THE BARE ESSENTIALS

# Headache

Richard Davenport

Correspondence to:
Richard Davenport, Department of Clinical Neurosciences, Western General Hospital, Crewe Road, Edinburgh EH4 2XU, UK; rjd@skull.dcn.ed.ac.uk

> "The patient with a headache often finds himself a medical orphan. He is fortunate indeed if his headache is transient, for otherwise he may find himself on an excursion to the ophthalmologist, otolaryngologist, neurologist, dentist, psychiatrist, chiropracter, and the latest health spa. He is *x* rayed, fitted with glasses, analysed, massaged, relieved of his turbinates and teeth and too often emerges with his headache intact." RC Packard 1979

Other than the most dedicated of superspecialists, all neurologists frequently see patients with headache. And yet headache has been, and perhaps is still, somewhat of a Cinderella area, with many myths perpetuated, and newer ones evolving. Faced with a patient with headache, the relevant questions are simple to ask, if not necessarily to answer (box 1). An accurate diagnosis almost always depends only on the history, and for most patients further investigations are supremely redundant—although many are investigated.

What headache patients most want is a comprehensible explanation of their symptom, which is perhaps the most important therapeutic step. This is a tricky task, especially when so little is understood about many headache syndromes, and many neurologists are not good at explaining concepts such as migraine or chronic daily headache. Far too often it is assumed that patients would like treatment and brain scans. Perhaps copying clinic letters to patients, and providing them with up-to-date review articles or useful websites, might help with our explanations.

To answer patients' questions, one needs time and energy—which are often in short supply; there is a temptation to reserve those resources for patients with "proper" neurological disorders such as multiple sclerosis or stroke. Invisible symptoms, such as pain, may lead to a degree of disbelief in doctors (and others)—something patients are acutely aware of. Neurologists need to remind themselves of this when faced with the healthy-looking chronic daily headache patient; an empathetic approach is as relevant here as anywhere else, if not more so.

This article is written mainly in the context of headache presenting in the outpatient clinic. But headache is also the primary symptom in 1–2% of patients presenting acutely to an emergency department. The skills required in each setting are essentially the same, but as the differential diagnosis is rather different (albeit with considerable overlap), headache in the emergency department is covered in a separate section. Although facial and neck pain are also common problems, this article will deal exclusively with pain and similar sensations felt in the head.

**Box 1 Questions to be answered during the consultation with a headache patient**

- Can I classify this headache, and if so how?
- Do I need to investigate?
- How can I best explain the diagnosis?
- What is the patient after—reassurance, explanation, treatment, brain scan, something else?
- Is treatment appropriate and if so, what is the most sensible approach?

## EPIDEMIOLOGY

Almost everyone experiences headache at some point. Fortunately, most do not seek medical attention. But some information is pertinent:

- Headache affects 95% of people in their lifetime.
- Headache affects 75% of people in any one year.
- One in 10 people have migraine.
- One in 30 people have headache more often than not, for 6 months or more.
- At least 90% of patients seen in a neurology outpatient clinic with headache will have migraine, tension type headache, or a chronic daily headache syndrome.
- Sinister causes of headache are rare, perhaps 0.1% of all headaches in primary care.

## DIFFERENTIAL DIAGNOSIS AND CLASSIFICATION

The International Classification of Headache Disorders (ICHD) provides a framework on which to base classification, although it is more suited to research than everyday clinical practice (box 2). Headache is divided into primary syndromes (predominantly migraine, and the trigeminal autonomic cephalalgias), but most patients and non-specialist doctors worry about the much rarer serious secondary headache syndromes (such as brain tumours).

The ICHD gives no indication as to the relative importance of what one sees in the clinic. In real life, the important headaches are:

- Chronic daily headache (most commonly medication overuse)

**Box 2 Framework of the International Classification of Headache Disorders**

**Primary headache syndromes**

- migraine
- tension type (see discussion)
- cluster and other trigeminal autonomic cephalalgias
- other primary headaches

**Secondary headache syndromes due to:**

- head and neck trauma
- cranial or cervical vascular disorder
- non-vascular intracranial disorder
- a substance or its withdrawal
- infection
- disorder of homeostasis
- disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures
- psychiatric disorder

- Migraine: disabling headache lasting hours or days, absent more often than present
- Trigeminal autonomic cephalalgias: characterised by strictly unilateral headache with ipsilateral autonomic ocular symptoms.

The frequency of headache is a useful diagnostic handle (box 3). Some headache syndromes, such as the trigeminal autonomic cephalalgias, often have a very specific and recognisable presentation (provided one has heard of them). Others, including many of the chronic headache disorders, are more featureless. There are however no shortcuts, and a careful history is the prerequisite.

