GENERAL APPROACH AND CLASSIFICATION OF HEADACHES

Headache is one of the most common medical complaints. Most of the population will have experienced headache, and over 5% will seek medical help.



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Pierre Bill was Professor and Head of Neurology from 1984 to 2003. He is involved in undergraduate and postgraduate teaching of neurology, development of curriculum for undergraduates in neurology, and in general neurology, neuromuscular and clinical neurophysiology services. Current research interests are in the molecular diagnosis and pathophysiology of Duchenne and limb girdle dystrophies. Most chronic headaches are symptomatic of a primary headache disorder, while secondary headaches are symptomatic of a wide variety of conditions, including ophthalmological, sinus and dental disorders, infection, brain tumour, cerebral haemorrhage and meningitis. Doctors should therefore be knowledgeable in the diagnosis and treatment of headache. The key to the diagnosis is the history. Pain and associated symptoms must be described by the patient. There are no specific tests for many of the headache syndromes; rather diagnostic tests help to exclude secondary causes.

Headaches are classified into 2 major groups: primary and secondary headache disorders. $^{\scriptscriptstyle \rm 14}$

PRIMARY HEADACHE DISORDERS

- Migraine
 - migraine without aura
- migraine with aura
- other forms of migraine.
- Tension-type headache
 - episodic tension-type headache
 - chronic tension-type headache
 - probable tension-type headache.
- Cluster headache and other trigeminal autonomic cephalgias
 - cluster headache
 - paroxysmal hemicrania
- short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
- probable trigeminal autonomic cephalgia.
- Other primary headaches
- primary stabbing headache
- primary cough headache
- primary exertional headache
- primary headache associated with sexual activity
- hypnoic headache
- primary thunderclap headache
- hemicrania continua
- new daily persistent headache (NDPH).

SECONDARY HEADACHE DISORDERS

- Headache attributed to head and/or neck trauma (acute and chronic post-traumatic headache).
- Headache attributed to cranial or cervical vascular disorders (subarachnoid haemorrhage and giant cell arteritis).
- Headache attributed to nonvascular intracranial disorders (idiopathic intracranial hypertension, hydrocephalus, neoplasm and postictal headache).

Around 50 - 60 % of migraineurs have a parent with the disorder and up to 80% have a first-degree relative with migraine.

It is well documented that frequent and regular use of acute antimigraine medication and/or analgesics by people with migraine or tension-type headache risks aggravation of the primary headache.

Associated symptoms and signs

The following are important: redness and tearing of the eyes as well as nasal congestion may occur with cluster headache and migraine. Anorexia, nausea and vomiting are most commonly associated with migraine but may occur with any severe headache. Polyuria is commonly associated with migraine. Signs of depression are frequently associated with longstanding headache. Mood changes may also be associated with the migraine attack. This may take the form of irritability, depression or euphoria.

Signs of neurological dysfunction

The physical examination is important to rule out systemic causes of headache and to identify neurological abnormalities that would suggest an intracranial lesion. Some of the important aspects, from a neurological point of view, are:

- auscultation of eyes, carotids, and vertebral arteries
- funduscopy
- palpation of structures in and about the face and head
- neurological examination.

Neurological dysfunction may be associated with a migraine attack or indicate a space-occupying lesion. Vertigo may occasionally be a forerunner of a migraine attack, and is sometimes associated with brain tumours (usually with other symptoms and signs).

Precipitating and relieving factors

Identifying precipitating factors may be useful. Alcohol, fatigue, stress, drugs, loss of sleep, food additives and menstruation can provoke migraine, as can exercise and orgasm. Migraineurs commonly volunteer that they must retire to a dark, quiet room: many patients find that sleep will relieve their attacks. Cluster headache patients note that sitting upright, rocking, or pacing to and fro lessens the pain. This behaviour is characteristic of cluster headache.

Effect of head jolt

Headaches that arise from dilatation of pain-sensitive intracranial vessels (fever, systemic infection, 'hangover', post lumbar puncture, hypoglycaemia, cluster headache, migraine) or inflammation of pain-sensitive structures (meningitis) are particularly sensitive to head jolting. Tension-type headaches are not intensified by head jolting.

