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Other primary headache disorders

Apart from migraine, tension-type headache and cluster headache, what are the other types of primary headache disorders?

Recommendation

In the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta),^{1,2)} primary headache disorders other than migraine, tension-type headache and cluster headache are grouped together as “*Other primary headaches disorders*”. They are classified into primary cough headache, primary exercise headache, primary headache associated with sexual activity, primary thunderclap headache, cold-stimulus headache, external-pressure headache, primary stabbing headache, nummular headache, hypnic headache, and new daily persistent headache.

Grade A

Background and Objective

In the first edition of the International Classification of Headache Disorders published in 1988 by the Headache Classification Committee (Chairman, Jes Olsen) of the International Headache Society,³⁾ these headaches were grouped under “*Miscellaneous headaches unassociated with structural lesion*”.

The headaches were classified into the following types: idiopathic stabbing headache, external compression headache, cold stimulus headache, benign cough headache, benign exertional headache, and headache associated with sexual activity. Cold stimulus headache was further divided into two subtypes: external application of a cold stimulus, and ingestion of a cold stimulus. Headache associated with sexual activity was classified into dull type, explosive type, and postural type.

When the first edition of the International Classification of Headache Disorders was undergoing complete revision, the Japanese Headache Society (International Classification Promotion Committee) in collaboration with the Ministry of Health, Labour and Welfare Study Group (Study Group for Chronic Headache Clinical Guideline) translated the revised guidelines⁴⁾ and published the Japanese Edition of the International Classification of Headache Disorders 2nd Edition.⁵⁾ In the second edition, headache disorders other than migraine, tension-type headache and cluster headache have been classified under the new term “*Other primary headaches*”.

Comments and Evidence

Headache disorders are classified according to the International Classification of Headache Disorders 3rd Edition (beta version) (ICHD-3beta), published in 2013.²⁾ In the ICHD-3beta, primary headache disorders other than migraine, tension-type headache and cluster headache are grouped under “*Other primary headaches disorders*”, and classified into ten types as follows: primary cough headache, primary exercise headache, primary headache associated with sexual activity, primary thunderclap headache, cold-stimulus headache, external-pressure headache, primary stabbing headache, nummular headache, hypnic headache, and new daily persistent headache.

Primary stabbing headache is transient and localized stab-like headache that occurs spontaneously in the absence of organic disease in local structures or in the cranial nerves.

Primary cough headache is headache triggered by coughing or straining, in the absence of intracranial diseases.

Primary exercise headache is headache triggered by exercise (regardless of type). Subforms such as “weight-lifters’ headache” are recognized.

Primary headache associated with sexual activity is headache precipitated by sexual activity, usually starting as a bilateral dull ache as sexual excitement increases and suddenly intensifies at orgasm, in the absence of intracranial diseases.

Hypnic headache manifests as dull headache attacks that always awaken the patient from asleep.

Primary thunderclap headache is high-intensity headache of abrupt onset mimicking that of ruptured cerebral aneurysm. Hemicrania continua is persistent, strictly unilateral headache responsive to indomethacin.

Hemicrania continua, originally grouped under “*Other primary headaches disorders*” in International Classification of Headache Disorders 2nd Edition (ICHD-II), is moved to “*Trigeminal autonomic cephalalgias: TACs*” in ICHD-3beta.

New daily persistent headache is headache that is daily and unremitting from very early after onset. The pain is typically

bilateral, pressing or tightening in quality, and of mild to moderate intensity. Photophobia, phonophobia or mild nausea may occur.

Some of these headaches are symptomatic. Careful evaluations using neuroradiological imaging such as MRI, and other tests are necessary.

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• Search terms and secondary sources

- Search database: Ovid (2011/12/21)
Headache and Headache disorders 560
{Headache and Headache disorders} and Classification 87
- Search database: Ichushi Web for articles published in Japan (2011/12/21)
Headache 5576
Headache and classification 166
Headache and classification and clinical guideline 9

How are primary stabbing headache, primary cough headache, and primary exercise headache diagnosed and treated?