Examining patients rarely contributes much to the diagnosis, but patients expect it, and thus it is an important part of the process of care. Occasionally, a diagnosis may hinge on the examination (for example, papilloedema in a persistent headache syndrome leading to a suspicion of idiopathic intracranial hypertension rather than new daily persistent headache). However, most reported focal signs turn out to be false positives, and the overwhelming importance of the history needs constant emphasising.

**Box 3 Headache patterns based on frequency**

**Headache on most days**

- Pain more often than not
  - Medication overuse*: abortive medication 3 or more days a week
  - Chronic migraine (bilateral or asymmetrical, associated features)
  - Hemicrania continua (strictly unilateral)
  - Chronic cluster headache (strictly unilateral)
- Pain-free more often than not
  - Cluster headache (strictly unilateral)
  - Paroxysmal hemicrania (strictly unilateral)
  - Icepick (anywhere in head)

**Headache on minority of days**

- Migraine (bilateral or asymmetrical, associated features)

*No other diagnosis can be made until medication overuse headache excluded.

## PRIMARY HEADACHE SYNDROMES

### Migraine

Migraine is best thought of as a "headache plus" syndrome, the "pluses" being prodrome, aura, nausea, vomiting, phonophobia and photophobia, and often systemic unwellness. This contrasts with the featureless chronic daily headache syndromes where headache (and misery) are the only true symptoms (once the adverse effects of medication are excluded).

- **Prodrome**: affects perhaps 1 in 10 patients at the most, usually 24–48 hours before headache; may include mood change, behavioural change, yawning, hunger, cravings, fatigue (or the opposite).
- **Aura**: affects up to 30%, although only about half this number regularly experience aura. It typically precedes the headache, evolving and subsiding over 5–60 minutes, and there is often a "no man's land" period between resolution of the aura and headache emergence, usually less than 60 minutes. Aura may sometimes intrude upon, or occur only during, the headache phase. Aura is typically visual, although almost any neurological symptom may occur.
  - The aura may occur in isolation, previously called migraine equivalents, now termed "typical aura without headache"; here focal epilepsy or transient ischaemic attacks (TIAs) enter the differential (the length and evolution of attacks are the most helpful discriminators, focal seizures usually lasting seconds to minutes, TIAs do not evolve, and cause negative rather than positive symptoms).
  - The nature of the aura may change over time and, when it does, it often alarms the patient; however, this remains entirely consistent with migraine, and does not indicate the need for urgent investigation.
  - People with aura who lose the associated headache as they get older, rarely complain; those who acquire aura in isolation, often in middle age, present typically to the TIA or eye clinic.
- **Headache**: migraine headache is typically thought of as severe, throbbing, and unilateral, but none of these is a requirement for diagnosis. Typically, it lasts for 24 hours or less, but can continue for 72 hours, and occasionally longer (hours to days). It often improves after vomiting and/or sleep and, in contrast to many chronic headache syndromes, generally improves with analgesia.
- **Associated features**: most migraine patients complain of at least one of nausea/vomiting, and dislike of noise/light/movement, and often all of these. Other autonomic features, such as diarrhoea, are uncommon. People with migraine feel (and look) unwell, and may complain of more global features such as mood change or lethargy. Rarely, more dramatic


2008;**8**:335–343. doi:10.1136/jnnp.2008.159095

features occur, including acute confusional states and even coma.

- **Frequency**: median is about 1.5 attacks per month, but at least 1 in 10 have weekly attacks
- **Triggers**: identifying reliable, reproducible triggers for migraine is over-rated, but most patients are keen to discuss this aspect. Hunger, sleep deprivation and "stress" are all recognised, and certainly an assessment of the patient's lifestyle is warranted. The relief (come down) of stress is probably a more common trigger than stress itself. Hormonal influences are important, and may open up a different therapeutic approach (see below), and getting up later than usual at weekends and other work-free days (that is, changes in body clock) is not always recognised by patients.
- **Hormones**: migraine during periods, and migraine emerging in pregnancy or with exogenous oestrogens (most commonly the combined oral contraceptive pill) are all well recognised. The difficulty is that the relationship is often inconsistent, with paradoxical effects (for example, some women lose their migraine during pregnancy; others only experience migraine during pregnancy). Patients who think there is a hormonal link for their migraine must keep a daily diary of headache and menstruation, and are often surprised to find there is no definite link. This is important because targeting a hormonal trigger is futile if it is not really present.

