Classification and Diagnosis of Secondary Headaches, Part III- Substances, Metabolic, EENT, and Neuralgias

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10. Headache attributed to disorder of homoeostasis

10.1 Headache attributed to hypoxia and/or hypercapnia
10.2 Dialysis headache
10.3 Headache attributed to arterial hypertension
10.4 Headache attributed to hypothyroidism
10.5 Headache attributed to fasting
10.6 Cardiac cephalalgia
10.7 Headache attributed to other disorder of homoeostasis

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Hypoxia and/or Hypercapnea: High altitude headache:

- Headache is most common neurologic syndrome arising from ascent to altitudes > 2500 meters
- Headache is bilateral, frontal or frontotemporal
- Dull or pressing
- Mild-moderate
- Aggravated by movement, exertion, straining, coughing, bending
- Develops within 24 hours of ascent
- Resolves within 8 hours of descent
- Affects younger people more often (lack of atrophy?)
- Women > Men
- Often awakens from sleep or present upon awakening

Hypoxia and/or Hypercapnea: Diving headache

- Diving below 10 meters
- Headache has no typical characteristics
- Associated with one or more features of CO2 intoxication
- Resolves within 1 hour of 100% O2
- Diving headache usually intensifies during decompression phase of dive or upon resurfacing
- Lightheadedness, confusion, dyspnea, flushing feeling, incoordination

Hypoxia and/or Hypercapnea: Sleep apnea headache

- These are recurrent headaches, >15 days/month
- Bilateral, pressing
- No associated symptoms
- Each headache resolves in 30 minutes
- Sleep apnea demonstrated by polysomnography
- Headache present upon awakening
- Headache ceases within 72 hours and doesn't recur after sleep apnea treated

Dialysis Headache:

- 70% of dialysis patients complain of headaches
- ICHD-II does not describe the headache
- Headache commonly occurs in association with hypotension and dialysis disequilibrium syndrome
- Dysequilibrium syndrome may begin as a headache progress to obtundation and coma with or without sz.
- As caffeine is removed by dialysis, consider diagnosis of caffeine withdrawal

Headache related to arterial hypertension:

- Mild or moderate hypertension does not cause headache
- Various disorders that lead to paroxysmal, abrupt, severe elevations are associated with headaches
  - **Pheochromocytoma**
  - **Hypertensive crises** (with or without encephalopathy)
  - **Pre-eclampsia and eclampsia**

Pheochromocytoma:

- These are catecholamine-producing tumors arising from chromaffin cells
- Consider in patients with headaches associated with hypertension, autonomic disturbances, panic attacks
- MEN II, Von-Hippel Landau, Neurofibromatosis type 1, familial carotid body tumors
- Usually found within adrenal medulla; 9-23% extra-adrenal

Pheochromocytoma:

- **Headache**
- 80% report sudden onset headache
- Severe, frontal or occipital
- Pulsating or steady
- Short duration of pain; < 15 mins in 50%; <60 mins in 70%
- Hypertension in 80%-paroxysmal in 50%
- 13% have normal blood pressure;8% asymptomatic
- ICHD-II states that headache develops concomitantly with abrupt rise in BP and that headache resolves or markedly improves within 1 hour of normalization of pressure

- **Associated features**
- Sweating-ICHD
- Palpitations-ICHD
- Anxiety-ICHD
- Facial pallor –ICHD or flushing
- Tachycardia
- Paroxysms of pain can occur spontaneously or provoked by exertion, stress, pressor medications, postural changes

Pheochromocytoma:

- **Diagnosis:**
- Increased 24 hour urinary excretion of metanepharine and normetanephrine, vanillymandelic acid and total catecholamines
- CT or MRI of neck, chest, abdomen and pelvis
- **Treatment:**
- Selective post synaptic α-1 adrenergic blockers (prazosin, terazosin,doxazocin)
- Calcium channel blockers

Headache attributed to hypertensive crises without hypertensive encephalopathy:

- **Headache:**
- Develops during hypertensive crises (rise in systolic >160 and/or diastolic >120)
- Bilateral
- Pulsating
- Precipitated by exertion
- Resolves within 1 hour after normalization of BP

Headache attributed to hypertensive encephalopathy:

