NEUROLOGICAL MANAGEMENT

Headache

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Current issues: general management**MORBIDITY: THE BURDEN OF HEADACHE TO PATIENT AND SOCIETY**

Morbidity due to headaches is a major problem. Tension headaches outnumber migraine by 5:1. Table 1 compares the more common causes of chronic or recurrent headaches in different age groups.

A recent study from Minnesota, United States, confirms the overall incidence of migraine to be about 10% in the population, with a marked variation related to age and a higher incidence in women in a defined population.¹ From 6400 patient records, 629 residents fulfilled the International Headache Society's (IHS) 1988 criteria² for migraine between 1979 and 1981. The overall age-adjusted incidence was 137 and 294 per 100 000 person years for males and females respectively. The highest incidence was in women aged 20-24 years (689 per 100 000 person years), whereas in boys aged 10-14 years, the incidence was 246 per 100 000 person years.

Based on a study of 1000 people using the International Headache Society's criteria, the overall lifetime prevalence of classic migraine was 5%, with a female to male ratio of 2:1.³ The overall lifetime prevalence of common migraine was 8%, with a female to male ratio of 7:1. Women were more likely to have common than classic migraine. Neither classic nor common varieties correlated with age, but in both types the most conspicuous precipitating factor was stress and mental tension.

The rate of consultation is a concern for those planning medical resources. Among subjects with classic and common migraine, 50% and 62%, respectively, consult their general practitioner because of migraine at

some time in their lives. Patients frequently attend for medical help, and lose much time from work which is often unrecorded. In a random sample of 740 subjects, aged 25-64 years living in the Copenhagen County, Denmark, 119 had migraine and 578 had tension headache (1:5).⁴ Among subjects with migraine 56% had consulted their general practitioner in the previous year for migraine; among subjects with tension headache 16% had had consultations. Specialists had been consulted by 16% of migraine sufferers and by 4% of subjects with tension-type headache. Less than 3% of all patients studied had required hospital admission and laboratory investigations for headache. Half the migraine sufferers and 83% of subjects with tension-type headache in the previous year had taken drug therapy. Thus migraine and tension headaches can be seen as potent sources of demand for medical attention and for the consumption of drugs.

In the Danish study, 43% of employed migraine sufferers and 12% of employed subjects with tension-type headache had lost working time in the preceding year. The total loss of work days due to migraine was estimated at 270 per 1000 persons per year; for tension headache the corresponding figure was 820. Women consult a doctor more often than men, but there is no sex difference in absenteeism.

PRINCIPLE OBJECTIVES AND INSTRUMENTS OF MANAGEMENT

If a patient with headache has cranial arteritis or a brain tumour, then all sociological intervention must take second place to prompt diagnosis and treatment. Over 95% of patients seen in a general practitioner's surgery or hospital clinic, however, have tension headache, migraine, or atypical headache without a structural lesion. For such patients whose symptoms are strongly influenced by social, personal, and family problems, several therapeutic commodities are needed.

For effective therapy doctors need to be accurate in diagnosis, clear in the direction of treatment, honest when they do not know how or why a symptom has developed or changed—yet reassuring. They should be understanding but not dismissive; and they should be prepared to see patients repeatedly until the headaches are controlled, and especially to continue to comfort and support when they are not cured. Such paragons are not ubiquitous. Painstaking efforts to communicate with patients from the start save many subsequent consultations.

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Table 1 Common headaches at different ages

3-16 (years)	17-60 (years)	60+ (years)
Migraine	Migraine	Referred from neck
Tension headache	Tension headache	Cranial arteritis
Psychogenic/fatigue	Cluster headache	Paget's disease of the skull
Post-traumatic	Post-traumatic	Glaucoma
Occasionally	Cranial and dural tumours	Cranial and dural tumours.
Tumours: posterior fossa, intraventricular	Cerebral tumours including abscess and subdural haematoma	Cerebral tumours including abscess and subdural haematoma
	Depression	Rare cluster headache
	Referred from neck	Paget's disease of the skull
	Paget's disease of the skull	Post-herpetic and cranial neuralgias
	Post-herpetic and cranial neuralgias*	Post-traumatic
		Continuing tension headache
		Continuing migraine

*Uncommon presentation at this age.

Political and financial constraints have failed to provide for the necessary number of doctor hours to cater for this ideal. In reality, this has been matched by the never-ending expectations of patients for the latest technical facilities, for a specialist's personal attentions (no longer is a consultant or even a neurologist adequate; it has to be a "headache expert"), and for clinical infallibility—or else, litigation may result.

Such issues of time and expertise have, by default, produced the notion that nurses, social workers, osteopaths, homeopaths, and psychologists, among others, might provide for these patients. Regrettably, they cannot. Although, under medical direction, they can help, they are no substitute for a doctor's training and knowledge. Physicians are unwise to abrogate their traditional role by delegating patients' management to let themselves off the hook. Such fashionable trends denigrate the physicians' role, but much more importantly, diminish their therapeutic effectiveness.

