The Diagnosis of Primary Headache Disorders

Learning Objective

After completing this activity, the participant should be better able to:

- Evaluate steps in the diagnostic workup of the patient with headache
Presentation Goals

- Identify primary headache disorders in clinical practice
- Improve history-taking skills for headache and understand key physical examination features for patients with primary headache disorders
- Understand diagnostic criteria for diagnosis of migraine and trigeminal autonomic cephalgias

My Head Hurts

- Ms R, 28-year-old woman presenting to office with chief complaint of headache
- Pain is frequent and severe, no medication is working, starting to miss work
- Patient worried something is really wrong
  - Mother sent her articles about young women who had daily headaches who had brain tumors
  - Friend suggested she see you and ask about getting a certain type of ear piercing to fix her headaches
  - Brother sent her a text message that his friend’s brother had headaches improve with medication, but not sure of the name
  - She needs to feel better
    - She has a big work event next week
    - She is worried she may lose her job
Diagnosis of Headache Disorders

- Based on history/normal physical examination
- Details can separate headache disorders
  - Associated symptoms
  - Description of pain and location
  - Relation to events (head trauma, start of new medicine/activity/hormonal changes, major life stressors, weight gain)
  - History of prior headaches and change over time
    - People can have more than 1 headache d/o, especially with migraine

The Headache History:
Focus on Headache Story

- Start at the beginning
  - Patients naturally discuss what bothers them NOW
- Ms R
  - “I can see you are really struggling with this headache. Tell me about other times you have had headaches, so I can understand these headaches better.”
  - “I would like to know if you ever had headaches when you were a child or teenager. What were they like? This will help me better understand your headaches now.”
The Headache History: Focus on Headache Story

- Move to current headache concerns
  - “Tell me how your headaches changed over time.”
  - “Did they become more frequent or more severe? Did they change?”
  - “The headache you have now, when did it start?”

- Identify life changes/stress points through the history

History Examples: Headache Story 1

- I began having headaches around age 15
- I remember the first one when I was at school after a big test. I had a bad headache and threw up and had to leave school early. I had to sleep the rest of that day
- After that I would get a similar headache every few months. I would take ibuprofen and it would usually make me feel better
- A few months ago, I started this new job. I have been staying up late and have been very stressed. I began to get more headaches, similar to the ones I have always had, but ibuprofen was not working so well anymore. My friend gave me some of her BC Powder and it worked a bit better
- I have been using BC Powder every day and it lets me function, but the headache is still there
History Examples: Headache Story 2

- I began having headaches around age 12. It was around the time I started to get periods. I would get a headache most months with my period. The headache would be severe and would last for 2 days. I would be a little sick to my stomach, but not throw up
- My mom had these, too, so she gave me ibuprofen and it worked
- The headaches started to become more frequent in college. Then I would get them with my period and also around big tests, or if I drank too much alcohol
- One time I had to go to Student Health for a bad headache that made me throw up, and it would not get better. After that I avoided alcohol and made sure to get plenty of sleep and changed my schedule, so I would plan out my classes better
- My headaches did better until this year. They started to come on a few times a week

History Examples: Headache Story 3

- I never had a headache before
- Two months ago I was at a concert and got pushed and fell down some stairs. I hit my head on the right side. I felt OK, but a few days later I began to notice a severe headache. It has not gone away
The Headache History: Focus on Headache Symptoms

- **Description of pain**
  - Location, severity, quality of pain

- **How long does it last (untreated)?**
  - Seconds/Minutes/Hours/Days

- **How often does it occur?**
  - How many days do you NOT have a headache?

