Emerging Orthopedic Care for Early Hip Dysplasia in Cerebral Palsy: Femoral Growth Modulation

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President, AACPDM
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October 26, 2022

NEMOURS CHILDREN'S HEALTH

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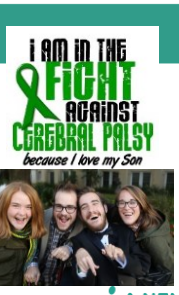
Disclosures

- Editorial Board, JAAOS, JPOSNA
- President, AACPDM
- Father of 2 young adults with CP



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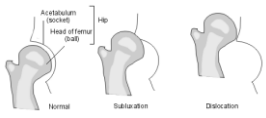
Pathology of Neuromuscular Hip Disease in CP

- NM Hip dysplasia very different than Developmental Dysplasia of the Hip (DDH)
- In CP, most babies hips are born normal
- The hips "grow out of the socket"



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Specific Anatomic Pathology of Hip Disease in CP

- Soft tissue contracture
  - Adductors and gracilis
  - Psoas
  - Weak Hip abductors and extensors
- Bony deformity
  - Neck shaft angle
  - Femoral neck anteversion

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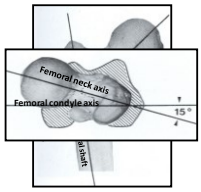
Pathology of Neuromuscular Hip Disease in CP

- Most Children with CP have normal hips at birth
- Neuromuscular dysplasia attributed to
  - Asymmetric muscle spasticity, contracture, and weakness
  - Lack of weight bearing, developmental delay, and growth
  - Bony deformity
    - Coxa Valga & Anterversion



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Radiographic Evidence of Subluxation

- Neck shaft angle
- Coxa vara, Normal, or Coxa Valga
- Head shaft angle (HSA)
  - Because the head can be in valgus relative to the neck

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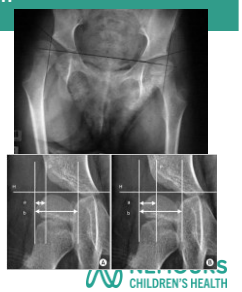
Radiographic Evidence of Subluxation

- Measure Subluxation Mostly with Reimer's index/Migration %
- Percentage of the femoral head NOT COVERED by the bony acetabulum
- Can be hard to measure with pelvic dysplasia
- Error of measurement thought to be around 5%
- "Surgical Indications" of what % vary: 30, 33, 40%....



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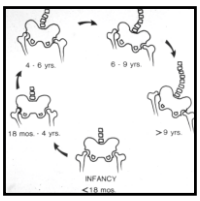
Example of Progressive Neuromuscular Hip Dysplasia in CP



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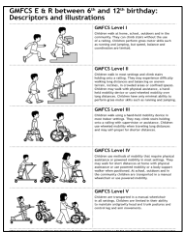
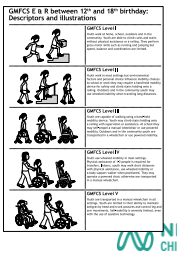
GMFCS: Gross Motor Functional Classification System





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GMFCS: Gross Motor Functional Classification System

- Valid: Based on GMFM
- Reliable
- Stable (Relatively)
- Prognostic: Predicts Natural History
- Goal Setting
- Monitoring but not Outcome Measure

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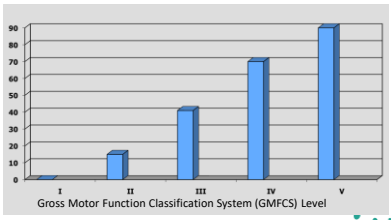
Hip Displacement (MP>30%) & GMFCS



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Soo and Howard, et al JBJS 2006

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Epidemiology: 3 Study Summary

- Hip Displacement = MP > 30%
- MP and NSA Linear relationship to GMFCS
- Not related to movement disorder
- Improve spontaneously in GMFCS I
- Usually progressive, GMFCS III to V

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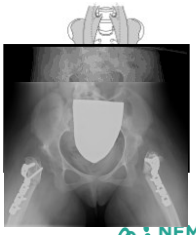
Surgical Treatment of CP Hip Dysplasia

- Preventative (Soft tissue releases)
- Reconstructive (VDRO and pelvic osteotomy
- Salvage (Resection, Arthroplasty)



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CP – Untreated Hip Dysplasia

- Untreated Hip Dysplasia in patients with CP can lead to frank dislocation over time
- Abnormal forces act on the femoral head
  - Contact/rubbing with Pelvis
  - Muscles "wearing" across cartilage surface
- Leads to degenerative joint disease and pain

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CP – Untreated Hip Dysplasia

- Most CP care providers acknowledge that the dislocated hip is more likely to be painful than the reduced hip (Shore JPO 2017)
- Salvage procedures are much less predictable than reconstruction
- Led to Formalized Hip Surveillance
- Most of us take a "preventive" strategy

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CP – Untreated Hip Dysplasia

- However, once severe hip dysplasia, hip arthritis, and pain are present, our treatment options are limited
- Pain from Hip DJD can lead to SEVERE loss of Quality of Life
- Salvage surgery is then considered

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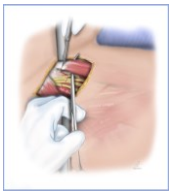
(Traditional) Preventative Hip Surgery in CP

- Indications:
  - MP > 40%
  - Age < 6
- Soft tissue releases:
  - Adductor longus
  - Iliopsoas (Fractional or Complete off the Lesser Trochanter)
  - Adductor brevis
  - Gracilis
- Obturator nerve chemo or mechanical neurolysis for GMFCS Level IV/V



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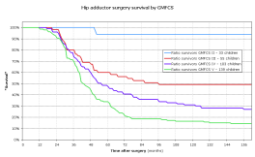
Preventative Hip Surgery in CP – Results

- duPont series showed 90% success rate for GMFCS Level II up to MP<60%
  - Less successful for higher GMFCS Levels and Higher MP
- But…..Melbourne group showed a more complete story….



Presedo A, et al, *JBJS-A*, 2005

Shore BJ, et al, *JBJS-A*, 2012

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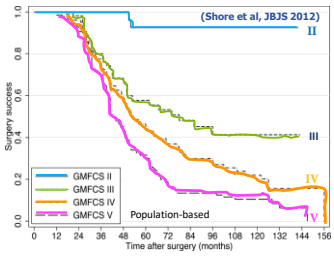
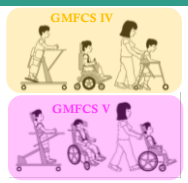
Adductors are not enough…We're stuck!





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Preventative Rx for early hip displacement

- Correction of muscle imbalance by adductor surgery alone has high failure rate
- Early reconstructive surgery (osteotomies) has high recurrence rate in < 6 yo
- Abnormal proximal femoral geometry ➜ acetabular dysplasia ➜ hip instability
- Can we modulate proximal femoral growth to reduce hip displacement?

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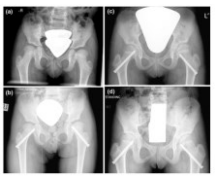
J Child Orthop (2015) 9:371–379
DOI 10.1007/s11832-015-0681-9

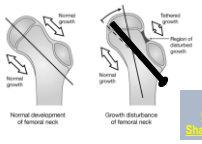
ORIGINAL CLINICAL ARTICLE

Caput valgum associated with developmental dysplasia of the hip: management by transphyseal screw fixation

Ian P. Torode[1] · Jeffrey L. Young[2]

- Type II AVN, 5-14yo
- Lateral physeal tilt
- Coxa valga ➜ subluxation
- Guided growth, reverse physeal tilt





hSA improved by 11°
Center-Edge Angle improved by 6°
Sharp's Acetabular Angle improved by 5°

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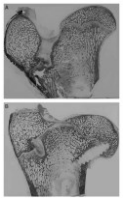
ORIGINAL ARTICLE

Guided Growth of the Proximal Femur
A Pilot Study in the Lamb Model

James J. McCarthy, MD, Kenneth J. Noonan, MD, Brett Nemke, BS, and Mark Markel, DVM



- **Basic science for guided growth: Lamb model**
- **NSA reduced to 132° vs 143° on control side**
- **"…a hemiepiphysiodesis … may be able to alter the growth and shape of the femur."**
- **"…may be of potential benefit, especially in …coxa valga… [for]children with cerebral palsy."**

Interesting but will it actually work in CP?

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Clin Orthop Relat Res (2019) 477:2568-2576
DOI 10.1097/CORR.0000000000000963

CORR 2019 Clinical Orthopaedics and Related Research

Clinical Research

Guided Growth Improves Coxa Valga and Hip Subluxation in Children with Cerebral Palsy

Hsiang-Chieh Hsieh MD, Ting-Ming Wang MD, PhD, Ken N. Kuo MD, Shier-Chieg Huang MD, PhD, Kuan-Wen Wu MD, PhD

- **24 children with CP, all GMFCS levels**
  - All with concomitant adductor sx
- **Reduction of HSA by 14° (p< 0.001)**
- **Reduction of MP by 10% (p < 0.001).**
- **Longer follow-up (r = 0.234; p < 0.001) and smaller preop MP (r = -0.258; p = 0.004) were associated with larger changes IN HSA**

NO CONTROL GROUP

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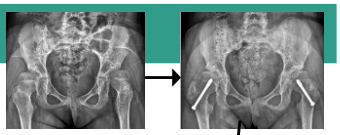
Journal of Orthopaedic Science
Volume 27, Issue 5, September 2022, Pages 1082-1088

2022

Original Article
The effectiveness of adding guided growth to soft tissue release in treating spastic hip displacement

Huan Shou, Wei C. Lee, Hsuan K. Kao, Wen E. Yang, Chia H. Chang



- **Adductor surgery (STR) + guided growth (GG) vs STR alone (comparative), *4.9y FU, age 8.9yo***
- **STR+GG had greater decrease in the MP than STR alone at 2y FU**
  - (MP; STR+GG: −14.8% vs STR: −11.8%, p < 0.05)
- **For patients w/ pre-op MP >50%:**
  - MP <40% at final FU was greater in Group GG (73%) than in Group STR (41%).

