

# Other primary Headaches



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# What is new for chapter 4: other primary headache

- Re-arrangement and rename of diagnostic entities
- 4 major groups
- Schema for definite criteria
- Schema for “probable”...headaches

# ICHD-2



# ICHD-3 (beta)

1. Primary stabbing headache
2. Primary cough headache
3. Primary **exertional** headache
4. Primary headache associated with sexual activity
5. Hypnic headache
6. Primary thunderclap headache
- 7. Hemicrania continua**
8. New daily-persistent headache (NDPH)

- 10. External compression headache**
- 11. Cold-stimulus headache**

**A13.7.1 Nummular headache**

## **3.4 Hemicrania continua**

1. Primary cough headache
2. Primary **exercise** headache
3. Primary headache associated with sexual activity
4. Primary thunderclap headache

## **5. Cold-stimulus headache**

## **6. External-pressure headache**

## **7. Primary stabbing headache**

## **8. Nummular Headache**

9. Hypnic headache
- 10. New daily persistent headache (NDPH)**

## **A4.11 Epicrania Fugax**

# 4 major groups

## Physical exertion

1. Primary cough headache
2. Primary exercise headache
3. Primary headache associated with sexual activity
4. Primary thunderclap headache

## Direct physical stimuli

5. Cold-stimulus headache
6. External-pressure headache

## Epicranial headaches

7. Primary stabbing headache
  8. Nummular Headache
- A4.11 Epicrania Fugax

## Others

9. Hypnic headache
10. New daily persistent headache (NDPH)

# Format for **definite** criteria

- Diagnostic schema for “definite” criteria as chapters 1-3
  - A. At least 2 episodes
  - B. Brought on by and occurring only in association with..
  - C. Headache pattern
  - D. Not accounted for by another ICHD-III..  
(secondary headache should be excluded especially for “other primary headache”)

## Format of diagnostic criteria for “Probable” other primary headaches

A. Either of the following:

1. a single headache episode fulfilling criteria B-D
2. at least two headache episodes fulfilling criterion B and either of criteria C and D

B-D. Headache characteristics

E. Not fulfilling ICHD-III criteria for any other headache disorder

F. Not better accounted for by another ICHD-III diagnosis.

## 4.1 Primary cough headache

- 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 manoeuvre
- C. Sudden onset
- D. Lasting between 1 sec and 2 hours
- E. Not better accounted for by another ICHD-III diagnosis.

## 4.1.1 Probable primary cough headache

- A. Either of the following:
  1. a single headache episode fulfilling criteria B-D
  2. at least two headache episodes fulfilling criterion B and either of criteria C and D
- B. Brought on by and occurring only in association with coughing, straining and/or other Valsalva manoeuvre
- C. Sudden onset
- D. Lasting between 1 second and 2 hours
- E. Not fulfilling ICHD-III criteria for any other headache disorder
- F. Not better accounted for by another ICHD-III diagnosis.



# Some cases!

# Case 1

- A 56-year-old man who had a history of well-controlled hypertension and episodic tension-type headache was awoken from sleep 13 nights per month for the past 6 months between 1:00 AM and 2:30 AM with a moderately severe bifrontal pressing headache, which persisted for 1 to 2 hours untreated and 10 minutes if treated with a caffeine-containing over-the-counter analgesic.

## Case 2

- A 45-year-old woman presents with a 3-month history of a scalp pain. She describes a burning, stinging, itching, and sore pain of the midposterior frontal and anterior parietal scalp in an elliptic distribution extending across both sides with a diameter of approximately 5 cm. The pain is present intermittently daily, lasting hours at a time with an intensity of 5/10. At times, the area is sensitive when she brushes her hair. Ibuprofen may reduce the discomfort. She has seen two dermatologists who found normal skin examinations. There is no history of migraine or other headaches. There is a medical history of hypertension. Neurologic examination was normal with no abnormality of scalp sensation. MRI of the brain was normal. Blood work was normal, including the following: erythrocyte sedimentation rate, 12 mm per hour; rheumatoid factor, 11 IU/mL; antinuclear antibody (ANA) screen, negative; The patient was placed on gabapentin (100 mg by mouth three times a day), which she took as needed with a reduction in the level of pain to a 2/10