Family history

Around 50 - 60 % of migraineurs have a parent with the disorder and up to 80% have a first-degree relative with migraine.

CAUSES FOR CONCERN

The doctor should be especially concerned if the patient has any of the following:

- a new-onset headache in a patient over the age of 50 years
- a sudden-onset headache
- a headache that is subacute in onset and gets progressively worse over days or weeks
- a headache associated with systemic illness (fever, neck stiffness, rash)
- a headache associated with focal neurological signs, other than the typical visual or sensory aura of migraine
- papilloedema

- headache triggered by cough, exertion or Valsalva
- headache during pregnancy or postpartum
- no obvious identifiable headache aetiology
- a new-onset headache in a patient with cancer or HIV.

Conditions that may cause the above include subarachnoid haemorrhage (SAH), bleeding, mass lesions, subdural haematoma, meningitis, collagen vascular disease, encephalitis, cortical vein thrombosis, carotid dissection, metastasis, and opportunistic infection.

PRIMARY HEADACHE SYNDROMES

The key to the diagnosis of a primary headache is a knowledge of the key features of the individual primary headache syndromes. The criteria for the diagnosis of the more common types of primary headache are given below.

The diagnosis of **migraine without aura** is made on the criteria of at least 5 attacks fulfilling the following:

- Headache lasting 4 72 hours
- Headache with 2 or more of the following characteristics:
 - unilateral location
 - pulsating quality
 - moderate or severe intensity
- aggravation by routine physical activity
- At least one of the following:
 nausea and/or vomiting
 photophobia and phonophobia
- No evidence of organic disease.
- No evidence of organic disease.

Criteria for diagnosis of **migraine** with aura:

- At least 2 attacks with 3 of the following:
 - reversible aura symptoms
 - aura symptoms develop gradually over more than 4 minutes
 - no aura symptom lasts more than 60 minutes
 - headache follows aura with a free interval of less than 60 minutes (however the headache may begin before or with the aura).

Criteria for **episodic tension-type headache** – at least 10 headache episodes with the following criteria:

- Number of days with such headaches ≤ 15/month
- Headache lasting from 30 minutes to 7 days
- At least 2 of the following pain characteristics:
 - pressing/tightening quality
 - mild or moderate intensity
 - bilateral location
 - no aggravation by routine physical activity
- Both of the following:
 - no nausea or vomiting
 - no more than one of photophobia or phonophobia
- Absence of organic disease.

Episodic tension headache may be associated with tenderness of the pericranial muscles.

Diagnostic criteria for chronic tension-type headache:

- Average headache frequency 15 or more days/month for 6 months
- At least 2 of the following:
 - pressing/tightening quality
 - mild or moderate severity
 - bilateral location
 - no aggravation by routine physical activity
- Both of the following:
 - no vomiting
- no or more than one of: nausea, photophobia, or phonophobia
- No evidence of organic disease.

Chronic tension-type headache may or may not be associated with tenderness of the pericranial muscles.

Diagnosis of **cluster headache** – at least 5 attacks fulfilling the following criteria:

- Severe unilateral orbital, supraorbital, and/or temporal headache lasting 30 – 180 minutes untreated
- Headache associated with one or more of the following: conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis, eyelid oedema
- frequency of attacks: 1 every other day – 8/day

Table I.Differentiating features of short-lasting chronic dailyheadaches

	Cluster	СРН	SUNCT
Sex F:M	1: 4	2:1	1:3
Attack duration	15 - 180 min	2 - 30 min	5 - 240 sec-
onds Attacks per day Autonomic features	2 - 8 Present	1 - 40 Present	3 - 200 Present
Site of maximal pain	Orbit, temple	Orbit, temple	Orbit, temple
Pain severity	Very severe	Severe	Moderate

• no evidence of organic disease.

Cluster headache may be episodic or chronic. In the episodic type, the cluster periods last from 7 days to 1 year, separated by at least 14 days. In chronic cluster headache there is an absence of remission for more than 1 year.

