

Giant cell arteritis (temporal arteritis)

What is giant cell arteritis?

Giant cell arteritis (GCA), also known as temporal arteritis, is a condition that can be a great threat to your sight. It affects large and medium sized arteries in your body, in particular, the arteries in your neck and head. GCA is sometimes referred to as temporal arteritis, as one of the more commonly affected arteries is the temporal artery at the side of your forehead (temple).

In GCA, the artery walls become inflamed (arteritis) with large inflammatory cells (giant cells). The inflammation causes the affected arteries to become swollen, greatly restricting the blood flow through them. If the blood flow becomes too restricted, the structures supplied by these arteries will be affected. If the artery which supplies the eye is affected, then this can cause sight loss in that eye.

How do we see?

Light enters our eyes by passing through the cornea, pupil, (the hole in the middle of the iris), and lens so that it is sharply focused onto the retina lining the back of our eye. The retina is made up of light-sensitive cells that convert light into electrical



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impulses which then travel along the optic nerve to our brain. Our brain processes these signals so that we are able to “see” the world around us.

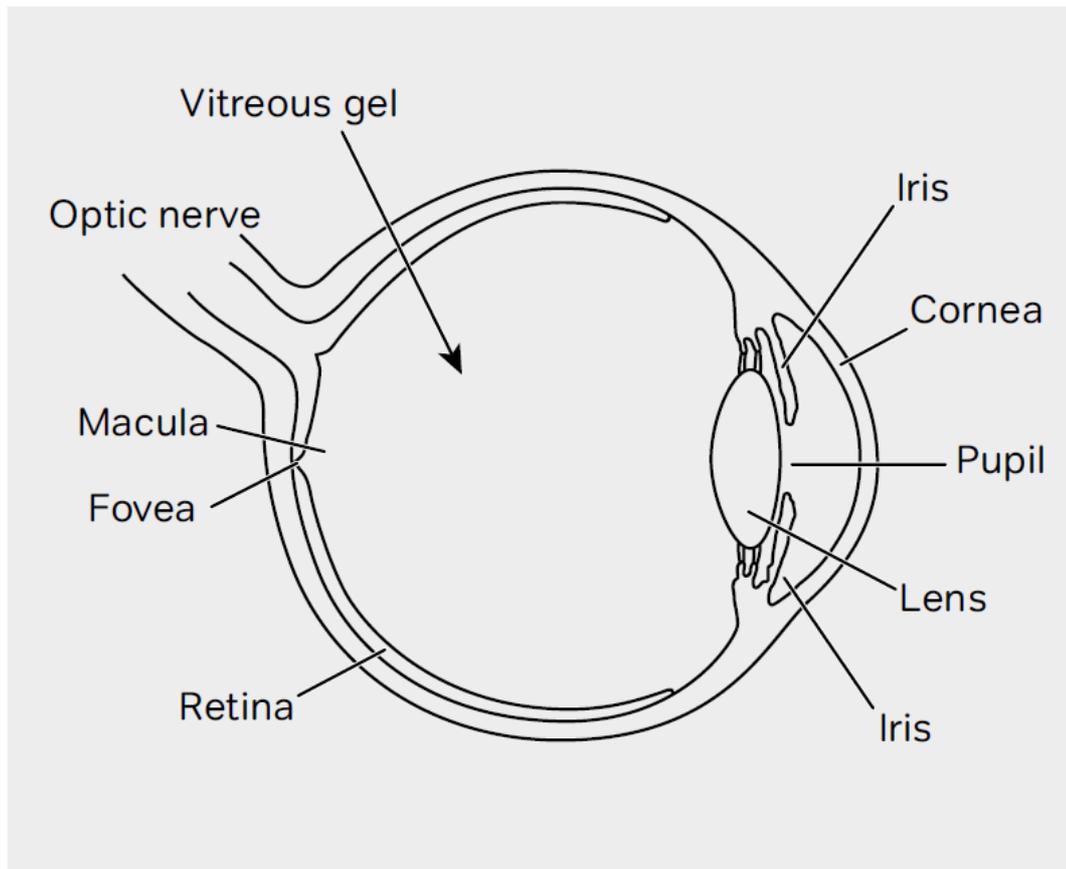


Image description: Diagram of cross section of eye (labels: cornea, pupil, iris, lens, vitreous gel, retina, macula, fovea, optic nerve)

How does giant cell arteritis affect vision?

