Headache Disorders

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Primary Headache Disorders

- Tension Type headache
- Migraine headache
- Trigeminal / Autonomic headaches
- Other Primary headache disorders

- Question: 43y/o male with diffuse head pain. Seems to occur toward the end of a busy work day. It does not necessitate him leaving work early, however. Has had similar headaches occasionally in the past. There is no associated photophobia or phonophobia. The headache is not clearly positional. He would most likely find acute symptomatic relief with:
- A. Oxygen
- B. Botulinum toxin injection
- C. Ibuprofen
- D. Topiramate
- E. Diltiazem

Tension Type Headache

- Episodic vs Chronic (≥15d/mos)
  - Episodic Infrequent (<1/mos) vs Frequent (1-14d/mos)
- Most Common 1˚ headache disorder
  - Bilateral, non-throbbing, mild-moderate severity ("dull", "pressure", "tight cap")
  - NOT aggravated by physical activity
  - NO photo/phonophobia
  - NO N/V

Tension Type Headache *cont'd*

- Precipitated by: stress and mental tension
  - Fatigue
  - Anxiety
  - Social
  - Economic
- Treat with:
  - NSAIDS (Ibuprofen 200-800mg, Naproxen 220-550mg, Diclofenac 25-100mg)
  - Aspirin 650-1000mg
  - Acetaminophen 1000mg

Tension Type Headache Txt *cont'd*

- Combination therapy:
  - ASA 500mg + Acetaminophen 500mg + Caffeine 130mg
  - NSAID + Caffeine 65-130mg +/- Acetaminophen
- Butalbital + APAP + caffeine – use only when NSAIDS or simple analgesics with caffeine are Ineffective
  - Butalbital + ASA in hepatic failure
- Biofeedback/Relaxation Techniques/Massage
- Osteopathic manual manipulation (ie. Suboccipital release)

Chronic Tension Type Headache

- ≥15 days/month (3+months)
- Peak in 40s
- F>M
- Assoc. with ↑ incidence comorbid psych disorders
- Tx: Antidepressants: Amitriptyline, Mirtazapine, Venlafaxine
- Topiramate -25mg qhs, titrate 25mg/wk
- Valproic Acid 250-1000mg
- Tizanidine (alpha2 agonist) – 2-24mg/day (TID)

Tension Type: ICHD Criteria

- HA lasting 30min – 7 days
- 2 of the 4
  - Pressing or tightening (non-pulsatile)
  - Mild/moderate (does not prohibit activity)
  - Bilateral location
  - Not aggravated by routine activity (walking, climbing stairs)
- 2 of 2
  - No N/V
  - No photophobia and phonophobia (one may be present)

Migraine Headache Disorders

ICHD Migraine Subtypes

I. Migraine
- 1.1 Migraine without aura
- 1.2 Migraine with aura
  - 1.2.1 Migraine with typical aura
    - 1.2.1.1 Typical aura with headache
    - 1.2.1.2 Typical aura without headache
  - 1.2.2 Migraine with brainstem aura
  - 1.2.3 Hemiplegic migraine
    - 1.2.3.1 Familial Hemiplegic migraine
    - 1..2.3.2 Sporadic Hemiplegic migraine
  - 1.2.4 Retinal migraine
- 1.3 Chronic migraine

- **Question: A 40y/o male with no history of cardiovascular risk factors. He notes the onset of scintillating visual patterns of 20min of duration followed by a throbbing head pain which improves somewhat with lying down in a dark quiet room. The pain is so severe that he must leave work. Which of the medications listed would be the most reasonable choice for abortive therapy?**
- A. Hydrocodone
- B. Topiramate
- C. Aspirin
- D. Sumatriptan
- E. Acetaminophen

Case: A 24y/o Female

- A 24y/o female with hx of h/a since her late teens. They occur 1-3x/month. She notes no clear exacerbating factor. They are preceded by some subtle visual changes lasting ~30min and then progress to involve right sided throbbing which is severe. These headaches improve when she lays in a quite dark room. When she wakes the next AM, the h/a has completely resolved.

