What is giant cell arteritis (GCA)?
Giant cell arteritis (GCA) is a form of vasculitis—a family of rare disorders characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Also called temporal arteritis, GCA typically affects the arteries in the neck and scalp, especially the temples. It can also affect the aorta and its large branches to the head, arms and legs. GCA is the most common form of vasculitis in adults over the age of 50.

The most common symptoms of GCA include persistent, throbbing headaches, tenderness of the temples and scalp, jaw pain, fever, joint pain, and vision problems. Early treatment is vital to prevent serious complications such as blindness or stroke.

GCA is typically treated with high doses of corticosteroids such as prednisone, sometimes in combination with other medications that suppress the immune system. Prompt treatment usually relieves symptoms, however GCA is a chronic condition with periods of relapse and remission, so ongoing medical care is usually necessary. Patients with GCA may also have symptoms of polymyalgia rheumatica (PMR), a closely related inflammatory disorder.

Causes
The cause of GCA is not yet fully understood by researchers. Vasculitis is classified as an autoimmune disorder—a disease which occurs when the body’s natural defense system mistakenly attacks healthy tissues. Researchers believe a combination of factors may trigger the inflammatory process. Studies have linked genetic factors, infectious agents, and a prior history of cardiovascular disease to the development of GCA.

Who gets GCA?
GCA is the most common form of vasculitis in older adults, affecting people over 50 years of age, with an average onset of 74 years of age. Women are more than twice as likely to get GCA than men. The condition is mostly seen in people of Northern European ancestry and is rare in other ethnic groups such as Asians and African Americans. GCA prevalence is estimated at 278 per 100,000 people in the United States over the age of 50.

Having PMR can put you at risk for getting GCA. Although estimates vary, approximately 15 percent of people with PMR develop GCA, and PMR symptoms occur in about 50 percent of patients with GCA.

How are GCA and PMR related?
GCA and PMR are considered linked inflammatory conditions that affect different parts of the body. Patients with either disease should be checked for symptoms of the other. PMR is the most common form of new-onset inflammatory disorder causing muscle and joint pain in elderly persons, and is characterized by severe pain and stiffness in the neck, shoulders and hips, which worsens in the morning.
GIANT CELL ARTERITIS

Symptoms of GCA
The most common symptoms of GCA are new, persistent headaches and tenderness of the temples due to inflammation of the temporal arteries on either side of the head. Severe throbbing pain is often accompanied by tenderness and swelling of the temporal artery and tenderness of the scalp.

Other common symptoms include:
- Flu-like symptoms at onset, including fatigue, fever and loss of appetite
- Jaw pain when chewing
- Sudden vision loss in one eye
- Vision loss in both eyes, or double vision
- Arm pain or weakness
- Aching and stiffness of shoulder or hip joints
- Dizziness
- Weight loss

Left untreated, serious complications can occur with GCA, including blindness, stroke or aortic aneurysm—an abnormal bulge in the wall of the aorta, which carries blood from the heart to the rest of the body. A burst aneurysm can be life-threatening.

Diagnosis
GCA can lead to vision loss early on, so it is essential that patients with suspected disease be evaluated promptly. Your doctor will consider a number of factors, including symptoms, medical history, physical exam findings, and results of blood tests and imaging studies. A biopsy of the arteries in one or both temples is usually obtained to confirm the diagnosis. If GCA is suspected, your doctor may begin steroid treatment even before the diagnosis is confirmed, to prevent complications such as vision loss.

Physical exam: Your doctor will check for tenderness, swelling, or decreased pulse in the temporal arteries on either side of the head, as well as tenderness in the temples or scalp. Also, decreased pulses in the arms or legs or discrepancy in blood pressure between any of the four extremities could suggest GCA.

Blood tests: The two main tests for GCA include the erythrocyte sedimentation rate (ESR), commonly called the “sed rate,” and the C-reactive protein test (CRP), both of which can detect inflammation. However, these tests are not conclusive on their own.

Imaging studies: For detailed images of the blood vessels, your doctor may order a magnetic resonance angiogram (MRA), which combines the use of magnetic resonance imaging (MRI) with contrast material; an ultrasound; or a PET scan, which uses a special dye injected into the arm to enhance detail in the images of your blood vessels.

Biopsy: The gold standard to confirm the diagnosis of GCA is biopsy of the temporal artery. A segment of the artery is surgically removed, and then examined under a microscope. In most cases of GCA, there will be evidence of inflammation that includes abnormally large cells—called giant cells—which give the
GIANT CELL ARTERITIS

disease its name. However, in some individuals the biopsy may be negative or normal, even though the disease is present.

Treatment
Until 2017, treatment of GCA was mostly limited to high-dose corticosteroids such as prednisone, which can bring dramatic relief of headache and other GCA symptoms. However, long-term use of steroids can cause serious side effects, and patients usually need to stay on high doses for at least a month, with most remaining on a lower dose up to two years or more.

In 2017, the FDA approved the use of the biologic drug tocilizumab to treat adults with GCA, signaling the first drug approved to treat this disease in more than 50 years. Biologic medications are complex proteins derived from living organisms. They target certain parts of the immune system to control inflammation. Tocilizumab targets an inflammation protein, interleukin 6 (IL-6), which is known to be involved in the disease GCA.

In clinical studies, tocilizumab—used in combination with steroids as they were being tapered off over a six-month period—helped patients achieve a sustained remission while significantly reducing their exposure to steroids.

Occasionally other medications are used to treat GCA. Methotrexate, a drug commonly used to treat rheumatoid arthritis, is sometimes used to help reduce relapses (flares) in GCA.

