

Pediatric Movement Disorders

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Bresnan Course – September 2020

Disclaimers

- I do not have financial relationships with any for-profit corporations or entities.
- All patients have consented for videotaping

Learning objectives

- Use language to describe abnormal movements
- Identify a few common movement disorders
- Identify a few uncommon but treatable movement disorders



Epidemiology

Public Domain, <https://commons.wikimedia.org/w/index.php?curid=3901667>

1

Tics
Stereotypies
Dystonia
Chorea
Tremor
Myoclonus

HYPERKINETIC

2

Cerebellar ataxia
Sensory ataxia

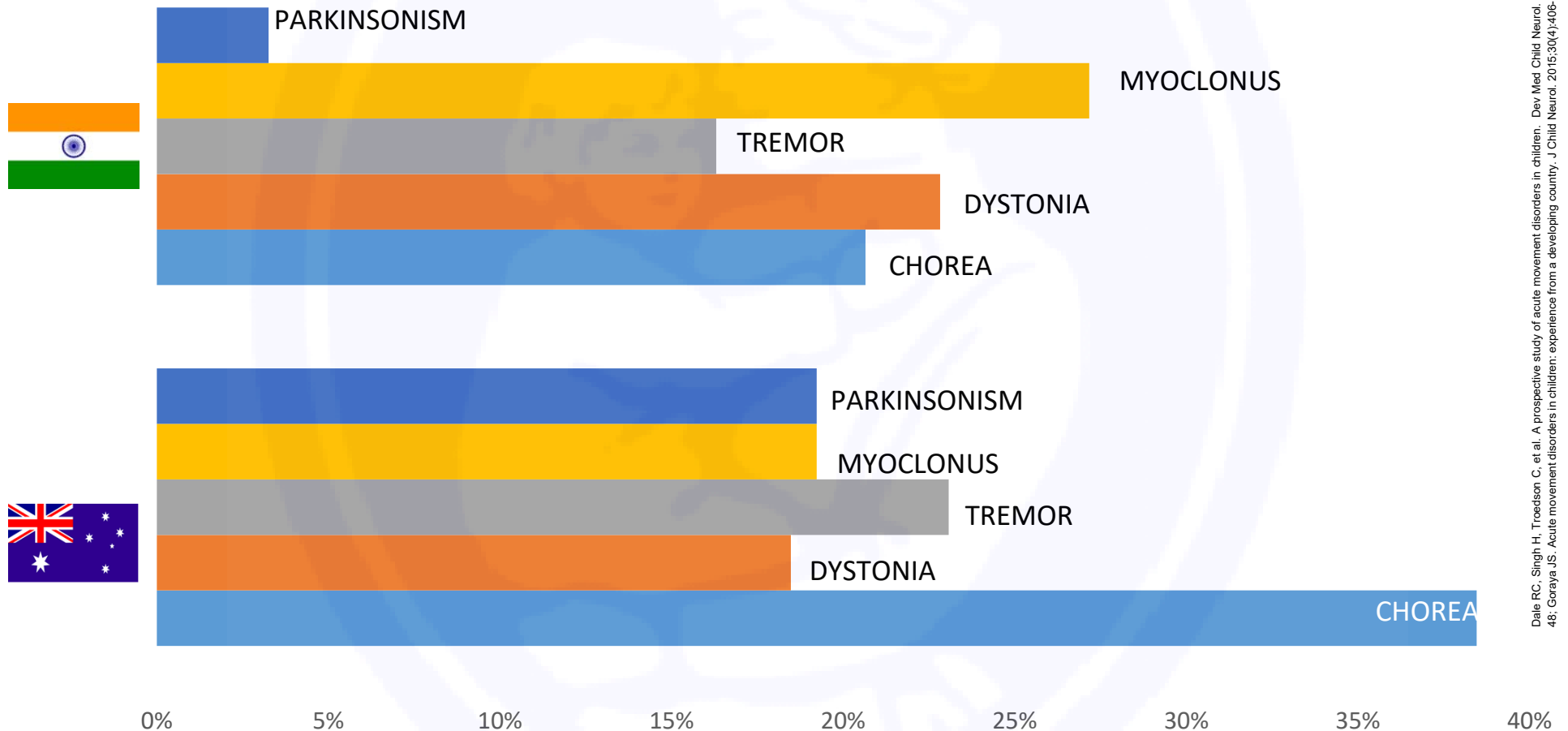
ATAXIC

3

Parkinsonism

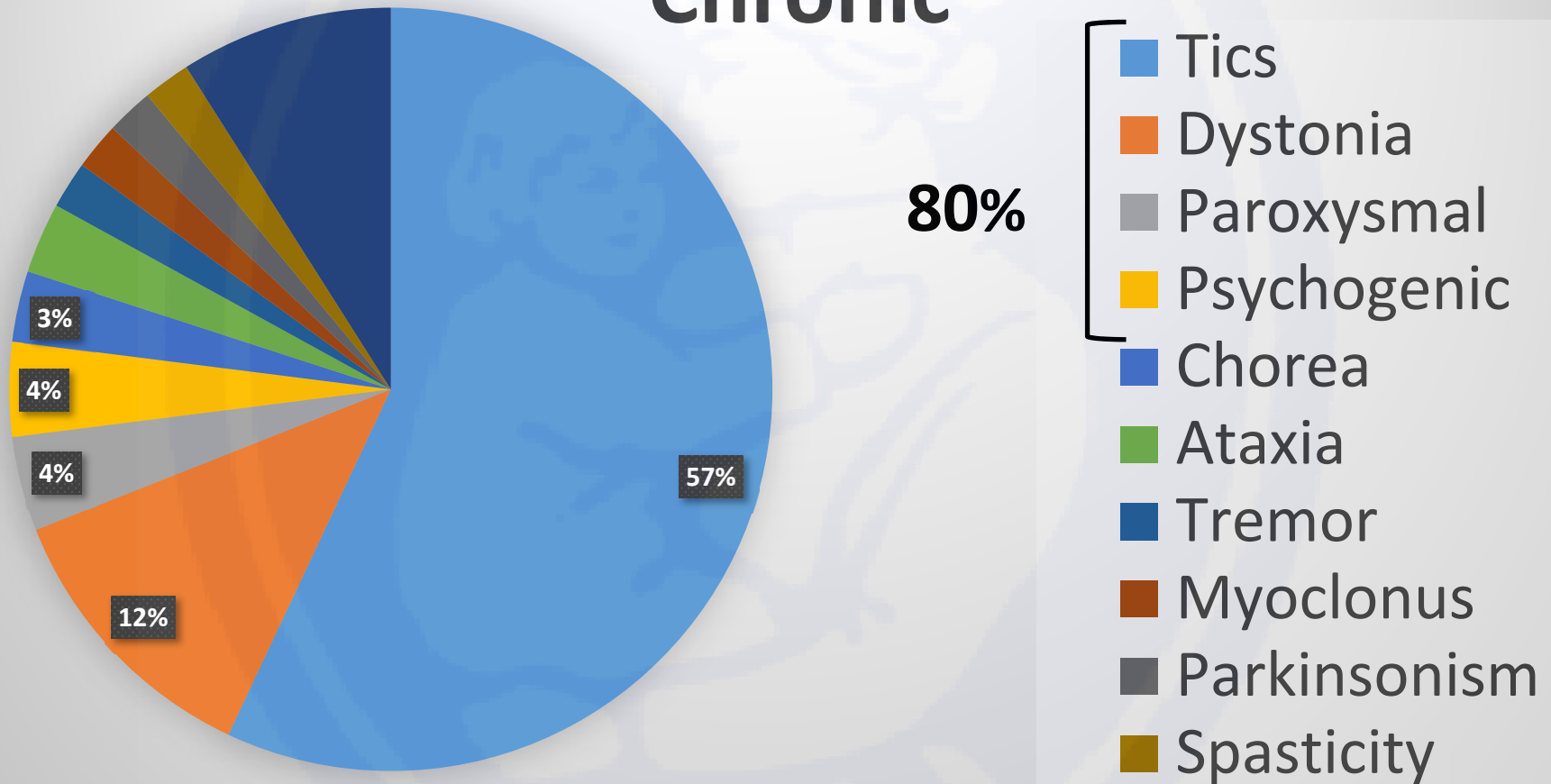
HYPOKINETIC

Acute Movement Disorders (NOT INCLUDING TICS)



Dale RC, Singh H, Troedson C, et al. A prospective study of acute movement disorders in children. Dev Med Child Neurol. 2010;52(8):739-48; Goraya JS. Acute movement disorders in children: experience from a developing country. J Child Neurol. 2015;30(4):406-11.

