RECOGNIZING ABNORMAL MOVEMENTS IN CHILDREN

Jeff Waugh, MD/PhD
Medical Director, Pediatric Movement Disorders and Deep Brain Stimulation Program
Boston Children’s Hospital and Massachusetts General Hospital
Jeff.Waugh@Childrens.Harvard.edu
DISCLOSURES

- I have no financial relationships with any for-profit corporations or entities
- Grant funding from non-profit entities to study pediatric dystonia
CC: Unable to eat or drink

5yo RH girl with lifelong “twitchiness,” now with worsening difficulty when running, writing, eating, drinking.
How are Movement Disorders Different?

- There are many types of abnormal movement:
  - ataxic
  - apraxic
  - spastic
  - paretic
  - functional / conversion
  - antalgic
  - myopathic

- In contrast to those types, movement disorders:
  - Originate centrally (brain or spinal cord)
  - Are more than just a failure of control or fine-tuning
  - Result from abnormal motor control networks

- Can be too much (hyperkinetic) or too little (hypokinetid)
Movement Disorders localize to a Network

Movement Disorders connote a disruption in motor control networks

- More than just a “routing” problem – an emergent property
- Lesions at multiple sites within a network can produce abnormal

Abnormal Motor Control Networks in Dystonia

Carbon and Eidelberg *Neuroscience* 164 (2009) 220–229
STEP 1: 
DIAGNOSIS BEGINS WITH PHENOMENOLOGY

Describe the patient based on:
- How much movement? (Hyper- or Hypokinetic)
- Type of movement
- Co-incidence with other Movement Disorders

If a picture is worth a thousand words, then a video is worth a million
EXAM – THINGS NOT SHOWN

- Writing: cramped and sloppy, progressive worsening - a pattern consistent with hand dystonia
- Dystonic posturing of the L. foot while walking, R. hand while writing.

STEP ONE:
Myoclonus affecting trunk, neck and R > L arm
Dystonia affecting mostly L body, task specificity
HYPERKINETIC MOVEMENT DISORDERS

- **Chorea** - “Dance like”, continuous, irregular, often incorporating planned movements
- **Tics**
- **Tremor**
- **Myoclonus**
- **Dystonia**
HYPERKINETIC MOVEMENT DISORDERS

- **Chorea**

- **Tics** - Sudden, rapid, purposeless, repetitive, stereotyped non-rhythmic, suppressible.
  
  - Rules for tics: 1. Suppressible  2. Evolving  3. Premonition  4. Associated factors (e.g, ADHD)

- **Tremor**

- **Myoclonus**

- **Dystonia**

  Too broad a topic to consider today.
HYPERKINETIC MOVEMENT DISORDERS

- Chorea
- Tics
- Tremor - Oscillating, rhythmic about a fixed point, usually a single joint but can also be an axis
- Myoclonus
- Dystonia
HYPERKINETIC MOVEMENT DISORDERS

- Chorea
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HYPERKINETIC MOVEMENT DISORDERS

- Chorea
- Tics
- Tremor
- Myoclonus
- Dystonia - Sustained muscle contraction, often leading to twisting movements or fixed postures. Co-contraction of agonist-antagonist muscles, overflow to surrounding muscles.
Paroxysmal Dyskinesias – multiple, often overlapping Movement Disorders

Motor Stereotypies

Developmental (NORMAL) Movement Disorders
- At least 12 distinct disorders for which the appropriate treatment is reassurance.

OFTEN FORGOTTEN:

Functional Movement Disorders (Conversion Disorder)
- Common, disabling, readily identified
- More frequent than any other disorder discussed here
What parts of the body are affected?

- Focal - Segmental - Multifocal - Generalized
- Often will start with one area and later spread
- Pattern of presentation and later spread aids diagnosis

Timing

- Early morning?
- Late in the day?
- Induced by action? After exercise?
STEP 2 FOR OUR TWITCHY 5 YEAR OLD:

- Bilateral and trunk involvement, upper and lower: Generalized
- No clear progression, though with age sx impacted her more
- Stimulus-induced myoclonus
- Multi-focal task-specific dystonia
STEP 3: APPROXIMATE THE ETIOLOGY

- Presence of cognitive decline or epilepsy
  - a broader encephalopathy?
- Primary movement disorder (symptom in isolation) vs.
  Secondary to injury or degeneration.
  - Will often require MRI
- Incorporate family history, response to prior med trials, results of imaging / EEG / etc.
**CHOREA**

- “dance like,” continuous, irregular, often incorporating planned movements
- Random movements of chorea rarely repeat, are not suppressible. Watch in 10s bins – can you predict movement in next 10s bin?
- Important to distinguish from akathisia – inner restlessness
- Most important clues on exam are motor impersistence: the inability maintain a fixed posture
  - Jack-in-the-box tongue
  - Milk maid’s grip
  - Touchdown sign
- Hypotonia
11yo boy with 8 years of adventitious movements
EXAM FINDINGS IN CHOREA: MOTOR IMPERSISTENCE

