Giant Cell Arteritis

What is Giant Cell Arteritis (GCA)?

Giant cell arteritis is a condition that can cause vision loss, new persistent headaches, scalp tenderness, and jaw pain with chewing. It is due to inflammation of blood vessels primarily of the head and neck. GCA is sometimes called “temporal arteritis,” as it frequently affects the blood vessels in the temples, causing headaches. GCA can affect other blood vessels in the body.

GCA almost only affects people older than 50 years of age, with an average age of 70 years. It affects both men and women and can occur in any race or ethnicity. The cause of GCA is unknown, though increasing age increases the risk of developing this condition.

How Does Giant Cell Arteritis Affect My Vision?

Because the inflammation in GCA causes blood vessels to narrow, blood may not get to the optic nerve or the retina, causing permanent damage. As a result, vision loss in one or both eyes may occur in up to 20-50% of patients. About 1/3 of patients who develop vision loss have episodes of transient vision loss (amaurosis fugax) about 7-10 days prior to the permanent vision loss. For this reason, early diagnosis and prompt treatment is extremely important. The vision loss can be very severe, and is almost always irreversible, even with treatment. Some people with GCA may have intermittent double vision because of reduced blood flow to the muscles or nerves which control eye movement.

What are Other Symptoms of Giant Cell Arteritis?

Because GCA can affect any large or medium-sized blood vessels in the body, there are many different symptoms. The most common initial symptom is new persistent headache, which is present in 40-90% of patients.

A less common, but important symptom is pain in the jaw, ear, tongue, throat, or neck while chewing or swallowing. This symptom, called jaw claudication, is thought to be caused by a decrease in blood flow to the muscles for chewing and swallowing.
Another common symptom of GCA is scalp tenderness, which most often is noted when brushing or washing hair, or sleeping on one side of the head. Thickened and inflamed blood vessels in the temple may even be seen, as shown in the figure below.

Many people do not have specific symptoms and may only have **fatigue**, generalized **malaise**, loss of appetite, and unintentional **weight loss**. Some people have difficulty with their memory or signs of dementia that could be related to giant cell arteritis. Other neurological symptoms include numbness or tingling, hearing loss, or dizziness. In severe cases, people may have heart attacks, strokes, or kidney damage.

**Why Do I Need to See a Neuro-ophthalmologist?**

**Neuro-ophthalmologists are the experts with the most experience and knowledge about GCA that affects vision.** Though other physicians also may see and manage people with GCA, neuro-ophthalmologists receive specific training to diagnose and assist in taking care of patients who have developed vision loss from GCA. Many specialists may be involved in your treatment:

- **A neuro-ophthalmologist** makes the diagnosis of GCA, monitors your visual status, and guides treatment based on your eye exam, symptoms, and lab values.

- **A rheumatologist** is an expert in inflammatory disease and use of anti-inflammatory medications and may co-manage the disease alongside the neuro-ophthalmologist in patients with vision loss and may manage patients with GCA with no vision loss.

- **An internal medicine or family medicine** physician monitors for potential side effects of your treatment and evaluates for GCA involvement of the body, especially the aorta.
Your neuro-ophthalmologist monitors your central and peripheral vision as well as your color vision, your eye movements, and your general eye health. During your exam he/she may perform several tests. The **visual acuity** measures the sharpness of your central vision (ability to read the eye chart). **Color vision** can be tested using various plates with figures or numbers on them. A **visual field** test maps your field of vision. Other tests may include a **dilated eye exam**, **photos**, and a special imaging test called **optical coherence tomography (OCT)**.

**How is Giant Cell Arteritis Diagnosed?**

If your doctor is concerned that you have GCA, you will need to get blood testing right away to screen for inflammation including **Erythrocyte Sedimentation Rate (ESR)**, **C-reactive protein (CRP)**, and **Complete Blood Count (CBC)**. People who have normal blood tests are much less likely to have GCA, though there are some exceptions.

If your tests are concerning for GCA, your doctor will start you on a **high dose of oral steroids** (i.e., prednisone), and plan for you to have a biopsy of an artery under your scalp (temporal artery biopsy) within 7-10 days. In some situations, your doctor may recommend that you get IV steroids for a few days before starting you on oral steroids.

A **temporal artery biopsy** is required for definitive diagnosis of giant cell arteritis. Some people need to have more than one biopsy. It is uncommon for the biopsy to be negative in patients in whom there is a high clinical suspicion of GCA. Thus, if, despite a negative temporal artery biopsy, your doctor believes you have a high risk for GCA, he/she may still recommend that you be treated with steroids.

**How is Giant Cell Arteritis Treated?**

GCA is primarily treated with **high-dose steroids**. Because this condition usually lasts for a long time (usually 1-2 years, but in some cases, up to 10+ years), people with GCA must be on steroids for a very long time. Most people with GCA have improvement of pain and headache within 24-72 hours after starting treatment. Though some people experience some improvement in their vision once treatment is begun, most people do not and may even have worsening vision despite treatment. Thus, the reason to treat as soon as possible is to prevent further visual loss or other complications of GCA (eg, stroke, heart attack).
Once you begin steroid treatment, your doctor will continue to check your lab tests (ESR and CRP) and follow your symptoms over the next several months to years. Depending on how well you are feeling and the levels of inflammation noted on your lab tests, your doctor will gradually lower the steroid dose. Most patients eventually can stop steroids completely.

Though other anti-inflammatory medications other than steroids have been studied in the treatment of giant cell arteritis, most have not been proven to be as effective. If a patient with GCA has significant side effects from steroids, treatment with other medications that suppress the immune system may be possible. However, these medications may have side effects as well. The choice of long term medication for GCA should be made in conjunction with your neuro-ophthalmologist, a rheumatologist, internist, family practitioner, or a combination of these doctors. In 2018, the FDA approved a medication called tocilizumab that, if taken in addition to steroids, can shorten the amount of time a patient is on the steroids.

What is My Prognosis?

With early treatment, GCA has a good life prognosis. Unfortunately, vision loss is typically permanent regardless of treatment. The goal of treatment is to avoid vision loss in the other eye, to limit worsening of vision, and to prevent damage to other organs.

Additional Resources:

- American Academy of Ophthalmology EyeWiki: [http://eyewiki.org/Giant_Cell_Arteritis](http://eyewiki.org/Giant_Cell_Arteritis)
- Vasculitis Foundation: [https://www.vasculitisfoundation.org/education/forms/giant-cell-arteritis/](https://www.vasculitisfoundation.org/education/forms/giant-cell-arteritis/)
- Actemra (tocilizumab) for GCA: [https://www.actemra.com/gca.html](https://www.actemra.com/gca.html)