The armamentarium is, however, limited. The frustration of implacable patients is reflected in their search for "alternative therapies". Medical science assesses its tools by rigorous analysis and scientific trials. Medicine should not be daunted by practitioners of alternative medicine since, with rare exceptions, they have so far failed to subject their unbounded claims of therapeutic triumphs to scientific scrutiny. When they do so, and when their methods produce validated benefit, physicians should welcome and use them.

General practitioners provide primary diagnostic facilities which essentially serve to separate those with major pathology (tumours, glaucoma, arteritis) from functional headaches. Many of the latter group are dealt with adequately by history taking, examination, reassurance, and selected analgesics and antimigraine drugs. Patients posing diagnostic problems and those with refractory headaches should be referred to a neurologist.

"Headache clinics" are convenient, and well suited to trials and research; but, to be of benefit to patients they should, ideally, be available on a daily basis, staffed by one or more experienced physicians to provide continuity of service: these ideals are not generally practicable.

Audit

This concept, woolly in definition and often inadequate in application, is not easy to impose on the management of patients with headaches. Patients and their general practitioners can be questioned about their satisfaction with the service; improvement in headache scores would need several applications per patient over a long period of time to demonstrate efficacy above the substantial placebo effect of seeing a new doctor, receiving the investigations that patients perceive as reassuring, and being offered new treatments. Without implementation of results of audit ("closing the loop") and further appraisal, the

exercise is meaningless. To instigate standards of good practice, yardsticks need to be established and agreed.

In a district or regional service, the assessment of wrong diagnoses—for example, missed tumours, subarachnoid haemorrhage, or missed cranial arteritis, is feasible. Similarly errors of diagnosis, choice of first-line treatment and patterns of secondary referral can be appraised in casualty departments and in general practitioner referrals.

The headache patient

This account is restricted to headache and excludes the related but separate facial pains and neuralgias. The clinical approach rests heavily on a detailed analysis of symptoms and a thorough general and neurological examination. Selective investigations are applicable to only a small number of patients who require neurological referral (table 2). Headaches may be acute, usually signifying meningeal irritation or, rarely, raised intracranial pressure (ICP), recurrent as in migraine, or chronic as in tension headaches.

HEADACHE AS A SYMPTOM OF INTRACRANIAL DISEASE

Few headaches fail to evoke some anxiety, which can distort, disguise, or magnify the primary clinical features. Confronted by a patient with headaches, the first clinical responsibility is to exclude a structural or dynamic cause. Any expanding mass—tumour, abscess, or haematoma—can cause raised pressure.

Headaches of abrupt onset may signify trauma, spontaneous intracranial haemorrhage, hydrocephalus, or acute meningeal irritation at any age; the elderly are not immune. The most common cause is acute meningeal irritation due to subarachnoid haemorrhage or to meningitis—bacterial or viral, (rarely HIV, fungal, or malignant). An abrupt onset, fever, neck stiffness, and Kernig's sign accompany the obvious severe pain, vomiting, and photophobia. Hospital referral for CSF examination is mandatory if haemorrhage or infection is considered. CT (or MRI) should be the first investigation to exclude a mass lesion, haematoma, or hydrocephalus, each of which contraindicates lumbar puncture. Subarachnoid haemorrhage may be shown, but can be missed by CT in up to 20% of patients on admission.

When a mass lesion is excluded, lumbar puncture may be necessary to examine the

Table 2 Indications for specialist referral and investigations

- Sudden onset, new headache
- Atypical symptoms or signs suggesting organic pathology
- Abnormal signs—for example, papilloedema to suggest raised intracranial pressure; a red, tender scalp vessel to indicate cranial arteritis
- An unremitting course, unresponsive to conventional treatment
- The advent of progressive physical signs

CSF for subarachnoid bleeding and meningitis. Acute migraine and tension headache uncommonly produce a meningitic picture, diagnosable only by exclusion. Acute headache should be distinguished from the common and totally benign "exploding head syndrome" in which patients are alarmed by a sudden, momentary, very loud noise in the twilight stage of sleep.⁵

Raised-pressure headache

This is an aching, throbbing headache aggravated by alcohol, exertion, and by coughing or straining. These features suggest a vascular or hydrodynamic mechanism which may be caused by a tumour or hydrocephalus. Similar symptoms, however, are also common in migraine and other vascular pains. The location is non-specific, although when a progressive headache radiates to the neck, tonsillar coning is imminent. The headache is: (a) worse in the morning and may waken the patient from sleep; (b) aggravated by sitting up or standing and relieved by lying down; (c) aggravated by coughing, straining, and vomiting; (d) relieved by aspirin or paracetamol in the early stages (in contrast to psychogenic headache); (e) associated with vomiting and eventually by papilloedema and progressing focal signs. By the stage of stupor and hemiplegia with a dilated (Hutchinson's) pupil, diagnosis has been delayed too long.

Chronic and recurrent headaches

REFERRED PAIN

The orbits, paranasal sinuses, cervical spine, mediastinum, and teeth can cause pain referred via branches of the trigeminal nerve to the forehead and temple. Sinusitis and toothache are commonplace examples, but otitis media, glaucoma, orbital cellulitis, and cavernous sinus thrombosis may produce referred frontotemporal pain. Secondary involuntary contraction of scalp and facial muscles further complicate the picture, causing a secondary generalised "tension headache" which may obscure the primary source. Primary care physicians can treat acute infection by antibiotics but, when cranial spread is suspected, patients should be immediately referred to the neurological/neurosurgical centre.