### Key questions in the headache history

- **Periodicity**: establish whether the headaches are episodic (at least a few days free from headache in between attacks), or present most of/all the time (at some point most/every day) (box 3)
- **Associated features**: any neurological symptoms and/or systemic problems: migrainous such as nausea, vomiting, dislike of noise, light, smell or movement; red flags such as persisting cognitive problems; or constitutional such as weight loss, malaise, fevers, etc? Autonomic features (lacrimation, redness, droopy eyelid) may need a witness.
- **Behaviour during headache (from patient and witness)**: episodic migraine usually leads the patient to a dark, quiet room, and the need to rest, and will stop a patient working. Patients typically look pale and unwell, contrasting with the healthy appearance of most chronic daily headache patients. Agitated behaviour during cluster is typical, and quite different from migraine behaviour.
- **Family history**: migraine is often familial, although the family diagnosis may be overlooked or erroneous and a family history is of no diagnostic value, but may be an interesting discussion point. Cluster is occasionally familial. Patients commonly offer a family history of brain tumour or haemorrhage: these raise concern but do not raise risk, in respect of headache diagnosis.
- **Current medication**: are patients taking prescription drugs that may contribute/cause headache (oral contraceptive, dipyridamole, etc)? Are they using/overusing analgesia (including over the counter drugs)? Are they using recreational drugs? People with medication overuse headache are typically reluctant to detail their medication use, and often under-report their usage.
- **Social situation/stressors**: building up a picture of the patient's social/psychological situation is important in any consultation, but my own experience is that most patients are stressed because they have a headache, not the other way round.
- **What the patient thinks**
  - Always ask patients what their own thoughts are; sometimes the answers are surprising, and many patients may suspect highly unlikely diagnoses (for example, multiple sclerosis). In such cases, it is easy to reassure them, but unless one asks, one will not know.
  - The inevitable brain tumour concerns emerge, sometimes with good cause (for example, family history or recent friend with the diagnosis; previous cancer elsewhere), although more often this is a concern of the referring doctor rather than the patient (not uncommonly it transpires the patient was unconcerned until such a diagnosis was raised by their doctor or other healthcare worker).

### Unusual migraine subtypes

Whether it is helpful to further categorise migraine is debateable, and the very existence of some of the subtypes below is questioned. However, the terms are still used by some:

- **Basilar-type migraine** (previously basilar migraine) is used for patients with migraine type headache accompanied by symptoms referable to the vertebrobasilar circulation, including dysarthria, vertigo, tinnitus, deafness, diplopia, ataxia, decreased level of consciousness, and bilateral sensory symptoms.
- **Hemiplegic migraine**: weakness which may last longer than 60 minutes (ie, much longer than a typical aura). It may be sporadic, or familial with an autosomal dominant inheritance. Up to 20% of the familial cases experience episodic cerebellar ataxia, and many develop more typical migraine (with and without aura).
- **Retinal migraine**: rare in practice, but identified as monocular visual aura (or blindness) followed by typical migraine headache. Other causes of transient monocular symptoms including TIA, and ophthalmic structural disorders (for example, retinal detachment) should be excluded. Patients find it difficult to distinguish monocular symptoms from the more typical binocular visual field symptoms.
- **Childhood periodic syndromes (migraine precursors)**: cyclical vomiting, abdominal "migraine" and benign paroxysmal vertigo are more common in children who subsequently develop migraine than in those who do not.
- **Ophthalmoplegic "migraine"**: is now reclassified as a cranial neuralgia (secondary headache syndrome), rather than a form of migraine. Typical migraine headache is associated with an external ophthalmoplegia (most commonly 3rd nerve palsy), but the headache often lasts a week or more, with a latency between headache onset and ophthalmic symptoms of up to four days.

### The complications of migraine

- **Medication overuse headache** (although more accurately this is a complication of the treatment).
- **Chronic migraine**: a common cause of chronic daily headache (see below).

- **Status migrainosus**: a debilitating migraine attack lasting >72 hours.
- **Persistent aura without infarction**: typical aura symptoms, usually experienced previously as a normal migraine attack for that patient, lasting >1 week (and often months to years, but with normal brain imaging).
- **Migrainous infarction**: typical migraine aura symptoms persisting >60 minutes in someone with previous migraine with aura, with radiological evidence of infarction in the relevant territory, not explained by other disease. This is not the same as an ischaemic stroke in someone who has (or has had in the past) migraine.
- **Migraine triggered seizure**: epileptic seizure occurring during or within 60 minutes of a typical migraine aura in someone with established migraine aura. Termed "migralepsy", it highlights the similarities between migraine and epilepsy. It can be difficult to distinguish aura from focal seizures in these circumstances.

### Tension-type headache

Although this diagnosis exists within the ICHD, and review articles on the subject continue to be published, the increasingly held view is that it is mild/moderate migraine. The headache is usually bilateral, non-pulsatile, and mild to moderate; although patients may describe it as severe, it rarely incapacitates them. In other words, it is a less disabling form of migraine.

### Trigeminal autonomic cephalalgias

These involve activation of the trigeminal and parasympathetic systems and are characterised by short-lasting headaches with variable autonomic features, cluster headache being the most common and most recognisable; the others are rare, or very rare.