- **What improves/worsens symptoms?**
  - Rest or movement
  - Dark, quiet

- **Associated symptoms**
  - Nausea/vomiting, light/sound sensitive
  - Tearing, congestion, conjunctival injection, ptosis
  - Pacing, agitation

- **Pre symptoms**
  - Vision changes, speech changes, other neurological symptoms
  - Fatigue, yawning, irritability, food cravings, dizziness

The Headache History: Focus on Quality of Life/Goals

- **Quality-of-Life Questions**
  - HIT-6/ MIDAS
  - Missed work/school
  - Missed social events
  - Impact during attacks

- **Patient Goals and Fears**
  - What are they concerned about?
  - What are their goals for treatment?
    - Cure (goal re-set will be important here)
    - “I want to make it to Thanksgiving dinner”
    - “I want to be more reliable when making plans”
    - “I want to be able to take my children to their swim meets”
The Headache History:
Focus on Headache Risk/Comorbidity

- Family history of headaches
- Comorbid conditions
  - Sleep disorder (insomnia/apnea), mood disorder (depression/anxiety); other pain (FM/neck pain),
  - GI (IBS/gastroparesis), pulmonary (asthma/allergies),
  - CV (HTN, history of stroke), endocrinological (thyroid/DM)

CV=cardiovascular; DM=diabetes mellitus; FM=fibromyalgia;
GI=gastrointestinal; HTN=hypertension; IBS=irritable bowel syndrome.

The Headache Physical Examination

- General and Neurological Examination
  - Focus on MSK
    - Jaw, neck, shoulders
    - Nerve root tenderness, trigger points
  - Focus on fundoscopic examination and visual fields
- Typical Examination Findings in Headache Patients
  - Neck or shoulder tightness
  - Trigger points in neck or shoulder
  - Tenderness over greater or lesser occipital nerve or a branch of trigeminal nerve

MSK=musculoskeletal.
The Headache Workup

- No laboratory or imaging warranted unless certain red flags

**Headache Red Flags**
- S-ystemic symptoms (fever, weight loss)
- N-eurologic symptoms or abnormal signs (confusion, impaired alertness or consciousness, ptosis, horners)
- O-nset is sudden, abrupt, or split second
- O-lde patient aged >50 years with new onset or progressive headache (GCA)
- P-revious headache history with new or different headache (change in frequency, severity, or clinical features)
- S-econdary risk factors (HIV, cancer)

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**Primary Headache Disorders**

Courtesy of David Dodick, MD
Mayo Clinic – Scottsdale, Arizona
GCA=giant cell arteritis; HIV=human immunodeficiency virus.
Primary Headache Disorders

- 90% of all headache disorders “Benign”
- Lack of clear underlying pathology, systemic disease, or trauma
- International Classification of Headache Disorders (ICHD)
  - Classification of headache disorders
  - Grouped into primary and secondary
    - 5 categories of primary headache
  - Uses most common reported symptoms to create criteria for diagnosis


Primary Headache Disorders

- Migraine
  - Episodic and Chronic
  - With and Without Aura
  - Status Migraine
  - Menstrual Migraine
- Tension Type
  - Episodic and Chronic
- Medication Overuse
- Trigeminal Autonomic Cephalgias
  - Cluster
  - Paroxysmal Hemicrania
  - SUNCT/SUNA

- Other
  - New Daily Persistent Headache
  - Primary Stabbing
  - Primary Cough
  - Primary Exertional
  - Primary Headache associated with sexual activity
  - Primary Thunderclap
  - Hemicrania Contiu
  - Nummular Headache

SUNA=short-lasting unilateral neuralgiform headache attacks with cranial autonomic features; SUNCT=short-lasting unilateral neuralgiform headache with conjunctival injection and tearing.
Primary Headache Disorders:
Focus on Length – Long Duration

Long Duration

- Hours
  - Associated Features
    - Migraine
    - Tension Type
  - No Associated Features
- Daily
  - In-Nerve Distribution With Burning/Tingling?
  - Daily From Onset
  - Transformation of Headache

Primary Headache Disorders:
Focus on Length – Short Duration

Short Duration

- Autonomic Features
  - Continuous
    - <15 minutes
      - HC
    - >15 minutes
      - Cluster
  - Activity Related
    - SUNCT/SUMA
    - Primary Exertion
    - Primary Cough
    - Primary Sneezing
- No Autonomic Features
  - Not Activity Related
  - Sudden Onset

NDPH=new daily persistent headache.