- acute cerebral syndrome caused by sudden severe hypertension
- the rate and extent of BP rise are most important factors
- In normotensives this may develop at level of 160?100 without retinopathy
- In patients with chronic hypertension, encephalopathy usually not before systolic > 250 and diastolic > 120 with grade 3 or 4 retinopathy
- ICHD-II persistent BP elevations to > 160/100 with at least 2 of:
- Confusion
- Reduced level of consciousness
- Visual disturbances including blindness
- Seizures

Headache attributed to hypertensive encephalopathy:

- Develops in close relation to BP elevation
- Diffuse
- Pulsating
- Inc with exertion
- Resolves within 3 months after control of hypertension
- Hypertensive encephalopathy is part of posterior leuko encephalopathy syndrome (PLES)
- Characterized by headache nausea vomiting visual disturbances altered mental status seizures focal signs and abnormal MRI
- MRI findings include:
- Posterior white matter edema (especially parietal-occipital) with spread to basal ganglia, brainstem and cerebellum

Headache attributed to Pre-eclampsia and eclampsia:

- Pre-eclampsia usually occurs after 20 weeks gestation
- Consists of hypertension, edema, and proteinuria (> .3 g/2 hours)
- Eclampsia – adds seizure
- ICHD-II says up to 4 weeks postpartum for both
- **Headache:**
- Bilateral, pulsatile, worse with exertion
- Thunderclap headache has been reported (not ICHD)
- Incidence of both disorders increased by presence of prior hypertension, nulliparity, multiple pregnancies, prior or family history of pre-eclampsia and chronic hypertension

Cardiac cephalgia

- This is a headache that occurs during acute myocardial ischemia
- Headache begins in close proximity to onset of vigorous exercise
- Headache subsides with rest or anti-anginal treatment
- Reports have occurred while at rest (not ICHD)
- Headache associated with nausea (ICHD)
- Often unilateral (not ICHD)
- Not precipitated by valsalva (not ICHD)

11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cranial structures

11.1 Headache attributed to disorder of cranial bone
11.2 Headache attributed to disorder of neck
11.3 Headache attributed to disorder of eyes
11.4 Headache attributed to disorder of ears
11.5 Headache attributed to rhinosinusitis
11.6 Headache attributed to disorder of teeth, jaws or related structures
11.7 Headache or facial pain attributed to temporomandibular joint (TMJ) disorder
11.8 Headache attributed to other disorder of the above

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Headache attributed to disorder of cranial bone:

- Most disorders of the skull not associated with headache
- Exceptions: Osteoarthritis, Pagets, multiple myeloma
- Headache can result from skull lesions that:
- involve periosteum
- are rapidly expansive
- are aggressively osteoclastic
- have inflammatory component
- Lesions of mastoid and petrositis cause headaches

Cervicogenic headache:

- 0.4-2.5% of general pop.
- 15-20% of people with chronic headaches
- Mean age 43
- F:M 4:1

Cervicogenic headache:

- Pain sensitive cervical structures:
- 1. C2 sensory root, greater and lesser occipital nerves-refer to back of head
- 2. C1 (?) root- refers to vertex or frontal head region
- 3. Connections between tentorial branches of V1 and posterior fossa branches of C2- pain referral from C2 structures to front of head
- Descending spinal tract of V intermingling impulses from upper cervical segments with those of V- referral to the head

Cervicogenic headache:

- **Accepted cervical causes of headache:**
- Developmental abnormalities and tumors of craniovertebral junction and upper cervical spine
- Pagets of skull with secondary basilar invagination
- Osteo of upper cervical vertebrae
- RA or ankylosing spondylitis of upper cervical spine
- Traumatic subluxation
- Retropharyngeal tendonitis
- Craniocervical dystonias

- **Controversial causes:**
- Cervical disk disease and spondylosis
- Osteochondritis
- "whiplash"

Cervicogenic headache: Clinical Features:

- Headache of cervical origin is non-specific
- Suggestive features include:
- Posterior location
- Triggering by flexion, coughing, straining
- Postural component may mimic low pressure
- Vertigo, facial numbness, limb weakness, ataxia
- Uni- or bi-lateral

