

# Primary Headache – Classification, Pathophysiology, and Basics of Lifestyle Modification

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### **Disclosures:**

I have no outside funding sources to claim.

## Objectives

- Define characteristics of primary headache
- Identify indications for ordering neuroimaging tests in headache patients
- Discuss management of primary headache disorders
- Discuss recommendations for referral to the Headache Program at Children's National



## Headache is Common

### Migraine without aura – 20-28% of adolescents

- ICHD-II definition
- Neurol Clin. 2009; 27: 481-501

### Chronic daily headache - 2-4% of adolescents

- Defined as >15 headaches per month > 3months
- Arch Pediatr. 2008 Dec;15(12):1805-14

### Chronic migraine - 1% of adolescents

- Defined as >15 migraines per month > 3months
- Headache. 2011 May;51(5):693-706.

### Medication overuse headache - 1.75% of adolescents

- Defined as >15 headaches per month in setting of abortive medication use > 15 days per month > 3months
- Headache. 2011 May;51(5):693-706.



## Primary or Secondary?



## Primary or Secondary Headache?

### Primary Headache

- No other causative disorder
- Migraine headache
- Tension headache
- Cluster headache

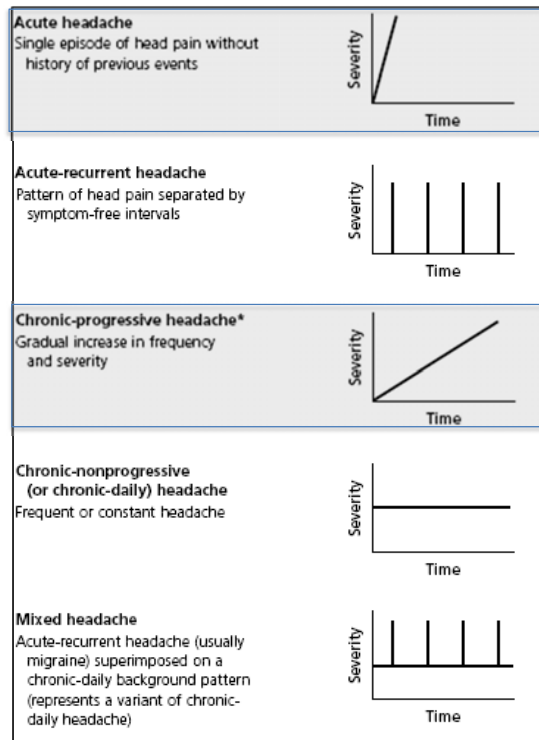
### Secondary Headache

- Caused by another disorder
- Medication overuse headache
- Hydrocephalus
- Brain tumor
- Hemorrhage
- Meningitis



# Temporal Profile of Primary and Secondary Headaches

Rothner AD. The evaluation of headaches in children and adolescents. *Semin Pediatr Neurol* 1995;2:109-118.



\*Possible Secondary Headache -Tumor, Hemorrhage



## Pediatric Headache: Consult and Referral Guidelines

### Provider's initial evaluation may include:

- Asking about common symptoms seen in primary headaches
  - Tension headaches are diffuse, non-throbbing, mild-to-moderate severity headaches without significant worsening with activity, light or sound sensitivity, or nausea
  - Migraine headaches are bifrontal or unilateral moderate-to-severe intensity headaches associated with a throbbing quality, worsening with activity, associated with light and/or sound sensitivity, nausea and/or vomiting
  - Migraine aura may occur before or during headaches lasting 5-60 minutes and include sensations of visual changes (dark or bright spots or lines), sensory changes (tingling, numbness), or speech changes
- Considering other common causes of headache
  - Sinus headache
  - Post traumatic/concussive headache
  - Allergic rhinitis
  - Ophthalmologic problems
  - Depression



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**Primary Headache Classification:  
International Classification of Headache Disorders  
3rd Edition (Beta Version)**



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**16-year-old Female with Headache**

- Frontal headache
- Throbbing quality
- "10/10"
- Needs to lay down in dark, quiet room
- Tingling left face, left arm, left leg at onset of headache for 10 min
- Occurring twice per week



## Migraine Has Two or More Major Criteria

- Bi-frontal or unilateral
- Throbbing or pounding
- Moderate to severe
- Worse with activity or relieved with rest



