Duke University School of Medicine

Presentation of Neurologic Disease in the Pediatrician's office

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Learning Objectives

- Review common neurologic concerns presenting to the pediatrician
- Identify red flags to prompt urgent evaluation
- Outline first steps in evaluation and management

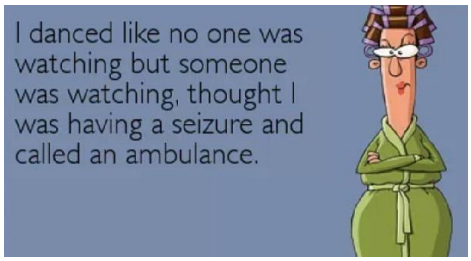
Disclosures

- There may be discussion of off-label and/or experimental medications in this presentation.
- I have no relevant financial disclosures.

Outline

- Spells
- Headaches
- Tics

Spells: Seizures vs. Everything else



Paroxysmal Non-Epileptic Events in Childhood

- Psychogenic non-epileptic spells
- Benign Neonatal Sleep Myoclonus
- Benign Non-Epileptic Myoclonus of Early Infancy
- Benign Paroxysmal Vertigo
- Shuddering Spells
- Hyperekplexia
- GERD with Sandiffer's Syndrome
- Pallid Syncope and Breath Holding Spells
- Self Stimulation or Stereotypies Hyperventilation
- Complex Tics
- Migraine/Cyclic Vomiting
- Narcolepsy/Cataplexy
- Dysomnias (Confused Arousal, Somnambulism, Night Terrors)

Seizure: Worrisome features

- Eye deviation
- Behavioral arrest
- Stiffening
- Post-event confusion
- Event duration
- Bladder or bowel incontinence

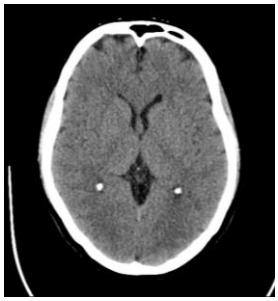
First time seizure: Evaluation

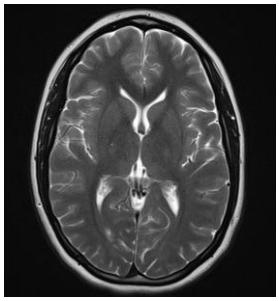
- Was it provoked?
  - Illness
  - Toxin ingestion
  - Metabolic abnormality
  - Trauma
- Labs: as indicated by history and exam
- LP: if suspicion of infection or less than 6 months
- EEG: As soon as possible

First time seizure: Imaging

- Lesions influencing treatment - 2%
- Indication for emergent imaging (CT or MRI):
  - Postictal focal deficit (Todd's paresis) not quickly resolving
  - Not returned to baseline within several hours after the seizure
- Indication for non-emergent imaging (MRI):
  - Cognitive or motor impairment of unknown etiology
  - Unexplained neuro exam abnormalities
  - Focal onset seizure with or without secondary generalization
  - Abnormal EEG that does not represent a benign partial epilepsy of childhood or primary generalized epilepsy
  - Children under 1 year of age

Head CT vs MRI





First Seizure: Recurrence Risk

- Cryptogenic - 5 Yr Recurrence 37%
  - Normal EEG, First Seizure Awake: 20%
  - Normal EEG, First Seizure in Sleep: 40%
  - Abnormal EEG, Awake: 55%
  - Abnormal EEG, Asleep: 65%
- Remote Symptomatic - 5 Yr Recurrence 66%
  - Prior Febrile Seizure: 90%
  - Age < 3 yrs 90%
- Timing of recurrences: 90% by 2 years

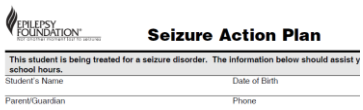
Shinnar et al. Peds, 98:216,1996.

Treatment Summary

- In general:
  - 1st seizure with normal EEG, nml exam: no
  - 1st seizure abn EEG, nml exam: maybe
  - 1st seizure abn EEG, abn exam or imaging: yes
  - 2nd seizure: yes
- All should be counseled on seizure first aid and given rescue med.

First time seizure: Counseling

- Review seizure first aid
- Seizure action plan
- Rectal diazepam or intranatal midazolam
- Limitations:
  - Driving
  - Swimming


EPILEPSY FOUNDATION

Seizure Action Plan

This student is being treated for a seizure disorder. The information below should assist y
school hours.

Student's Name | Date of Birth

Parent/Guardian | Phone

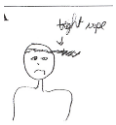
Headaches

"Headache may be equally intense, whether its implications are malignant or benign, and though there are few instances in human experience where so much pain can mean so little in terms of tissue injury, failure to separate the ominous from the trivial may cost life or create paralyzing fear."