Pretty modest gains…older kids

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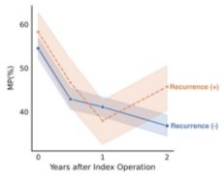
ORIGINAL ARTICLE

J Pediatr Orthop 07/2023 published ahead of print

Soft Tissue Releases With Simultaneous Guided Growth Decrease Risk of Spastic Hip Displacement Recurrence

Cheng-Min Hsu, MD, Huan Shen, MD, Wei-Chun Lee, MD, PhD, Hsuan-Kai Kao, MD, PhD, Wen-E Yang, MD, and Chia-Hsieh Chang, MD, PhD



- 66 patients with adductor releases (AR), 20 w/ +guided growth (GG)
- HSA decreased from 169° to 159° in AR+GG, no change in AR alone
- ↓Recurrence/rebound in GG group vs AR alone (39% vs 5%, p=0.012)
- ↑Recurrence/rebound and high MP risk factors for MP>40% at 2y FU

Mean age at surgery = 6.8 yo

Proximal femoral growth slow after age 5

Will younger patients have even better outcomes?

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Early Evidence for guided growth as prophylactic treatment: A Systematic Review already?

- Percutaneous placement, inferomedial physis best
- Only Level IV evidence (case series) thus far but promising
- Systematic review, 2 yr FU: Lebe et al, Children 2022
  - MP improved from 35% to 26% (p<0.01) [178 hips]
  - HSA improved from 162° to 149° (p<0.01) [178 hips]
  - AI improved more modestly, from 22° to 18° (p<0.01) [165 hips]

Mean age at surgery = 7.2 yo (range 4-12)
Older than 6 yo where hip reconstruction typical
What about younger patients?

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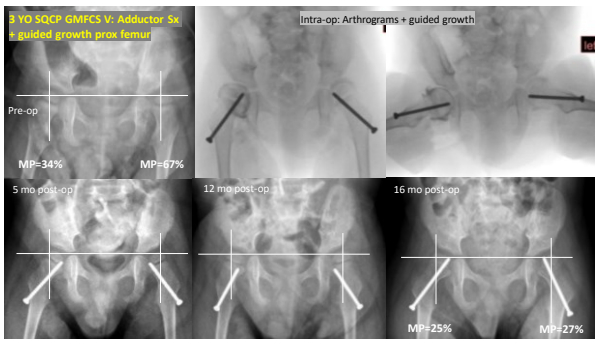

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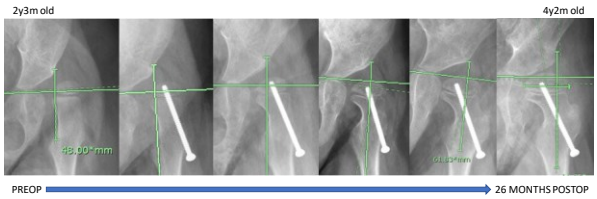
Guided growth may prevent future osteotomies for younger kids



ALSO WORKS IN HYPOTONIC CHILDREN
ONLY PROPHYLACTIC TREATMENT FOR THAT GROUP

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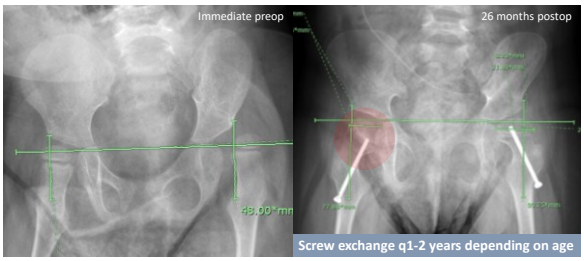
Younger patients may have better correction



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Nothing is for free….

- **Proximal femur changes w/ growth**
  - Coxa breva
  - Coxa magna
  - Acceptable trade-off?
- **Older children may have less contribution from interepiphyseal growth plate***
- **Needs further study**

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DuPont Guided Growth Indications

- **Primary treatment: MP>40% to <70%, GMFCS IV/V, 18mo to 5-6 yo, +/- adductor spasticity.**
  - Add traditional adductor, gracilis, iliopsoas releases if contractures present.
- **Secondary treatment: Rescue or prevention after VDRO, at/after time of blade plate removal.**
  - Documented lateral tilting of physis and MP progression.
  - Perhaps beneficial for early VDROs as standard to prevent rebound but unknown at this point (risk of fracture!)

UNDER INVESTIGATION

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Summary

- **Prophylactic/preventative treatment of hip displacement (HD) changing based on new/old ideas re: etiology**
- **Abnormal growth of proximal femur the key, lateral physeal tilt as per HSA secondary to abductor insufficiency/lack of WB**
- **Early treatment with guided growth of the proximal femur +/- adductor releases hold promise for younger patients with early onset HD**
- **Definitive treatment in the young vs delaying until after age 6y in older patients? Need longitudinal comparative studies (?RCT) to know for sure but early results are promising.**

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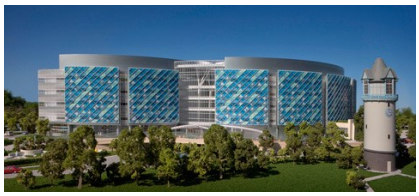
Thank You



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Delivery and Dosing of Evidence-Based Therapy Intervention in Youth with Cerebral Palsy

October 23, 2023

Amy F Bailes PT, PhD
Director of Physical Therapy Research, Division of Occupational Therapy and Physical Therapy, Cincinnati Children's Hospital Medical Center
Associate Professor Department of Rehabilitation, Exercise and Nutrition Sciences, University of Cincinnati
Amy.bailes @cchmc.org

1

- Cerebral Palsy Research Network supports some of my time to work on quality improvement initiatives for the network.
- NICHD NIH R01HD103654

2

Objectives

- Describe what is meant by dose of therapy intervention
- Reflect on how you might apply the principles of dose to your sessions
- Recognize and understand the benefits for standardizing documentation of what happens in our treatment sessions

3

What is DOSE?

Do 2 straight leg raises and call me in the morning…

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American College of Sports Medicine: Defines dose as FITT

F Frequency - how often

I Intensity – how hard you work
(Rehabilitation Intensity of therapy Scale (RITs) level of effort of the child during the session)

T Time - how long

T Type – what intervention was delivered

American College of Sports Medicine. *ACSM's Guidelines for Exercise Testing and Prescription*. 9th ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2013.

Kolobe TH, Christy JB, Gannotti ME, et al. Research summit III proceedings on dosing in children with an injured brain or cerebral palsy: executive summary. *Phys Ther*. 2014;94(7):907-920. doi:10.2522/

Saur, R. T., Corrigan, J. D., Dijkers, M. P., Barrett, R. S., Bogner, J., Smout, R. J., ... & Horn, L. D. (2015). Patient effort in traumatic brain injury inpatient rehabilitation: course and associations with age, brain injury severity, and time postinjury. *Archives of physical medicine and rehabilitation*, *96*(8), S235-S244.

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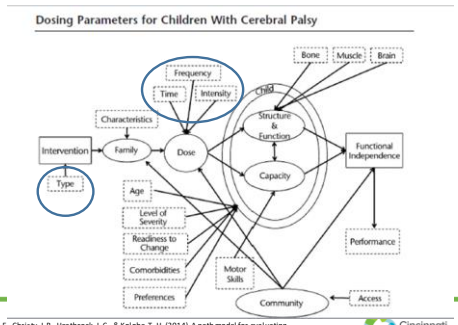
Framework



Gannotti, M. E., Christy, J. B., Heathcock, J. C., & Kolobe, T. H. (2014). A path model for evaluating dosing parameters for children with cerebral palsy. *Physical therapy*, *94*(3), 411-421.

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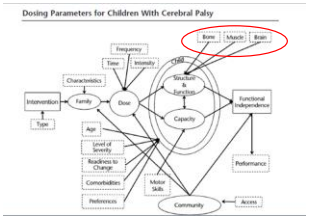
What is currently known about

- Bone
- Muscle
- Brain



Cincinnati Children's changing the outcome together

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Designing Exercise to Improve Bone Health Among Individuals With Cerebral Palsy

Mary E. Gannotti, PT, PhD; Brianna M. Liquori, MS; Deborah E. Thorpe, PT, PhD; Robyn K. Fuchs, PhD, FACSM, FASBMR

Department of Research, Shriners Hospitals for Children (Dr Gannotti and Ms Liquori), Springfield, Massachusetts; Department of Rehabilitation Services (Dr Gannotti), University of Hartford, West Hartford, Connecticut; Department of Allied Health Services (Ms Thorpe), The University of North Carolina at Chapel Hill, North Carolina; Department of Physical Therapy (Ms Fuchs), Indiana University-Purdue University, Indianapolis, Indiana.

Pediatr Phys Ther 2021;33:50–56)

- Osteogenic Index (OI) takes into account
  - Ground reaction force or load
  - Frequency or number of loading cycles
  - Number of times per week
- Age at time of intervention matters
  - Preadolescence is a critical period due to hormonal factors
- Osteogenic activities should be considered throughout the lifespan

Cincinnati Children's changing the outcome together

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Gannotti et. al. Pediatr Phys Ther 2021;33:50–56)

TABLE 2
Hierarchy of Osteogenic Activities

Examples
Highly
Squash
Tennis
Soccer
Ice hockey
Badminton/volleyball
Volleyball
Weight lifting
Moderately
Long distance running
Stair stepping
Rowing machines
Least
Walking
Swimming
Cycling
Yoga
Calisthenics

9

Gannotti et. al. Pediatr Phys Ther 2021;33:50–56)

- Bone does not adapt to loads unless they are applied in short bursts of repeated loading and unloading (standing still is not as beneficial as jumping rope)
- Short intense frequent bouts of movement throughout the day that safely load the skeleton is a challenge for clinicians and ancillary staff especially for those with severe impairment
- Takes 6 months to impact skeletal adaptation.

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Even more challenging is improving bone health for non ambulatory individuals

Systematic Review and Evidence-Based Clinical Recommendations for Dosing of Pediatric Supported Standing Programs

(Pediatr Phys Ther 2013;25:232–247)

SCOPING REVIEW

Supported-standing interventions for children and young adults with non-ambulant cerebral palsy: A scoping review

McLean, L. J., Paleg, G. S., & Livingstone, R. W. (2022). Supported-standing interventions for children and young adults with non-ambulant cerebral palsy: A scoping review. *Developmental Medicine & Child Neurology*.