## Case 3

- A 35-year-old man while taking a shower, had an acute onset of severe dull headache in the occipital region spreading anteriorly, reached its peak in minutes and subsided in about 30 minutes. He was seen in the clinic the next day when he felt well. He was a normotensive subject. He had no nausea/vomiting during or after the headache spell. His neurological examination was normal. He volunteered on directing questioning of spells of dull occipital and nuchal headache at height of orgasm on many occasions. However, he was not having sex in the shower. Investigations revealed a normal computed tomography (CT) scan of brain, a normal clear cerebrospinal fluid (CSF) and clean CT angiography using a subtraction software. Again a missing history was revealed.

## Case 4

- A 56-year-old lady presented with history of daily several spells of brief (few seconds) severe burning sensation spreading from near about the right angle of jaw along in a linear fashion over the lateral side of right face and then over the right ear to the right nape of the neck. This sensation was associated with a feeling of fullness in the right ear. In between, the area remained sore to touch. She had no headache at all; neither she had any associated classical autonomic features. There was no sensory loss. She had no neurological deficit. ENT examination was normal. Her ECG was normal. MRI brain with contrast and internal auditory meatus views were normal. She was treated with carbamazepine 400 mg daily with near complete relief of symptoms.

# Case 1: hypnic headache

- Hypnic headache is characterized by recurring headache attacks developing **only during sleep**.
- Hypnic headache is more frequent in **women**, the onset **typically** occurs **after the age of 50 years** (mean approximately 60) but may occur in younger people.
- By definition, attacks occur at night during sleep (or during a nap in 10% of cases), waking the patient at constant time intervals (**“alarm clock headache”**).
- Headache is usually mild-moderate in intensity, being severe in 34% of the cases, and lasts from 15 to 240 minutes after waking, but longer attacks of up to 10 hours also have been described
- Triggers or **autonomic phenomena are not** part of the clinical picture of hypnic headache.

# Case 1: hypnic headache

- Pain location is not characteristic, being bilateral in approximately two-thirds of cases.
- Pathophysiological mechanisms of hypnic headache:
  - A) dysfunction within the suprachiasmatic nucleus in the hypothalamus, which is considered the brain pacemaker.
  - B) A decrease in nocturnal secretion of melatonin.
  - C) most hypnic headaches attacks arise from non-REM sleep stages, mainly sleep stage N2.
- Secondary causes of headache must be ruled out before the diagnosis of hypnic headache:
  - nocturnal hypertension, increased intracranial pressure (mass lesion or idiopathic intracranial hypertension), trigeminal autonomic cephalalgias (specifically cluster headache), medication-overuse (rebound) headache, and sleep apnea headache.

# Case 1: hypnic headache

- There are **no controlled trials** for the treatment of hypnic headache, so treatment recommendations are based on case reports or small open case series.
- Main treatment options include **caffeine, lithium, indomethacin, and melatonin.**
- There are scattered case reports documenting the usefulness of topiramate, amitriptyline, verapamil, prednisone, acetazolamide, gabapentin, and pizotifen



## 4.9 Hypnic headache

- A. Recurrent ~~dull~~ headache attacks fulfilling criteria B-E
- B. Developing only during sleep, and causing wakening
- C. Occurring on  $\geq 10$  days per month for >3 months
- D. Lasting  $\geq 15$  minutes and for up to 4 hours after waking  
~~— first occurs after age of 50 years~~
- E. No cranial autonomic symptoms or restlessness
- F. Not better accounted for by another ICHD-III diagnosis.

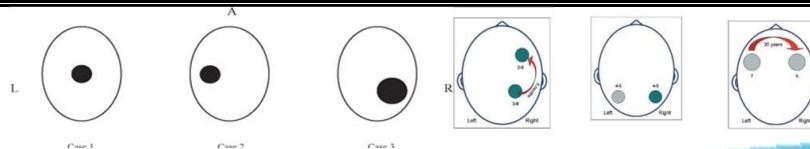
\* Hypnic headache syndrome; ‘alarm clock’ headache.