Chronic



Adapted from: Baumer, T., Sajin, V., & Munchau, A. (2017). Childhood-Onset Movement Disorders: A Clinical Series of 606 Cases. *Movement Disorders Clinical Practice*, 4(3), 437–440.



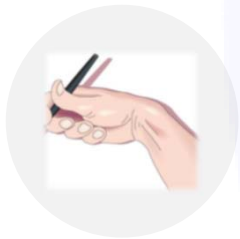
Phenomenology

https://upload.wikimedia.org/wikipedia/commons/1/12/Physician_setting_a_dislocated_arm.jpg



Dystonia

Dystonia



Patterned, sustained or intermittent **muscle contractions** causing abnormal, often repetitive, movements and/or postures

Albanese, 2013 | Breakfield 2008

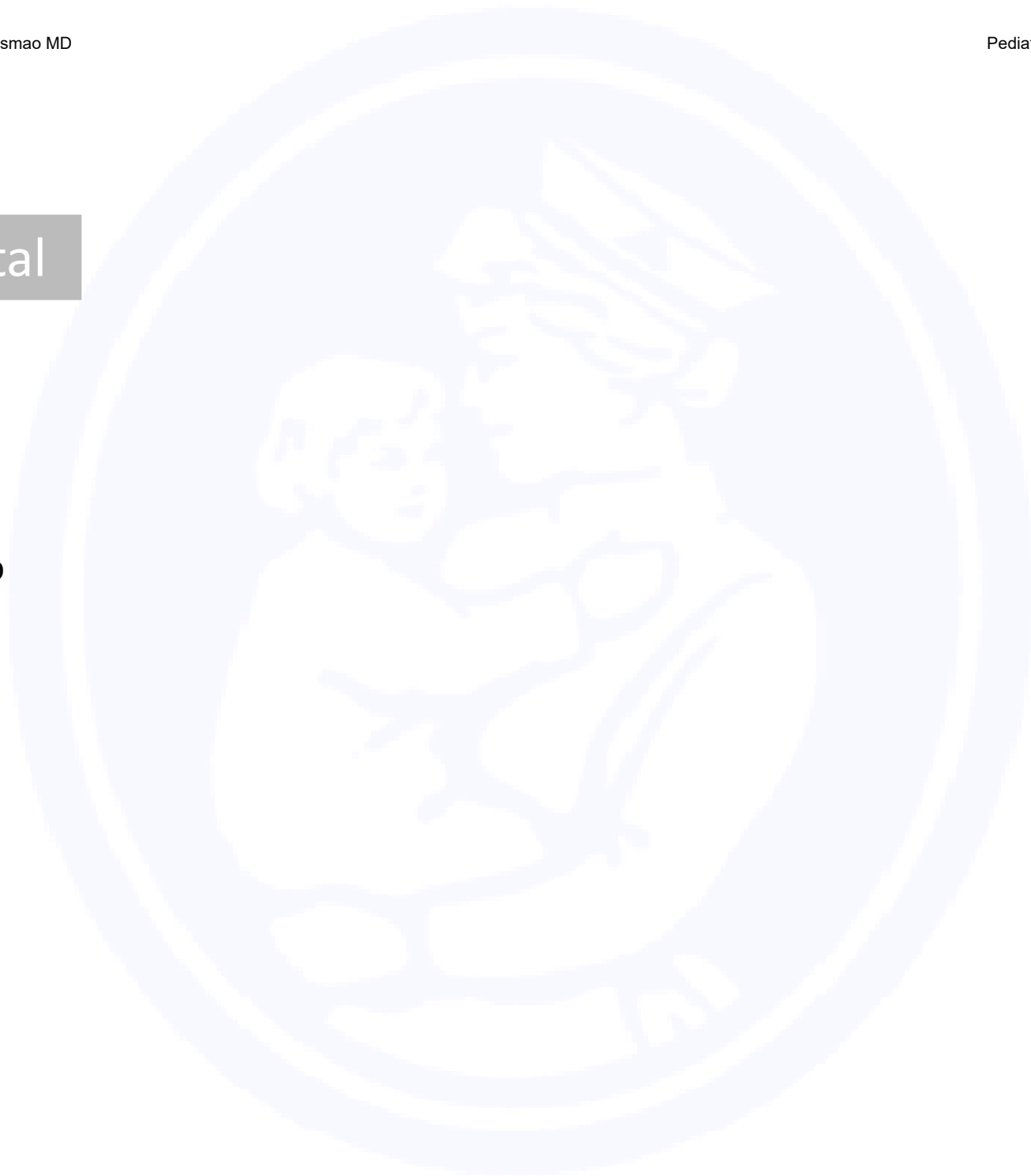
Focal

Video



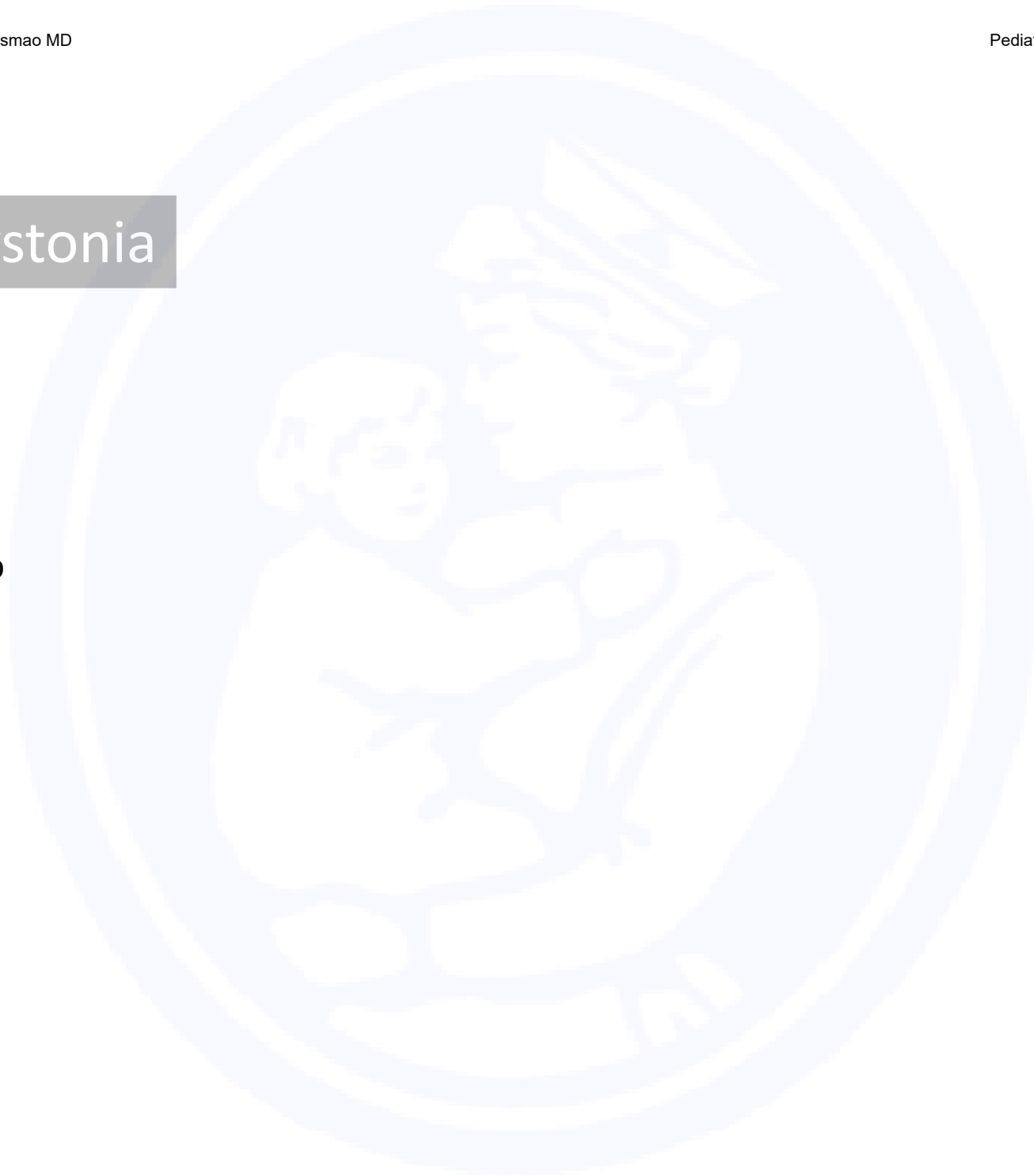
Segmental

Video



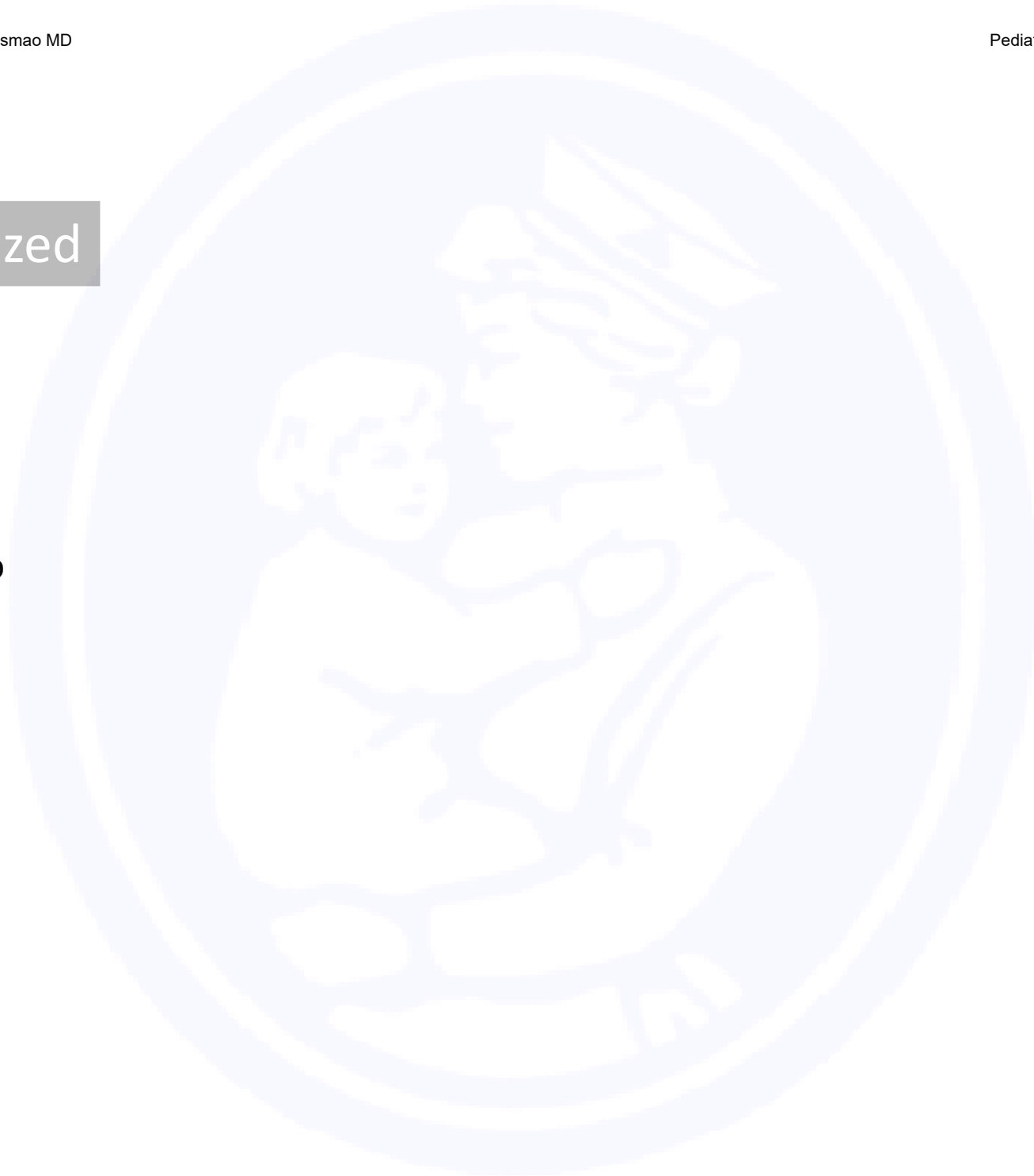
Hemidystonia

Video



Generalized

Video



Overflow

Video

Adapted from Meyer E, Carss KJ, Rankin J, et al. Mutations in the histone methyltransferase gene KMT2B cause complex early-onset dystonia. *Nat Genet* 2017;49(2):223–37.

Sensory trick



Brissaud - 1894

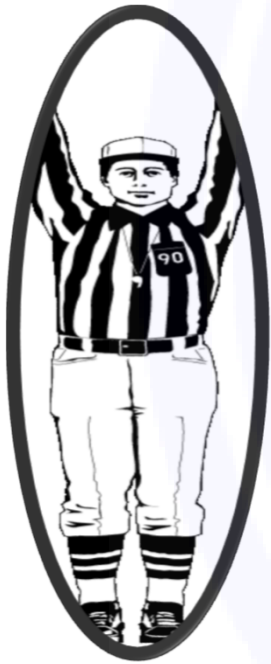
Video

Martins J, Darling A, Garrido C, et al. Sensory Tricks in Pantothenate Kinase-Associated Neurodegeneration: Video-Analysis of 43 Patients. *Mov Disord Clin Pract* 2019;6(8):704–7.