Insufficient high-quality studies measuring the same outcome:

At the very least to maintain BMD, stand 1 hour day 5 days week in at least 30 degrees hip abduction , starting at 9-12 months of age

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Vibration therapy to affect bone

Original Investigation

Effect of Low-Magnitude, High-Frequency Mechanical Stimulation on BMD Among Young Childhood Cancer Survivors
A Randomized Clinical Trial

Rona J. Mogil, PhD; Sue C. Kaste, DO; Robert J. Ferry Jr, MD; Melissa M. Hudson, MD; Daniel A. Mulrooney, MD; Carrie R. Howell, PhD; Robyn E. Partin, MS; Deo K. Srivastava, PhD; Leslie L. Robison, PhD; Kirsten K. Ness, PhD

JAMA Oncology 2016

Those who participated in prescribed LMS gained total BMD where placebo group lost BMD

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Muscle

In typically developing children

Position statement on youth resistance training: the 2014 International Consensus

Rhodri S Lloyd,[1] Avery D Faigenbaum,[2] Michael H Stone,[3] Jon L Oliver,[1] Ian Jeffreys,[4] Jeremy A Moody,[1] Clive Brewer,[5] Kyle C Pierce,[6] Teri M McCambridge,[7] Rick Howard,[8] Lee Herrington,[9] Brian Hanline,[10] Lyle J Micheli,[11,12,13] Rod Jaques,[14] William J Kraemer,[15] Michael G McBride,[16] Thomas M Best,[17] Donald A Chu,[18,19] Brent A Alvar,[18] Gregory D Myer[11,20]

Lloyd RS, et al. Br J Sports Med 2014;48:498–505. doi:10.1136/bjsports-2013-092952

Resistance training can start as young as 5 years old

Abb. 2 Potential benefits of youth resistance training.

Faigenbaum AD et al. The Promise of … Bewegungstherapie und Gesundheitssport 2021; 37: 47–51 | © 2021.

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Stricker PR, et. al. AAP Council on Sports Medicine and Fitness. **Resistance Training for Children and Adolescents. Pediatrics**. 2020;145(6):e20201011

TABLE 2 Misconceptions Versus Evidence

Misconceptions	Evidence
A child is unable to increase strength before puberty.	Prepubertal children are able to gain strength by an increase in neurologic recruitment of muscle fibers, and gains in strength can be made with low injury rates if resistance training programs are well supervised with an emphasis on proper technique.
Young boys and girls may get "muscle bound" if they resistance train.	Prepubertal strength gains occur by neurologic mechanisms, and pubertal gains may augment muscle growth by actual muscle hypertrophy enhanced by pubertal hormones.
Resistance training may decrease aerobic performance in youth.	Improvements in aerobic performance have been shown with combined aerobic and resistance training programs, and combined aerobic and resistance programs do not appear to impair strength gains in children.
Resistance training may stunt growth.	Well-designed resistance training programs have not been shown to have a negative effect on physeal (growth plate) health, linear growth, and cardiovascular health in youth.
Children are stronger now than ever before.	There is a need to target strength deficits and build strength reserves due to declining measures of muscular fitness in modern-day youth.
1 RM testing is unsafe for youth.	1 RM testing may be a safe method for assessing muscular strength in youth provided that qualified supervision is present and appropriate testing guidelines are followed.

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Muscle Deficits in children with CP

Moreau 2022

INVITED REVIEW

Effects of voluntary exercise on muscle structure and function in cerebral palsy

Noelle G. Moreau[1] | Richard L. Lieber[2]

Dev Med Child Neurol. © 2022 Mac Keith Press 2022;64:700–708.

- Muscle weakness AND decreased muscle length
- Decreased ability to produce force fast or (POWER)
- Increasing strength does not necessarily translate to increased activity and participation
- Need to train power

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Power = force x velocity Moreau 2022

- Resistance training at high velocities can affect fascicle length and muscle size
- Moreau and colleagues are demonstrating that increasing power can have an effect on activity and participation (walking in the community, more)
- Must be dosed properly

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Moreau 2022

TABLE 1 Optimal dosing parameters for strength vs power training

Parameter	Intensity	Volume	Speed	Frequency	Duration	Rest
Muscle strength	70% to 85% 1RM	3 sets of 6–10 repetitions	Slow and controlled to moderate	2–3 × per wk (non-consecutive days)	8–20wks	1–2min between sets; 48h between sessions
Muscle power	60% to ≥80% 1RM	3–6 sets of 1–6 repetitions	*Concentric*: fast as possible *Eccentric*: slow and controlled over 2–3s	2–3 × per wk (non-consecutive days)	8–20wks	1–2min between sets; 48h between sessions

Reproduced with permission from Moreau.[10]
Abbreviation: 1RM, one repetition maximum.

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Show power training video

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Strength or Power?

Strength	Power
Use strength training to specifically increase underlying muscle bulk for stability or slow and controlled functional movement goals or when base level strength needs to be developed	Use power training when goals are centered around power activities such as walking speed, balance, running/jumping , standing transitional movements, stair negotiation and efficiency.

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Applying this to the School Setting:

Pediatric Physical Therapy, *34*(1), 73-80.

SPECIAL COMMUNICATION

TABLE 1
Key Differences Between Traditional SBPT Service Delivery and a PRE Model

	Traditional SBPT Delivery Model	School-Based Intensive PRE Model
Frequency	Varies, ranging from 1 time per week to 1 time per month	At least 2 times per week consistently
Intensity	Varies on the basis of student effort, subjectively determined by the SBPTs	Student works at 70%-80% of the 1RM for selected exercises
Time	Varies depending on student needs, ranges from 15 to 30 min (average: 26.7 min)[4]	Most student intervention sessions were at least doubled in minutes to complete the program protocol (ranged from 25 to 60 min depending on student needs)
Type	Typically geared toward student IEP goals, may depend on student preferences, could cover several activity limitations and impairments	Activities and exercises focused on increasing strength and power to improve functional mobility in the school environment

Abbreviations: IEP, individualized education program; 1RM, 1-repetition maximum; PRE, progressive resistance exercise; SBPTs, school-based physical therapists.

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Brain

- Guided by theories of motor control and neuroplasticity
- Timing- there are critical periods we don't' know exactly when these are but early intervention >response
- Examples
  - Constraint induced movement therapy
  - Locomotor training
  - Early Treadmill training (pre-ambulators)

Gannotti, M. E. (2017). *Pediatric physical therapy: the official publication of the Section on Pediatrics of the American Physical Therapy Association*, 29(Suppl. 3 IV STEP 2016 CONFERENCE PROCEEDINGS), 537.

21

CIMT Hoare et al. 2019

- Range of modes and setting
- Various constraints (cast, removable cast, mitt)
- 2 key ingredients restraint and intensive structured training
- Avg hours across studies = 129, range 20-504, longest period 10 weeks.

Future research focus: 1) the effect of age on the treatment effect; 2) the effect of repeated CIMT; and 3) the minimum dosage of CIMT required to impact outcomes

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Locomotor Training






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Ambulators with CP:

Evidence for locomotor training to improve gait speed

Locomotor training
on/off treadmill, with or without BWS

- Dose in general 20 training sessions 2-3 x week
- Goal of 30 minutes of walking
- Can be child active or passive
  - Younger is better
  - Active better than passive

(Novak, 2014)

- ***Recommended speed:*** *in the past, we had trained at 1 speed* Maintain good gait kinematics, no crouch if BWS,

24

Ambulators with CP:
Evidence for locomotor training, -cont.-

Dev Neurorehabil. 2019 February ; 22(2): 126–133. doi:10.1080/17518423.2018.1462270.

Short-burst interval treadmill training walking capacity and performance in cerebral palsy: a pilot study

Kristie F. Bjornson[a], Noelle Moreau[b], Amy Winter Bodkin[c]

- Typical children do not walk at one speed all the time but have bursts of fast and slow speeds
- Children with CP do **not** have bursts of fast walking like typically developing
- This has led to newer research on **short burst interval treadmill training**

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Pre Ambulators
Early Treadmill Training

- Most literature has focused on young pre ambulatory children with Ds to facilitate earlier attainment of walking
  (Damiano 2009, Fiss 2006, Valentin-Gudiol 2011, Angulo-Barroso 2008, Angulo-Barroso 2008, Looper 2010, Ulrich 2001, Ulrich 2008, Wu 2007, Wu 2008, Wu 2010, Lloyd 2010, Kokkoni 2020)
- This is important because literature in typical children suggests early walking is associated with:
  - Stronger bones (Ireland, 2014)
  - Improved language skills (Ireland 2014, Iverson 2010)

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Pre-ambulatory children with CP
Early treadmill training

- Newer literature = intensive treadmill training can also accelerate walking in pre-ambulatory children with CP younger children < 2 years old who are expected to ambulate
- SOOOOOO Earlier detection of CP allows us to identify these children that are likely to walk (i.e., GMFCS I, II, III)

[Mattern Baxter 2013, Mattern Baxter 2020]

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Pre-ambulatory

Dose of Early Treadmill training

- Showing readiness
  - Can sit independently for 30 seconds
  - Can take 5-7 steps when held over treadmill
- Start slow: 0.3 mph; increase speed, as tolerated
- ***Dose:*** 2x week for 10-20 minutes is as effective as 5x week in CP, don't know in DS if 2x week is as effective as 5x week
- Stop when taking independent steps (Mattern-Baxter, 2020)

At Cincinnati Children's, we deliver 2x week clinic and/or at home, depending on treadmill availability

- Re assess every 12 weeks, repeat episode as able; discontinue when the child can take 10 steps independently over ground

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Locomotor training Dose

Walking status	Goal	Level of Evidence	Frequency	Total Time Stepping (minutes)	Length of Episode (# sessions)	Location
Pre ambulators (Infant)	Earlier attainment of independent walking	Moderate	2x week,	As long as possible, up to 20 mins.	Reassess every 12 weeks	Home and clinic
Ambulators*	Increase speed & endurance	Moderate	2-3x week	25-30 mins.	20-24	Clinic
Non-ambulators	Provide experience of walking, weight bearing, trunk/head control	Weak	??	Up to 30 mins	No specific length; typically utilizing 12 week episodes at CCHMC	Home and Clinic

*In ambulatory children with CP: Newer evidence supports short burst interval training on treadmill to increase walking speed and endurance (Bjornson, 2019)

29

The 2021 NIH Research Plan on Rehabilitation:

National Institutes of Health (NIH) Research Plan on Rehabilitation

states that **generating consistent clinical data from ongoing care is essential to advancing the field of rehabilitation care**

What if ……

- we could change how and what we are documenting
- so that we could generate data at the point of care
- the name for this Practice Based Evidence (PBE)

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Practice Based Evidence

- A ***practice-based evidence approach*** offers a systematic method for gathering discrete information on therapy interventions and offers learnings at both the patient and population level that differ from an ***evidence-based practice*** approach.

