Referral Support Service

Rheumatology & Ophthalmology

OP01
Giant Cell Arteritis (GCA) / Temporal Arteritis

Definition

Giant cell arteritis is an immune-mediated form of vasculitis affecting medium and large vessels. While it can affect all medium to large arteries, the involvement of the temporal artery and the ability to detect physical changes on examination give rise to the alternative name of temporal arteritis. It is the most common form of vasculitis in adults. It is more common in females, age > 50, Caucasians and those with a history of Polymyalgia Rheumatica

GCA should be suspected if:

- Age at disease onset **>50 years**
- **New headache** - New onset of or new type of localized pain in the head
- **Temporal artery abnormality** - Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
- **Elevated CRP**
- **Abnormal artery biopsy** - Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

For purposes of classification, a patient shall be said to have giant cell (temporal) arteritis if at least 3 of these 5 criteria are present.

Other specific and important symptoms & signs

Visual disturbance (see below)
Jaw claudication
Scalp tenderness
Past history of PMR

Other symptoms and signs suggestive of giant cell arteritis:

- **Systemic features** (fever, fatigue, anorexia, weight loss, and depression) — affect most people. Fever is usually low grade, but may occasionally be higher.
- **Features of polymyalgia rheumatica**— present in about 40% of cases.
- **Scalp tenderness** in about 50% of people.

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- **Intermittent jaw claudication** — occurs in nearly half of people with giant cell arteritis, causing pain in the jaw muscles while eating.
- **Visual disturbances** — permanent partial or complete loss of vision in one or both eyes occurs in up to 20% of people and is a common early symptom. Typically it is described as a feeling of a shade covering one eye, which can progress to total blindness. The eye is not painful.
    - Double vision and visual field defects may occur. On fundoscopy there may be pallor and oedema of the optic disc, and 'cotton-wool' patches and (less ?commonly) small haemorrhages in the retina — these features are usually seen following loss of vision.
    - Untreated, the second eye is likely to become affected within 1–2 weeks, although it can be affected within 24 hours.
    - **Once visual impairment is established, it is usually permanent.**

- **Neurological features** — occur in about 30% of people and include: **Transient ischaemic attack or stroke, Mono- or polyneuropathy** and Upper **cranial nerve palsies**
- **Peripheral arthritis** and distal swelling with pitting oedema — occurs in about 25% of people. The swelling is most prominent over the dorsum of the hands and wrists, the ankles, and the tops of the feet.
- **Respiratory symptoms** — for example cough, sore throat, and hoarseness (present in about 10% of people).

Document the person's symptoms, signs, and level of function both before and after the onset of the condition to use as a baseline to compare response to treatment.

Manage Red Flags

- GCA can cause preventable blindness
- Treat whilst waiting for diagnostic confirmation
- If there is visual impairment — arrange an urgent (same day) assessment by an ophthalmologist.
- Abrupt cessation of long term steroids can precipitate an Addisonian crisis! Warn patients about this.
- Provide patients with a steroid card / information
- Share care with rheumatologists to ensure careful prescribing of steroids.

General Points

If there are visual symptoms or visual impairment — arrange an urgent (same day) assessment by an ophthalmologist. Phone the eye department if visual symptoms (link only viewable on NHS connected computers)

If there are NO visual symptoms or visual impairment: suspected GCA needs urgent referral for TA biopsy via the maxillofacial surgeons – see pathway below.

- Arrange urgent blood tests – that day or the next day
- Start high dose steroids as detailed below

A positive result confirms the diagnosis, but a negative result does not always rule it out.