TENSION HEADACHE

Tension headache is the most common of human complaints, constituting 70% of referrals to a "headache clinic". It is often a short-lived complaint with an obvious preceding cause: overwork, lack of sleep, or an emotional crisis. This is benign and often designated by patients as "my normal headaches".

Current systems classify the most common recurring headaches as either migraine or tension type. This traditional approach can be questioned. Some suggest that these two headache patterns are but different expressions of the same pathophysiological process, having overlapping symptomatic presenta-

tions with certain features emphasised to a greater or lesser extent. Additionally, the same therapies have been shown to be effective for patients in either headache group.⁶ An alternative continuum classification model has been suggested, as there is an undoubted overlap between common migraine and tension headache, although their common co-existence has added to the confusion.

By contrast, Solomon and Lipton propose that the diagnosis of migraine without aura (common migraine) is warranted if any two of the following symptoms are present: unilateral site, throbbing quality, nausea, photophobia, or phonophobia.⁷ These criteria are derived from a study comparing the features of 100 patients with migraine without aura and 100 patients with chronic daily ("tension") headache. The authors' proposed criteria for the diagnosis of migraine without aura were highly sensitive and adequately specific in discriminating the two groups. There are also differences in laboratory data. Binding of ³H-labelled imipramine to platelets was measured and a significant reduction was found in migraine compared with controls but not in tension headache. In migraine, there was no significant relationship between imipramine binding and depression or anxiety score suggesting that the reduction in platelet imipramine binding is a concomitant of migraine itself.⁸ A significant reduction in peripheral blood mononuclear cell β -endorphin concentrations was observed in migraine patients with and without aura, but not in patients with tension-type headache.⁹ Adult and childhood migraineurs without aura have an increased amplitude of the contingent negative variation between attacks.¹⁰ Likewise vascular phenomena, well described in migraine, contrast with transcranial Doppler ultrasound of blood flow velocities in chronic tension headache which show no significant differences from controls.

The case for classic migraine as a separate entity is even more persuasive. The teenager with infrequent but prostrating attacks of photopsiae, dysphasia, vomiting, and hemicrania, is at least clinically distinct from the angst-ridden 40-year-old patient with continuous, daily, vertex, pressure headache for 20 years which does not prevent remunerative work.

Acute tension headache

This type of headache rarely presents as an emergency. When it does, the headache has increased over a few hours, but has become very severe, simulating subarachnoid bleeding. Lumbar puncture may be necessary to exclude meningeal irritation by blood or infection. More often the emotional basis is obvious and recovery ensues quickly after reassurance, analgesia, and sedation. Liaison with the general practitioner should lead to continued support and prompt return to work.

Chronic tension headache (chronic daily headache)

More common than the acute syndrome, pain is diffusely felt all over the head, often located

on the vertex, or may start in the forehead or in the neck. Primary tension headache is psychogenic; its mechanisms are not wholly understood.¹¹ It is commonly bilateral, but may be unilateral. Patients characteristically complain of pressure, a feeling of tightness, or a heavy weight pressing on the crown. "A tight band like a skullcap" or "as if a clamp or vice was squeezing my head" are common descriptions. Many say "it is not really a pain, but a pressure". The symptoms may also seem to derive from inside the cranium: "as if my head is bursting" or "about to explode". A "creeping" sensation (formication) may be felt under the scalp, or a sense of sharp knives or burning hot needles driven in, may be related.

Tension headache occurs daily, worse in the evenings. Visual disturbance, photophobia, and vomiting seldom occur. Most patients continue their normal work. Symptoms continue for years without evident deterioration of general health. Symptoms are worse when the patient is tired or under pressure of work, or domestic stresses.¹² Most sufferers have insight, so that a carefully taken history clarifies both the diagnosis and the aggravating circumstances; many are emotional and anxious with fears of brain tumours, hypertension or 'clots in the brain'. I enquire specifically about fears of serious brain disease, whether the fears are voiced by the patient or not. Illnesses seen on television programmes, like maladies of relatives and acquaintances, are often "contagious"; these too need careful appraisal. Thorough clinical examination is of the utmost therapeutic value and provides a rational basis for effective reassurance that is denied to the psychologist or counsellor.

Treatment is most effective when the history is short. To cure such headaches after many years is a daunting and often unsuccessful task.¹³ An important step is to enquire about the events that determined the onset. These are often forgotten, or perhaps suppressed, yet repeated enquiry at subsequent consultations will often unravel the apparent mystery, the consequent knowledge being sufficient to explain the cause to the patient. Sensitive patients with fragile personalities may be unable to cope with life's stresses and unconsciously use headaches to escape responsibilities with which they can't cope. Sedatives, tranquilizers, and tension-relieving drugs are of limited value unless the psychological issues are adequately handled. Glib reassurance will not eradicate headache if fundamental psychological problems are unresolved. When the history is short and if a cause is exposed, explanation and reassurance may suffice.

