Secondary Headache Disorders

Secondary Headaches
- When a new headache occurs for the first time in close temporal relation to another disorder that is known to cause headache, the new headache is coded as a secondary headache attributed to the causative disorder.
- This remains true even when the headache has the characteristics of a primary headache (migraine, tension-type headache, cluster headache or one of the other trigeminal autonomic cephalalgias).

Headache attributed to trauma or injury to the head and/or neck
- The most common secondary headache disorders.
- During the first 3 months from onset they are considered acute; if they continue beyond that period they are designated persistent.
- Most often these resemble tension-type headache or migraine. Consequently their diagnosis is largely dependent on the close temporal relation between the trauma and headache onset.
- Headache must be reported to have developed within 7 days of trauma or injury, or within 7 after regaining consciousness and/or the ability to sense and report pain when these have been lost following trauma or injury.

Headache attributed to cranial or cervical vascular disorders:
- The close temporal relationship between the headache and neurological signs is crucial in establishing causation. In many of these conditions, such as ischaemic or haemorrhagic stroke, headache is overshadowed by focal signs and/or disorders of consciousness. In others, such as subarachnoid haemorrhage, headache is usually the prominent symptom.
- In a number of other conditions that can induce both headache and stroke, such as dissections, cerebral venous thrombosis, giant cell arteritis and central nervous system angiitis, headache is often an initial warning symptom.

Ischemic stroke
- Headache attributed to ischaemic stroke (cerebral infarction) is usually of moderate intensity, and has no specific characteristics. It can be bilateral or unilateral (ipsilateral) to the stroke. Rarely, an acute ischaemic stroke, notably a cerebellar infarction, can present with an isolated sudden (even thunderclap) headache.
- It accompanies ischaemic stroke in up to one-third of cases; it is more frequent in basilar- than in carotid-territory strokes.
- Headache attributed to transient ischaemic attack is more common with basilar- than carotid-territory TIA, it is rarely a prominent symptom of TIA.

Intracerebral Haemorrhage
- Headache attributed to non-traumatic intracerebral haemorrhage is more often a result of associated subarachnoid blood and local compression than intracranial hypertension.
- Headache can occasionally present as thunderclap headache. The headache is usually overshadowed by focal deficits or coma, but it can be the prominent early feature of some
intracerebral haemorrhages, notably cerebellar haemorrhage, which may require emergency surgical decompression.

**Subarachnoid haemorrhage**

- Headache is typically severe and sudden, peaking in seconds (thunderclap headache) or minutes. It can be the sole symptom of SAH.
- Subarachnoid haemorrhage (SAH) in the absence of head trauma has been diagnosed.
- Evidence of causation demonstrated by: clinically headache has developed in close temporal relation to other symptoms and/or clinical signs of SAH, or has led to the diagnosis of SAH.

**Unruptured cerebral aneurysm**

- Headache is reported by approximately one-fifth of patients with unruptured cerebral aneurysm.
- Headache usually has no specific features. On the other hand, a new-onset headache can reveal a symptomatic but unruptured saccular aneurysm. A classic variety is acute IIIrd nerve palsy with retro-orbital pain and a dilated pupil, indicating an aneurysm of the posterior communicating cerebral artery or termination of the carotid artery. Such painful IIIrd nerve palsy is an emergency.

**Warning leaks**

- Half of patients with an aneurysmal subarachnoid haemorrhage reported the occurrence of a sudden and severe headache within the 4 weeks prior to the diagnosis of aneurysmal rupture.
- This suggests these headaches are a result of sudden enlargement of the arterial malformation (‘sentinel headache’) or to mild subarachnoid haemorrhage that is not diagnosed as such (‘warning leak’).
- Patients should undergo complete investigation, including cerebral imaging, CSF study and cerebral angiography.

**Arteritis**

Giant cell arteritis is the disease most conspicuously associated with headache, which is a result of inflammation of cranial arteries, especially branches of the external carotid artery.

The variability in the features of headache attributed to giant cell arteritis and in the other symptoms of GCA (polymyalgia rheumatica, jaw claudication) are such that any recent persisting headache in a patient over 60 years of age should suggest GCA and lead to appropriate investigations.