AN APPROACH TO PRIMARY HEADACHE SYNDROMES⁵

The majority of chronic daily headaches is attributable to benign primary headache disorders. A thorough history is the most critical aspect of the evaluation and provides the diagnosis in the majority of cases. However, one must be vigilant for secondary causes of headaches, e.g. disorder of intracranial pressure (increased or decreased), infectious causes, sphenoid sinusitis, obstructive sleep apnoea, temporomandibular joint disorders and dental pathology, and cervical spine disorder.

A useful approach to primary chronic daily headache (headache present on 15 or more days per month) is to consider 2 categories – headaches that are short-lasting (< 4 hours if untreated), and those that are long-lasting (> 4 hours if left untreated) (see Fig. 1).

Short-lasting chronic daily headaches (CDH) are chronic clus-

ter, chronic paroxysmal hemicrania (CPH), and SUNCT (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing), and hypnoic headache (Table I). The essential features of these conditions can be summarised as follows: Hypnoic headache (alarm-clock headache) is a rare benign syndrome characterised by a dull headache, occurring only during sleep, and waking the patient at a consistent time, usually between 01h00 and 03h00. It lasts approximately 1 hour, and occurs from once per week to 6 per night. It is distinct from cluster headache in its lack of unilaterality or cranial autonomic symptoms. It is associated with REM sleep.

Long-duration chronic daily

headaches are diagnosed according to the criteria given above. In chronic migraine, the headaches fulfil the criteria for migraine without aura on more than 15 days per month for more than 3 months in the absence of medication overuse. Hemicrania continua is a continuous unilateral headache defined by its absolute response to indomethacin. Autonomic symptoms occur during painful exacerbations but are less prominent than in cluster headache. New daily persistent headache begins without an evolution from episodic headache and then persists.

Medication overuse headache, classified as a secondary headache disorder, is an additional complication to the above types and is a common and disabling disorder, defined as the generation, perpetuation, or maintenance of chronic head pain in headache sufferers, caused by the frequent and excessive use of immediate relief medications. It is well documented that frequent and regular use of acute antimigraine medication and/or analgesics by people with migraine or tensiontype headache risks aggravation of

CLASSIFICATION

There are no specific tests for many of the headache syndromes; rather diagnostic tests help to exclude secondary causes.

The most important tool for making a diagnosis is a detailed and relevant history, as most patients have a normal neurological and physical examination.

Headache associated with brain tumours is usually only moderately severe.

- Headache attributed to a substance or its withdrawal (carbon monoxideinduced headache, medicationinduced headache, and oestrogen withdrawal headache).
- Headache attributed to infection (intracranial infection, meningitis).
- Headache attributed to disorder of homeostasis (metabolic disorders).
- Headache or facial pain attributed to disorders of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures.
- Cranial neuralgias and central causes es of facial pain (trigeminal neuralgia, occipital neuralgia, post-herpetic neuralgia, and ophthalmoplegic migraine).
- Other headache, cranial neuralgia, central or primary facial pain.
- Headache attributed to psychiatric disorder (somatisation and psychotic disorder).

The most important tool for making a diagnosis is a detailed and relevant history,^{4,5} as most patients have a normal neurological and physical examination. If the patient has more than one type of headache, begin with the most important one for the patient before exploring the other headache types. The following are the important features to elicit when taking a history:^{4,5}

Age of onset

Primary headaches often begin in childhood, adolescence, or the second and third decade of life. Headaches that begin after the age of 50 more frequently have an organic aetiology. Tension-type headaches however can start at any age.

Location of headache

The trigeminal nerve is the major source of innervation for pain-sensitive structures in the supratentorial space. Lesions in this region therefore often cause frontal headache. Lesions in the pituitary or parasellar area often cause bifrontal or bitemporal headache. The headache of tooth, sinus or eye disease is usually frontal. Trigeminal neuralgia may cause pain in any part of the face innervated by the trigeminal nerve. Infratentorial pain-producing structures are innervated by the upper cervical and the glossopharyngeal and vagus nerves, resulting in pain being referred to the occipital and posterior cervical region.

