

CEFALEE PRIMARIE RARE

GIOVANNI D'ANDREA

**CENTRO CEFALEE “VILLA MARGHERITA”
VICENZA**

CEFALEE PRIMARIE RARE

- **Emicrania parossistica cronica**
- **SUNCT**
- **Cefalea trafittiva idiopatica**
- **Cefalea da tosse**
- **Cefalea da sforzo**
- **Cefalea associata ad attività sessuale**
- **Cefalea ipnica**
- **Primary thunderclap headache**
- **Hemicrania continua**
- **New daily persistent headache**

3. Cluster headache and other trigeminal autonomic cephalalgias

3.1 Cluster headache

3.1.1. Episodic cluster headache

3.1.2. Chronic cluster headache

3.2 Paroxysmal hemicrania

3.2.1. Episodic paroxysmal hemicrania

3.2.1. Chronic paroxysmal hemicrania (CPH)

3.3 Short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)

3.4 Probable trigeminal autonomic cephalalgias

3.4.1. Probable cluster headache

3.4.2. Probable paroxysmal hemicrania

3.5.3. Probable SUNCT

3.4 Probable trigeminal autonomic cephalalgia

Description:

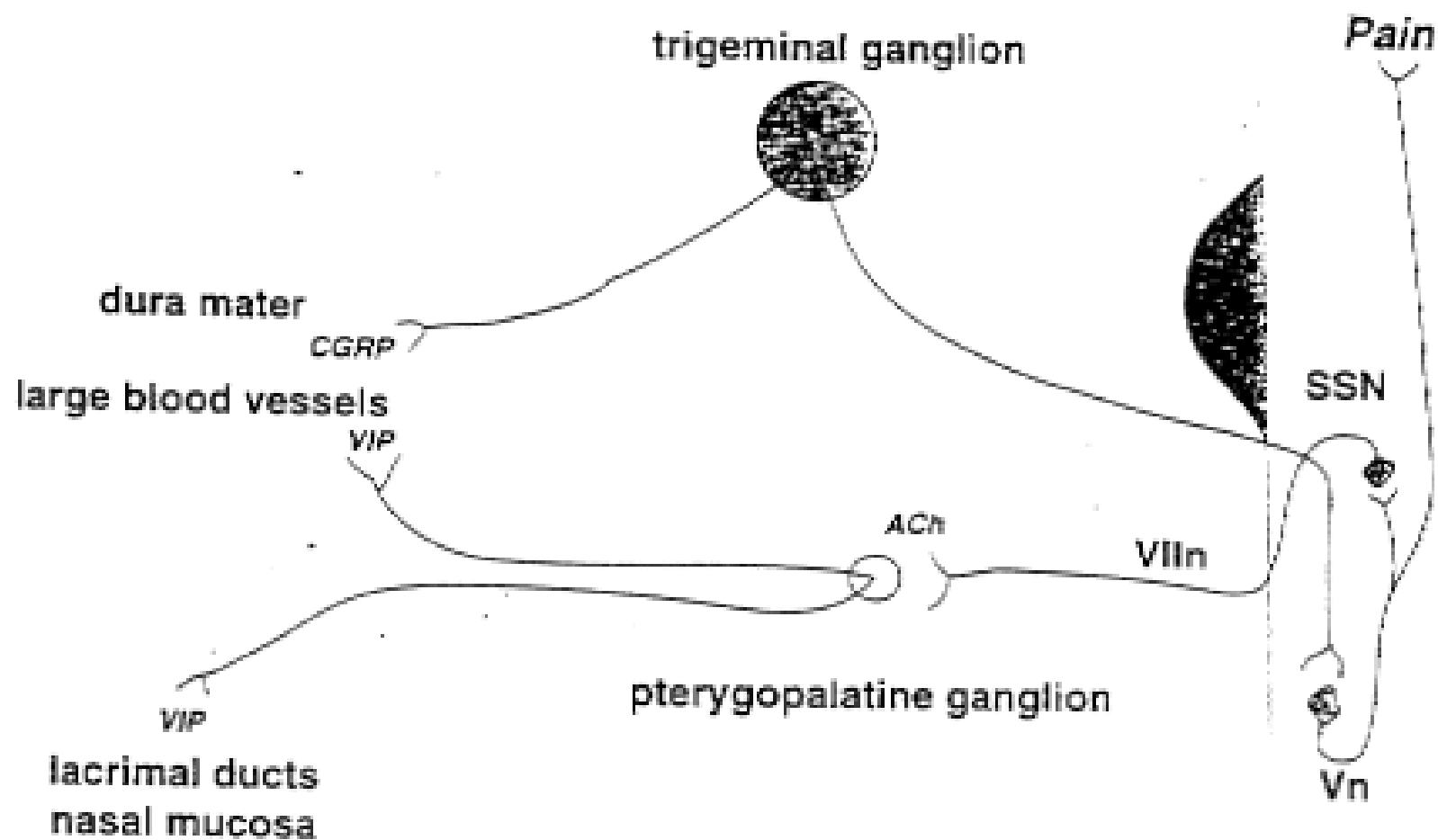
Headache attacks that are believed to be a subtype of trigeminal autonomic cephalalgia but which do not quite meet the diagnostic criteria for any of the subtypes described above

