

Referral Pathways for Headache in Adults

This pathway is not inclusive of all headache types.

IMMEDIATE assessment required:

Ref Hospital

Thunder clap headache

(including orgasmic headache)
Exclude subarachnoid haemorrhage

- Severe headache rising to maximum crescendo within a minute
- Worst ever headache

Headache associated with possible Meningo/encephalitis

Malignant hypertension

- Retinal changes
- BP > 200 systolic, 120 diastolic

Significant head injury

URGENT assessment required

Urgent investigation or urgent neurology referral

Temporal arteritis

Check inflammatory markers

- Always consider in patients over 50 years
- Inflammatory markers are normal in 5% of cases
- May need urgent biopsy to confirm

Exercise headache

(including pre-orgasmic headache)

iNeed image/scan

- 10% will have a secondary cause

Carbon monoxide poisoning

Measure CO-haemoglobin

- Non-specific headache
- Enquire re heating devices

Space occupying lesion

Red flags (risk >1%)

Image/scan or refer neurologist

- Associated relevant neurological signs
- Associated with new onset seizure

Orange flags (risk >0.1%-1%)

Need careful monitoring and low threshold for Image/scan or referral to GPwSI or neurologist

- Significant unexplained change in headache character
- Migraine aura >1 hour
- Headache precipitated by Valsalva manoeuvre
- New headache in a patient older than 50 years
- Headache that wakes from sleep (not migraine or cluster)
- Headache where diagnosis can not be made 6 weeks from presentation
- Primary cancer elsewhere
- Immunosuppressed or HIV

Diagnose Primary Headache

Exclude medication overuse headache

If treatment resistant refer to GPwSI

- Any analgesia including Triptans taken on more than 3 days of the week on a regular basis
- Non specific headache with a history of a prior primary headache
- Can obscure diagnosis of primary headache

Cluster

Refer to GPwSI. All new cases will need MRI.
(Can be relaxed if stable cluster present for some time)

- Excruciating unilateral peri-orbital pain lasting up to 3 hours – the cluster attack
- Unilateral autonomic features
- Number of cluster attacks in a cluster period – classically 6-8 weeks
- 10% are chronic

Migraine

Refer to GPwSI if:

- Difficult to manage
- Chronic migraine
- Uncertain diagnosis

- Recurrent severe, unilateral (30%) or bilateral pain with (30%) or without aura lasting 4-72 hours (can be longer).
- May be associated with nausea
- May be associated with phonophobia, photophobia or movement sensitivity
- Two out of three positive has high sensitivity: three months recurrent headache; associated with nausea; light sensitivity more pronounced with headache.

Tension type headache

Refer to GPWSI:

- Difficult to manage
- Uncertain diagnosis

- Dull, featureless, bilateral pain
- Cause unknown but often associated with anxiety/depression
- Reassurance and amitriptyline

APPENDIX

Further information from links above

Classification of Headache

Headache was first formally classified in 1986 and revised 2004. The brain has no sensory fibres. Intracranial pain arises from invasion, stretching, pressure on or inflammation of meninges. The two main classifications of headache are primary and secondary.

Primary headache

No underlying cause demonstratable - 90% of GP presentations.

Migraine 85% of GP presentations. See fact sheet 3.

- Severe episodic pain with or without aura associated with nausea, photophobia and phonophobia.
- 5% chronic migraine, >15 days each month. Usually history of episodic migraine.

Tension type headache - 10% of GP presentations but high population prevalence. Poorly understood. If occurs in migraine sufferer probably part of migraine spectrum.

- Dull, pressing pain usually bilateral with no nausea, photophobia or phonophobia.
- Episodic or chronic. Reassurance and amitriptyline first line approaches.

Cluster headache and other autonomic cephalalgias (<1% of GP presentations)

- Very severe unilateral pain with autonomic features rarely with nausea, photophobia or phonophobia. See fact sheet 4.

Secondary headache

Underlying cause demonstratable (5% of GP presentations). Some important headaches listed below. See (www.i-h-s.org) for full list.

Headache attributed to vascular disorder.

