Norsk kvalitetsregister for alvorlige primære hodepiner

ICDH3: Diagnostiske kriterier for trigeminal autonomic cephalalgias (TACs)

Initial comments: The TACs share the clinical features of unilateral headache and, usually, prominent cranial parasympathetic autonomic features, which are lateralized and ipsilateralto the headache. Experimental and human functional imaging suggests these syndromes activate a normal human trigeminal-parasympathetic reflex, with the clinical signs of cranial sympathetic dysfunction being secondary. Typical migraine aura can be seen, rarely, in association with TACs.

3.1 Cluster headache

Coded elsewhere: Symptomatic cluster headache, secondary to another disorder, is coded as a secondary headache attributed to that disorder.

Description: Attacks of severe, strictly unilateral pain which is orbital, supraorbital, temporal or in any combination of these sites, lasting **15–180 minutes** and occurring from once every other day to eight times a day. The pain is associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis and/or eyelid oedema, and/or with restlessness or agitation.

Diagnostic criteria:
A. At least five attacks fulfilling criteria B–D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting **15–180 minutes** (when untreated)
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 oedema
d) forehead and facial sweating
e) miosis and/or ptosis
2. a sense of restlessness or agitation
D. Occurring with a frequency between one every other day and eight per day
E. Not better accounted for by another ICHD-3 diagnosis.

3.1.1 Episodic cluster headache

Description: Cluster headache attacks occurring in periods lasting from seven days to one year, separated by pain-free periods lasting at least three months.

Diagnostic criteria:

A. Attacks fulfilling criteria for 3.1 Cluster headache and occurring in bouts (cluster periods)
B. B. At least two cluster periods lasting from seven days to one year (when untreated) and separated by pain-free remission periods of ≥3 months.

Comment: Cluster periods usually last between two weeks and three months.

3.1.2 Chronic cluster headache

Description: Cluster headache attacks occurring for one year or longer without remission, or with remission periods lasting less than three months.

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.1 Cluster headache, and criterion B below
B. Occurring without a remission period, or with remissions lasting <3 months, for at least one year.

Comment: 3.1.2 Chronic cluster headache may arise de novo (previously referred to as primary chronic cluster headache), or evolve from 3.1.1 Episodic cluster headache (previously secondary chronic cluster headache). In some patients, change occurs from 3.1.2 Chronic cluster headache to 3.1.1 Episodic cluster headache

3.2 Paroxysmal hemicrania

Description: Attacks of severe, strictly unilateral pain, which is orbital, supraorbital, temporal or in any combination of these sites, **lasting 2–30 minutes** and occurring several or many times a day. The attacks are usually associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, forehead and facial sweating, miosis, ptosis and/or eyelid
oedema. They respond absolutely to indomethacin.

Diagnostic criteria:
A. At least 20 attacks fulfilling criteria B–E
B. Severe unilateral orbital, supraorbital and/or temporal pain lasting 2–30 minutes
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 oedema
d) forehead and facial sweating
e) miosis and/or ptosis
2. a sense of restlessness or agitation
D. Occurring with a frequency of >5 per day
E. Prevented absolutely by therapeutic doses of indomethacin
F. Not better accounted for by another ICHD-3 diagnosis.

Notes:
1. During part, but less than half, of the active timecourse of 3.2 Paroxysmal hemicrania, attacks may be less frequent.
2. 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. Smaller maintenance doses are often employed.

Comment: In contrast to cluster headache, there is no male predominance. Onset is usually in adulthood, although childhood cases are reported.

3.2.1 Episodic paroxysmal hemicrania

Description: Attacks of paroxysmal hemicrania occurring in periods lasting from **seven days to one year**, separated by *pain-free periods lasting at least three months*.

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.2 Paroxysmal hemicranias and occurring in bouts
B. At least two bouts lasting from seven days to one year (when untreated) and separated by pain-free remission periods of ≥3 months.

3.2.2 Chronic paroxysmal hemicrania (CPH)

Description: Attacks of paroxysmal hemicrania occurring for more than one year without remission, or with remission periods lasting less than three months.

