Movers and Shakers:
Pediatric Movement Disorders

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Disclosures

Relevant Financial Relationship(s)
None

Off Label Usage
None

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Learning Objectives

- Review common and treatable hyperkinetic movement disorders in children
- Highlight tics and tic disorders, including diagnosis and treatment
- Identify when tremor and myoclonus are more concerning/pathologic
- Characterize dystonia, chorea, athetosis, and ballismus

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Tics

- Sudden brief intermittent movements or vocalizations
- Involuntary but can be voluntarily suppressed
- Premonitory urge, feeling, sensation
- Temporarily relieved after execution
- Waxes and wanes (stressors)

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Motor tics

- Simple
  - Eye blinking, facial movements, shoulder shrugging, head jerking
- Complex
  - Bizarre gait, kicking, jumping, seductive or obscene gestures (copropraxia), echopraxia

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Vocal tics

- Simple
  - Sniffing, throat clearing, grunting, moaning, barking, hollering, yelping, etc.
- Complex
  - Coprolalia <10%
  - Echolalia: repeating words said by others
  - Palilalia: repeating words or phrases rapidly

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Tics

- Transient tics of childhood
- Can evolve over time with several different tics
  - Motor or vocal tic disorder
  - Motor and vocal tic disorder
  - Tourette syndrome (TS)

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Tics

- Onset between 2 and 15 years of age
- Average age of onset 6 years
- 96% before 11 years
- Severity peaks 10-12 years
- Improvement in majority late adolescence or early adulthood
  - Rule of thirds

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Tics

- Genetics poorly understood, often familial
- Prevalence <1% population
- Males>female 4:1
- Uncommon in African American population
- Prenatal maternal smoking thought to be associated risk factor

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Tics: Common comorbidities

- ADHD 30-60%
- OCD 10-50%
- Depression 30%
- Anxiety 30%
- ODD and disruptive behaviors 30%

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Tics: Other comorbidities

- Learning disorders
- Sleep
  - Insomnia, EDS, parasomnias, bruxism
  - Motor tics can be seen in sleep
- Obesity, type 2 DM, cardiac disease
- Migraine, TTH, cervical spine d/o?

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Tics: Diagnosis

- Clinical
- Can be delayed or misdiagnosed
- Tourette syndrome
  - 2 or more motor tics
  - At least one vocal tic
  - Multiple per day, nearly daily
  - More than 1 year

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Tics: Differential

- Inherited (Huntington, Wilson)
- Infections (encephalitis, Sydenham chorea)
- Drugs (stimulants, levodopa, antipsychotics)
- Toxin (carbon monoxide)
- Head injury, stroke

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Tics: Differential

- Stereotypies
- Hyperactivity and impulsivity
- Compulsions
- Startle response
- Self-injurious behavior

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Tics: Treatment

- No cure
- Treatment rarely leads to complete cessation
- Many patients don't need more than education
- Comorbidities

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Tics: When to treat?

- Affecting academic/job performance or ADLs
- Psychosocial concerns
- Discomfort, pain, or injury

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Comprehensive Behavioral Intervention for Tics (CBIT)

- Habit reversal training (HRT)
  - Tic awareness
  - Competing response
- Relaxation training and functional intervention
- Limitations: access and coverage

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Medication: Alpha adrenergic agonists

- ADHD or behavioral symptoms
- Clonidine
  - Start .05 mg qhs, titrating to 3-4 doses per day, 0.1-0.4 mg per day
- Guanfacine: Less sedating, extended release
  - Start .5 mg qhs, titrating to 2-3 doses per day, max 4 mg per day

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Medication: Antidopaminergic drugs

- Dopamine receptor antagonists
  - Tardive dyskinesia
  - Parkinsonism, akathisia, acute dystonic reaction
  - **Risperidone**, *haloperidol*, *aripiprazole*, *pimozide*, fluphenazine

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Medication: Antidopaminergic drugs

- VMAT2 inhibitors
  - Deplete dopamine
  - Do not cause tardive dyskinesia?
  - Less risk of depression and suicidality
  - Tetrabenazine
    - Sedation, akathisia, parkinsonism

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Medication: Topiramate

- Small randomized controlled trials show promise

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Botulinum toxin injection

- Focal motor tics
  - "Whiplash" tics
- Vocal tics
  - Loud screaming

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Tics: Treatment

- CBIT first-line treatment
- Medication second-line?
- Refractory or severe
  - Botulinum toxin injection
  - Deep brain stimulation
- Duration: weaning during nonstressful time after good control for prolonged period

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PANDAS: Fact or Fiction?