Recommendation

1. Diagnosis

Primary stabbing headache, primary cough headache, and primary exercise headache are diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3 beta). **Grade A**

2. Treatment

Although no randomized controlled trials of treatment for these headaches have been reported, indomethacin is considered effective in most cases for these headaches. As adverse effect of indomethacin, gastrointestinal symptoms may be an issue when used long-term. Other drugs have been tried, but are limited to case reports and small case series. **Grade C**

Background and Objective

Primary stabbing headache, primary cough headache, and primary exercise headache are included in primary headaches other than migraine, tension-type headache, and cluster headache. The objective of this section is to review the reports on the diagnosis and treatment of these disorders.

Comments and Evidence

1. Diagnosis

(1) Primary stabbing headache¹⁾²⁾

- A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B-D
- B. Each stab lasts for up to a few seconds
- C. Stabs recur with irregular frequency, from one to many per day
- D. No cranial autonomic symptoms
- E. Not better accounted for by another ICHD-3 diagnosis

(2) Primary cough headache¹⁾²⁾

- A. At least two headache episodes fulfilling criteria B-D
- B. Brought on by and occurring only in association with coughing, straining and/or other Valsalva maneuver
- C. Sudden onset
- D. Lasting between 1 second and 2 hours
- E. Not better accounted for by another ICHD-3 diagnosis

(3) Primary exercise headache¹⁾²⁾

- A. At least two headache episodes fulfilling criteria B and C
- B. Brought on by and occurring only during or after strenuous physical exercise
- C. Lasting <48 hours
- D. Not better accounted for by another ICHD-3 diagnosis

2. Treatment

(1) Primary stabbing headache

Several uncontrolled studies have reported response to indomethacin,³⁾⁴⁾ but there are also reports of partial or even no response. Mathew⁵⁾ treated 5 patients with 50 mg indomethacin 3 times a day and reported drastic reduction in mean headache frequency in a week compared to aspirin and placebo. On the other hand, Pareja et al.⁶⁾ studied the clinical features of 38 patients, and reported that among 17 patients treated with 75 mg/day indomethacin for 15 days, 6 patients (35%) achieved complete remission and 5 patients had partial remission, while 6 patients (35) were refractory to treatment. Several case reports are available for drugs other than indomethacin. They include a report of a 71 year-old woman responding to

nifedipine sustained release tablet 90 mg/day;⁷⁾ a report of 3 cases recommending a treatment regimen of melatonin starting at a dose of 3 mg/day and increasing gradually;⁸⁾ a report of 4 young onset cases responding to gabapentin 400 mg/day;⁹⁾ and 3 cases responding to celecoxib, a cyclooxygenase-2 inhibitor.¹⁰⁾

(2) Primary cough headache

This headache usually responds to indomethacin. Mathew⁵⁾ conducted a double-blind study in 2 patients, and reported the effectiveness of indomethacin 150 mg/day. Raskin¹¹⁾ treated 16 patients with indomethacin 50 to 200 mg (mean 78 mg) per day, and observed complete remission in 10 patients, moderate improvement in 4 patients and no response in 2 patients. In the report of Pascual et al.,¹²⁾ response was observed in 6 of 13 patients treated with indomethacin 75 mg/day. Indomethacin is considered to be the most effective drug for symptomatic relief.¹³⁾ As for the other treatments, Calandre et al.¹⁴⁾ reported cases responding to propranolol 120 mg and also cases responding to methysergide. In one case reported by Mateo and Pascual,¹⁵⁾ naproxen 550 mg given every 12 hours achieved partial relief. Wang et al.¹⁶⁾ studied the usefulness of acetazolamide in 5 indomethacin responsive patients. Acetazolamide was started at a dose of 125 mg three times a day and titrated until maximum effect was obtained, up to a maximum of 2,000 mg/day. The outcome was complete response in 2 patients, favorable response in 2 patients and no response in 1 patient. Raskin¹¹⁾ treated 14 patients by performing lumbar puncture to remove 40 mL of cerebrospinal fluid, and reported response in 6 patients; with response observed immediately after the procedure in 3 patients, and 2 days or longer later in the other 3 patients.

