

# Polymyalgia Rheumatica and Giant Cell Arteritis

## Questions and Answers about Polymyalgia Rheumatica and Giant Cell Arteritis

### What Is Polymyalgia Rheumatica?

Polymyalgia rheumatica is a rheumatic disorder associated with moderate-to-severe musculoskeletal pain and stiffness in the neck, shoulder, and hip area. Stiffness is most noticeable in the morning or after a period of inactivity, and typically lasts longer than 30 minutes. This disorder may develop rapidly; in some people it comes on literally overnight. But for most people, polymyalgia rheumatica develops more gradually.

The cause of polymyalgia rheumatica is not known. But it is associated with immune system problems, genetic factors, and an event, such as an infection, that triggers symptoms. The fact that polymyalgia rheumatica is rare in people under the age of 50 and becomes more common as age increases, suggests that it may be linked to the aging process.

Polymyalgia rheumatica usually resolves within 1 to 2 years. The symptoms of polymyalgia rheumatica are quickly controlled by treatment with corticosteroids, but symptoms return if treatment is stopped too early. Corticosteroid treatment does not appear to influence the length of the disease.

### What Is Giant Cell Arteritis?

Giant cell arteritis, also known as temporal arteritis and cranial arteritis, is a disorder that results in inflammation of arteries of the scalp (most apparent in the temporal arteries, which are located on the temples on each side of the head), neck, and arms. This inflammation causes the arteries to narrow, impeding adequate blood flow. For a good prognosis, it is critical to receive early treatment, before irreversible tissue damage occurs.

### How Are Polymyalgia Rheumatica and Giant Cell Arteritis Related?

It is unclear how or why polymyalgia rheumatica and giant cell arteritis frequently occur together. But some people with polymyalgia rheumatica also develop giant cell arteritis either simultaneously, or after the musculoskeletal symptoms have disappeared. Other people with giant cell arteritis also have polymyalgia rheumatica at some time while the arteries are inflamed.

When undiagnosed or untreated, giant cell arteritis can cause potentially serious problems, including permanent vision loss and stroke. So regardless of why giant cell arteritis might occur along with polymyalgia rheumatica, it is important that doctors look for symptoms of the arteritis in anyone diagnosed with polymyalgia rheumatica.

Patients, too, must learn and watch for symptoms of giant cell arteritis, because early detection and proper treatment are key to preventing complications. Any symptoms should be reported to your doctor immediately.

### What Are the Symptoms of Polymyalgia Rheumatica?

In addition to the musculoskeletal stiffness mentioned earlier, people with polymyalgia rheumatica also may have flu-like symptoms, including fever, weakness, and weight loss.

### What Are the Symptoms of Giant Cell Arteritis?

Early symptoms of giant cell arteritis may resemble flu symptoms such as fatigue, loss of appetite, and fever. Symptoms specifically related to the inflamed arteries of the head include headaches, pain and tenderness over the temples, double vision or visual loss, dizziness or problems with coordination, and balance. Pain may also affect the jaw and tongue, especially when eating, and opening the mouth wide may become difficult. In rare cases, giant cell arteritis causes ulceration of the scalp.

### Who Is at Risk for These Conditions?

Caucasian women over the age of 50 have the highest risk of developing polymyalgia rheumatica and giant cell arteritis. Although women are more likely than men to develop the conditions, research suggests that men with giant cell arteritis are more likely to suffer potentially blinding eye involvement. Both conditions almost exclusively affect people over the age of 50. The incidence of both peaks between 70 and 80 years of age.

Polymyalgia rheumatica and giant cell arteritis are both quite common, according to the National Arthritis Data Work Group. 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 50 years of age develop giant cell arteritis.

## How Are Polymyalgia Rheumatica and Giant Cell Arteritis Diagnosed?

A diagnosis of polymyalgia rheumatica is based primarily on the patient's medical history and symptoms, and on a physical examination. No single test is available to definitively diagnose polymyalgia rheumatica. However, doctors often use lab tests to confirm a diagnosis or rule out other diagnoses or possible reasons for the patient's symptoms.

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 inflammation by determining 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. Although 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, but this is also a symptom of many forms of arthritis and other rheumatic diseases.

Before making a diagnosis of polymyalgia rheumatica, the doctor may order additional tests. For example, the C-reactive protein test is another common means of measuring inflammation. There is also a common test for rheumatoid factor, an antibody (a protein made by the immune system) that is sometimes found in the blood of people with rheumatoid arthritis. Although polymyalgia rheumatica and rheumatoid arthritis share many symptoms, those with polymyalgia rheumatica rarely test positive for rheumatoid factor. Therefore, a positive rheumatoid factor might suggest a diagnosis of rheumatoid arthritis instead of polymyalgia rheumatica.

As with polymyalgia rheumatica, a diagnosis of giant cell arteritis is based largely on symptoms and a physical examination. The exam may reveal that the temporal artery is inflamed and tender to the touch, and that it has a reduced pulse.