## Case 2 : nummular headache

## 4.8 Nummular headache

- A. Continuous or intermittent ~~mild to moderate~~ head pain fulfilling criterion B
- B. Felt exclusively in an area of the scalp, with **all** of the following four characteristics:
1. sharply contoured
  2. fixed in size and shape
  3. round or elliptical
  4. **1**-6 cm in diameter
- C. Not better accounted for by another ICHD-III diagnosis.

\* Coin-shaped headache



## Case 2 : nummular headache

- Also called **coin-shaped headache**, nummular headache is an unusual headache disorder characterized by pain in a small circumscribed area of the scalp **in the absence** of any underlying structural lesion.
- nummular headaches are more common in **women** with a mean age of **onset at 45 years**.
- A preexisting headache diagnosis has been described in **approximately 50% of patients**, the most common **migraine**.

## Case 2 : numular headache clinical features:

- mild to moderate intensity and confined to a round or elliptical unchanging area from 1 to 6 cm in diameter.
- the pain being continuous or intermittent
- Superimposed on the continuous pain, lancinating pain may occur.
- The pain remains focal and well circumscribed and characteristically never radiates.
- The affected area may also suffer sensory dysfunction (allodynia, paresthesia, or hyperesthesia) with the parietal region being the area of scalp most often affected.
- Typically, nummular headache is not accompanied by photophobia, nausea, vomiting, or autonomic symptoms.

# Case 2: nummular headache

## The differential diagnosis

- **Secondary headaches:**
  - underlying structural lesions as **cranial bone lesions** caused by metastatic cancer, multiple myeloma, Paget disease, or osteomyelitis
- **Primary headaches:**
  - primary stabbing headache
  - epicrania fugax
  - other cranial neuralgias.

## Case 2 : nummular headache treatment

- Little evidence exists for nummular headache treatment
- 60% of patients respond to analgesics and nonsteroidal anti-inflammatory drugs(NSAID).
- For patients with more severe, refractory, or continuous pain, **preventive** treatment should be offered.  
**Gabapentin and tricyclic antidepressants** have been reported to be effective
- Other options include **indomethacin**, and even **transcutaneous electrical nerve stimulation**.
- New approaches include the use of **local onabotulinumtoxinA**
- However, nummular headache **often becomes refractory** to prophylactic and analgesic therapies.