3. Primary exercise headache

Indomethacin has long been used as the drug of choice for prophylactic treatment of exertional headache. Diamond¹⁷⁾ treated 15 patients with indomethacin starting from 25 mg/day and titrating to a maximum dose of 150 mg. Response was obtained in 13 patients (87%). After headache was controlled, indomethacin was discontinued and headache recurred within 7 days in 12 of 13 patients. As for the other drugs, Pascual et al.¹²⁾ tried ergotamine tartrate in 16 patients who took the drug before exertion started, and 4 patients reported subjective response showing potential prophylactic effect. They also treated 5 patients with propranolol prophylactically; 3 patients had irregular attacks, 1 patient showed clear response, while 1 patient did not respond but improved with indomethacin. A study in Japan also reported the usefulness of propranolol as a prophylactic drug.¹⁸⁾ Furthermore, flunarizine was administered to 2 patients, and response was obtained in 1 patient.¹²⁾

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- **Search terms and secondary sources**

1. Diagnosis

- Search database: PubMed (2012/1/30)
{Headache and Headache disorders} and Classification 170

2. Treatment

- Search database: PubMed (2012/1/30)
{Stabbing headache} 60
{Primary cough headache} or {Benign cough headache} or {Valsalva manoeuvre headache} 119
{Exertional headache} 68

How is primary headache associated with sexual activity diagnosed and treated?

Recommendation

1. Diagnosis

Primary headache associated with sexual activity is diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta). This headache is precipitated by sexual activity, and is diagnosed after excluding intracranial disorders by brain imaging study and cerebrospinal fluid examination.

Grade A

2. Treatment

To treat primary headache associated with sexual activity, it is necessary for the patient and the partner to understand the disorder. Pharmacotherapy using indomethacin, triptans and propranolol is effective in some cases.

Grade C

Background and Objective

Statistical data from headache clinics suggest that primary headache associated with sexual activity is rare. However, potential patients probable exist in relatively large numbers. Appropriate approach to this disorder is necessary.

Comments and Evidence

1. Diagnosis

The diagnostic criteria for primary headache associated with sexual activity are as follows¹⁾:

- A. At least two episodes of pain in the head and/or neck fulfilling criteria B-D
- B. Brought on by and occurring only during sexual activity
- C. Either or both of the following:
 1. increasing in intensity with increasing sexual excitement
 2. abrupt explosive intensity just before or with orgasm
- D. Lasting from 1 minute to 24 hours with severe intensity and/or up to 72 hours with mild intensity
- E. Not better accounted for by another ICHD-3 diagnosis

When occurring at the first time, it is mandatory to exclude subarachnoid hemorrhage and internal carotid artery or vertebral artery dissection. Differential diagnosis also includes intracerebral hemorrhage, subdural hematoma, unruptured aneurysm, cerebral venous sinus thrombosis, Arnold-Chiari I malformation, posterior fossa neoplasm, increased intracranial pressure, decreased intracranial pressure, and cervical spinal cord disease.²⁾ Reversible cerebral vasoconstriction syndrome (RCVS) has also been reported, emphasizing the necessity of diagnostic imaging study.³⁾ Headache clinic surveys reported that patients with primary headache associated with sexual activity occupied 0.2 to 1.3% of all headache patients.⁴⁾ A more recent case-control study estimated a prevalence of 0.9% in the general population.⁵⁾ It is possible that the headache is underdiagnosed because patients are embarrassed to describe the circumstances in detail, and that the true prevalence may be considerably higher. The prevalence is 3 to 4 times higher in men than in women. The age at onset has two peaks, one in the early twenties and the other around 40 years of age.^{4,6)} Type 1 and type 2 in the first edition of the International Classification of Headache Disorders are equivalent to preorgasmic headache (dull type, approximately 20%) and orgasmic headache (explosive type, approximately 80%), respectively, in the International Classification of Headache Disorders 2nd Edition (ICHD-II). Type 3 in the first edition, which is positional headache, is caused by cerebrospinal fluid leak and is coded as "headache attributed to spontaneous low CSF pressure" in ICHD-II.⁷⁾ The pathogenetic mechanism has not been fully elucidated, but onset of preorgasmic headache is associated with tension-type headache and muscular contraction mainly in the neck,⁸⁾ while orgasmic headache is associated with increased intracranial pressure accompanying an abrupt increase in blood pressure or heart rate.²⁾ Patients' blood pressure increases markedly during sexual activity, and the existence of metabolism-related impaired cerebrovascular autoregulation is speculated.⁹⁾ Headache is bilateral and commonly occur in