Any doctor who suspects giant cell arteritis should order a temporal artery biopsy. In this procedure, a small section of the artery is removed through an incision in the skin over the temple area and examined under a microscope. A biopsy that is positive for giant cell arteritis 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.

## How Are They Treated?

The treatment of choice for both polymyalgia rheumatica and giant cell arteritis is corticosteroid medication, usually prednisone.

Polymyalgia rheumatica responds to a low daily dose of prednisone that is increased as needed until symptoms disappear. At this point, the doctor may gradually reduce the dosage to determine the lowest amount needed to alleviate symptoms. 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 (Advil, Motrin<sup>1</sup>), also may be used to treat polymyalgia rheumatica. The medication must be taken daily, and long-term use may cause stomach irritation. For most patients, NSAIDs alone are not enough to relieve symptoms.

<sup>1</sup> Brand names included in this booklet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

Even without treatment, polymyalgia rheumatica usually disappears in 1 to several years. With treatment, however, symptoms disappear quickly, usually in 24 to 48 hours. If prednisone doesn't bring improvement, the doctor is likely to consider other possible diagnoses.

Giant cell arteritis is treated with high doses of prednisone. If not treated promptly, the condition carries a small but definite risk of blindness, so prednisone should be started as soon as possible, perhaps even before confirming the diagnosis with a temporal artery biopsy.

As with polymyalgia rheumatica, the symptoms of giant cell arteritis quickly disappear with treatment; however, high doses of prednisone are typically maintained for 1 month.

Once symptoms disappear and the sed rate is normal, there is much less risk of blindness. At that point, the doctor can begin to gradually reduce the prednisone dose.

In both polymyalgia rheumatica and giant cell arteritis, an increase in symptoms may develop when the prednisone dose is reduced to lower levels. The doctor may need to hold the lower dose for a longer period of time or even modestly increase it again, temporarily, to control the symptoms. Once the symptoms are in remission and the prednisone has been discontinued for several months, recurrence is less common.

Whether taken on a long-term basis for polymyalgia rheumatica or for a shorter period for giant cell arteritis, prednisone carries a risk of side effects. Although long-term use and/or higher doses carry the greatest risk, people taking the drug at any dose or for any length of time should be aware of the potential side 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
- increase in infections.

People taking corticosteroids may have some side effects or none at all. Anyone who experiences side effects should report them to his or her doctor. When the medication is stopped, the side effects disappear. Because prednisone and other corticosteroid drugs reduce the body's natural production of corticosteroid hormones, which are necessary for the body to function properly, it is important not to stop taking the medication unless instructed by a doctor to do so. 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?**

Research is providing new information that will help scientists better understand polymyalgia rheumatica and giant cell arteritis. The following issues are being studied:

**Causes and mechanisms**—Researchers studying possible causes of polymyalgia rheumatica and giant cell arteritis are investigating the role of genetic predisposition, immune system problems, and environmental factors. In one study supported by the National Eye Institute, researchers are attempting to better understand the immunobiology of inflamed arteries and advance the understanding of events that initiate vasculitis. Other research funded by the National Eye Institute is using a mouse model of giant cell arteritis to examine interactions between the immune system and blood vessels, and to explain the tissue damage that results.

**Prognostic indicators**—By examining characteristics of people with and without the conditions, doctors are starting to understand some factors that are associated with both the disease, and its prognosis and manifestations. For example, one study has shown that women are more likely than men to have jaw involvement from giant cell arteritis, while men are more likely to have eye involvement that can lead to blindness.

**Treatment**—Although treatment with prednisone is almost always effective for both conditions, the drug carries the risk of potentially serious side effects. For that reason, one area of investigation involves looking for treatments that are safe while still being effective. A study funded by the National Center for Research Resources is looking at whether high doses of intravenous corticosteroid drugs given at the time of diagnosis can control giant cell arteritis more quickly, make it possible to prescribe lower subsequent doses of oral steroids, and control the disease with fewer drug side effects than current prednisone regimens.

**Longitudinal information**—As part of the NIH-funded Rare Diseases Clinical Research Network, scientists participating in the Vasculitis Clinical Research Consortium are collecting clinical and laboratory information from patients with giant cell arteritis to follow the disease over an extended period of time. Data from these studies will be used to examine the genetics and causes of giant cell arteritis, find new ways to track disease and predict responses, understand how to treat patients, and much more.

Revised December 2006

**This information was written by National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS).**

### **Acknowledgements**

*NIAMS gratefully acknowledges the assistance of the following individuals for reviewing this booklet: Susana Serrate-Sztejn, M.D., and Libby Gretz, Ph.D., of NIAMS; Gene G. Hunder, M.D., and Cornelia M. Weyland, M.D., of the Mayo Clinic; and Louis A. Healey, M.D. (retired).*

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