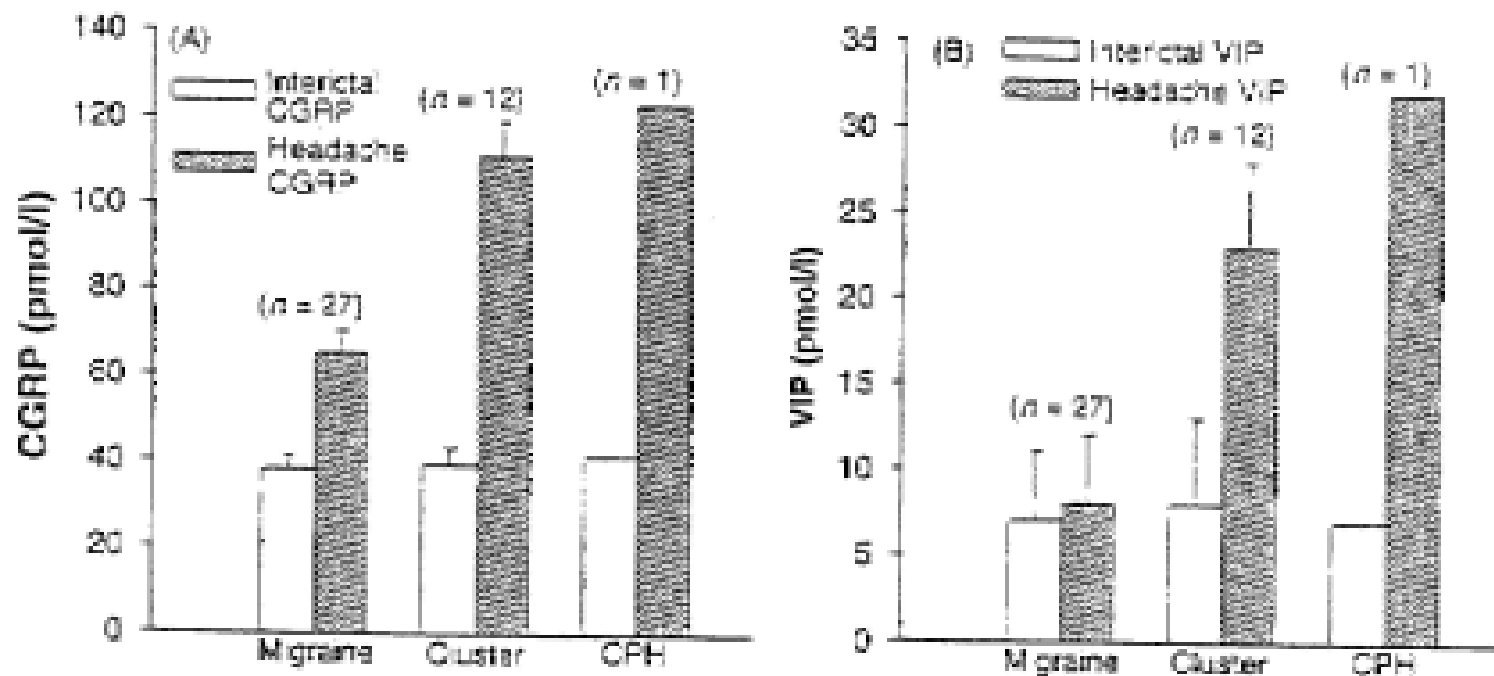
Diagnostic criteria

- A. Attacks fulfilling all but one of specific criteria for one of the subtypes of trigeminal autonomic cephalalgia**
- B. Not attributed to other disorder**

CEFALEE AUTONOMICHE TRIGEMINALI

- **Sindromi dolorose unilaterali**
- **Segni autonomici**
- **Attivazione del sistema trigeminale**





The figure demonstrates the changes in (A) calcitonin gene-related peptide (CGRP) and (B) vasoactive intestinal polypeptide (VIP) in cranial venous blood during attacks of migraine (with and without aura), cluster headache and chronic paroxysmal hemicrania (CPH). The level of elevation of CGRP and VIP in the CPH patient is comparable with that seen in cluster headache patients (Goadsby and Edvinsson, 1994b; Fanciullacci *et al.*, 1995).