the occipital region. The pain lasts from several minutes to several hours or one day, and headache is severe usually during the first 5 to 15 minutes. The headache duration is longer in orgasmic headache than in preorgasmic headache. Headache occurs during coitus with the usual partner and also during masturbation. Comorbidity with migraine, tension-type headache, and primary exertional headache has been reported.²⁾⁶⁾

2. Treatment

To treat primary headache associated with sexual activity, the patient's and partner's understanding of the disorder is necessary.¹⁰⁾ In preorgasmic headache, headache is usually relieved by discontinuing sexual activity. Therefore patients are advised to remain sexually inactive as much as possible until they are completely free of headache.⁴⁾ The usefulness of taking indomethacin (50 to 100 mg) 1 to 2 hours before coitus,⁴⁾ and the use of triptans (such as naratriptan) have been reported. Treatment with ergotamine and benzodiazepine compounds has also been used.¹¹⁾¹²⁾ For patients with prolonged headache duration, prophylactic therapy using propranolol, metoprolol, and diltiazem has been attempted.²⁾⁸⁾ A report has shown the usefulness of greater occipital nerve blockade by injection of a steroid and local anesthetic combination.¹³⁾ The prognosis of headache associated with sexual activity is relatively good. In the majority, the headache appears in a bout and remits, but 25% of patients have a chronic course.¹⁰⁾

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- Search database: PubMed (2011/12/5)
Headache
& {Sexual activity} 134
& {Migraine} 27
Sexual Headache 649
& {Migraine} 474
& {Treatment} 21
- Search database: PubMed (2011/12/5)
Sexual headache 340
& {Migraine} & {Treatment} 9
- Search database: Ichushi Web for articles published in Japan (2011/11/11)
Sexual headache 5

How is hypnic headache diagnosed and treated?

Recommendation

1. Diagnosis

Hypnic headache is diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta).

Grade A

2. Treatment

Caffeine is used not only as an acute treatment but also as a prophylactic drug. Lithium is another frequently used prophylactic drug.

Grade C

Background and Objective

Although hypnic headache is a rare headache disorder, over 170 cases have been reported. Reported for the first time by Raskin in 1988, this headache is also called “alarm clock headache” because it awakens the patient from sleep. In the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3 beta), hypnic headache is classified in the group “*Other primary headache disorders*”.¹⁾ The pathophysiology has not been fully elucidated.

Comments and Evidence

1. Diagnosis

The diagnostic criteria of hypnic headache are as follows¹⁾:

- A. Recurrent headache attacks fulfilling criteria B-E
- B. Developing only during sleep, and causing wakening
- C. Occurring on ≥ 10 days per month for >3 months
- D. Lasting ≥ 15 minutes and for up to 4 hours after waking
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-3 diagnosis

Hypnic headache is a rare headache, and is estimated to occupy 0.07 to 0.35% of headache patients.²⁾³⁾ The male to female ratio is 1: 1.2 to 1: 1.7, with a female preponderance. The mean age of onset is around 60 years. although hypnic headache occurs typically in older persons,⁴⁾⁻⁸⁾ pediatric cases have also been reported.³⁾ Only a small number of cases have been reported in Japan.⁹⁾ Headache is typically mild to moderate in intensity, dull and bilateral, but one-third is pulsating with severe intensity. The duration ranges from 15 to 180 minutes (mean 80 minutes), although headache may last 6 hours. The frequency of attack is 1 to 2 times per night, and the mean frequency of headache episodes is 23 per month. When patients are woken up by the headache at night, they read books, watch television, drink or eat, or walk inside the room. These characteristics are in contrast to the excited and restless states in cluster headache.⁵⁾⁻⁸⁾ Polysomnographic studies have reported that headache arises during REM sleep,¹⁰⁾⁻¹³⁾ but recent research contradicts the association between hypnic headache and sleep stage.⁷⁾¹⁴⁾ A MRI study with voxel-based morphometry (VBM) has reported a decrease in posterior hypothalamus gray matter.¹⁵⁾ The characteristic clinical picture of chronobiological abnormality in addition to pain suggests impairments of pain sensation and sleep rhythm at the trigeminal nerve in the hypothalamo-pituitary system. It is important to conduct imaging studies to differentiate from secondary headaches such as posterior fossa tumor, pontine infarction and pituitary tumor. Other headache disorders that should be differentiated include cluster headache, trigeminal-autonomic cephalalgias, and hemicrania continua.

