

Polymyalgia Rheumatica and Giant Cell Arteritis

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) National Institutes of Health Public Health Service • U.S. Department of Health and Human Services

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What Are Polymyalgia Rheumatica and Giant Cell Arteritis?

Polymyalgia rheumatica is a rheumatic disorder that is associated with moderate to severe muscle pain and stiffness in the neck, shoulder, and hip area. Stiffness is most noticeable in the morning. This disorder may develop rapidly—in some patients, overnight. In other people, polymyalgia rheumatica develops more gradually. The cause of polymyalgia rheumatica is not known; however, possibilities include immune system abnormalities and genetic factors. The fact that polymyalgia rheumatica is rare in people under the age of 50 suggests it may be linked to the aging process.

Polymyalgia rheumatica may go away without treatment in 1 to several years. With treatment, the symptoms of polymyalgia rheumatica are quickly controlled, but relapse if treatment is stopped too early.

Giant cell arteritis, also known as temporal arteritis and cranial arteritis, is a disorder that results in swelling of arteries in the head (most often the temporal arteries, which are located on the temples on each side of the head), neck, and arms. This swelling causes the arteries to narrow, reducing blood flow. Early treatment is critical for good prognosis.

How Are Polymyalgia Rheumatica and Giant Cell Arteritis Related?

It is unclear how or why polymyalgia rheumatica and giant cell arteritis are related, but an estimated 15 percent of people in the United States with polymyalgia rheumatica also develop giant cell arteritis. Patients can develop giant cell arteritis either at the same time as polymyalgia rheumatica or after the polymyalgia symptoms disappear. About half of the people affected by giant cell arteritis also have polymyalgia rheumatica.

When a person is diagnosed with polymyalgia rheumatica, the doctor also should look for symptoms of giant cell arteritis because of the risk of blindness. With proper treatment, the disease is not threatening. Untreated, however, giant cell arteritis can lead to serious complications including permanent vision loss and stroke. Patients must learn to recognize the signs of giant cell arteritis, because they can develop even after the symptoms of polymyalgia rheumatica disappear. Patients should report any symptoms to the doctor immediately.

Who Is at Risk?

White women over the age of 50 are most at risk of developing polymyalgia rheumatica and giant cell arteritis. Women are twice as likely as men to develop the conditions. Both conditions almost exclusively affect people over the age of 50. The average age at onset is 70 years. Polymyalgia rheumatica and giant cell arteritis are quite common. In the United States, it is estimated that 700 per 100,000 people in the general population over 50 years of age develop polymyalgia rheumatica. An estimated 200 per 100,000 people over the age of 50 develop giant cell arteritis.

What Are the Symptoms?

The primary symptoms of polymyalgia rheumatica are moderate to severe stiffness and muscle pain near the neck, shoulders, or hips. The stiffness is more severe upon waking or after a period of inactivity, and typically lasts longer than 30 minutes. People with this condition also may have flulike symptoms, including fever, weakness, and weight loss.

Early symptoms of giant cell arteritis also may resemble the flu. People are likely to experience headaches, pain in the temples, and blurred or double vision. Pain may also affect the jaw and tongue.

How Are Polymyalgia Rheumatica and Giant Cell Arteritis Diagnosed?

No single test is available to definitively diagnose polymyalgia rheumatica. To diagnose the condition, a physician considers the patient's medical history, including symptoms that the patient reports, and results of laboratory tests that can rule out other possible diagnoses. The most typical laboratory finding in people with polymyalgia rheumatica is an elevated erythrocyte sedimentation rate, commonly referred to as the sed rate. This test measures how quickly red blood cells fall to the bottom of a test tube of unclotted blood. Rapidly descending cells (an elevated sed rate) indicate inflammation in the body. While the sed rate measurement is a helpful diagnostic tool, it alone does not confirm polymyalgia rheumatica. An abnormal result indicates only that tissue is inflamed, which also is a symptom of many forms of arthritis and/ or other rheumatic diseases. Before making a diagnosis of polymyalgia rheumatica, the doctor may perform additional tests to rule out other conditions, including rheumatoid arthritis, because symptoms of polymyalgia rheumatica and rheumatoid arthritis can be similar.

The doctor may recommend a test for rheumatoid factor (RF). RF is an antibody sometimes found in the blood. (An antibody is a special protein made by the immune system.) People with rheumatoid arthritis are likely to have RF in their blood, but most people with polymyalgia rheumatica do not. If the diagnosis still is unclear, a physician may conduct additional tests to rule out other disorders.

