

# Other primary headaches

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## Abstract

The 'Other Primary Headaches' include eight recognised benign headache disorders. Primary stabbing headache is a generally benign disorder which often co-exists with other primary headache disorders such as migraine and cluster headache. Primary cough headache is precipitated by valsalva; secondary cough has been reported particularly in association with posterior fossa pathology. Primary exertional headache can occur with sudden or gradual onset during, or immediately after, exercise. Similarly headache associated with sexual activity can occur with gradual evolution or sudden onset. Secondary headache is more likely with both exertional and sexual headache of sudden onset. Sudden onset headache, with maximum intensity reached within a minute, is termed thunderclap headache. A benign form of thunderclap headache exists. However, isolated primary and secondary thunderclap headache cannot be clinically differentiated. Therefore all headache of thunderclap onset should be investigated. The primary forms of the aforementioned paroxysmal headaches appear to be Indomethacin sensitive disorders. Hypnic headache is a rare disorder which is termed 'alarm clock headache', exclusively waking patients from sleep. The disorder can be Indomethacin responsive, but can also respond to Lithium and caffeine. New daily persistent headache is a rare and often intractable headache which starts one day and persists daily thereafter for at least 3 months. The clinical syndrome more often has migrainous features or is otherwise has a chronic tension-type headache phenotype. Management is that of the clinical syndrome. Hemicrania continua straddles the disorders of migraine and the trigeminal autonomic cephalalgias and is not dealt with in this review.

## Key Words

Cough headache, exertional headache, hypnic headache, primary headache disorders, stabbing headache

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*Ann Indian Acad Neurol 2012;15:66-71*

## Introduction

The International Headache Society Classification for Headache Disorders provides a guide to diagnosing Head and Facial Pain disorders. The classification is divided into three parts: "Primary," "Secondary Headaches," and "Cranial neuralgias, central and primary facial pain and other headaches."

The primary headaches are supported by a healthy evidence base, while the rest of the classification is less well scientifically backed. There are four sections within the primary headaches section: "Migraine," "Tension-Type Headache," "Cluster Headache and other Trigeminal Autonomic Cephalalgias," and "Other Primary Headaches."

The "other primary headaches" defines eight headache

## Table 1: Classification of the international headache society: Other primary headaches

Primary stabbing headache
Primary cough headache
Primary exertional headache
Primary headache associated with sexual activity
Preorgasmic headache
Orgasmic headache
Hypnic headache
Primary thunderclap headache
Hemicrania continua
NDPH

phenotypes [Table 1]. Hemicrania continua has clinical and functional imaging features which are shared by both migraine and the trigeminal autonomic cephalalgias. This review will focus on the seven other primary headaches in this group.

## Primary Stabbing Headache

### Epidemiology

The prevalence of primary stabbing headache has varied from 2 to 35%. The discrepancy in part is likely to be related to the populations studied. In more than 50% of cases, the disorder is

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10.4103/0972-2327.100012

associated with other primary headaches. Stabbing headache is reported to occur in 40% of migraineurs and 30% of cluster headache sufferers.<sup>[1]</sup>

### Clinical syndrome

Primary stabbing headache has been previously termed “Ice-pick headache” or “Jabs and jolts.” Patients can also describe the pain as needle-, nail-, or pinprick-like. The head pain occurs as a single jab or a series of jabs. In the majority of patients, jabs last 1–2 seconds at a time, at most for 10 seconds.<sup>[2]</sup> Although the pain is reported to be predominantly in the distribution of the first division of the trigeminal nerve, in patient cohorts the pain is frequently experienced in the distribution of C2.<sup>[3,4]</sup> The pain can be unifocal or multifocal in site. Attacks occur with irregular frequency, from once to many times a day. The pain is usually spontaneous and without additional features. However, in patients who also have migraine, described precipitants include physical exertion, postural change, dark-light transition, and head motion. The jabs can be associated with the migraine attacks and/or occur independently; similarly location may be restricted in some patients to that affected by the coexisting primary headache disorder.<sup>[1]</sup> Aura symptoms can occur in association with the jabs of pain alone.<sup>[2]</sup>

