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

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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

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Overview of Movement Disorders

Hyperkinetic

- **Tremor**
- **Dystonia**
- **Chorea**
- **Tics**
- Myoclonus
- Athetosis
- Ballism
- Ataxia

Hypokinetic

- Parkinsonism
  - Bradykinesia
  - Rigidity

More frequent in children

Uncommon in children

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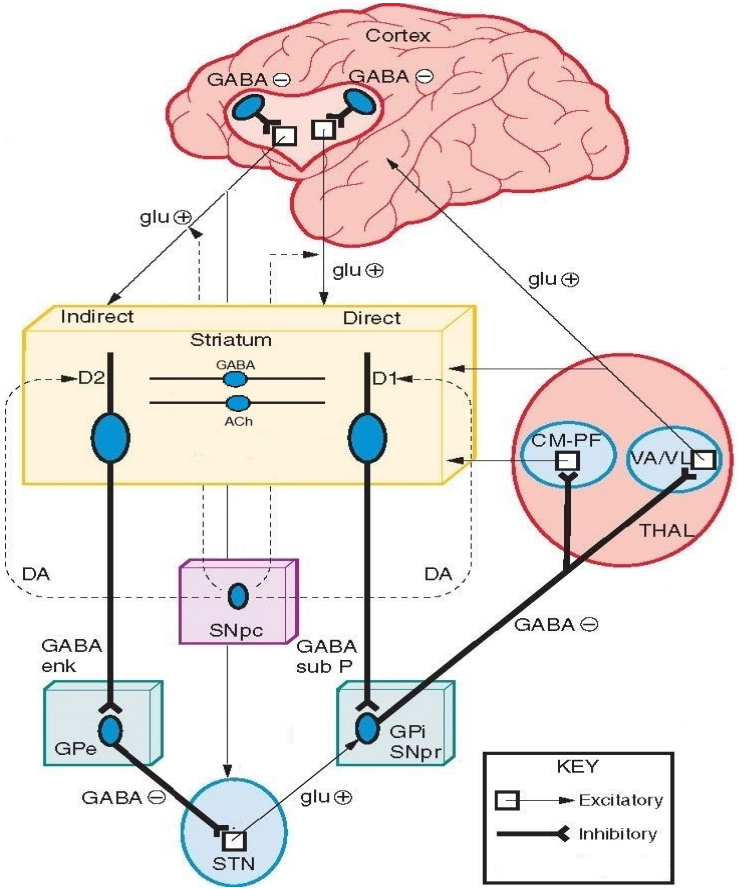
Pathophysiology



- Cortico-striato-thalamo-cortical circuitry
  - Indirect pathway
  - Direct pathway

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Video 1

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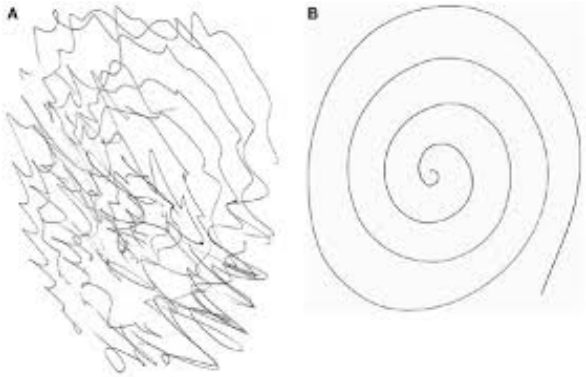
Tremor- *"rhythmic back-and-forth or oscillating involuntary movement about a joint axis"* (Sanger et al. Mov Disord. 2010 August 15; 25(11): 1538–1549.)

