Diagnosis of Primary Headache Syndromes

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Goals

- Distinguish primary from secondary headaches
- Recognize typical histories of migraine, tension-type, and cluster
- Learn the prevalence and impact of migraine
- Learn red flags for worrisome headaches
- Recognize indications for neuroimaging in suspected primary headaches
- Know when to refer patients for further evaluation
Why is Headache Important?

• Headache is part of the human experience
• Lifetime prevalence of headache:
  – 99% in women
  – 95% in men
• One half of the population experiences headaches severe enough to prevent work in a given year
• We must distinguish rare serious headaches from all the rest!
“Lord, how my head aches!
What a head have I!
It beats as it would fall in twenty pieces.”

Romeo and Juliet
Act II, scene v
William Shakespeare
Ten Most Common Symptoms in Primary Care Office Visits

Number of patients with given symptom of 410 surveyed
Source: Arch Intern Med 1990;150:1685
Headache is Common
Proportion of Outpatient Visits to Neurologists

- Migraine and Headache: 20%
- Epilepsy: 17%
- Alzheimer's Disease: 4%
- Stroke: 3%
- Parkinson's Disease: 1%
- Other: 1%

Source: IMS Audit. January 1999
There are Only Two Types of Headaches

• Old Headaches

• New Headaches
Vignette: Is This an Old or a New Headache?

• 34 year old woman with a 15 year history of throbbing unilateral headaches that are worse before menses
• Over past 6 months, has had an increasing frequency of headaches, now occurring 2-3 times per week and preventing her from working.
• Pain is unilateral and throbbing
• What if the headache was bifrontal and non-throbbing?
First Step: Distinguish Old From New Headaches

- Old headaches are usually benign
- New headaches are often benign, but must exclude serious causes
- A new headache is
  - Any headache of recent onset
  - A change in the pattern or character of a chronic headache
- A change in severity is not a new headache
- The longer a headache has been present, the more likely it is benign
Primary versus Secondary Headaches

- Primary headaches
  - Migraine
  - Tension-type headache
  - Cluster headache
- Secondary headaches
  - Medication induced headache
  - Cervicogenic headache
  - Pseudotumor cerebri
  - Chronic sinusitis
  - Benign exertional or sexual headaches
  - TMJ dysfunction
Taking a Headache History

- Pain character
- Location
- Onset
- Aura
- Duration

- Associated symptoms
- Frequency
- Evolution of headache over time
Bedside Manner

“The physician must have a certain degree of sociability, for a morose disposition is inaccessible both to those who are well and those who are sick.”

Hippocrates (460 B.C.-370 B.C.)
Physical Exam is Usually Normal

- Suggests the rare serious cause
- Reassures the patient that evaluation is thorough
  - Vital signs (fever, hypertension)
  - Sinus exam (tenderness)
  - Pupillary exam
  - Assess optic discs for papilledema
  - Neck exam (rigidity, muscle tension)
  - Screening neurologic exam to exclude focality suggestive of intracranial lesion
Papilledema
“An honest neurologist would admit that the most useful function of an ophthalmoscope is to provide a quiet time in the consultation, during which he may dwell upon the implications of the history just obtained. On a few occasions each year his ruminations will be interrupted by the appearance of gross papilledema in a patient who appears surprisingly well.”

The Lancet 1976
Migraine

- Most chronic headaches are migraine or tension-type
- Migraine is less common but results in more disability and lost work days
- A correct diagnosis minimizes unnecessary work-ups, and leads to effective treatment
- Three times as prevalent in women as in men
- Migraine is a *clinical* diagnosis
- The more historical features present, the more confident one can be in diagnosis
- Exam and imaging are normal
Life History of Migraine Patient

• Infancy
  – “Colicky” baby
  – Recurrent childhood vomiting attacks predict 3-fold increase in migraines as adult

• Childhood
  – Car sickness
  – ?migraine equivalent

• Adolescence
  – Onset of headaches usually in teens or early 20’s
  – Average age of onset 16

• Family history
  – Present in 60% of patients
Age- and Gender-Specific Prevalence of Migraine

Source: Neurology 1993;43(suppl 3);S6
Migraine Precipitants

- Caffeine withdrawal
  - Common cause of weekend headaches
- Sleep deprivation or oversleeping
- Menses
- Strong odors, perfume
- Weather change
- Foods
  - Red wine, chocolate, cheese, MSG, aspartame
- Missing a meal
- Stress
  - Headache often after stressful period
## Diagnostic Value of Headache Precipitants