The two main differential diagnoses are the syndrome of SUNCT (short-lasting neuralgiform attacks with conjunctival injection and tearing) and trigeminal neuralgia. In SUNCT, the attacks of pain are strictly unilateral and occur primarily in the distribution of the first division of the trigeminal nerve, characteristically orbital, supra-orbital, and temporal. Duration is longer, each jab lasting up to 250 seconds. There are prominent accompanying autonomic features ipsilateral to the pain (typically conjunctival injection, and lacrimation, although other symptoms such as nasal congestion, rhinorrhea, and eye-lid swelling can occur). Although the attacks occur spontaneously, they are often precipitated by movements such as chewing, brushing teeth, or talking. In trigeminal neuralgia, the key feature is that the pain is in the distribution of the second and third division of the trigeminal nerve and characteristically triggered. There are no autonomic features. Each of these syndromes responds to different medical treatments.

### Secondary Stabbing Headache

There are few case reports of isolated stabbing headache being attributed to an alternative pathology. A Meningioma<sup>[5]</sup> and pituitary tumor<sup>[6]</sup> are reported to have precipitate stabbing headache. Whether a relationship exists is presumptive; in these reports evolution of tumor and onset of headache cannot be adequately causally time-locked. Moreover, resolution of headache following surgery is not necessarily an indication of causal relationship; it cannot be established whether removal of the lesion, the actual surgery, or the anesthetic resulted in the resolution.

### Management

Primary stabbing headache shows a complete or partial response to indomethacin, usually between 25 and 150 mg daily.<sup>[3,7]</sup> Alternative preventatives with reports of efficacy include the COX-II inhibitors<sup>[8]</sup> and melatonin.<sup>[9]</sup>

## Primary Cough Headache

### Epidemiology

The lifetime prevalence of cough headache is reported to be 1%.<sup>[10]</sup> Age of onset tends to be more than 40 years with a male predominance.

### Clinical syndrome

The head pain is sudden onset, bilateral and *precipitated* (rather than aggravated, as occurs in migraine) by coughing, straining, or Valsalva maneuver. The duration ranges from 1 second to 30 minutes, although about 10% have been reported to have longer attacks.<sup>[11]</sup> Typically the pain arises moments after coughing, reaches a peak almost instantaneously, and then subsides over several seconds or minutes. Most patients are pain free between attacks but some may have a dull headache afterward which persists for hours. Typically migrainous features, such as nausea, photophobia, and phonophobia, are uncommon. Rarely the pain can be unilateral.<sup>[12]</sup>

### Secondary Cough Headache

Symptomatic cough headache is reported for a number of pathologies which include Chiari I malformation, cerebrospinal fluid (CSF) volume depletion, basilar impression, platybasia, medulloblastoma, middle and posterior fossa meningioma, and chromophobe adenoma.<sup>[12,13]</sup> Although some reports of symptomatic cough headache show a clear association, the association in other cases is tentative. Symonds' original observations from 1956 are most informative. He presented 17 patients with cough headache who were monitored longitudinally.<sup>[14]</sup> All had a normal neurological examination and skull x-rays; four had further investigations. Nine underwent spontaneous remission between 5 weeks and 10 years, six improved over 18 months to 5 years, and two died of cardiac disease within 2–3 years of follow-up. He reported six patients with cough headache who were found to have associated pathology: A posterior fossa meningioma, acoustic neuroma, brain stem cyst, and in three cases Paget's disease of the skull with moderate or severe basilar impression. Isolated cough headache was present in four and two had abnormal neurological signs. One patient underwent spontaneous remission periods and another remission after removal of acoustic neuroma. One patient had resolution after “deep X-ray treatment” and one following lumbar puncture and air encephalogram. Chen and colleagues reported on their 83 patients with cough headache, all of whom were imaged.<sup>[11]</sup> Normal imaging was found in 89%; neurological examination was normal in all patients. Nine patients had abnormal imaging: three patients had posterior fossa meningioma greater than 4 cm and with associated hydrocephalus, two had metastatic deposits, one in the posterior fossa and another diffuse, two had Chiari I malformations (4 and 5 mm below foramen magnum), one had acute sinusitis with mucous retention in the sphenoid sinus, and one a right frontal subdural hematoma and aneurysm at the supraclinoid portion of the left internal carotid artery. Notably, only three patients had an abnormal examination, the two with metastatic disease and one with meningioma (4 mm) and hydrocephalus; all had gait ataxia with or without limb ataxia. At mean follow-up of 51 months, 84% of the primary cough headache group