Migraine Headache

- Without aura: with aura – 5:1
- Some Family Hx (but NO mendelian inheritance)
- F>M
- 18% women, 6% men
- Onset in adolescence
  - Most <30y/o
  - Decreased severity with age
- Perimenstrual = catamenial migraines
- Dietary Triggers (tyramine, ETOH, MSG, chocolate)

Migraine HA without Aura: ICHD Criteria

- ≥5 Headaches lasting 4-72hrs (untreated or unsuccessfully treated)
- **H/A has at least 2 of the 4**
  - Unilateral – can vary side to side
  - Pulsating
  - Moderate/severe
  - Aggravated by routine activity (walking or climbing stairs)
- **During the H/A at least 1 of the 2**
  - Phonophobia and Photophobia
  - Nausea and/or vomiting

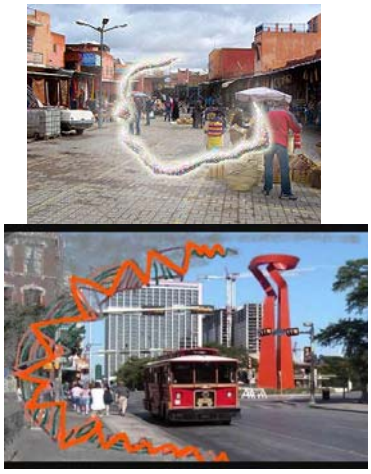
Migraine HA with Aura

- Classic / complicated migraine
- 4-72hrs
- Unilateral
  - Varies from side to side
- Throbbing/pulsating
- Aggravated with activity
- Photo/phonophobia
- Mod-severe
- Nausea +/- vomiting

Migraine HA with Aura: ICHD Criteria

- A. At least 2 attacks fulfilling B and C
- B. Aura consisting of visual, sensory and/or speech/language symptoms; each fully reversible. **NO motor, brainstem or retinal symptoms**
- C. at least 2 of the 4:
  - 1. at least 1 aura symptom spreads gradually over ≥5min and /or 2 or more sxs occur in succession
  - 2. Each aura symptom lasts 5-60min.
  - 3. At least 1 symptom is unilateral
  - 4. Aura is accompanied, or followed w/in 60min by H/A

Scintillating Scotoma and Fortification Spectra



Migraine With Aura Without Headache

- Fulfills criteria for migraine with Aura (visual, sensory, speech/language symptoms)
- No headache accompanies or follows the aura within 60minutes
- Distinction between TIA and aura requires further evaluation.

Hemiplegic Migraine

- Migraine with aura including motor weakness
- A. At least 2 attacks fulfilling following criteria B and C
- B. Aura consisting of both of the following:
  - 1. Fully reversible motor weakness
  - 2. Fully reversible visual, sensory and/or speech/language sx
- C. At least 2 of the 4:
  - At least 1 aura symptom spreads gradually over ≥5min, and/or 2 or more sxs occur in succession
  - Each non-motor aura sx lasts 5-60min, motor sxs <72hr*
  - At least 1 aura sx is unilateral
  - Aura is accompanied, or followed within 60min by h/a.

Hemiplegic Migraine (cont'd)

- **Familial**- autosomal dominant –at least one 1st or 2nd degree relative meets criteria
  - FHM1 – 50% cases, Chromosome 19
    - CACNA1A gene mutation (P/Q Ca channel) – this gene also assoc with EA2, SCA6
  - FHM2 – <25% cases, Chromosome 1
    - ATP1A2 gene mutation (Na/K ATPase)
  - FHM3 - ?prevalence, Chromosome 2
    - SCNA1 (Na channel)
  - Additional sxs: fever, seizures, prolonged weakness, coma and rarely death.
  - **Sporadic** – no 1st or 2nd degree relative with attacks