## Migraine Has One or More Minor Criteria

- Nausea
- Vomiting
- Photophobia
- Phonophobia



## Migraine with Aura

### Migraine With Aura

- A. At least two attacks fulfilling the criteria B-D (below)
- B. Aura consisting of at least one of the following, but no motor weakness:
1. Fully reversible visual symptoms, including positive features or negative features (eg, flickering lights, spots, or lines)
  2. Fully reversible sensory symptoms, including positive features (ie, pins and needles) or negative features (ie, numbness)
  3. Fully reversible dysphasic speech disturbances
- C. At least two of the following:
1. Homonymous visual symptoms or unilateral sensory symptoms
  2. At least one aura symptom develops gradually over  $\geq 5$  min or different aura symptoms occur in succession over  $\geq 5$  min
  3. Each symptom lasts  $\geq 5$  min and  $\leq 60$  min
- D. Not attributable to another disorder

### Table 4. Diagnostic Criteria for Basilar-type Migraine\*

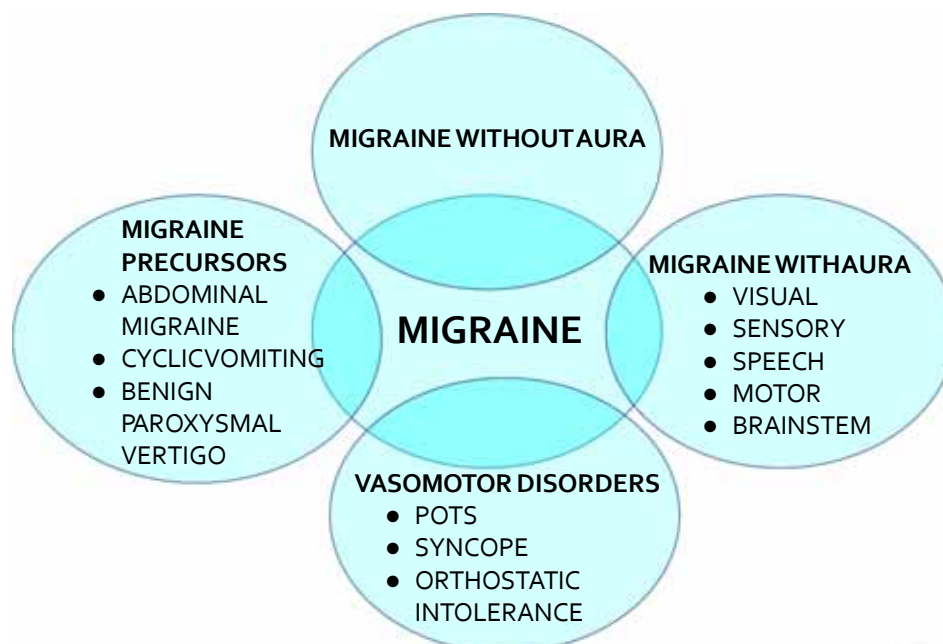
- A. Fulfills criteria for migraine with aura
- B. Accompanied by two or more of the following types of symptoms:
1. Dysarthria
  2. Vertigo
  3. Tinnitus
  4. Hypacusia
  5. Diplopia
  6. Visual phenomena in both the temporal and nasal fields of both eyes
  7. Ataxia
  8. Decreased level of consciousness
  9. Decreased hearing
  10. Double vision
  11. Simultaneous bilateral paresthesias
- C. At least one of the following:
1. At least one aura symptom develops gradually over  $\geq 5$  min or different aura symptoms occur in succession over  $\geq 5$  min
  2. Each aura symptom lasts  $> 5$  and  $\leq 60$  min
- D. Headache fulfills criteria for migraine without aura and begins during the aura or follows aura within 60 min

\*Adapted from Olesen. (4)