A large, faint, light blue circular logo of Boston Children's Hospital is centered in the background. It features a classical illustration of a child and a woman, possibly a nurse or parent, in a tender embrace.

Chorea

Chorea – Motor impersistence



Touchdown sign



Piano playing



Milkmaid grip



Darting tongue

Video

12yo boy. Has had movements of his fingers, arms and sometimes trunk since 1 yo. When young, would wake up with twitching and very evident movements early morning, but over the years became more permanent. His father has similar movements.

Athetosis

Chorea

Ballismus

Severity

The diagram consists of a horizontal bar with a color gradient from light orange on the left to dark brown on the right. The bar is divided into three sections, each containing a text label. Below the bar is a large yellow arrow pointing to the right, with the word 'Severity' written in italics inside it. The background of the slide features a large, faint, light blue circular seal of Boston Children's Hospital.

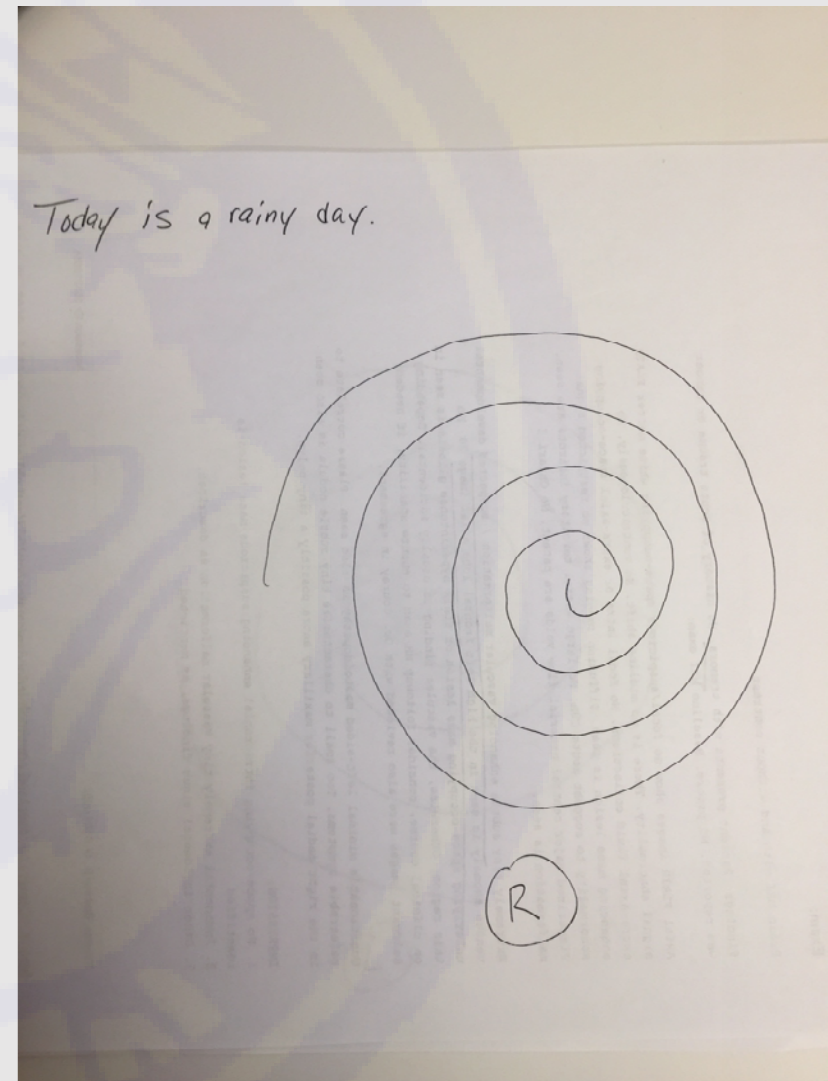
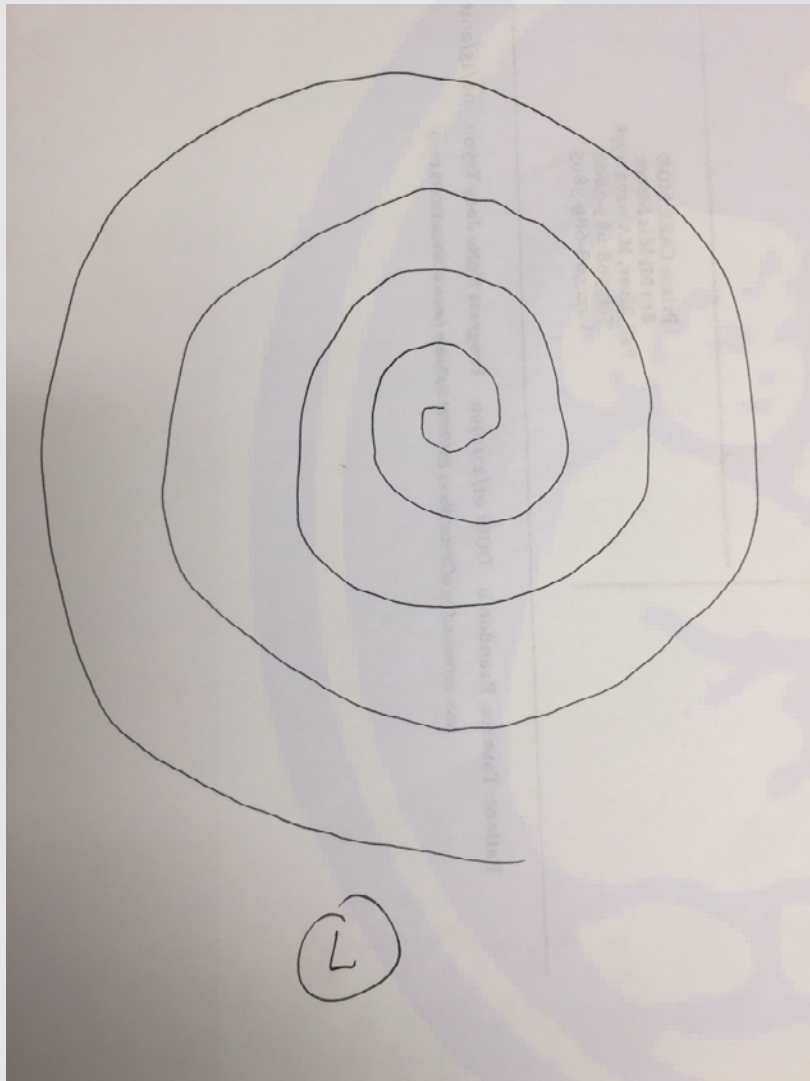
What about dyskinesia?