**Giant Cell Arteritis**

Recent repeated attacks of amaurosis fugax associated with headache are strongly suggestive of GCA, and should prompt urgent investigations.

The major risk is of blindness as a result of anterior ischaemic optic neuropathy, which can be prevented by immediate steroid treatment; the time interval between visual loss in one eye and in the other is usually less than 1 week. Patients with GCA are also at risk of cerebral ischaemic events and of dementia.

CADASIL
CADASIL is an autosomal dominant disease with some sporadic cases, involving the smooth muscle cells in the media of small arteries of the brain; the diagnosis is made by screening for NOTCH-3 mutations or by a simple skin biopsy with immunostaining of NOTCH-3 antibodies. CADASIL is characterized clinically by recurrent small deep infarcts, subcortical dementia, mood disturbances and, in one-third of cases, by migraine with aura. In such cases, this is usually the first symptom of the disease, appearing at a mean age of 30. Migraine attacks are typical of an unusual frequency of prolonged aura. MRI shows striking white matter changes on T2-weighted images.

MELAS (Mitochondrial Encephalopathy, Lactic Acidosis and Stroke-like episodes) is a genetically heterogeneous mitochondrial disorder with a variable clinical phenotype, central nervous system involvement as (seizures, hemiparesis, hemianopia, cortical blindness, sensorineural deafness and/or episodic vomiting) and, frequently headache, which is either recurrent in migraine-like attacks or a presenting symptom of stroke-like episodes.

Headache attributed to non-vascular intracranial disorder

Headache attributed to increased cerebrospinal fluid pressure:
- Headache attributed to idiopathic intracranial hypertension (IIH)
- Headache attributed to intracranial hypertension secondary to metabolic, toxic or hormonal causes
- Headache attributed to intracranial hypertension secondary to hydrocephalus

Syndrome of transient Headache and Neurological Deficits with cerebrospinal fluid Lymphocytosis
It is of 1–12 discrete episodes of transient neurological deficits accompanied or followed by moderate to severe headache. Most of the episodes last hours, but some may last for more than 24 hours. The neurological manifestations include sensory symptoms in about three-quarters of cases, aphasia in two-thirds and motor deficits in a little. The syndrome resolves within 3 months.

In addition to CSF lymphocytosis (up to 760 cells/ml), there are elevations of CSF total protein (up to 250 mg/dl) in >90% of cases and of CSF pressure (up to 400 mm CSF) in more than 50% of cases. The presence of a viral prodrome in at least one-quarter of cases has raised the possibility of an autoimmune pathogenesis of Syndrome.

Colloid Cyst of third ventricle
The vast majority of colloid cysts of the third ventricle are discovered incidentally, having been asymptomatic.
Nevertheless, their position immediately adjacent to the foramen of Monro can, on occasion, result in sudden obstructive hydrocephalus, causing headache with thunderclap onset and reduced level or loss of consciousness.

This highly characteristic presentation should lead to rapid diagnosis

Pituitary tumor
Headache caused by a pituitary adenoma and hypothalamic or pituitary hyper- or hyposecretion, usually accompanied by disorder of temperature regulation, emotional state and/or altered thirst or appetite. It remits after successful treatment of the underlying disorder.

Pre ictal headache
Headache is frontotemporal, ipsilateral to the focus with temporal lobe epilepsy (TLE) and/or some times frontal lobe epilepsy.

Hemicrania epithleptica
Headache occurring during a partial epileptic seizure, ipsilateral to the epileptic discharge, and remitting immediately or soon after the seizure has terminated.

Medication overuse headaches
Medication-overuse headache is an interaction between a therapeutic agent used excessively and a susceptible patient.

Among those with a previous primary headache diagnosis, most have Migraine or Tension-type headache (or both); only a small minority have other primary headache diagnoses such as Chronic cluster headache or New daily persistent headache.

Approximately half of people with headache on 15 or more days per month for more than 3 months have Medication-overuse headache.

Majority of patients improve after discontinuation of the overused medication, as does their responsiveness to preventative treatment.

Prevention is especially important in patients prone to frequent headache. However, the behavior of some patients with Medication-overuse headache is similar to that seen with other drug addictions.

Thank you