2. Treatment

Caffeine is used not only as an acute treatment but also as a prophylactic drug.³⁾¹⁶⁾¹⁷⁾ Drinking a cup of coffee when awoken by pain or before going to sleep is effective. As prophylactic drugs, lithium is usually effective, while topiramate, indomethacin, melatonin, and amitriptyline have also been used. Some cases remit spontaneously, while others remit upon treatment but relapse later.

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• Search terms and secondary sources

- Search database: PubMed (2011/12/31)
Hypnic headache 118
- Search database: Ichushi Web for articles published in Japan (2011/11/30)
Hypnic headache 4

How is primary thunderclap headache diagnosed and treated?

Recommendation

1. Diagnosis

Primary thunderclap headache is diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta).

Grade A

2. Treatment

Differentiating primary thunderclap headache from diseases that cause thunderclap headache secondarily is most important. There is no established treatment.

Grade C

Background and Objective

In the diagnosis of thunderclap headache, the first and foremost step is to exclude a wide variety of secondary headaches. Accurate diagnosis and treatment by headache specialists are important.

Comments and Evidence

1. Diagnostic criteria

The diagnostic criteria of primary thunderclap headache are as follows.¹⁾

- A. Severe head pain fulfilling criteria B and C
- B. Abrupt onset, reaching maximum intensity in <1 minute
- C. Lasting for ≥ 5 minutes
- D. Not better accounted for by another ICHD-3 diagnosis

The most important step in diagnosis is to differentiate from disorders that may cause secondary thunderclap headache. It is mandatory to exclude subarachnoid hemorrhage due to ruptured cerebral aneurysm,²⁾ unruptured saccular cerebral aneurysm,^{3,4)} carotid or vertebral artery dissection,⁵⁾ intracerebral hemorrhage,⁶⁾ cerebral infarction,⁷⁾ cerebral venous sinus thrombosis,⁸⁾ and pituitary apoplexy.⁹⁾ Other disorders that require differentiation include central nervous system angitis, colloid cyst of the third ventricle, cerebrospinal fluid hypotension, acute sinusitis (especially barotrauma), retroclival hematoma, primary cough headache, primary exertional headache, primary headache associated with sexual activity,¹⁰⁾ and bath-related headache.¹¹⁾ In recent years, reversible cerebral vasoconstriction syndrome (RCVS)¹²⁾¹³⁾ as a cause of secondary thunderclap headache has drawn attention. For the diagnosis of headaches associated with subarachnoid hemorrhage, dissecting aneurysm and pituitary apoplexy, see the CQs for “Headache: General Considerations”.

Primary thunderclap headache is known to occur commonly in female adults, and is diagnosed only after all organic underlying diseases have been excluded. Secondary thunderclap headaches are treated according to the treatments for the underlying diseases, while treatment for primary thunderclap headache has not been established. The pathophysiology of primary thunderclap headache remains largely unclear, although failure of the afferent sympathetic nerve system that modulates intracranial vascular tone causing acute vasoconstriction or alteration in vascular tone has been suggested to cause the headache.¹⁰⁾

2. Treatment

Nimodipine has been reported to be effective,¹⁴⁾ but there is no established treatment.

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- Search database: PubMed (2011/12/5)
thunderclap headache 215
- Search database: Ichushi for articles published in Japan
thunderclap headache 948

How is hemicrania continua diagnosed and treated?