had gone into remission. Response to indomethacin was seen in 73% of the primary group and 38% of the secondary group. Thus, it was not possible to clinically differentiate primary from secondary cough headache. At a variance to the aforementioned cohorts, Pascual and colleagues reported a primary to secondary ratio of 13:17.<sup>[12]</sup> All 17 secondary cough headache patients had a Chiari I malformations and five also had syringomyelia. All but three had abnormal neurological signs. The three presenting with isolated cough headache developed abnormal signs within 1–5 years. Indomethacin response was not tested in the secondary headache group. Eight underwent suboccipital craniectomy and C1-3 laminectomy; seven had improved headache postoperatively.

Thus, the clinical message from these studies is that primary and secondary cough headache cannot be distinguished clinically nor by medical treatment response.

### Management

The most consistently reported effective treatment is with indomethacin with doses range between 25 and 250 mg daily.<sup>[15,16]</sup> Treatment should be withdrawn periodically as symptoms may naturally remit. Open-label trials and case reports of effective treatments are published for acetazolamide,<sup>[17]</sup> methysergide,<sup>[18,19]</sup> parenteral dihydroergotamine,<sup>[20,21]</sup> naproxen,<sup>[21,22]</sup> propranolol,<sup>[19]</sup> and lumbar puncture.<sup>[14,23]</sup> The latter involved removal of 40 ml of CSF. Constituents are normal. Responses can be dramatic and long term.<sup>[16]</sup>

### Primary Exertional Headache

#### Epidemiology

Prevalence figures for exertional headache range between 12 and 30%.<sup>[24]</sup> In contrast to primary cough headache, the disorder seems to affect a younger population (generally below age 50 years). The disorder is more prevalent in individuals with a personal or family history of migraine.<sup>[12]</sup>

#### Clinical syndrome

Exertional headache is headache brought on by, and occurring during or after, prolonged physical exertion. The pain can be prevented by avoidance of physical exertion. The pain can be of thunderclap or gradual onset, bilateral, and less commonly unilateral, throbbing in quality, and with or without migrainous features. Symptoms persist from 5 minutes to 48 hours.<sup>[12,13,23,25]</sup>

#### Secondary exertional headache

All patients with exertion-precipitated thunderclap headache must be investigated for a symptomatic headache (as for secondary thunderclap headache). Patients are more likely to have benign exertional headache if the headache is of gradual onset during exertion. In Pascual's cohort of 12 secondary exertional headache, all presented with sudden onset severe headache. Ten were diagnosed with subarachnoid hemorrhage on CSF examination and/or computed tomography (CT); only two were found to have an aneurysm on angiography. One patient of the remaining two had metastatic carcinoma of the breast and on examination had papilledema and a sixth nerve palsy. The other patient was diagnosed with pansinusitis; symptoms settled following a course of antibiotics.