Arteries are blood vessels that carry blood full of oxygen from the heart to the different tissues in the body. A good blood supply with oxygen is essential for these tissues to survive. If an artery becomes inflamed (arteritis) and restricts the blood supply to a particular tissue, the function of that tissue will be affected because not enough oxygen is getting to the cells. This reduction in blood supply is known as ischaemia, which results in a lack of oxygen, known as hypoxia.

The ophthalmic artery supplies blood to the various tissues of the eyes, including the optic nerve, and this artery is prone to being

affected by GCA. If there is ischaemia of the optic nerve, there can be sudden and severe sight loss as a result. Unfortunately, the optic nerve cannot recover from the lack of blood supply, so the sight loss is usually permanent.

What causes giant cell arteritis?

The cause of GCA is unknown. It is not a common condition and it rarely affects people under the age of 50. It can affect people from 60 years of age, but more usually, a person will be in their 70s when they experience GCA. Women are more commonly affected by GCA than men.

What are the signs and symptoms of giant cell arteritis?

Headache

One of the first signs of GCA is a severe headache which may come on suddenly or gradually. It is often at the side of your forehead, at your temples, and it may affect one or both sides of your head. The headache associated with GCA will probably not feel like any headache you've had before.

Scalp and temple tenderness

You may also feel that your temples and scalp are tender to touch, so that combing or brushing your hair feels uncomfortable.

Pain around the eyes

Some people with GCA also experience pain around their eyes within the eye sockets.

Pain on chewing

Another early sign of GCA is pain on chewing. This can be a cramp-like pain in your jaw which can make you stop eating. You may hear this referred to as jaw claudication and it is due to ischaemia in the muscles that help you to chew. Ischaemia can also cause pain in your tongue or mouth.

If you have any of the symptoms outlined above, you should contact your GP urgently.

Vision disturbances

You may experience a temporary loss of vision in one eye which returns after a while. This is called amaurosis fugax and it is another sign of ischaemia. You may also experience double vision that you haven't had before because the muscles controlling your eye movements are affected.

Loss of vision (arteritic anterior ischaemic optic neuropathy)

About 30-50 per cent of people who have untreated GCA will develop a permanent and severe loss of vision in one eye. Without immediate treatment, about a third of these people will develop sight loss in the other eye, often within a week of the first eye.

Therefore, it is vital that you see an ophthalmologist (hospital eye doctor) immediately in A&E if you experience sudden sight loss in one eye.

If you have sight loss caused by GCA, your ophthalmologist may refer to this eye condition as arteritic anterior ischaemic optic neuropathy (arteritic AION), so called because the arteritis of GCA has led to ischaemia, causing damage the optic nerve (optic neuropathy).

Loss of vision due to GCA is usually permanent and severe. Very rarely, prompt treatment with a high dose of steroids can lead to a partial recovery of sight.

Sudden loss of vision should always be investigated immediately by an ophthalmologist, even if your sight returns after a while, and even if you are not experiencing any of the other symptoms of GCA. Some people lose their sight without having previously noticed any other symptom of

GCA. If you experience sudden sight loss, **you should go to A&E or to a hospital eye casualty department straight away to be seen urgently so that if treatment is needed, it can be given as soon as possible.**

Other general health issues

Some people with GCA also experience weight loss, tiredness, night sweats, fever and depression.

Usually, people have some symptoms of GCA before their sight is affected, so it's very important to see your GP straight away if you experience any of them.

How is giant cell arteritis diagnosed?

If it is suspected that you have GCA, you will be given treatment straight away, even before all the results of the diagnostic tests are obtained. This is because it is important to try and reduce any possible inflammation as quickly as possible, and to prevent blood flow in other arteries becoming restricted.

There are several things that your ophthalmologist or GP will check:

- The artery in your temple (the superficial temporal artery) may look and feel raised so that it cannot be flattened down on your head and you may not be able to feel a pulse there when you touch it.
- Blood tests which check for signs of inflammation in your blood:
 - (1) **Erythrocyte sedimentation rate (ESR).** A sample of your blood is taken and placed in a test tube with a chemical to stop it clotting. The red cells (erythrocytes) in your blood gradually sink down in the tube, like sediment. The distance in millimetres that they fall in

one hour is measured (mm/hr). When there is inflammation somewhere in your body, certain proteins will be found in your blood. These proteins bind to your red blood cells, making them stick together in clumps which are heavier than individual cells. These heavier clumps of cells will fall further in the test tube in an hour, giving a higher ESR result. Your doctor will be able to explain your ESR result and whether or not it suggests you have GCA. A measurement of 50mm/hr or more is considered to be high.