HASA=headaches associated with sexual activity; HC=hemicrania continua; PH=paroxysmal hemicrania.
Migraine Without Aura

- At least 5 attacks
- Attacks last 4 to 72 hours untreated
- At least 2 of the following 4 characteristics:
  - Unilateral location
  - Pulsating quality
  - Moderate or severe pain intensity
  - Aggravation by or causing avoidance of routine physical activity
- During headache, at least 1 of the following:
  - Nausea and/or vomiting
  - Photophobia and phonophobia

Migraine With Aura

- At least 2 attacks
- 1 or more of the following fully reversible aura symptoms
  - Visual, sensory, speech/language, motor, brainstem, retinal
- At least 2 of the following 4 characteristics:
  - At least 1 aura symptom spreads gradually over 5 minutes, and/or 2 or more symptoms occur in succession
  - Each individual aura symptom lasts 5 to 60 minutes
  - At least 1 aura symptom is unilateral
  - Aura is accompanied, or followed within 60 minutes, by headache
- Transient ischemic attack has been excluded
Chronic Migraine

- Headache (tension-type-like and/or migraine-like) on ≥15 days per month for >3 months in a year

- Fulfiling criteria
  - Occurring in a patient who has had at least 5 attacks fulfilling criteria for migraine with or without aura
  - On at least 8 days per month for >3 months, fulfilling any of the following:
    - Migraine symptoms of nausea or vomiting or photophobia and phonophobia
    - Believed by the patient to be migraine at onset and relieved by a triptan or ergot derivative

Medication Overuse Headache

- Headaches occurring on 15+ days/month in a patient with preexisting primary headache disorder
- Regular overuse for >3 months of 1 or more drugs that can be taken for acute and/or symptomatic headache
- Not better accounted for by another ICHD-3 diagnosis
Trigeminal Autonomic Cephalgias (TACs)

- Repetitive
- Short-duration attacks
- Unilateral head pain in the distribution of the first division of the trigeminal nerve
- Accompanied by ipsilateral cranial autonomic symptoms

<table>
<thead>
<tr>
<th>Feature</th>
<th>CH</th>
<th>PHC</th>
<th>SUNCT/SUNA</th>
</tr>
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<tbody>
<tr>
<td>One-year prevalence</td>
<td>0.02%-0.1%</td>
<td>0.05%</td>
<td>0.1%</td>
</tr>
<tr>
<td>One-year incidence</td>
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<td>Unknown</td>
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<tr>
<td>Female: Male</td>
<td>1:4.3</td>
<td>1.6 to 2.4 :1</td>
<td>1.5-2:1</td>
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<td>Peak age onset</td>
<td>3rd decade</td>
<td>3rd-4th decade</td>
<td>5th decade</td>
</tr>
<tr>
<td>Episodic: Chronic</td>
<td>6-9:1</td>
<td>1:4</td>
<td>1:1-9</td>
</tr>
</tbody>
</table>

CH=cluster headache; PHC=paroxysmal hemicrania continua.
Differential Diagnosis

- Aneurysm
- Dissection
- Structural Lesion – posterior fossa or pituitary
- One of the other TACs
- Trigeminal Neuralgia
- Primary Stabbing Headache
- MRI/MRA all patients with suspected TAC

Cluster Headache

- At least 2 cluster periods lasting from 7 days to 1 year (untreated)
  - For episodic separated by pain-free periods lasting at least 3 months (for episodic), or less than 3 months (for chronic)
- At least 5 attacks fulfilling below:
  - Severe or VERY severe unilateral orbital/supraorbital and/or temporal pain, lasting 15 min to 180 min if untreated
  - 1 attack every other day up to 8 attacks per day
- Either or both of the following:
  - A sense of restlessness or agitation
  - At least 1 of the following symptoms or signs, ipsilateral to the headache
    - conjunctival injection and/or lacrimation
    - nasal congestion and/or rhinorrhea
    - eyelid edema
    - forehead and facial sweating
    - miosis and/or ptosis

MRI/MCA=magnetic resonance imaging/magnetic resonance arteriogram.