#### Cluster headache (previously called migrainous neuralgia)

- The patient is more often male than female, age 20–50, and a smoker.
- He describes recurrent attacks of very severe unilateral pain, usually centred around the orbit, with ipsilateral autonomic features including conjunctival injection, tearing, miosis/ptosis, and nasal stuffiness.
- Patients are typically agitated during attacks (contrasting with standard migraine behaviour).
- The attacks peak rapidly (within minutes), and last 30–180 minutes (typically about an hour). The offset is usually quite abrupt.
- During a cluster, attacks occur at least once every 24 hours, and often more frequently, and usually wake patients from sleep at the same time ("alarm clock headache").
- Most cluster headache is episodic, clusters lasting weeks to months, with months to years remission, but 10–15% have a more chronic variant, with clusters lasting longer than a year without remission, or with remission lasting less than a month.

#### Paroxysmal hemicrania

- These patients describe frequent (>5 attacks/day) attacks of unilateral pain lasting 2–30 minutes, with autonomic features as in cluster headache, with a definite response to indometacin.
- As with cluster headache, the attacks may be episodic (periods of remission lasting at least 1 month) or chronic, and trigeminal neuralgia has been described in association with them.
- It is not the same as hemicrania continua (see below), though both are responsive to indometacin

#### Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)

- One of the shortest lasting headache syndromes (but with the longest name), it is very rare.
- Patients complain of frequent (up to 200/day) attacks of unilateral pain lasting 5–240 seconds, with ipsilateral autonomic features.

### Other primary headache syndromes

While these may not be common, it is important to recognise them to avoid unnecessary investigations and to reassure the sufferer (and their family).

- **Hemicrania continua**: a continuous (as opposed to paroxysmal hemicrania) unilateral headache usually without any other features, of moderate intensity, which responds to indometacin.
- **Primary stabbing headache**: previously called "icepick" or "jabs and jolts", refers to brief (seconds) attacks of stabbing pain, usually within the distribution of the ophthalmic division of the trigeminal nerve, but without autonomic features. It often accompanies other forms of headache, especially migraine.
- **Primary cough headache**: typically short-lasting, triggered by straining or Valsalva manoeuvre. It usually leads to imaging of the posterior fossa, especially to exclude Chiari malformation but what one should do if one is identified is far from clear.
- **Primary exertional headache**: headache is very similar to migraine, and lasts minutes to 48 hours.
- **Primary headache associated with sexual activity**: previously called benign orgasmic cephalalgia or benign sex headache, there are two varieties; pre-orgasmic which sounds most like tension-type headache and evolves as the act progresses, and orgasmic which may mimic subarachnoid haemorrhage, with an abrupt onset at orgasm. This kind of headache seems less common in women.
- **Hypnic headache**: the patients are elderly, and describe headache awakening them from sleep, usually at the same time of the night, and lasting typically 30 minutes, of mild to moderate severity.


2008;**8**:335–343. doi:10.1136/jnnp.2008.159095

- **Primary thunderclap headache**: this diagnosis can only reliably be made in secondary care, after exclusion of subarachnoid haemorrhage, as the clinical presentation is identical. Symptoms may last from hours to days, and may recur, but are benign.

### New daily persistent headache

The symptoms are similar to tension-type headache, but evolve de novo, without previous episodic headache. It responds poorly (if at all) to treatment, and is often a source of considerable concern to patients and their doctors, yet rarely indicates a sinister cause. Nevertheless, secondary causes usually require exclusion:

- **Raised intracranial pressure**: a space-occupying cause is unlikely if the only symptom is headache of several months, but anyone with a suggestive history requires brain imaging. More likely is idiopathic intracranial hypertension (almost only affects obese women); the most common sign is papilloedema, although a variety of cranial nerve palsies have also been described, secondary causes such intracranial venous thrombosis should be excluded.
- **Low cerebrospinal fluid volume (or pressure) headache**: previously called spontaneous intracranial hypotension, the clue is in the history, with a story typical of post lumbar puncture headache (ie, headache worse on sitting or standing), but without the lumbar puncture. This postural relationship becomes less clear with time and patients may forget their initial symptoms, so the physician must enquire carefully about this aspect. Only about 10% of patients have the typical MR appearances of a "sagging" brain (meningeal enhancement and sometimes subdural hygromas/haematomas); it is sometimes possible to identify the source of the cerebrospinal fluid leak.
- **Chronic meningoencephalitis**: infectious and non-infectious causes of meningoencephalitis may cause persistent headache, but these are rare, and usually have other features as well as headache.
- **Post brain insult headache**: patients who have an index brain insult (trauma, meningoencephalitis, subarachnoid haemorrhage, etc) may subsequently develop new daily persistent headache. There is great uncertainty about how the index event leads to the subsequent headache (if it really does), and in many cases there is retrospective recall bias.

## CHRONIC DAILY HEADACHE SYNDROMES

Chronic daily headache is a more useful term for doctors than patients. It is defined as headache lasting >4 hours on >15 days per month, for >3 months, but in practice means someone who has a headache more often than not. Patients typically describe symptoms over many weeks, months, years and sometimes decades.

