

ACUTE HEADACHE IN THE EMERGENCY DEPARTMENT

Richard Davenport

ii33

J Neurol Neurosurg Psychiatry 2002;**72**(Suppl II):ii33–ii37

About one fifth of the patients seen by a general neurologist in the UK will present with headache; the majority have a chronic or stable syndrome (mostly migraine and chronic daily headache). Most patients are assessed in the relative peace and calm of the outpatient clinic, and their management is dealt with elsewhere in this supplement. A small proportion of headache patients will present acutely, either directly to the emergency department (ED), or referred via their general practitioner (GP). They account for between 1–2% of admissions to an ED.¹ This article focuses on the diagnosis and early management of acute or recent onset headache, which I have arbitrarily defined as the onset of a new headache syndrome within the last few days, hours or even minutes. I recognise that in many areas of the UK patients presenting as an emergency with a headache are rarely seen by a neurologist (of any grade) within the acute phase, if indeed at all; nevertheless, this article is written primarily as a guide for the neurological trainee when faced with a “headache in the ED”.

OBJECTIVES

The primary objectives for the neurologist are deceptively simple—make the diagnosis, relieve the headache, and investigate and manage appropriately. As many as a third of patients presenting to the ED with acute headache will be harbouring potentially fatal or disabling intracranial conditions such as subarachnoid haemorrhage (SAH), but all will be in distress, alarmed and often frightened. These emotions may be reflected in their medical attendants, and there is a tendency to channel the patient immediately towards the nearest scanner, in the hope that this will identify the cause. Certainly it will delay the need for active thought; it may also delay the patient receiving appropriate treatment. The neurologist must maintain his or her calm, and remember that their greatest asset is the ability to take a careful history, even in the chaos of a busy ED on a Saturday night. The history alone will identify who needs reassurance and discharge, and who requires further investigation. Figure 1 summarises the initial assessment.

THE DIFFERENTIAL DIAGNOSIS

The differential diagnosis for a patient presenting with headache in the ED is intimidating (table 1); yet most of these causes are uncommon. The first priority is to identify those patients who are presenting with sudden onset headache (about a third of whom will be harbouring serious intracranial pathology) and are likely to require urgent investigations. It is far more dangerous not to recognise sudden onset headache than to over diagnose it; this applies especially to GPs who will have initial contact with most SAHs who survive the first few minutes. Education of doctors is therefore a key factor in improving recognition.³ The remaining patients will be those with a pre-existing condition who have presented for a variety of reasons (for example, the chronic daily headache syndrome patient presenting “at the end of their tether”), who require sympathetic treatment but rarely admission or investigation; a readily identifiable primary headache syndrome (for example, first ever migraine); and those patients with a non-neurological explanation for their symptoms.

The history is essential to the diagnosis. Clearly resuscitation of an ill patient takes precedence, and in some circumstances the institution of immediate treatment may be appropriate (for example, administering antibiotics to a febrile patient with headache and a purpuric rash—see fig 2), but the crucial importance of the history cannot be over emphasised. The history will allow the neurologist to recognise primary headache syndromes immediately (for example, first ever migraine headache, or previously unrecognised cluster syndrome); while alarming and distressing to the patient, a confident diagnosis in such cases will avoid unnecessary investigations, direct appropriate treatment, and indicate a benign prognosis. Because of the wide differential diagnosis, the neurologist must be prepared to consider headache causes outwith his or her own organ of interest. For example, unioocular or retro-orbital pain with an associated visual disturbance should trigger consideration not just of migraine, a transient ischaemic attack (perhaps caused by ipsilateral carotid dissection) or optic neuritis, but also acute glaucoma, requiring the urgent attention of an ophthalmologist.