Recommendation

1. Diagnosis

Hemicrania continua is diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3 beta).

Grade A

2. Treatment

Complete remission is obtained by treatment with indomethacin.

Grade A

Background and Objective

Hemicrania continua is a rare disorder.^{1,2)} Since the disorder was first described by Sjaastad in 1984, over 150 cases have been reported.³⁾⁻¹¹⁾ Although hemicrania continua is characterized by association with autonomic symptoms and marked response to indomethacin, the pathophysiology, clinical picture, treatment and prognosis remain undefined.

Comments and Evidence

1. Diagnostic criteria

The diagnostic criteria of hemicrania continua are as follows.^{1,2)}

- A. Unilateral headache fulfilling criteria B-D
- B. Present for >3 months, with exacerbations of moderate or greater intensity
- C. Either or both of the following:
 1. at least one of the following symptoms or signs, ipsilateral to the headache:
 - a) conjunctival injection and/or lacrimation
 - b) nasal congestion and/or rhinorrhoea
 - c) eyelid edema
 - d) forehead and facial sweating
 - e) forehead and facial flushing
 - f) sensation of fullness in the ear
 - g) miosis and/or ptosis
 2. a sense of restlessness or agitation, or aggravation of the pain by movement
- D. Responds absolutely to therapeutic doses of indomethacin^{Note 1}
- E. Not better accounted for by another ICHD-3 diagnosis

Note:

1. In an adult, oral indomethacin should be used initially in a dose of at least 150 mg daily and increased if necessary up to 225 mg daily. The dose by injection is 100-200 mg. Smaller maintenance doses are often employed.

Hemicrania continua is a rare disorder and evidence is limited to case series.³⁾⁻¹¹⁾ In summary, the male to female ratio is approximately 1: 2, with a female preponderance. The mean onset age is in the thirties. Headache is unilateral and does not shift to the other side, and is lasting pain with mild to moderate intensity. The sites of headache are mainly in the frontal, temporal, orbital and occipital regions. Exacerbation of headache occurs sometimes and intense pain greatly impairs daily living. During exacerbation, ipsilateral autonomic symptoms including lacrimation and conjunctival injection often occur. Headache may be accompanied by the associated symptoms seen in migraine. Hemicrania continua is characterized by chronically persistent pain. When recurrence occurs after a remission, the pain usually takes a chronic course thereafter. Complete remission is obtained by indomethacin. Only a few cases of hemicrania continua have been reported in Japan.¹²⁾⁻¹³⁾ However, cases of pain shifting to the other side, cases not responsive to indomethacin, cases with no autonomic symptoms, and cases manifesting autonomic symptoms not listed in the diagnostic criteria of ICHD-3beta have been reported. Although PET examination demonstrated activation in contralateral posterior hypothalamus and ipsilateral dorsal rostral pons, the

exact pathophysiology remains unknown.¹⁴⁾ Both hemicrania continua and paroxysmal hemicrania exhibit indomethacin responsiveness and autonomic symptoms, thereby raising a possibility that they share a common pathophysiological basis.⁸⁾¹⁰⁾

Differential diagnosis includes unilateral localized chronic migraine, new daily persistent headache, cervicogenic headache, trigeminal-autonomic cephalalgias, chronic post-traumatic headache, headache attributed to arterial dissection, and headache attributed to brainstem infarction.

In ICHD-3 beta, hemicrania continua is classified as one of the trigeminal-autonomic cephalalgias.

2. Treatment

Headache responds absolutely to therapeutic doses of indomethacin. In Japan, the maximum dose is 75 mg/day for oral formulation, and 100 mg/day for rectal administration.²⁾ In overseas countries, however, indomethacin is used at a starting dose of 25 to 75 mg/day, increasing gradually if there is no response, and the responsive dose has been reported to range from 50 to 300 mg/day.³⁾⁻⁷⁾¹⁰⁾ Oral indomethacin has to be taken for long term, and adverse effects including vertigo and gastrointestinal symptoms are an issue. To reduce gastrointestinal adverse effects, the use of indomethacin farnesil, a prodrug of indomethacin, is sometimes effective from experience. Most of the other analgesics are not effective. Ibuprofen, naproxen, and aspirin have been tried, but results are inconsistent.⁵⁾ Supraorbital nerve or greater occipital nerve block has been reported to be effective in patients with tenderness.¹⁵⁾ In a crossover study of occipital nerve stimulation therapy in 6 patients, good result was reported¹⁶⁾ but the method has not been established for general use.

