

Unusual Headache Disorders

Primary headache phenotypes continue to expand. This review will focus on some of the more uncommon primary headache disorders defined in chapter 4 of the International Headache Society (IHS) Classification for Primary Headaches: Primary stabbing headache, Primary cough headache, Primary exertional headache, Hypnic headache, and New persistent daily headache.¹ The aetiology remains speculative. There is increasing evidence for neurally driven mechanisms as integral to the development of the more common primary headaches. The same is likely to be true for the uncommon disorders. Most primary headache phenotypes may be precipitated by a secondary pathology. The appropriate context in which to investigate will be discussed.

Primary Stabbing Headache

Phenotype: The head pain occurs as single jabs or a series of jabs, predominantly in the distribution of the first division of the trigeminal nerve. The pain lasts for a few seconds. There are usually no additional features. Attacks occur with irregular frequency, from once to many times a day. In more than 50% of cases the disorder is associated with other primary headache disorders (migraine, cluster headache, paroxysmal hemicrania, hemicrania continua).

Management: The syndrome shows a complete or partial response to Indomethacin.²

Primary Cough Headache

Phenotype: The head pain is sudden onset, bilateral and precipitated (rather than aggravated) by coughing, straining or other Valsalva manoeuvres. The duration ranges from one second to 30 minutes. Typically the pain arises moments after coughing, reaches a peak almost instantaneously, then subsides over several seconds or minutes. Most patients are pain free between attacks but some may have a dull headache afterwards which persists for hours. Typically migrainous features, such as nausea, photophobia and phonophobia, are uncommon. Rarely the pain can be unilateral.³

Investigation: Symptomatic cough headache is reported for a number of pathologies which include Chiari I malformation, cerebrospinal fluid (CSF) volume depletion, basilar impression, medulloblastoma, middle and posterior fossa meningioma and pituitary adenoma.³ Although some reports of symptomatic cough headache show a clear association, the association in other cases is tentative. Clinical characteristics and treatment responses cannot differentiate between primary and secondary cough headache.⁴ Thus, until the literature becomes more clear, all patients with cough headache should have magnetic resonance imaging. A diagnosis of primary cough headache remains that of exclusion.

Management: The most consistently reported effective treatment is with indomethacin.⁵ Doses range between 25 and 250mg daily. Treatment should be withdrawn periodically as symptoms may naturally remit. Open-label trials, often case reports, of effective treatments are published for acetazolamide,⁶ methysergide,⁷ parenteral dihydroergotamine,⁸ naproxen,⁸ propranolol⁷ and lumbar puncture.⁹ The latter involves removal of 40ml of CSF. Constituents are normal. Responses can be dramatic and long-term.⁵

Primary Exertional Headache

Phenotype: Exertional headache is distinguished from exercise-induced migraine. The headache is brought on by and

occurs during or after physical exertion. The pain can be prevented by avoidance of physical exertion. The pain can be of thunderclap or gradual onset, bilateral, less commonly unilateral, throbbing in quality, and with or without migrainous features. Symptoms persist from 5 minutes to 48 hours.^{3,9,10}

Investigation: All patients with exertion-precipitated thunderclap headache must be investigated for symptomatic headache, most commonly subarachnoid haemorrhage (SAH).¹¹ Patients are more likely to have benign exertional headache if the headache is of gradual onset during exertion.

Management: In situations where exertion cannot be predicted, treatment is regular prophylaxis. If exertion can be predicted, pre-emptive therapy 30-60 minutes before exercise can be used. The most consistent responses have been reported for Indomethacin 25-250mg.¹² Aim to start at the lowest dose and titrate up as required and tolerated. Reports also exist for propranolol, naproxen and ergotamine derivatives.^{3,8}

Walk Headache

Walk headache is currently not defined by the IHS classification. The headache occurs with exertion, may be unilateral or bilateral and with or without additional features (eg nausea). There may be concomitant chest or left arm discomfort. The headache settles with rest and can be eased by anti-anginal treatment such as nitroglycerine spray. The diagnosis can be confirmed by an exercise electrocardiogram or thallium scan. The headache responds to treatment of the cardiac ischaemia. All reported cases except one have been over 50 years old (one 40 years). Older patients with a supportive history, particularly with cardiovascular risk factors, should be investigated accordingly.¹³

Primary Headache Associated With Sexual Activity

Phenotype: There are two clinical syndromes of headache associated with sexual excitement (coitus and masturbation): (1) Thunderclap headache just before or at orgasm. (2) Bilateral, often occipital, pressure-headache which gradually increases in severity towards orgasm. Sexual activity may be a precipitant for 'spontaneous' low CSF volume headache.¹⁴ This is presumed to be due to a ruptured developmental malformation eg perineural cyst or meningeal diverticulum. Sexual headaches are not experienced with every sexual encounter.^{3,10,14,15}

Investigation: As for exertional headache all patients with sexual excitement-precipitated thunderclap headache must be investigated for symptomatic headache.¹⁶ Patients are more likely to have benign sexual headache if the headache is of gradual onset and develops during excitement.

