Common Movement Disorders in Childhood

David Moon, MD
Section Chief – Pediatric
Movement Disorders
Disclosures

None
Objectives

- By the end of this presentation participants should:
  - Know the clinical presentation of common movement disorders in childhood
  - Know indications for referral for common movement disorders of childhood
  - Know key exam findings for common movement disorders of childhood
Movement Phenomenology

- Ataxia
- Chorea/Athetosis/Ballism
- Dystonia
- Myoclonus
- Parkinsonism – bradykinesia, rest tremor, rigidity
- Stereotopies
- Tics
- Tremor
Benign Movement Disorders of Early Childhood

- Benign Sleep Myoclonus
- Benign Myoclonus of Early Infancy
- Jitteriness
- Shuddering
- Paroxysmal Tonic Upgaze (downgaze) of Infancy
- Spasmus Nutans
- Head nodding
- Benign paroxysmal Torticollis
- Benign idiopathic dystonia of infancy
- Infantile/Toddler gratification syndrome
Benign Sleep Myoclonus

- Repetitive myoclonic jerks occurring during sleep
  - Can be focal, multifocal, unilateral or bilateral
  - Can be rhythmic or non-rhythmic
- Onset typically in 1st month of life
  - Will decrease in frequency significantly by 6mo
  - Can persist prominently into toddler age
- **Exam/History Pearls:**
  - Myoclonus ceases on awakening
- Treatment: Reassurance (typically does not require referral)
Benign Myoclonus of Infancy

- Myoclonic spasms involving flexion of trunk, neck and extremities
  - Can resemble Infantile Spasms (West Syndrome)
  - Typically occur in clusters without alteration of consciousness
  - Occur only in waking state
- Onset typically 3-9mo but can occur earlier
  - Typically self-resolve in several months (8 weeks to 1-2 years)
- Requires referral to Neurology to rule out Myoclonic Seizures and Infantile Spasms
- Treatment: Reassurance after negative work-up (EEG)
Jitteriness

- Generalized, symmetric rhythmic oscillatory movements
  - Can resemble tremor or clonus
  - Highly stimulus sensitive
- Onset in the neonatal period
  - Typically resolves by 6 months or sooner with gradual improvement over time

Exam Pearls
- Often can have prominent moro reflex, symmetric ankle clonus, mild hypertonia, mild symmetric hyperreflexia

Differential
- NAS, HIE, Electrolyte abnormalities, Hyperekplexia
- Generally a normal finding in young infants
  - Referral generally not required if improving gradually over time and no other concerns
Hyperekplexia (aka Stiff Baby Syndrome)

- Disorder of exaggerated startle response
  - Sensory or emotional stimuli can trigger stiffening
  - Can also trigger apnea
  - Often have failure to thrive to startle responses interrupting feeding
- Onset can be in infancy or early childhood
- Generally caused by mutation in glycine receptor (GLRA1)

**Exam Pearls**
- Exaggerated head retraction or blink reflex with tapping on the nose or forehead (does not habituate)
- Often have jitteriness and mild hyperreflexia or hypertonia on exam

**Requires referral to Neurology**
- Treatment: clonazepam, valproate, levetiracetam
Shuddering

- Rapid low amplitude tremor of head, shoulder and arms
  - Resembles shivering
  - Often accompanied with facial grimace
  - No alteration of consciousness
  - Episodes can occur 100’s of times a day

- Onset typically in infancy or early childhood (can be as late as 10yo)
  - Typically resolve at some point in later childhood (prior to puberty)

- Referral to Neurology is generally warranted to rule out seizure

- Treatment: reassurance, propranolol has reported benefit in some case reports
Spasmus Nutans

- Slow (~2Hz) head tremor
  - Can be “no-no” or “yes-yes”
  - Accompanied with nystagmus
- Onset Late Infancy (3-8mo)
  - Generally resolves in several months
- Must distinguish between Congenital Nystagmus
  - Can also have head tremor
  - Earlier onset: early infancy onset, neonatal onset
  - Nystagmus should be symmetric (spasmus nutans can be asymmetric)
  - Persists beyond a few months
  - Visual acuity is abnormal in 90%

Requires referral to Ophthalmology
Benign Paroxysmal Movement Disorders of Early Childhood

- **Benign Paroxysmal Torticollis**
  - Onset in 1st year of life, typically resolves by 2-3 yo (5yo at latest)
  - Considered a migraine variant
  - Differential: CN4 palsy, early onset genetic dystonia
  - **Requires referral to Neurology**
  - Treatment: diphenhydramine, topiramate