Goadsby P, & Lipton R, 1997

3.2 PAROXYSMAL HEMICRANIA

Description:

Attacks with similar characteristics of pain and associated symptoms and signs of cluster headache, but they are short lasting, more frequent, occur more commonly in **females** and respond **absolutely to indomethacin**.

PAROXYSMAL HEMICRANIA

Diagnostic criteria:

- A. At least 20 attacks fulfilling criteria B-D
- B. Attacks of **severe unilateral** orbital, supraorbital or temporal pain lasting **2-45 minutes**
- C. Headache is accompanied by at least one of the following:
 - 1. **Ipsilateral conjunctival injection and/or lacrimation**
 - 2. **Ipsilateral nasal congestion and/or rhinorrhoea**
 - 3. **Ipsilateral eyelid oedema**
 - 4. **Ipsilateral forehead and facial sweating**
 - 5. **Ipsilateral miosis and/or ptosis**
- D. Attacks have a **frequency above 5 per day** for more than half of time, although periods with lower frequency may occur
- E. Attacks are prevented **completely by indomethacine**
- F. Not attributed to another disorder



PAROXYSMAL HEMICRANIA

Notes:

1. In order to rule out incomplete response, indomethacin should be used in a dose of ≥ 150 mg daily orally or rectally, or ≥ 100 mg by injection , but for maintenance smaller doses often sufficient.
2. History and physical and neurological examination do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examination do suggest such disorder but is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relationship to the disorder.

3.2.1 EPISODIC PAROXYSMAL HEMICRANIA

Description:

Attacks of paroxysmal hemicrania occurring in periods lasting 7 days or one year separated by pain-free periods lasting 1 month or longer.

PAROXYSMAL HEMICRANIA

Diagnosi differenziale:

- Cefalea a grappolo (per le crisi brevi)
- SUNCT (per la crisi lunghe)
- Risposta alla indometacina

3.3 SUNCT

Description:

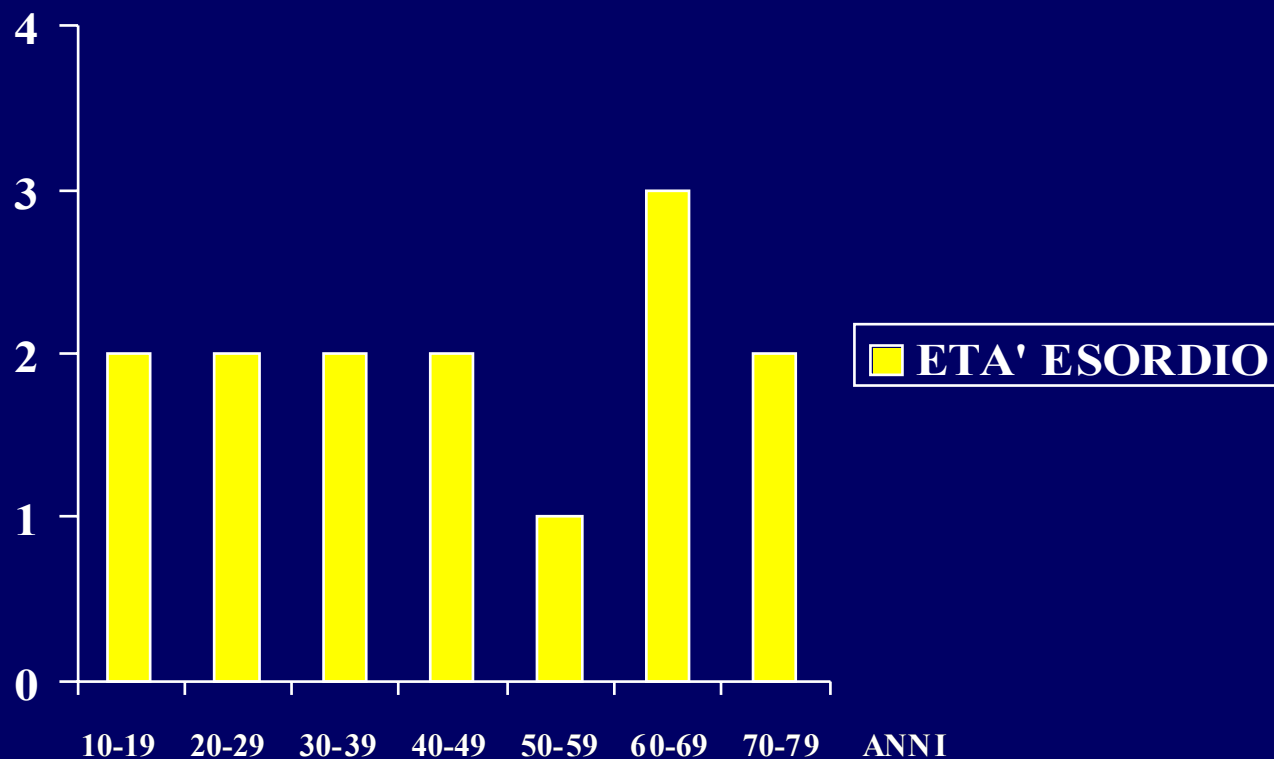
This syndrome is characterised by short-lasting attacks of unilateral pain that are much briefer than those seen in any other TAC and very often accompanied by prominent lacrimation and redness of the ipsilateral eye.

