

Giant Cell Arteritis Protocol

Background

Giant cell arteritis (GCA) is a granulomatous vasculitis commonly of the temporal artery associated with polymyalgia rheumatic that classically presents in those above the age of 50 with a new temporal headache; it may be associated with rapid irreversible bilateral visual loss and thoracic aortic aneurysm (late).

Diagnostic Criteria – American College Rheumatology¹

Presence of 3 or more symptoms yields sensitivity 93.5% and specificity 91.2%:

- 1. > 50 years old.
- 2. New onset headache.
- 3. Temporal artery abnormality (e.g. thickened/beaded, reduced/absent pulse, tenderness).
- 4. ESR > 50 mm/hr (Westergren method).
- 5. Abnormal arterial biopsy demonstrating a necrotizing vasculitis with predominance of mononuclear cells and granulomatous inflammation.

If patients fulfil the above diagnostic criteria, they should be DIAGNOSED and TREATED for GCA as per this protocol.

If patients DO NOT fulfil the above diagnostic criteria, consider the following differentials²:

- 1. Herpes zoster.
- 2. Migraine or other causes of headaches.
- 3. Serious intracranial pathology (e.g. infiltrative).
- 4. Retro-orbital or base of skull lesions.
- 5. Other causes of acute vision loss (e.g. transient ischaemic attack).
- 6. Cervical spine disease.
- 7. ENT pathology (e.g. sinus, temporo-mandibular joint and ear disease).
- 8. Systemic vasculitides.
- 9. Connective tissue diseases.

If you still suspect GCA, please treat as GCA as per this protocol.

Visual Symptoms³

The following count as "visual symptoms":

- 1. NEW onset blurring of vision (not explained by other causes), either intermittent or constant.
- 2. NEW onset binocular (i.e. only present with BOTH eyes open) diplopia.
- 3. NEW pain in or around the eye.

High Risk Symptoms for Visual Loss²

- 1. Jaw claudication (not the same as temporo-mandibular joint dysfunction).
- 2. Tongue claudication.
- 3. Scalp necrosis (must be differentiated from herpes zoster ophthalmicus, which only affects CN V1).



Management If A Diagnosis of GCA Has Been Made²⁻⁵ - ALL PATIENTS

Investigations (should be done/requested immediately)

- 1. Observations BP, pulse, weight (if patient unsure).
- 2. Bloods CRP, ESR, FBC, LFTs, U&Es, blood glucose, HbA1c.
- 3. Imaging CXR.

Basic Management (should be started immediately)

- 1. PO Adcal D3 (or equivalent) once tablet daily continue whilst on steroids.
- 2. PO Lansoprazole 30mg (or equivalent) once daily continue whilst on steroids.
- 3. Consider starting PO Aspirin 75mg once daily if no contraindications.
- 4. Follow glycaemic management (see attached) as per Trust guidelines.

Referrals (should occur immediately in ALL cases)

1. Urgent letter faxed to Rheumatology (– call Calderdale Royal Hospital (CRH) Rheumatology secretaries to confirm receipt.

AND

2. Urgently referral faxed to Vascular Surgery for temporal artery biopsy of symptomatic side (TAB) – form available later in this document.

Steroids (start immediately)

If no high risk symptoms AND no visual symptoms:

1. Start PO Prednisolone 0.75 mg/kg (minimum 40mg, maximum of 60mg) once daily (without delay). This should continue until reviewed by Rheumatology.

OR

If visual symptoms:

- 1. Start IV Methylprednisolone (IVMP) 1 gram (500mg if <50kg) STAT and repeat once daily for TWO further days. The first dose should be given without delay at the point of diagnosis (i.e. it is not acceptable to wait until patient is transferred / seen elsewhere because this time lost could be the cause of sight being lost permanently). Subsequent doses can then be given as in-patient.
- 2. **PLUS** After THREE days of IVMP, convert to PO Prednisolone as stated above. This should continue until reviewed by Rheumatology.
- 3. **PLUS** Urgent phone referral to Ophthalmology for review (if already under another speciality) / admission (if from A+E, GP, other). Discuss DIRECTLY with:
 - a) Eye Clinic staff before 5pm in the first instance (CRH 01422 347218; HRI 01484 343235)
 - b) The 1st on-call doctor via Switchboard out of hours or in the second instance.
 - c) The Consultant on-call via Switchboard in the third instance.