<table>
<thead>
<tr>
<th>Precipitant</th>
<th>Sensitivity Migraine %</th>
<th>Sensitivity Tension %</th>
<th>LR +</th>
<th>LR -</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chocolate</td>
<td>33</td>
<td>5</td>
<td>7.1</td>
<td>0.70</td>
</tr>
<tr>
<td>Cheese</td>
<td>38</td>
<td>8</td>
<td>4.9</td>
<td>0.68</td>
</tr>
<tr>
<td>Stress</td>
<td>60</td>
<td>43</td>
<td>1.4</td>
<td>0.70</td>
</tr>
<tr>
<td>Alcohol</td>
<td>29</td>
<td>23</td>
<td>1.3</td>
<td>0.92</td>
</tr>
<tr>
<td>Menses</td>
<td>56</td>
<td>46</td>
<td>1.2</td>
<td>0.82</td>
</tr>
<tr>
<td>Hunger</td>
<td>62</td>
<td>54</td>
<td>1.1</td>
<td>0.83</td>
</tr>
<tr>
<td>Lack of sleep</td>
<td>31</td>
<td>38</td>
<td>0.83</td>
<td>1.1</td>
</tr>
</tbody>
</table>

*Arch Intern Med 2000;160;2729*
Historical Features of Migraine

- Throbbing pain
- Unilateral
- Nausea
- Photophobia
- Phonophobia
- Exacerbation by physical activity
- Duration between 4 and 72 hours
Value of Historical Features in Migraine vs. Tension-Type Headache

<table>
<thead>
<tr>
<th>Feature</th>
<th>Positive LR</th>
<th>Negative LR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea</td>
<td>19.2</td>
<td>0.20</td>
</tr>
<tr>
<td>Photophobia</td>
<td>5.8</td>
<td>0.25</td>
</tr>
<tr>
<td>Phonophobia</td>
<td>5.2</td>
<td>0.38</td>
</tr>
<tr>
<td>Exacerbation by activity</td>
<td>3.7</td>
<td>0.24</td>
</tr>
<tr>
<td>Unilateral pain</td>
<td>3.7</td>
<td>0.43</td>
</tr>
<tr>
<td>Throbbing pain</td>
<td>2.9</td>
<td>0.36</td>
</tr>
</tbody>
</table>

Arch Intern Med 2000;160:2729
Does this Patient Have a Migraine?

POUND

Is the headache
1. **P**ounding?
2. 4-72 **h**ours?
3. **U**nilateral?
4. Nausea?
5. Disabling?

<table>
<thead>
<tr>
<th>Number of Features</th>
<th>LR+ (CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>≥ 4</td>
<td>24 (1.5-388)</td>
</tr>
<tr>
<td>3</td>
<td>3.5 (1.3-9.2)</td>
</tr>
<tr>
<td>≤ 2</td>
<td>0.41 (0.32-0.52)</td>
</tr>
</tbody>
</table>

For definite or possible migraine by IHS criteria

Rational Clinical Exam Series  JAMA 2006;296:1274
Types of Migraine Headaches

• Migraine without aura (common)

• Migraine with aura (classic)

• Migraine with typical aura (complicated)
Migraine Without Aura

- Common migraine
- Two thirds of all migraineurs
- Prodrome of difficulty concentrating, irritability may last for one day before migraine
- Often, but not always, centered around eye or temple
Migraine with Aura

- Classic migraine
- Aura usually lasts 20 minutes (4-60 minutes)
- Time course is an important clue
- 85% of all aura are visual
  - Scintillating scotoma
  - Zigzags
  - Hemianopsia
  - Scotoma
  - Distortions of vision (Alice in Wonderland effect)
- Headache occurs as aura ending, or may not occur at all
Fortification Spectra

“Another time I saw a fine zigzag line running up and down and a coarse one running below it, horizontally. Later, the two ran together, end to end, and bowed out to the right. The line resembles a snake fence, or an old-style fortification with projecting angles. In some spells, the line is so brilliant one can see it easily with the eyes open…”

Migraine with Typical Aura

• Complicated migraine
• Same as classic migraine except:
• Aura also includes complex neurologic features:
  – Unilateral paresthesias
  – Unilateral weakness
  – Aphasia
  – Diplopia
• Neurologic features may outlast headache
Frequency of Aura Features Among Patients with Migraine with Aura

<table>
<thead>
<tr>
<th>Aura feature</th>
<th>Frequency %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any visual aura</td>
<td>84</td>
</tr>
<tr>
<td>• Fortification spectra</td>
<td>56</td>
</tr>
<tr>
<td>• Stars or flashes</td>
<td>83</td>
</tr>
<tr>
<td>• Scotoma</td>
<td>40</td>
</tr>
<tr>
<td>• Hemianopsia</td>
<td>7</td>
</tr>
<tr>
<td>Sensory</td>
<td>20</td>
</tr>
<tr>
<td>Aphasia</td>
<td>11</td>
</tr>
<tr>
<td>Motor</td>
<td>4</td>
</tr>
</tbody>
</table>

