

# Common Myths and Misconceptions in Pediatric Neurology

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# Disclosures

- None

# Objectives

- To outline 4 common complaints or reasons for referral to Neurology, common misconceptions associated with them, and how (most) child neurologists see these issues.
- To introduce or reintroduce clinical pathways for headache and other complaints.

# Misconception #1

Kids with headaches might have a brain tumor

# Two types of headaches

## Primary Headache

- Tension type HA
- Migraine
- TAC's
  - Cluster HA
  - SUNCT
- “Other” primary headaches
  - NDPH
  - Idiopathic stabbing HA
  - Hemicrania continua...

## Secondary Headache

- **Tumor/Mass**
- Vascular lesion
- Increased ICP due to other causes
- Post-ictal
- Paranasal sinus disease
- Other illnesses

# Characteristics of secondary HA

## Also known as “*red flags*”

- Any new/unexplained neurological exam abnormality
- Constant, slowly increasing headache
- Neurocutaneous syndrome
- Age less than 3 years (+/-)
- +/- Sudden onset of headache
- Acute “worst headache of life”
- +/- Headache with exertion
- +/- Headache on waking in morning or during night
- +/- Posterior location
- ++/- Presence of VP shunt





# Time Course:

## Acute

- In otherwise healthy child, usually due to viral illness
- With focal neurological signs, can be ICH
- Severe, with fever can be due to meningitis

## Acute, recurrent

- Attacks of headache separated by symptom-free intervals.
  - Migraine, Tension Type Headache
  - Partial seizures, substance abuse, Cluster HA, recurrent Trauma

## Chronic, progressive

- Most ominous, can imply increased intracranial pressure
  - Tumor, hydrocephalus, ICH, chronic meningitis, abscess, SDH

## Chronic, non progressive

- Over 4 months, >15 HA days/month, >4hrs/day

## Mixed

- Usually Acute, recurrent on top of chronic daily/ non progressive HA.

# Brain Tumor Headache

Chronic and progressive pattern

AM or nocturnal onset/occurrence

Pernicious vomiting, especially in morning

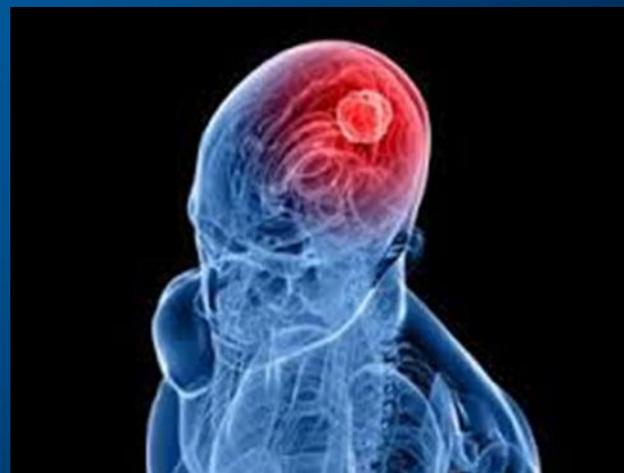
Personality change

Declining school performance

Diplopia

Head tilt

Gait changes



# Exam Findings to check

BP measurement

Sinus tenderness

TMJ clicks or pain

Neck range of motion, shoulder muscle tenderness

Mental status exam suggesting depression

**OFC measurement**

**Optic disks**

**Eye movements**

**Pronator drift**

**Ataxia**

**Abnormal DTR's**



# When to Image?

- From --AAN Guideline on Evaluation of Headache in Children and Adolescents: <http://www.neurology.org/content/59/4/490.full#ref-1>
- Data on 605 children out of 1275 who had:
  - Undergone neuroimaging
  - Been examined by a neurologist

## Found:

14 (2.3%) with nervous system lesions that required surgical treatment.