# Topography of nummular headache (NH).





# Topography of nummular headache (NH)

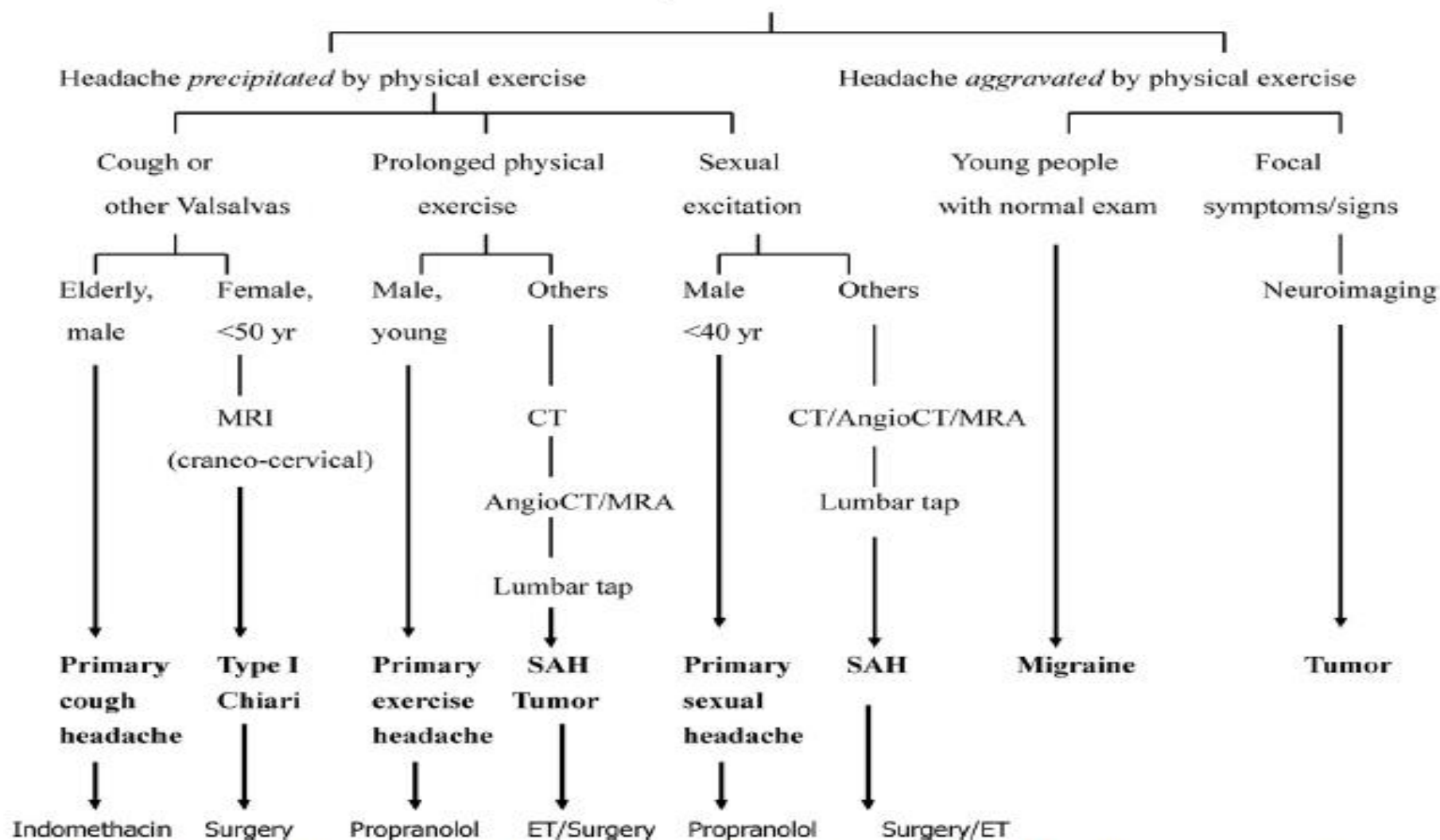


# Case 3 : ??

# QUESTIONS

- 1. What is the initial diagnosis in the this case?
  
  
  
  
  
  
  
  
  
  