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

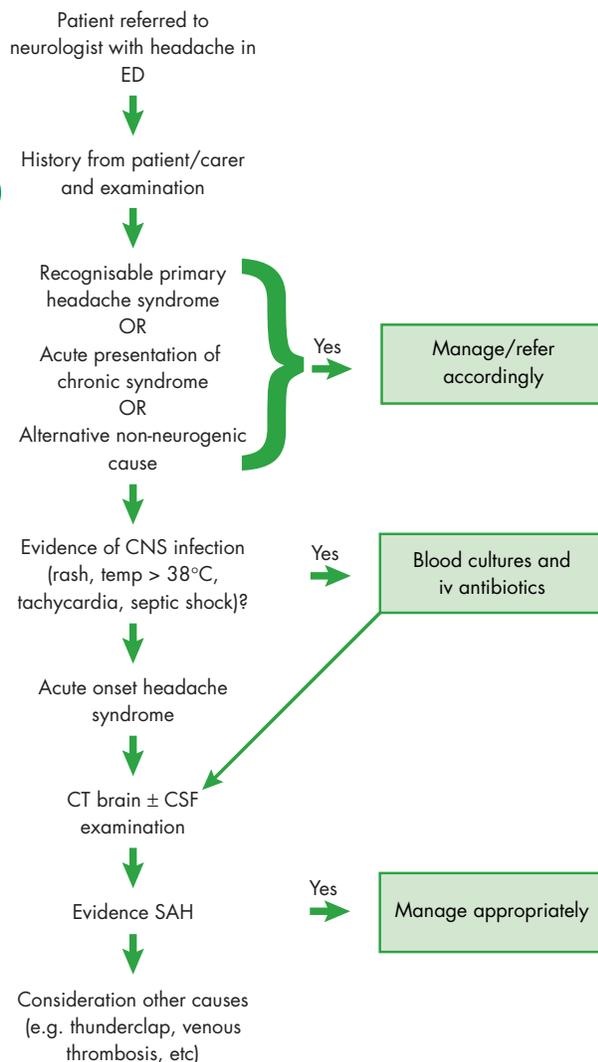


Figure 1 Initial assessment of acute headache in the emergency department (ED).

EXAMINATION

The neurologist must perform a general medical examination in addition to the neurological assessment. A skin rash and raised temperature must be actively sought, and signs not usually encountered in neurology considered (for example, enlarged cervical lymph nodes or an inflamed tympanic membrane). Neurological signs will suggest a primary neurogenic cause, but may be non-specific (for example, a Horner's syndrome may occur in migraine, cluster headache, or carotid dissection), and the absence of signs is of little comfort in the presence of a suspicious history—many patients with SAH will present with headache alone. The presence of neck stiffness (meningism) indicates irritation of the meninges by blood or inflammation, but cannot differentiate between the two; meningism may be absent in coma, and the absence of neck stiffness does not exclude SAH or meningo-encephalitis. The classic sign of SAH, subhyaloid haemorrhage, may be difficult to see, and probably is most common in patients with notably increased intracranial pressure and a decreased level of consciousness (in whom urgent investigations are required anyway).

Table 1 Causes of headaches presenting in the emergency department

Primary headache syndromes
▶ Migraine
▶ Cluster headache and related syndromes (including paroxysmal hemicranias, SUNCT)
▶ Thunderclap headache
▶ Hypnic headaches
▶ Benign exertional/sex headache
▶ Cough headache
▶ Exploding head syndrome (note this is a sensation, not headache)
Secondary headache syndromes
▶ Traumatic
–simple closed head injury
–complex with subdural/extradural/subarachnoid/intracerebral haemorrhage
Vascular disease
–subarachnoid haemorrhage (aneurysmal, perimesencephalic, other vascular anomalies)
–unruptured aneurysms
–acute cerebral ischaemia (TIA or stroke)
–non-traumatic subdural/extradural/intracerebral haemorrhage
–dissection of carotid/vertebrobasilar arteries
–cerebral venous thrombosis
–vasculitis (including giant cell arteritis)
▶ CNS infection
–meningo-encephalitis (bacterial, viral, fungal)
–cerebral abscess
▶ Non-vascular intracranial disease
–intermittent hydrocephalus (e.g. colloid cyst)
–idiopathic intracranial hypertension
–intracranial hypotension (spontaneous or post LP)
–intracranial tumour
–pituitary apoplexy
–Arnold Chiari malformations
–optic neuritis
▶ Metabolic or toxic disturbances
–phaeochromocytoma
–thyroid disease
–drug induced
–withdrawal syndromes
–hypercarbia
▶ Hypertensive encephalopathy
▶ Cervical spine disease
▶ Dental, ENT or ophthalmic disease (e.g. sinusitis, acute glaucoma)
▶ Secondary to general medical conditions (e.g. ischaemic heart disease, infection other than cranial, epilepsy)