Patients often abuse analgesics which aggravate the situation, but the abuse, with persuasion, can be reversed with dramatic benefit (see below). More often, when daily pain has persisted for years, the prognosis is poor, but short courses of benzodiazepines or amitriptyline may be helpful. Supportive psychotherapy in liaison with a practice nurse or

psychotherapist may help: much depends on the quality, experience, and good sense of the individual available.

Latent depression presenting as tension headache is easily overlooked. Early morning insomnia, negativism, anhedonia, guilt, and diurnal mood swings are suggestive. The headache is worse in the morning (resembling that of raised pressure), and a cause for the misery is not always apparent. Full doses of tricyclic antidepressants or fluoxetine are needed.^{4,6} The prognosis for depression is often good.

MIGRAINE

Classic migraine

Classic migraine is synonymous with migraine with aura and occurs in 20% of patients. It is a paroxysmal disorder with headaches, often unilateral at the onset, associated with nausea, anorexia, and often vomiting; it is preceded or accompanied by visual, sensory, motor, and mood disturbances and is often familial.

Common migraine

This is synonymous with migraine without aura and occurs in 75% of patients. The term refers to similar paroxysmal headaches without the aura. Both types of attack may occur at various times in the same patient. It is common for migraineurs to have tension headaches between their migraines: these should be identified to prevent misdirected treatment. Daily headaches are never migrainous.

Migraine variants

These are hemiplegic, basilar, and ophthalmoplegic, *migraine sine cephalgia*, etc, and occur in less than 5% of patients, usually requiring a neurologist's appraisal and sometimes brain imaging.

Natural history and management

In childhood, attacks begin before the age of 10 years in a third of patients. They may be overlooked if the child is unable to describe headaches and strange visual or sensory experiences clearly. They are also concealed by labels of "bilious attacks" or "periodic syndrome". Diagnosis can be difficult as affected children may simply appear pale, ill, limp, and inert, complaining of poorly localised abdominal pain. Headache is usually present, vomiting is common, and there may be a fever of up to 38.5°C so that the suspicion of appendicitis or mesenteric adenitis often arises. It should be remembered that over 80% of migraineurs have their first attacks before the age of 30 years, and the diagnosis should therefore be viewed with suspicion if onset is after the age of 40, although an increased frequency of attacks at the menopause is common. Some subjects have only a few attacks in a lifetime but most have several attacks each year. Promises of remission at the menopause are often ill-founded, though attacks tend to lessen after the age of 50 years. Remission occurs in 70% of

pregnancies. It is well recognised that exacerbation, or complicated migraine with infarction, may result from oestrogen-containing contraceptives.¹⁴

Many attacks in adults end gradually after 24 to 48 hours but attacks in children often last only two to six hours. In later life headaches may disappear completely, attacks presenting with teichopsiae and no headache (*migraine sine cephalgia*). The mechanism is uncertain. Indeed, the apparent onset of migraine in the elderly implies atherosclerotic, thromboembolic disease. Food idiosyncrasies, food allergies, red wines, specific dietary amines, and omission of food are occasional precipitants. Emotional stress and fatigue is the principal aggravating factor but is not causal.

Migraine is determined by a primarily cerebral (neural) mechanism with a fluctuating threshold which determines the timing and pattern of attacks.¹⁵ A neural trigger activates the trigeminovascular reflex, releasing vaso-genic amines from blood vessel walls accompanied by their painful, pulsatile distension. The cerebral mechanism responds to mood, emotions, tiredness, relaxation, hormonal changes, and to bright lights, and noise. Its threshold is susceptible to hypothalamic function which, in turn, is modulated by seasonal patterns, diurnal and biological clocks, and by hormonal factors and coitus.¹⁶ Personality and variations of mood and behaviour also influence the pattern of attacks, remissions, and treatment.

SYMPTOMATIC TREATMENT

Assessment of the patient's habits, work, personality, and stresses is important. Suitably trained clinic nurses can assist in the time-consuming elucidation of these factors in selected cases. Known precipitants should be sought, with the aid of a diary and should, when possible, be eliminated (table 3). The patient's recognition of stressful patterns and the acceptance of a benign disorder will achieve some benefit. Good rapport enhances reassurance and facilitates the marked placebo effect of all therapy which often confounds the analysis of drug trials.

The aims of treatment are the control of symptoms, and the prevention or reduction of attacks. Many prophylactic drugs act by central serotonin (5HT₂) antagonism,¹⁷ whereas control of an attack relies on constriction of cranial vessels mediated by α -adrenergic or 5HT₁ receptors.¹⁸

Table 3 Common precipitants of migraine

●	Fatigue, overwork, travel
●	Relaxation after stress—holiday and Saturday morning headache
●	Bright lights, discos
●	Sleep excess or shortage—Sunday morning headache
●	Missing meals
●	Rare dietary sensitivity
●	Alcohol, red wines
●	Menstruation
●	Exercise-related vascular headaches: footballer's migraine, coital cephalgia

Analgesics

Rest, dark, and quiet, where practicable, are supplemented by simple analgesics (paracetamol 1 g or soluble aspirin 0.6 g) or non-steroidal anti-inflammatory drugs such as naproxen 500 mg. The addition of caffeine and spasmolytics add to expense but not to benefit. Codeine 15–30 mg may enhance pain relief. Analgesics should be taken immediately the attack begins, then repeated four to six hourly as needed. Absorption is improved by ingestion with metoclopramide 10 mg or domperidone 10–20 mg. If vomiting is severe, suppositories of domperidone, prochlorperazine, or chlorpromazine are valuable. Analgesic abuse can cause headaches.