- Subarachnoid haemorrhage. 85% due to aneurismal bleed. Characterised by thunderclap headache. (Worst headache ever rising to a maximum within a minute.) 10% of thunderclap headaches are due to SAH. Medical emergency with high mortality. Sentinel headaches may be recognised in retrospect.
- Temporal arteritis. Occurs over the age of 50. 40% will have polymyalgia rheumatica. Can be systemically unwell. May have jaw claudication. Raised ESR or CRP in 97% of cases. Temporal artery biopsy may be necessary to confirm diagnosis. Treat with steroids 1mg per kilogram, maximum of 60mg a day and reduce to a maintenance dose. (Higher doses may be needed).
- Carotid/vertebral artery dissection. Can radiate anywhere in face and neck. Can have an associated Horner's syndrome. Can occur after trauma e.g. RTA. Collagen disease is a risk factor.
- Stroke. Non-specific headache can be associated with stroke
- Cerebral venous thrombosis. Can mimic any headache. Watch cancer, prothrombotic states, pregnancy, infection of facial structures. Can be fatal.
- Hypertension. Apart from malignant hypertension, the contribution of hypertension to headache is over-rated and in practice negligible.

Headache attributed to space occupying lesions

- Tumour. Primary tumour: 70% glioma - prognosis poor; 30% meningioma - 80% 5 year survival. Although headache is common during course of illness, only 10% of tumours present with isolated headache. Pain usually featureless.
- Secondary tumour particularly breast, lung, prostate.
- Non-malignant space occupying lesions e.g. AV malformations, cysts.

Disorders of intracranial pressure

- Idiopathic intracranial hypertension. Commoner in young obese women. Headache and papilloedema often with pulsatile tinnitus. Can lead to permanent loss of vision. Refer.
- Intracranial hypotension. Occurs due to CSF volume depletion as a result of leakage. Headache is worse on standing and alleviated by lying down. Classically post lumbar puncture but spontaneous leaks can occur. Refer.

Headache attributed to head trauma.

- Can come on up to 7 days after trauma. Intensity of pain may not be related to degree of trauma. Most resolve < 6 months but 25% can go on for longer. Watch for development of depression.
- Headache can be part of a post-concussive syndrome associated with other non-specific symptoms. Treatment difficult but amitriptyline drug of choice. Watch medication overuse headache. Underlying aetiology unknown but watch for secondary causes - haematoma, low CSF pressure due to dural tear, carotid or vertebral artery dissection.

Headache referred from other structures.

- Cervico-genic pain most common. Probably over rated as cause of headache. Neck pain in a migraine sufferer often part of migraine process.
- Eyes (refractive errors, glaucoma), temporo-mandibula joint, teeth, sinus (85% of diagnosed chronic sinusitis is migraine) are all possible but overestimated as causes of headache.

Headache associated with activity

- Sexual activity. Pre orgasmic - dull, gradually increases with sexual activity. Orgasmic – sudden severe at orgasm. Needs investigation. Treatment of both pre-emptive (indomethacin) or (betablocker).
- Exercise induced headache. May be a co-existing primary headache induced by exertion, e.g. migraine but cause of most exercise headache unknown. Need to exclude an underlying pathology. Treatment as for sex headache.

Headache attributed to infection

- Meningitis, encephalitis, systemic infection, HIV, brain abscess, T.B.

Headache attributed to metabolic causes, disorders of homeostasis or drugs

- Obstructive sleep apnoea. May be due to CO2 retention or poor sleep exacerbating primary headache. Reversible with treatment of problem.
- Carbon monoxide. Still fatalities each year.
- Alcohol, drugs including many prescribed drugs.
- Renal failure, thyroid disease, raised calcium.
- Medication overuse headache. Up to 3% of population. Occurs with regular analgesia or triptan use. (More than three days a week). All analgesics and NSAIs implicated, particularly codeine compounds. Non-specific, dull pain but usually starts from an underlying primary headache. (Enquire for previous headache history). Abrupt cessation of analgesics rather than gradual withdrawal after starting relevant preventative medication. Steroids can be useful to cover withdrawal symptoms but limited evidence base to support. High relapse rates.

Cranial neuralgias

- Trigeminal neuralgia most common. Burning or stabbing pain lasting less than two minutes. Often provoked by mild pressure or other triggers. Vascular compression of nerve most common cause.
- Glossopharyngeal nerve less common involving tongue, throat, jaw ear.
- Treatment with carbamazepine or lamotrigine.
- Trigeminal neuralgia may be confused with idiopathic stabbing headache particularly with co-existing migraine. Seconds, “jabs and jolts” anywhere in head.