(Horn 2012)

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EBP or PBE

Evidence Based Practice	Practice Based Evidence
• Read article and rate it • Often a limited group of patients included • May or may not apply to your clinical population	• Data is collected from actual real-life practice • All patients included • May separate data into groups to learn about different subgroups • Longitudinal and ongoing

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PBE Example: TBI

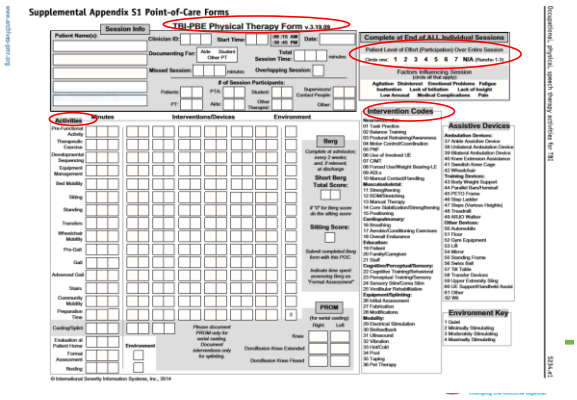
ACRM Archives of Physical Medicine and Rehabilitation

ORIGINAL RESEARCH

Occupational, Physical, and Speech Therapy Treatment Activities During Inpatient Rehabilitation for Traumatic Brain Injury

Cynthia L. Beaulieu, PhD,[a] Marcel P. Dijkers, PhD,[b] Ryan S. Barrett, MS,[c] Susan D. Horn, PhD,[c] Clare G. Gluffrida, PhD, OTR/L, FAOTA,[d] Misti L. Timpson, PT, DPT, NCS,[e,f] Deborah M. Carroll, MS, CCC-SLP,[e] Randy J. Smout, MS,[c] Flora M. Hammond, MD[g,h]

33



Supplemental Appendix S1 Point-of-Care Forms

TBI-PBE Physical Therapy Form v.3.10.09

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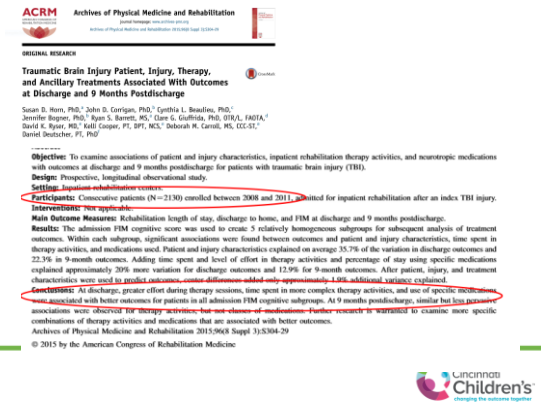

ACRM Archives of Physical Medicine and Rehabilitation

ORIGINAL RESEARCH

Traumatic Brain Injury Patient, Injury, Therapy, and Ancillary Treatments Associated With Outcomes at Discharge and 9 Months Postdischarge

Susan D. Horn, PhD,[a] John D. Corrigan, PhD,[b] Cynthia L. Beaulieu, PhD,[c] Jennifer Bogner, PhD,[d] Ryan S. Barrett, MS,[a] Clare G. Gluffrida, PhD, OTR/L, FAOTA,[d] David K. Ryser, MD,[a] Kalli Cooper, PT, DPT, NCS,[a] Deborah M. Carroll, MS, CCC-SLP,[a] Daniel Deutscher, PT, PhD[a]

Objective: To examine associations of patient and injury characteristics, inpatient rehabilitation therapy activities, and neurotropic medications with outcomes at discharge and 9 months postdischarge for patients with traumatic brain injury (TBI).

Design: Prospective, longitudinal observational study.

Setting: Inpatient rehabilitation centers.

Participants: Consecutive patients (N=2130) enrolled between 2008 and 2011, admitted for inpatient rehabilitation after an index TBI injury.

Interventions: Not applicable.

Main Outcome Measures: Rehabilitation length of stay, discharge to home, and FIM at discharge and 9 months postdischarge.

Results: The admission FIM cognitive score was used to create 5 relatively homogeneous subgroups for subsequent analysis of treatment outcomes. Within each subgroup, significant associations were found between outcomes and patient and injury characteristics, time spent in therapy activities, and medications used. Patient and injury characteristics explained on average 35.7% of the variation in discharge outcomes and 22.3% in 9-month outcomes. Adding time spent and level of effort in therapy activities and percentage of stay using specific medications explained approximately 20% more variation for discharge outcomes and 12.9% for 9-month outcomes. After patient, injury, and treatment characteristics were used to predict outcomes, some differences added only approximately 1.0% additional variance explained.

Conclusions: At discharge, greater effort during therapy sessions, time spent in more complex therapy activities, and use of specific medications were associated with better outcomes for patients in all admission FIM cognitive subgroups. At 9 months postdischarge, similar but less pervasive associations were observed for therapy activities, but not classes of medications. Further research is warranted to examine more specific combinations of therapy activities and medications that are associated with better outcomes.

Archives of Physical Medicine and Rehabilitation 2015;96(8 Suppl 3):S304-29

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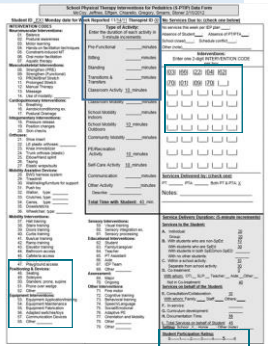
Pediatric Example: PT COUNTS

- Effgen SK, Westcott McCoy S, Chiarello LA, Jeffries LM, Bush H. Physical therapy-related child outcomes in school: an example of practice based evidence methodology. *Pediatr Phys Ther*. 2016;28(1):47-56. doi:/PEP.0000000000000197.
- https://www.uky.edu/chs/academic-programs/department-rehabilitation-sciences/physical-therapy/pt-counts



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What school therapists are delivering

Jeffries 2019 *Physical therapy*, *99*(1), 98-108.

Methods: PBE school therapists completed paper form weekly on what they delivered to 5-12 year old children over an academic year (mostly with CP)

TIME: More severely involved children received more sessions or minutes

TYPE: The most frequent over the year were in neuromuscular, mobility, and musculoskeletal; and the least frequent interventions were positioning, equipment, cardiopulmonary, sensory, and integumentary.

Limited use of some EBIs in the school setting, (constraint-induced movement therapy, body weight–supported treadmill training, and cardiopulmonary/fitness interventions).

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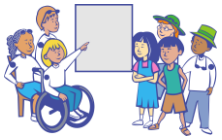
What school therapists are delivering

McCoy 2018 *Developmental Medicine & Child Neurology*, *60*(11), 1140-1148.

Active mobility interventions and increased child effort related to better outcomes on School Function Assessment



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Opportunity for us to engage

- Are you documenting the important features of dose from the intervention session?
- Are we measuring what we are doing?
- How can we use data we collect to improve our knowledge about dose and improve care?

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Limitations to current documentation

- There are limitations to using current documentation and billing codes.
- Need more details to learn how each variable contributes to outcomes, what matters most, do variables interact ?
- So that we can deliver the right intervention to the right person at the right time.

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So, lets get FITT

- F — Frequency: how often
- I — Intensity: Rehab intensity scale/child effort
- T — Time: duration of intervention
- T — Type: kind of intervention

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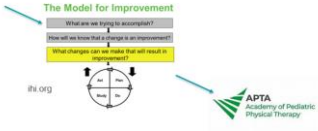
How can we do this?



Langley G, et al. *The Improvement Guide: A Practical Approach to Enhancing Organizational Performance.* 2nd ed. San Francisco, CA: Jossey-Baas; 2009

42

The Village

- National team that adapted PT Counts form: Amy Bailes, Adam Brown, Danni Bellows, Mary Gannotti, Chris Joseph, Maureen Nahormiak, Lisa Steed and Andi Todd
- Posted on NINDS Common Data elements https://commondataelements.ninds.nih.gov/ Physical therapy individual session form
- Locally tested and adapted through PDSA cycles use of individual PT session form into an EPIC flowsheet→ allows for comprehensive capture of PT dose (Bailes et al. 2019)

  Funded by the Academy of Pediatric Physical Therapy Research Grant
- Created an electronic database specific to details of PT intervention

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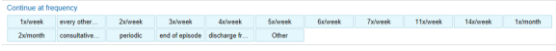
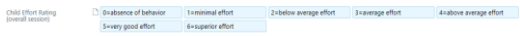
PT FITT flowsheet

Frequency: How often you do intervene



Intensity: Child's level of effort towards meeting the session objectives (as rated by you)



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Rehabilitation Intensity of Therapy Scale (RITS)

Records an overall rating for "Child Effort" across the entire session by circling a number on the visual analogue scale; choose the number that best fits your initial impression (this is, in essence, recording what you believe to be the child's effort/intensity in the session towards meeting the objectives of the session):

0= absence of effort
1= minimal effort
2=below average effort
3=Average effort
4=above average effort
5=very good effort
6=superior effort

Should reflect normal distribution of the population, a score of 0 or 6 is uncommon. Most people tend to fall into the categories of 2, 3, and 4 **where 3 would be average effort**.

- This is not a measure of the patient's ability! **Choose the number that best** fits what you observed the patient DO - not whether they did their "best"

Several examples in the literature
Seel, R. T., (2015). *Archives of physical medicine and rehabilitation*, *96*(8), S235-S244.
Beaulieu, C. L., (2015). *Archives of physical medicine and rehabilitation*, *96*(8), S222-S234.
Effgen, S. K., (2016). *Pediatric Physical Therapy*, *28*(1), 47-56.