Ergotamine

Ergotamine (or dihydroergotamine¹⁹) is an effective remedy for acute attacks in about 50% of cases and should be tried before more expensive agents. Ergotamine has no place as a prophylactic, however, and, when overused, can lead to habituation with ergotamine-dependent headache, similar to chronic analgesic-dependent headaches which mimic migraine status. When suspected, the drugs should be withdrawn, often in hospital under short-term sedation.

Ergotamine is an α -adrenergic agonist with potent 5HT₁ receptor affinity, a potent vasoconstrictor. Absorption is erratic, oral doses often producing sub-therapeutic blood levels. Suppositories (1–2 mg), inhalation (0.36 mg), or sublingual (1–2 mg) are the most effective acceptable routes, but injections are not generally tolerated. If two doses at intervals of two to six hours are ineffective, no more should be given for that attack. If a patient is taking ergotamine more than twice each week, there is a major risk of habituation. Many patients experience vague malaise, nausea, and cramps with ergotamine, but coronary, cerebral, and limb (St Anthony's fire) ischaemia are rare, but well-proven, hazards. Vascular claudication, angina, and pregnancy are contraindications.

Sumatriptan

This is a specific and selective agonist of 5HT₁ receptors on cranial blood vessels causing vasoconstriction.²⁰ It has negligible effects on other receptors. Sumatriptan does not penetrate the blood-brain barrier and has no CNS effects. Intravenous sumatriptan does not change regional cerebral blood flow; it constricts the carotid vascular bed, but has no effect on pial vessels in cats. It has a plasma half-life of two hours.

The subcutaneous preparation (6 mg) gives relief of headache in 77% patients at 60 minutes, and in 83% at two hours with corresponding improvement in nausea, vomiting, and photophobia.²¹ In cluster headache too, it is effective with relief of symptoms at 15 minutes in 74% compared with 26% given placebo.²²

Oral medication (100 mg) provides relief in about 70% of attacks within two hours.²³

Second and third attacks respond as well as the first. Comparative trials have shown slight but significant superiority to aspirin 900 mg plus metoclopramide 10 mg. Recurrence of headache occurs within 48 hours in 42% of patients.²⁴ In comparisons of sumatriptan and Cafergot, oral sumatriptan 100 mg relieved headache after two hours in 66% compared with 48% of those given oral Cafergot.²⁵ The poor oral absorption of ergotamine, however, was a serious limitation of this study.

Another study showed a good response to oral sumatriptan in 51% of patients at two hours compared with 9% given placebo; rescue medication was needed in 41% of the sumatriptan group, but in 88% of the placebo group.²⁶ A total of 39%, however, had recurrent headache within 24 hours—a significant “rebound effect” which may relate to the natural history of migraine, or to a pharmacological effect.

In over 100 000 treated attacks, toxicity has not been a problem.²⁷ There are now proven cardiovascular ischaemic sequelae with angina, infarction²⁸ and ventricular arrhythmias, but these are rare if established ischaemic heart disease, Prinzmetal angina, and arrhythmias are respected as contraindications. Vague non-ischaemic chest discomfort occurs in some patients. Nausea, vomiting, and tingling, deemed mild and transient, occur in 38% patients within four hours. Rebound in a third of patients responds well to further doses, but compounds the pressing issue of cost. Sumatriptan is an effective, safe, and prompt remedy, suppressing all the symptoms, not headache alone.²⁹ It works in 70% of sufferers, however—not in every patient. The present high cost limits its use.

Good communication with family doctors, advice by clinic or practice nurses on the use of self-injectors and inhalers and the frequency of tablets are important. Audit of the results of treatment by completing headache charts can be invaluable, but patients should not overuse diary cards which can engender excessive introspection and neurosis.

PROPHYLAXIS

Prophylaxis should be considered if attacks occur more often than twice each month. Non-pharmacological techniques are successful in certain subjects, but claims for their general application should be viewed with scepticism. Current data support no significant difference in the efficacy of hypnosis, biofeedback, and relaxation training. Prophylactics aim to reduce the frequency, but none is a panacea. They should be given for three to six months, then reassessed. In patients with exacerbations related to stress, amitriptyline 100–150 mg, at night, introduced gradually, is often effective. Which of its actions—sedative, antidepressant, anti-serotonergic, or calcium channel blockade—is implicated, is not known. β -blockers without intrinsic sympathomimetic activity propranolol,³⁰ atenolol, and metoprolol—reduce the frequency in about 60% of cases,

and are most effective in the tense or hypertensive subject with tachycardia and overt physical signs of autonomic “overdrive”. Their action is probably central.