**All 14 of those children had definite abnormalities on exam.**

**No patient with a normal examination had a lesion that required surgical treatment.**

# Misconception #1

## Summary:

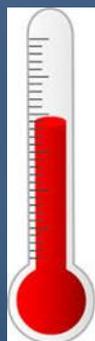
- Patients presenting with recurrent, isolated headaches who have a normal neurological exam and no red flags have an extremely low chance of having a brain tumor
- The time course is important
- The exam is important

# Misconception #2

Febrile seizures in an otherwise normal child could indicate epilepsy and require an EEG.

# Types of Febrile Seizures

## Simple Febrile Convulsions



- Generalized/ non focal
- Less than 15 minutes
- Most are much shorter
- Once in a 24 hour period

***In any of these, the fever may come well after the seizure!***

## Complex Febrile Seizures

- Focal
- Over 15 minutes
- Multiple within 24 hours

## *Febrile Status Epilepticus (FSE)*

- Over 30 minutes*

# Simple Febrile Seizures

Risk of future epilepsy is between 2-3%

That of the general pediatric population is about 1-2%

EEG is not shown to be helpful in identifying who is at higher risk for febrile seizure recurrence or future epilepsy\*.

Kuturec et al found that simple febrile seizure recurrence rate was not significantly different between patients with normal or abnormal EEGs\*\*.

\***Febrile Seizures: Guideline for the Neurodiagnostic Evaluation of the Child With a Simple Febrile Seizure:** <http://pediatrics.aappublications.org/content/127/2/389.long>

\*\***Kuturec M et al. *Febrile seizures: is the EEG a useful predictor of recurrences?* Clin Pediatr (Phila). 1997;36(1):31-36**

# Complex Febrile Seizures

More disagreement within the literature, among neurologists.

There is a higher risk for epilepsy among these patients:

*Annegers et al\* studied 687 children with FS into adulthood; 3 risk factors for developing epilepsy were identified: focal FS, prolonged FS, repeated within 24 hrs. Risk of developing epilepsy was 2.4%, 6-8%, 17-22%, and 49% among patients with zero, 1, 2, or 3 of those, respectively*

*Nat'l Collaborative Perinatal Project\*\* found children with abnormal neurological development and whose first FS was complex (for any reason) has 9.2% chance of afebrile seizures by 7 years of age, 18x higher than children w/o h/o FS.*

\*Annegers et al. Factors prognostic of unprovoked seizures after febrile convulsions. N Eng J Med. 1987 Feb 26;316(9): 493-8.

\*\*Nelson KB, Ellenberg JH. Predisposing and causative factors in childhood epilepsy. Epilepsia. 1987;28 Suppl 1: S16-24.

# Complex Febrile Seizures

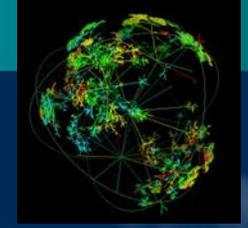
While it is tempting to recommend more testing (EEG) in patients with complex febrile seizures, the small amount of literature has not supported that it is helpful:

Maytal et al found on retrospective review of 33 patients with CFS that the rate of EEG abnormality is low and similar to the reported rate of EEG abnormality in children with simple febrile seizures (about 8.6%)\*.

**No study has been able to find any significant differences between EEG abnormalities seen in patients with simple vs complex febrile seizures.**

**No study has been able to show that EEG abnormalities in patients with CFS reliably predicts epilepsy.**

\* Maytal et al. The Value of Early Postictal EEG in Children with Complex Febrile Seizures. [Epilepsia](#). 2000 Feb;41(2):219-21



# Complex Febrile Seizures

Conclusion: We do not recommend EEG for patients with complex febrile seizures.

If an EEG is completed however, we might consider altering management if one or more of the following is present:

- a. Generalized epileptiform discharges
- b. Focal slowing
- c. Very frequent or abundant focal epileptiform discharges

HOWEVER, the EEG should never be used in place of a good developmental history and neurological exam. In other words, abnormalities on either of those is a much better guide of diagnostic workup than an EEG abnormality

# Misconception #2

## Summary:

- The risk of epilepsy among otherwise normal patients with febrile seizures is low
- Complex febrile seizures carry a higher (though still usually low) risk of future epilepsy.
- EEG is not recommended for patients with febrile seizures
- A good developmental history and examination is important.