OR

If ANY high risk but NO visual symptoms:

- 1. Start IV Methylprednisolone (IVMP) 1 gram (500mg if <50kg) STAT and repeat once daily for TWO further days. The first dose should be given without delay at the point of diagnosis (i.e. it is not acceptable to wait until patient is transferred / seen elsewhere because this time lost could be the cause of sight being lost permanently). Subsequent doses can then be given as in-patient.
- 2. **PLUS** After THREE days of IVMP, convert to PO Prednisolone as stated above. This should continue until reviewed by Rheumatology.
- 3. **PLUS** Urgent phone referral to Rheumatology (instead of fax referral) if within hours OR referral to Medicine for admission (to provide further IVMP doses, and related monitoring).
- 4. **PLUS** Urgent phone referral to Ophthalmology for review (to confirm no ophthalmic symptoms). Discuss DIRECTLY with:
 - a) Eye Clinic staff before 5pm in the first instance (CRH 01422 347218; HRI 01484 343235).
 - b) The 1st on-call doctor via Switchboard out of hours or in the second instance.
 - c) The Consultant on-call via Switchboard in the third instance.

References

- 1. Hunder G, Bloch, DA, Michel, BA. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. Arthritis Rheum 1990;33:1122-1128.
- 2. https://www.rcplondon.ac.uk/guidelines-policy/diagnosis-and-management-giant-cell-arteritis
- 3. Hayreh SS, Podhajsky PA, Zimmerman B. Ocular manifestations of giant cell arteritis. Am J Ophthalmol. 1998 Apr;125(4):509-20.
- 4. Dasgupta B, Borg FA, Hassan N, Alexander L, Barraclough K, Bourke B, Fulcher J, Hollywood J, Hutchings A, James P, Kyle V, Nott J, Power M, Samanta A; BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR guidelines for the management of giant cell arteritis. Rheumatology (Oxford). 2010 Aug;49(8):1594-7. doi: 10.1093/rheumatology/keq039a. Epub 2010 Apr 5.
- 5. Hayball P, Cosh, DG, Ahern, MJ, Schultz, DW, Roberts-Thomson, PJ. High-dose oral methylprednisolone in patients with rheumatoid arthritis: pharmokinetics and clinical response. Eur J Clin Pharmacol 1992;42:85-88.

Diagnosis of Giant Cell Arteritis made



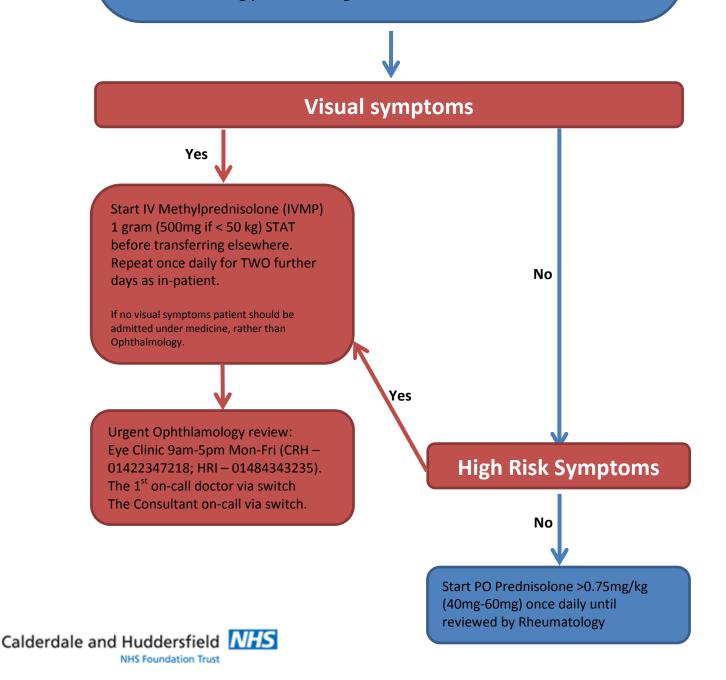
- Rheumatology +/- Medicine
- Vascular Surgery for temporal artery biopsy of symptomatic side

Investigations:

- Observations: BP, pulse, weight (if patient unsure)
- o Bloods: CRP, ESR, FBC, LFTs, U&Es, blood glucose, HbA1c
- o Imaging: CXR

• Immediate management:

- o PO Adcal D3 (or equivalent) one tablet daily
- o PO Lansoprazole 30 mg (or equivalent) one daily
- Consider starting PO Aspirin 75 mg once daily if no contraindications
- Follow glycaemic management





Referral Form to Vascular Surgery for Temporal Artery Biopsy (for inpatient and outpatient use)

Patient Sticker		Date & Time of Referral:	
		Referring Doctor:	
		Consultant:	
Patient contact number:		Copy Histology results to:	
Type of Transport, if Required?			
This procedure is carried out using Local Anaesthestic. Is the patient suitable?			
Please tick:			
Anticoagulant			
If yes, which one:			
Antiplatelet			
If yes, which one:			
Steroid			
If yes, date started:			
Co-morbidities			
Allergies			
If yes, please state:			
Infection risk			
If yes, please state:			
Which side is the biopsy to be performed	Left	Right	