- 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

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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

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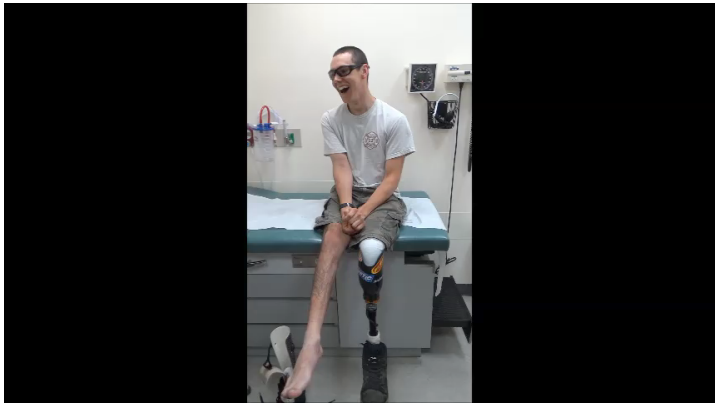
Video 2



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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

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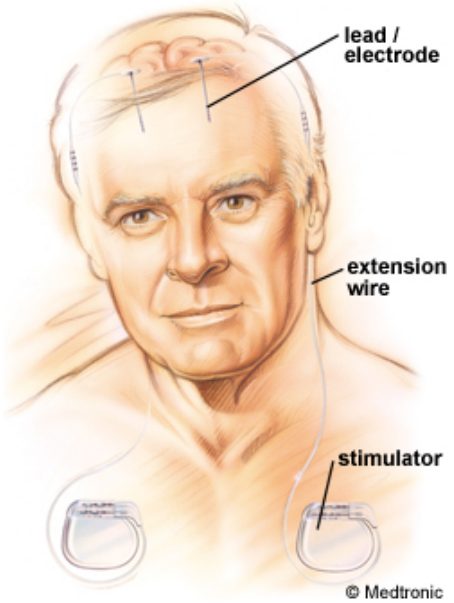
Dystonia treatment

- Pharmacologic
- Surgical

Table 5
Commonly used oral medications for dystonia

Class of Medication	Examples
Anticholinergics	Benztropine, biperiden, ethopropazine, orphenadrine, procyclidine, trihexyphenidyl
Dopaminergics	Levodopa, pramipexole, ropinirole, tetrabenazine
GABAergics	Alprazolam, baclofen, chlordiazepoxide, clonazepam, diazepam
Muscle "relaxants"	Baclofen, benzodiazepines, carisoprodol, chlorzoxazone, cyclobenzaprine, metaxalone, methocarbamol, orphenadrine
Others	Carbamazepine, cannabidiol, cyproheptadine, gabapentin, lithium, mexiletine, nabilone, riluzole, tizanidine, zolpidem

Jinnah & Factor, 2015



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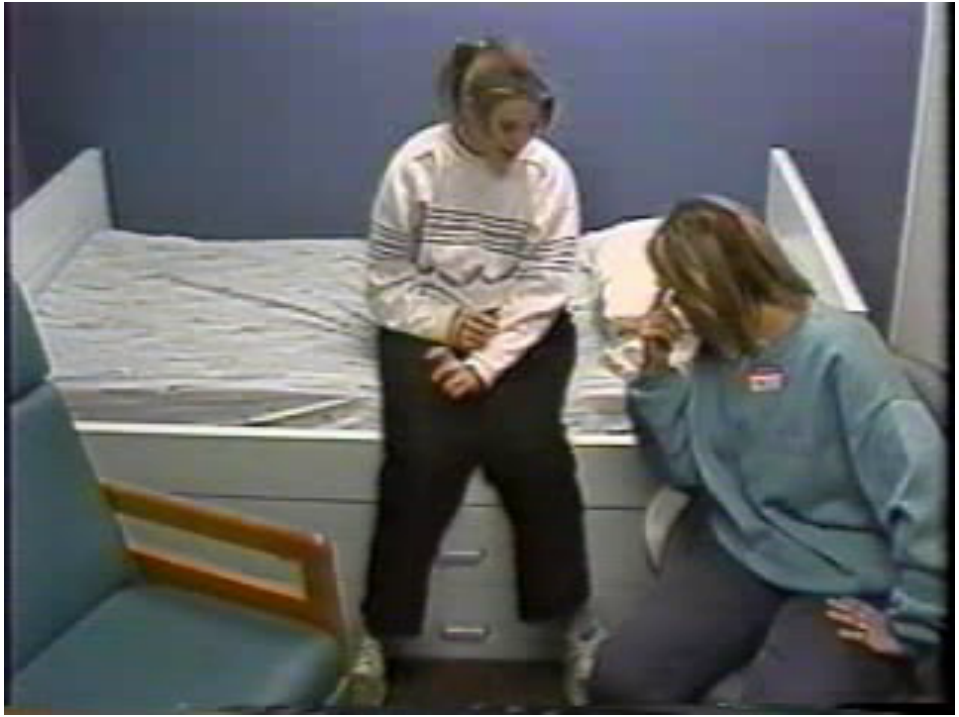
Video 3