- 2. What was the final diagnosis in this case?

## Activity-related headache



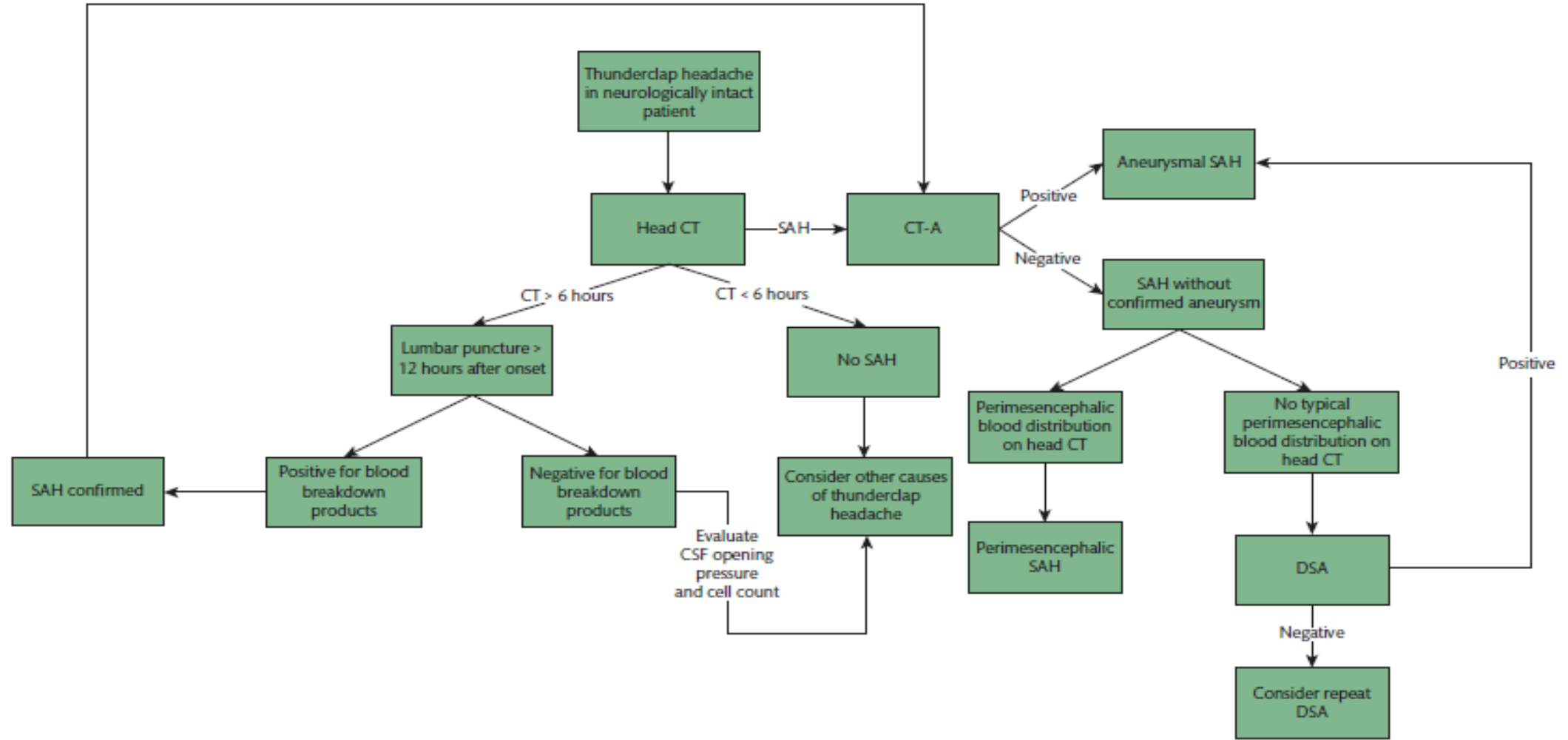
**Fig. 4.** Differential diagnosis and management of activity-related headaches. ET, endovascular treatment; SAH, subarachnoid hemorrhage.