45

PT
TIME and TYPE

Time (minutes) spent in each focus area:

- Pre-functional/Preparatory
- Sitting
- Standing
- Transitions/Transfers
- W/C mobility
- Gait
- Fitness/Health/Rec Management
- Gross Motor/Developmental
- Pain/Effusion
- Formal Assessment
- Other

Intervention Type :
select choices within categories

- Test/assessment
- Neuromuscular
- Musculoskeletal
- Modalities
- Adaptive Equipment/Orthotic Management
- Casts/Orthoses/Prosthetics/ Supports
- Equipment
- Assistive Tech
- Cardiopulmonary
- Integumentary
- Education/Training
- Other

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OT TIME Focus Areas

- Pre-functional/preparatory
- Pain/effusion
- Bathing
- Toileting
- Dressing
- Grooming/hygiene
- Eating swallowing
- Feeding
- Functional Mobility/Transfers
- Household Chores
- Meal Prep
- Safety Maintenance
- Rest and Sleep
- Education School
- Play Leisure
- Social Participation
- Health Management Fitness
- Formal Assessment

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OT Intervention TYPE categories

- Assessments
- Neuromuscular (includes things like CIMT, Bimanual, Fine motor dexterity etc)
- Musculoskeletal
- Cardiopulmonary
- Sensory-Perceptual
- Visual
- Mental Functions
- Emotional Regulation(psych only)
- Rehearsal of daily life activities
- Modalities
- Integumentary
- Adaptive Equipment/orthotic/prosthetic management
- Casts/orthoses/prosthetics/supports
- Equipment
- Assistive technology/devices
- Education/training

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Let me show you an example: 9 year old with CP, GMFCS I

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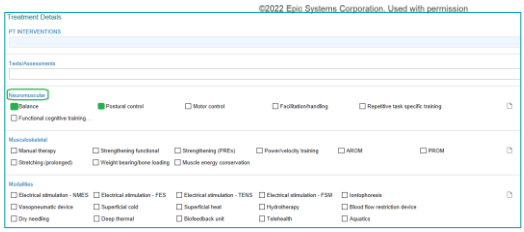
Example- Intervention Type



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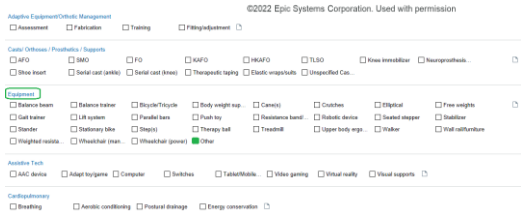
Example: Intervention Type, -cont.-



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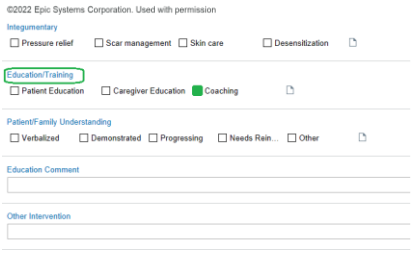
Example: Intervention Type, -cont.-



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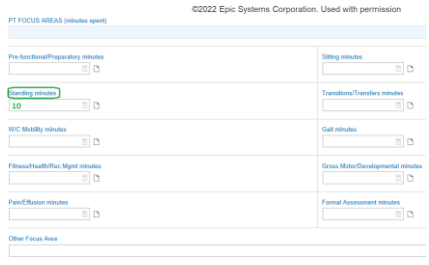
Example: Time (Focus Areas)



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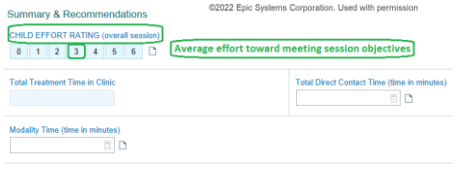
Example: Intensity (Child Effort Rating)



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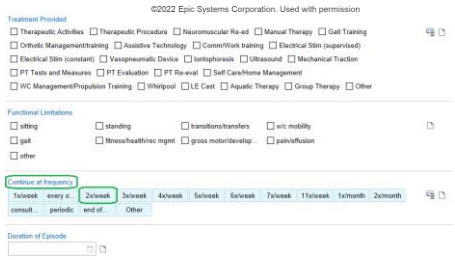
Example: Frequency



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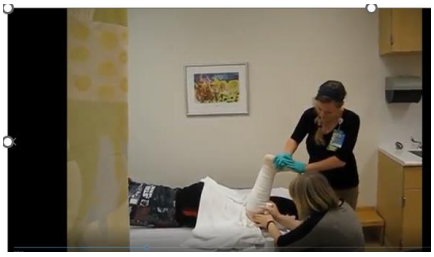
Another example: 18 y.o. GMFCS I



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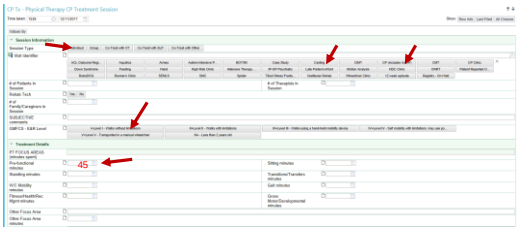
Documentation



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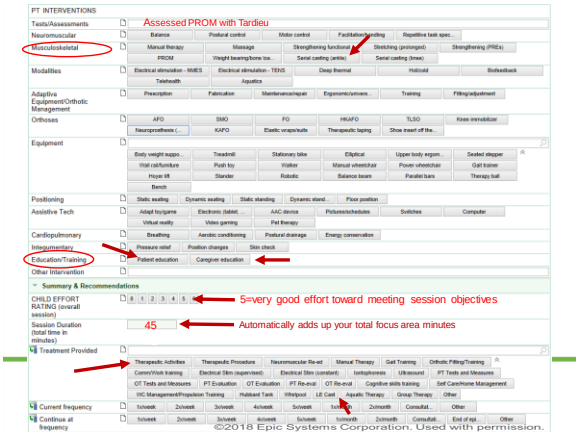

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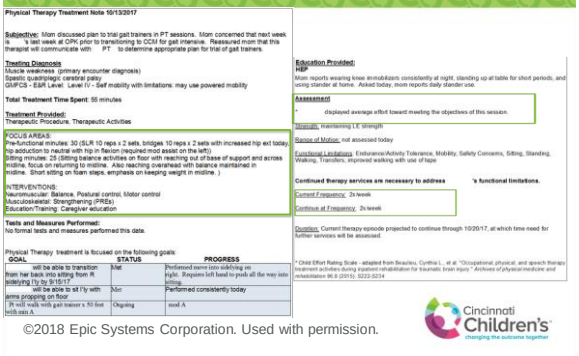
Abracadabra!



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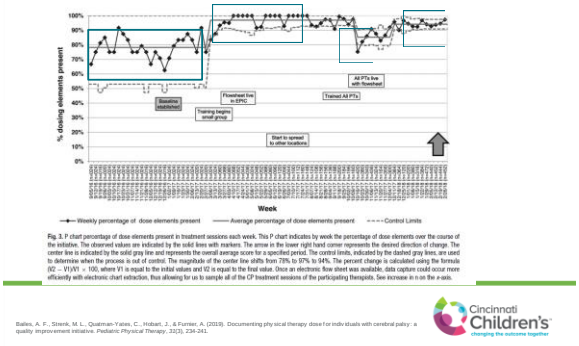
% of dose elements (FITT) present in treatment notes each week



Bailes, A. F., Strenk, M. L., Quatman-Yates, C., Hobart, J., & Fiorilli, A. (2019). Documenting physical therapy dose for individuals with cerebral palsy: a quality improvement initiative. *Pediatric Physical Therapy, 31*(2), 234-241.

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What are we learning and spreading

Outpatient DD PT and OT

Inpatient PT and OT

Sports PT, Home Health OT&PT Psych OT

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Cerebral Palsy: Percent of evidence-based interventions (EBI) per person

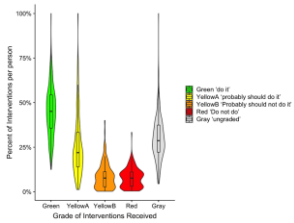
465 individuals with CP over 1 year

4335 treatment visits

28, 344 interventions delivered

48% green "do it"

18% yellow" probably should do"



Bailes, A. F., Greve, K., Long, J., Kurowski, B. G., Vargus-Adams, J., Aronow, B., & Mitelpunkt, A. (2021). Describing the Delivery of Evidence-Based Physical Therapy Intervention to Individuals With Cerebral Palsy. *Pediatric Physical Therapy*, *33*(2), 65-72.

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Delivery of EBI continued

Most frequently delivered EBIs

- Caregiver education
- Motor control
- Functional strengthening
- Ankle foot orthoses
- Treadmill training
- Adaptive equipment fitting

Room for improvement

- Low volume of fitness interventions in outpatient setting

Bailes, A. F., Greve, K., Long, J., Kurowski, B. G., Vargus-Adams, J., Aronow, B., & Mitelpunkt, A. (2021). Describing the Delivery of Evidence-Based Physical Therapy Intervention to Individuals With Cerebral Palsy. *Pediatric Physical Therapy*, *33*(2), 65-72.

Cincinnati Children's

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Delivery of EBIs continued

- Individuals GMFCS V received lowest counts of EBI, GMFCS III the most
- Individuals GMFCS IV more than V but not different than other Levels.
- Different than school, individuals at GMFCS I did not receive lowest counts
- No difference for age categories.

Could not study relationship with outcomes due to variety of measures administered to the children and time points throughout the year

Bailes, A. F., Greve, K., Long, J., Kurowski, B. G., Vargus-Adams, J., Aronow, B., & Mitelpunkt, A. (2021). Describing the Delivery of Evidence-Based Physical Therapy Intervention to Individuals With Cerebral Palsy. *Pediatric Physical Therapy*, 33(2), 65-72.

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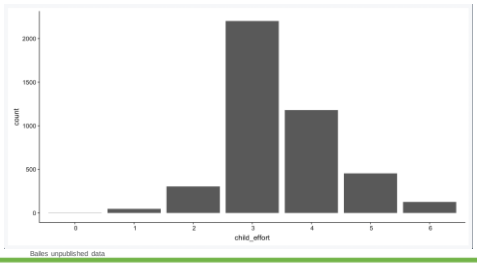
Intensity varied over the 4335 sessions



Bailes unpublished data

65

Variation in physical therapy dose after single event multi level surgery in children with CP **CSM 2022 Poster:**
Greve K, Bailes A, Long J, Aronow B, Zhang N, Mitelpunkt A

17 children with CP (mean 9 yr)
- 10 ambulatory (GMFCS I-III)
- 12 high surgical burden (> 2 osteotomies).

Ambulatory children vs non ambulatory

F: greater frequency of visits (231 vs. 114, p<0.001)
I: higher intensity (5 vs. 3, p<0.001)
T: more time (minutes) in pre-functional activities (5899 vs. 1975, p=0.000) and gait (4138 vs. 1318, p=0.005).
T: Intervention type did not differ by ambulatory status

No differences in FITT by surgical burden

Abstracts of the Academy of Pediatric Physical Therapy Poster Presentations at the Combined Sections Meeting, Pediatric Physical Therapy: January 2022 - Volume 34 - Issue 1 - p 97-140
doi: 10.1097/PEP.0000000000000860

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Low Back Pain: CSM 2023 Poster PT Intervention Delivered to Children with Low Back Pain Hobart J, Strenk, M., Allen M., Hugentobler, K. Bailes AF.

Objectives:

- Characterize the population
- Dose: Frequency and Intervention types
- How does treatment for children compare to adult guideline?

http://commons.wikimedia.org/wiki/File:Backbone_%28PSF%29.png?uselang=de

Cincinnati Children's changing the outcome together

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Characteristics of Children with LBP over 6-month period

CSM 2023 poster Hobart J, Strenk, M., Allen M., Hugentobler, K. Bailes AF.