- Pediatric autoimmune neuropsychiatric disorder associated with group A streptococci (PANDAS)
- No definitive evidence of antibodies affecting basal ganglia
- No evidence of improvement with antibiotics
- My treatment plan
  - Treat acute GAS infection
  - Treat OCD and tics
  - No chronic antibiotics

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Stereotypies

- Repetitive simple or complex movements, such as hand flapping, twirling, rocking
- Exacerbated by stressors
- Can be seen in kids with developmental delay (ASD) or typical development
- Toddler/preschool age
- Becomes less frequent with age

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Tremor

- Rhythmic back and forth or oscillating movement about a joint axis
- Frequency relative constant, amplitude may be variable
- May be caused or exacerbated by psychological stressors

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Tremor: Evaluation

- History and physical
  - Resting vs. postural vs. intentional
- Labs: CBC w/ smear, CMP, TSH/free T4, heavy metals screen
- MRI brain with and without contrast

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Essential Tremor

- Most common cause of tremor in childhood
- Tremor only neurologic manifestation
- Usually benign, but can impact ADLs
- Often idiopathic, inherited forms
- Tx: Occupational therapy +/- medication

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Shuddering attacks

- Infancy or early childhood
- Bursts of rapid trembling of entire body
- May see head turning, sniffing, throat clearing
- Fall to the floor if standing
- May occur during sleep
- Resolve over time

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Myoclonus

- Brief shock-like jerks
- Often clusters, typically nonrhythmic
- Physiologic or pathologic
- Seizures may need to be ruled out

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Benign Myoclonus in Sleep

- Onset first month after birth
- Can occur at any age, more common the younger you are
- Early stages of sleep
- Stimulus sensitive
- Restless leg symptoms can be similar

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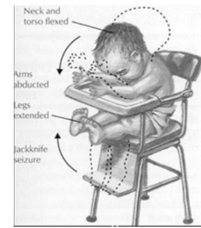
Infantile Spasms

- Onset 3-7 months, after 18 months rare
- Developmental regression
- EEG: Hypsarrhythmia
- Tx: ACTH and Vigabatrin



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Essential Myoclonus

- No associated neurologic deficit
- May be associated with essential tremor
- Autosomal dominant inheritance or sporadic
- Typically begins <20 years of age

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Myoclonus: Treatment

- Antiepileptic medications
- Immune therapy?
- Botulinum toxic injections (focal)
- Transcranial magnetic stimulation?

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Dystonia

- Sustained or intermittent muscle contraction
- Repetitive movements and/or postures
- Typically patterned and twisting
- +/- tremulousness
- Often initiated or worsened by voluntary action

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Dystonia

- Children: often begins distal and later generalizes
- Early onset can be genetic (sporadic or inherited)
  - DYT1 dystonia one of the most common
  - Dopa-responsive dystonia rare but treatable

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Dystonia

- CP most common cause of acquired dystonia
- Thyrotoxicosis
- Metabolic disorders such as Wilson disease
- Pseudodystonias: Sandifer syndrome, torticollis, and scoliosis

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Acute dystonic reaction

- Dopamine receptor-blocking drugs
  - Antipsychotics (haloperidol, chlorpromazine)
  - Antiemetics (metoclopramide, phenothiazines)
- Antidepressants (SSRIs)
- Levodopa
- Anticonvulsants
- Ergots
- Tx: Parenteral diphenhydramine 1-2 mg/kg, max 50 mg

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Chorea, Athetosis, and Ballismus

- Chorea: random appearing involuntary movements or fragments
- Athetosis: slow, continuous, involuntary writhing movements (slow chorea)
- Ballismus: large amplitude movements of limbs
  - Hemiballismus w/ contralateral STN lesion

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Physiologic chorea

- All young infants, often resolves by 8 months of age
- ADHD and distal chorea (chorea minima)

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Sydenham chorea

- Major clinical manifestation of acute rheumatic fever
- Most common form of acquired chorea in children
- 5-13 years of age
- 1-8 months after infection
  - Carditis and arthritis often present in first 21 days

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Sydenham chorea

- Distal movements of hands→generalized face and feet jerking
- Rapid, irregular, nonstereotyped
- “Piano fingers,” tongue fasciculations, “mild maid”
- Continuous while awake, improve w/ sleep

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Sydenham chorea

- Emotional lability may precede
- Regression in school performance can also be seen
- Typically improves gradually, mean duration 12-15 weeks
- Tx other than antibiotics
  - AEDs and antipsychotics
  - Corticosteroids may shorten duration

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Other forms of chorea

- Post-pump chorea
- Kernicterus much less common
- Huntington disease most common hereditary chorea in children
- Benign hereditary chorea

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Questions?

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