### Management

In situations where exertion cannot be predicted, treatment is prophylactic. If exertion can be predicted, preemptive therapy 30–60 minutes before exercise can be used. The most consistent responses have been reported for indomethacin 25–250 mg.<sup>[15,26]</sup> The aim is to start at the lowest dose and titrate up as required and tolerated. Reports of efficacy also exist for propranolol, naproxen, and ergotamine derivatives.<sup>[12,21]</sup> The natural history is of spontaneous quiescence. In Rooke's cohort of 103 patients 30 had complete relief within 5 years and 73 were free from headache or improved after 10 years.<sup>[23]</sup>

### Cardiac cephalgia

Cardiac cephalgia is a rare exertional headache secondary to cardiac ischemia. It is currently not defined by the International Headache Society classification. The headache occurs with exertion, may be unilateral or bilateral and with or without additional features (e.g., 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 ischemia. All reported cases except one have been more than 40 years old. Older patients with a supportive history, particularly with cardiovascular risk factors, should be investigated accordingly.<sup>[27,28]</sup>

### Primary Headache Associated with Sexual Activity

#### Epidemiology

The estimated prevalence of headache related to sexual activity is 1%. There is a male preponderance.<sup>[29]</sup> The mean age of onset is in the fourth decade.

There does appear to be an association between exertional and sexual headache; this is reported in 10–40% of patients.<sup>[12,25,30]</sup> 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.<sup>[31]</sup> There is also comorbidity reported between migraine and sexual headache.<sup>[32]</sup>

#### Clinical syndrome

There are two clinical syndromes of headache associated with sexual excitement (coitus and masturbation):

*Preorgasmic headache:* A bilateral (less commonly unilateral), occipital, or generalized pressure headache which gradually increases in severity toward orgasm. Preorgasmic headache is seen in about 20% of cases of sexual headache.

*Orgasmic headache:* Sudden onset severe pain (thunderclap onset) immediately before or at orgasm. A bilateral (less commonly unilateral), occipital or generalized, dull or throbbing pain. Orgasmic headache is seen in about 80% of cases.

Sexual headaches are not experienced with every sexual encounter.<sup>[12,25,30,31]</sup> The attacks of headache tend to be short-lived. In Frese's cohort of 51 patients, the range was 1 minute to 24 hours. The median duration of severe pain

was 30 minutes and of residual milder pain 4 hours.<sup>[29]</sup> Accompanying symptoms are uncommon. Most commonly sexual headache occurred with sexual activity with the usual partner (94%) and also during masturbation (35%), with a new partner (14%), and during an extramarital affair in one patient. Twenty patients (40%) could terminate the headache by stopping sexual activity. Twenty-six patients (51%) could ease the headache by taking a more passive role during sexual activity. In five patients, sexual headache occurred only with specific sexual practices.

### Secondary Sexual Headache

As for exertional headache, all patients with sexual excitement-precipitated thunderclap headache must be investigated for symptomatic headache.<sup>[33]</sup> Patients are more likely to have benign sexual headache if the headache is of gradual onset and develops during excitement.<sup>[12]</sup>

Sexual activity may be a precipitant for spontaneous low CSF volume headache;<sup>[30]</sup> this is presumed to be due to a ruptured developmental malformation, e.g., perineural cyst or diverticulum.

#### Management

The most effective therapies are propranolol (40–200 mg) or indomethacin (25–225 mg) taken as prophylaxis or preemptively before sexual intercourse.<sup>[2,12]</sup> Triptans can be used both acutely and preemptively before sexual activity.<sup>[34,35]</sup>

In Frese's cohort of 45 patients with a single or recurrent episodes of sexual headache at presentation, 37 were in complete remission at 3-year follow-up.<sup>[36]</sup> Thus, the disorder appears to be self-limiting in the majority of individuals.

#### Primary thunderclap headache

Thunderclap headache has been characteristically associated with the dramatic presentation of subarachnoid hemorrhage. However, longitudinal observation of patients with subsequent normal investigations revealed a benign form of the syndrome,<sup>[37-39]</sup> hence termed primary thunderclap headache.

