Polymyalgia rheumatica (PMR) is a rheumatic disease characterized by aching and morning stiffness in the shoulder, hips, neck, and torso that rarely occurs in individuals under the age of 50. Symptoms usually affect both sides of the body equally, but asymmetrical pain or stiffness can occur. Some patients also complain of malaise, fatigue, appetite loss, weight loss, and fever. It is a relatively common disorder. Among Caucasians over age 50 about 7 out of 1000 people are affected.

Giant cell (or temporal) arteritis (GCA) is a chronic inflammation of medium and large sized arteries that also occurs most commonly among individuals over 50 years of age. In this age group, the prevalence is approximately 2 per 1000 persons. Although the inflammation may be anywhere, it most frequently involves the branches of arteries that originate from the neck and go into the head. Polymyalgia rheumatica occurs in about 50 percent of patients with GCA, while approximately 5 to 15 percent of patients with PMR as the primary diagnosis have GCA. Some patients who have one disorder and may later develop evidence of the other.

There are two main issues that arise when the diagnosis of PMR is being considered:

- How is the diagnosis established and distinguished from other disorders that can produce similar symptoms?
• Does the patient also have giant cell arteritis (GCA)?

SYMPTOMS OF POLYMYALGIA RHEUMATICA — The cause of the discomfort and stiffness found among patients with PMR is inflammation. The location of the inflammation is disputed, it may be in or near large joints such as the hip and shoulder. Peripheral joints may also be involved and arthritis can occur in the small joints of the hands and feet. When these smaller joints are affected the arthritis is usually mild, may be transient, and resolves promptly upon treatment with anti-inflammatory steroids, such as prednisone. (See "Polymyalgia rheumatica").

Some patients develop swelling due to increased tissue fluid in the hands, wrists, ankles, or the top of the feet. Pitting edema is when a depression remains in swollen tissue after it is pressed. The edema usually occurs with other signs of polymyalgia rheumatica, but can be the first sign of the disease. The edema appears to represent the inflammation in certain structures. Swelling of the tissues of the wrist may lead to symptoms of carpal tunnel syndrome, which occurs in approximately 10 to 15 percent of patients with PMR.

There may be a decreased ability to actively move the shoulders, neck, and hips. The shoulders may be tender to touch, but this is usually less prominent than expected given the severity of the symptoms. Muscle strength is usually normal, though weakness may become a problem if the muscles atrophy from disuse due to pain.

DIAGNOSIS OF POLYMYALGIA RHEUMATICA — A combination of approaches are used to diagnose polymyalgia rheumatica, including physical examination, laboratory tests, and imaging tests (X-ray, MRI, etc).

A person should meet the following three criteria for the clinical diagnosis of PMR:

• Age 50 years or older at onset of symptoms.
• Aching and morning stiffness (lasting 30 minutes or more) on both sides of the body, persisting for at least one month and involving at least two of the following three areas: neck or torso, shoulders or proximal regions of the arms, and hips or proximal aspects of the thighs.
• Erythrocyte sedimentation rate (a blood test which indirectly measures inflammation by the rate at which red blood cells fall through blood plasma) of more than 40 mm per hour.

Some doctors will consider the prompt response of symptoms to corticosteroids as an additional criterion. On the other hand, the presence of another disease to explain the findings excludes PMR as the diagnosis. Examples of other diseases which must be excluded are: rheumatoid arthritis and chronic infections.

The diagnosis is more difficult to establish in those with atypical presentations. These include those patients aged 40 to 50 years, those with symptoms affecting one side of the body more than the other, and those with an erythrocyte sedimentation rate less than 40 mm/h.

**SYMPTOMS OF GIANT CELL ARTEMIS** — The onset of the symptoms in GCA tends to be gradual, but they can occur abruptly. Symptoms that affect the whole body are often present and include fever (in up to one-half of cases), fatigue, and weight loss. Although the fever is usually low grade, it can reach 102°F to 104°F (39°C to 40°C) in about 15 percent of affected individuals, and may cause confusion with infectious diseases. (See "Clinical manifestations and diagnosis of giant cell (temporal) arteritis").