It is important to establish whether the headache arose de novo, or began as an episodic syndrome, as this helps in classification; unfortunately, this is not always easy to establish, especially when symptoms have been present for a long time. There are seldom any systemic or focal neurological features—it is simply headache, although adverse effects of analgesia are common and may confuse the picture, and depression may also be present.

It is best divided into primary and secondary forms; primary may be further divided into:

- transformed migraine
- chronic tension type headache
- new daily persistent headache
- hemicrania continua.

There is confusion over the use of the terms chronic and transformed migraine:

- Chronic migraine requires typical migraine without aura occurring on more than 15 days of the month.
- Transformed migraine has a rather looser definition and only requires one of: prior history of migraine, a period of escalating headache frequency, and/or concurrent and superimposed attacks of migraine.

What is not in any doubt is the relevance of establishing whether there is medication overuse. All analgesics may contribute (including triptans), and a careful history is required; like for alcohol, patients may either deliberately or unwittingly underestimate their intake. They may also dismiss over-the-counter medications as irrelevant. Asking how long a packet of paracetamol might last, and how many separate stores of analgesia they keep, is one way of getting a more accurate picture, and the answer may surprise the patient, if not the physician.

## HEADACHE IN THE EMERGENCY DEPARTMENT

The emergency department environment is not conducive to a carefully considered history, and yet nowhere is it more vital. Patients and their families are often distressed and frightened, and may not appreciate that talking can be more important than immediate brain scans, so particular sensitivity on the physician's part is required.

The key issue is how the headache evolved. One which reaches its maximum intensity immediately or within minutes always suggests subarachnoid haemorrhage (SAH), even though only about 10–25% of such patients will prove to have this. Unfortunately, there are no accurate discriminators in the history, and although other neurological symptoms, neck stiffness, vomiting, seizures or transient disturbances of consciousness all indicate an increased likelihood of SAH, their absence does not exclude it. Other important secondary causes of abrupt onset headache include:

- ischaemic stroke, especially due to arterial dissection
- intracerebral haemorrhage, sometimes with no focal localising signs

- intracranial venous thrombosis
- intermittent hydrocephalus
- meningoencephalitis.

Headaches which evolve more slowly (over hours) are less likely to be sinister, but meningoencephalitis must always be considered, even in the absence of fever, rash (which must be looked for carefully, involving full inspection of the skin), neck stiffness or photophobia.

It remains the case that most "emergency" headaches will prove to be benign (most commonly migraine), but investigations are more often necessary than in the outpatient setting, and the treatment approach is also different.

### Investigations and treatment in the emergency department

- All patients with abrupt onset headache need an unenhanced CT brain scan as soon as possible (ideally within an hour of presentation).
- On the other hand, patients with suspected meningitis without focal symptoms or signs, and no disturbance of consciousness should not have their lumbar puncture delayed by a CT "to exclude raised intracranial pressure", which of course a CT never can.
- All patients should have baseline bloods tests early, including blood cultures if bacterial meningitis is suspected (and those patients should then immediately receive an appropriate antibiotic with corticosteroids).
- Pain relief—including triptans if indicated—is an obvious, but often neglected, part of management, and unwarranted concerns about "masking evolving symptoms or signs" with analgesia persist.
- A not uncommon mistake is to abandon searching for a possible secondary cause if the CT is "normal". If this result is surprising on the basis of the story, review the films, and consider other "CT normal" explanations (for example, intracranial venous thrombosis, missed signs of SAH).
- If a lumbar puncture is required, ensure it is done by someone with the appropriate skills to measure and record the opening pressure, request the correct investigations, and pursue and interpret the results correctly.

## RED FLAGS

During the history, symptoms suggestive of a secondary cause should become obvious, but only if the right questions are asked. Remember that patients with SAH may present late in the outpatient department, so be ready to consider this and other potentially sinister secondary causes, even though very few outpatients harbour such pathology. Ensure the following have been covered:

- **Other neurological symptoms**: unless part of typical migraine aura or autonomic features, these should always suggest a secondary cause.
- **Systemic features**: weight loss, fever and other systemic features should stimulate concern. Patients with benign headache are not persistently unwell.

## SECONDARY HEADACHE SYNDROMES NOT TO MISS

### Giant cell (temporal) arteritis:

This tends to be overdiagnosed, which is understandable given the potential consequences of missing it, but the considerable adverse effects of prolonged steroid treatment mean that strict diagnostic rigour should be applied. It almost never occurs under the age of 55, and one should have biopsy evidence to support the diagnosis in anyone younger. It is a disease of the elderly who frequently, but not always, have systemic features (such as malaise, polymyalgia), with a very high ESR (>75 mm in first hour), and whose headache disappears within 48 hours (often less) of adequate (60–80 mg prednisolone/day) steroid treatment. However, basing the diagnosis purely on steroid response is inadvisable, as steroids (or simply time) can lead to improvement in many other headache syndromes.