CNS, central nervous system; ENT, ear, nose, and throat; LP, lumbar puncture; SUNCT, short lasting unilateral neuralgiform headache with conjunctival injection and tearing; TIA, transient ischaemic attack

SPECIFIC ACUTE HEADACHE SYNDROMES

Subarachnoid haemorrhage

As many as one in four patients presenting to their GP with a sudden onset headache will have suffered an intracranial haemorrhage, although this figure drops to about one in eight if headache occurs in the absence of other symptoms (that is, excluding nausea, vomiting, neck stiffness, transient disturbances of consciousness or focal neurological symptoms).⁴ The cardinal symptom of an SAH is headache, present in 85–100% (those without a headache are usually unconscious, and therefore not able to give a history). Approximately 85% of SAHs are secondary to a ruptured intracranial saccular aneurysm, a further 10% are caused by the benign perimesencephalic syndrome,⁵ and the remainder are caused by arteriovenous anomalies and rarities.

The prognosis of SAH is poor; about 25% of patients will die within 24 hours of their ictus (that is, either before they reach the ED, or very shortly thereafter); a further 25% will die within hospital, and of the survivors, up to 50% will be

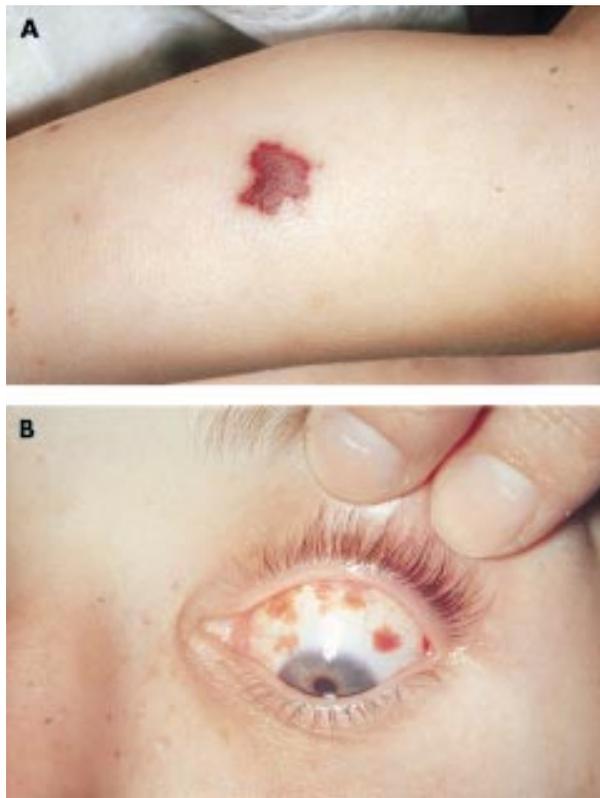


Figure 2 Purpuric rash in meningococcal meningitis.

disabled. Those who survive their aneurysmal SAH but do not have the aneurysm secured (either by clipping or endovascular treatment) have a 3% per year risk of rebleeding. An early and accurate diagnosis is therefore imperative, and it is salutary that in the UK, failed diagnosis or management of SAH represents the largest single area of neurological litigation. However, most patients presenting with headache, even in the ED, will have an entirely benign explanation. Who therefore needs investigation? It is easy to state that anyone with a “sudden” headache needs investigation. Yet there is remarkably little written about what exactly qualifies as a “sudden” headache and what does not, with many articles restricting themselves to instant, apoplectic headaches.² Real life is often less clear, and some of the problems faced by both GPs and hospital doctors when confronted with headache are illustrated in the case described in the adjacent box.