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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

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

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

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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

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Video 4

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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

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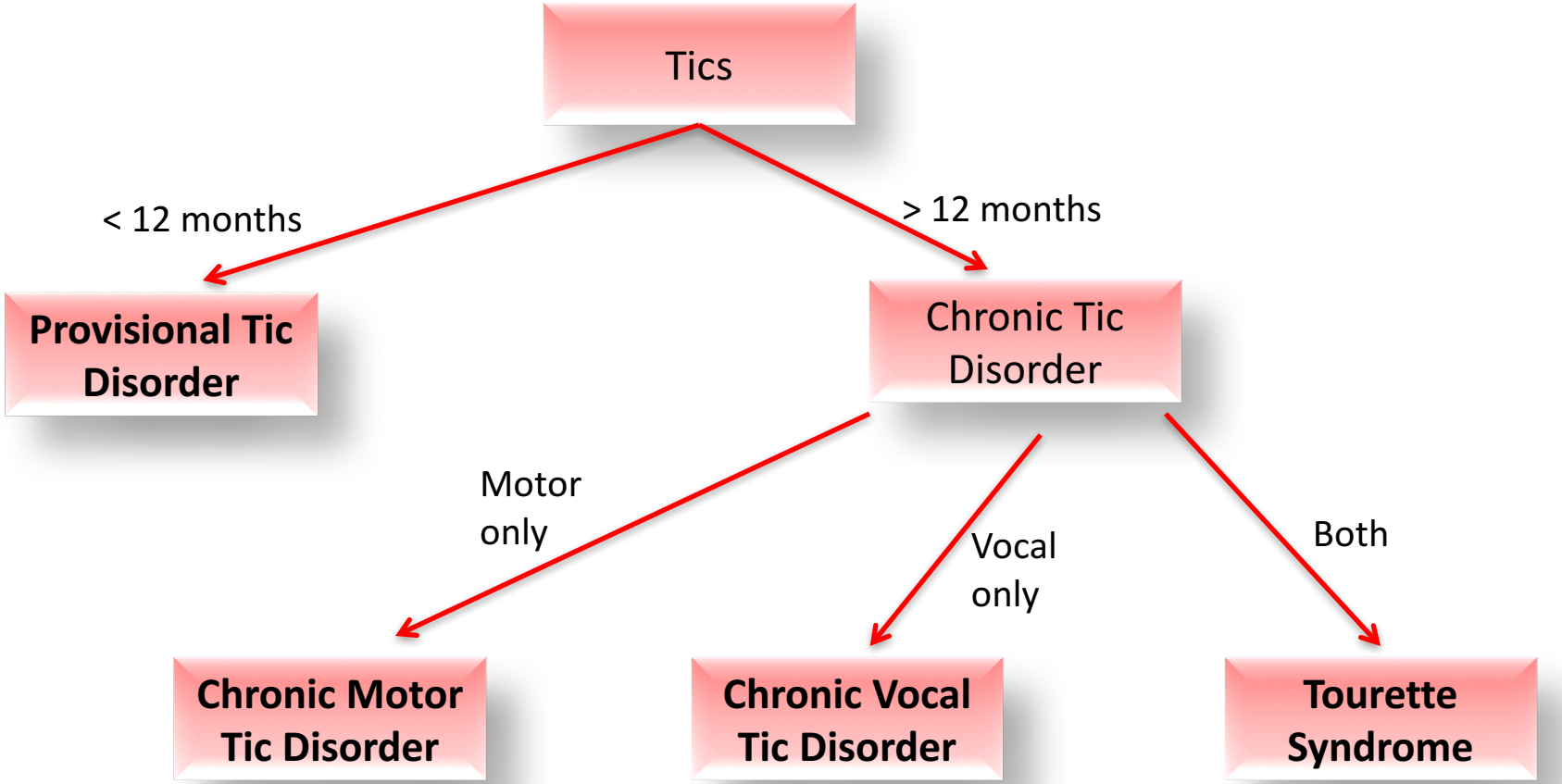
Additional Characteristics

- 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



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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!