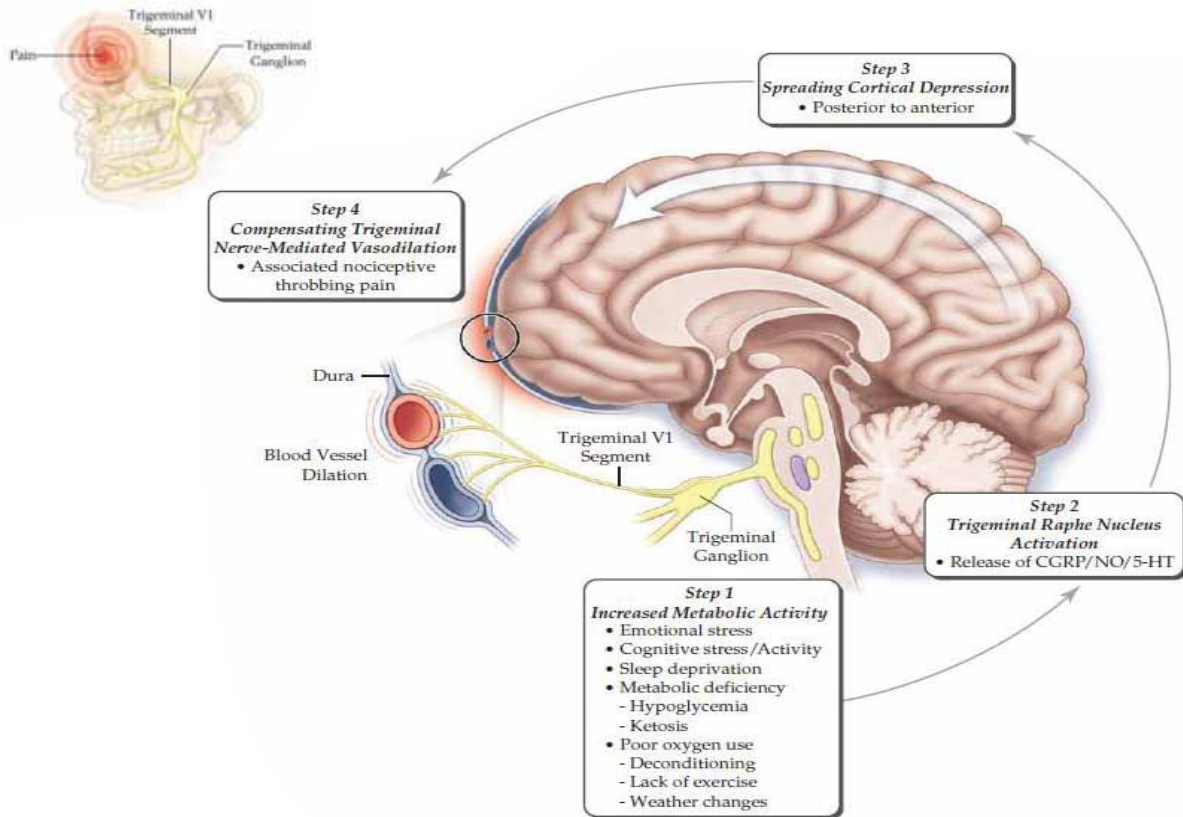
### Table 5. Diagnostic Criteria for Familial Hemiplegic Migraine\*

- A. Fulfills criteria for migraine with aura
- B. Aura consists of fully reversible motor weakness and at least one of the following:
1. Fully reversible visual symptoms, including positive features (eg, flickering lights, spots, or lines) and negative features (eg, loss of vision)
  2. Fully reversible sensory symptoms, including positive features (eg, pins and needles)
  3. Fully reversible dysphasic speech disturbance
- C. At least two of the following:
1. At least one aura symptom develops gradually over  $> 5$  min
  2. Aura symptom lasts  $> 5$  min and  $< 24$  h
  3. Headache that fulfills criteria for migraine without aura begins during the aura or follows the onset of aura within 60 min
- D. At least one first-degree or second-degree relative has had an attack
- E. At least one of the following:
1. History and physical and neurologic examination findings not suggestive of any organic disorder
  2. History or physical or neurologic examination findings suggest such a disorder, but it is ruled out by appropriate investigations

## Overlapping Migraine Disorders



## Pathophysiology of Migraine – Stress and the Trigeminovascular System



### 15-year-old Female with Headache

- Frontal and occipital headache
- "3/10"
- No change with activity
- Dull, non pulsating



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## Tension Type Headache

- Headache lasting from 30 minutes to 7 days
- Headache has at least two of the following characteristics:
  - Bilateral location
  - Pressing/tightening (non-pulsating) quality
  - Mild or moderate intensity
  - Not aggravated by routine physical activity
- Both of the following
  - No nausea or vomiting (anorexia may occur)
  - No more than one of photophobia or phonophobia



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## So Who Needs Imaging?