Criteri Diagnostici della SUNCT (IHS '04)

- Almeno 20 attacchi che soddisfano i criteri B-E
- Attacchi di dolore severo unilaterale orbitario o temporale trafittivo o pulsante della durata di 5-240 secondi
- Il dolore è accompagnato da iniezione congiuntivale e lacrimazione presenti dal lato del dolore
- Frequenza degli attacchi: da 3 a 200 al giorno
- Esclusione di cause organiche mediante diagnostica strumentale



PAZIENTI SUNCT



SISC, 2001

LAMOTRIGINA

DOSI	n° PAZIENTI	EFFICACIA	SODDISFAZIONE
100 mg/die	2	***	§§§
125 mg/die	1	**	§§§
150 mg/die	5	***	§§§
200 mg/die	1	**	§§§

** riduzione 80% degli attacchi

§§§ completa soddisfazione

*** scomparsa degli attacchi

SISC, 2001

SUNCT

PROBLEMI DIAGNOSTICI:

- **Emicrania cronica parossistica (per le crisi più brevi: 2-3 min.)**
- **Nevralgia del trigemino**

4. Other primary headaches

4.1 Primary stabbing headache

4.2 Primary cough headache

4.3 Primary exertional headache

4.4 Primary headache associated with sexual activity

4.4.1 Preorgasmic headache

4.4.2 Orgasmic headache

4.5 Ipnictic headache

4.6 Primary thunderclap headache

4.7 Hemicrania continua

4.8 New daily persistent headache

4.1 PRIMARY STABBING HEADACHE

Diagnostic criteria:

- A. Head pain occurring as a single stab or a series of stab and fulfilling criteria B-D
- B. Exclusively or predominantly felt in the distribution of the first division of trigeminal nerve (orbit, temple and parietal area)
- C. Stabs last for up to a few seconds and recur with irregular frequency ranging from one to many per day
- D. Not accompanying symptoms
- E. Not attributed to another disorder

PRIMARY STABBING HEADACHE

Comments:

- a) In one study 80% of stabs lasted 3 seconds or less. In rare cases, stabs occur repetitively over days, and there has been one description of **status** lasting one week.
- b) Stabs may **move from one area to another** in either the same or opposite hemicranium. When they are **strictly localised to one area, structural changes at this site** and the distribution of affected cranial nerve must be excluded.
- c) Stabbing pain are more commonly experienced by **people subject to migraine (about 40%)** or cluster headache (about 30%), in which cases are felt in the site habitually affected by these headaches.
- d) A positive response to **indomethacin** has been reported in same uncontrolled studies, whilst others have partial or no responses.

4.2 PRIMARY COUGH HEADACHE

Description:

Headache precipitated by coughing or straining in the absence of any other intracranial disorder.

4.2 PRIMARY COUGH HEADACHE

Diagnostic criteria:

A. Headache fulfilling criteria B and C

B. Sudden onset, lasting from **one to 30 minutes**

C: Brought on by and occurring only in association with coughing, straining and/or Valsalva manoeuvre

D. Not attributed to another disorder

IHS, 2003

PRIMARY COUGH HEADACHE

Note:

1. Cough headache is symptomatic in about 40% of cases and the large majority of these present Arnold-Chiari malformation type I°. Other reported causes of symptomatic cough headache include carotid and vertebrobasilar diseases and cerebral aneurysms. Diagnostic neuroimaging plays an important role in differentiating secondary cough headache from 4.2 *Primary cough headache*.