Most patients have symptoms due to the inflammation, narrowing, and/or blockage of characteristic arteries at some time during the course of the illness. Common findings include:

**Headache** — A new headache occurs in two-thirds of patients with GCA. The pain, which may be mild or severe, tends to be located over the temples, but may be in the front or back of the skull. The headaches have a variable course; they may become progressively worse, but sometimes they can subside before treatment is started. The arteries in the temples or back of the skull are tender to touch in approximately one-third of patients.

**Jaw pain and/or weakness** — Nearly one-half of patients suffer from jaw pain and/or weakness (claudication) as a result of decreased blood flow to the muscles involved in
chewing. In some cases, muscle spasms occur rather than fatigue of the chewing muscles. Tongue or throat pain may occur as well.

**Visual loss** — Permanent partial or complete loss of vision in one or both eyes has been observed in 15 to 20 percent of affected individuals. Impaired vision is often an early manifestation of the disease. Affected persons typically note an abrupt partial loss of vision in one eye, which may progress to total blindness. If untreated, the second eye is likely to become affected within one to two weeks. It is rare, however, for patients to become completely blind in both eyes. Transient visual loss or double vision are each infrequent symptoms, but may precede permanent visual loss.

**Arm claudication** — Arm weakness that develops with use and resolves with resting of the arms occurs in approximately 15 percent of cases, due to the narrowing of arteries.

**Polymyalgia rheumatica** — As noted above, polymyalgia rheumatica is closely linked to GCA, occurring in about one-half of patients. On the other hand, GCA is found in about 5 to 15 percent of patients with PMR. The exact nature of the relationship between these two syndromes is not completely understood. The population of patients affected and the genetic susceptibility are identical. However, some patients have symptoms and findings limited to GCA or to PMR throughout the course of their illness, while others have manifestations of both, either simultaneously or at different times.

**Upper respiratory complaints** — About 10 percent of patients have upper respiratory symptoms. These include a cough, which is usually nonproductive, and sore throat, which may be very severe.

**Thoracic aortic aneurysms** — The development of a thoracic aortic aneurysm, dilation in the main artery carrying blood from the heart, is a late and potentially serious complication of GCA. Screening for a developing aneurysm is done using periodic chest X-rays. If an aneurysm is detected by chest X-ray serial measurements of the diameter of the aorta using computer aided tomography (CT scan) is helpful in determining whether surgical repair of the aneurysm is necessary.
**Diagnosis of Giant Cell Arteritis** — A combination of approaches are used to diagnose GCA, including physical examination, laboratory tests, biopsy of an affected artery, and imaging tests (eg, ultrasonography or magnetic resonance imaging).

Persons with giant cell arteritis should have at least three of the following criteria that are suggestive of arteritis:

- Age 50 years or older at onset of symptoms
- New, localized headache
- Tenderness or decreased pulse of the temporal artery (artery in the temple)
- Erythrocyte sedimentation rate greater than 50 mm/h
- Biopsy that includes an artery, and reveals signs characteristic of giant cell arteries

Some physicians exclude an elevated ESR as part of the diagnostic criteria, but include scalp tenderness, claudication of the jaw and/or tongue, or claudication with swallowing.

**Biopsy of Involved Artery** — Biopsy of one or both of the arteries in the temples is suggested in all cases of suspected GCA. A tender or swollen segment of the artery should be chosen for biopsy if present, since the inflammation may be limited to that area. Although the arteries in the temples are most accessible for biopsy, other arteries can be biopsied if they are clearly abnormal. Whenever possible, it is preferable to perform a biopsy prior to beginning treatment in order to maximize the possibility of an accurate diagnosis. However, since resolution of the inflammation occurs slowly after treatment, the arteries may show a persistent inflammation several weeks after the start of anti-inflammatory steroid therapy.