Management of Hemiplegic Migraine

- May require hospital care.
- Initial presentation requires work up for TIA
- Prevention meds: First line: Verapamil 120-360mg day, Acetazolamide 250mg bid, Lamotrigine 25-100mg.
- Second line: Topiramate, Depakote, Amitriptyline
- Abortive therapy: NSAIDS.
- Avoid Vasoconstrictors
- Limited evidence suggests that triptans do not increase risk of ischemic events when used in HM

Migraine w/ Brainstem Aura (Basilar Migraine)

- At least 2 attacks fulfilling following criteria B-D
- B. Aura consisting of fully reversible visual, sensory and/or speech/language sxs. **NO motor weakness**
- C. At least 2 of the following brainstem symptoms: Dysarthria, vertigo, tinnitus, hyperacusis, diplopia, ataxia, decreased consciousness.
- B. At least 2 of the 4:
  - 1. ≥1 aura symptom gradually develops over ≥5min and / or 2 or more aura symptoms in succession.
  - Each aura symptom lasts ≥5min and ≤60min.
  - At least 1 symptom is unilateral
  - Aura is accompanied, or followed w/in 60min by h/a

Retinal Migraine (Ocular, ophthalmic)

- At least 2 attacks fulfilling criteria A&B
- A. Fully reversible monocular positive and/or negative visual phenomena (e.g scintillations, scotoma, or blindness) confirmed by examination during an attack or (after proper instruction) by the patient's drawing of a monocular field defect after an attack.
- B. HA begins during the visual symptoms or follows them within 60 minutes
- C. Normal ophthalmological exam between attacks

Chronic Migraine

- HA (tension-type-like and/or migraine-like) on ≥15 days/month (3+ months) with ≥8 days features of migraine HA
- 3%-14% migraines transform / year
- Neurogenic inflammation→Central neurosensitization
- Increased incidence of comorbid psychiatric disorders
- Txt: topiramate, gabapentin, tizanidine, SSRI, TCAs, levetiracetam

Migraine HA Treatment

- Prophylactic
  - TCAs
  - B-Blockers (**metoprolol, Propranolol, Timolol**)
  - Ca Channel Blockers (per AAN data insufficient)
  - ACEI (lisinopril), ARB (candesartan)
  - AEDs (**Topiramate, Valproic acid**)
  - SNRI (Venlafaxine)
  - Triptans (frovatriptan for short term prophylaxis)
  - Botuljnum toxin (approved for chronic migraines >15d/mos, 4+ hrs)
  - NSAIDS (ibuprofen, ketoprofen, naproxen)
- Deemed to be ineffective:
  - Telmisartan (ARB), Lamotrigine, Oxcarbazepine, Gabapentin

Future Migraine HA treatment

- Calcitonin Gene-related peptide (CGRP) antibodies - CGRP is a therapeutic target in migraine b/c of its presumed role in mediating trigeminovascular pain transmission and the vasodilatory component of neurogenic inflammation.
- Stimulation of the trigeminal ganglion induces release of CGRP. Infusions of CGRP trigger migraine attacks in migraineurs.
- Elevated levels of CGRP are normalized in patients with migraine following administration of the serotonin 1b/1d agonist (Sumatriptan) suggesting that triptan may act to control migraine, at least in part, by blocking release of CGRP.
- At least 3 different drug companies developing CGRP Abs

Migraine HA Treatment *cont'd*

- COMPLIMETARY THERAPIES
  - OMM
  - Cefaly – TENS unit to the head to decrease incidence of migraines
- PROPHYLACTIC
  - Petasites (butterbur) 75mg bid (No longer recommended by AAN due to pyrrolizidine alkaloids = hepatotoxicity/carcinogenic, lack of regulation)
  - MIG-99 (feverfew) 50-150mg (has antiplatelet effects)
  - Magnesium oxide 400-500mg/day
  - Riboflavin 400mg/day (3 months)
- Inadequate data for : Omega 3, Hyperbaric oxygen