Video

7yo girl with hypoxic ischemic encephalopathy with dyskinetic CP

Tremor



Tremor qualifiers



Myoclonus

Video

21 yo man with a history of obsessive compulsive symptoms and autism spectrum disorder. He has had “twitches” since age 4.5.

Ataxia

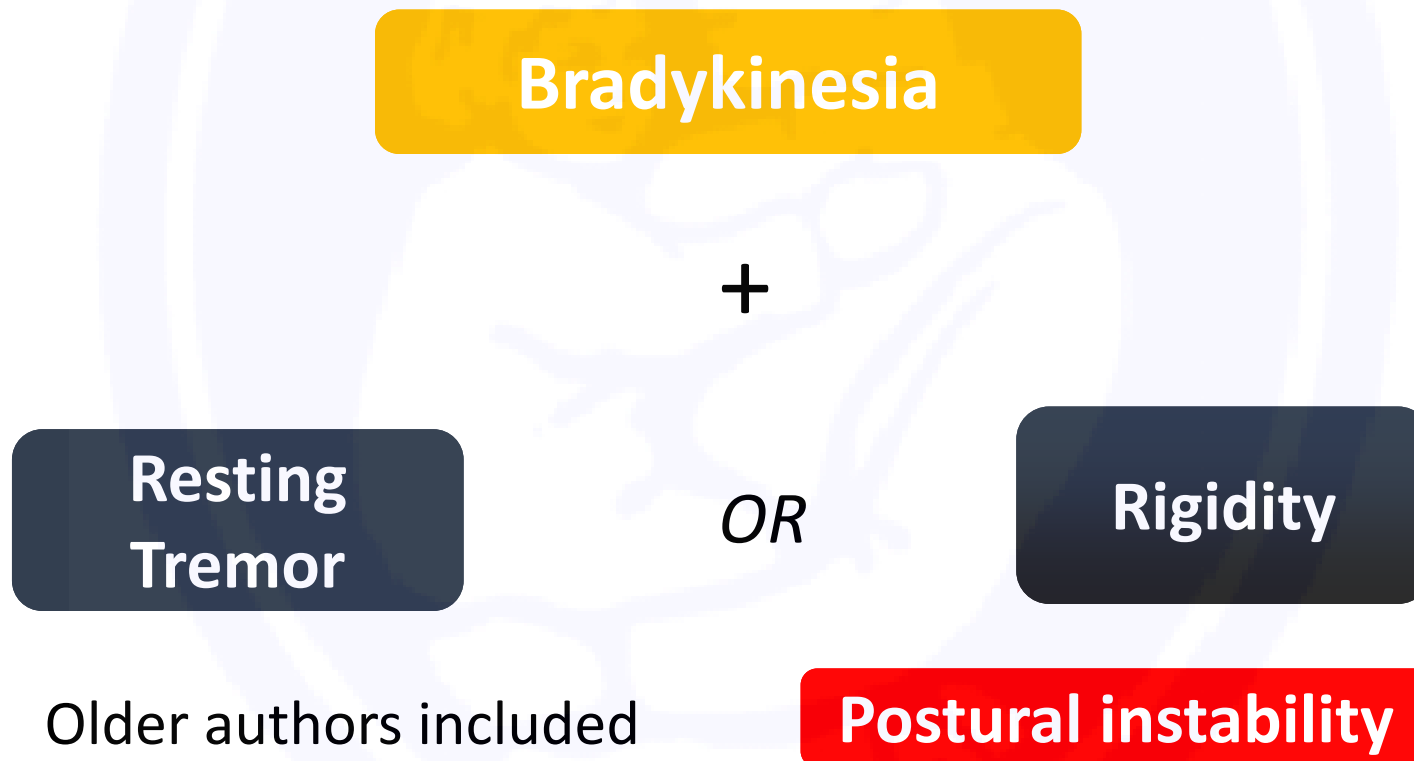


Video



Parkinsonism

Core criteria



Pediatric parkinsonism

Video

- **Hypokinesia >> Bradykinesia**
- **Often with associated dystonia**



clinical scenarios

By https://wellcomeimages.org/indexplus/obf_images/04/aa/df6e2ce39c21c846a68959d65e62.jpg Gallery:
<https://wellcomeimages.org/indexplus/image/V0011195.html> Wellcome Collection gallery (2018-03-23): <https://wellcomecollection.org/works/jrkte3n> CC-BY-4.0, CC BY 4.0, <https://commons.wikimedia.org/w/index.php?curid=36461615>

Benign developmental disorders



4 month old boy, normal development. Pregnancy and labor unremarkable.

Intermittent, abnormal movement in his legs since birth.

Video

Jitteriness

Video courtesy Dr. Daniel Davila

**1 month old girl,
normal pregnancy
and delivery.**

**Mom noted
movements while
asleep.**

Video

Benign Neonatal Sleep Myoclonus

Video Courtesy Dr. Jeff Waugh

Video

4yo boy with minor speech delay. “Hand movements” often when engaged in activity (such as legos) or idle.

Movements occur mostly when :

- engaged in activity (eg, play)
- excited
- bored

Easily distractible.

Adapted from Singer H, Mink J, Gilbert D, Jankovic J. **Movement Disorders in Childhood**. 1st Ed, Philadelphia, PA: Saunders; 2010.



Variability	change over time	Generally stable
Comorbid	ADHD, Anxiety, OCD	None OR developmental disorders
Treatment	α_2 -agonists, anti-dopaminergics, topiramate, CBIT, Botox, (DBS?)	(Behavioral therapy ?)

Video

Normally developing 6 month old boy has developed these spells. Begins with sudden staring, then opening of the mouth, shivering movements. They last few seconds, abrupt onset/offset. Having up to 10 of those/day. 1. Often occurred when eating.

Adapted from: Tibussek D, Karenfort M, Mayatepek E, Assmann B. Clinical reasoning: shuddering attacks in infancy. *Neurology* 2008;70(13):e38-41.