- (2) **C-reactive protein (CRP).** This test measures the level of C-reactive protein (CRP) in a sample of your blood. CRP is released from your liver into your bloodstream when inflammation starts within your body or when a long-standing inflammation flares up. CRP is not typically found in your blood when you have no inflammation in your body. Your doctor will be able to tell you whether your CRP level indicates that you may have GCA.
- (3) **Plasma viscosity (PV).** This test gives very similar information as the ESR test and may be done instead of the ESR test at some hospitals. When there is inflammation in your blood, there is an increase in certain proteins. This causes the viscosity (the thickness of the liquid) of the plasma to rise. A higher result means a higher level of inflammation present in your body.

These blood tests are non-specific tests, meaning they don't pinpoint exactly where inflammation is in your body. Also, in some people the results are inconclusive. Therefore, other tests such as a temporal artery biopsy or an ultrasound scan are used to confirm the diagnosis.

- **Temporal artery biopsy.** A small sample of the artery is taken from your temple and examined under a microscope by a specialist doctor called a histopathologist. They look for signs of inflammation in the tissue sample that indicate GCA is present.
- **An ultrasound scan** of the superficial temporal artery can indicate whether or not the artery walls are swollen.

What is the treatment for giant cell arteritis?

Steroids

If it is suspected that you have GCA, emergency treatment will be started immediately with a high dose of steroids.

Steroids are drugs that reduce inflammation in the body and, if GCA is suspected, you will be given steroid tablets to take even if your test results have not come through yet. The aim of this emergency treatment is to prevent sight loss and other general health concerns such as stroke and heart attack.

You will probably have to take steroid tablets for between one and two years or even longer, with the dose being reduced gradually over time as the inflammation subsides. It is possible to experience relapses of the condition (where the inflammation flares up again) so you may have to restart medication again or your dose may have to be increased.

Biological therapy

A treatment involving biological therapy has recently been approved for treating GCA. Biological therapy treatments involve the use of drugs that alter a biological process occurring in your body.

With GCA, a number of proteins are produced by the body. One particular protein has been found to be very high in people who

have GCA. Biological therapy treatment blocks the action of this protein so that the inflammation is reduced.

It is given as a weekly injection into the skin and you may need to have these injections for up to a year, but your specialist will advise you about this. It may be possible for you to self-inject your treatment at home, but you will be advised as to whether this is appropriate for you.

These injections can be given in addition to your steroid tablets or on their own when steroids have been stopped. Steroids can lead to several unwanted side effects when taken at a high dose for a long time. By adding biological therapy to your treatment, lower doses of steroids can be given while still controlling your GCA.

The advantages of adding biological therapy to your treatment is that it can reduce the amount of steroid you need to take and can also reduce the amount of time to relapse.

It is possible to experience relapses of the condition, so treatment may need to be restarted or increased.

Who will manage my condition?

If your GP suspects that you have GCA, they will carry out the necessary blood tests but treat you immediately with steroids as a precautionary measure. Then they will refer you to a rheumatologist or ophthalmologist to confirm your diagnosis.

A rheumatologist is a hospital doctor who treats people with inflammatory conditions such as GCA and they will manage your condition until they are happy to return you to the care of your GP.

If your sight has been affected, you will also be examined by an ophthalmologist, who will diagnose the effects of GCA on your optic nerve. The ophthalmologist will be the first specialist to

examine you if you go to A&E with sight loss in the first place. The ophthalmologist can arrange blood tests and a biopsy to diagnose GCA. They can also refer you to the rheumatologist for further tests and for treatment.

What if I already have sight loss due to giant cell arteritis?

If it is suspected that your sight loss is due to GCA, you may be admitted to hospital for treatment or given treatment to use at home straight away.

If you are admitted, you may be given a high dose of steroids directly into your bloodstream via a drip over the next few days. If you are not admitted, you'll be given high dose steroid tablets to use at home and then followed up by the hospital. The high dose steroids are given to try and bring down the swelling in your arteries as quickly as possible. Unfortunately, there is no treatment available that can bring back the sight loss caused by GCA so it is unlikely that the vision you have lost will return. The aim of this treatment is to prevent further sight loss in your other eye.

How will I manage with sight loss due to giant cell arteritis?