Management: The most effective therapies are propranolol (40-200mg) or Indomethacin (25-225mg) taken as regular prophylaxis or pre-emptively before sexual intercourse.³ There is a single successful report of Naratriptan 2.5mg taken 2 hours pre-emptively.¹⁷

A significant proportion of patients presenting with cough, exertional and sexual headaches have symptomatic headache (up to ~60%), thus the recommendation is to image all patients. Since the pain occurs in paroxysms and naturally remits, figures for symptomatic forms no doubt remain confounded by referral bias. Both primary and secondary forms of cough, sexual and exertional headache are more common in migraineurs.¹⁸ There does appear to be an association between exertional and sexual headache; this is



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Table: Summary of Unusual Headache Disorders

	Cough headache	Exertional headache	Sexual headache	Hypnic headache	Stabbing headache
Site of pain	Usually bilateral	Bilateral	Bilateral	Bilateral	Varying site – mainly V1 trigeminal
Character of pain	Sharp, stabbing	Throbbing	Thunderclap – before or at orgasm or bilateral pressure headache gradually increasing in severity towards orgasm.	Dull	Single jabs or series of jabs
Additional features	Nausea, photophobia and phonophobia uncommon	With or without nausea, photophobia and phonophobia	None	Usually featureless. Autonomic features, photophobia and phonophobia uncommon.	None
Duration	Seconds to 30 minutes	5 minutes to 48 hours	A minute to 3 hours	60 to 180 minutes	Seconds
Frequency	Sudden and precipitated by cough, straining or other types of Valsalva	Pain precipitated by and occurs during or after physical exertion	Associated with orgasm	Exclusively during sleep. Up to 6 times / night	Irregular frequency
Preventative Treatment	Indomethacin	Indomethacin, propranolol, ergotamine derivatives	Indomethacin, propranolol	Lithium, caffeine, indomethacin	Indomethacin

reported in 10–40% of patients.^{3,10,14} However a distinction between sexual excitement and exertion associated with sexual activity is not consistently made. Therefore the association may merely reflect the commonality of headache associated with exertion.¹⁵

Hypnic ('Alarm-clock') Headache

Phenotype: Attacks of head pain occur exclusively during sleep and wake the patient, often at consistent times during the night. The pain is typically moderately severe, generalised, dull and featureless. Attacks usually last an hour (range 15–180 minutes) and can occur up to 6 times per night.¹⁹ The pain can be unilateral, throbbing, with nausea and uncommonly autonomic features, photophobia and phonophobia can be present.

Investigation: Since all primary headache phenotypes can be precipitated by a secondary pathology, there remains an argument for imaging all patients with unusual headache disorders until there is adequate data to support doing otherwise.

Management: Treatment responses are from case reports which are few. Sumatriptan and oxygen do not seem to be effective. Aspirin is the most consistently reported effective abortive treatment. Preventative efficacy is reported for lithium, caffeine, indomethacin and flunarizine.

New Persistent Daily Headache (NPDH)

Phenotype: NPDH is a relatively newly recognised head pain disorder. The headache is daily and unremitting from onset (within 3 days at most), and lasts more than 3 months. The IHS classify the phenotype as that of tension-type headache but with new onset and chronic evolution; this is not supported by the literature. From a cohort of 56 patients, 80% could pin-point the exact date of headache onset.²⁰ In 30% onset was associated with a 'flu-like illness, in 12% extracranial surgery, and 12% a stressful life event.

Thirty-eight percent had a prior history of episodic headache, most commonly migraine. None had a prior history of chronic headache. The daily pain was continuous in 80% and bilateral in 64%. The prevalence of nausea was 68%, photophobia 66%, phonophobia 61%, throbbing pain 54% and visual aura 9%.

Investigations: The diagnosis of secondary NPDH is guided by additional neurological and systemic clinical features eg subarachnoid haemorrhage, meningitis, arterial dissection, head injury. Most of these patients present initially to the acute medical and surgical teams. The patient group who present to the neurologist is that represented by the cohort of Rozen,²⁰ often many months or years from the onset. Neurological examination pertinent to the headache and imaging (CT or MRI) are normal. One study found 85% of patients had evidence of active Epstein-Barr infection compared to 13% of controls. A treatable cause of NPDH is spontaneous low CSF volume headache. By the time the patient is seen postural features are often absent. Most patients do not have MRI with gadolinium to address pachymeningeal enhancement. This is the investigation of choice in patients presenting with NPDH.

Management: There are no randomised controlled trials of treatment in NPDH. Consistently clinical experience suggests these patients are refractory to abortive and preventative treatment. Despite withdrawal of all abortive medication, patients remain resistant to preventative treatment. Management usually involves minimisation of acute-relief medication, establishment of preventative treatment (often those used in the management of daily migraine), with or without local anaesthetic blockades such as greater occipital nerve injection. The initial series of 45 patients quoted 86% headaches had disappeared by 2 years, while many patients from the cohort of Rozen continued to suffer for more than 5 years.

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