In migraine, the headache is often unilateral, often alternating sides, but may be bilateral. The pain can occur anywhere in the head or face. Cluster headache is almost always unilateral. Tension-type headache is often most intense in the neck, shoulder and occipital area, but may also be frontal. The headache may be unilateral or bilateral. Focal headaches imply focal disease, such as local infection, sinusitis or inflammation or disease of the facial organs, unless the headache is typically migrainous or suggests cluster headache.

Frequency

The frequency and pattern provide clues to the diagnosis. Cluster headaches occur in brief attacks lasting 30 - 90 minutes and recurring 2 -6 times a day. Migraine also occurs at sporadic intervals. Chronic tensiontype headaches occur on more than 15 days per month, whereas episodic tension-type headache occurs on less than 15 days per month. Organic headaches do not occur with any set pattern. Headaches of increasing frequency require diagnostic evaluation.

Onset, duration, character and severity

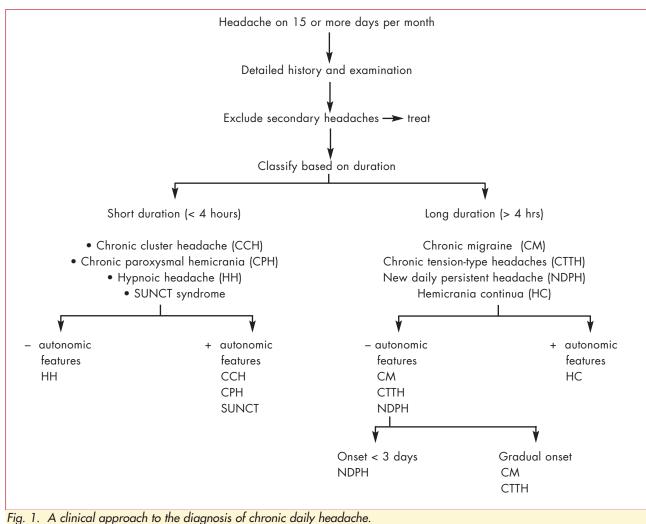
The severity and rapidity of onset and resolution and character are diagnostically important. The headaches of fever and migraine are characteristically throbbing or pulsating in quality. The headaches of brain tumour and meningitis usually have a steady aching quality. Tension-type headache is nagging, dull and persistent, often described as a band around the head. The most intense headaches are those associated with migraine, malignant hypertension, meningitis, and ruptured intracranial aneurysm. Headache associated with brain tumours is usually only moderately severe.

Chronological features

Migraine headaches typically last 4 -72 hours, but may be as brief as 20 -30 minutes. They frequently occur in the early morning hours. There is no headache between the attacks. The headaches of brain tumours are intermittent and vary in severity, and may be worse in the early part of the day. Cluster headache frequently occurs after sleep onset. Headaches that progressively worsen may have an organic cause. However the longer a headache has existed in its present form, the more likely it is to be benign.

Prodromes, auras and visual disturbances

Prodromes such as mood changes and change in appetite may occur 1 - 2 days before a migraine attack. Auras such as scintillating scotomata, hemianopia or paraesthesias precede and define classic migraine, but may also occur with lesions in the occipital lobes or adjacent to the visual pathways. In migraine the visual disturbances usually precede the headache and last less than 1 hour. When the visual defects outlast the headache attack it is likely that a cerebrovascular disturbance or brain tumour is the cause. Glaucoma may also cause visual disturbances.



rig. 1. A clinical approach to the diagnosis of chronic daily headac

the primary headache. The limits indicated for triptans, ergotamine, opioids, or combination analgesics are used on 10 or more days per month, and simple analgesics and non-steroidal antiinflammatory drugs are used on 15 or more days per month.

Making an accurate diagnosis is necessary as different headache disorders may require different treatments. A good understanding of the classification of headache disorders enables better understanding of headache and ultimately betters management of this set of disabling conditions.

References available on request.

IN A NUTSHELL

Headache is a common presenting complaint.

History is the most important tool for diagnosis.

There are 2 major types of headache disorder – primary and secondary.

There are certain features that are cause for concern, for example, onset of a new headache after the age of 50, new-onset headache in a patient with HIV, headache associated with pregnancy.

Most chronic daily headaches are benign.

Analgesic overuse is a common cause of chronic daily headache.