# Misconception #3

Tics always need to be treated (with medication)

# Tics

## *Definition:*

A repeated movement or noise that is typically done frequently, is suppressible, and stereotyped. It is non-rhythmical and discrete. There is a pre-monitory urge. The movement or noise is involuntary or semi-voluntary.



# Tics

**Motor**: Eye blinking, facial grimacing, nose wrinkling, eyebrow raising, eye rolling, shoulder shrugs, head shakes or quick jerks, abdominal movements, wrist or ankle movements...

*Transient motor tics occur in 10-25% of all children*

**Vocal**: Sniffs, coughs, hums, groans, screeches, gasps, clicks, bilabial trills, screams, words or brief phrases...

*Rarely coprolalia*

# Misconception #3 and 1/2

Having both motor and vocal tics means you have Tourette syndrome  
(and must be treated with medication)

# Tourette Syndrome

Definition:

## DSM-V Diagnostic Criteria

- Onset before 18 years of Age
- Multiple Motor and 1 or More Vocal Tics (do not need to be present concurrently)
- Tics not secondary to substance or other medical condition
- Duration > 1year (may wax and wane in frequency)

**TOURETTE = (motor + vocal) x 1 year**

Tourette Syndrome occurs in 1-4% of mainstream children with a 25% incidence in children with special needs

4:1 male to female

# When to medicate?

## IF

- *Experiencing distress or impairment of function.*

## ONLY FOR

- *Tics causing impairment/distress – not ALL tics*
- *Keep in mind medications **may** negatively impact QOL also*

## DO NOT MEDICATE ONLY BECAUSE

- Tics present
- Parents concerned
- Teachers complain
- Peers tease



# Medications for tics

*Are intended to reduce the urge and reduce the tics*

***Are not expected to eliminate all tics!***

***Can take several weeks to begin having a beneficial effect***

- Guanfacine
- Clonidine
- Risperidone
- Topiramate
- Other typical or atypical antipsychotics



# Misconception #3

## Summary:

- The presence of tics in a child is quite common.
- If the tics are not bothersome or are only rarely bothersome, treatment with medication is not recommended.
- Many children have tics that they are unaware of, and hence cannot be bothered by them.
- Medication is intended to improve the QOL of the child who has the tics.

# Misconception #4

Kids do not get migraines

# Migraine prevalence

Children 5-15 yrs: 11%

Incidence rises after age of 10 yrs

Adult prevalence is about 20%

# Migraines in Kids

## Common and Classic Migraine

Without and With aura, respectively

## Variants of Migraine

Benign Paroxysmal Vertigo

Cyclic Vomiting Syndrome

Alice in Wonderland Syndrome

Benign Paroxysmal Torticollis of Infancy

Basilar Migraine



# Pediatric Migraine w/o Aura

## Diagnostic Criteria

- A. At least 5 attacks fulfilling criteria B through D.
- B. Headache lasts 1-72 hours
- C. Has at least 2 of the following:
  - Unilateral location OR bilateral frontotemporal (not occipital)
  - Pulsating quality
  - Moderate or severe pain
  - Aggravation by or causing avoidance of routine physical activity
  - At least one of the following present during HA:
    - Nausea/Vomiting
    - Photophobia and phonophobia (may be inferred from behavior)
- D. Not attributed to another structural or metabolic disorder.

# Pediatric Migraine with Aura

- About 10-20% of migraineurs
- Carries a slightly higher lifetime risk of stroke.
  - Even higher if estrogen-containing OCP is taken
  - Much higher if smoker



# Pediatric Migraine:

- Should have a normal inter-ictal neurological exam.
- Should be improved– if not entirely relieved –by sleep.
- Often occurs with a positive family history.
  - about 80-90 % of the time
- Often has specific triggers:
  - Irregular and/or inadequate sleep
  - Missed meals
  - Heat
  - Bright lights, loud noise
  - Poor hydration
  - STRESS!
  - Let-down periods after stress
- May be significantly improved or aborted by Acetaminophen or Ibuprofen
  - About 50% and 60% of the time, respectively, if taken promptly.