It is important to strive as hard as possible to confirm the diagnosis with temporal artery biopsy, if necessary bilateral, accepting that a negative result does not exclude it, and only persist with steroids if there is an unequivocal, immediate and sustained therapeutic response in both the symptoms and the ESR.

### Depression

Headache is a not uncommon symptom of depression, although clearly there is overlap with polymyalgia and giant cell arteritis.

### Cervicogenic headache

This is usually obvious from the history and examination, although neck pain is common in migraine and other primary headache syndromes, so the presence of neck pain should not automatically lead to this diagnosis. The pain often radiates frontally, behind the orbit(s). Examining neck movements is an important part of the clinical assessment, and finding a stiff and painful neck with reduction of movement opens up an obvious (if not evidence-based) therapeutic avenue (neck manipulation/massage), and identifying occipital neuralgia is useful, as this suggests a specific therapy (local anaesthesia of the occipital nerve).

## INVESTIGATIONS

By definition, only patients who are suspected of having a secondary cause of headache require investigation, which underlines the essential requirement of an accurate clinical diagnosis.

### Blood tests

Other than an ESR or CRP in older patients with new headache, blood tests are rarely helpful, although any suggestion of a systemic component


2008;8:335–343. doi:10.1136/jnnp.2008.159095

to the history should stimulate at least basic haematological and biochemical analyses (as well as a proper physical examination of more than just the nervous system).

### Brain imaging

Only a tiny proportion of patients with non-acute headache have a structural explanation. On the other hand, up to 10% of asymptomatic people may have an incidental abnormality of some sort on MR brain imaging, which can raise very difficult treatment dilemmas (for example, unruptured aneurysm, meningioma). Thus brain imaging in non-acute headache patients should be approached with considerable caution. Unfortunately, brain imaging is becoming ever more available, and patients (and some doctors) have misplaced faith in "doing a scan". Furthermore, organising a scan is often quicker and more attractive to the harassed doctor than carefully explaining what the diagnosis is, and why brain imaging is not appropriate. The end result is that far too many patients with headache are imaged unnecessarily—some with damaging consequences if a worrying "incidentaloma" is revealed.

Therefore, avoid imaging unless there is a very persuasive reason, and do not image patients with a robust diagnosis of a primary headache syndrome. Situations where imaging might be required are:

- headache syndromes where a structural explanation is plausible
- localising neurological symptoms or signs not explained by migrainous aura
- constitutional or systemic symptoms or signs
- recent diagnosis/treatment of cancer, especially where cerebral metastasis plausible
- unclassifiable headaches.

Sufficient time with patients will allow them to describe their story fully, and for neurologists to provide an understandable explanation of the diagnosis and why tests will not helpful. Warn patients that the risk of an incidental finding on MR imaging is up to 1 in 10. MR imaging carries no risk of radiation (or indeed any known harm), the disadvantage being in its very sensitivity, increasing the risk of identifying non-relevant abnormalities, so one might argue that CT is a more appropriate choice. However, patients are increasingly aware that MR is more detailed than CT, and thus may still request MR even after a normal CT.

### Other tests

These depend entirely on the clinical scenario, and may include examination of the CSF, more specialised imaging such as CT/MR venography, etc.

## TREATMENT

Only the treatment of primary headache syndromes will be discussed; of these, only migraine and trigeminal autonomic cephalalgias are effectively helped by drugs. Always assess whether any treatment at all is required—an explanation and reassurance may be sufficient.

### Migraine

- The few patients with avoidable triggers will mostly have recognised them long before they see a neurologist, but cyclical/menstrual migraine in women deserves special mention.
- Providing simple "housekeeping" tips, such as not skipping meals, adequate sleep and so on is appropriate, and providing more information (most easily by referring the patient to a patient support organisation) is often appreciated.
- Discuss alternative treatments such as acupuncture because it is remarkable how many patients have already tried these, which they were disinclined to mention until raised by the doctor; tell them there is no robust evidence to support or refute most alternative treatments.
- Explain that drug treatment is one avenue.
- Before adding drugs, look at the patient's current prescription, and consider whether drug withdrawal is appropriate (particularly women with migraine who are taking the combined oral contraceptive pill where withdrawal for at least 6 months is a potential treatment).
- Explain how treatments should be used, particularly symptomatic versus preventative treatment, so often confused by patients (box 4).
- Symptomatic treatment is only effective for the headache/nausea elements; there is no symptomatic treatment for aura.
- The decision whether to use the oral or parenteral route is important, but often not considered; early nausea and vomiting are likely to reduce the absorption of oral medication, and the parenteral route might be better.
- A "stepped" approach, using simple analgesia first, is appropriate, as this is highly effective for many patients.

#### Preventative treatments

The decision to use preventative therapy as well as symptomatic drugs is for the patient, depending on the frequency and severity of attacks, their consequences for their life, as well as the relative efficacy of the chosen symptomatic therapy. A number of drugs are available, and the choice is mainly governed by potential and actually experienced adverse effects (box 5).