Comment:

Primary cough headache is usually bilateral and predominantly affects patients **older than 40 years of age**. Whilst **indomethacin is usually effective in the treatment of primary cough headache**, a positive response to this medication has also been reported in some symptomatic cases.

4.3 PRIMARY EXERTIONAL HEADACHE

Description:

Headache precipitated by any form of exercise. Subform such as “**weight-lifters**” **headache** are recognised.

4.3 PRIMARY EXERTIONAL HEADACHE

Diagnostic criteria:

- A. Pulsating headache fulfilling criteria B and D**
- B. Lasting from 5 minutes to 48 hours**
- C. Brought on by and occurring only during after physical exertion**
- D. Not attributed to another disorder**

4.3 PRIMARY EXERTIONAL HEADACHE

Note:

1. On the first occurrence of this headache type it is mandatory to exclude subarachnoid haemorrhage and arterial dissection.

Comment:

Primary exertional headache occurs particularly in hot weather or at high altitude. There are reports of prevention in some patients by ingestion of ergotamine tartrate. Indomethacin has found effective in the majority of cases.

Headache described in weight-lifters has been considered a subform of 4.3 *Primary exertional headache*; because of its sudden onset and presumed mechanism it may have more similarities to 4.2 *Primary cough headache*.

4.4 PRIMARY HEADACHE ASSOCIATED WITH SEXUAL ACTIVITY

Description:

Headache precipitated by sexual activity , usually starting as a **dull bilateral ache** as sexual excitement increases and suddenly becoming **intense at orgasm**, in absence of any intracranial disorder.

4.4.1 PREORGASMIC HEADACHE

Diagnostic criteria:

- A. **Dull ache in the head and neck** associated with awareness of **neck and/or jaw muscle contraction** and fulfilling criterion B
- B. Occurs during sexual activity and increases with sexual excitement
- C. Not attributed to another disorder

4.4.2 ORGASMIC HEADACHE

Diagnostic criteria:

A. Sudden severe (**explosive**) headache fulfilling criterion B

B. Occurs at orgasm

C. Not attributed to another disorder

Note:

On first onset of orgasmic headache it is mandatory to exclude conditions such as **subarachnoid haemorrhage and arterial dissection.**

4.5 HYPNIC HEADACHE

Description:

**Attacks of dull headache that always awaken the patient
from the sleep**

4.5 HYPNIC HEADACHE

Diagnostic criteria:

- A. Dull headache fulfilling criteria B-D**
- B. Develops only during sleep, and awakens patient**
- C. At least two of following characteristics:**
 - 1. Occurs >15 times per month**
 - 2. Last \geq 15 minutes after waking**
 - 3. First occurs after age of 50 years**
- D. No autonomic symptoms and no more than one of nausea, photophobia or phonophobia**
- E. Not attributed to another disorder**

4.5 HYPNIC HEADACHE

Note:

1. Intracranial disorders must be excluded. Distinction from one of the trigeminal autonomic cephalalgias is necessary for effective management.

Comment:

The pain of Hypnic headache is usually **mild to moderate**, but severe pain is reported by approximately 20% of patients. Pain is **bilateral** in about two-thirds of cases. The attack usually lasts from **15 to 180 minutes**, but longer duration have been described.

Caffeine and lithium have been effective treatment in several reported cases.

4.6 PRIMARY THUNDRCLAP HEADACHE

Description:

High-intensity headache of abrupt onset mimicking that of ruptured cerebral aneurysm.

4.6 PRIMARY THUNDERCLAP HEADACHE

Diagnostic criteria:

A. Severe head pain fulfilling criteria B and C

B. Both of the following characteristics:

1. sudden onset, reaching maximum intensity in <1 minute
2. lasting from 1 hour to 10 days

C. Does not recur regularly over subsequent weeks or months

D. Not attributed to other disorder

note:

1. Headache may recur within the first week after onset
2. Normal CSF and normal brain imaging are required

4.6 PRIMARY THUNDERCLAP HEADACHE

Comment:

Evidence that thunderclap headache exists as a primary condition is poor: the search of an underlying cause should be expedient and exhaustive. Thunderclap headache is frequently associated with serious vascular intracranial disorder, particularly **subarachnoid haemorrhage**: it is **mandatory to exclude this** and a range of other such conditions including **intracerebral haemorrhage, cerebral venous thrombosis, unruptured vascular malformation** (mostly aneurysm), **arterial dissection** (intra and extracranial), **CNS angiitis, reversible benign CNS angiopathy** and **pituitary apoplexy**. Other organic cause of thunderclap headache are **colloid cyst of third ventricle, CSF hypotension** and **acute sinusitis** (particularly with barotrauma). **4.6 primary thunderclap headache** should be the diagnosis only when all organic causes have been excluded.