# Pediatric Migraine:

*May* have dietary triggers (worth mentioning or asking about):

- Caffeine (missed/ dependence)
- Aged cheeses
- MSG
- Aspartame/ artificial sweeteners
- Nuts
- Chocolate
- Nitrites/nitrates
- Aromatic foods (bananas, strawberries)
- Alcohol



# Pediatric Migraine - Chronic

## Chronic migraine

Headache occurring on  $\geq 15$  days per month on average for  $>3$  months ( $\geq 180$  days per year) and fulfilling criteria B-D

Headache lasts hours or may be continuous

At least 5 attacks<sup>1</sup> fulfilling criteria B-D

Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)<sup>2;3;4</sup>

Headache has at least two of the following characteristics:

- unilateral location
- pulsating quality
- moderate or severe pain intensity
- aggravation by or causing avoidance of routine physical activity (eg, walking or climbing stairs)
- During headache at least one of the following:
  - nausea and/or vomiting
  - photophobia and phonophobia
- Not attributed to another disorder
- WITH OR WITHOUT ANALGESIC OVERUSE

# Migraine treatment – 3 facets

## 1. Headache Hygiene

- Sleep
- Meals
- Hydration
- Exercise
- Caffeine
- Ergonomic/ Posture



## 2. Abortive Medication

- NSAIDs (Ibuprofen, Naproxen)
- Acetaminophen
- Triptans (Sumatriptan)

## 3. Preventative Medication

- Amitriptyline
- Periactin
- Topiramate
- Others: Valproate, Gabapentin
- Riboflavin, Magnesium

# Clinical Pathways

## Headache

Aim to have them available in EPIC as Smart-Sets

## Tics

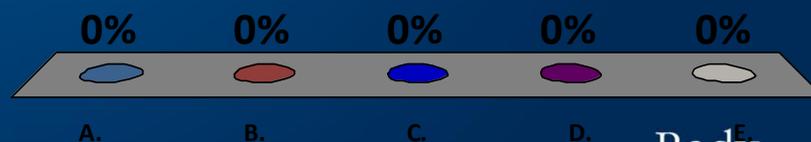
Goal is to provide pediatricians with more structured guidelines and steps that may take the place of referral to Neurology, increase quality of care.

## Febrile Seizures

## Question #1

Which of the following features is considered a “*red flag*” for children with headache?

- A. Presence of visual changes before or during headache
- B. Occurrence of photophobia and/or phonophobia
- C. Presence of neck or shoulder muscle tenderness
- D. Any new or unexplained abnormal finding on neurological exam
- E. Lack of relief of headache with OTC analgesics



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4. **Any new or unexplained abnormal finding on neurological exam**
5. Lack of relief of headache with OTC analgesics

## Question #2

Which of the following is/are true regarding children with tics and Tourette syndrome?

- A. Most are boys
- B. Tics are common
- C. Presence of multiple motor and 1 or more vocal tics x 1 yr = Tourette syndrome
- D. All of the above
- E. Only A and C



## Question #2

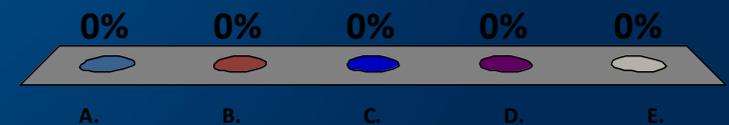
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## Question #3

Which of the following is/are true regarding febrile seizures?

- A. Patients who have simple febrile seizures should have an EEG to see if they have epilepsy
- B. Patients who have recurrent febrile seizures of any type are at a very high risk of having epilepsy
- C. Patients with febrile seizures who also have abnormal neurological exams and/or delayed development are at higher risk of epilepsy
- D. Seizures should be called febrile seizures only if a temperature of 100.4 degrees F or higher is noted at the time of a seizure (in the appropriate age group).
- E. A febrile seizure is called a complex febrile seizure if the fever comes after the seizure



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# Thank You!

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