**Imaging Studies** — Ultrasonography has been used to detect swelling of temporal arteries and can sometimes reveal narrowing and decreased blood flow. Not all imaging centers have experience with the use of ultrasound for this purpose and an abnormal ultrasound test is not considered sufficient for diagnosis. It has not replaced temporal artery biopsy.

Another test called ocular pneumoplethysmography can reveal abnormalities in blood flow to the eyes that may suggest the presence of GCA. Like ultrasound, this technique does not supplant biopsy.
Angiography involves using X-ray guidance to pass a small catheter through an artery; a dye is injected that allows the arteries to be seen so the anatomy of the artery can be analyzed. This technique can be combined with balloon dilation of narrowed segments of arteries, especially those that carry blood to the arms and legs.

Computerized tomographic (CT) scanning and magnetic resonance angiography (MRA), an MRI of the blood vessels, can also detect large artery involvement, and MRI has the potential to follow changes in the thickness of arterial walls that can not be appreciated by X-ray angiography.

DISTINGUISHING PMR FROM GCA — As noted above, there is considerable overlap between PMR and GCA. Nevertheless, patients with "pure" PMR lack the classic symptoms and findings of GCA such as headache, jaw pain, visual loss, or tenderness of the temporal artery.

An important issue that arises is whether temporal artery biopsy should be performed. Although this is recommended in patients with GCA, the yield of information that it provides is quite low in patients with "pure" PMR. Thus, temporal artery biopsy may not be necessary in patients with MIR who have no symptoms suggestive of GCA. Furthermore, steroid therapy should be effective treatment for both disorders and does not appear to affect the amount of information gathered by temporal artery biopsy should it be performed later. Still, ongoing monitoring for signs suggestive of GCA is warranted.

TREATMENT — Polymyalgia rheumatica characteristically responds promptly to anti-inflammatory steroids. Naturally occurring anti-inflammatory steroids, such as cortisol, and other synthetic corticosteroids such as prednisone and prednisolone may be used, usually in low doses. Giant cell arteritis is also treated with corticosteroids. The initial dose of prednisone needed to alleviate musculoskeletal symptoms in PMR is lower than that required to control the vascular inflammation associated with GCA.

Treatment for polymyalgia rheumatica — Depending upon the patient's weight and the severity of symptoms, a starting dose of corticosteroids between 7.5 and 20 mg per day can be tried. Patients usually respond quickly and often note improvement after the first dose. The dose should be increased if the symptoms are not well controlled within one week. (See "Polymyalgia rheumatica").
The effective steroid dose should be maintained for two to four weeks after the aching and stiffness have resolved. The dose should then be gradually reduced every two to four weeks by approximately 10 percent to find the minimum amount that is needed to maintain the suppression of symptoms. Once the dose has fallen below 10 mg/day, it is best to reduce no faster than 1 mg per month.

Relapse occurs in as many as 25 to 50 percent of patients after therapy. This is more likely to occur in patients in whom steroid tapering has occurred more rapidly than suggested. Relapse may be less common if other agents are used along with corticosteroids in the initial treatment, but this point is still controversial.

Side effects with corticosteroids in polymyalgia rheumatica — Treatment of patients with PMR with corticosteroids may result in a high incidence of side-effects. The risk of diabetes mellitus and fractures of the vertebral bodies and hip, was increased two- to fivefold among patients with PMR treated with steroids compared to people who were disease-free.

Methotrexate — Among those with PMR, who are at increased risk for corticosteroid-induced side effects, the addition of the drug methotrexate may control symptoms while the steroids are being tapered and has been suggested to be able to reduce the rate of relapse. However, the vast majority of patients with PMR but without GCA seldom need methotrexate, since adverse effects are relatively mild with the low doses of prednisone required to adequately control the disease.