What is an “acute onset” headache?

We have all been taught that the typical headache of SAH arises instantaneously or within seconds (“like a blow to the head”), and is immediately of maximal severity. Any patient presenting with such a description must be investigated, and should provide little dilemma for the neurologist, or any other physician. However, SAH may also present with a headache evolving over minutes rather than seconds, and benign syndromes (such as thunderclap or benign sex headache) may present in an apoplectic manner indistinguishable clinically from SAH—so although a truly abrupt onset increases the likelihood of SAH, it is not sufficiently good at discriminating from other benign syndromes.⁶ Quite simply, it is unclear over what time a headache may crescendo and still be caused by an SAH; furthermore, patients sometimes find it difficult to be sure whether their headache did get worse over time, especially if it was bad to start with and they are vomiting and

Case study

A 42 year old woman became aware of a mild global headache while warming up for her aerobic class. Several minutes later (before the class started), she had sudden exacerbation of her headache, followed by vomiting and photophobia. She was seen by her GP, who diagnosed migraine and gave her intramuscular morphine and prochlorperazine. She spent the next four days in bed with her worst ever headache (she had a previous history of migraine without aura, but this was much worse). On day 6 she was seen by a different GP who recognised the significance of her symptoms and referred her urgently.

On examination she looked mildly unwell and distressed, but had no signs. A computed tomographic (CT) brain scan was reported initially as normal (in retrospect, a neuroradiologist identified a probable anterior communicating artery aneurysm and blood in the interhemispheric fissure—see fig 3); a lumbar puncture revealed 4000 red cells in three successive bottles, but no visible xanthochromia. A diagnosis of late presenting SAH was made, the aneurysm was confirmed on catheter angiography, and this was successfully coiled. She was discharged with no neurological deficit.

This case demonstrates several points:

- ▶ the patient’s symptoms were initially misinterpreted—sudden onset headache, even in migraineurs, is an SAH until proved otherwise
- ▶ SAH does not always present exactly like “a bolt from the blue”
- ▶ a normal clinical examination and CT (especially those reported on call by junior or non-specialist radiologists) do not exclude SAH
- ▶ xanthochromia to the naked eye is an unreliable sign.

distressed. A low index of suspicion is required, and all “first or worst” headaches arising within a few minutes or so require referral and investigation, although the odds of SAH fall with virtually every passing minute of a truly crescendo headache.

The other unresolved issue is how short a headache may be and still be caused by an SAH. Most SAH headaches last days to weeks, but some are shorter. The best observational studies of acute headache have defined duration of at least one hour, and traditionally many specialists use the “one hour” rule. A group of neurologists with considerable experience of SAH have stated that they have never seen anyone presenting with SAH with a headache of less than one hour—although they qualify this by admitting that no one truly knows what the shortest duration of headache might be, and conclude that perhaps SAH should be considered in anyone with a sudden severe headache, even if it resolves within one hour, particularly if there is any impairment of consciousness.⁷

In summary therefore, an operational definition of a headache suggestive of SAH might be as follows: a sudden onset “first or worst” headache which is usually maximal within moments, but may sometimes develop over a few minutes, and which lasts at least one hour. SAH may present with a headache out with these parameters, but it is likely to be rare indeed.