Shuddering attacks

But... Consider
getting an EEG

Frontal lobe epilepsy with atypical seizure semiology resembling shuddering attacks or wet dog shake seizures

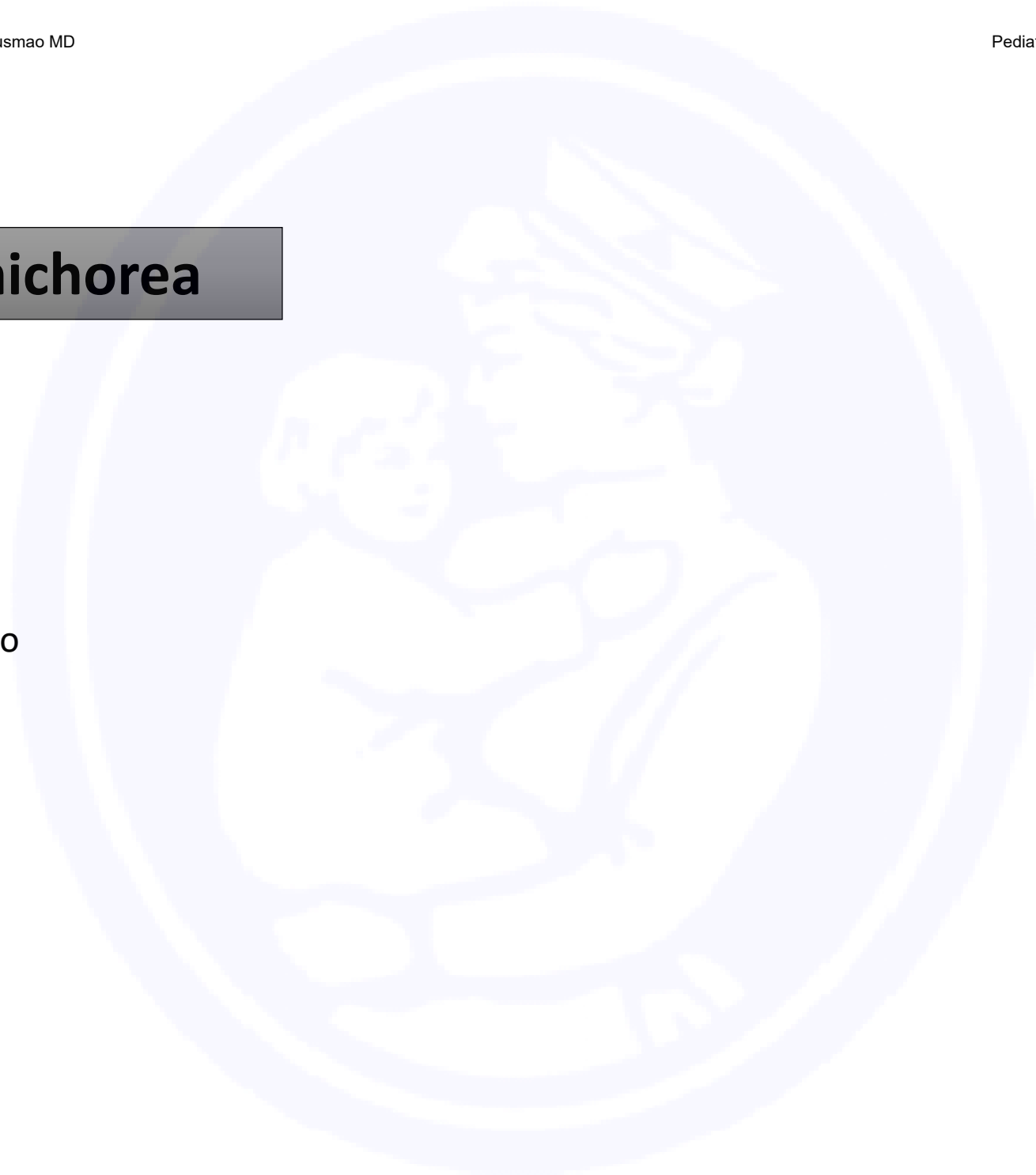
Alena Jahodova¹, Pavel Krsek¹, Vladimir Komarek¹,
Martin Kudr¹, Martin Kyncl², Josef Zamecnik³, Michal Tichy⁴

Epileptic Disord 2012; 14 (1): 69-75

Acquired immune-mediated

Hemichorea

Video

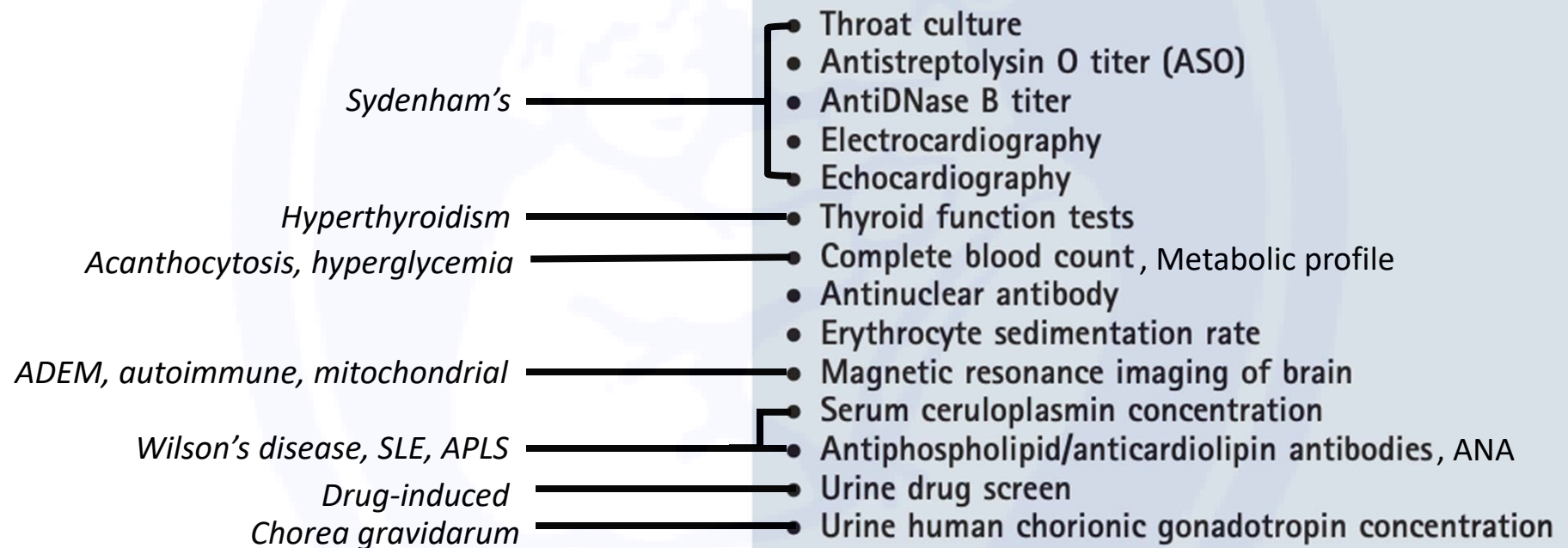


Sydenham's Chorea

- Weeks or months after GABHS infection
- May occur without carditis or arthritis
- May be asymmetric in 10-30% of cases!!
- BUT : think about neuroimaging, SLE and APLS


Chorea differential

Diagnostic Testing



Neurometabolic and genetic tests in select cases

Adapted | Mink, 2010



Genetic - treatable

Video

12 yo boy with spells of abnormal movements. They seem to occur after initiating movement or standing up from sitting position. Some dizziness before the spells.
Happening about 10 times/day. EEG was negative



東京醫科歯科大学付属病院

The attacks of this 23-year-old male had their onset at **10 years of age**, and gradually **increased in frequency** until reaching a rate of almost once each day. The attacks consisted of peculiar, purposeless, irregular **involuntary movements**, with a very **short duration**. These were **triggered by sudden movement**, and initiated from the legs sometimes spreading to the body with right-side dominance. They were preceded by an **odd sensation**, a kind of sensory aura (...) The patient had **never lost consciousness**, and abnormal neurological signs were totally absent.

Atypical case of Thomsen's disease; Kure S. **1892** Tokyo Igakukai Zasshi 6, 505-514



Paroxysmal kinesigenic dyskinesia

- Age of onset between 1-20 yo
- Triggered by movement
- Short duration (< 1min)
- No LOC or pain
- Responds to AED (eg. phenytoin, carbamazepine)



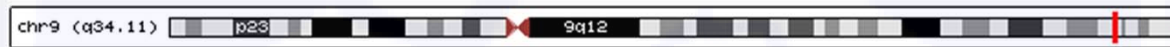
PRRT2

Bruno MK, Hallett M, Gwinn-Hardy K, et al. Clinical evaluation of idiopathic paroxysmal kinesigenic dyskinesia: new diagnostic criteria. *Neurology*; 2004;63:2280–2287.