If the biopsy is POSITIVE refer to rheumatology urgently via RSS

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If the biopsy is NEGATIVE reduce steroids appropriately and monitor patient's symptoms
If a patient is deemed to need hospital admission due to frailty refer to Ambulatory Medical Care (telephone and internal pathway will be followed)

Investigations

Where there is clinical suspicion arrange a biopsy at the same time as ordering blood tests

- C-reactive protein (CRP) The CRP level is typically elevated and may be a more sensitive indicator of inflammation than ESR in some people with giant cell arteritis. Do not order an ESR as well.
- Full blood count - Normochromic normocytic anaemia and an elevated platelet count are common and indicate chronicity.
- U+E
- Liver function tests

How to get a Temporal Artery Biopsy

Manage patients as detailed above

Arrange urgent bloods if not yet done

Send email referral for temporal artery biopsy (TAB), same day as commencing oral steroids, to: yhs-tr.tabreferrals@nhs.net

Head and Neck Admin team (Monday to Friday) will contact Max Fac 1st on call to arrange TAB within 10 days of commencement of oral steroids (specific TAB slot or acute theatre list).

If there are visual symptoms or visual impairment — arrange an urgent (same day) assessment by an ophthalmologist. Phone the eye department if visual symptoms (link only viewable on NHS connected computers)

If biopsy is positive GPs maintain responsibility to continue steroids and refer urgently via RSS to Rheumatology. Rheumatology will advise patients and GPs on long term steroid management

Seek rheumatology input via A&G if biopsy and / or CRP negative but clinical suspicion is high. Advise patients on maintaining or carefully reducing steroids as appropriate.

The diagnosis is rare with normal acute phase markers and this should remain a determinant of management. Local audit of biopsy results showed there is a high pick up rate - about 20% (compared to single figures in other centres), so GPs are sending the right patients and the surgeons are skilled at taking biopsies of? the artery.

It does mean that 80% have a headache that the rheumatologists cannot help with, i.e. the management of patients who *do not* have Giant Cell Arteritis.

A normal bloods and biopsy rules out disease with a high degree of confidence. Whilst there are always exceptional cases GPs should routinely follow headache guidance for these patients where biopsy is negative. The rheumatologists are happy to discuss cases via Advice and Guidance if needs be.

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Ensure **oral corticosteroids** are started immediately while awaiting the biopsy. See regime below.

Consider alternative conditions (see differential diagnoses below) that may present with similar features to giant cell arteritis, especially if there is a poor response to oral corticosteroids after 48 hours. If appropriate, arrange tests to exclude them.

Treatment:

Oral prednisolone (not enteric coated or soluble):
For people with visual symptoms — 60 mg as a one-off dose (they should be seen by an ophthalmologist the same day).
For people without visual symptoms — 40 to 60 mg daily (minimum 0.75 mg/kg).

Assess the person's response to prednisolone within 48 hours (the response of symptoms to corticosteroids is usually rapid).

If response to prednisolone is poor, seek specialist advice and consider an alternative diagnosis (see differential below).

Advise the person:

1. To seek urgent (same-day) medical attention if they develop any visual disturbances (such as visual loss, double vision, or visual field defects).
2. That the dose of prednisolone is normally reduced very slowly over several months. Treatment is often required for 1–2 years, but people may require low doses of corticosteroids for several years.
   A NICE suggested schedule:
   a. Continue high-dose prednisolone 40–60 mg daily until the person has been in remission for 4–8 weeks, then
   b. Reduce the dose by 10 mg every 2 weeks to 20 mg daily, then
   c. Reduce the dose by 2.5 mg every 2–4 weeks to 10 mg daily, then
   d. Reduce the dose by 1 mg every 1–2 months, provided there is no relapse.
3. Relapses may occur while taking prednisolone and are more common while the dose is being reduced.
4. Frequent follow-up visits are required to monitor for relapses and adverse effects of corticosteroids.

See BNF section on Patient and carer advice under Prednisolone:

Give the patient a steroid emergency card, see below, and warn them not to abruptly stop their pills or run out of them. Community pharmacies and GP practices should ensure they supply Steroid Emergency Cards to relevant patients. They should also replace those lost by patients or which become damaged. Supplies of the card can be ordered from Primary Care Support England via their online portal . A National Patient Safety Alert on this was published in August 2020 The National Reporting and Learning System (NRLS) in a recent two-year period identified four deaths, four patients admitted to critical care, and around 320 other incidents describing issues with steroid replacement therapy for patients with adrenal insufficiency or emergency treatment for adrenal crisis. At present it is advised by the Society for Endocrinology that the old steroid card is still given to patients. These too are available through Primary Care Support England.
Consider co-prescription of bone protection e.g. oral bisphosphonate and calcium and vitamin D if level of suspicion is high or high risk of osteoporosis and PPI due to risk of gastric irritation whilst on steroids.