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- Search database: PubMed (2012/6/4)
Hemicrania continua 254
& Indomethacin 138
Indomethacin farnesyl 13
- Search database: Ichushi Web for articles published in Japan (2012/6/4)
Hemicrania continua 9
Hemicrania continua (Japanese) 5
Hemicrania continua9

How is new daily persistent headache diagnosed and treated?

Recommendation

1. Diagnosis

New daily persistent headache is diagnosed according to the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3 beta).

Grade A

2. Treatment

There are no clearly established treatment criteria, and also no treatments with established efficacy. There are two types; a type that resolves spontaneously, and a refractory type that is resistant to aggressive treatments.

Grade C

Background and Objective

The International Classification of Headache Disorders 2nd Edition (ICHD-II) recognizes *New daily persistent headache* (NDPH) as a new separate entity. However, details of the headache properties, treatment effects and prognosis are not known. The mode of onset is important in diagnosis, and excluding secondary headaches is important.

Comments and Evidence

1. Diagnostic criteria

The diagnostic criteria of new daily persistent headache described in the ICHD-3beta are as follows.¹⁾²⁾

- A. Persistent headache fulfilling criteria B and C
- B. Distinct and clearly remembered onset, with pain becoming continuous and unremitting within 24 hours
- C. Present for >3 months
- D. Not better accounted for by another ICHD-3 diagnosis

New daily persistent headache is a relatively rare disorder, and evidence is limited to case series.³⁾⁻⁵⁾ This headache has also been reported in Japanese, but the number of cases is relatively small.⁴⁾⁶⁾ In summary, the male to female ratio is slightly higher in female. The mean age of onset is in the thirties. The day of headache onset is usually clearly remembered by the patient. While the headache often has features resembling those of tension-type headache, it may also manifest characteristics of migraine such as nausea, photophobia and phonophobia. The headache may remit, or recur and remit repeatedly, or persist, but many patients follow a chronic course. Robbin et al.⁵⁾ divided new daily persistent headache according to headache properties into two groups: a group with migraine-like headache that has a female preponderance and frequently a history of anxiety or depressive disorder, and a group with features of tension-type headache in which patients recall accurately the day of headache onset. Their report emphasizes that new daily persistent headache may manifest migraine-like headache. In a Norwegian population-based study of a sample aged 30 to 40 years, the 1-year prevalence was 0.03%.⁷⁾ Among children and adolescents who are less likely to overuse medications than adults, onset of new daily persistent headache is typically secondary to infection and trauma.⁸⁾⁹⁾

Differential diagnosis includes chronic migraine, chronic tension-type headache, hemicrania continua, headache attributed to low cerebrospinal fluid pressure, headache attributed to increased cerebrospinal fluid pressure, headache attributed to head and/or neck trauma, and headache attributed to infection. Although many symptoms resemble those of chronic tension-type headache, the unique features are that headache is not evolved from episodic tension-type headache and that headache is daily and unremitting from the day of onset.

2. Treatment

No prospective placebo-controlled trial has been reported, and clear treatment criteria have not been established.³⁾⁻⁵⁾ New daily persistent headache has two types: a self-limiting type that resolves spontaneously, and a refractory subtype that is resistant to aggressive treatment. In line with tension-type headache and migraine, abortive drugs and prophylactic drugs such as gabapentin and topiramate have been tried, with no consistent results.

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- Search database: PubMed (2012/6/4)
New daily persistent headache 144
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New daily persistent headache 7

How is chronic daily headache diagnosed?