Arch Intern Med 2000;160:2729
Differential Diagnosis of Migraine

- These “new” throbbing headaches do not have all typical migraine features
  - AVM
    - New onset after age 30
    - May be always on same side
    - Prolonged aura
  - Temporal arteritis
    - New onset after age 50
  - Brain tumor
    - “New” headache, progressive
  - Carotidynia - Pain centered over carotid
    - Need to exclude carotid dissection
  - Carbon monoxide exposure
The Impact of Migraine

Original artwork by patients with migraine

Source: American Council for Headache Education
Cluster Headaches

- Cluster headache
  - M:F ratio 6:1
  - Average age of onset 30 years
  - Episodic (90%)
  - Chronic (10%)
    - Cluster period lasts for more than one year
Circadian Periodicity of Cluster Headaches

Source: Cephalalgia 1993;13(suppl 13):196

- 1-3 attacks daily (up to 8 attacks/day)
- Peak time periods

REM sleep
Cluster Headache: Clinical Features

- Strictly unilateral orbital / temporal
- Often always on same side for individual patient
- Severe, excruciating pain intensity
- Rapid onset (5-15 minutes)
- Shorter duration than migraine (45-90 minutes)
- Agitated, pacing, restless patient
- Occasional migrainous symptoms (nausea, photophobia)
- Autonomic features
Cluster Headache: Autonomic Features

- Conjunctival injection
- Lacrimation
- Nasal congestion
- Rhinorrhea
- Partial Horner’s syndrome
  - Ptosis
  - Miosis
- Facial flushing or sweating
## Frequency of Cluster Headache

### Clinical Features

<table>
<thead>
<tr>
<th>Feature</th>
<th>Sensitivity</th>
<th>Feature</th>
<th>Sensitivity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male sex</td>
<td>86%</td>
<td>Duration &lt;1hr</td>
<td>54%</td>
</tr>
<tr>
<td>Location:</td>
<td></td>
<td>Duration 1-2 h</td>
<td>32%</td>
</tr>
<tr>
<td>Ocular</td>
<td>80%</td>
<td>Cluster 4-8 wks</td>
<td>57%</td>
</tr>
<tr>
<td>Temporal</td>
<td>72%</td>
<td>Ipsilateral lacrimation</td>
<td>76%</td>
</tr>
<tr>
<td>Frontal</td>
<td>69%</td>
<td>Rhinorrhea</td>
<td>51%</td>
</tr>
<tr>
<td>Right sided</td>
<td>48%</td>
<td>Nausea</td>
<td>35%</td>
</tr>
<tr>
<td>Left sided</td>
<td>38%</td>
<td>Partial Horner</td>
<td>37%</td>
</tr>
</tbody>
</table>

*Arch Intern Med 2000;160:2729*
Tension-Type Headaches

- Most common type of old or primary headache
- Dull, non-throbbing
- Bilateral
- Usually temporal and occipital: “bandlike”
- No complex or focal neurologic features
- No nausea
- Lasts from 30 minutes to 1 week
- Often precipitated by emotional stress
- Anxiety or depression may be present
  - Consider if daily headaches
Differential Diagnosis of Tension-Type Headache

- Cervicogenic headache
- Occipital neuralgia
- TMJ dysfunction
- Giant cell arteritis
  - Consider in patients over age 50
Less Common Old Headaches

• Cervical degenerative arthritis
  – Common cause of chronic headache in the elderly
  – Features much like tension-type headache
  – Occipital neuralgia may be a clue

• Giant cell arteritis
  – Always consider in new or old headache after age 50
  – May be throbbing temporal, but more often non-specific
Old Headaches: Giant Cell Arteritis

- Polymyalgia rheumatica symptoms may be present
- Scalp tenderness, jaw claudication
- ESR as screening test.
- Any visual symptoms with headache in patient > 50 y.o.
- Temporal arteries may be beaded or tender, more often normal
- This is a medical emergency requiring immediate corticosteroids and elective biopsy within 5 days
# Value of Clinical Features for Temporal Arteritis

<table>
<thead>
<tr>
<th>Feature</th>
<th>Sensitivity %</th>
<th>LR+</th>
<th>LR-</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diplopia</td>
<td>9</td>
<td>3.4</td>
<td>0.95</td>
</tr>
<tr>
<td>Temporal h/a</td>
<td>52</td>
<td>4.5</td>
<td>0.82</td>
</tr>
<tr>
<td>Jaw claudication</td>
<td>34</td>
<td>4.2</td>
<td>0.72</td>
</tr>
<tr>
<td>PMR</td>
<td>34</td>
<td>0.97</td>
<td>0.99</td>
</tr>
<tr>
<td>Abnormal fundus</td>
<td>31</td>
<td>1.1</td>
<td>1.0</td>
</tr>
<tr>
<td>Any abnormal TA</td>
<td>65</td>
<td>2.0</td>
<td>0.53</td>
</tr>
<tr>
<td>Beaded TA</td>
<td>16</td>
<td>4.6</td>
<td>0.93</td>
</tr>
</tbody>
</table>