Cluster Headache Features

- Cluster “attack” – episode of cluster headache
  - Most often same time of day (late afternoon/after dinner/middle of night)
  - Most often during REM sleep (under debate)
- Cluster “cycle” or “period” – period of time patient will have attacks
  - Lasting between 2 and 12 weeks
  - Seasonal variation
  - 90% have at least 1 month remission between cycles
  - 10% without remission period (chronic cluster headache)

Paroxysmal Hemicrania

At least 20 attacks fulfilling below:

- Attacks are:
  - Severe, unilateral orbital, supraorbital, or temporal pain lasting 2 to 30 minutes
  - Headache is accompanied by either or both of following:
    - At least 1 unilateral autonomic feature:
      - conjunctival injection and/or lacrimation
      - nasal congestion and/or rhinorrhea
      - eyelid edema
      - forehead and facial sweating
      - miosis and/or ptosis
      - A sense of restlessness or agitation
  - Attacks have a frequency of >5 per day
  - Attacks prevented completely by therapeutic doses of indomethacin
  - Not better accounted for by another ICHD-3 diagnosis
Paroxysmal Hemicrania Features

- Pain is often sharp, stabbing, throbbing, shooting, burning, or boring
- Abrupt onset and cessation
- One-third of patients have interictal discomfort
- Typical headache lasts 2 minutes to 2 hours; average is 13 to 21 minutes
- Most common autonomic features are lacrimation, conjunctival injection, rhinorrhea, nasal congestion, ptosis, or facial flushing

Paroxysmal Hemicrania Features

- High frequency of attacks – 1 to 40 daily with mean frequency around 11 attacks
- Can occur any time of day, including during REM
- Attacks are unilateral with less than 5% of patients reporting side alternating attacks or bilateral attacks
- 80% have chronic form (no attack-free, or less than 1 month attack-free period in 1 year)
Hemicrania Continua

Unilateral headache fulfilling below:
- Present for 3+ months, with moderate or severe exacerbations
- Either or both:
  - At least 1 sign or symptom, ipsilateral to headache
    - conjunctival injection and/or lacrimation
    - nasal congestion and/or rhinorrhea
    - eyelid edema
    - forehead and facial sweating
    - miosis and/or ptosis
  - A sense of restless or agitation, or aggravation of the pain by movement
- Responds absolutely to therapeutic doses of indomethacin
- Not better accounted for by another ICHD-3 diagnosis


Short-lasting Unilateral Neuralgiform Headache Attacks

At least 20 attacks fulfilling criteria:
- Attacks are:
  - Moderate to severe unilateral head pain in orbital, supraorbital, temporal, and/or other trigeminal distribution
  - Lasting 1 to 600 seconds, occurring as single stabs, series of stabs, or in a saw-tooth pattern
  - Pain is accompanied by at least 1 of the following symptoms or signs, ipsilateral to the pain:
    - conjunctival injection and lacrimation
    - nasal congestion and/or rhinorrhea
    - eyelid edema
    - forehead and facial sweating
    - miosis and/or ptosis
  - Attacks occur with a frequency of at least 1 a day
- Not better accounted for by another ICHD-3 diagnosis

SUNCT
Short-lasting Unilateral Neuralgiform Headache Attacks With Conjunctival Injection and Tearing

- As prior
- Pain is accompanied by ipsilateral conjunctival injection and lacrimation

SUNCT Features

- Ultra-short attacks that are very frequent
  - Single stabs of pain with mean duration of 58 seconds
  - Groups of stabs with mean duration of 396 seconds
  - Saw-tooth attack of continuous pain with multiple superimposed stabs with mean attack duration of 1160 seconds

- SUNCT with coexistent trigeminal neuralgia
  - Such patients should receive both diagnoses
  - This differentiation is clinically difficult
SUNCT Triggers

- Touching the face or scalp
- Bathing or showering
- Washing or brushing hair
- Shaving
- Nose blowing
- Chewing or eating
- Brushing teeth
- Talking
- Coughing
- Exercise
- Light (including sunlight and fluorescent lights)

SUNA
Short-lasting Unilateral Neuralgiform Headache Attacks With Cranial Autonomic Symptoms

- Attacks as in short-lasting unilateral neuralgiform headache attacks
- Has only conjunctival injection or lacrimation or neither
Conclusions

- Diagnosis of primary headache disorders based on headache history
- Key features of history can help in differential diagnosis
  - Can also identify red flags for further workup
- Associated symptoms and length of attacks can lead you to appropriate primary headache disorder
- Use ICHD criteria to make diagnosis