Treatment of GCA — Glucocorticoid treatment should begin once the diagnosis of GCA is strongly suspected, even if it has not yet been proven by biopsy. Patients with visual loss, high dose therapy given intravenously may be started immediately. If artery biopsies performed later reveal no evidence of arteritis, but there is still a strong suspicion of GCA as a result of vascular symptoms or findings, glucocorticoid treatment may still be warranted. For those patients with no evidence of arteritis on biopsy and for whom the diagnosis of GCA was considered only moderately likely or unlikely, steroid therapy may be quickly reduced to that needed for PMR (if present) or stopped entirely. (See "Treatment of giant cell (temporal) arteritis").

Initial dose of glucocorticoids in GCA — An initial dose of 40 to 60 mg of prednisone or its equivalent, in single or divided daily doses, is adequate in nearly all
cases of GCA; some patients have responded to doses as low as 20 mg/day. The dose should be increased if the patient does not respond promptly, and therapy should be initiated with intravenous pulse methylprednisolone (eg, 1 g intravenously daily for three days) in those with recent visual loss.

The effective starting dose of steroids should be continued until all reversible symptoms and findings have disappeared and laboratory tests have reverted to normal. This response usually occurs within two to four weeks after therapy is begun. The diagnosis of GCA should be reevaluated in patients who are resistant to steroid therapy in usual doses.

**Withdrawal of steroids in GCA** — Tapering of steroids can begin once the disease begins to go into remission. Tapering continues as long as the disease continues to be in remission. As an example, if the initial dose of prednisone was 60 mg/day, it can generally be reduced to 50 mg/day after two weeks and to 40 mg/day at the end of a month. After that, the dose can be gradually reduced by approximately 10 percent of the total daily dose each one or two weeks.

At some point, the results of monitoring tests may rise above normal, so further reductions in the dose may be temporarily halted. When this happens, subsequent reductions may have to be smaller and at longer intervals. It is not uncommon, for example, to find that it is necessary to continue doses of 10 to 20 mg or more of prednisone daily for several months before making a further reduction. This regimen of gradual reductions lets the doctor 'find the minimum dose required to suppress the disease and also helps to avoid "exacerbations" resulting from too rapid withdrawal of prednisone.

Taking corticosteroid therapy on alternate days is less effective than daily administration in GCA and cannot be depended upon to control symptoms. It may be tried, however, in those who have disturbing side effects with daily treatment.

**Methotrexate** — The addition of methotrexate may or may not control symptoms during steroid tapering in patients with GCA. Studies of this have given contradictory findings. The routine addition of methotrexate to glucocorticoid therapy for GCA is not recommended. However, among those who have developed or are at high risk for adverse effects of prednisone, the addition of methotrexate may be considered.
PROGNOSIS — In most patients, polymyalgia rheumatica tends to run a self-limited course over months to years, and steroid therapy can eventually be discontinued. There is no evidence of increased mortality associated with this disorder.

GCA also tends to run a self-limited course over several months to several years. The glucocorticoid dose can eventually be reduced and discontinued in most patients. A small group of those with GCA have more chronic disease and require low doses of prednisone for a number of years. Spontaneous relapses can occur in an unpredictable fashion and are seen more frequently in the first year or two of the disease. GCA does not seem to adversely affect mortality. The presence of thoracic aortic aneurysms does effect prognosis.

WHERE TO GET MORE INFORMATION — Your doctor is the best resource for finding out important information related to your particular case. Not all patients with polymyalgia rheumatica or giant cell arteritis are alike, and it is important that your situation is evaluated by someone who knows you as a whole person.

This discussion will be updated as needed every four months on our web site (http://www.uptodate.com). Additional topics as well as selected discussions written for healthcare professionals are also available for those who would like more detailed information.

A number of other sites on the intermit have information about polymyalgia rheumatica and giant cell arteritis. Information provided by the National Institutes of Health, national medical societies and some other well-established organizations are often reliable sources of information, although the frequency with which their information is updated is variable.

- National Library of Medicine
  (http://www.nlm.nih.gov/medlineplus)

- National Institute of Arthritis and Musculoskeletal and Skin Disease
  (http://www.niams.nih.gov)

- American College of Rheumatology
  (http://www.rheumatology.org)