#### Other treatments for migraine

- Acupunture: the only "alternative" treatment for migraine for which there is any evidence, it should be considered as a non-drug option, although limited availability.
- Psychological intervention: no scientific evidence to support its use, but a "pain management" approach may be helpful in patients with severe, drug resistant migraine provided medication overuse headache is kept in mind.

## Box 4 Symptomatic treatments for migraine

### Oral route

| | |
|---|---|
| Aspirin 900 mg plus anti-emetic (eg, domperidone 10–30 mg) | Supported by Class A evidence* |
| Ibuprofen 400 mg/ketoprofen 100 mg plus anti-emetic | Supported by Class A evidence* |
| Other non-steroidal anti-inflammatory drugs licensed for migraine in UK: diclofenac, tolfenamic acid, flurbiprofen, naproxen | |
| Triptans (eg, almotriptan 12.5 mg, rizatriptan 10 mg, eletriptan 40–80 mg) | Supported by Class A evidence*;<br>must be taken at onset of headache, *not* of prodrome/aura;<br>patients who do not respond to one triptan should be offered at least one other |
| Ergotamine | Less effective, more adverse effects |
| Opiates | Ideally should be avoided |

*Scottish Intercollegiate Guidelines Network, Class A: at least one meta-analysis, systematic review or high quality randomised trial.

### Parenteral route

| | |
|---|---|
| Diclofenac 100 mg suppositories +/− domperidone 30 mg suppositories or buccal prochlorperazine 3–6 mg | |
| Sumatriptan (10–20 mg) or zolmitriptan (5 mg) nasal spray | Many find subcutaneous easier to tolerate than nasal. Class A evidence |
| Sumatriptan 6 mg subcutaneous | |
| Haloperidol 5 mg iv | Effective, but adverse effects, and rarely used in UK outside hospital. |
| Metoclopromide 10 mg iv | Effective at relieving pain and nausea/vomiting (Class A evidence) |

- Botulinum toxin: not effective in at least one randomised trial, not recommended.
- Closure of patent foramen ovale (migraine with aura): only published randomised trial to date did not achieve primary endpoint, not recommended on current evidence.

### Cyclical/menstrual migraine

- Should be diagnosed only after recording at least 3 months' menstrual and migraine diary.
- Supplemental oestrogen in the premenstrual phase, and "tricycling" the oral contraceptive pill have been recommended.
- The symptomatic use of mefenamic acid, aspirin/paracetomol/caffeine, or triptans have Class A evidence to support them.

## Box 5 migraine preventative treatment

| Drug | Comments |
|---|---|
| Propranolol 40–240 mg/day | Class A evidence<br>Beta-blockers should be avoided in asthma |
| Other beta-blockers (eg, metoprolol, atenolol, timolol, nadolol) | Probably as effective as propranolol |
| Sodium valproate (800–2000 mg/day) | Class A evidence<br>Teratogenicity and weight gain limit usefulness in women |
| Topiramate (100 mg/day) | Class A evidence |
| Gabapentin (up to 1800 mg/day) | Evidence less robust than for valproate/topiramate |
| Anti-depressant drugs (start amitryptyline 10 mg or dothiepin 25 mg and titrate up as necessary) | Tricyclics/venlafaxine appear effective, but SSRIs are not useful |
| Pizotifen (1.5–3 mg/day) | Appears less effective in adults than children, adverse effects limit usefulness |
| Flunarizine (5–10 mg/day) | As effective as propranolol, not licensed in UK |
| Methysergide (up to 12 mg/day) | Effective, but rare serious fibrotic adverse effects: should not be used for longer than 6 months without at least 1 month break |

- Frovatriptan and naratriptan are effective as preventative treatment when started 2 days before menses, and continued for a few days, although are not licensed for such use; clearly a patient must be able to accurately predict her period to use this strategy effectively.
- Hormone replacement therapy to combat menopausal symptoms is safe to use in migraine patients, but there is no evidence that it is effective in treating migraine, and can sometimes exacerbate it.

## Treatment of trigeminal autonomic cephalalgias

- The only class A evidence for the treatment of cluster headache is sumatriptan 6 mg subcutaneous.
- High flow 100% oxygen (requiring a specific high flow oxygen regulator and face mask) or intranasal lidocaine 10% drops may be considered for those patients who do not respond to sumatriptan.
- The published evidence to support preventative therapies is poor. One (non-evidence based) approach is to start prednisolone (60 mg/day) and verapamil (up to 240 mg/day, sometimes higher) or methysergide (up to 12 mg/day) together at the beginning of a cluster, tailing the steroids after 2–3 weeks, but continuing verapamil/methysergide until the cluster has resolved. ECG monitoring is recommended when using verapamil, especially in high doses.
- A positive response to indometacin (up to 225 mg/day) in paroxysmal hemicrania is part of the diagnostic definition. Start at 25 mg three times daily for first week, then 50 mg three times daily for second week, and 75 mg three times daily in third week, but continue


2008;**8**:335–343. doi:10.1136/jnnp.2008.159095

## USEFUL WEBSITES

- British Association for the Study of Headache (BASH): For healthcare professionals with interest in headache www.bash.org.uk
- International Headache Society (download ICHD-2): www.i-h-s.org
- Migraine Action Association: www.migraine.org.uk
- Migraine Trust: www.migrainetrust.org
- Organisation for the Understanding of Cluster Headache (OUCH UK): www.ouchuk.org/html/

only if positive response. If successful, continue for at least a few weeks before titrating down.