#### Clinical syndrome

Thunderclap headache is a sudden onset severe headache which reaches maximum severity within a minute.

### Secondary Thunderclap Headache

Clinically it is not possible to differentiate between primary and secondary thunderclap headache.<sup>[38]</sup> Secondary thunderclap headache has been reported with a number of other cerebral pathologies including cerebral venous sinus thrombosis,<sup>[38]</sup> arterial dissection,<sup>[40]</sup> pituitary apoplexy,<sup>[41]</sup> and spontaneous intracranial hypotension.<sup>[42]</sup> Therefore, *all* patients with thunderclap headache should be investigated.

In patients presenting with isolated thunderclap headache, CT will pick up subarachnoid blood within 12 hours in 98% of cases, dropping to 86% by 24 hours.<sup>[43]</sup> Thereafter, CSF xanthochromia spectroscopically is 100% reliable for subarachnoid blood between 12 hours and 2 weeks, dropping down to 70% by the third

week.<sup>[44,45]</sup> The difficulty of angiographic examination thereafter is that there is no reliable way of differentiating an incidental from a ruptured aneurysm. Other pathologies precipitating thunderclap headache will be guided by history and clinical signs.

#### Management

Primary thunderclap headache is generally self-limiting, but can recur over months or year. Attacks can occur spontaneously or be provoked by valsalva, exertion, or bathing.<sup>[46]</sup> About a third hence develop migraine headache.<sup>[37,47,48]</sup>

### Hypnic (Alarm-clock) Headache

#### Epidemiology

Hypnic headache was first reported in 1988.<sup>[49]</sup> Tertiary clinic prevalence is reported at 0.1%.<sup>[50,51]</sup>

#### Clinical syndrome

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, generalized, dull, and featureless. Attacks usually last an hour (range 15–180 minutes) and can occur up to six times per night; the mean number of attacks is one.<sup>[51]</sup> The pain can be unilateral, throbbing, with nausea and uncommonly autonomic features, photophobia, and phonophobia.

#### Secondary hypnic headache

As eventually becomes apparent with all newly recognized primary headache syndromes, various independent cerebral pathologies can precipitate the same syndrome. There are few reports of secondary hypnic headache. The most convincing is a case report of a patient presenting with hypnic headache and a few months later additional symptoms and signs of acromegaly. The patient had transphenoidal resection of the growth hormone secreting microadenoma, with resolution of headache; it would have been interesting to see whether resolution had occurred with medical management of the microadenoma.<sup>[52]</sup> A tentative reported precipitant is mild to moderate hypertension in two cases. The patients initially had an absolute response to indomethacin. Thereafter, treatment of the hypertension with nifedipine also resulted in complete resolution of the pain. Whether nifedipine or treatment of the hypertension resulted in this improvement was not addressed.<sup>[53]</sup>

#### Management

Treatment responses are from case reports which are few. Simple analgesics are effective abortive treatments. Sumatriptan and oxygen do not seem to be effective. Preventative efficacy is reported for lithium, caffeine, indomethacin, and flunarizine.<sup>[51,53]</sup>

### New Daily Persistent Headache

#### Epidemiology

New daily persistent headache (NDPH) was first reported as a "new" entity in 1986.<sup>[54]</sup> The disorder is distinguished from new onset daily persistent headache which is obviously precipitated, such as posttraumatic headache following traumatic brain injury or other pathology, such as meningitis or cerebral hemorrhage.<sup>[55]</sup> The 1-year prevalence of NDPH is reported at 0.03%.<sup>[56]</sup>