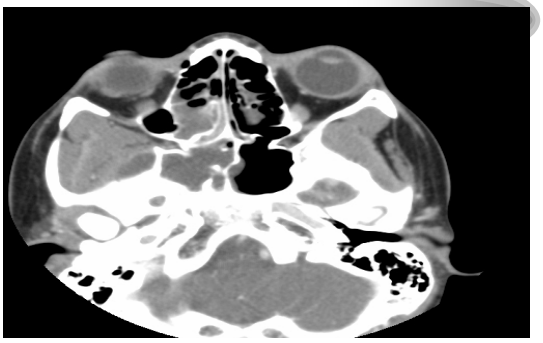
Headache attributed to rhinosinusitis:

- Systemic diseases that predispose to sinusitis:
- cystic fibrosis
- immune deficiency (HIV, chemotherapy, post-transplant, IDDM)
- bronchiectasis
- immobile cilia syndrome
- Local factors that predispose to sinusitis:
- URI
- Allergic rhinitis
- Overuse topical decongestants
- Hypertrophied adenoids
- Deviated septum
- Nasal polyps/tumors
- Smoking
- **The most common predisposing factor is mucosal inflammation from viral URI or allergic rhinitis**

Rhinosinusitis:

- Acute sinus infections usually manifested by facial pain and tenderness, congestion and purulent discharge
- Other signs include anosomia, pain on chewing, halitosis
- Fever present 50% adults, 60% children
- Headache location:
- Frontal sinusitis-directly over sinus, may radiate to vertex or retro-orbitally
- Maxillary- Over antral area, may radiate to upper teeth or forehead
- Ethmoid—between and behind eyes, may radiate temporally
- Sphenoid-Occipital area, vertex, frontal, or retro-orbitally

Sinusitis

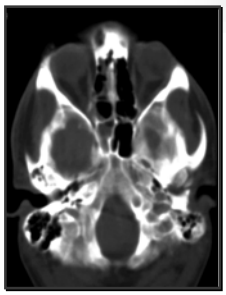


Sphenoid Sinusitis

- Headache is major complaint in all patients who can complain
- Worsened by standing, walking, bending etc
- Interferes with sleep
- Resistant to opioids
- Usually occipital, frontal, temporal headache
- Periorbital pain is common
- Nausea/vomiting common
- Discharge, congestion not common
- Pain/paresthesias in facial distribution of V, photophobia, tearing

- **Consider diagnosis in any severe, intractable, new onset headache that interferes with sleep and not relieved with simple analgesics**

Sphenoid Sinusitis



11.2.1 Cervicogenic headache

A. Pain, referred from a source in the neck and perceived in one or more regions of the head and/or face, fulfilling criteria C and D

B. Clinical, laboratory and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache

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11.2.1 Cervicogenic headache

C. Evidence that the pain can be attributed to the neck disorder or lesion based on ≥1 of the following:
   1. demonstration of clinical signs that implicate a source of pain in the neck
   2. abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls

D. Pain resolves within 3 mo after successful treatment of the causative disorder or lesion

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11.5 Headache attributed to rhinosinusitis

A. Frontal headache accompanied by pain in one or more regions of the face, ears or teeth and fulfilling criteria C and D

B. Clinical, nasal endoscopic, CT and/or MRI imaging and/or laboratory evidence of acute or acute-on-chronic rhinosinusitis

C. Headache and facial pain develop simultaneously with onset or acute exacerbation of rhinosinusitis

D. Headache and/or facial pain resolve within 7 d after remission or successful treatment of acute or acute-on-chronic rhinosinusitis

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11.5 Headache attributed to rhinosinusitis
Notes

- 11.5 *Headache attributed to rhinosinusitis* is differentiated from “sinus headaches”, a commonly-made but non-specific diagnosis. Most such cases fulfil the criteria for 1.1 *Migraine without aura*, with headache either accompanied by prominent autonomic symptoms in the nose or triggered by nasal changes
- *Chronic sinusitis* is not a cause of headache or facial pain unless relapsing into an acute stage

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Part 3: Cranial neuralgias, central and primary facial pain and other headaches

13. Cranial neuralgias and central causes of facial pain
14. Other headache, cranial neuralgia, central or primary facial pain

13. Cranial neuralgias and central causes of facial pain

13.1 Trigeminal neuralgia
13.2 Glossopharyngeal neuralgia
13.3 Nervus intermedius neuralgia
13.4 Superior laryngeal neuralgia
13.5 Nasociliary neuralgia
13.6 Supraorbital neuralgia
13.7 Other terminal branch neuralgias
13.8 Occipital neuralgia
13.9 Neck-tongue syndrome
13.10 External compression headache
13.11 Cold-stimulus headache