Cluster headache and other autonomic cephalopathies

Background

Cluster headache is arguably one of the most painful conditions a GP will ever see. It is invariably misdiagnosed and usually inadequately treated. Often sufferers will have been to eye Departments with suspected glaucoma, ENT Departments with suspected sinusitis and Dental departments to have their wisdom teeth extracted. Cluster headache has been known by a number of terms including cluster migraine but it is very distinct from migraine and one of a group of headaches known as the trigeminal autonomic cephalalgias characterised by strictly unilateral pain and autonomic features. The pathophysiology is unknown.

Epidemiology

- 0.1- 0.2%
- Male: female ratio is 5:1
- It can begin at any age though the most common age of onset is the third or fourth decade.
- 10% of patients have chronic cluster headache (CCH) where remissions last less than one month
- Cluster headache is a life long disease but attacks invariably get less frequent with age

Making the diagnosis

The cluster attack

- The attacks are strictly unilateral although the headache may alternate sides with attacks.
- The pain is excruciatingly severe and is invariably associated with restlessness or agitation.
- Located mainly around the orbital and temporal regions.
- Headache lasts from 15 minutes to 3 hours. It has an abrupt onset and cessation.
- The cluster attack frequency varies one on alternate days to up to eight daily. The condition can have a striking circadian rhythmicity, with some patients reporting that the attacks occur at the same time each day.
- There are associated cranial autonomic symptoms on the side of the pain and lasting with it. For example, conjunctival injection, lacrimation, miosis, ptosis, eyelid oedema, rhinorrhoea, nasal blockage and forehead or facial sweating.
- Nausea, photophobia and phonophobia usually absent.
- Alcohol, exercise, and elevated environmental temperature can precipitate an attack but not outside a cluster period. Allergies, food sensitivities, reproductive hormonal changes and stress do not appear to have any significant role in precipitating attacks.

The Cluster Period or Bout

A cluster period is an episode during which there are frequent cluster attacks following which the individual is in remission.

- The average cluster period lasts between 6 and 12 weeks but there is considerable variation between patients.
- Most patients have one or two annual cluster periods, each lasting between one and three months. Often, a striking circannual periodicity is seen with periods occurring in same month of the year, often spring or autumn.

Important differences between Migraine and Cluster Headache	
Migraine	Cluster Headache
Prodrome or aura can occur	Prodrome or aura very rare
Pain can occur in any location	Pain is mainly periorbital
Pain is severe and throbbing. Patients want to lie down.	Pain is very severe and piercing. Patients pace the room.
Attack lasts 4-72 hours	Attack lasts 15-180 minutes and come in clusters
No autonomic features	Autonomic features around the eye on side of pain
Nausea, vomiting, photophobia or phonophobia	Rare

Differential diagnosis of cluster headache – other rare autonomic cephalopathies					
Diagnosis	Type of pain	Severity	Location	Duration	Frequency
Cluster headache	Boring	High	Orbital	15-180 minutes	1-8 a day
Paroxysmal hemicrania	Boring	High	Orbital	1-30 minutes	3-30 a day
*SUNCT	Stabbing	Moderate	Orbital	15-240 seconds	1 a day to 30 an hour

*short acting unilateral neuralgia form headaches with conjunctival injection and tearing

Investigation

Approximately 1% of cluster headache presentations will have an underlying pathology. Pituitary tumours are most common. All new cluster headache should be imaged. If a patient presents with a history of many years of stable cluster this can be relaxed.

Treatment

Acute treatment

- Subcutaneous sumatriptan 6mgs is the drug of choice. Unlike in migraine, it can be prescribed at a frequency of twice daily without reduction in effectiveness, side effects or rebound.
- Oral triptans are ineffective but there is evidence to support the nasal route.
- Oxygen. The mechanism of its action is unknown.

- 100% oxygen is required for a therapeutic effect. Ordering physicians should specify: A delivery of at least 10-12 litres per minute, a non-breathable mask. Specify "100% oxygen for cluster" on the Home Oxygen Order Form (HOOF)
- A static cylinder will provide up to 200 minutes supply depending on the cylinder pressure supplied.
- For portable use, an ambulatory cylinder can be ordered providing up to 40 minutes of treatment.
- Oxygen should inhaled for 10 to 20 minutes depending on the clinical response.
- For other co-existing pulmonary conditions where 100% oxygen may be harmful, advice should be taken from a respiratory physician.
- Patients should be made aware of the dangers of continuing to smoke in the presence of oxygen therapy. The majority of cluster patients are smokers at presentation and smoking cessation intervention should be given.