*JAMA 2002;287:92*
New (Acute) Headaches

• Headaches which are either new or changed from a previous pattern

• Though often benign, most serious causes of headache are “new” headaches
Worrisome Headaches: Red Flags “SNOOP”

Source: American Headache Society

- **S**ystemic symptoms (fever, weight loss) or **S**econdary risk factors (HIV, h/o malignancy)
- **N**eurologic symptoms or abnormal signs (eye pain, visual loss, confusion, impaired alertness or consciousness)
- **O**nset: sudden (thunderclap)
- **O**lder: new onset and progressive headache, especially in patients > 50 y.o. (giant cell arteritis)
- **P**revious headache history: first or worst headache or different (change in attack severity or clinical features)
When To Image?

- Change in pattern of old headache
- New migraine headache >40 y.o. (R/O AVM)
- Migraine headache side-locked, always on same side (R/O AVM)
- Focal neurologic exam
- New progressive headache in person without chronic headache history
- New persisting headache after age 50 (also ESR)
- Personality change
Abnormal Neuroimaging is Rare if Normal Neurologic Exam

<table>
<thead>
<tr>
<th>Type of headache</th>
<th>1994 Guideline</th>
<th>2000 Guideline</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>0.4%</td>
<td>0.2%</td>
</tr>
<tr>
<td>Tension-type</td>
<td></td>
<td>0%</td>
</tr>
<tr>
<td>Unspecified</td>
<td>2.0%</td>
<td>1.2%</td>
</tr>
</tbody>
</table>

## Meta-Analysis: Influence of Clinical Features on Likelihood of Abnormal Neuroimaging

<table>
<thead>
<tr>
<th>Feature</th>
<th>LR+</th>
<th>LR-</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cluster-type headache</td>
<td>11</td>
<td>0.95</td>
</tr>
<tr>
<td>Abnormal neuro exam</td>
<td>5.3</td>
<td>0.71</td>
</tr>
<tr>
<td>“Undefined” headache</td>
<td>3.8</td>
<td>0.66</td>
</tr>
<tr>
<td>Aura</td>
<td>3.2</td>
<td>0.51</td>
</tr>
<tr>
<td>Focal symptoms</td>
<td>3.1</td>
<td>0.79</td>
</tr>
<tr>
<td>↑ by Valsalva or exertion</td>
<td>2.3</td>
<td>0.70</td>
</tr>
<tr>
<td>Vomiting</td>
<td>1.8</td>
<td>0.47</td>
</tr>
<tr>
<td>New headache</td>
<td>1.2</td>
<td>0.89</td>
</tr>
<tr>
<td>Migraine type headache</td>
<td>0.55</td>
<td>1.2</td>
</tr>
</tbody>
</table>

Rational Clinical Exam Series  JAMA 2006;296:1274
When To Image?

- Exertional or sexual headache
- Symptoms suggest seizure
- Patient reassurance/insistence (??)

Study selection:
  - Non-contrast CT if acute hemorrhage suspected or head trauma
  - MR in all other cases due to higher yield of abnormalities
When Should We Refer Patients?

- Suspected complicated migraine
- Chronic daily headache
- Diagnosis unclear
- Suspected giant cell arteritis
- Abnormal imaging study consistent with pathologic cause for headache
- Acute glaucoma suspected
- Acute headache with worrisome features
An expert is a man who tells you a simple thing in a confused way; in such a fashion as to make you think the confusion is your own fault.

William B. Castle
Summary

• Two types of headaches
  – New
  – Old
• Most old headaches are primary headaches and are either migraine or tension-type
• Three types of migraine
  – Migraine without aura
  – Migraine with aura
  – Migraine with typical aura
## Summary

<table>
<thead>
<tr>
<th>Most useful distinguishing features of migraine</th>
<th>Other migrainous features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea</td>
<td>Throbbing pain</td>
</tr>
<tr>
<td>Photophobia</td>
<td>Unilateral</td>
</tr>
<tr>
<td>Phonophobia</td>
<td>Exacerbation by physical activity</td>
</tr>
<tr>
<td>Aura</td>
<td>Duration 4-72 hours</td>
</tr>
</tbody>
</table>
Summary

• Diagnosis of chronic headache syndromes, and degree of certainty, rests entirely on history taking
• Always ask if time course fits the working diagnosis
• Screen for red flags for worrisome headache “SNOOP”
• Consider GCA if > 50 y.o.
• Reserve neuroimaging for atypical features, abnormal neurologic exam, and advanced age