- **Paroxysmal Tonic Upgaze (or downgaze) of Infancy**
  - Repeated episodes of sustained gaze deviation without alteration of consciousness
    - Often accompanied by head tilting
    - Episodes can be brief or prolonged
  - Onset 1st of life typically (but can be as late as 7yo)
    - Typically resolves within 1-4 years
  - Differential: seizures, oculogyric crisis (dystonia), tics
  - **Requires referral to Neurology**
  - Treatment: diphenhydramine, topiramate
Movement Disorder Mimics in Early Childhood

- **Sandifer Syndrome**
  - Flexion of the neck, arching of the back, or opisthotonic posturing, associated with either gastroesophageal reflux
    - Can be associated with limb stiffening
    - Can also have breathing pause/apnea, color changes
    - Can also be associated with poor responsiveness (vagal response)
    - Often confused with seizures or dystonia
  - Onset typically in infancy/early childhood
  - **Exam/History Pearls**: association with feeds, supine position, frequent spit-up
  - Treatment: reassurance, reflux measures
Movement Disorder Mimics in Early Childhood

- **Infantile/Toddler Gratification syndrome**
  - Early childhood masturbatory behavior
  - Considered a normal behavior in early childhood
  - Often accompanied with abnormal postures, facial grimaces or flushing, diaphoresis, grunting, leg scissoring, eye rolling, rocking
  - Episodes can last minutes or hours
  - Often confused for seizures or dystonia
  - May appear poorly responsive/groggy during or after

- **Exam/History Pearls**: Can be interrupted with distraction or noxious stimulus
  - Treatment: education and reassurance
Common Movement Disorders in Older Children

- Tremor
- Tics
- Stereotopies
Tremor

- Tremor is typically a benign phenomenon in children
- Most common causes of tremor in childhood are:
  - Drug-induced tremor (SSRIs, ADHD medications, seizure medications)
  - Essential tremor
  - Psychogenic tremor
- Most important part of examination of the tremor is to distinguish action/postural tremor from a rest tremor
- Exam Pearls:
  - If you are concerned about a rest tremor, always examine tremor with the patient supine
  - A postural tremor is often confused as a rest tremor
  - Finger-to-nose testing should be done very slowly, when done fast, can confuse an action tremor for an intention tremor
  - Other types of tremor: dystonic, intention, enhanced physiologic
- All rest tremors require referral to Neurology
Essential Tremor

- Action/kinetic and postural tremor
  - Typically presents in bilateral hands first
  - Typically runs in families (+FHx in ~50%)
  - Severity can be highly variable between individuals in the same family
  - Generally stable but can progress slowly/gradually
- Onset highly variable (typically adolescence/early adulthood)
  - Can be in infancy or late adulthood
- Differential: Enhanced physiologic tremor (anxiety), thyroid dysfunction, neuropathy, developmental coordination disorder
- Relatively stable mild bilateral action hand tremors do not require additional work up or labs
  - Tremor and history consistent with Essential Tremor do not necessarily require referral to Neurology (though always reasonable to refer)

Treatment: OT, propranolol, primidone
Tics

- Repetitive motor movements or vocalizations
  - In practice any movement, sound or behavior can potentially be a tic
  - Mostly simple (purposeless, quick) but can be complex (purposeful appearing, orchestrated bouts/clusters)

- Onset typically between 4-8yo (but can be as early as 2yo or late as early adulthood
  - Typically will resolve in <12mo
  - If >12mo (chronic tics) will wax/wane and generally peak in adolescence before resolution or significant reduction in late teens or early adulthood

- Hallmark features
  - Waxing/waning course
  - Change over time
  - Suppressibility (at least briefly)
  - Reduction during focused or engaging tasks (sports, music, art)
Myths about tics

- Tics do not occur in sleep
- Tics are caused or worsened by medications
  - ADHD medications worsening or causing tics is an enduring myth amongst providers
- Tourette syndrome is worse than other chronic tic disorders
- Tics need to be treated if they are frequent or very noticeable
  - Tics should only be treated if they cause significant functional impairment, pain or are socially inappropriate
Chronic Tics Disorders and Tourette Syndrome