Harold G. Wolff

Clinician's Goals

- Primary or secondary headache disorder?
- Secondary headache disorders
  - Identify, refer if needed, and treat
- Primary headache disorders:
  - Decrease pain
  - Improve Quality of Life
  - Decrease disability



Children are different…

- Younger children's reactions
  - Crying, rocking, hiding
  - Chronic-regression, depression, anxiety, behavioral difficulties
  - May affect sleeping, eating, playing
- Older child dealing w/ chronic pain
  - Manifest as developmental disorder-problems sleeping, eating, playing, school dysfunction
  - Absenteeism, problems with interaction with peers, family, authority
  - Depression, anxiety

Red Flags

- Worst
- First
- Change in pattern
- Pain that awakens patient from sleep
- Early morning headaches
- Accompanied by new neurologic or psychiatric symptoms

Secondary Headache Disorders

- Mass lesions
- Venous sinus thrombosis
- Toxic metabolic disturbance, hypothyroidism
- Post traumatic
- Idiopathic intracranial hypertension
- Chiari malformation

IHS Criteria-Pediatric Migraine

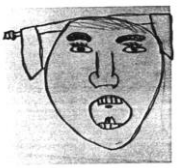
- A. At least 5 attacks fulfilling B-D
- B. HA lasting 1-48 hours **(includes sleep)**
- C. HA has 2 of the following:
  - bilateral/unilateral location
  - pulsating quality
  - mod-severe intensity
  - aggravation w/nl activity
- D. During HA, at least 1 of the following:
  - nausea and/or vomiting
  - photophobia and phonophobia

IHS Criteria - Tension-type Headache

- Fewer than 15/month
- Duration 30min-7 days
- Characteristics (2/4)
  - Pressing/tightening (Non-pulsating)
  - Mild/mod intensity
  - Bilateral
  - Not aggravated by activity
- Both of following
  - No N/V
  - No more than one of photophobia or phonophobia

History

- Headache history
- Lifestyle factors: sleep, exercise, diet, hydration
- Specifically query:
  - Concern for sleep apnea
  - Medication use
  - Substance abuse
  - Stressors

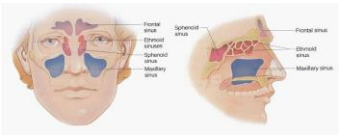


- Tools
  - Questionnaires
  - Disability Measurements-PedMIDAS, PedQL
  - Drawings
  - Calendars & Diaries

Examination

- General Examination:
  - Weight, BP (with orthostatics)
  - Fundoscopic exam
  - Tonsils
  - Palpate sinuses

- Neurological Exam



Evaluation

- Complete Headache history
- Physical (including funduscopic) and neurological exam
- Headache calendar
- Consider comorbidities (sleep apnea, orthostasis, hypothyroidism, blood pressure, depression)
- Imaging
  - Focal neurologic findings
  - Concern for secondary cause
  - Young children
- LP
  - Papilledema
  - Consider for persistent headache of unclear etiology
  - Note: MRI/MRV prior to lumbar puncture in majority

Treatment

- Accurate headache calendar
- For more than 1/week- consider prophylactic therapy
- Identify triggers
- Communication with parents and patient about realistic expectations

Preventive Medications

- Antihistamine
- Antidepressants
  - Tricyclic Antidepressants
  - SSRI's
- Antihypertensive medications
  - Beta blockers
  - Ca+ Channel blockers
- Anticonvulsants
- Vitamin supplementation (riboflavin)
- Melatonin

Preventive Pearls

- Even when they work, not guaranteed to get rid of all headaches
- All treatments likely to be approximately 50% effective
- Large placebo effect in children
- May take 4-8 weeks to notice effect
- Meds do not take place of other interventions

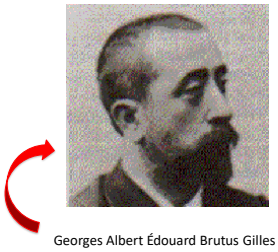
Episodic treatment

- NSAIDS
- Triptans
- Antiemetics (+/- antihistamine)
- Combination Products
- Limit to 2x per Week

Non-Pharmacologic Treatment

- Alternative or supplement to medications
  - Biofeedback
  - Acupuncture
  - Physical Therapy
  - Massage
- Disadvantages
  - Lack of insurance coverage
  - Requires a time commitment

Tic Disorders



Georges Albert Édouard Brutus Gilles de la Tourette

What is a tic?

- Involuntary, sudden, rapid, abrupt, repetitive, recurrent, and non-rhythmic movement or vocalization
- Previously described as "stereotyped"

Tic Types

Simple tics

- Single muscle or localized group
- Motor: eye blinking, head jerking, shoulder shrugging
- Vocal: grunts, barks, yelps, sniffs, throat clearing

Complex tics

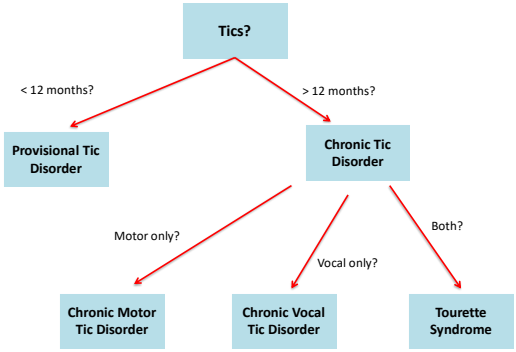
- Cluster of simple actions or more coordinated sequence
- Motor: touching, smelling, repeating observed movements (echopraxia), making obscene gestures (copropraxia)
- Vocal: repetition of words, phrases, echolalia (repeating other people's words), coprolalia (repeating obscene words)