13. Cranial neuralgias and central causes of facial pain

13.12 Constant pain caused by compression, irritation or distortion of cranial nerves or upper cervical roots by structural lesions
13.13 Optic neuritis
13.14 Ocular diabetic neuropathy
13.15 Head or facial pain attributed to herpes zoster
13.16 Tolosa-Hunt syndrome
13.17 Ophthalmoplegic 'migraine'
13.18 Central causes of facial pain
13.19 Other cranial neuralgia or other centrally mediated facial pain

Nomenclature Issues Regarding TN and GN

- Secondary- When pain result of vascular loop found at surgery
- Classical- Since many patients do not have surgery there is uncertainty as to if neuralgia is primary or secondary
- Symptomatic- A causative lesion, other than vascular compression has been demonstrated

Trigeminal neuralgia

- Classical
- **Symptomatic**- Diagnostic criteria as for classical PLUS:
- Persistence of aching between paroxysms may or may not be present **and**
- A causative lesion other than vascular compression has been demonstrated
- With symptomatic there may be sensory impairment
- No refractory period in symptomatic form

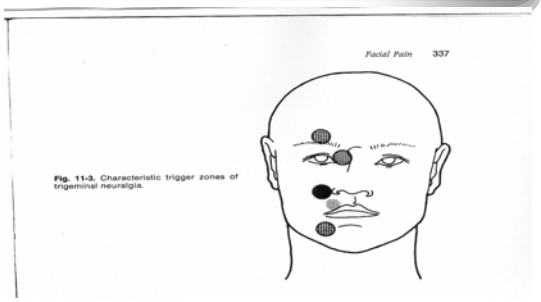
Classical Trigeminal Neuralgia:

- Largest subset of TN patients
- Probably due to vascular cross-compression of REZ
- Yearly incidence:
  - 2.7/100,000 men
  - 5.0/100,00 women
- Age of onset usually after 4th decade
- Peak onset 5th and 6th decades

Classical Trigeminal Neuralgia: Clinical Features

- Unilateral pain, usually in V_2 distribution
- Pain is agonizingly severe, paroxysmal, and lancinating
- Tic doloreux
- Pain is brief, lasting seconds but may occur in volleys lasting several minutes
- Refractory periods
- Perioral trigger zones
- Relief by firm pressure with hands around (but not touching) trigger zones

Characteristic Trigger Zones



13.1.1 Classical trigeminal neuralgia

A. Paroxysmal attacks of pain lasting from a fraction of 1 sec to 2 min, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C

B. Pain has ≥1 of the following characteristics:
   1. intense, sharp, superficial or stabbing
   2. precipitated from trigger areas or by trigger factors

C. Attacks are stereotyped in the individual patient

D. There is no clinically evident neurological deficit

E. Not attributed to another disorder

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Trigeminal Neuralgia: Radiographic Work-Up

- All new patients get contrast-enhanced MRI
- Attention to CP angle
- Sequencing to evaluate arterial and venous relationships

Trigeminal Neuralgia: Medical Therapies

- Carbamazepine 400-800 mg/day Begin 100-200/day, increase 100-200 q 2-3 days
  - Leukopenia, bone-marrow suppression (aplastic anemia)
- Clonazepam 1.5-8mg/day
- Baclofen 40-80mg/day Begin 5-10 tid, increase 5-10 qod
- Gabapentin 900-2400mg/day. Start 300mg and inc qod
- Lamotrigine 150-400mg/day
- Topiramate 25-400mg/day
- No role for opioids

Trigeminal Neuralgia: Surgical Options

- Failure of multiple, appropriate medication trials, at maximum dosages
- Pre-op MRI and MRA to identify vascular compression of trigeminal root
- Choice of major vs. minor procedure
  - MRI/MRA evidence of vascular association supports MVD
  - Elderly or ill patients supports minor procedure

Trigeminal Neuralgia: Minor Surgical Options

- Percutaneous retrogasserian glycerol rhizolysis (PRGR)
- Percutaneous radiofrequency trigeminal gangliolysis (PRTG)
- Percutaneous trigeminal ganglion compression (PTGC)
- Stereotactic radiosurgery