Migraine HA treatment *cont'd*

- Abortive
  - **Triptans** (5-HT-1 agonists)
  - **NSAIDS**
  - **ASA**
  - **Ergotamines**
  - Valproic acid infusion
  - Steroids (i.e. Medrol dose pack)
  - Fioricet (butalbital, acetaminophen, caffeine)

Triptans

- 5-HT1 Agonists --- vessel constriction
- PO, SL, nasal spray, SQ
- May repeat x 1 dose
- Pts ≥12y/o
- Contraindications
  - Hemiplegic/basilar migraine
  - Cardio/cerebro/peripheral vascular disease
  - MAOI within 2 weeks
- Side effects
  - Abnormal vision
  - Angina
  - Ischemia

Ergotamines

- 5-HT1 agonist/antagonist
  - Also effects on D2 and NE
- PO, SL, Nasal spray, IM, SQ, PR
- Max dose = 6mg/24hrs
- Pts ≥12y/o
- Contraindications: angina/CV dz, glaucoma, hepatic dz, uncontrolled HTN, cerebrovascular dz, PVD, renal impairment
- SEs: Ischemia, arrhythmia, retroperitoneal fibrosis, gangrene

Steroids

- Less data to support this!
- Decrease cerebral edema
- Decrease perivascular inflammation
- Used to break HA cycles
- PO methylprednisolone (Medrol) dose pack
- IV Solumedrol/dexamethasone
- Used for atypical migraines, used for HA due to increased ICP

Trigeminal/Autonomic Headaches

Cluster Headache

- Men 20-40y/o
- Severe
- Unilateral
- Peri-orbital
- 15-180min
- Up to 8x/day
- Ipsilateral symptoms: Conjunctival injection, lacrimation, rhinorrhea, forehead sweating, miosis (small pupil), ptosis

Cluster HA: ICHD Criteria

- A. At least 5 attacks fulfilling criteria B-D.
- B. Severe unilateral orbital, supraorbital and/or temporal pain lasting 15-180 minutes untreated.
- C. Either or both of the following:
  - **1**. At least 1 of these symptoms/signs:
  - Conjunctival injection and/or lacrimation
  - Nasal Congestion and/or Rhinorrhea
  - Forehead/facial sweating and/or flushing
  - Miosis and/or Ptosis
  - Eyelid Edema
  - Sensation of fullness in ear
  - **2**. a sense of restlessness or agitation
- Frequency: QOD to 8x/day

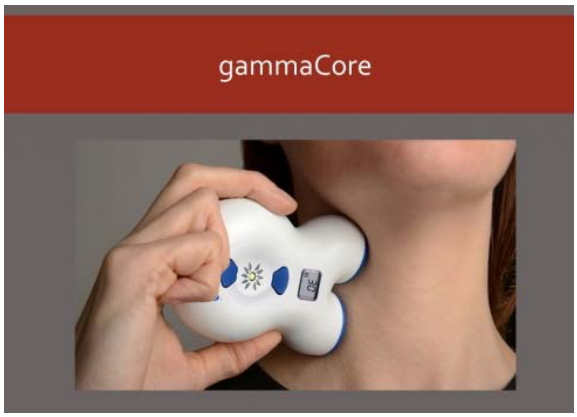
Cluster HA: ICHD Criteria *cont'd*

- Episodic – attacks occurring in bouts (cluster periods). At least 2 cluster periods lasting from 7 days to 1 year (when untreated) and separated by pain-free periods of ≥1 month.
- Chronic - attacks occurring without a remission period, or with remission lasting less than 1 month, for at least 1 year

Cluster Headache Treatment

- Abortive therapy:
  - Oxygen (12-15L/min x 15min),
  - Triptans
  - Ergotamines
  - Intranasal lidocaine (1ml) in 4%soln.
- Prevention therapy:
  - Verapamil – start 240mg/day, increase by 80mg q 10-14days, up to 960mg/day. Monitor ECG for bradycardia, heart block
  - Lithium- 300mg bid, work up to 900-1200mg/day.
    - Monitor liver, renal, thyroid fxn,ECG. Li^{2+} level 0.4-0.8mmol/L
  - Steroids- prednisone 60mg-100mg/day x 5 days, taper 10mg/day
  - Topiramate- often add on txt, start at 25mg/day work up