Table 1: Characteristics of Individuals Seen in Outpatient Physical Therapy Division for Low Back Pain during the 6-month Study Period (7/1/21-12/31/21)

	n=274
Age (years)	
Mean (SD)	15 (2.0)
Gender, female, n (%)	187 (68.2)
Race, n (%)	
White	204 (74.5)
Black or African American	50 (18.2)
Ethnicity, n (%)	
Non-Hispanic	262 (95.6)
Total Number of Visits	1480
Number of Visits per Participant	
Mean (SD)	9 (5.4)
Median (range)	8 (1-24)

Cincinnati Children's changing the outcome together

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LBP TYPES

Adult guideline for low back pain

George *J Orthop Sports Phys Ther.* 2021

Table 2: Description of Delivered PT Intervention Type Grouped by Evidence Grade

Interventions by CPG Recommendation	Flow Sheet Intervention Category	Flow Sheet Intervention Type	Intervention Type, n (%)
Should Use			5427 (71.8)
Patient Education	Education	Patient Education	1164 (15.4)
General exercise	Musculoskeletal	Strengthening functional	1038 (13.7)
Trunk muscle strengthening & endurance exercise	Musculoskeletal	Strengthening (PREs)	1025 (13.6)
Specific trunk muscle activation exercise	Neuromuscular	Motor control	907 (12.0)
Specific trunk muscle activation exercise	Neuromuscular	Postural control	860 (11.4)
Movement control exercise	Neuromuscular	Balance	216 (2.9)
Trunk muscle strengthening & endurance exercise	Musculoskeletal	Power/velocity training	92 (1.2)
Thrust or nonthrust joint mobilization	Musculoskeletal	Manual Therapy (thrust or nonthrust joint mobilization)	65 (*)
Movement control exercise	Neuromuscular	Repetitive task specific training	32 (*)
Aerobic exercise	Cardiopulmonary	Aerobic conditioning	28 (*)
Aquatic exercise	Modality	Aquatic	0 (*)
May Use			1122 (14.9)
Trunk mobility exercise	Musculoskeletal	Stretching	802 (10.62)
Trunk mobility exercise	Musculoskeletal	Active Range of Motion	230 (3.04)
Soft tissue mobilization	Musculoskeletal	Manual Therapy (soft tissue mobilization)	90 (1.19)
Massage	Musculoskeletal	Massage	0 (*)
Can Use			61 (*)
Dry needling	Modalities	Dry needling	61 (0.81)
Unlisted			945 (12.5)
	Modalities	Superficial cold	51 (*)
		Superficial heat	33 (*)
		Electrical stimulation (all)	31 (*)
		Iontophoresis	7 (*)
		Blood Flow Restriction	3 (*)
		Vasopneumatic device	1 (*)
	Musculoskeletal	Weight bearing/Bone loading	89 (1.2)
		Passive Range of Motion	81 (1.1)
		Positioning	35 (*)
	Neuromuscular	Facilitation/handling	9 (*)
	Cardiopulmonary	Breathing	74 (*)
	Adapt Equip Ortho Mgmt	Fitting/adjustment	3 (*)
		Fabrication	1 (*)
		Training	1 (*)
	Education	Caregiver education	502 (6.6)
		Coaching	24 (*)
Total Count of Interventions Delivered			7555

*Less than 1%. Sum of percentages do not equal 100 due to rounding.

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LBP Types: Primarily aligned with the adult recommendations

CSM 2023 poster Hobart J, Strenk, M., Allen M., Hugentobler, K. Bailes AF

- Majority (71.8%) of the intervention types delivered were mapped to the "Should Use"
- Most common "Should Use" interventions delivered
  - patient education
  - functional strengthening
  - progressive resisted exercise strengthening
  - motor control.
- Least frequent "Should Use" interventions delivered
  - manual therapy (thrust or non thrust joint mobilization)
  - repetitive task specific training
  - aerobic conditioning
  - aquatic therapy

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Ongoing clinician engagement

- Share findings regularly
- Elicit feedback on what we are learning
- Measure agreement among therapists every 6 months

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Next

- Continue our work in CP how dose relates to outcomes
- All conditions and therapists at Cincinnati (OT and PT), including inpatient, outpatient, mental health, and home health
- Discrete fields allow us to track delivery of EBI we want to increase (CIMT, early treadmill training, power training and others)

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Next

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Thank you

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With technical support from Julie Badylak, Jason Long, Jessie Hobart (student), and Sarah Bidwell CRC.

Susan Horn PhD.
PT counts: University of Kentucky

- FITT flowsheet is available at EPIC community library.

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Therapy and Rehabilitation Medicine Strategies to Promote Independence in Activities of and Instrumental Activities of Daily Living for Youth with Disabilities

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Nemours Children's Health Cerebral Palsy Conference for Pediatric Therapists 2023

Monday, October 23, 2023

NEMOURS CHILDREN'S HEALTH

1

What is occupation and how does it pertain to this discussion?

As defined by the World Federation of Occupational Therapists, the term *occupation* "refers to the everyday activities that people do as individuals, in families, and with communities to occupy time and bring meaning and purpose to life. Occupations include things people need to, want to and are expected to do."[1]

This is central to the work of an occupational therapist, often forming the basis for goals that are meaningful and relevant to the child/adolescent/adult and parent/caregiver.

2

Occupational Therapy Practice Framework

The 4th edition of the *Occupational Therapy Practice Framework: Domain and Process (OTPF – 4)* identifies a broad range of occupations [2]

Activities of daily living (ADL) – activities oriented toward taking care of one's own body and completed on a routine basis

Showering, toileting, dressing, eating, feeding, functional mobility, personal hygiene, sexual activity

Instrumental Activities of Daily Living (iADL) – activities to support daily life within the home and community

Care of others, care of pets, child rearing, communication management, driving & community mobility, financial management, home establishment and management, meal preparation & clean up, religious and spiritual expression, safety management, shopping

3

Occupational Therapy Practice Framework

The 4th edition of the Occupational Therapy Practice Framework: Domain and Process (OTPF – 4) identifies a broad range of occupations [2]

Health Management – activities related to developing, managing, and maintaining health and wellness routines

Social & emotional health promotion and maintenance, symptom and condition management, communication with health care system, medication management, physical activity, nutrition management, personal care device management

Rest and Sleep – activities related to obtaining restorative rest and sleep to support healthy, active engagement in other occupations

Rest, sleep preparation, sleep participation

4

Occupational Therapy Practice Framework

The 4th edition of the Occupational Therapy Practice Framework: Domain and Process (OTPF – 4)[2] identifies a broad range of occupations [2]

Education – activities needed for learning and participating in the educational environment

Formal education participation, informal personal education needs or interests exploration (beyond formal education), information educational participation

Work – labor or exertion related to the development, production, delivery or management of objects or services

Employment interests and pursuits, employment seeking and acquisition, job performance and maintenance, retirement preparation and adjustment, volunteer exploration and participation

5

Occupational Therapy Practice Framework

The 4th edition of the Occupational Therapy Practice Framework: Domain and Process (OTPF – 4)[2] identifies a broad range of occupations [2]

Social Participation – activities that involve social interaction with others, including family, friends, peers and community members and that support social interdependence

Community participation, family participation, friendships, intimate partner relationships, peer group participation

Play – activities that are intrinsically motivated, internally controlled and freely chosen and that may include suspension of reality

Play exploration & participation

Leisure – non-obligatory activity that is intrinsically motivated and engaged in during discretionary time

Leisure exploration & participation

6

Occupation: What is it and how does it pertain to this discussion?

- Occupational Therapy (OT) may not address each occupation specifically in treatment, as another discipline may be suited to address the issue in greater depth.
- This evaluation process may bring specific aspects of an adolescent's participation in occupations to light.
- Well-chosen outcome measures, in the form of patient and parent-reported outcomes, can be one tool used to guide the conversation.

These outcomes are intended to guide goal setting and priorities in treatment

- Parent-reported and patient-reported outcomes

Family-centered care (FCC) framework [3]

7

Patient-Centered Care & Meaningful Occupations

Patient-Centered Care: "an individual's specific health needs and desired health outcomes are the driving force behind all health care decisions and quality measurements. Patients are partners with their health care providers, and providers treat patients not only from a clinical perspective, but also from an emotional, mental, spiritual, social, and financial perspective." [4]

Occupational therapy plan of cares should always include goals related to occupations that are meaningful to both the patient and their parent or caregiver. [5]

8

Plan of Care Focused on Meaningful Occupations

Adolescence is a time where individuals are beginning to find their own identity so encouraging patients to decide which occupations are most important for them to work on is crucial to helping foster their self-identity. [6]

Patients have increased engagement in therapy and make increased progress when they are interested and motivated during therapy sessions.

Families and patients demonstrate increased carry over when they are working on occupations that are meaningful to them. [7]

9

Facilitating Collaborative Conversations

Build rapport by asking about current interests and hobbies

Ask about daily routine including ADLs, school, leisure, and anything else that may be important to the patient and their family

Ask guiding questions about occupations that they have brought up including their strengths and areas of improvement for those occupations

The two parent- and patient-reported outcomes highlighted have similarities yet may be used for different purposes and at different times

10

Canadian Occupational Performance Measure (COPM) – 5th edition [8]

- The COPM-5 is an outcome measure designed to detect change in an individual's self-perception of occupational performance in areas of self-care, productivity and leisure.
- Used primarily by occupational therapists with individuals ages 8+ at the start of skilled intervention to establish intervention goals and at the conclusion of intervention to determine the progress and outcome.
- Guides conversation through a 5-step process
  1. Identify occupational performance "problems"
     - "problem" is an occupation that a person wants to do, needs to do or is expected to do much can't do, doesn't do or is not satisfied with the way he/she does it.
  2. Once occupational performance problems have been identified, individual is asked to rate each in terms of it's importance in his/her life
     - 1 = not important at all, 10 = extremely important
  3. Individual is asked to choose up to 5 problems that seem most important

11

Canadian Occupational Performance Measure (COPM) – 5th edition [8]

- Guides conversation through a 5-step process
  4. Individual is asked to rate performance
     - "How would you rate the way you do this activity now?"
     - "How satisfied are you with the way you do this activity now?"
  5. Establish a date for re-assessment

12

(CP-CHILD) Caregiver Priorities and Child Health Index of Life with Disabilities [9]

- Reliable and valid measure of caregiver's perspectives on health status, functional limitations and well-being of children with severe cerebral palsy.
- Six domains are considered using this measure
  - Activities of daily living/personal care (9 items)
  - Positioning/transferring and mobility (8 items)
  - Comfort and emotions (9 items)
  - Communication and social interaction (7 items)
  - Health (3 items)
  - Overall quality of life (1 item).