Serotonin inhibitors are valuable in 60–70% of patients. Cyproheptadine 4 mg three times daily inhibits calcium channels, and serotonin and histaminergic activity. Pizotifen 0.5 mg three times daily or 1.5 mg at night is moderately effective and free of hazards other than sedation and weight gain.³¹ Methysergide 1–2 mg three times daily is the most effective drug in this group but should be used under hospital supervision in courses not exceeding three to four months. Pleural, pericardial, and retroperitoneal fibrosis are rare but serious, side effects that result from prolonged use; they usually regress when the drug is stopped. Myocardial and peripheral vascular ischaemia are uncommon complications. Although calcium antagonists can cause a vasodilator-type headache, some—for example, flunarizine and verapamil, have been established as useful prophylactic drugs.³²

Menstrual migraine

Menstrual migraine, defined as occurring exclusively within 48 hours of menstruation, is uncommon but may be relieved by sumatriptan, or oestrogen patches or implants. Migraine between periods but worse with menstruation, is common (35%) but resistant to diuretics and hormonal manipulation.

CLUSTER HEADACHE

Synonyms are migrainous neuralgia, Harris's syndrome, and Horton's syndrome.

Often misdiagnosed, this is a distinct syndrome separable from migraine; it predominantly affects men (male:female ratio 10:1). It begins at any age, most often 20 to 50 years, and is manifest as daily bouts of unilateral headache of great severity lasting 30–120 minutes. The brevity, severity, lack of aura, and vomiting occurring daily in clusters, lasting usually for 4–16 weeks, clearly separate it from migraine. The pain is boring, aching, or stabbing and is centred on one orbit with radiation to the forehead, temple, or cheek and jaw (“lower-half headache”). It characteristically strikes at night, an hour or so after sleep, and may recur during the day, often at the same time (“alarm-clock headache”). In many cases the ipsilateral eye becomes red and bloodshot, watering profusely. The nostril may be blocked or run. A transient Horner's syndrome is seen in 25% of cases and occasionally persists.

Restlessness, crying, and head banging betray the frightening severity of the pain and, in contrast to migraine, most patients get out of bed and pace the floor, even taking nocturnal walks. Alcohol and other vasodilators precipitate attacks: nitroglycerine is useful as a provocative test—a typical attack following within an hour of a sublingual 0.5 mg tablet. “Acute episodic cluster” lasts for one to four months although occasionally they continue for a year or more, when it is

Table 4 Diagnostic criteria for cluster headaches

Episodic
(a) At least five attacks fulfilling (b)–(d)
(b) Severe unilateral orbital or supraorbital pain, with or without temporal pain, lasting 15–180 minutes untreated
(c) Headache is associated with at least one of the following signs which have to be present on the painful side
• Conjunctival injection
• Lacrimation
• Nasal congestion
• Rhinorrhoea
• Forehead and facial sweating
• Miosis
• Ptosis
• Eyelid oedema
(d) Frequency of attacks: from one every other day to eight per day
(e) History and/or physical and neurological examinations do not suggest other disorders associated with head trauma
Chronic
Attacks occur for more than one year without remission, or with remission lasting less than 14 days. The attacks are clinically indistinguishable from episodic cluster headache.

known as “chronic migrainous neuralgia”. Remissions are complete but the clusters recur every year or two. The quality, timing, duration, and distribution of pain separate it from trigeminal neuralgia, migraine, and other cephalgias. There is no family history, and it is uncommon for a patient to have both migraine and cluster headache. There is an unexplained high incidence of peptic ulcer.³³

The diagnostic criteria (IHS) are summarised in table 4.

Treatment

The aim is prevention of attacks.³⁴ During clusters, alcohol is prohibited. Ergotamine is given one hour in *anticipation* of daytime attacks, and at bedtime for nocturnal attacks. Suppositories are the most useful preparation. Control is good in 75% patients and the drug is stopped each Sunday to see if the cluster re-emerges; if so, ergotamine is continued for a further week, until the cluster ends. If ergot is unsuccessful, sumatriptan 6 mg subcutaneously or 100 mg orally,²² methysergide 1–2 mg three times daily or verapamil 40–80 mg three times daily are useful alternatives. Oxygen 5–10 litres per minute for 10 minutes at the onset is often effective²⁹ but β -blockers and pizotifen are not. Lithium is useful in the chronic variant if other methods fail. In intractable cases, a short course of steroids often provides relief. Surgery is seldom indicated, but trigeminal lesions meet with occasional success in refractory patients.

Chronic paroxysmal hemicrania

This is a rare variant³⁵ of cluster headache, occurring predominantly in females, with identical attacks, often five to 20 per day, which last from three to 15 minutes; they respond almost invariably to indomethacin, 75–150 mg daily.