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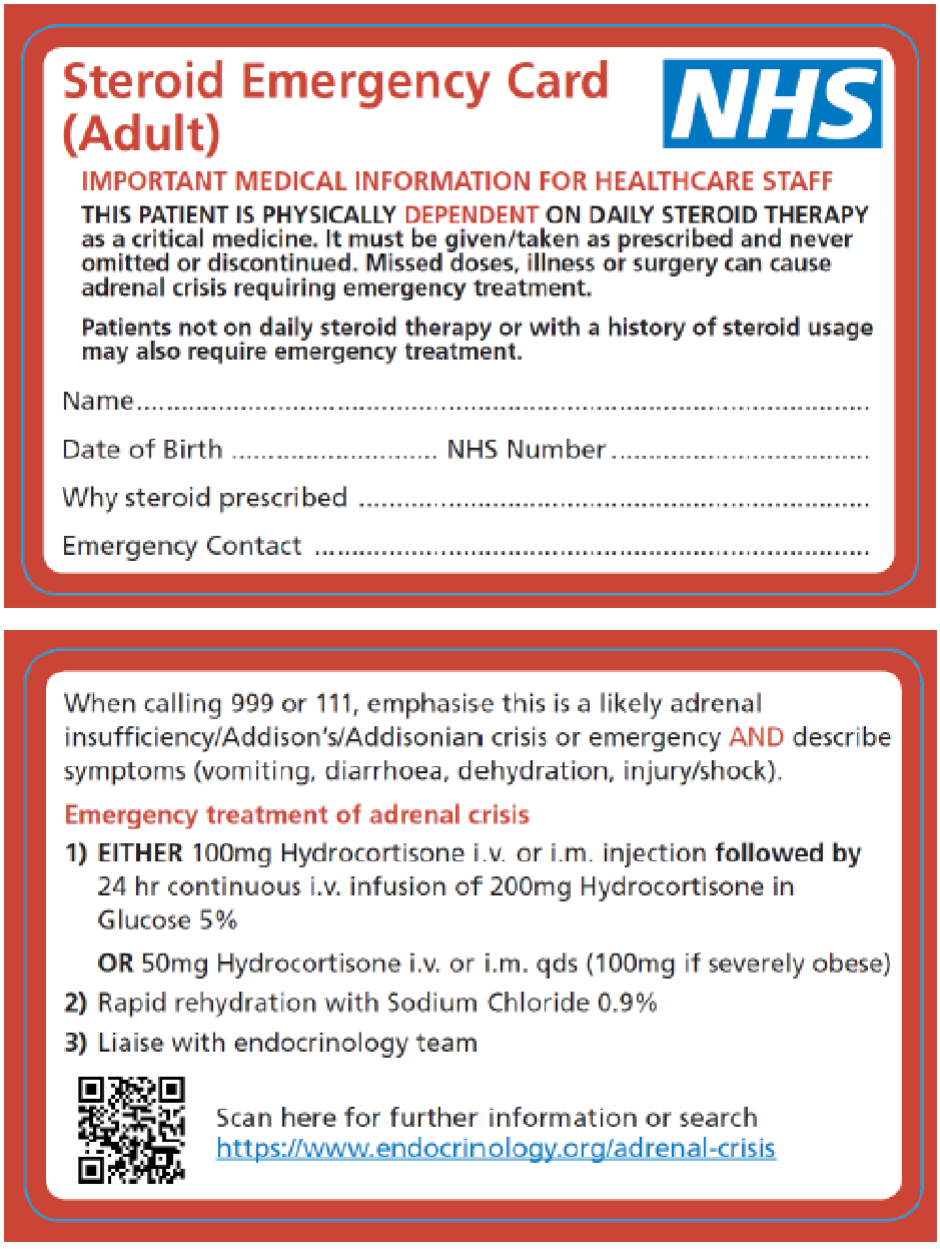
Aspirin 75 mg daily is recommended, unless there are contraindications such as active peptic ulceration or a bleeding disorder. It is uncertain how long a person with giant cell arteritis should remain on aspirin. If not indicated for other reasons (such as coexistent cardiovascular disease), consider seeking specialist advice regarding duration of treatment.