# Thunderclap headache

**Table 34.1** Causes of thunderclap headache.

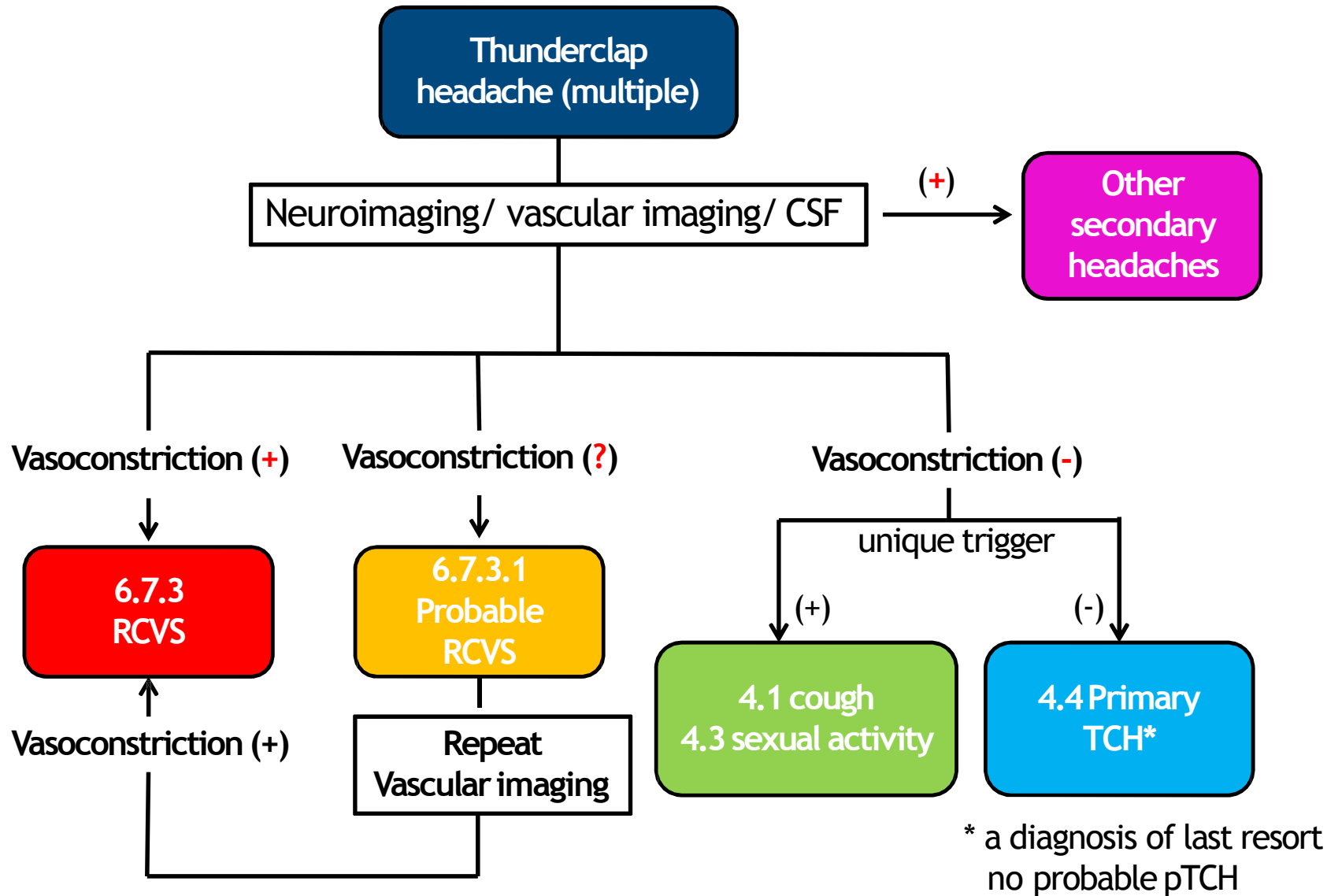
Thunderclap headache in secondary headaches (4)	Thunderclap headache in primary headaches
<p><b>More common causes</b></p> <ul style="list-style-type: none"><li>• Subarachnoid haemorrhage (8)</li><li>• Reversible cerebral vasoconstriction syndrome (39,41)</li><li>• Cervical artery dissection (45–47)</li><li>• Cerebral venous sinus thrombosis (48–51)</li><li>• Spontaneous intracranial hypotension (52–54)</li><li>• Stroke (ischaemic, haemorrhagic) (56–60)</li><li>• Pituitary apoplexy (61–63)</li></ul>	<ul style="list-style-type: none"><li>• Primary cough headache (4)</li><li>• Primary headache associated with sexual activity (4)</li><li>• Primary thunderclap headache (4)</li><li>• Primary exercise headache (4)</li></ul>
<p><b>Miscellaneous causes</b></p> <ul style="list-style-type: none"><li>• Retroclival haematoma (64,65)</li><li>• Meningitis or vasculitis (59)</li><li>• Acute hydrocephalus (aqueduct stenosis (67)/colloid cyst (68)</li><li>• Sinusitis (66)</li><li>• Cardiac cephalalgia (70,71)</li><li>• Pheochromocytoma (69)</li></ul>	

# Thunderclap headache



**Figure 34.1** Diagnostic evaluation of neurologically intact patients with thunderclap headache.

CT, computed tomography; CT-A, computed tomographic angiography; SAH, subarachnoid haemorrhage; CSF, cerebrospinal fluid; DSA, digital subtraction angiography.





## 4.3 Primary headache associated with sexual activity

- 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-III diagnosis.

**Note:** 1. No more subtypes: ~~Preorgasmic~~ ~~Orgasmic~~  
2. Vascular lesions esp. RCVS should be excluded first



# 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 the absence of any intracranial disorder

## *Notes*

- 1. The 4.3 Primary headache associated with sexual activity is **not associated** with disturbance of consciousness, vomiting or visual, sensory or motor symptoms, **whereas symptomatic sexual headache may be**. On the first onset of headache with sexual activity, it is **mandatory** to exclude subarachnoid hemorrhage (SAH), intra- and extracranial arterial dissection and RCVS.
- 2. **Multiple explosive** headaches during sexual activities should be considered as 6.7.3 **Headache attributed to RCVS** until proven otherwise by angiographic studies (including conventional, magnetic resonance or CT angiography) or transcranial Doppler ultrasonography. Of note, **vasoconstrictions may not be observed in the early stage of RCVS**; therefore, follow-up studies may be needed.