4.7 HEMICARIA CONTINUA

Description:

Persistent strictly unilateral headache responsive to indomethacin.

Criteri diagnostici dell'hemicrania continua (IHS '04) 4.7

- cefalea presente da più di 3 mesi
- 1) localizzazione del **dolore strettamente unilaterale**
2) **dolore quotidiano e continuo** senza periodi liberi
3) **intensità moderata** ma con **esacerbazioni** di dolore severo
- Almeno 1 dei seguenti **segni autonomici** si verifica **durante le esacerbazioni** e dallo stesso lato del dolore:
 - **iniezione congiuntivale e/o lacrimazione**
 - **congestione nasale e/o rinite**
 - **ptosi e/o miosi**
- **assoluta risposta all'indometacina**
- esclusione di cause organiche mediante diagnostica strumentale

4.7 HEMICARIA CONTINUA

Note:

1. History and physical and neurological examinations do not suggest any of the disorders listed in groups 5-12, or history and/or physical and/or neurological examinations do suggest such disorders but it is ruled out by appropriate investigations, or such disorders is present but attacks do not occur for the first time in close relationship to the disorder.

Comment:

Hemicrania continua is usually unremitting, but **rare case of remission are reported**. Whether this headache type can be subdivided according to length of history and persistence is yet to be determined.

4.8 NEW DAILY-PERSISTENT HEADACHE (NDPH)

Description:

Headache that is daily and unremitting from very soon after onset (within 3 days at most). The pain is **typically bilateral, pressing or tightening** in quality and **of mild to moderate intensity**. There may be photophobia, phonophobia or mild nausea.

4.8 NEW DAILY-PERSISTENT HEADACHE (NDPH)

Diagnostic criteria:

- A. Headache for >3 months fulfilling criteria B-D
- B. Headache is daily and unremitting from onset or from <3 days from the onset
- C. At least two or the following pain characteristics :
 - 1. **bilateral location**
 - 2. **pressing/tightening (non pulsating) quality**
 - 4. **not aggravated by routine physical activity such walking or climbing stairs**
- D. Both of the following:
 - 1. no more than one of photophobia, phonophobia or mild nausea
 - 2. neither moderate or severe nausea nor vomiting
- E. Not attributed to other disorder

4.8 NEW DAILY-PERSISTENT HEADACHE (NDPH)

Note:

1. Headache may be unremitting from the moment of onset or very rapidly build up to continuous and unremitting pain. Such onset or rapid development must be clearly recalled and unambiguously described by the patient. Otherwise code as 2.3 *Chronic tension-type headache*.
2. History and physical and neurological examination do not suggest any of the disorders listed in groups 5-12 (including 8.2 *Medication-over use headache and its subforms*), or history and/or physical and/or neurological examination do suggest such disorder but is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relationship to the disorder.

4.8 NEW DAILY-PERSISTENT HEADACHE (NDPH)

Comment (1):

This second edition of the classification recognises 4.8 *New daily-persistent headache* as a separate entity from 2.3 *Chronic tension-type headache*. Although it has many similarities to tension-type headache, NDPH is unique in that headache is daily and unremitting from or almost from the moment of onset, typically in individuals without a prior headache history. A clear recall of such onset is necessary for the diagnosis of *New daily-persistent headache*.

The headache of NDPH can have associated features suggestive of either migraine or tension-type headache. **Secondary headaches** such as **low CSF volume headache**, **raised CSF pressure headache**, **post-traumatic headache** and **headache attributed to infection** (particularly viral) should be ruled out by appropriate investigations.

4.8 NEW DAILY-PERSISTENT HEADACHE (NDPH)

Comment (2):

NDPH may take either **two subforms**: a **self-limiting subform** which typically resolves without therapy **within several months** and a **refractory subform** subform which is **resistant to aggressive treatment programmes** . The subcommittee aims to stimulate further clinical characterisation and pathophysiological research of this entity, especially studies comparing 4.8 *new daily-persistent headache* with 2.3 *Chronic tension-type headache*.