- Lamotrigine has been suggested as an effective treatment for SUNCT.

### Treatment of other primary headache syndromes

Indometacin should be considered as a treatment, often prophylactically, for many of the other headache syndromes (see dosage schedule above). No response should lead to rapid abandonment of this strategy.

### Treatment of chronic daily headache syndromes

This is not a rewarding area for drug treatment, and indeed withdrawing drugs is probably more important. Patients who are using any analgesia three times or more each week are likely to have medication overuse headache, and withdrawal is the treatment. This strategy will never work unless the theory is adequately explained to a patient, and even then frequently fails.

## FURTHER READING


- Davenport RJ. Sudden Headache in the Emergency Department. *Pract Neuro* 2005;**5**:132–43.
- EFNS guideline on the drug treatment of migraine – report of an EFNS task force. *Eur J Neurol* 2006;**13**:560–572.
- Elrington G. Migraine: diagnosis and management *J Neurol Neurosurg Psychiatry* 2002;**72**:10–15.
- Frearson R, Cassidy T, Newton J. Polymyalgia rheumatica and temporal arteritis: evidence and guidelines for diagnosis and management in older people. *Age Ageing* 2003;**32**:370–4.
- Goadsby PJ. Recent advances in the diagnosis and management of migraine. *BMJ* 2006;**332**:25–9.
- Lipton RB, Bigal ME, Steiner TJ, *et al*. Classification of primary headaches *Neurology* 2004;**63**:427–35.
- MacGregor EA. Oestrogen and attacks of migraine with and without aura. Lancet Neurol 2004;**3**:354–61
- Matharu MS, Goadsby PJ. Trigeminal autonomic cephalgias *J Neurol Neurosurg Psychiatry* 2002;**72**:19–26.
- Packard RC. What Does the Headache Patient Want? *Headache* 1979;**19**:370–4.
- Schoenen J, Sándor PS. Headache with focal neurological signs or symptoms: a complicated differential diagnosis. *Lancet Neurol* 2004;**3**:237–45.
- Stovnera LJ, Zwarta J, Hagena K, *et al.* Epidemiology of headache in Europe. *European Journal of Neurology* 2006;**13**:333–45.
- Sudlow S. US guidelines on neuroimaging in patients with non-acute headache: a commentary *J Neurol Neurosurg Psychiatry* 2002;**72**:16–18.


- Explain why complete withdrawal from analgesia is essential.
- Tell them they are likely to get worse before they get better, and that many weeks may pass before they improve.
- Perhaps use low dose tricyclics to cover the period of withdrawal (amitryptyline 10 mg or dothiepin 25 mg), and occasionally topiramate or propranolol if convinced they have chronic/transformed migraine.
- For patients with long standing, intractable chronic headache, a pain management programme may help.

### What to do if treatment is not working

- Reconsider the diagnosis carefully. Have you missed medication overuse headache?
- Is the patient taking the treatment? If not, why not (commonly because he or she does not understand the rationale for treatment, or adverse effects)?
- Is medication overuse complicating the picture?
- Is the treatment being taken correctly (for example, triptans commonly fail because they are taken at onset of aura/prodrome rather than onset of headache, or is preventative treatment being taken in a symptomatic manner)?
- Recognise that many headache syndromes do not respond well to medication (less common in migraine, very common in chronic daily headache syndromes).
- Consider non-drug approaches.

## CONCLUSIONS

- Most headaches are due to a primary headache syndrome.
- Most patients who reach a neurologist have either migraine or a chronic daily headache syndrome.
- Patients want an adequate hearing of their symptoms, followed by a diagnosis, and understandable explanation.
- Most patients need reassurance, some will benefit from treatment, and few require investigations.
- Some headache syndromes are amenable to medical treatment (e.g. migraine), but others are much less so (chronic daily headache), and an honest explanation is usually appreciated.
- Above all, patients want someone who is interested in their headache and who will listen to their story.

**Acknowledgements:** This article was reviewed by Giles Elrington, London, UK.

**Competing interests:** Within the last 5 years, I have received honoraria for speaking or chairing meetings from various companies involved with headache treatments, including Merck Sharp & Dohme, Menarini, Janssen-Cilag, and Pfizer.