- 3. Recent studies have shown that **up to 40%** of all cases run a chronic course over more than a year.
- 4. Some patients experience **only one attack** of 4.3 Primary headache associated with sexual activity during their lives; they should be diagnosed as 4.3.1 **Probable** primary headache associated with sexual activity.
- 5. Epidemiological research has further shown that 4.3 Primary headache associated with sexual activity can occur at any sexually active age, is **more prevalent in males** than in females (ratios range from **1.2:1 to 3:1**).

- 6. It occurs **independently of the type of sexual activity**, is not accompanied by autonomic or vegetative symptoms in most cases, is **bilateral** in two-thirds and unilateral in one-third of cases and is **diffuse or occipitally localized in 80% of cases**
- 7. Attack frequency of 4.3 Primary headache associated with sexual activity should always be related to the frequency of sexual activity.
- 8. **The differential diagnosis** for sexual headache includes: RCVS, arterial dissection, SAH caused by aneurysm rupture or arteriovenous malformation (AVM), nonhemorrhagic strokes, meningitis, encephalitis, hemorrhage into a cerebral tumor and pheochromocytoma

- 9. Given that **myocardial ischemia** may occur during sexual intercourse, **referred cardiac pain** should be considered as a potential cause of sexual headache in individuals with risk factors for coronary artery disease.
- 10. The use of several drugs has also been linked in case reports to sexual headaches associated with neurologic symptoms:
  - **amiodarone, birth control pills, pseudoephedrine and cannabis**
- 11. Several other non-neurologic disorders have been suggested as possible causes for sexual headache, including **glaucoma, myxedema, anemia, chronic obstructive pulmonary disease, sinusitis, hypoglycemia, Cushing disease** and **occlusion of the abdominal aorta**.

# treatment

- Some patients can lessen the severity or abort the headache by **stopping sexual activity** when the headaches begin or by taking a more passive role.
- **Weight loss** and an **exercise programme** might also be helpful.
- It is suggested that patients should remain sexually **inactive during the acute stage** as recurrent attacks are frequent when they resume sexual activities early.
- **Daily beta blockers**, such as propranolol, metoprolol, and bisoprolol, or a **calcium channel blocker**, such as verapamil or diltiazem, may be effective for prophylaxis in patients with frequent attacks
- **Indomethacin, 25– 100 mg 1– 2 hours before** intercourse, is effective as a pre- emptive treatment
- **Ergotamine tartrate, methysergide, or naratriptan** have also shown positive results.

# treatment

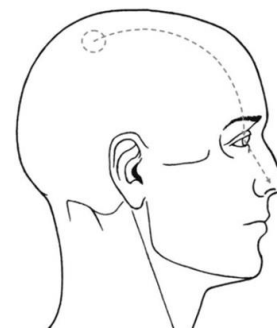
- Because there is substantial overlapping of sexual headache and RCVS, **nimodipine**, a calcium channel blocker, might also be considered.
- In contrast, **vasoconstrictive drugs such as triptans**, although possibly effective in aborting the pain, might aggravate vasoconstriction and **should not be used** until RCVS or dissection has been excluded.
- **Other NSAIDs** (ibuprofen and diclofenac), aspirin or paracetamol, given after the onset of headache, are **generally ineffective**.

# Case 4: *epicrania fugax*



## A4.11 Epicrania fugax

- A. Recurrent stabbing head pain attacks lasting 1-10 seconds, fulfilling criterion B
- B. The pain is felt to move across the surface of one hemicranium in a linear or zig-zag trajectory, commencing and terminating in the territories of different nerves
- C. Not better accounted for by another ICHD-III diagnosis.