Side effects
The medications used to treat GCA have potentially serious side effects, such as lowering your body’s ability to fight infection, and potential bone loss (osteoporosis), among others. Therefore, it’s important to see your doctor for regular checkups. Medications may be prescribed to offset side effects. Infection prevention is also important. Talk to your doctor about getting a flu shot, pneumonia vaccination, and/or shingles vaccination, which can reduce your risk of infection.

Relapse
Even with effective treatment, relapse of GCA is common. Causes of relapse are not fully understood, although infections can be a trigger. If your initial symptoms return or you develop new ones, report them to your doctor as soon as possible. Regular doctor visits and ongoing monitoring of lab and imaging tests are important in detecting relapses early.

Your medical team
Effective treatment of GCA may require the coordinated efforts and ongoing care of a team of medical providers and specialists. In addition to a primary care provider, you may need to see the following specialists: rheumatologist (joints, muscles, and immune system); neurologist (brain and nervous system); ophthalmologist (eyes); or others as needed.

The best way to manage your disease is to actively partner with your health care providers. Get to know the members of your health care team. It may be helpful to keep a health care journal to track medications, symptoms, test results and notes from doctor appointments in one place. To get the most
GIANT CELL ARTERITIS

out of your doctor visits, make a list of questions beforehand and bring along a supportive friend or family member to provide a second set of ears and take notes.

Remember, it’s up to you to be your own advocate. If you have concerns with your treatment plan, speak up. Your doctor may be able to adjust your dosage, or offer different treatment options. It is always your right to seek a second opinion.

Living with GCA
Living with GCA can be challenging at times. Fatigue, pain, emotional stress and medication side effects can take a toll on your sense of well-being, affecting relationships, work and other aspects of your daily life. Sharing your experience with family and friends, connecting with others through a support group, or talking with a mental health professional can help.

Outlook
There is no cure at this time for GCA, but with early treatment and careful monitoring, most patients with GCA have a good prognosis. Symptoms generally improve within days of starting treatment, and with proper medical care the disease can run its course in one to two years. Left untreated, however, GCA can lead to serious complications including blindness, stroke and aneurysms. Newer medications, such as the biologic drug tocilizumab, offer hope for treating this disease with less reliance on steroids.

Clinical studies are ongoing at multicenter research centers, including the Vasculitis Clinical Research Consortium (VCRC), to better understand the risk factors and causes of vasculitis, investigate more effective and safer treatments, and work toward a cure. The Vasculitis Foundation encourages patients to consider participating in clinical research studies to help further understanding of vasculitis. Patients are also encouraged to join the Vasculitis Patient Powered Research Network (VPPRN), where they can provide valuable disease insight and information.

For more information on vasculitis research, visit: www.vasculitisfoundation.org/research
GIANT CELL ARTERITIS

About Vasculitis
Vasculitis is a family of nearly 20 rare diseases characterized by inflammation of the blood vessels, which can restrict blood flow and damage vital organs and tissues. Vasculitis is classified as an autoimmune disorder, which occurs when the body’s natural defense system mistakenly attacks healthy tissues. Triggers may include infection, medication, genetic or environmental factors, allergic reactions, or another disease. However, the exact cause is often unknown.

A Family of Diseases
• Anti-GBM (Goodpasture’s) disease
• Aortitis
• Behcet’s syndrome
• Central nervous system vasculitis
• Cogan’s syndrome
• Cryoglobulinemia
• Cutaneous small-vessel vasculitis (formerly hypersensitivity/leukocytoclastic)
• Eosinophilic granulomatosis with polyangiitis (EGPA, formerly Churg-Strauss syndrome)
• Giant cell arteritis
• Granulomatosis with polyangiitis (GPA, formerly Wegener’s)
• IgA vasculitis (Henoch-Schönlein Purpura)
• Kawasaki disease
• Microscopic polyangiitis
• Polyarteritis nodosa
• Polymyalgia rheumatica
• Rheumatoid vasculitis
• Takayasu’s arteritis
• Urticarial vasculitis

About the Vasculitis Foundation
The Vasculitis Foundation (VF) is the leading organization in the world dedicated to diagnosing, treating, and curing all forms of vasculitis. The VF provides a wide range of education, awareness and research programs and services.

To learn more, and get the most updated disease and treatment information, visit www.vasculitisfoundation.org

Connect with the VF on Social Media
• Instagram: vasculitisfoundation
• Twitter: @VasculitisFound
• VF Facebook Discussion Group: www.facebook.com/groups/vasculitisfoundation
GIANT CELL ARTERITIS

Ways to Get Involved
- Participate in research
- Join social media
- Host or participate in an event
- Attend a regional conference and/or symposium
- Give a donation toward long-term solutions
- Become an advocate for yourself, or for others
- Share your journey

Vasculitis Foundation Mission
Building upon the collective strength of the vasculitis community, the Foundation supports, inspires and empowers individuals with vasculitis, and their families, through a wide range of education, research, clinical, and awareness initiatives.

Join the VPPRN!
The Vasculitis Patient-Powered Research Network (VPPRN) seeks to improve the care and health of patients with vasculitis by exploring research questions that matter most to patients, and by advancing medical knowledge about vasculitis. For more information, visit: www.VPPRN.org

The Vasculitis Clinical Research Consortium (VCRC) is an integrated group of academic medical centers, patient support organizations, and clinical research resources dedicated to conducting clinical research in different forms of vasculitis. For more information, visit: www.rarediseasesnetwork.org/cms/vcrc

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