Video

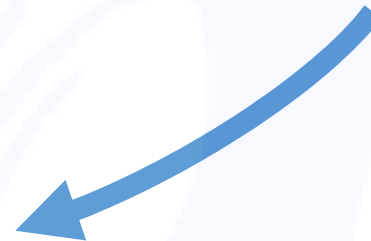
This 9 yo girl started having trouble in the summer while walking on the beach. She had a previous history of anxiety, no medications. Parents are healthy. Her father was of Ashkenazi Jewish ancestry.

TOR1A

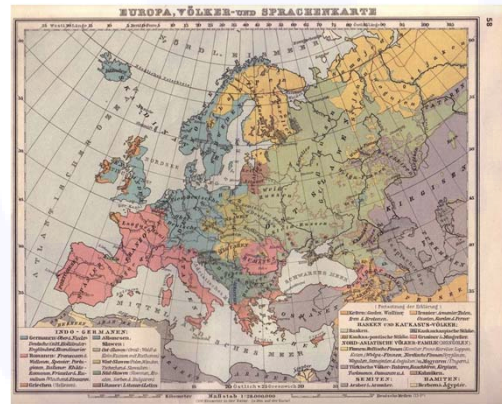
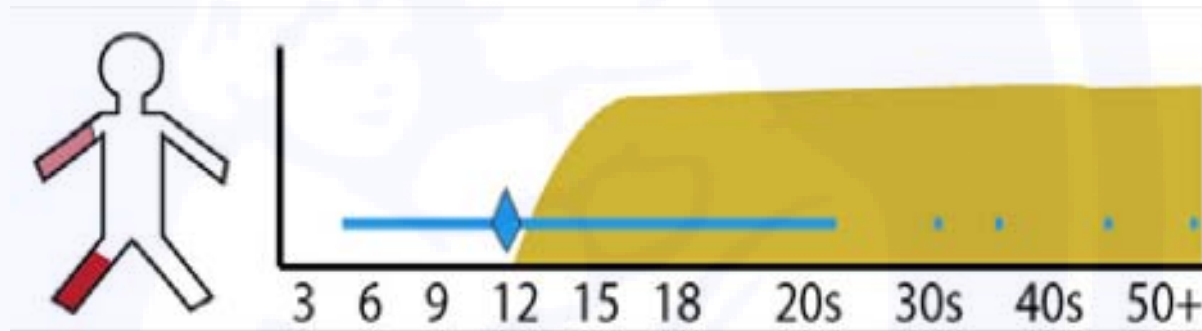


H. Gross

c907_909delGAG



DYT-TOR1A (DYT1, Oppenheim's dystonia)

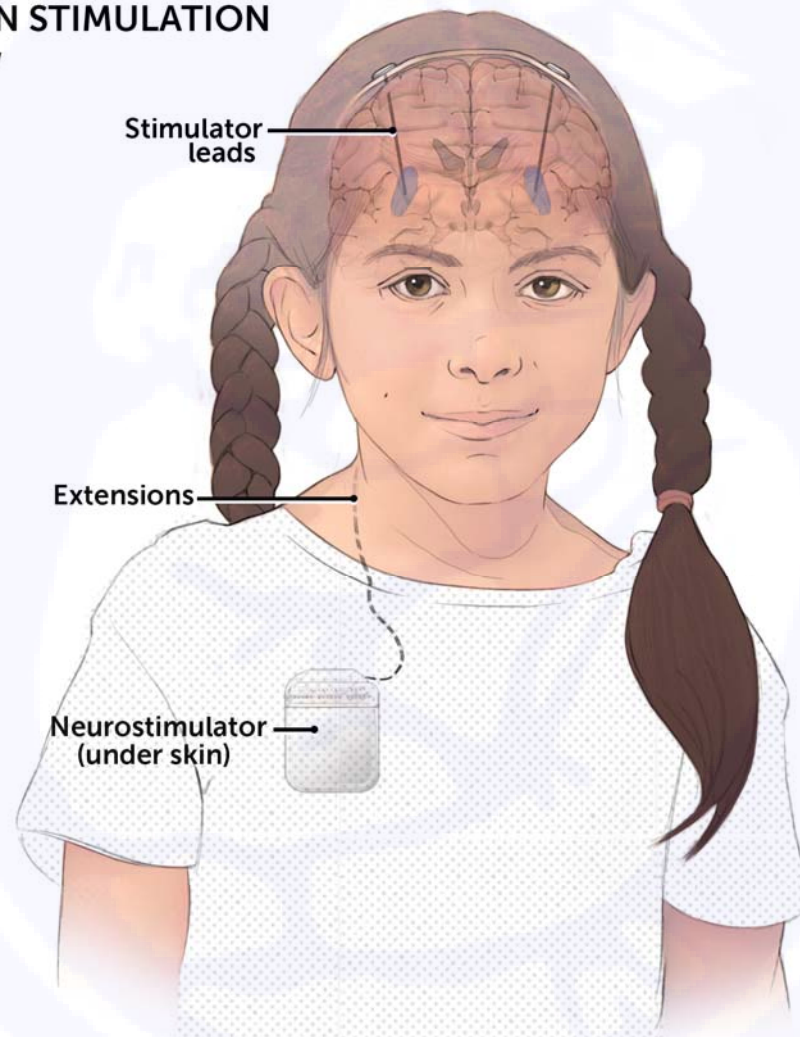


Inheritance: AD
Penetrance ~
30%

Panov, 2013
 Waugh, 2013
 Muller, 2009

16-43%

DEEP BRAIN STIMULATION OVERVIEW



<http://www.childrenshospital.org/conditions-and-treatments/treatments/deep-brain-stimulation>

1 MONTH POST SURGERY

Doctor using wireless programmer to test device



Programmer

<http://www.childrenshospital.org/conditions-and-treatments/treatments/deep-brain-stimulation>



3 months after DBS

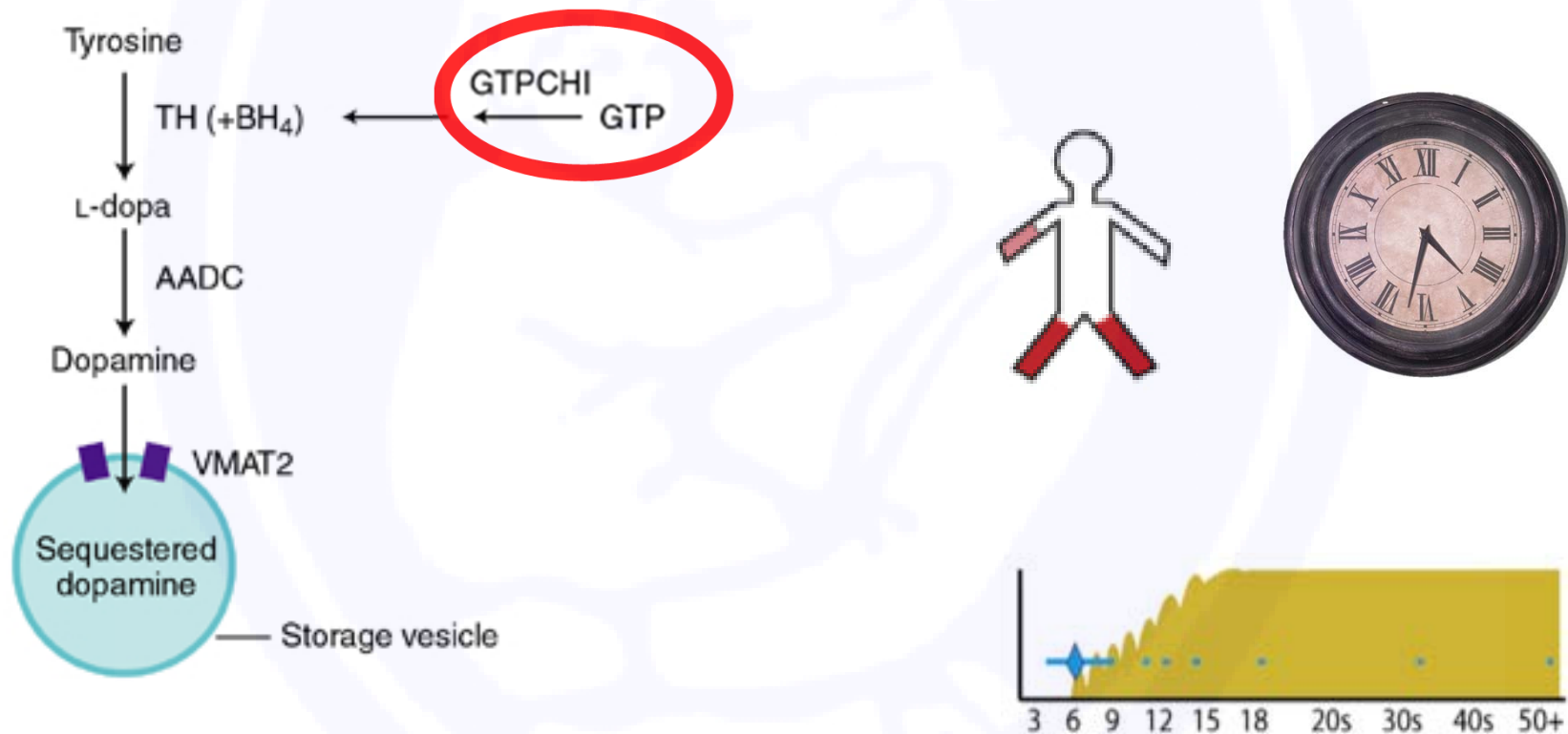
Video

Video



Adapted from Singer H, Mink J, Gilbert D, Jankovic J. **Movement Disorders in Childhood**. 1st Edition Philadelphia, PA: Saunders; 2010.

