OVERVIEW, CLASSIFICATION, AND BASIC APPROACH TO PEDIATRIC MOVEMENT DISORDERS

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Disclosures

• No financial disclosures
• I will discuss off-label use of medications/treatments
Objectives

• Describe appearance of different movement disorders presenting during childhood
• Review some basic treatment strategies for common movement disorders
• Discuss when to refer patients to the movement disorders program
Overview of Movement Disorders

Hyperkinetic
• Tremor
• Dystonia
• Chorea
• Tics
• Myoclonus
• Athetosis
• Ballism
• Ataxia

Hypokinetic
• Parkinsonism
  – Bradykinesia
  – Rigidity

More frequent in children
Uncommon in children
Pathophysiology

- Cortico-striato-thalamo-cortical circuitry
  - Indirect pathway
  - Direct pathway
Tremor- “rhythmic back-and-forth or oscillating involuntary movement about a joint axis” (Sanger et al. Mov Disord. 2010 August 15; 25(11): 1538–1549.)

• Classification of tremor
  – Resting (parkinsonian)
  – Postural
  – Action/kinetic

• Differential diagnosis
  – Enhanced physiologic tremor
  – Essential tremor (familial tremor)
  – Medication-induced: SSRIs, valproate, lithium, albuterol, stimulants, caffeine
  – Structural lesions
  – Other neurological processes
Diagnostic studies

• **Thyroid function**, CMP, vitamin levels (B12), ceruloplasmin (Wilson’s), medication levels when applicable

• When to get an MRI?
  – Unilateral tremor
  – Other associated neurologic features besides tremor
Dystonia- “involuntary sustained or intermittent muscle contractions cause twisting and repetitive movements, abnormal postures, or both”  

(Sanger et al. Mov Disord. 2010 August 15; 25(11): 1538–1549.)

Classification by location
- Focal
- Segmental
- Multifocal
- Hemidystonia
- Generalized

Classification by type
- “old” classification
  - Primary
  - Secondary
- “new” classification
  - Isolated: genetic (DYT1, DYT6)
  - Combined
    - neurodegenerative
    - CP
    - traumatic
Dystonia treatment

- Pharmacologic
- Surgical

<table>
<thead>
<tr>
<th>Class of Medication</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anticholinergics</td>
<td>Benztpine, biperiden, ethopropazine, orphenadrine, procyclidine, trihexyphenidyl</td>
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<tr>
<td>Dopaminergics</td>
<td>Levodopa, pramipexole, ropinirole, tetrabenazine</td>
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<tr>
<td>GABAergics</td>
<td>Alprazolam, baclofen, chloridiazepoxide, clonazepam, diazepam</td>
</tr>
<tr>
<td>Muscle “relaxants”</td>
<td>Baclofen, benzodiazepines, carisoprodol, chlorzoxazone, cyclobenzaprine, metaxalone, methocarbamol, orphenadrine</td>
</tr>
<tr>
<td>Others</td>
<td>Carbamazepine, cannabidiol, cyproheptadine, gabapentin, lithium, mexiletine, nabilone, riluzole, tizanidine, zolpidem</td>
</tr>
</tbody>
</table>

Jinnah & Factor, 2015
Chorea- “ongoing random-appearing sequence of one or more discrete involuntary movements or movement fragments”

**Acquired**
- Vascular
  - Stroke
  - Moya Moya
- Immunologic
  - Lupus
  - Anti-phospholipid Ab syndrome
  - **Sydenham Chorea (post strep)**
  - Paraneoplastic (NMDA R Ab)
- Chorea gravidarum
- Metabolic
  - Hyperglycemia
  - Thyroid

**Inherited**
- Huntington Disease
- Wilson Disease
- PKAN
- Benign hereditary chorea

(Sanger et al. Mov Disord. 2010 August 15; 25(11): 1538–1549.)
Sydenham Chorea aka “St Vitus Dance”

- Post-streptococcal phenomenon
- Chorea, emotional lability, irritability, OCD, psychosis

**Evaluation**
- Labs: ASO titer, anti-DNAse B (may not be helpful, chorea can be months after presentation!)
- Cardiac work up

**Treatment**
- secondary PCN prophylaxis
- Chorea: immunotherapy, VPA, carbamazepine
Video 4
What is a tic?

• “repeated, individually recognizable, intermittent movements or movement fragments that are almost always briefly suppressible”

  (Sanger et al. Mov Disord. 2010 August 15; 25(11): 1538–1549.)

• Simple vs complex tics
• Motor (movement) vs vocal (noise-producing) tics
Additional Characteristics

- Waxing/waning course
- Exacerbating factors
  - Anxiety, stress
  - Fatigue
- Premonitory urge/sensory phenomena
  - 90% of adults, 37% young children
  - urge, itch, tingling, tension, feeling, or other sensation
  - intensifies until tic is performed
  - relieved following the completion of tic
- Suppressible
- Suggestible
Tics

Provisional Tic Disorder

Chronic Tic Disorder

Chronic Motor Tic Disorder

Chronic Vocal Tic Disorder

Tourette Syndrome

< 12 months

Motor only

> 12 months

Vocal only

Both
DSM-5 Criteria for Tourette

• Both multiple motor and one or more vocal tics have been present, not necessarily concurrently
• The tics may wax and wane in frequency but have persisted for more than 1 year since first tic onset.
• Onset is before age 18 years.
• The disturbance is not attributable to the physiological effects of a substance (e.g., cocaine) or another medical condition (e.g., Huntington’s disease, postviral encephalitis).

• Does NOT include SEVERITY or PROGNOSIS!
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
Treatment algorithm

Are tics causing problems?

Education and observation

no

yes

First Line

Behavioral therapy
CBIT

not effective or not available

Pharmacotherapy
α2-agonists
clonidine
guanfacine
anti-convulsants
topiramate

Second Line

Pharmacotherapy
Neuroleptics
Atypicals
aripiprazole*
risperidone
Typicals
pimozide*
fluphenazine
haloperidol*

Third Line +

Pharmacotherapy
tetrabenazine
chemodenervation

Surgical therapy
DBS

*FDA approved
When should a patient be seen in the movement disorder program?

• For any help in classifying and evaluating involuntary movements
• For determining if additional work-up is necessary
• If any additional education needs to be provided
• For initiation of any treatments
Referrals

• Child Neurology Movement Disorder Program  
  (202) 476-3611  
• Itochen2@childrensnational.org

Additional Resources

• www.tourette.org
• https://www.cdc.gov/ncbddd/tourette/index.html
Thanks!

**Tourette/Tic, Movement Program**
- Neurology
  - Laura Tochen
  - Bennett Lavenstein
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  - Julia Dorfman
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- Neuropsych
  - Yael Granader
- Research
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  - Mary Furda

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- Orthopedics
  - Sean Tabaie
- Neurosurgery
  - Robert Keating