**Table 2: Differential diagnostic features**

	<b>Stabbing headache</b>	<b>Cough headache</b>	<b>Exertional headache</b>	<b>Sexual headache</b>	<b>Hypnic headache</b>
Site of pain	Varying site within V1 trigeminal and C2	Bilateral diffuse	Bilateral diffuse	Bilateral diffuse	Bilateral diffuse
Character of pain	Single jabs or series of jabs	Sharp, stabbing	Throbbing	Thunderclap-before/at orgasm or bilateral pressure headache gradually increasing in severity towards orgasm.	Dull
Additional features	None	Nausea, photophobia and phonophobia uncommon	With or without nausea, photophobia and phonophobia	None	Usually featureless. Autonomic features, photophobia and phonophobia uncommon.
Duration	Seconds (maximum 10 s)	Seconds to 30 minutes	5 minutes to 48 hours	A minute to 3 hours	60 to 180 minutes
Frequency	Irregular frequency	Sudden and precipitated by coughing, straining or other valsalva	Pain precipitated by and occurs during or after physical exertion	Associated with orgasm	Exclusively during sleep. Up to 6 times/night (usually once)
Preventative treatment	Indomethacin	Indomethacin	Indomethacin, propranolol, ergotamine derivatives	Indomethacin, propranolol	Lithium, caffeine, indomethacin, flunarizine

### Clinical syndrome

The headache is daily and unremitting from onset (within 3 days at most), and lasts more than 3 months. Eighty percent of patients can pin-point the exact date of headache onset.<sup>[57]</sup> The international classification for NDPH unfortunately does not reflect the evidence from reported cohorts;<sup>[57-59]</sup> the phenotypic classification provided is that of tension-type headache but with new onset and chronic evolution. From a cohort of 56 patients, 30% associated onset with a “flu-like illness,” 12% with extracranial surgery, and 12% a stressful life event.<sup>[57]</sup> 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%.

In 71 patients reported by Robbins, 44% fulfilled the current international classification criteria, while 56% had too many migrainous features.<sup>[59]</sup> Twenty-five percent had a preexisting history of a primary headache disorder (episodic tension-type headache in 18% and migraine in 7%), and almost half gave a family history of a primary headache disorder.

### Secondary New Daily Persistent Headache

In patients presenting with NDPH, neurological examination pertinent to the headache and imaging (CT or magnetic resonance imaging) is normal. One study found that 85% of patients had evidence of active Epstein-Barr infection (otherwise asymptomatic) compared with 13% of controls.<sup>[60]</sup> An uncommon but treatable cause of NDPH is spontaneous low CSF volume headache. In such cases, the diagnosis is aided by typical postural features and supportive imaging abnormalities. However, postural features are not consistent and imaging can be normal.<sup>[61]</sup>

### Management

There are no randomized controlled trials of treatment in NDPH. Patients can be prone to medication-overuse headache. Despite withdrawal of abortive medication, patients often remain resistant to preventative treatment. Management involves minimization of acute-relief medication and

establishment of preventative treatment guided by headache phenotype, and use of local anesthetic blockade such as greater occipital nerve injection. The initial series of 45 patients quoted 86% headaches had disappeared by 2 years.<sup>[54]</sup> In Robbins cohort of 71 patients, about 76% had a persistent subform (median duration in migrainous group 31 months and in the group defined by the International Headache Society classification, 18 months), 15.5% had a remitting form (median duration 21 months) and 8.5% a relapsing remitting form (median time to first remission 5.5 months).

### Summary

The “other primary headaches” encompasses a group of uncommon but distinct headache disorders. A number of these are characterized by short-lived paroxysmal attacks of pain which are often indomethacin responsive [Table 2]. While the new presentation of stabbing headache or preorgasmic headache is usually benign, that of cough headache or headache with thunderclap onset is more frequently reported to be precipitated by other cranial pathologies. As larger patient cohorts are reported, the relative frequency of primary to secondary presentations will become clearer.

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**How to cite this article:** Bahra A. Other primary headaches. *Ann Indian Acad Neurol* 2012;15:66-71.

**Received:** 31-05-11, **Revised:** 07-06-11, **Accepted:** 25-07-11

**Source of Support:** Nil, **Conflict of Interest:** Nil