DYT-GCH1 (DYT 5 or Segawa's disease)

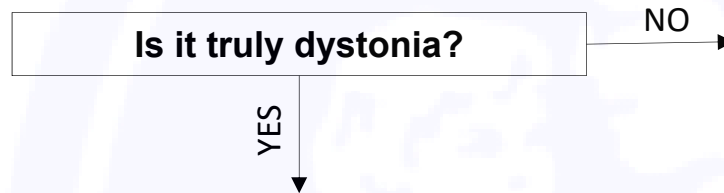


Think about dopa-responsive dystonia

- “Cerebral palsy” without a suggestive history or imaging findings
- Fluctuating symptoms
- Early-onset parkinsonism, oculogyric crises
- Dysautonomia, palpebral ptosis
- Migraines, Anxiety, Depression

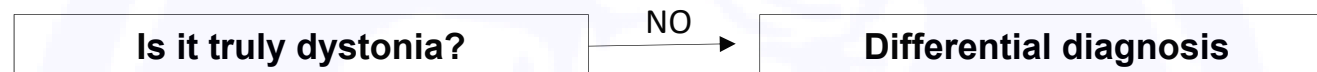
Some patients respond to Levodopa/Carbidopa even if no neurotransmitter defect is found

Diagnostic algorithm – dystonia in childhood



Adapted from van Egmond, M. E., Kuiper, A., Eggink, H., Sinke, R. J., Brouwer, O. F., Verschuuren-Bemelmans, C. C., ... De Koning, T. J. (2015). Dystonia in children and adolescents: A systematic review and a new diagnostic algorithm. *Journal of Neurology, Neurosurgery and Psychiatry*, 86(7), 774–781.

Diagnostic algorithm – dystonia in childhood



Tics

Stereotypies

Spasticity

Psychogenic/Functional

Musculoskeletal disorders

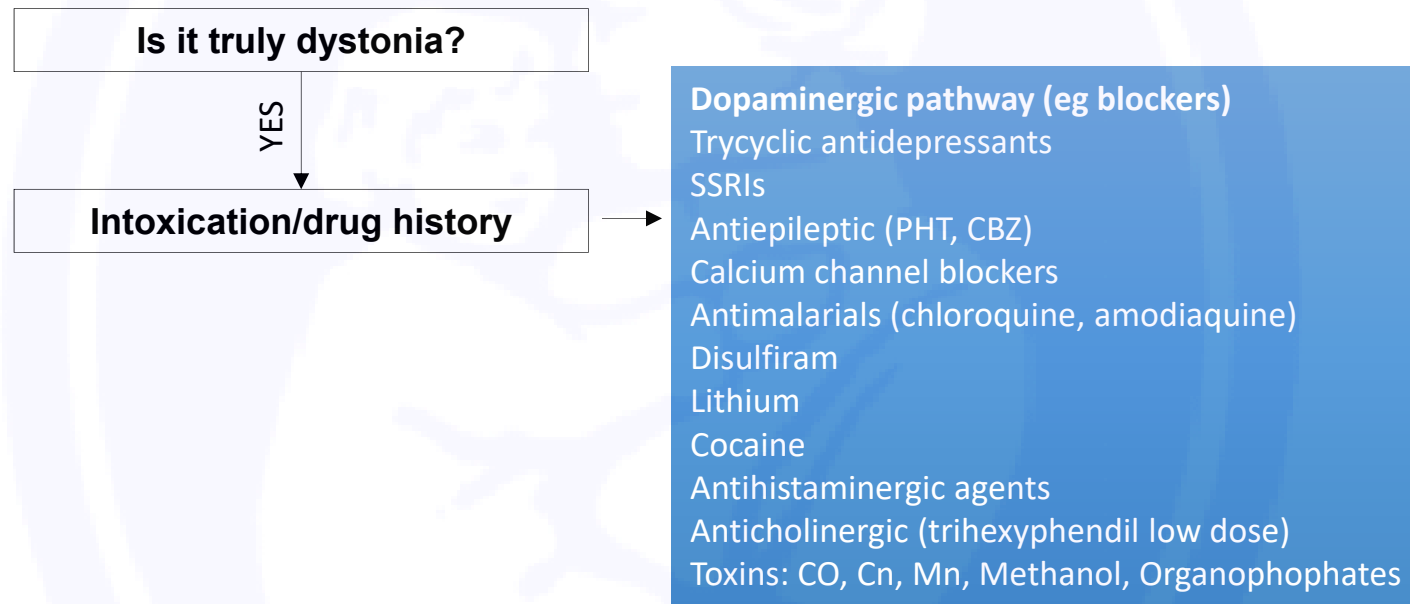
Benign developmental movements

Limb: *myotonia/neuromyotonia, Satoyoishi syndrome, tonic seizures, metabolic disturbances(Ca, Mg, pH)*

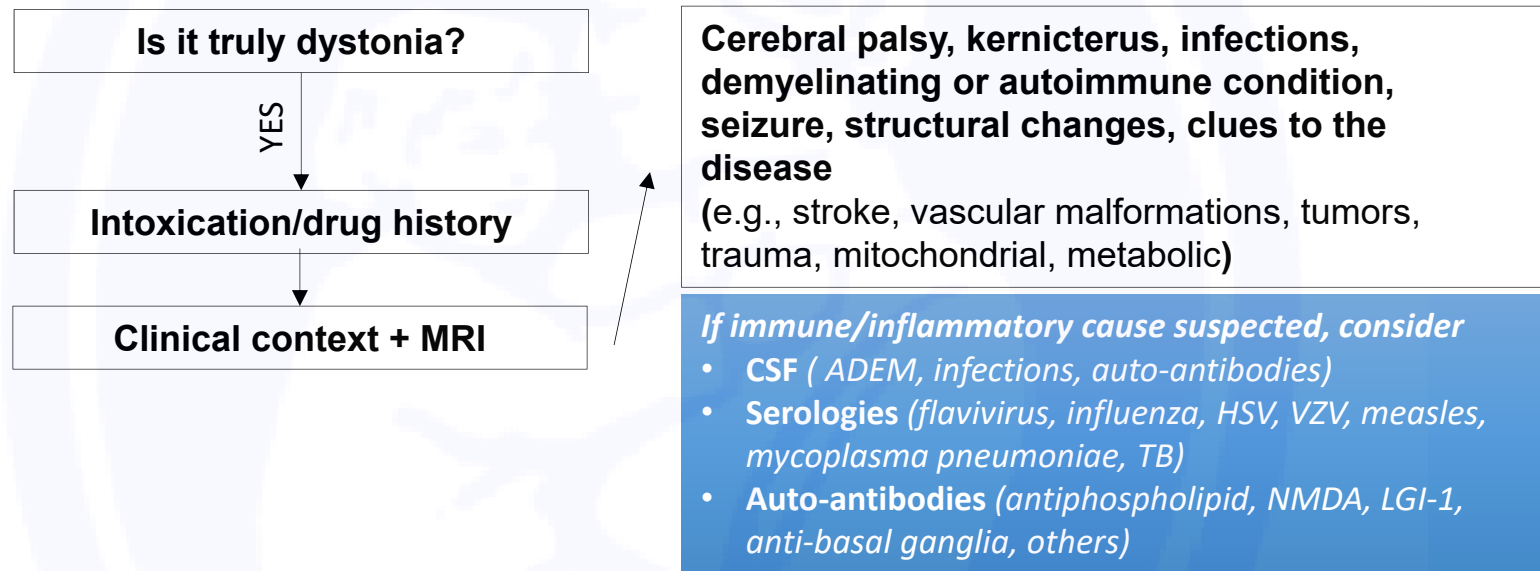
Torticollis: *cranial neuropathy (IV, VIII), spasmus nutans, posterior fossa tumor, Parinaud syndrome, Chiari I, atlantoaxial subluxation, Klippel-Feil, Syndrome de Sandifer*