CERVICOGENIC HEADACHE

Head pain referred from cervical spondylosis is undoubtedly common, with pain on one or both sides of the neck radiating not only to the occiput but also to the temples and frontal region. It may be a dull “toothache” pain, worse in the morning when the neck has

been kinked on high pillows during sleep; it can initiate migraine. It can last throughout the day, aggravated by neck movement and tension and is a nondescript pain, without accompanying vomiting or physical signs other than restriction of lateral flexion and rotation of the neck. Such signs are common, however, in those without headache. Vague and intermittent symptoms of tinnitus, dizziness, and visual disturbance are sometimes attributed to compression of the vertebral arteries, but this is unproven. Pain arises from the posterior zygapophyseal joints and related ligaments as the result of osteophytes with irritation of the C₂ root or greater occipital nerves.³⁶ Manipulation endangers the vertebral arteries and is contraindicated. Collars are comforting but of little value. Injections of the facet joint region anatomically related to pain will induce useful temporary remission in about 70% of subjects.³⁷ Hydrocortisone 25 mg or methylprednisolone are used, with 1 ml of 2% lignocaine on three occasions at weekly intervals. The benefit may wane in a few months, when the injections can be repeated.

GIANT CELL ARTERITIS

Synonyms are cranial or temporal arteritis.

This condition is crucially important as a treatable cause of headache, it is also a preventable cause of blindness and strokes. The diagnostic features are shown in table 5.

Pain is generalised or may be sited over the classic, but rare, reddened, tender, superficial temporal or occipital artery. The history is of a few weeks' duration. The patient is unwell with aches and pains in the shoulder and pelvic girdle muscles (polymyalgia rheumatica) and there may be fever, sweats, and masseter claudication. Visual involvement (50% patients) is due to an ischaemic optic neuropathy which presents with unilateral blindness or due to a branch retinal or ciliary occlusion and is irreversible. Posterior cerebral artery lesions cause a hemianopia.

Ophthalmoplegia and diplopia may be the presenting sign, based on ischaemic lesions in the third, fourth or sixth cranial nerves, before the onset of headache and malaise: hence the importance of early diagnosis. It affects the vertebral, and less often the carotid, arteries and may present as a stroke or transient ischaemic attack.

Every elderly subject with recent headaches or unilateral visual loss should be suspected of harbouring this condition. Serial erythrocyte sedimentation rates (ESR; usually

Table 5 Diagnostic criteria for cranial arteritis³⁸

• Age ≥ 50 years at onset
• New onset of localised headache
• Temporal artery tenderness or decreased temporal artery pulse
• Elevated ESR ≥ 50 mm/h
• Biopsy showing necrotising arteritis

The presence of three or more of these five criteria provides a sensitivity of 93.5% and a specificity of 91.2%. Scalp tenderness and claudication of the jaw or tongue or on deglutition increases sensitivity to 95.3%

ESR = erythrocyte sedimentation rate.

70–120 mm/h), sometimes supplemented by an adequate biopsy of the clinically affected scalp vessel, which should be serially sectioned, will prove the diagnosis. Cranial arteritis may spontaneously remit and the ESR then falls; thus a single, normal ESR does not exclude arteritis. Biopsy is indicated in clinically borderline cases.

Atypical presentations that should prompt assiduous investigation include: (a) patients with minimal headache; (b) headache in an appropriately aged patient with, initially, a normal ESR; (c) fever of unknown origin; (d) psychiatric symptoms of hallucinations, depression, and “confusional state”; (e) isolated third or sixth nerve palsy; rarely internuclear ophthalmoplegia.

Steroids will avert blindness in almost every case, and should be started immediately the patient is seen and the blood sample for an ESR taken. They do not affect the biopsy changes for at least 48 hours. Headaches often abate within 24 hours of treatment.

The initial dose of 60 mg prednisolone daily is quickly reduced as symptoms abate and the ESR falls. The maintenance dose of 5–10 mg daily is usually reached within a month or two and is governed by clinical progress and ESR measurements. Late relapses are common³⁹ and treatment may be required for many years with gradual reductions by 1 mg per month, only when the patient and the ESR have been normal for over two years. In many, small doses are necessary for life, as proved by serious relapses months after the dose of prednisolone is reduced.³⁹

HYPNIC HEADACHE (SOLOMON'S SYNDROME)

This is a curious headache, seen mainly in those over 60 years. Patients are woken by pulsating headache, sometimes accompanied by nausea, at the same time, one to three times each night.⁴⁰ This occurs most nights, lasts about 30 minutes and may coincide with rapid eye movement sleep. It is uncommon and differs from chronic cluster headache in age, generalised location, and absence of autonomic features. There are no physical signs and the disorder is benign. The response to lithium carbonate 300 mg at night is often spectacular.

POST-TRAUMATIC HEADACHE AND ITS MANAGEMENT

Headache after head injury is a common complaint. In most circumstances a knock on the head will cause local bruising and abrasions no different from those resulting from a kick on the shin; local pain subsides in three to 10 days without sequelae. The emotional vulnerability of the head and the easy recourse to medicolegal compensation complicate both symptoms and mechanisms. Many victims of severe head injury, with post-traumatic amnesia of 24 hours or more, waken with no headache. Similarly, the headache of patients after major craniotomy seldom lasts more than three to seven days.

The commonest complaints are heard from those with minor injury (loss of consciousness less than 20 minutes; Glasgow coma score 13–18; stay in hospital less than 48 hours). Despite minor cognitive deficits, most cases leave hospital within a few days, have no organic signs, recover quickly, and return to work without further complaints. This is particularly true of those suffering injury during contact sports. Attributable, post-traumatic headaches persist for a few weeks, and seldom more than six months, unless complicated by other issues.