Recommendation

Chronic daily headache is a headache classification proposed by Silberstein, Lipton and colleagues,^{1,2)} and is defined as headache that lasts 4 or more hours per day and occurs on 15 or more days per month. This disorder is classified into four types: transformed migraine, chronic tension-type headache, new daily persistent headache, and hemicrania continua. There is no clear evidence. With the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta) now being established, each headache type and medication-overuse headache should be diagnosed according to ICHD-3 beta, and chronic daily headache should be used as an umbrella term that includes various types of chronic headache.

Grade C

Background and Objective

Since the International Headache Society first published the diagnostic criteria for headache disorders in 1988, the debate on how to diagnose and classify headaches that occur daily has continued. Chronic daily headache is a headache classification proposed by Silberstein, Lipton and colleagues^{1,2)} in 1994. It is defined as headache that lasts 4 or more hours per day and occurs on 15 or more days per month, and is classified into four types. The International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3 beta) published in 2013³⁻⁶⁾ does not recognize chronic daily headache as a separate entity. However, the name is still used due to the convenience of allowing evaluation of all the headaches that occur on a daily basis.

Comments and Evidence

In 1994, Silberstein, Lipton and colleagues^{1,2)} defined chronic daily headache as headache that lasts 4 or more hours per day and occurs on 15 or more days per month. They classified this group into four types and set out diagnostic criteria. The four types are:

1. Transformed migraine (TM)
2. Chronic tension-type headache (CTTH)
3. New daily persistent headache (NDPH)
4. Hemicrania continua (HC)

The above classification is currently used worldwide. The criterion of at least 4 hours a day excludes cluster headache. Regarding the duration of headache, various articles have described durations ranging from 1 month to 1 year. In accordance with the diagnostic criteria of the International Classification of Headache Disorders, 3rd Edition (beta version) (ICHD-3beta) for chronic migraine, new daily persistent headache and hemicrania continua,³⁻⁶⁾ a duration of over 3 months is generally accepted.⁷⁻⁹⁾ The prevalence of chronic daily headache in the general population has been reported to be approximately 3 to 4%, while a prevalence of approximately 1.5% has been reported among subjects aged 12 to 14 years in population-based studies.^{7,9)-13)} In a study comparing 638 adults aged 18 years or older and 170 adolescents aged 13 to 17 years, transformed migraine associated with medication overuse was significantly more frequent in adults while transformed migraine without medication overuse and chronic tension-type headache were significantly more common in adolescents.¹²⁾ In a prospective cohort 8-year follow-up study of 122 adolescents aged 12 to 14 years, one-fourth of the patients continued to have disability in daily living due to chronic daily headache.¹⁴⁾

In the ICHD-3beta, chronic daily headache is not recognized as a separate entity, and transformed migraine is handled as chronic migraine and classified as various types in the group of primary headaches, differentiated from medication-overuse headache. Compared with the Silberstein-Lipton diagnostic criteria, the criteria in the ICHD-3beta are stricter. Transformed migraine with increased headache frequency due to medication overuse is considered almost equivalent to the ICHD-3beta codes of “migraine” + “medication-overuse headache”, while transformed migraine with no medication overuse or no increased headache frequency even though there is medication overuse is considered similar to the ICHD-3beta code of “chronic migraine”. Likewise, chronic tension-type headache with increased headache frequency due to medication overuse is considered to be “tension-type headache” + “medication-overuse headache”, while chronic tension-type headache with no

medication overuse or no increased headache frequency even though there is medication overuse to be “*chronic tension-type headache*”. Furthermore, the ICHD-3beta places importance on the presence of autonomic symptoms in hemicrania continua, and the elements of tension-type headache in new daily persistent headache. With the ICHD-3beta being well established, it has been recommended to discontinue using the term chronic daily headache and to diagnose according to ICHD-3beta. However, the term continues to be used currently, because when individual headaches cannot be classified accurately, it offers the convenience to evaluate these headaches under the umbrella term of chronic daily headache.

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- Search database: PubMed (2012/6/4)
Daily headache 4502
chronic daily headache 31338
& definition 224
& diagnostic criteria 2155
& prevalence 4655
& frequency 6238
- Search database: Ichushi Web for article published in Japan (2012/6/4)
Chronic daily headache 14
Chronic daily headache (Japanese) 5329