Trigeminal Neuralgia: Major Surgical Options

- Microvascular decompression
  - Only procedure that addresses the presumed etiology
  - Provides the longest lasting relief
  - Mortality rate 1%
  - Morbidity:
    - Hearing loss 1%
    - CSF leakage 1-2%
    - Transient (or permanent) CN damage very rare

13.1.2 Symptomatic trigeminal neuralgia

As 13.1.1 *except*:

A. Paroxysmal attacks of pain lasting from a fraction of 1 sec to 2 min, with or without persistence of aching between paroxysms, affecting one or more divisions of the trigeminal nerve and fulfilling criteria B and C

D. (*replacing criteria D and E*)
A causative lesion, other than vascular compression, has been demonstrated by special investigations and/or posterior fossa exploration

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Glossopharyngeal Neuralgia

- Rare syndrome
- Usually occurs 6th decade
- Pain occurs in areas supplied by somatosensory branches of glossopharyngeal and vagus nerves
- May be caused by vascular cross-compression
- May occur together with TN

Glossopharyngeal Neuralgia: Clinical Features

- Paroxysmal pain in tonsil, tongue and larynx
- Pain radiates from oropharynx backwards to ear
- Abrupt onset pain persists for 1 minute and abruptly ends ("square wave" pain)
- Dull, deep continuous pain
- Paroxysms recur 5-30 times/day
- Nocturnal attacks
- Associated coughing, hoarseness, syncope

Glossopharyngeal Neuralgia: Clinical Features

- Triggers:swallowing, esp. cold liquids, chewing, clearing throat, coughing talking, rapid head movements, yawning, jaw movement, elevating ipsilateral arm
- Bouts occur in clusters lasting weeks to months
- Remission periods of months to years

Glossopharyngeal Neuralgia: Treatment

- Carbamazepine
- Exploration and MVD of IX and X
- If no cross-compression demonstrated, division of entire IX and upper half of X
- Thermocoagulation of petrous ganglion also effective

Superior Laryngeal Neuralgia

- Branch of vagus nerve
- Innervates cricothyroid muscle of larynx
- Cricothyroid muscle stretches, tenses, and adducts vocal cord
- Unilateral paralysis causes hoarse, weak, fatigued voice
- SLN may be damaged during carotid endarterectomy

Superior Laryngeal Neuralgia

- Middle-aged men
- Paroxysmal unilateral submandibular pain
- Radiation into ear, eye, or shoulder
- Pain paroxysms last seconds to minutes
- Precipitated by swallowing, straining voice, head turning, coughing, sneezing, yawning or blowing nose
- Pain accompanied by irresistible urge to swallow (which exacerbates pain)
- Impossible to speak

Superior Laryngeal Neuralgia: Treatment

- Local blockade of superior laryngeal nerve is diagnostic
- Neurectomy effectively cures condition

Tolosa-Hunt Syndrome:

- Rare syndrome
- Due to granulomatous inflammation of cavernous sinus, superior orbital fissure or orbit
- One or more episodes of unilateral orbital pain with paralysis of III,IV,VI
- Lasts 8 weeks untreated
- Relief within 72 hrs with steroids
- Rule out aneurysm, DM, paranasal mucocele, parasellar tumor, CC fisula, sphenoid sinusitis

Recommended Readings:

- ICHD-II
- Wolff's Headache
- Headache in Clinical Practice
- The Headaches

13.17 Ophthalmoplegic 'migraine'

A. At least 2 attacks fulfilling criterion B

B. Migraine-like headache accompanied or followed within 4 d of its onset by paresis of ≥1 of the third, fourth and/or sixth cranial nerves

C. Parasellar, orbital fissure and posterior fossa lesions ruled out by appropriate investigations

13.17 Ophthalmoplegic 'migraine'
Reclassification 1988-2004

- 13.17 *Ophthalmoplegic* '*migraine*' was previously classified as 1.3 *Ophthalmoplegic migraine*
- It is unlikely to be a variant of migraine since the headache often lasts for ≥1 wk and there is a latent period of up to 4 d from headache onset to ophthalmoplegia
- 13.17 *Ophthalmoplegic* '*migraine*' may be a recurrent demyelinating neuropathy
- It is very rare