phenomenon.** This is a condition in which your fingers, toes, cheeks, nose and ears turn pale when exposed to cold temperatures.
- **Other connective tissue diseases.** Other conditions, such as lupus, rheumatoid arthritis, scleroderma and Sjogren's syndrome, can occur in combination with polymyositis.
- **Cardiovascular disease.** Polymyositis may cause the muscle of your heart to become inflamed (myocarditis). In a small number of people who have polymyositis, congestive heart failure and heart arrhythmias may develop.
- **Lung disease.** A condition called interstitial lung disease may occur with polymyositis. Interstitial lung disease refers to a group of disorders that cause scarring (fibrosis) of lung tissue, making lungs stiff and inelastic. Signs and symptoms include a dry cough and shortness of breath.

Concerns during pregnancy
Pregnancy may worsen signs and symptoms in women whose disease is active. Active polymyositis can also increase the risk of premature birth or stillbirth. If the disease is in remission, the risk isn't as great.

What to expect from your doctor
Your doctor will ask you detailed questions about your symptoms and family history, as well as completing a thorough physical examination that includes different types of strength tests. If your doctor suspects polymyositis, more tests will be ordered.

Tests and diagnosis

Diagnosis of polymyositis isn't always easy and can be a lengthy process. Even though the attempt to diagnose your condition may be frustrating, remember that an accurate diagnosis is necessary to receive appropriate treatment. In addition to a thorough physical exam, your doctor will likely use other tests to confirm a diagnosis of polymyositis.

Muscle tests

- **Electromyography.** A doctor with specialized training inserts a thin needle electrode through the skin into the muscle to be tested. Electrical activity is measured as you relax or tighten the muscle, and changes in the pattern of electrical activity can confirm a muscle disease. The doctor can determine the distribution of the disease by testing different muscles.
- **Muscle biopsy.** A small piece of muscle tissue is removed surgically for laboratory analysis. A muscle biopsy may reveal abnormalities in your muscles, such as inflammation, damage or infection. The tissue sample can also be examined for the presence of abnormal proteins and checked for enzyme deficiencies. In polymyositis, a muscle biopsy typically shows inflammation, dead muscle cells (necrosis), and degeneration and regeneration of muscle fibers.

Blood tests

- **Blood analysis.** A blood test will let your doctor know if you have elevated levels of muscle enzymes, such as creatine kinase (CK) and aldolase. Increased CK and aldolase levels can indicate muscle damage. A blood test can also detect specific autoantibodies associated with different symptoms of polymyositis, which can help in determining the best medication and treatment.

Treatments and drugs

Although there's no cure for polymyositis, treatment can improve your muscle strength and function. The earlier treatment is started in the course of polymyositis, the more effective it is, leading to fewer complications.

Drugs

- **Corticosteroids.** These medications suppress your immune system, limiting the production of antibodies and reducing muscle inflammation, as well as improving muscle strength and function. Corticosteroids, especially prednisone, are usually the first choice in treating inflammatory myopathies such as polymyositis.

  Your doctor may start with a very high dose, and then decrease it as your signs and symptoms improve. Improvement generally takes about two to four weeks, but therapy is often needed for years.

  Prolonged use of corticosteroids can have serious and wide-ranging side effects, so your doctor may recommend supplements to combat them, such as calcium and vitamin D, and may prescribe bisphosphonates, such as alendronate (Fosamax), risedronate (Actonel) or zoledronic acid (Reclast). Bisphosphonates in pill form may not be recommended if you have difficulty swallowing.

- **Corticosteroid-sparing agents.** Your doctor may recommend other medications, either to decrease side effects or if your condition doesn't respond to corticosteroids. These medications include azathioprine (Imuran) or methotrexate (Rheumatrex). Your doctor may prescribe these alone or in combination with corticosteroids.

  When in combination, these additional immunosuppressants can be used to lessen the dose and potential side effects of the corticosteroid. Immunosuppressants, such as cyclophosphamide (Cytoxan) and cyclosporine (Neoral, Sandimmune), may improve signs and symptoms of polymyositis and interstitial lung disease.