Proton pump inhibitor (e.g. lansoprazole 30mg daily) for gastrointestinal protection.

This is the new card



Steroid Emergency Card (Adult)

NHS

IMPORTANT MEDICAL INFORMATION FOR HEALTHCARE STAFF

THIS PATIENT IS PHYSICALLY DEPENDENT ON DAILY STEROID THERAPY as a critical medicine. It must be given/taken as prescribed and never omitted or discontinued. Missed doses, illness or surgery can cause adrenal crisis requiring emergency treatment.

Patients not on daily steroid therapy or with a history of steroid usage may also require emergency treatment.

Name.....

Date of Birth ..... NHS Number .....

Why steroid prescribed .....

Emergency Contact .....

When calling 999 or 111, emphasise this is a likely adrenal insufficiency/Addison's/Addisonian crisis or emergency AND describe symptoms (vomiting, diarrhoea, dehydration, injury/shock).

Emergency treatment of adrenal crisis

1) **EITHER** 100mg Hydrocortisone i.v. or i.m. injection **followed by** 24 hr continuous i.v. infusion of 200mg Hydrocortisone in Glucose 5%

   OR 50mg Hydrocortisone i.v. or i.m. qds (100mg if severely obese)

2) Rapid rehydration with Sodium Chloride 0.9%

3) Liaise with endocrinology team

Scan here for further information or search https://www.endocrinology.org/adrenal-crisis

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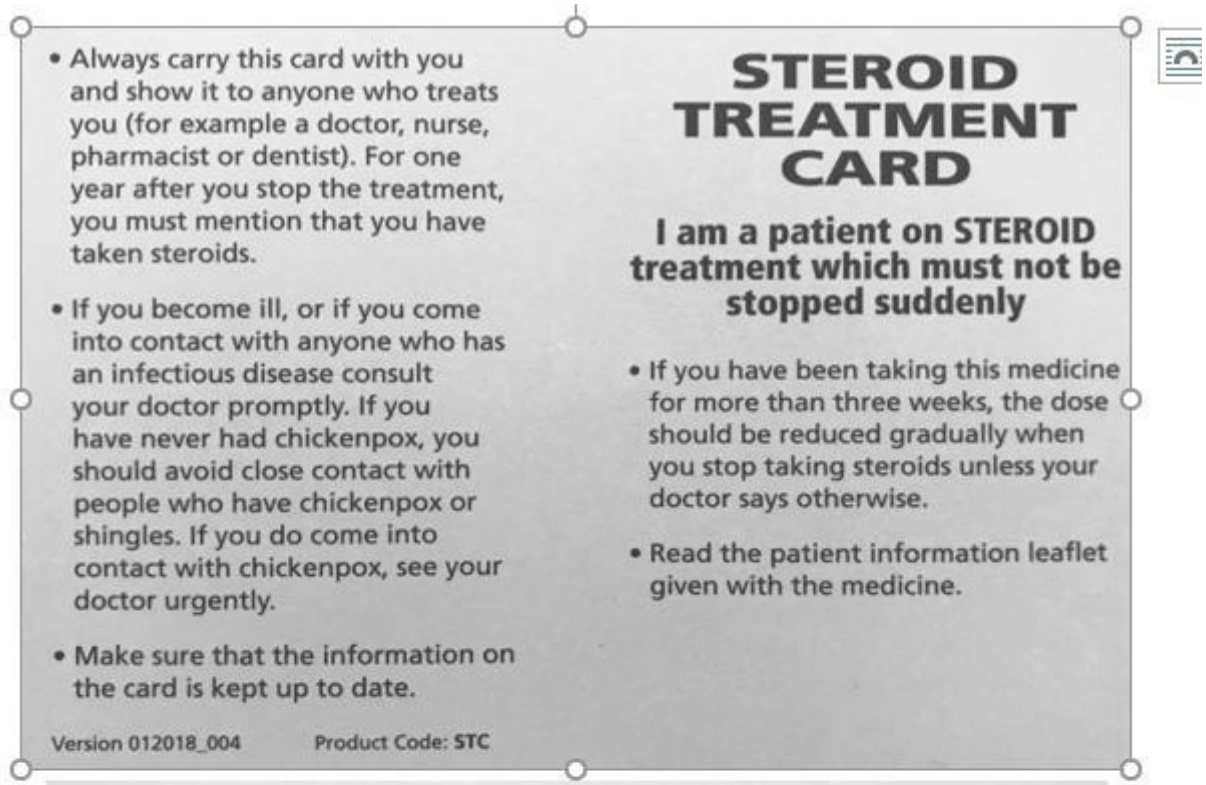