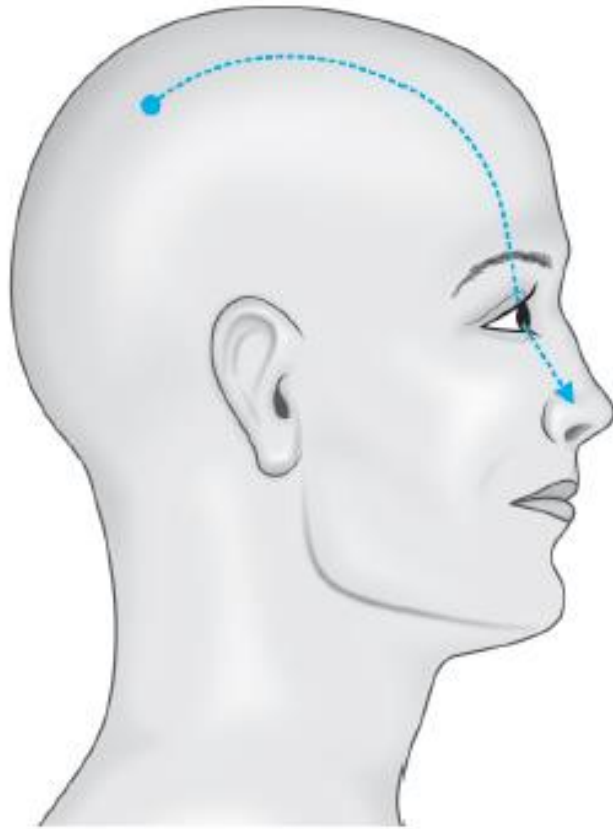
# Description

- Brief paroxysmal head pain, with stabbing quality, describing a **linear or zig-zag trajectory** across the surface of one **hemispheric**.
- Patients with A4.11 Epicrania fugax describe their pain in terms of its trajectory between two distant points on the head surface, with motion from onset to termination taking just a few seconds.
- Such **dynamic topography** is a distinctive attribute that differentiates A4.11 Epicrania fugax from other epicranial headaches and neuralgias

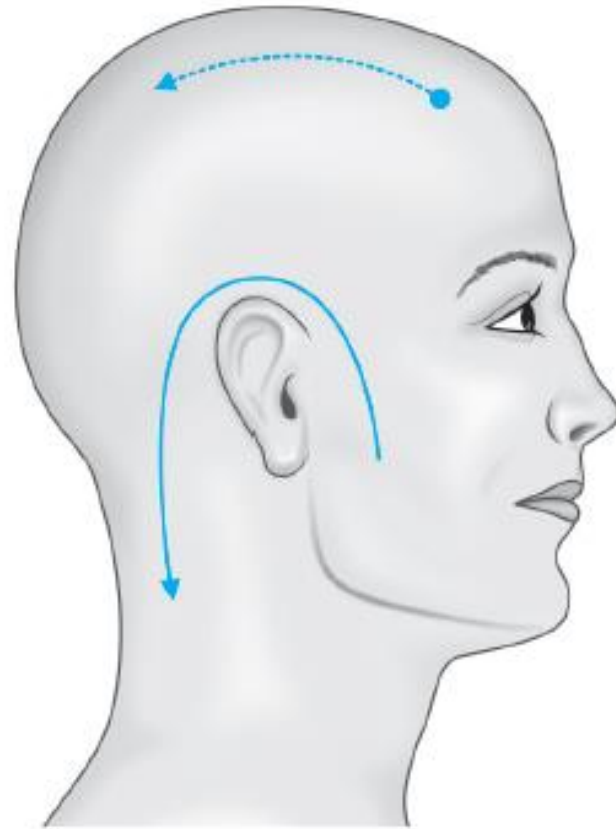
- The onset and termination points remain constant in each patient, with pain usually moving forward from a posterior hemicranial area toward the ipsilateral eye or nose, but backward radiation is also possible from a frontal or periorbital area toward the occipital region.
- In all cases, pain is strictly unilateral, although some patients have
  - shifting sides.
  - At the end of attacks, ipsilateral autonomic signs such as lacrimation,
  - conjunctival injection and/or rhinorrhea may occur

- Although attacks are mostly spontaneous, they may occasionally be triggered by touch on the point of onset, which may remain tender between attacks.

# Distribution of pain in epicrania fugax



**A**



**B**

# Management

- Gabapentin and lamotrigine have been reported as providing partial or complete relief for some patients
- A few patients have benefited from pregabalin, levetiracetam, and carbamazepine
- Amitriptyline, indomethacin, occipital nerve blocks, and trochlear injections have also been occasionally effective

THANK YOU