Using oxygen in cluster headache. Based on the British Association for the Study of Headache guidelines for Oxygen in Cluster Headache (www.bash.org.uk)

Short term prevention

- Steroids give rapid relief and are useful where there are only 2-3 attacks each year. Prednisolone 1mg/kg, to a maximum of 60mg once daily for 5 days and thereafter decrease over a three week period. Relapse can occur as the dose is tapered and in this case steroids are used as an initial therapy in conjunction with preventatives until the latter are effective.

Long term prevention

- Verapamil is the preventative drug of choice in both episodic and chronic cluster headache but higher doses than those used in cardiological indications are needed. After performing

a baseline ECG, start on 80mgs three times daily and thereafter the total daily dose is increased in increments of 80mgs every 10-14 days until the cluster attacks are suppressed with an ECG performed prior to each increment up to a maximum of 960mgs daily.

- Lithium, topiramate, sodium valproate, gabapentin are used but their impact is often marginal. Lithium is the most effective aiming for a serum level in the upper part of the therapeutic range. Occipital nerve injection can be helpful.
- Methysergide is a potent agent but as prolonged treatment has been associated with fibrotic reactions these are used only under specialist supervision.
- Surgery is a last-resort measure, either destructive procedures or neuromodulatory procedures with implanted electrodes.

Useful patient support sites

- The patient support group - the Organisation for the Understanding of Cluster Headache (www.clusterheadache.org)

Migraine

Introduction

Migraine is the main cause of high impact headache and affects 7.6% of males, 18.3% of females and 12% of children. Unfortunately, the majority of sufferers are reluctant to seek help and when they do the condition is less than optimally managed. Migraine is co-morbid with depression and anxiety disorders, epilepsy and asthma.

Making the diagnosis

Migraine is the most common headache presentation in primary care. Although the International Headache Society criteria (see figure 1) are quite specific, from a clinical perspective they may be relaxed. Answering yes to two out of three simple questions effectively identifies migraine sufferers:

- Has a headache limited your activities for a day or more in the last three months?
- Are you nauseated or sick to your stomach when you have a headache?
- Does light bother you when you have a headache?

- a) At least 5 attacks fulfilling criteria b-d
 - b) Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)

- c) Headache has at least two of the following characteristics:
- Unilateral location
 - Pulsating quality
 - Moderate or severe pain intensity
 - Aggravating or causing avoidance of routine physical activity
- d) During headache, at least one of the following:
- Nausea and or vomiting
 - Photophobia or phonophobia
- e) Not attributed to other disorder (A formal diagnosis of migraine therefore must include an examination to exclude another disorder.)

Fig 1. Formal criteria for a migraine diagnosis. Can be relaxed in practice.

The stages of migraine

- Prodrome. 30-50% of migraineurs – sensory or psychological features that can occur up to 48 hours prior to attack. Prodrome, e.g. craving for a particular food can be confused with a trigger.
- Aura. 30% of migraineurs – a reversible sensory or motor phenomena of cortex or less commonly brainstem lasting under 60 minutes. Visual auras are the most common followed by paraesthesia. Atypical auras can occur, e.g. vertigo, hemiplegia. Aura can occur in the absence of headache. As they are caused by a spreading cortical depression, their evolution with time distinguishes from T.I.A.
- Headache. 60% unilateral, throbbing or pulsating. Associated with nausea or vomiting, photophobia or phonophobia. Increased skin sensation can also occur.
- Postdrome – tiredness, elation.

Migraine triggers

- Often inconsistent. The importance of allergy remains unproven.
- The often unrecognised trigger is sensitivity to change. E.g. glucose, hydration, stress, oestrogen, sleep patterns – keep everything as constant as possible.

Pathogenesis

- Poorly understood. Mid brain migraine generator activates trigeminal system which causes dural inflammation and pain.
- Migraine generator close to nausea and vomiting centre. Gastric stasis and inhibition of drug absorption major problem.
- Migraine generator overlaps nuclei of upper cervical nerves. Neck and shoulder pain common in migraineurs and probably represents efferent signals and not primary neck problem.