Jack-in-the-box tongue                     Milk maid’s grip                     Touchdown sign
IDIOPATHIC CHOREA

11yo F, progressive chorea over 5 years, now with cognitive decline and mood lability
TREMOR

- Oscillating, rhythmic about a fixed point, usually a single joint but can also be an axis (neck, trunk)
- Varies with position (rest, kinetic, postural, etc.)
- Uncommon in kids, but makes up 10-20% of MvDis clinic visits
- Most common in systemically-ill child – more frequently, an inpatient consult, rarely contributes to diagnosis
12yo boy with autism and tremor
WHEN IS TREMOR... NOT TREMOR?
MYOCLONUS

- A brief, involuntary muscle jerk that is non-suppressible and generally has no premonitory features.
- May be an isolated finding (primary myoclonus) or can be a symptom of many diseases.
- Physiologic myoclonus occurs episodically throughout life: hiccups, hypnic jerks, fatigue-related benign myoclonus.
- Developmental conditions (e.g., benign neonatal sleep myolonus, benign myoclonus of infancy)
- May come as a referral for ataxia!
18yo man with epilepsy and non-epileptic myoclonus
19yo man with autism and myoclonus
Dystonia

- Sustained muscle contraction, often leading to twisting movements or fixed postures. Often painful.
- Co-contraction of agonist-antagonist muscles; overflow to surrounding muscles not typically involved in that action.
- Predilection for over-learned actions – writing, typing, walking, speech, musical instruments
- Normal motor function in between triggering-tasks, normal in other body parts

3rd-most common movement disorder
9yo boy with abnormal gait
DYSTONIA OFTEN LEADS TO BIZARRE MOVEMENTS
WRITING: DYSTONIA VS. CHOREA

1. Walfre
2. Walfre
3. Walfre

Dystonia: Progressive loss of control, Tightening of script

Chorea: No Progression, but irregular intrusions into text

Today is a sunny day

Today is a sunny day

Today is a sunny day
DYSTONIA CAN BE FOCAL…

14yo with task-based dystonia of R hand
Patients in dystonic storm can suffer fractures, severe muscle breakdown, dehydration, hyperthermia…

Typically requires barbiturate or benzodiazepine coma to break dystonic status
**Dyskinesias**

- “Dyskinesias” simply means abnormal movements – nonspecific term, but useful in that these conditions are recognized by the shifting combination of movement disorders.
- Intermittent, involuntary, often of a “strange/bizarre” character
- Note that “dyskinetic CP” refers to a combination of dystonic and choreiform movements – but in practice is usually highlighting the dystonic feature.

**Paroxysmal Dyskinesias**

- 3 subtypes, all autosomal dominant with high penetrance
- Key ?’s: How often? How long? Just before?
PKD – ELICITED BY RAPID MOVEMENT
HYPOKINETIC MOVEMENT DISORDERS

Parkinsonism

- Any two of the Parkinsonian quartet:
  - Bradykinesia
  - Tremor at rest
  - Rigidity (velocity IN-dependent resistance)
  - Postural instability

- Idiopathic Parkinson disease is exceedingly rare in kids, still rare in the 20s.
- Most commonly due to medications
- Post-infectious, stroke
- Many inherited causes, usually degenerative
20yo woman with slow movements
A SPECTRUM EXISTS BETWEEN NORMAL AND ABNORMAL MOVEMENTS

- When to reassure?
- When to test?
- When to treat?

- The only way to get perspective on what are normal variants is to spend time with (many) children outside of a clinical setting.
5yo girl with repetitive movements
9yo girl with repetitive movements
1st Younger Sister of Patient with Stereotypies
2nd Younger Sister of Patient with Stereotypedies
FAMILY HISTORY:
AWKWARD SOCIAL INTERACTIONS
Stereotypies are common, found in:

- 40% of typically-developing children and
- 85% of developmentally-delayed children

- Hoch et al., Journal of Pediatric Psychology 2016

In Stereotypies:

- Movements can evolve over time but typically preserve a core feature(s).
- Rarely distressing to the child - often soothing, enjoyable, or simply below the level of awareness.
IN SUMMARY:

- Capture a video whenever possible
- Organize the patient in a hierarchical fashion:
  - Phenomenology First!
  - Distribution and timing
  - Preliminary guess at etiology, inheritance pattern
- Learn to recognize the patterns of movement that are common in childhood and abnormal only by adult standards
**SPASTICITY VS. DYSTONIA**

Very common consult question. How to distinguish?

**Spasticity**
- Severity increases with RATE of movement
- Typically large body-area involved: hemibody or at least most of limb
- Always referable to known/suspected injury or slowly-progressive process

**Dystonia**
- Increases with TYPE of movement  
  e.g., may resolve when walking backward in foot dystonia, may resolve with drumming of fingers in dystonic writers cramp
- Often very specific for particular muscles. May generalize to other body areas, but even then is muscle-specific
- Usually on a background of stable function X years

End of the day: they just look different. Try to see lots of cases!