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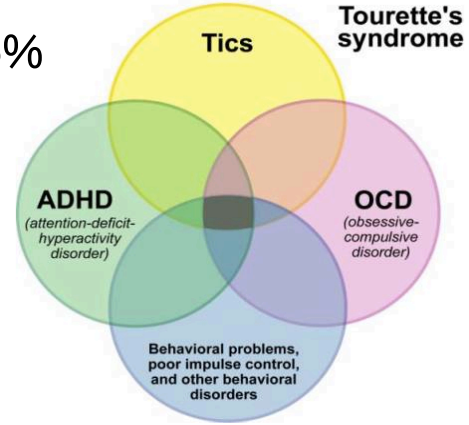
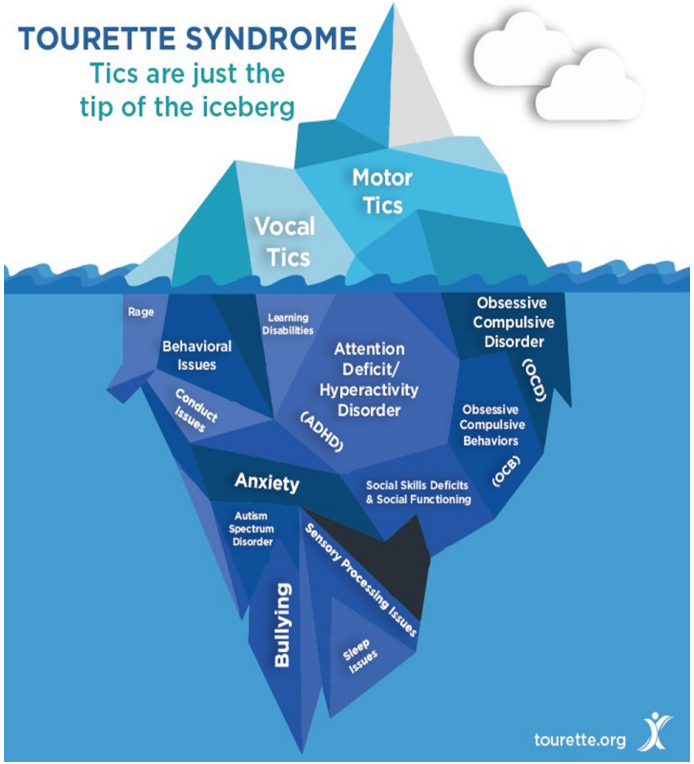
Comorbidities