## Does the Patient Require Neuroimaging to Rule Out Secondary Causes of Headache?

- MRI findings in 315 children, ages 3 to 20 who had headaches
- The neurologic examinations were abnormal in 89 patients (28%).
- Thirteen (4%) had surgical space-occupying lesions
  - All had abnormal exams

Medina S, Pinter JD, Zurakowski D, et al. Children with headache: clinical predictors of surgical space-occupying lesions and the role of neuroimaging. *Radiology* 1997;202:819-24.



## Recommendations for MRI in Headache

*Division of Child Neurology at Children's National Health System*

- Headaches for less than 6 months duration not responding to lifestyle changes and first line medications
- Headaches associated with abnormal neurologic exam findings, especially papilledema, nystagmus, gait or motor changes
- Absent family history of headache
- Headaches associated with substantial confusion or emesis
- Headaches that awaken a child from sleep repeatedly
- A family history or disorders that predispose child to central nervous system lesions such as brain tumors or cerebral aneurysms

Adapted from Medina S, Pinter JD, Zurakowski D, et al. *Radiology* 1997;202:819-24



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## Pediatric Headache: Consult and Referral Guidelines

### Provider may consider testing in patients who:

- Patients with recurrent headache and a normal neurologic exam generally do not require additional testing
- Brain imaging studies are suggested for patients who have:
  - Headaches for less than 6 months duration not responding to lifestyle changes and standard first line treatment (ibuprofen, triptans, cyproheptadine),
  - Headaches associated with abnormal neurologic exam findings, especially papilledema, nystagmus, gait or motor changes
  - Absent family history of headache
  - Headaches associated with substantial confusion or emesis
  - Headaches that awaken a child from sleep repeatedly
  - A family history or disorders that predispose child to central nervous system lesions such as brain tumors or cerebral aneurysms
- Specific testing for children with other systemic complaints including arthralgias, rash, sleep complaints

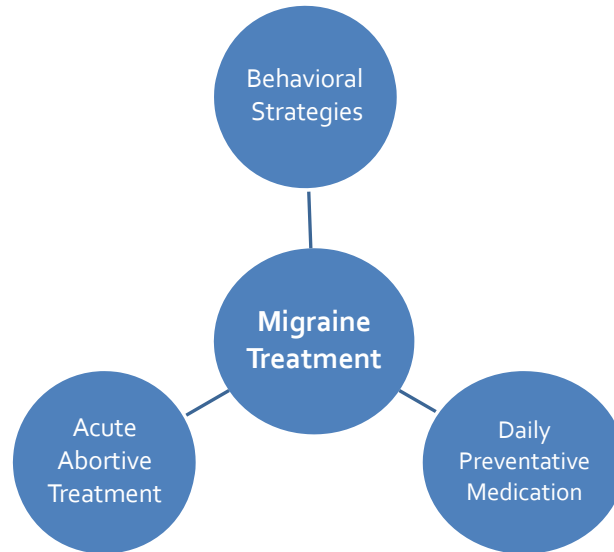


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## What Basic Treatment Should be Given for All Primary Headaches?



## Treatment Arms in Migraine



## Headache Treatment Approach

**Table 2** How to Explain Migraine and a Treatment Plan to a Patient and Family

1. Discuss the criteria for migraine and specify which are fulfilled by the child's symptoms. A standardized handout may be used for this.
2. Discuss migraine pathophysiology to explain to the child and family that increased brain needs, whether due to dehydration, sleep disruptions, poor caloric intake, barometric weather fronts, emotional stress, or increased cognitive demands, result in vasodilation to the brain, which results in activation of the V1 subdivision of the trigeminal nerve, causing head pain.
3. Discuss the treatment strategy for the patient's migraine, including separate foci on lifestyle modification, behavioral strategies, abortive therapies, and preventive therapy when indicated. A standardized handout with clear delineation of each of these areas can act as a self-administered treatment plan at home.
4. Discuss the indications for diagnostic testing and whether or not the patient fulfills these. If the patient does not require testing, discuss the historical and examination findings that are reassuring and make the likelihood of more serious central nervous system pathology unlikely.
5. Provide appropriate contact information for interim support for headaches. Consider providing a specific emergency department protocol for status migrainosus.
6. Provide specific follow-up timing.