Doctors and patients both need to be aware of the risk of giant cell arteritis in people with polymyalgia rheumatica and should be on the lookout for symptoms of the disorder. Severe headaches, jaw pain, and vision problems are typical symptoms of giant cell arteritis. In addition, physical examination may reveal an abnormal temporal artery: tender to the touch, inflamed, and with reduced pulse. Because of the possibility of permanent blindness, a temporal artery biopsy is recommended if there is any suspicion of giant cell arteritis.

In a person with giant cell arteritis, the biopsy will show abnormal cells in the artery walls. Some patients showing symptoms of giant cell arteritis will have negative biopsy results. In such cases the doctor may suggest a second biopsy.

What Are the Treatments?

Polymyalgia rheumatica usually disappears without treatment in 1 to several years. With treatment, however, symptoms disappear quickly, usually in 24 to 48 hours. If there is no improvement, the doctor is likely to consider other possible diagnoses.

The treatment of choice is corticosteroid medication, usually prednisone. Polymyalgia rheumatica responds to a low daily dose of prednisone. The dose is increased as needed until symptoms disappear. Once symptoms disappear, the doctor may gradually reduce the dosage to determine the lowest amount needed to alleviate symptoms. The amount of time that treatment is needed is different for each patient. Most patients can discontinue medication after 6 months to 2 years. If symptoms recur, prednisone treatment is required again. Nonsteroidal anti-inflammatory drugs (NSAIDs) such as aspirin and ibuprofen also may be used to treat polymyalgia rheumatica. The medication must be taken daily, and longterm use may cause stomach irritation. For most patients, NSAIDs alone are not enough to relieve symptoms.

Giant cell arteritis carries a small but definite risk of blindness. The blindness is permanent once it happens. A high dose of prednisone is needed to prevent blindness and should be started as soon as possible, perhaps even before the diagnosis is confirmed with a temporal artery biopsy. When treated, symptoms quickly disappear. Typically, people with giant cell arteritis must continue taking a high dose of prednisone for 1 month. Once symptoms disappear and the sed rate is normal and there is no longer a risk of blindness, the doctor can begin to gradually reduce the dose. When treated properly, giant cell arteritis rarely recurs.

People taking low doses of prednisone rarely experience side effects. Side effects are more common among people taking higher doses. But all patients should be aware of potential effects, which include:

- fluid retention and weight gain
- rounding of the face
- delayed wound healing
- bruising easily

- diabetes
- myopathy (muscle wasting)
- glaucoma
- increased blood pressure
- decreased calcium absorption in the bones, which can lead to osteoporosis
- irritation of the stomach

People taking corticosteroids may have some side effects or none at all. A patient should report any side effects to the doctor. When the medication is stopped, the side effects disappear. Because prednisone and other corticosteroid drugs change the body's natural production of corticosteroid hormones, the patient should not stop taking the medication unless instructed by the doctor. The patient and doctor must work together to gradually reduce the medication.

What Is the Outlook?

Most people with polymyalgia rheumatica and giant cell arteritis lead productive, active lives. The duration of drug treatment differs by patient. Once treatment is discontinued, polymyalgia may recur; but once again, symptoms respond rapidly to prednisone. When properly treated, giant cell arteritis rarely recurs.

What Research Is Being Conducted To Help People Who Have Polymyalgia Rheumatica and Giant Cell Arteritis?

Researchers studying possible causes of polymyalgia rheumatica and giant cell arteritis are investigating the role of genetic predisposition, immune system abnormalities, and environmental factors. Scientists also are looking for markers of the diseases, exploring treatments, and studying why the two disorders often occur together.

With funding from the National Eye Institute, a new mouse model of giant cell arteritis is being used to examine interactions between the immune system and blood vessels to explain tissue damage.

Where Can People Get More Information About Polymyalgia Rheumatica and Giant Cell Arteritis?

 National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)

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- National Eye Institute Information Clearinghouse 2020 Vision Place Bethesda, MD 20892–3655 Phone: 301–496–5248 Fax: 301–402–1065 www.nei.nih.gov
- National Heart, Lung, and Blood Institute

31 Center Drive, MSC 2480 Bethesda, MD 20892–2480 Phone: 301–496–4236 Fax: 301–402–2405 www.nhlbi.nih.gov

American College of Rheumatology

1800 Century Place, Suite 250 Atlanta, GA 30345 Phone: 404–633–3777 Fax: 404–633–1870 www.rheumatology.org

Arthritis Foundation 1330 West Peachtreet Street Atlanta, GA 30309 Phone: 404–872–7100 or 800–283–7800 (free of charge) or call your local chapter (listed in the telephone directory) www.arthritis.org

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