The emergency call out

- Parenteral sumatriptan treatment of choice.
- IM diclofenac 50mg with IM antiemetic second line.
- Avoid opiates. Dependency soon develops

Management of the acute attack

- A useful first line of treatment that can be bought cheaply OTC, all of which should be taken together is: domperidone 10mg, paracetamol 1gram, ibuprofen 400 mg or aspirin 600mg. Soluble preparations act quicker. Can be taken prior to Triptan. Larger initial doses may reach therapeutic levels quicker. (Domperidone 20mg, paracetamol 1.5 gram, ibuprofen 600 mg or aspirin 900mg) providing maximum daily doses are not exceeded.
- If vomiting or severe nausea use domperidone suppositories 30mg and diclofenac suppositories 100mg.
- Triptans. Cornerstone of the acute attack. See figure 2. Nasal forms (Sumatriptan and Zolmatriptan) useful when gastric stasis is a problem or injectable (Sumatriptan) for intractable cases. Wafer formulations (Zolmatriptan and Rizatriptan) are for convenience only and not absorbed through the oral mucosa. Sumatriptan 50mg is now available OTC. Failure of response to Triptans is not a class effect. If one doesn't work rotate choices. Treat at onset of pain. May not be effective if taken during aura phase. More effective if taken with anti emetic. Vascular disease an absolute contraindication.

Group A (Higher speed onset)	Group B (Lower headache recurrence, lower side effect profile)
Sumatriptan 100/50mg	Naratriptan 2.5mg
Rizatriptan 10/5mg	Frovatriptan 2.5mg
Zolmatriptan 2.5mg	
Eletriptan 20mg/40mg	
Almotriptan 12.5mg	

Figure 2 - Available Triptans

Preventative treatment

- No specific indications for using preventative treatment. The impact upon the patient is the best guide.
- Preventative medication should be given for at least 8 weeks at its maximum tolerated dose before its impact should be assessed and if successful continued for at least 6 months.

- Beta-blockers are the drug of first choice and Propranolol, Metoprolol, Timolol and Nadolol are licensed for use in migraine. Atenolol 25 mg increasing to 100mg or the highest tolerable dose appears effective, is cheap and convenient to take. Nebivolol is a useful if side-effects are problematic.
- Amitriptyline 10mg increasing to 100mg or the highest tolerable dose is the preventative medication of second choice. Works well with a beta-blocker.
- Anti-epilepsy drugs form the third line choices, particularly sodium valproate (un-licensed) and topiramide (licensed). Gabapentin and lamotrigine are sometimes used.
- Pizotifen is licensed but weight gain can be troublesome.

Alternative treatments

- Physical therapies such as biofeedback, relaxation therapy, cervical manipulation, and cranial massage have are not supported by firm evidence.
- Positive trials have been reported on acupuncture but have been criticised on methodological grounds.
- Butterbur, feverfew, coenzyme Q10 and magnesium have a weak evidence base.

Migraine in women

- Avoid combined oral contraception with aura or migraine without aura with other vascular risk factors.
- Look for hormone sensitive migraine. Perimenstrual (Mefenamic acid or Naproxen –2d to +2d; transcutaneous oestrogen gel 1.5g/day –5d to +2d; frovatriptan 2.5mg/day – 2d to+4d. Perimenopausal (low dose HRT, not oral).

The majority of women get fewer migraines in pregnancy. Paracetamol and antiemetics are safe. Seek expert advice otherwise.

Useful resources

SIGN guidelines. www.sign.ac.uk. An excellent review of evidence based headache care.

The British Association for the Study of Headache (www.bash.org.uk). Contains UK headache management guidelines.

Exeter headache clinic web site www.exeterheadacheclinic.org.uk. Contains clinical guidance and comprehensive patient information treatment sheets that can be downloaded.

Useful information can be obtained from patient support groups on:

Migraine Action - www.migraine.org.uk

Migraine Trust - www.migrainetrust.org

Tension type headache

- 10% of GP presentations but high population prevalence.
- Dull, pressing pain usually bilateral with no nausea, photophobia or phonophobia.

- Poorly understood. Pathophysiology unknown.
- If occurs in migraine sufferer probably part of migraine spectrum.
- Episodic or chronic.
- Reassurance and amitriptyline first line approaches.
- Other non-pharmacological approaches such as biofeedback, relaxation can be useful.