Associated features

The presence of nausea, vomiting, neck stiffness, transient loss of consciousness, or focal neurological symptoms are supportive of the diagnosis of SAH, but are not sufficient to distinguish from more benign syndromes (or even to distinguish between aneurysmal SAH and the considerably more benign syndrome of perimesencephalic SAH).⁶ Absence



Figure 3 Non-contrast CT brain on day 6 of subarachnoid haemorrhage; note subtle outline of anterior communicating artery aneurysm, with blood in interhemispheric fissure. CT reported initially as normal.

of these symptoms cannot be taken as reassurance that the patient has not had an SAH, as a proportion will present with headache as the only symptom.

The notion of a warning or “sentinel” headache (a term coined by an Edinburgh neurosurgeon) is perpetuated in textbooks and review articles. The “evidence” comes from hospital based, retrospective studies. A prospective, community based study has provided a more realistic estimate of headache preceding SAH. A total of 148 patients presenting within the community with an acute onset headache (defined as reaching maximal severity within one minute, lasting at least one hour) were assessed. Of these 37 (25%) had suffered an SAH, yet only two of these 37 recalled previous sudden headache at the time of presentation to their GPs. Furthermore, the pattern of blood distribution on CT was similar when compared to a previous hospital cohort of SAH patients, and the outcomes between the two groups was not significantly different.⁴ In a further prospective study, the clinical and radiological features of a group of patients presenting to hospital with aneurysmal SAH were compared; there were no differences between those with preceding episodes of sudden headache and those without.⁵ The term “warning leak” is therefore misleading, and should be abandoned. The emphasis must be on educating doctors to recognise the significance of an acute onset headache, and ensure that such patients are referred and investigated appropriately at the time of presentation.³ Attempting to make a diagnosis several weeks after the ictus is far more difficult than during the acute phase, as all neurologists will testify (see below).

Vascular causes other than SAH

Intraventricular or primary intracerebral haemorrhage, and ischaemic stroke may present with headache. By definition,

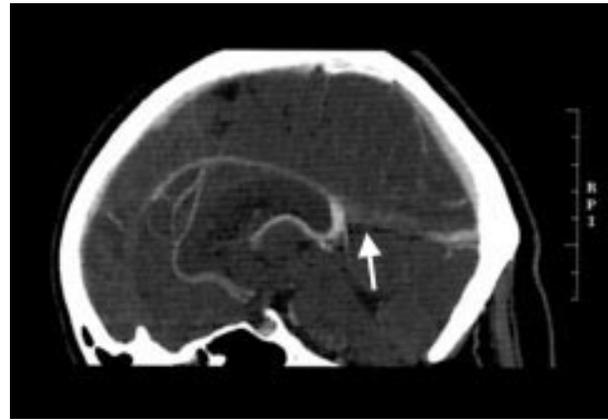


Figure 4 CT venogram in intracranial venous thrombosis. Note the absence of contrast in the straight sinus.

however, there will be other focal symptoms, to alert the neurologist. Some patients will be comatose early (usually those with haemorrhagic stroke, particularly in the posterior fossa), and urgent CT is required for the diagnosis. Dissection of the carotid or vertebral arteries may present with predominantly head, neck or facial pain. Dissection of the internal carotid artery causes ipsilateral facial, dental or cervical pain; neurological features may be minimal or absent initially, but an ipsilateral Horner’s syndrome should be looked for. More obvious features are ipsilateral monocular blindness or contralateral motor or sensory symptoms. Vertebral dissection may cause ipsilateral pain in the back of the neck and/or occiput, and may lead to brainstem symptoms. The neurological symptoms in arterial dissection may fluctuate considerably—for example, alternating hemiparesis and variable levels of consciousness in a patient with vertebral dissection. Dissection may also be a rare cause of SAH.

Intracranial venous thrombosis presents in a diverse manner, ranging from intracranial hypertension to rapid onset coma and death. A proportion will present with acute onset headache indistinguishable from SAH,⁹ the diagnosis only becoming clear with further investigations, such as CT angiography (fig 4).