- In practice we do not treat Tourette Syndrome any different than other chronic tic disorders
- Most patients with chronic tics do not need treatment for their tics
- Treatment of co-morbid conditions is often a more important measure
  - Anxiety, OCD, **Anger outbursts**, Repetitive behaviors, Stuttering
  - Due to co-morbid conditions, chronic tic patients are often labeled as “high-functioning autism”
  - Do not be afraid to treat co-morbid conditions due to fear of “making tics worse”
- Referral to a movement disorder neurologist is often beneficial to provide education and normalize tics to prevent over treatment of tics and over-analysis of transient increases in tics
Treatment of Tics

- Only in cases of:
  - Significant impairment in function
  - Persistent or frequent pain related to tics
  - Socially inappropriate tics

- Treatments:
  - CBT
  - Alpha agonists (clonidine, guanfacine)
  - Topiramate
  - Muscle relaxants
  - Antipsychotics
  - VMAT inhibitors (tetrabenazine)

- Would refer to neurology if fails alpha agonist
Inflammation in Tic Disorders and Obsessive-Compulsive Disorder: Are PANS and PANDAS a Path Forward?

Donald L. Gilbert, MD, MS

A Pediatric Neurology Perspective on Pediatric Autoimmune Neuropsychiatric Disorder Associated with Streptococcal Infection and Pediatric Acute-Onset Neuropsychiatric Syndrome

Donald L. Gilbert, MD, MS, FAAP, FAAN, Jonathan W. Mink, MD, PhD, FAAN, FANA, FAAP, and Harvey S. Singer, MD

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PANDAS/PANS

PANDAS 1998 (Swedo et al)
• OCD and/or tic syndrome
• Prepubertal onset
• Episodic (sawtooth course)
• Association with GABHS infections
• Association with neurologic signs

PANS 2012 (Swedo et al)
• Abrupt, dramatic onset
• OCD or severely restricted food intake
• 2 or more of the following

- Anxiety
- Emotional lability and/or depression
- Irritability, aggression and/or severely oppositional behavior
- Behavioral regression
- Deterioration in school performance
- Sensory or motor abnormalities
- Somatic symptoms, including sleep disturbances, enuresis or urinary frequency
- Symptoms not better explained by neurological or medical disorder
PANDAS/PANS: What’s the current consensus?

- Two main schools of thoughts in mainstream child neurology provider community
  - 1- PANDAS/PANS does not exist as a distinct clinical entity from primary psychiatric disorders and tic disorders
  - 2- PANDAS/PANS may exist as a distinct auto-immune entity but if it does it is exceedingly rare (not the 1 in 200 that Dr. Swedo claims)
PANDAS/PANS: What’s the current consensus?

- Routine evaluation for GABHS or ASO titers in the absence of symptoms of an active GABHS infection is not indicated in patients who present with tics or OCD (unless there is chorea).

- Antibiotic therapy is not recommended in the absence of a positive culture for GABHS.

- Chronic antibiotic therapy is not recommended.

- Immunotherapy is not recommended unless CNS inflammation is demonstrated or other neurologic signs (seizures, chorea, dystonia, encephalopathy, etc.) develop.
Stereotopies

- Repetitive motor movements in childhood
  - Can be simple (hand flapping, hand twirling, hand wringing)
  - But can also be very complex, coordinated and bizarre in appearance
- Onset as early as 6mo to 3yo *(generally earlier than tics)*
  - Typically resolve by 10yo though some carry them into adulthood
    (with less frequency of occurrence)
- Unlike tics, they do not wax and wane (daily) or change over time
- Like tics, they are suppressible, often can be triggered by emotion
- Treatment: reassurance
Cerebral Palsy

- Though often classified as spastic, hypotonic, dystonic or ataxic, movement phenomenology is often mixed.
- Always take a diagnosis of Cerebral Palsy with a grain of salt.
  - Many patients carry historical diagnoses of CP and the etiology of their symptoms is never properly investigated.
  - Without a compelling birth history or MRI neuroimaging, these patients should always be referred to neurology for evaluation.
  - Some patients with undiagnosed treatable causes of their symptoms go years untreated.
Referrals

- Any concern for chorea, dystonia, myoclonus or ataxia should be referred to Neurology
  - Acute onset or rapidly progressive symptoms often need ED evaluation

- Rest tremors should always be referred to Neurology

- Under-treated CP or CP without a clear etiology

- Mild Tics and Essential Tremor can reasonably be handled by primary providers
  - Referrals for education/reassurance can often be helpful

- For most movement disorders, laboratory work-up and neuro-imaging can be safely deferred until seen by a neurologist
Questions?