Additional Characteristics

- Waxing/waning course
- Exacerbating factors
  - Anxiety, stress
  - Fatigue
- Premonitory urge/sensory phenomena
  - 90% 0f adults, 37% young children
  - Urge, itch, tingling, tension, feeling, or other sensation
  - Intensifies until tic is performed
  - Relieved following the completion of tic
- Supressibility & Suggestibility

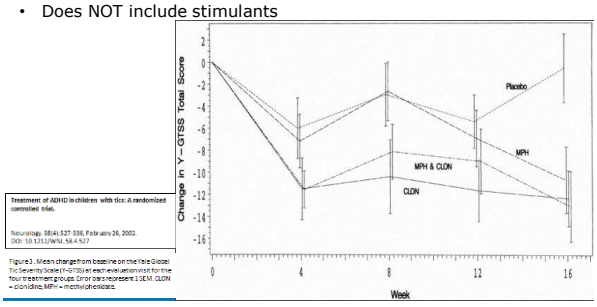
DSM-5 Tic Disorders

- Provisional Tic Disorder
- Chronic Motor or Vocal Tic Disorder
- Tourette's Disorder (Tourette Syndrome)
- Substance-Induced Tic Disorder
- Tic Disorder Due to a General Medical Condition
- Tic Disorder, Not Otherwise Specified



Substance induced tic disorder

- Motor and/or vocal tics developed during, or within 1 month of substance intoxication, withdrawal, or use
- Does NOT include stimulants



Tic disorder due to a general medical condition

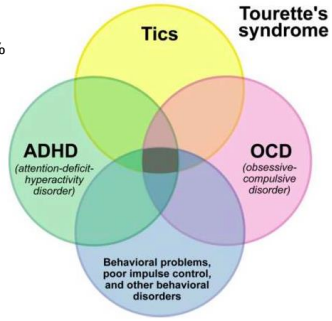
- Neurodegenerative/genetic
  - Neuroacanthocytosis
  - Huntington disease
  - NBIA
- Stroke
- Carbon monoxide exposure
- Head trauma
- Infection: Encephalitis

Epidemiology

- Tics: ~15%
- Tourette Syndrome: 0.5%
- Male:Female 3:1
- Onset 4-7 years
- Peak 11-12 years
- Outcome/prognosis:
  - 1/3 resolve by teenage/adulthood
  - 1/3 significant improvement
  - 1/3 continue to have fluctuations into adulthood

Comorbidities

- Lifetime prevalence of psychiatric comorbidity: 85%
  - ADHD 54%
  - OCD 50%
  - Anxiety 36%
  - Mood disorder 30%
  - Disruptive behavior 30%
- 57% met criteria for 2 diagnoses

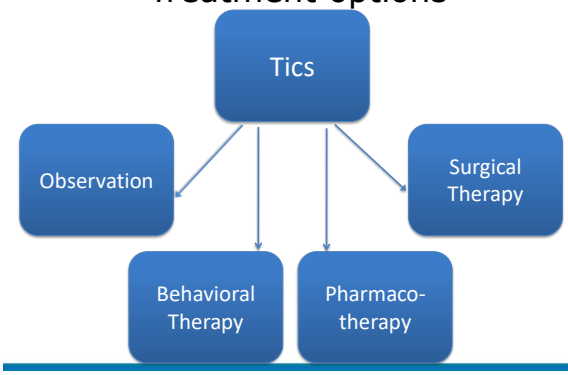


Hirschtritt et al. 2015

Auto-immune disease

Not this talk!!

Treatment options



Behavioral Treatments

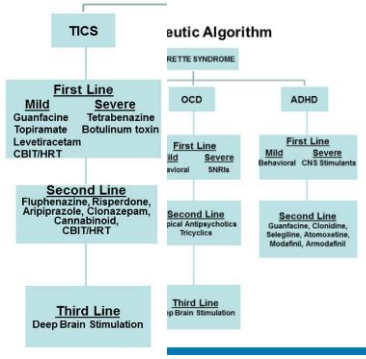
- Habit Reversal Training
  - Awareness training + competing response training
- CBIT
  - HRT + function based intervention + relaxation training

Pharmacotherapy

- α2 agonists
  - Clonidine
  - Guanfacine
- Anticonvulsants
  - Topiramate
  - Levetiracetam
  - Benzodiazepines
- Atypical neuroleptics
  - Aripiprazole*
  - Risperidone
- Typical neuroleptics
  - Fluphenazine
  - Pimozide*
  - Haloperidol*
- Others
  - Tetrabenazine
  - Botulinum toxin

* FDA approved

Treatment Algorithms



Jankovic, *Mov Disord* 2015

Summary

- Neurologic conditions most commonly present in your office.
- Children with neurologic concerns require a targeted history and exam.
- Comorbidities are common.
- When in doubt, refer!

Questions?

Acknowledgements

- Laura Tochen, MD
- Liz Rende, NNP
- Richard Chung, MD

Additional references available on request