Acute cervicogenic pain

A number of conditions may cause acute neck pain, which is not synonymous with neck stiffness. Neck stiffness (that is, meningism) is elicited by attempting passive flexion of the patient’s neck—rotation of the neck is not usually affected by true meningism, in contrast to a patient with a neck problem caused by arthritis or mechanical disease. Absence (particularly in a comatose patient) does not exclude subarachnoid blood or meningeal inflammation. Other conditions, including local (for example tonsillitis, pneumonia, cervical lymphadenitis), or non-central nervous system systemic (for example, viraemia) infection may also cause neck stiffness, so it is neither a sensitive nor specific sign.

Meningo-encephalitis

Although meningeal infection usually presents in a subacute manner, patients may either present comatose (that is, with no history available from them), or occasionally with true sudden onset headache indistinguishable from SAH. I have seen both bacterial and viral meningo-encephalitis present in this manner, the diagnosis only becoming clear on investigation. Untreated bacterial meningitis has a poor prognosis,

and immediate treatment with antibiotics is essential. The presence of a high fever ($> 38^{\circ}\text{C}$), features of septic shock, or a purpuric rash should be sought (the rash in particular may not be obvious, and careful inspection of the whole body is crucial—see fig 2).

Benign headache syndromes

Physical exertion (including sexual activity) may precede SAH or benign syndromes such as thunderclap, benign exertional or coital headache, but it is rarely possible to differentiate between these on clinical grounds alone. Thunderclap headache is a diagnosis of exclusion, and is only acceptable once SAH and other serious intracranial causes have been excluded. The prognosis is excellent, although about half will have a pre-existing history of migraine, or will subsequently develop it.¹⁰ A history of recurrent headache in association with sexual activity is a reassuring one for the neurologist (if not the patient), but to diagnose a “first ever” coital headache in the ED without further investigation is to invite disaster.

MANAGEMENT IN THE ED

A detailed description of the management of the many different types of headache encountered in the ED is beyond the scope of this article. The immediate responsibilities of the neurologist are to provide an accurate differential diagnosis, initiate any immediate treatment required (including analgesia), and organise appropriate investigations. Virtually all sudden onset headache patients require admission for cerebral imaging and possibly lumbar puncture. The choice of imaging will depend upon availability and the differential diagnosis, but CT remains the investigation of choice for patients presenting with an acute onset headache within 72 hours of ictus (identification of subarachnoid blood on CT starts to fall rapidly after a few days).

It is easy to forget analgesia in the excitement of the ED and while organising investigations, but this is important. Patients with a proven or suspected SAH should receive adequate analgesia, as pain with agitation may increase intracranial pressure. Drugs with antithrombotic properties (that is, aspirin and other non-steroidal anti-inflammatory drugs) should be avoided, and codeine phosphate (or paracetamol for less severe headaches) is the drug of choice. Other opiate narcotic drugs should ideally be avoided, although some patients may require the judicious use of diamorphine. Stool softeners should be prescribed simultaneously with codeine.

Ideally patients presenting with an acute headache who require admission to hospital should be managed on a neurological ward. Although there is no direct evidence to suggest that outcome is improved, there are plenty of studies in other areas (including acute asthma, myocardial infarction, gastrointestinal haemorrhage, and stroke) which indicate that management on specialist units improves outcome. In the UK only a small proportion of such patients are admitted directly to neurological units, although there is considerable variation between regions.

DELAYED PRESENTATION OF ACUTE ONSET HEADACHE

Occasionally the neurologist is faced in the clinic (these patients rarely present to the ED for obvious reasons) with a patient describing a sudden severe headache which occurred several weeks or even months beforehand, in whom the significance of the symptoms was not recognised by the

patient or his doctors at the time. The difficulty facing the neurologist is whether to investigate (and if so, how) or not. The usefulness of CT scanning and cerebrospinal fluid (CSF) examination declines rapidly within three weeks of the ictus (CT images often return to normal within 1–2 weeks of initial SAH, and CSF xanthochromia may be seen in 40% or less after three or four weeks), making these tests almost pointless in many cases. The neurologist must first decide whether to investigate or not, and if so whether to employ invasive conventional catheter angiography, or non-invasive, but less specific and sensitive magnetic resonance angiography. My own practice is to undertake urgent magnetic resonance angiography unless there are particular aspects which significantly increase the chance of an aneurysm, such as a family history, in which case I would recommend catheter angiography.