The main concern of physicians is the assessment of symptoms in those with headaches without accompanying neurological signs, but often with a collection of intrinsically subjective symptoms often called the “post-traumatic syndrome”.⁴¹ The complaints are: forgetfulness, irritability, slowness, poor concentration, fatigue, dizziness (usually not vertigo), somnolence, intolerance of alcohol, light, and noise, loss of initiative, depression, anxiety, loss of interests, and impaired libido. The number of complaints is often inversely related to the severity of the injury.

In 2493 individuals examined as part of a nationwide general population survey, post-traumatic stress disorder was found in 1% of the total population, about 3.5% in civilians exposed to physical attack and in Vietnam veterans who were not wounded, and 20% in veterans wounded in Vietnam.⁴² Although hyperalertness and sleep disturbances, occurred commonly in the general population, the full syndrome (DSM-III-R) was common only among veterans wounded in Vietnam. This argues against the validity of the widespread use of this label in those subjected to minor injuries.

Trauma probably never causes migraine, but pre-existing migraine may be temporarily worse, usually for up to three to six months, probably as a non-specific reaction to stress or to disabilities.⁴³

The failure of doctors to provide complete reassurance soon after injury is important in determining patients' fears of brain damage or subdural haematoma; it also delays return to work and induces iatrogenic morbid anxiety. Headaches are at the site of trauma, often with scalp tenderness, or more often—like tension headaches—are diffuse, aching, tight, or heavy. They resist analgesics, and investigations in most patients are both unrevealing and unwarranted. Headaches sometimes improve when the patient is counselled and returns to work, but do not invariably disappear even when satisfactory settlement is attained. Anxieties, phobias, loss of self esteem, resentment, and depression are genuine accompanying features in some cases, and serve to induce or to aggravate headache. Deliberate exaggeration, or malingering in occasional cases, are motivated by quest for attention and financial gain. In litigants there is great pressure from trades union officials and lawyers which, together with the common delay in settlement, serve to prolong and exaggerate the symptoms.

Table 6 Simplified plan of management for headache

Patient presents with headaches of abrupt onset

- Exclude trauma. If signs of meningeal irritation, suspect intracranial haemorrhage or meningitis and admit to hospital. Perform CT or MRI and, if no tumour, haematoma, or hydrocephalus found, perform lumbar puncture
- NB: lumbar puncture within six hours of the ictus may miss an early bleed
- If no signs of meningeal irritation, consider a mass lesion, arrange CT or MRI and refer to neurosurgery if necessary

Patient presents with increasing headache, short history

- Exclude local cranial pathology, such as glaucoma, sinusitis, dental disease; look for signs of raised intracranial pressure, suspect mass or hydrocephalus. Arrange CT or MRI before lumbar puncture

Patient presents with chronic or intermittent headache with no neurological signs

- Exclude local cranial pathology: glaucoma, sinusitis, dental disease. If continuous, suspect tension headache; if paroxysmal, suspect migraine with or without aura. Look for features of cluster headache, other headache syndromes, and cranial neuralgias

POST-HERPETIC NEURALGIA

This dreaded complication of trigeminal herpes zoster, particularly common in the elderly (50% of over 70s) and rare under the age of 60, is defined by pain persisting for more than one month after the eruption. Two-thirds spontaneously recover in one year, leaving a minority with protracted and intractable pain. Errors in diagnosis are avoided by insisting on the presence of healed post-herpetic hyperpigmented or depigmented areas, or both, in a segmental distribution. An underlying neoplasm of the affected roots and systemic immunosuppression should be excluded by judicious investigation.

The mechanism of pain is uncertain. The preferential loss of large diameter neurons, according to the "gate theory" of Melzack and Wall, permits increased transmission of nociceptive information through the dorsal horn, thus evoking pain. There can be no general recommendation for acyclovir or steroids in the acute stages, but in the frail and elderly who are highly vulnerable, it may be justifiable to employ both drugs⁴⁴ at the onset of herpes zoster infection in an attempt to avert this potentially grave affliction, in which suicide is well-known. In the established case, symptomatic improvement is disappointing. It is worth trying tricyclics building slowly up to high doses (such as imipramine or amitriptyline 125 to 200 mg daily). Additional regular non-opiate analgesics by day and, in severe cases, oral morphine or heroin at night will often be justifiable. Local counterirritants, now called "neuroaugmentation", in the form of freezing sprays or topical capsaicin 0.025% cream can be helpful. Transcutaneous electrical nerve stimulators find occasional success.

Conclusion

Most headaches presenting to the general practitioner and hospital physician have no ominous intracranial cause, but are a source of suffering and loss of working time. Many headaches are infrequent and self-limiting. A painstaking clinical approach by a good listener will resolve many problems and will prevent the refractory course in some of those in whom a cursory initial examination fails to

secure reassurance. Patients presenting problems in diagnosis, and those unresponsive to appropriate treatment will benefit by referral to a neurologist.

Table 6 shows a simplified plan of management.

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