13

(CP-CHILD) Caregiver Priorities and Child Health Index of Life with Disabilities [9]

- Separate caregiver report and adolescent report. One or both can be completed
- Asks families to consider the level of difficulty of an activity over previous 2 weeks on a scale of 0-6
  - 0 = not possible, 6 = no problem at all
- Asks caregiver and child to consider level of assistance needed on scale of 0-3.
  - 0 = total, 3 = independent
- An organization or individual user must register prior to distributing to caregivers or using in any capacity clinically.
- https://lab.research.sickkids.ca/pscoreprogram/cpchild

14

Goal Attainment Scale (GAS) [10,11,12]

- Goal Attainment Scaling (GAS) is a person-centered and collaborative approach, allowing to assess the effectiveness of an intervention on personally relevant goals.
- The GAS has its roots in mental health, as it was created by Kiresuk and Sherman in 1968 to determine effectiveness of community mental health programming.
- The GAS was widely embraced by the rehabilitation community, children and adults alike, due to its person-centered approach to goal writing and effectiveness of intervention. [11]
- Goal achievement is measured using a 5-point scale, ranging from +2 through -2.
  - -2: much less than expected
  - -1: somewhat less than expected
  - 0: expected level of outcome
  - +1: somewhat more than expected
  - +2" much more than expected

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Goal Attainment Scale (GAS)

- Basic criteria required for goal writing in GAS format, the SMART format.
  - Specific
  - Measurable
  - Achievable
  - Realistic
  - Timely
- To define levels using the GAS, the clinician changes one variable of the goal

16

Intensive Plan of Care

Frequency	
Schedule of Visits	3-5x per week
Plan of Care Length	2-6 weeks
Reassessment	Reassess need for continued therapy at least every 12 visits

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Episodic Plan of Care

Frequency	
Schedule of Visits	1-2x per week
Plan of Care Length	1-3 months
Reassessment	Reassess need for continued therapy at least every 12 visits

18

Why Episodic Care for Adolescents?

Psychological benefits of taking a break from intervention, which ultimately increases patient participation during each episode of care.

Breaks from therapy allow for opportunities to practice their new skills in daily routines and environments to increase generalization of skills across various settings.

Provides increased opportunities to explore peer-related activities in the community especially for leisure activities.

19

Frequency During Episode of Care

- When considering recommendations for therapy frequency, clinicians need to view the time dedicated to therapy in the context of the teen and family's full life.
- How much skilled intervention is needed to create a shift in activity participation?
- How feasible is it for a family to attend the frequency of sessions recommended?
  - Transportation considerations
  - Financial considerations
- Does this frequency still allow the teen to participate in activities outside of skilled therapy?

20

Considerations for GMFCS IV & V

ACUTE POST-OP:

Patients with GMFCS levels of IV & V often have specific routines that they have established with their caregivers. However, these routines often need to be modified following a surgery because of pain and surgical precautions.

It is important to provide patients with choices and as much control as possible over this new change in routine.

21

Considerations for GMFCS IV & V

OUTPATIENT

- Collaboration with teen and caregiver
- Use of caregiver-report tools and conversation with caregiver to identify areas of daily routines that are the highest priority based on current circumstances.
- Collaboration with caregiver to figure out which aspects of daily routines are problematic.
  - Is it timing?
  - Is it the level of involvement of the teen in the routine?
- Considerations for physical involvement of caregiver.
  - Strategies for supporting the teen to maximize independence while caregiver does "less".

22

Treatment Interventions

Activities of Daily Living (ADLs)

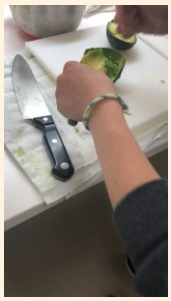
- It is important that all treatment interventions recreate patient's natural environment to the extent possible.
- Practice tasks with patient's own supplies
- Using adaptive equipment that patient has access to at home
- Bringing in pictures or video of environment

23

Treatment Interventions

Instrumental Activities of Daily Living (iADLs)

- It is important that all treatment interventions recreate patient's natural environment to the extent possible.
  - Practice tasks with patient's own supplies
- Using adaptive equipment that patient has access to at home
  - Bringing in pictures or video of environment



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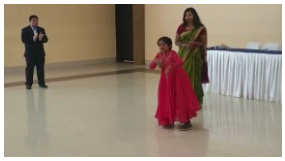
Treatment Interventions



Leisure

- It is important that all treatment interventions recreate patient's natural environment to the extent possible.
  - Practice tasks with patient's own supplies
  - Using adaptive equipment that patient has access to at home
  - Bringing in pictures or video of environment

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Treatment Interventions

Leisure

- Address the underlying skills needed for the leisure activity
  - Recreate the leisure activity in therapy space
  - Assist with ways to finding ways for the patient to participate in this leisure activity within the community

26

Self Advocacy in Adolescents

Adolescence as a time of transition in responsibilities and taking on an increased role in advocacy in their own healthcare and a time to work with families to prepare for the transition into adult-based care.

We should start preparing patients to ask questions, confidently explain their opinions, and advocate for any accommodations that they may need in various environments and throughout their life.

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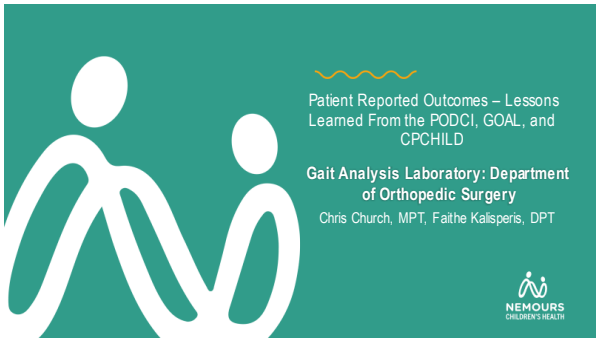

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Disclosures

- Authors have no relevant disclosures related to this presentation
- Photos / Videos used with patient-family permission

NEMOURS CHILDREN'S HEALTH

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3

Patient Reported Outcomes

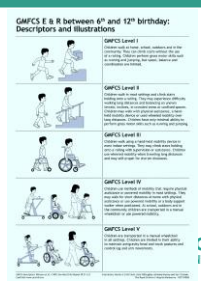
- Questionnaires
- Why use?
- What have we learned?

4

Gross Motor Function Classification System

- Provides a common language to communicate about CP
- Essential when discussing gross motor function in children with CP
- Provides the context for considering the individual child's prognosis goal setting management

Palisano et al. (1997)
CanChild www.canchild.ca
Illustrations Version 2.0 Rad, Willoughby, Harvey and Graham, The Royal Children's Hospital Melbourne ERC/1880



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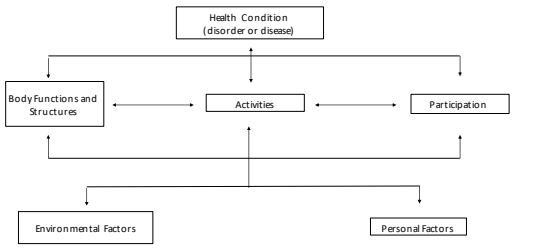
International Classification of Functioning, Disability, and Health (ICF) Model



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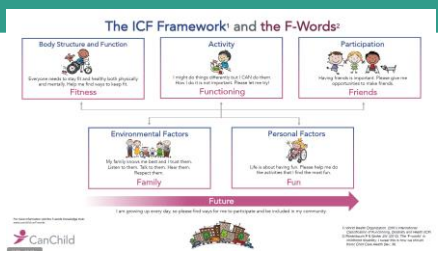

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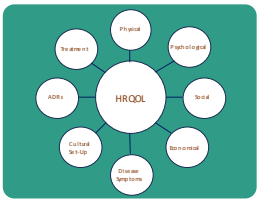
Health – Related Quality of Life



- "Perceived wellbeing in physical, mental, and social domains of health"

(Haas et al., 2010)

- Understanding more about HRQOL can allow clinicians to make changes to treatment
- Self-reported measurements provide a more comprehensive assessment of patient well-being
  - Patient-reported outcomes

(Kaimi et al, 2016)

8

Upper Extremity and Physical Function

Transfer and Basic Mobility

Sports and Physical Functioning

Pain and Comfort

Happiness

Global Functioning

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Pediatric Outcomes Data Collection Instrument: PODCI

- Both parents and patients themselves can take the test
- GMFCS I-III, IV?
- Child vs. Adolescent
  - Child: 2 – 10
  - Adolescent: 11 - 18
- Questions ask patient/parent to rank ease of completing tasks as well as feelings about different subjects over the last week

(Daltroy et al, 1998)
Scott et al, 2021

9

Objectives

1. To compare patient reported outcomes between children with physical disabilities and typically developing children
2. To compare patient reported outcomes between children with CP, arthrogryposis, Morquio syndrome, and achondroplasia
3. To assess correlations between gross motor skills, happiness, and pain

Church et al, not yet published

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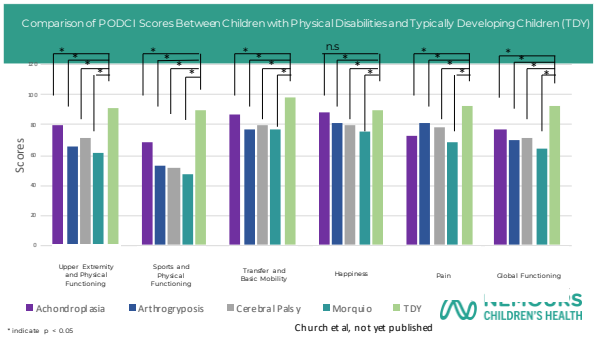

Church et al, not yet published

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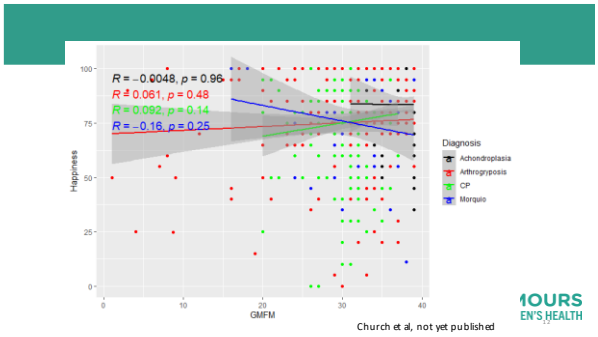

Church et al, not yet published

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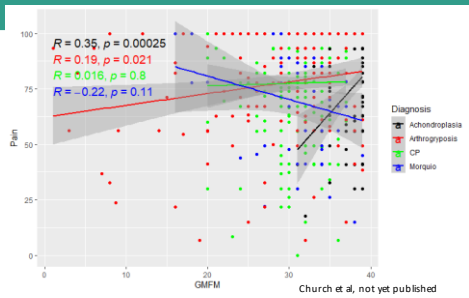

Church et al, not yet published

13

What did we learn?