Trunk: *Scoliosis, stiff person syndrome*

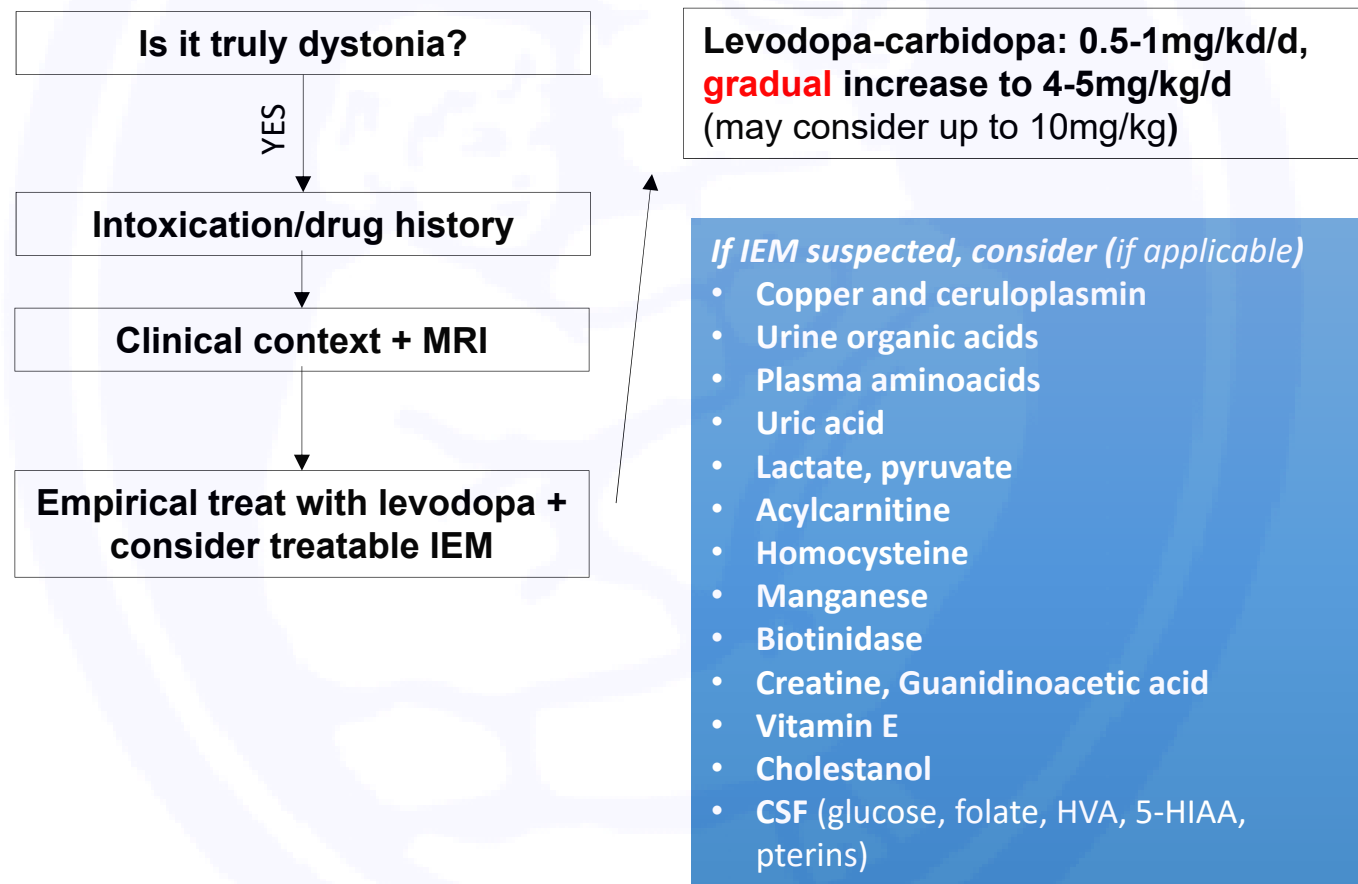
Diagnostic algorithm – dystonia in childhood



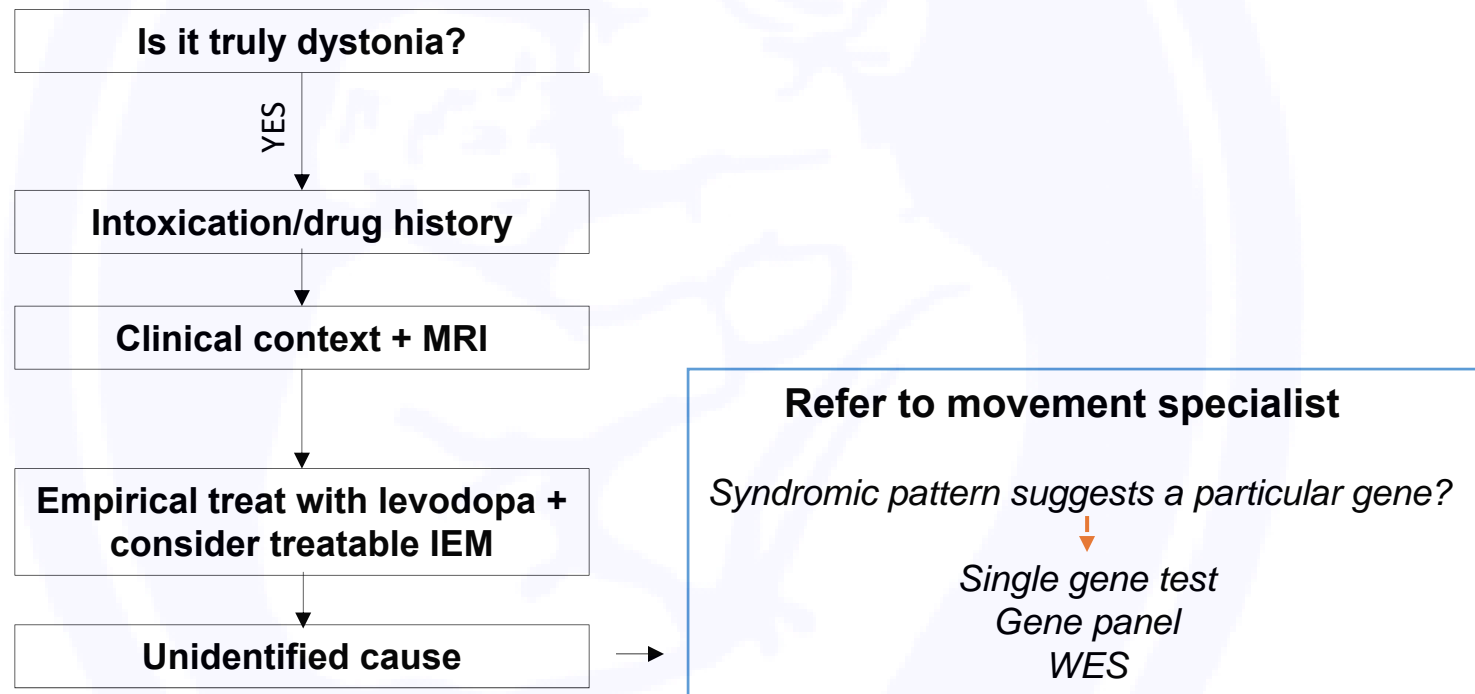
Diagnostic algorithm – dystonia in childhood



Diagnostic algorithm – dystonia in childhood



Diagnostic algorithm – dystonia in childhood



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WRAP UP



THANK YOU!

Suggested reading / references

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Movement Disorders in Childhood, 2nd Ed. Singer HS, Mink J, Gilbert DL, Jankovic J. Saunders, 2010

Principles and Practice of Movement Disorders, 2nd Ed. Fahn S., Jankovic J., Hallet M. Saunders, 2011

Koy A, Lin J-P, Sanger TD, Marks WA, Mink JW, Timmermann L. Advances in management of movement disorders in children. Lancet Neurol 2016;15(7):719–35.

van Egmond ME, Kuiper A, Eggink H, et al. Dystonia in children and adolescents: a systematic review and a new diagnostic algorithm. J Neurol Neurosurg Psychiatry 2015;86(7):774–81.

Mohammad SS, Paget SP, Dale RC. Current therapies and therapeutic decision making for childhood-onset movement disorders. Mov Disord 2019;34(5):637–56.