# Comprehensive Headache Treatment Plan


  
**My Headache Treatment Plan**

**Diagnoses:** Migraine with aura    Migraine without aura    Chronic Migraine    Status Migrainosus  
 Basilar-Type Migraine    New Daily Persistent Headache    Medication Overuse Headache  
 POTS/Orthostatic Intolerance    Amplified Pain Syndrome    Post-Concussive Headache  
 Tension-Type Headache    Cluster Headache    Trigeminal Neuralgia

**Preventative Treatment – Do these every day to prevent headaches**

- Fluids – \_\_\_\_\_ ounces per day, none with caffeine or artificial sweeteners
- Exercise – 5 times a week for 30 minutes of aerobic activity (running, biking, swimming)
- Sleep – \_\_\_\_\_ hours each night, with no more than 2hrs change (do not stay up or sleep in)
- Diet – 3 healthy meals a day plus snacks if needed
- Screens – Take rest breaks with prolonged use (i.e. 30 min on, 30 min break)
- Participate – Do not avoid activities because of headache
- Distract yourself – When you have pain do something you enjoy
- Desensitize – Work through pain to teach your brain to ignore amplified pain signals
- Don't ask or talk about pain – Avoid focusing on pain and do not "check-in" about pain

Take the following medication every day to prevent headache:

Week	# Pills Left	# Pills PM
1		
2		
3		
4		

\_\_\_\_\_ mg AM \_\_\_\_\_ mg PM

\_\_\_\_\_ mg

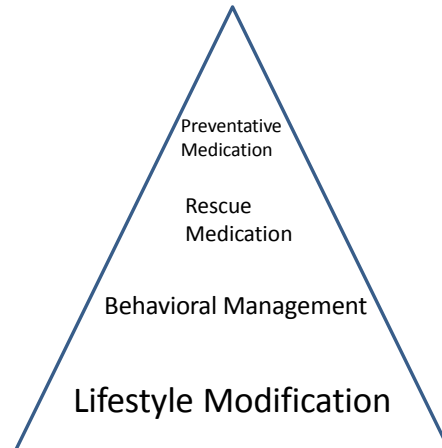
Riboflavin 300mg once daily for four consecutive months

**Acute Treatment – Do this immediately at the first sign of headache (<1 hour from onset)**

- Fluids (sports drink) \_\_\_\_\_ oz. Drink quickly every time you get a headache. Avoid O2/Propof.
- \_\_\_\_\_ mg at headache onset. Do not take more than \_\_\_\_\_ days/week.
- \_\_\_\_\_ mg
- If your child has a headache longer than 3 days and the above treatment fails, go to the nearest Emergency Department for the migraine protocol on the back of this sheet.

**Diagnostic Testing – Email your provider once completed to notify them the test was done for results**

- Neuroimaging    MRI Brain    MIV Brain    MRA Brain
- Other Testing \_\_\_\_\_



## Pediatric Headache: Consult and Referral Guidelines

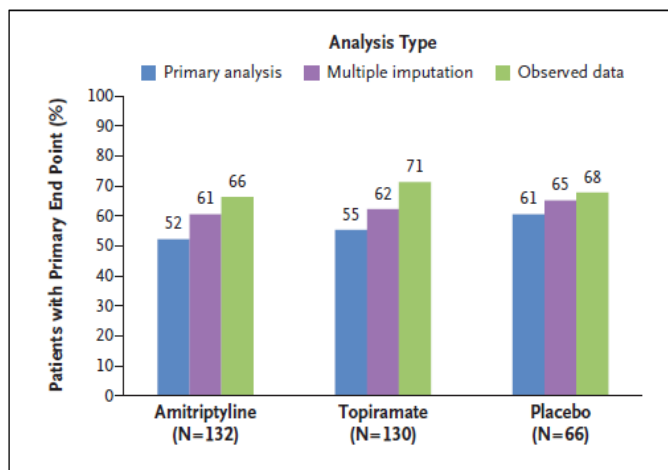
### Provider should instruct family on basic first-line treatment for headaches including:

- Lifestyle modification for prevention of headaches including:
  - Hydration – goal ounces per day = weight in pounds to a max of 100 oz. per day, none with caffeine or artificial sweeteners
  - Exercise at least five days per week for 30 minutes
  - Sleep per AAP guidelines with no more than two hours of variability in sleep or wake timing
  - Eat three healthy well balanced meals per day
- Abortive therapy when child gets a headache includes:
  - Ibuprofen 10mg/kg per dose up to three days per week
  - 8-12 oz. fluid bolus with medication, sports drinks preferable in those without contraindications (obesity, diabetes)
  - Triptans may be considered up to twice weekly if no contraindication
- Preventative therapy may be considered in those with frequent headaches and include cyproheptadine (max 0.25mg/kg/day) and amitriptyline (max 1mg/kg QHS)



# Trial of Amitriptyline, Topiramate, and Placebo for Pediatric Migraine – CHAMP

*N Engl J Med* 2017; 376:115-124

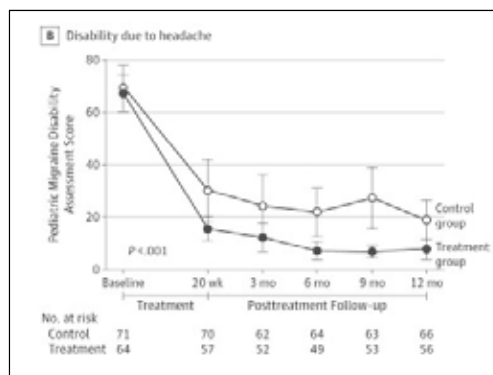
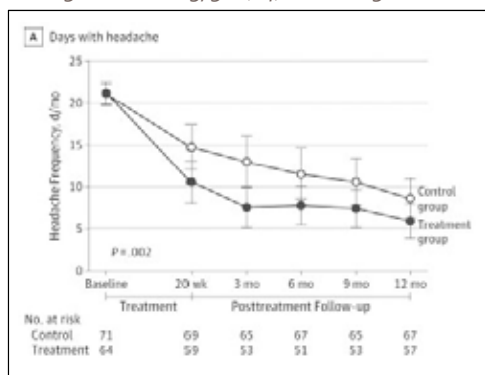


**Figure 2.** Patients with a Relative Reduction of 50% or More in the Number of Headache Days.

Shown is the percentage of patients with a relative reduction of 50% or more in the number of headache days in the comparison of the 4-week baseline period with the last 4 weeks of a 24-week trial (primary end point). Results are shown for the primary analysis and two a priori sensitivity analyses to assess the effect of missing data. Sample sizes for the trial groups represent the primary analysis population. For observed data, the population is the subgroup with observed data at week 24.

# Cognitive Behavioral Therapy Plus Amitriptyline for Chronic Migraine in Children and Adolescents: A Randomized Clinical Trial

*JAMA.* 2013 December 25; 310(24): 2622–2630



- At the 20-week end point, **days with headache** were reduced by 11.5 for the CBT plus amitriptyline group vs 6.8 for the headache education plus amitriptyline group (difference, 4.7 [95% CI, 1.7–7.7] days; *P* = .002).
- The **PedMIDAS** decreased by 52.7 points for the CBT group vs 38.6 points for the headache education group (difference, 14.1 [95% CI, 3.3–24.9] points; *P* = .01).

# Pediatric Headache: Consult and Referral Guidelines

Division of Child Neurology at Children's National

Providers may consider initiating referral to child neurology when:	Providers may instruct families to bring the following to the evaluation:
<ul style="list-style-type: none"> <li>•Patients with a new severe headache of acute onset, headache with focal neurologic deficit, or papilledema should be referred to the Emergency Department for neuroimaging</li> <li>•Recurrent headache that has been present for at least six months and is not responding to standard medical treatment including lifestyle modification and acute abortive treatment</li> <li>•Headache that is resulting in missed school days or worsening of school participation (declining grades, extracurricular activity limitation)</li> </ul>	<ul style="list-style-type: none"> <li>•A headache calendar for at least one month including dates of headaches, location, severity, associated symptoms, time at onset and resolution, activities preceding headaches including diet, and treatment provided</li> <li>•A complete list of medications used for headache treatment including doses and frequency of use. Include any abortive or preventative medications used.</li> <li>•Copies of testing done including other referrals, labs, imaging films/CDs (not just reports), and any other additional information that may be helpful.</li> </ul>




# Pediatric Urgent Headache Program

Division of Child Neurology at Children's National

**In an effort to improve access and ease of referral,**

Children's National is expanding its Comprehensive Headache and Concussion Programs with a centralized referral process, staffed by a program coordinator available from 8:30 am - 4:30 pm Monday through Friday.