Vagal Nerve Stimulator for Acute Cluster H/A treatment



- First noninvasive hand held VNS applied at the neck.
- FDA approved April 2017 for acute txt of episodic cluster in Adults
- ACT1 trial data- decrease in h/a pain in 34% vs 11% placebo
- Available late Fall 2017

- **Question: A 36y/o female develops a new headache which predominantly involves her left face and neck. She develops conjunctival injection, unilateral rhinorrhea, and lacrimation on the same side as the headache. The symptoms last 30-45 min and seem to increase slightly with head turning. MRI brain is unremarkable. The first therapy to consider is:**

- A. Oxygen
- B. Hydrocodone
- C. Fentanyl patch
- D. Indomethacin
- E. C2 steroid injection

Paroxysmal Hemicrania

- Unilateral
- Face +/- neck
- Autonomic symptoms- lacrimation, conjunctival injection, rhinorrhea, nasal congestion, ptosis or facial flushing
- Minutes to 1 hour; Abrupt onset and cessation
- Remitting (similar to cluster)
  - Hemicrania continua discussed later with CDH
- F>M (different than cluster)
- May be exacerbated by head movement
- Txt w/ Indomethacin – 25mg TID x 3 days, then 50mgTID
  - Can also use Calcium channel blockers

ICHD criteria for Paroxysmal Hemicrania

- A. At least 20 attacks fulfilling B-E
- B. severe unilateral orbital, supraorbital, and or temporal pain lasting 2-30min
- C. H/A accompanied by at least 1 of the following ipsilateral to the pain:
  - 1. Conjunctival injection and or lacrimation
  - 2. Nasal congestion and/or rhinorrhea
  - 3. Eyelid edema
  - 4. Forehead and facial sweating
  - 5. Forehead and facial flushing
  - 6. Ipsilateral miosis and/or ptosis.
- D. Attack Frequency of >5/day for more than ½ the time.
- E. Attacks are prevented by therapeutic dose of Indomethacin
- F. Not better accounted for by another ICHD disorder

SUNCT

- **S**hort-lasting, **U**nilateral, **N**euralgiform headache with **C**onjunctival injection and **T**earing
- Men, >50
- Usually during day
- Seconds-minutes
- Multiple attacks per hour
- Tx with AEDs or Steroids (not Oxygen)
- Very RARE

SUNCT (cont'd)

- At least 20 attack fulfilling criteria A-C:
- A. Attacks of unilateral orbital, supra-orbital, or temporal stabbing for pulsating pain lasting 5-240 seconds
- B. Pain accompanied by ipsilateral conjunctival injection and a lacrimation
- C. Attack frequency from 3-200 per day

Trigeminal / Autonomic Headaches

	Cluster	SUNCT	Paroxysmal Hemicrania
Sex	Male	Male	Female
Age	20-40	>50	10-50
Location	Periorbital	Periorbital	Face/neck
Duration	15-180 minutes	Seconds-minutes	Minutes-1 hour
Treatment	Oxygen	AED/Steroid	Indomethacin

2° Causes of Trigeminal/Autonomic HA

- Associated cranial lesions have been reported in pts with clinical attacks that resemble cluster, causal relationship uncertain.
  - Intracranial large artery aneurysm
  - Meningioma
  - Brain arteriovenous malformation
  - Pituitary macroadenoma
  - Nasopharyngeal carcinoma
  - Metallic foreign body in maxillary sinus
  - Aspergilloma in sphenoid sinus
  - Benign posterior fossa tumor
  - Cavernous hemangioma
- In these cases the h/a improved after txt of the lesion
- Trigeminal autonomic cephalgias due to structural lesions: a review of 31 cases. Favier I, van Vilet JA, Roon KI, Witteveen RJ, Verschuuren JJ, Ferrari MD, Haan J. Arch Neurol 2007:64(1):25