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.2 Paroxysmal hemicrania, and criterion B below
B. Occurring without a remission period, or with remissions lasting <3 months, for at least one year.

3.3 Short-lasting unilateral neuralgiform headache attacks

Description: Attacks of moderate or severe, strictly unilateral head pain lasting ***seconds to minutes***, occurring at least once a day and usually associated with prominent lacrimation and redness of the ipsilateral eye.

Diagnostic criteria:
A. At least 20 attacks fulfilling criteria B–D
B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting for **1–600 seconds** and occurring as single stabs, series of stabs or in a saw-tooth pattern
C. At least one of the following five cranial autonomic symptoms or signs, ipsilateral to the pain:
1. conjunctival injection and/or lacrimation
2. nasal congestion and/or rhinorrhoea
3. eyelid oedema
4. forehead and facial sweating
5. miosis and/or ptosis
D. Occurring with a frequency of at least one a day1
E. Not better accounted for by another ICHD-3 diagnosis.

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

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3 Short-lasting unilateral neuralgiform headache attacks, and criterion B below
B. Both of the following, ipsilateral to the pain:
1. conjunctival injection
2. lacrimation (tearing).

3.3.1.1 Episodic SUNCT

Description: Attacks of SUNCT occurring in periods lasting from **seven days to one year**, separated by pain-free periods lasting three months or more.

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3.1 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing and occurring in bouts
B. At least two bouts lasting from seven days to one year (when untreated) and separated by pain-free remission periods of ≥3 months.

3.3.1.2 Chronic SUNCT

Description: Attacks of SUNCT occurring for more than one year **without remission**, or with remission periods lasting **less than three months**.

Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3.1 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing, and criterion B below
B. Occurring without a remission period, or with remissions lasting <3 months, for at least one year.

3.3.2 Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)
Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3 Short-lasting unilateral neuralgiform headache attacks, and criterion B below
B. Not more than one of the following, ipsilateral to the pain:
1. conjunctival injection
2. lacrimation (tearing) .

3.3.2.1 Episodic SUNA

Diagnostic criteria:

A. Attacks fulfilling criteria for 3.3.2 Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms and occurring in bouts
B. At least two bouts lasting from seven days to one year (when untreated) and separate by pain-free remission periods of ≥3 months.

3.3.2.2 Chronic SUNA

Diagnostic criteria:

A. Attacks fulfilling criteria for 3.3.2 Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms, and criterion B below
B. Occurring without a remission period, or with remissions lasting <3 months, for at least one year.

3.4 Hemicrania continua

Diagnostic criteria:
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 oedema
d) forehead and facial sweating
e) 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
E. Not better accounted for by another ICHD-3 diagnosis.

Note: 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. Smaller maintenance doses are often employed.

3.4.1 Hemicrania continua, remitting subtype

Diagnostic criteria:
A. Headache fulfilling criteria for 3.4 Hemicrania continua, and criterion B below
B. Headache is not daily or continuous, but interrupted (without treatment) by *remission periods of ≥24 hours.*

3.4.2 Hemicrania continua, unremitting subtype
Diagnostic criteria:
A. Headache fulfilling criteria for 3.4 Hemicrania continua, and criterion B below
B. Headache is daily and continuous for at least one
year, *without remission* periods of 24 hours.

3.5 Probable trigeminal autonomic cephalalgia

Diagnostic criteria:
A. Headache attacks fulfilling all but one of criteria A–D for 3.1 Cluster headache, criteria A–E for 3.2 Paroxysmal hemicrania, criteria A–D for 3.3 Shortlasting unilateral neuralgiform headache attacks or criteria A–D for 3.4 Hemicrania continua
B. Not fulfilling ICHD-3 criteria for any other headache disorder
C**.** Not better accounted for by another ICHD-3 diagnosis.

Comment: Patients may be coded;
3.5.1 Probable cluster headache,
3.5.2 Probable paroxysmal hemicrania,

3.5.3 Probable short-lasting unilateral neuralgiform headache attacks or
3.5.4 Probable hemicrania continua.