Urgent appointments will be available at a Children's National Regional Outpatient Center or the main hospital.



A team of trusted experts in neurology and neuropsychology will provide urgent evaluations of your patients with headaches and concussions.

**HEADACHE 202-476-HEAD (4323)**

- Patients with urgent needs for headache care should call 202-476-HEAD (4323) or email [headache@childrensnational.org](mailto:headache@childrensnational.org). Symptoms for referral include:
  - Headaches that are recurrent (more than four times per month) or are increasingly severe
  - Headaches that have been present for over four weeks and are not responding to intervention.
  - Headaches resulting in missed school days
- Patients with headaches that are difficult to control will be referred by a Children's National provider to our Interdisciplinary Intractable Headache Clinic. This clinic utilizes lifestyle modification and healthy habits, abortive and preventative medications, complementary and alternative medicine, pain-focused cognitive behavioral therapy, biofeedback, nerve blocks, and infusions to provide additional treatment options for your patients.
- Emergency Department referral is recommended for patients with new onset of acute, severe headaches or new neurological deficits. For less severe or more intermittent headaches, please call the Neurology appointment line at 202-476-3611 to schedule an appointment with one of our Neurology care providers.

URGENT HEADACHE APPOINTMENTS are available within **72 HOURS.**



# Comprehensive Interdisciplinary Headache Program

## Children's National Health System

**Children's National Health System  
Headache and Concussion Referral Sheet**

**Comprehensive Headache Program**  
**202-476-HEAD (4323), Fax 410-266-0943**





Patients with urgent needs for headache care should call 202-476-HEAD (4323) or email [headache@childrensnational.org](mailto:headache@childrensnational.org). Symptoms for referral include:

<ul style="list-style-type: none"> <li>• Headaches that are recurrent (more than four times per month) or are increasingly severe</li> </ul>	<ul style="list-style-type: none"> <li>• Headaches that have been present for over four weeks and are not responding to intervention</li> </ul>	<ul style="list-style-type: none"> <li>• Headaches resulting in missed school days</li> </ul>
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DATE: \_\_\_\_\_

PATIENT: \_\_\_\_\_

REFERRING PHYSICIAN: \_\_\_\_\_

 <p><b>Marc Disabella, DO</b> Director, Neurology Annapolis Outpatient Center Children's National Main Campus Washington, DC</p>	 <p><b>William McConlock, MD</b> Neurology Northern Virginia Outpatient Center Children's National Main Campus Washington, DC</p>
 <p><b>Raquel Langston, MD</b> Co-Director, Neurology Montgomery County Outpatient Center Northern Virginia Outpatient Center Children's National Main Campus Washington, DC</p>	 <p><b>Angela Fletcher, PsyD</b> Director, Behavioral Pain Medicine Montgomery County Outpatient Center Northern Virginia Outpatient Center Children's National Main Campus Washington, DC</p>