Other Primary Headache Disorders

- **A 70y/o male with HTN and hypercholesterolemia recurrently awakes during the night with unilateral throbbing h/a that lasts about 1 hour. He notes no photo/phonophobia, no N/V, no rhinorrhea. There is no ptosis or pupillary asymmetry when he looks in the mirror. He denies any weight loss or muscle aches. There is no pain with chewing food and no tortuosity of the temporal artery is noted on exam. MRI brain w/wo is unremarkable. A reasonable treatment option would be:**
  - A. O2
  - B. Opioid analgesics
  - C. Lithium
  - D. Orthodontic surgery
  - E. Prednisone followed by temporal A. biopsy

Hypnic Headache

- Typically age >50 (avg onset 63y/o +/- 11yrs)
- Moderate, Throbbing, often same time nightly
- Awake patients from sleep (?onset during REM)
- Unilateral/bilateral
- 15-180 min
- NO: N/V, photo/phonophobia, or autonomic symptoms
- Tx with: Lithium 200-600mg at bedtime
  - Indomethacin 50mg qhs or caffeine 40-60mg tab
  - Verapamil 80-240mg, Topiramate 25mg-100mg

Cough/Exertional Headache

- Sudden onset with Valsalva
- Severe
- Short duration – seconds/minutes
- Frontal/occipital
- Bilateral/unilateral
- Abrupt onset, resolves after months/years
- ?etiology (no increase in CSF pressure)
- With weight lifting/running, sex
- Treatment: Indomethacin (25mg-150mg 30-60min before activity),
  - NSAIDs, ergots, propranolol, phenelzine (MAOI)

Chronic Daily Headache (CDH)

- Includes chronic tension, chronic migraine ("transformed migraine), new daily persistent headache (NDPH), hemicrania continua
- ~4% adults
- Headache occurs at least ½ the days of the month for ≥ 3 months.
- Assoc with major life changes (moving, divorce, etc.)
- F>M, low socioeconomic status, obesity, sleep disorders, smoking, ↑caffeine use
- No secondary etiology

New Daily Persistent Headache (NDPH)

- CDH Subtype
- Distinct and clearly remembered onset, with pain becoming continuous and unremitting w/in 24hrs
- Must be present for ≥3 months
- Rare
- Very txt refractory
- F>M
- Onset 20s-30s F, 50s M
- Must do Imaging, think about 2˚ etiologies
- Variable clinical characteristics
- Tx the dominant h/a characteristic

Hemicrania Continua

- CDH subtype (similar to paroxysmal hemicrania)
- Unilateral
- Moderate severity
- Chronic background pain with severe exacerbations + autonomic symptoms
- Peak onset 30s
- F:M 2:1
- Responds to Indomethacin 25mg-300mg QD

Secondary Headache Disorders

Medication Overuse Headache

- Only in patients with underlying HA disorder
- Suspect if prn analgesics >2-3x/week
- Central sensitization
- ~1.5% of general population
- F:M 3.5:1
- Most assoc w/ migraine (~65%)
- High risk: opioids, butalbital, ASA/APAP/Caffeine
- Mod-risk: triptans
- Lowest risk: NSAIDs

Mediation Overuse Headache (cont'd)

- Headache present on ≥15days/month
- Headache *worsens* during med overuse
- Regular overuse of >3 months of 1 or more drugs that can be taken for acute and/or symptomatic treatment of headache
  - Ergotamine, Triptan, opioids, butalbital combination >10 days/month
  - Simple analgesic or combo of analgesics >15 days/month

Management of Medication Overuse HA

- Withdrawal of offending meds
- Limit abortives to 2 days/week
- Bridging with analgesics during withdrawal
  - 2-30 days
  - Steroids, naproxen, antiemetics, dihydroergotamine, valproic acid
- Pt education
- Prophylaxis for primary h/a disorder

THE END