For Hospital Use:

Appointment on: at:

Please send completed form to Email VascularSecs.HRI@cht.nhs.uk or Fax Number 01484 347218

If you need to speak to someone please call the Secretary on 01484 355415 or 01484 342481

Should there be any complex issues or concerns then please contact the secretaries or Vascular Surgery Team directly on the above numbers



Management of Hyperglycaemia and Glucocorticoid (Steroid) Therapy

Predisposing factors leading to increased risk of hyperglycaemia with steroid therapy

- Pre-existing type 1 or type 2 diabetes
- People at increased risk of diabetes (e.g. obesity, family history of diabetes, previous gestational diabetes, ethnic minorities, polycystic ovarian syndrome)
- Impaired fasting glucose or impaired glucose tolerance, HbA1c 42-47mmol/mol
- People previously hyperglycaemic with steroid therapy
- Those identified to be at risk utilising the University of Leicester/Diabetes UK diabetes risk calculator (riskscore.diabetes.org.uk)

Steroid induced diabetes (NEW ONSET) on once daily steroids

No known diabetes

- Check HbA1c in patients perceived to be at high risk prior to commencing steroids
- On commencement of steroids, recommend checking CBG once daily pre or post lunch or evening meal in those at "high risk" or having symptoms suggestive of "hyperglycaemia
- If CBG < 12 mmol/L, consider patient low risk and continue once daily CBG
- If a CBG is > 12 mmol/L, then increase monitoring to 4 times daily
- If CBGs are consistently > 12 mmol/L (1e over 2 readings in 24 hours), the commence treatment algorithm below

CBG above desired target eg 4-10 mmol/L

Start Gliclazide 40mg with breakfast and increase dose by 40mg increments regularly if targets not achieved

If no symptoms of hypoglycaemia and targets not achieved despite Gliclazide 160mg with breakfast, consider increasing to Gliclazide 240mg with breakfast (consider seeking specialist advice)

If target still not achieved, consider

- Adding in evening dose of Gliclazide (eg 40mg) and/or add human basal insulin (Humulin I, Insulatard, Insuman Basal) once daily in the morning
- If insulin commenced, suggest starting dose of 10 units and increase by 10-20% regularly to achieve desired CBG target

Discharge

- CBG monitoring will need to be continued if patients to remain on steroids after discharge
- If steroids stopped before discharge and hyperglycaemic resolves then CBG can discontinue with GP follow up
- If steroids stopped before discharge and hyperglycaemic persists then continue with CBG monitoring and seek specialist advice on further investigations/follow up



Managing Glucose control in people with <u>KNOWN DIABETES</u> on once daily steroids

Known Diabetes - Reassess glucose control and current therapy

- Set target blood glucose eg, 4-10 mmol/L
- Check CBGs ("BMs") 4 times daily and follow flowchart to adjust treatment accordingly
- In Type 1 Diabetes, also check daily ketones if CBG > 12 mmol/L
- In Type 2 Diabetes, check for ketones if CBG > 12 mmol/L with osmotic symptoms

Type 2 DM – diet/oral therapies/GLP-1

Insulin controlled (Type 1 or Type 2)

If no "hypo" symptoms and not on sulphonylurea

- Commence Gliclazide 40mg am, titrate accordingly to a maximum of 240mg am or until target reached
- Seek specialist advice if glycaemic control not improving despite Gliclazide 160mg am
- If on BD Gliclazide and targets not reached, consider specialist referral to consider titration to 240mg am and 80mg pm

Once daily night time insulin

- Transfer to morning
 Titrate by 10-20%
- Titrate by 10-20% according to pre-evening meal CBG readings
- If target not achieved, consider BD insulin

Twice daily insulin

 Increase morning dose by 10-20% according to pre-evening meal CBG readings Basal bolus insulin

- Consider transferring night time basal insulin to morning
- Increase quick acting insulins by 10-20% to target each pre-meal and bedtime readings accordingly

If no "hypo" symptoms and taking max dose 320mg/day

- Add Humulin I, Insulatard or Insuman Basal 10 units am
- Titrate by 10-20% accordingly until target achieved
- If target not achieved, consider BD insulin

If steroids are reduced or discontinued

- CBG monitoring may need to be continued in inpatients, and in discharged patients, assessed by their usual team (GP/Specialist)
- Any changes should be reviewed and consideration given to reverting to previous therapy or doses