CONCLUSIONS

The neurologist may encounter a wide variety of patients in the ED presenting with headache. Of those patients presenting with their “first or worst” sudden onset headache, approximately one third will have a serious intracranial cause, most commonly SAH; however, most of these patients will require investigation, as even the best history and examination cannot accurately distinguish between sinister and benign syndromes. The neurologist’s key task is to identify and manage these patients.

REFERENCES

- 1 Ward T, Morris L, Phillips JM. Evaluation and management of headache in the emergency department. *Med Clin North Am* 2001;**85**:971–96.
- ▶ **A useful and up to date summary.**
- 2 van Gijn. Pitfalls in the diagnosis of sudden headache. *Proc R Coll Physicians Edinburgh* 1999;**29**:21–31.
- ▶ **All you ever needed to know about sudden headache, and entertainingly written by the foremost neurological authority on SAH and sudden headache syndromes.**
- 3 Fridriksson S, Hillman J, Landtblom AM, et al. Education of referring doctors about sudden onset headache in subarachnoid haemorrhage – a prospective study. *Acta Neurol Scand* 2001;**103**:238–42.
- ▶ **Emphasising that education is the key to recognising the relevance of sudden headache in the community (although their claim that this improves outcome is difficult to accept on the data available).**
- 4 Linn F, Wijidicks E, van der Graaf Y, et al. Prospective study of sentinel headache in aneurysmal subarachnoid haemorrhage. *Lancet* 1994;**344**:590–3.
- ▶ **A key study from the Dutch group.**
- 5 van Gijn J, van Dongen KJ, Vermeulen M, et al. Perimesencephalic haemorrhage: a nonaneurysmal and benign form of subarachnoid haemorrhage. *Neurology* 1985;**35**:493–7.
- ▶ **Identification and description of perimesencephalic SAH.**
- 6 Linn F, Rinkel G, Algra A, et al. Headache characteristics in subarachnoid haemorrhage and benign thunderclap headache. *J Neurol Neurosurg Psychiatry* 1998;**65**:791–3.
- ▶ **Important paper indicating the limitations of history in distinguishing between benign and malignant “sudden” headache syndromes.**
- 7 Warlow CP, Dennis MS, van Gijn J, et al. What pathological type of stroke is it? In: *Stroke: a practical guide to management*. Oxford: Blackwell Science, 1996.
- ▶ **The only reference you need to learn about SAH, including evidence based diagnosis, prognosis, and management.**
- 8 Linn FHH, Rinkel GJE, Algra A, et al. The notion of “warning leaks” in subarachnoid haemorrhage: are such patients in fact admitted with a rebleed? *J Neurol Neurosurg Psychiatry* 2000;**68**:332–6.
- ▶ **Hopefully the final nail in the coffin of the “warning leak” myth.**
- 9 De Bruijn SFTM, Stam J, Kappelle LJ. Thunderclap headache as the first symptom of cerebral venous sinus thrombosis. *Lancet* 1996;**348**:1623–5.
- ▶ **Describing another potentially serious cause of sudden onset headache to worry about—if only we knew how to best treat it once we have identified it.**
- 10 Wijidicks EFM, Kerkhoff H, van Gijn J. Long-term follow up of 71 patients with thunderclap headache mimicking subarachnoid haemorrhage. *Lancet* 1988;**2**:68–70.
- ▶ **One of several studies confirming the benign outcome of thunderclap headache.**