It is rare to lose sight in both eyes at the same time due to GCA. However, if you have lost your sight in one eye, it may take some time to get used to this. You use two eyes together to see in three dimensions (3D) and to judge distances. When you have sight loss in one eye, you may misjudge steps and kerbs, and have difficulty judging the position of things, such as pouring liquids into cups. However, over time your brain will adapt to these changes by taking more notice of the vision in your better eye, so tasks you may have found difficult initially start to get easier.

Can I still drive?

You may be able to continue driving a car or motorcycle if the vision in your other eye is unaffected by other eye conditions, and can meet the visual requirements for driving. You are required by law to tell the Driver and Vehicle Licensing Authority (DVLA) if you have any eye conditions which may affect your vision in both eyes. Ask your optometrist or your ophthalmologist for advice about whether your sight meets DVLA standards and whether you can continue driving. Even if you are told that your sight does meet DVLA standards, you may be advised to wait until you have adapted to having poorer vision in one eye before you resume driving.

What if both my eyes are affected by sight loss?

If both eyes are affected by GCA or the affected eye was your good eye and you have another sight problem in your other eye, you may need to make changes or use aids to make the most of your remaining sight. This may mean making things bigger, using brighter lighting or using colour to make things easier to see. We have a series of leaflets with helpful information on living with sight loss, including how to make the most of your sight. You can find out more about our range of titles by calling our Helpline 0303 123 9999.

You should ask your ophthalmologist, optometrist or GP about low vision aids and getting a low vision assessment. During this assessment with a low vision specialist, you'll be able to discuss the use of magnifiers and aids to help you to see things more clearly.

You should also ask your ophthalmologist whether you're eligible to be registered as sight impaired (partially sighted) or severely sight impaired (blind). Registration can act as your passport to expert help and sometimes to financial concessions. Even if you aren't registered, a lot of this support is still available to you.

Local social services should be able to give you information on staying safe in your home and getting out and about safely. They should also be able to offer you some practical mobility training to give you more confidence when you are out.

Are there any other conditions associated with giant cell arteritis?

About half the people with GCA have another condition called polymyalgia rheumatica (PMR) where the blood vessels supplying some of the larger muscles in your body are inflamed. The muscles particularly affected by PMR are those in your shoulders, neck, hips and back. These muscles can be painful and stiff, particularly on waking. It is possible to have GCA without having PMR as well. It is thought they might be different conditions with the same underlying cause and they are both treated with steroids.

Coping

If you have been diagnosed with GCA with or without sight loss, it's normal to find yourself worrying about the future and how you will manage if your vision changes.

It can sometimes be helpful to talk about these feelings with someone outside of your circle of friends or family. At RNIB, we can help with our telephone Helpline and our Sight Loss Counselling team. Your GP or social worker may also find a counsellor for you if you feel this might help.

Your eye clinic may also have an Eye Clinic Liaison Officer (ECLO), who can be on hand to provide you with further practical and emotional support about your eye condition.

Further help and support

If you have questions about anything you've read in this factsheet, or just want to speak to someone about your eye

condition, please get in touch with us. We're here to support you at every step.

Our Helpline is your direct line to the support, advice and services you need. Whether you want to know more about your eye condition, buy a product from our shop, join our library, find out about possible benefit entitlements, or be put in touch with a trained counsellor, we're only a call away.

It's also a way for you to join RNIB Connect, our community for anyone affected by sight loss. RNIB Connect is free to join and you'll have the chance to meet other people with similar experiences in our helpful, welcoming and supportive community.

Give us a call today to find out how we can help you.

RNIB Helpline
0303 123 9999
helpline@rnib.org.uk

We're ready to answer your call Monday to Friday 8am to 8pm and Saturday 9.30am to 1pm.

You can also get in touch by post or by visiting our website:

RNIB
105 Judd Street
London WC1H 9NE
rnib.org.uk

Other useful organisations

PMRGCA UK (Polymyalgia Rheumatica & Giant Cell Arteritis UK) is a charity supporting people affected by PMR or GCA.
Helpline: 0300 111 5090
Address: BM PMRGCAuk, London WC1N 3XX
Website: pmrgca.co.uk

PMR-GCA Scotland is a charity supporting people affected by PMR or GCA in Scotland
Support Line: 0300 777 5090
Email: Secretary@PmrGcaScotland.com
Address: PMR GCA Scotland, 7 Hamilton Place, Perth, PH1 1BB
Website: www.PmrGcaScotland.com

Vasculitis UK

Helpline: 0300 365 0075
Email: john.mills@vasculitis.org.uk
Address: John Mills, West Bank House, Winster, Matlock, DE4 2DQ
Website: vasculitis.org.uk

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