- Always carry this card with you and show it to anyone who treats you (for example a doctor, nurse, pharmacist or dentist). For one year after you stop the treatment, you must mention that you have taken steroids.
- If you become ill, or if you come into contact with anyone who has an infectious disease consult your doctor promptly. If you have never had chickenpox, you should avoid close contact with people who have chickenpox or shingles. If you do come into contact with chickenpox, see your doctor urgently.
- Make sure that the information on the card is kept up to date.

Version 012018_004 Product Code: STC

STEROID TREATMENT CARD

I am a patient on STEROID treatment which must not be stopped suddenly

- If you have been taking this medicine for more than three weeks, the dose should be reduced gradually when you stop taking steroids unless your doctor says otherwise.
- Read the patient information leaflet given with the medicine.

Name	
Address	
Tel No	
GP	
Why steroid is used	
Emergency contact	
NHS number	

Date	Drug	Dose

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Potential differential diagnoses

- **Herpes zoster.**
  - For more information, see the CKS topic on Shingles.
- **Cluster headache or migraine.**
  - For more information, see the CKS topics on Headache - cluster and Migraine.
- **Acute angle closure glaucoma.**
  - For more information, see the CKS topic on Glaucoma.
- **Retinal transient ischaemic attacks and embolic visual deficits.**
  - For more information, see the CKS topic on Stroke and TIA.
- **Temporomandibular joint pain, sinus disease, and ear problems.**
  - For more information, see the CKS topics on TMJ disorders and Sinusitis.
- **Cervical spondylosis or other upper cervical spine disease.**
  - For more information, see the CKS topics on Neck pain - cervical radiculopathy and Neck pain - non-specific.
- **Ankylosing spondylitis.**
  - For more information, see the CKS topic on Ankylosing spondylitis.
- **Myeloma** with cervical or cranial deposits, or other cranial malignancy.
- **Serious intracranial pathology**, such as infiltrative retro-orbital or base of skull lesions.
- **Connective tissue disease.**

There is usually little difficulty in distinguishing giant cell arteritis from other forms of arteritis.

Patient information

https://www.nhs.uk/conditions/temporal-arteritis/
https://www.nhs.uk/conditions/polymyalgia-rheumatica/
Versus Arthritis charity booklet on GCA

References

NICE CKS July 2014 accessed May 2019 https://cks.nice.org.uk/giant-cell-arteritis#!topicSummary
Giant Cell Arthritis – Keep it in Your Head
Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 1990;33:1122---8.

Useful Reading

https://www.themdu.com/guidance-and-advice/journals/mdu-journal-april-2013/giant-cell-arteritis-risks

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