# Complementary Therapies

## Children's National Health System



**Sean Alexander, MD**

- Pain Medicine
- Occipital Nerve Blocks
- Trigger Point Injections



**Chima Oluigbo, MD**

- Neurosurgery
- Occipital nerve stimulator
- Neurointerventional Procedures



**Sarah Reece, MD**

- Pain Medicine
- Acupuncture





# Pediatric Headache: Consult and Referral Guidelines

Division of Child Neurology at Children's National

Provider's initial evaluation may include:	Provider should instruct family on basic first line treatment for headaches including:	Provider may consider testing in patients who:	Providers may consider initiating referral to child neurology when:	Providers may instruct families to bring the following to the evaluation:
<p>•Asking about common symptoms seen in primary headaches:</p> <ul style="list-style-type: none"> <li>–Tension headaches are diffuse, non-throbbing, mild-to- moderate severity headaches without significant worsening with activity, light or sounds sensitivity, or nausea</li> <li>–Migraine headaches are bifrontal or unilateral moderate- to-severe intensity headaches associated with a throbbing quality, worsening with activity, and light or sound sensitivity, nausea and/or vomiting</li> <li>–Migraine aura may occur before or during headaches lasting 5-60 minutes and include sensations of visual changes (dark or bright spots or lines), sensory changes (tingling, numbness), or speech changes</li> </ul> <p>•Considering other common causes of headache:</p> <ul style="list-style-type: none"> <li>–Sinus headache</li> <li>–Post traumatic/concussive headache</li> <li>–Allergic rhinitis</li> <li>–Ophthalmologic problems</li> <li>–Depression</li> </ul>	<p>•Lifestyle modification for prevention of headaches including:</p> <ul style="list-style-type: none"> <li>–Hydration , goal ounces per day = weight in pounds to a max of 100 oz per day, none with caffeine or artificial sweeteners</li> <li>–Exercise at least three days per week for 30 minutes</li> <li>–Sleep per AAP guidelines with no more than two hours of variability in sleep or wake timing</li> <li>–Eat three healthy well-balanced meals per day</li> </ul> <p>•Abortive therapy when child gets a headache includes:</p> <ul style="list-style-type: none"> <li>–Ibuprofen 10mg/kg per dose up to three days per week</li> <li>–8-12oz fluid bolus with medication, sports drinks preferable in those without contraindications (obesity, diabetes)</li> <li>–Triptans may be considered up to twice weekly if no contraindication</li> </ul> <p>•Preventative therapy may be considered in those with frequent headaches and include cyproheptadine (max 0.25mg/kg/day) and amitriptyline (max 1mg/kg QHS)</p>	<p>•Patients with recurrent headache and a normal neurologic exam generally do not require additional testing.</p> <p>•Brain imaging studies are suggested for patients who have:</p> <ul style="list-style-type: none"> <li>–Headaches for less than 6 months duration not responding to lifestyle changes and first line treatment (ibuprofen, triptans, cyproheptadine),</li> <li>–Headaches associated with abnormal neurologic exam findings, especially papilledema, nystagmus, gait or motor changes</li> <li>–Absent family history of headache</li> <li>–Headaches associated with substantial confusion or emesis</li> <li>–Headaches that awaken a child from sleep repeatedly</li> <li>–A family history or disorders that predispose child to central nervous system lesions such as brain tumors or cerebral aneurysms</li> </ul> <p>•Specific testing for children with other systemic complaints including arthralgias, rash, sleep complaints</p>	<p>•Patients with a new severe headache of acute onset, headache with focal neurologic deficit or papilledema should be referred to the Emergency Department for neuroimaging</p> <p>•Recurrent headache that has been present for at least six months and is not responding to standard medical treatment including lifestyle modification and acute abortive treatment</p> <p>•Headache that is resulting in missed school days, worsening of school participation (declining grades, extracurricular activity limitation)</p>	<p>•A headache calendar for at least one month including dates of headaches, location, severity, associated symptoms, time at onset and resolution, activities preceding headaches including diet, and treatment provided</p> <p>•A complete list of medications used for headache treatment including doses and frequency of use. Include any abortive or preventative medications used.</p> <p>•Copies of testing done including other referrals, labs, imaging films/CDs (not just reports), and any other additional information that may be helpful.</p>



## Conclusions

- Classify headache type based on standard pain questions
- Trigeminovascular system implicated in headache disorders
- Instruct all patients on healthy habits including hydration, exercise, sleep, and diet goals
- Provide every patient a rescue plan
- Cognitive Behavioral Therapy and lifestyle modification are most effective treatment
- Daily preventative medications with questionable efficacy



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## Register Now for the 27<sup>th</sup> Annual Pediatric Neurology Update Movement Disorders

Wednesday, April 19, 2017  
7:45 am -5 pm

This year's course will be focused on the new understandings, molecular biology, and novel treatments of childhood movement disorders ranging from Tourette's syndrome to incapacitating neuro-genetic and auto-immune conditions.

We invite you to join us for presentations from renowned experts in the field in this full-day, CME accredited event.

Learn more and register now at  
[ChildrensNational.org/NeurologyUpdate](http://ChildrensNational.org/NeurologyUpdate).