- 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





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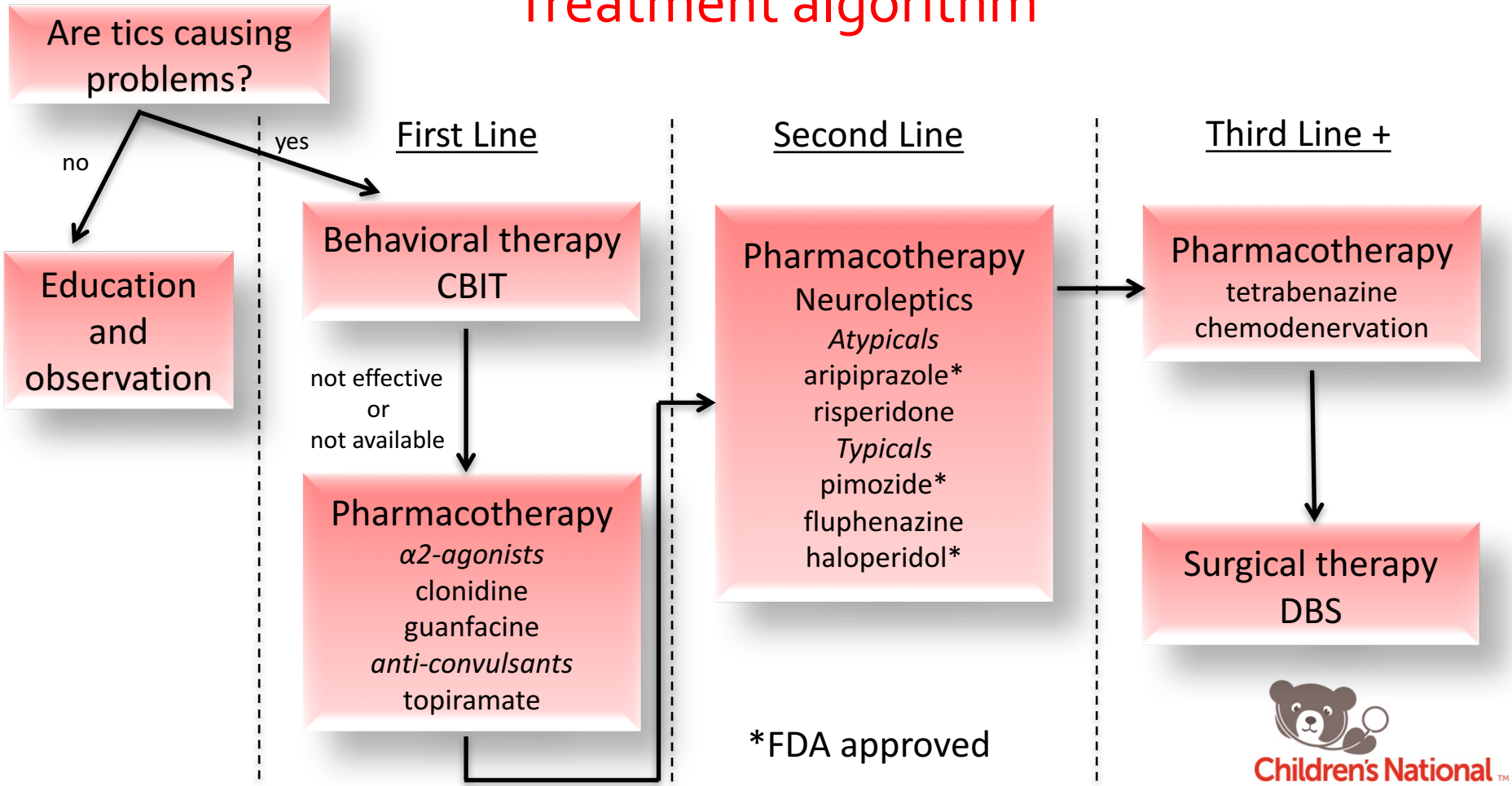
Treatment algorithm



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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

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Referrals

- Child Neurology Movement Disorder Program

  (202) 476-3611
- ltochen2@childrensnational.org

Additional Resources

- www.tourette.org
- https://www.cdc.gov/ncbddd/tourette/index.html

Tourette Association of America

Awareness. Research. Support.

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Thanks!

Tourette/Tic, Movement Program

- Neurology
  - Laura Tochen
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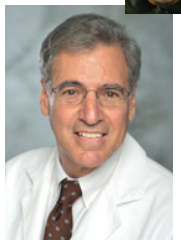
- Psychiatry
  - Julia Dorfman



- CBIT providers: Lauren Dome, Laura Gray, Sarah Hornack, Mi-Young Ryee

DBS Program

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Chima Oluigbo
-Neuropsych
Yael Granader
-Research
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Mary Furda

Cerebral Palsy Program

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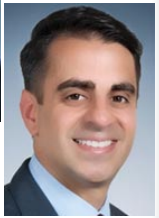
Jeff Rabin
-Orthopedics
Sean Tabaie
-Neurosurgery
Robert Keating



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