- Individuals with the most common disabilities of childhood present with limitations in HRQOL
- Happiness and pain both tend not to be associated with motor function
  - Future research should study factors that affect mental health in children with these disabilities
- It is essential to utilize patient reported outcomes to best understand and assist in the management of HRQOL in children and adolescents with lifelong physical disabilities.

Church et al, not yet published

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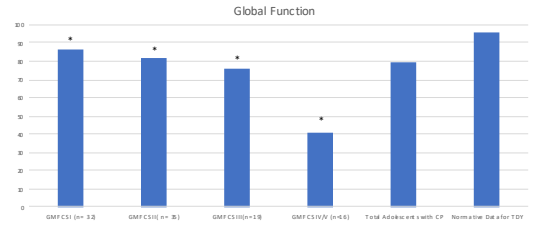
Objectives

1. To compare children with CP between GMFCS levels
2. To compare adolescent self-report to parent report
3. To assess correlations between gross motor skills, happiness, and pain

Lennon et al, 2023

15

Patient/Family Reported Outcomes

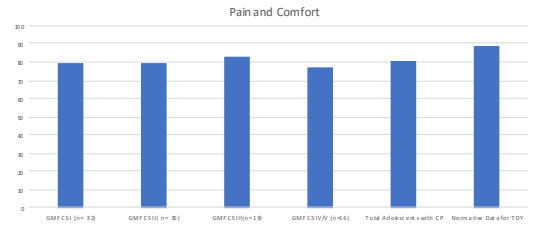


Lennon et al, 2023

* P<0.05

16

Patient/Family Reported Outcomes

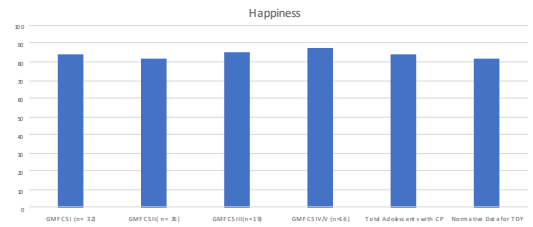


Lennon et al, 2023

No significant differences

17

Patient/Family Reported Outcomes

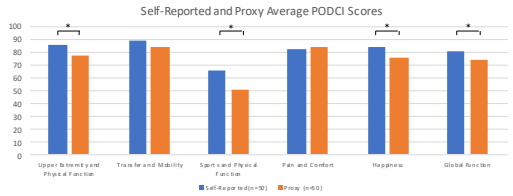


Lennon et al, 2023

No significant differences

18

Patient/Family Reported Outcomes



Lennon et al, 2023

* P<0.05

What did we learn?

- Greater functional ability doesn't = more happiness
- Adolescents score themselves higher than their parents

Lennon et al, 2023

PODCI Manual

Pediatric Outcomes Questionnaire

Developed by: American Academy of Orthopaedic Surgeons® Pediatric Orthopaedic Society of North America(POSNA) American Academy of Pediatrics Shriner's Hospitals

To be completed by the parent for children 2 –10 years old Based on the Version 2.0 Pediatrics-Parent/Child Outcomes Instrument Also commonly referred to as the **PODCI ("Pediatric Outcomes Data Collection Instrument")**

Revised, renumbered, reformatted August 2005

PODCI Manual

https://www5.aaos.org/research/outcomes/Pediatric.pdf

GOAL (Gait Outcomes Assessment List)

Stout, et al 2023
Munger, et al 2023

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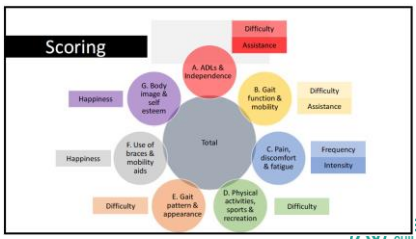
GOAL (Gait Outcomes Assessment List)



Stout, et al 2023
Munger, et al 2023

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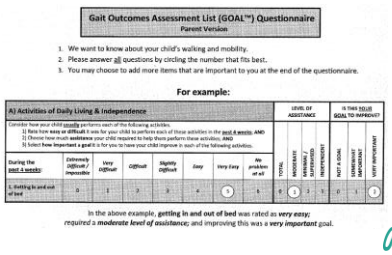
GOAL



Gait Outcomes Assessment List (GOAL™) Questionnaire
Parent Version

1. We want to know about your child's walking and mobility.
2. Please answer all questions by circling the number that fits best.
3. You may choose to add more items that are important to you at the end of the questionnaire.

For example:

A) Activities of Daily Living & Independence							LEVEL OF ASSISTANCE			IS THIS YOUR GOAL TO IMPROVE?		
Consider how your child usually performs each of the following activities. 1) Rate how easy or difficult it was for your child to perform each of these activities in the past 4 weeks; AND 2) Choose how much assistance your child required to help them perform these activities; AND 3) Select how important a goal it is for you to have your child improve in each of the following activities.							TOTAL	MODERATE / MAXIMUM / SUPERVISED	INDEPENDENT	NOT A GOAL	SOMEWHAT IMPORTANT	VERY IMPORTANT
During the past 4 weeks:	Extremely Difficult / Impossible	Very Difficult	Difficult	Slightly Difficult	Easy	Very Easy	No problem at all					
1. Getting in and out of bed	0	1	2	3	4	5	6	4	2	0	1	2

In the above example, **getting in and out of bed** was rated as ***very easy;*** *required a* ***moderate level of assistance;*** and improving this was a ***very important*** goal.

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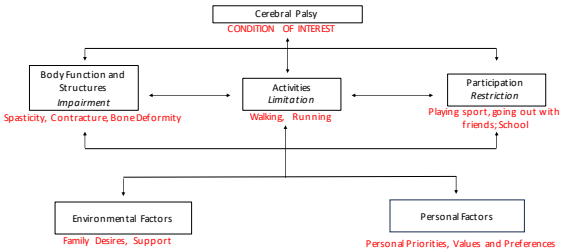
GOAL



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GOAL - Objective

1. *What goals are important to ambulatory children with CP?*

Boyer et al, 2022

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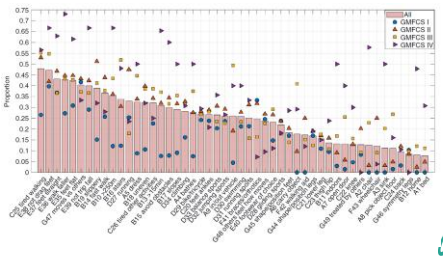
GOAL



Different people have different goals

Boyer et al, 2022

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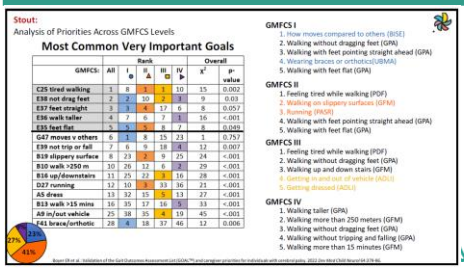
GOAL – What is important to our patients?



Stout:
Analysis of Priorities Across GMFCS Levels

Most Common Very Important Goals

GMFCS:	All	Rank I	Rank II	Rank III	Rank IV	Overall χ^2	Overall p-value
C25 tired walking	1	8	1	1	10	15	0.002
E38 not drag feet	2	2	10	2	1	9	0.03
E37 feet straight	3	1	4	17	6	8	0.057
E36 walk taller	4	7	6	7	1	16	<.001
E35 feet flat	5	3	5	8	7	8	0.049
G47 moves v others	6	1	8	15	23	1	0.757
E39 not trip or fall	7	6	9	18	4	12	0.007
B19 slippery surface	8	23	2	9	25	24	<.001
B10 walk >250 m	10	26	12	6	2	29	<.001
B16 up/downstairs	11	25	22	3	16	28	<.001
D27 running	12	10	3	33	36	21	<.001
A9 dress	13	32	15	5	13	27	<.001
B13 walk >15 mins	16	35	17	16	5	33	<.001
A9 in/out vehicle	25	38	35	4	19	45	<.001
E41 brace/orthotic	28	4	18	37	46	12	0.006

Boyer SR et al. Validation of the Gait Outcomes Assessment List (GOAL™) and caregiver priorities for individuals with cerebral palsy. 2022 Dev Med Child Neurol 64:378-86.

GMFCS I
1. How moves compared to others (BSE)
2. Walking without dragging feet (GPA)
3. Walking with feet pointing straight ahead (GPA)
4. Wearing braces or orthotics(UBMA)
5. Walking with feet flat (GPA)

GMFCS II
1. Feeling tired while walking (PDF)
2. Walking on slippery surfaces (GFM)
3. Running (WASR)
4. Walking with feet pointing straight ahead (GPA)
5. Walking with feet flat (GPA)

GMFCS III
1. Feeling tired while walking (PDF)
2. Walking without dragging feet (GPA)
3. Walking up and down stairs (GFM)
4. Getting in and out of vehicle (ADLI)
5. Getting dressed (ADLI)

GMFCS IV
1. Walking taller (GPA)
2. Walking more than 250 meters (GFM)
3. Walking without dragging feet (GPA)
4. Walking without tripping and falling (GPA)
5. Walking more than 15 minutes (GFM)

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What did we learn?

- Considering your patients/families goals is important
- Goals vary but may be related to GMFCS level

Boyer et al, 2022

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GOAL manual

- **GOAL™**
- **Gait Outcomes Assessment List (GOAL™)**
- The Gait Outcomes Assessment List (GOAL™) is a patient reported outcome measure for ambulatory children with cerebral palsy (CP).
- The GOAL comprises of 48 items distributed across 7 subscales, and spans all domains of the International Classification of Functioning, Disability and Health (ICF).
- Learn more about the GOAL Project.

- https://lab.research.sickkids.ca/pscoreprogram/goal/

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CPCHILD – Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD)

- GMFCS levels IV and V
- Age 5-19 years
- 37 items distributed over 6 sections
  - Activities of Daily Living/Personal Care
  - Positioning, Transferring and Mobilty
  - Comfort and Emotions
  - Communication and Social Interaction
  - Health
  - Overall Quality of Life

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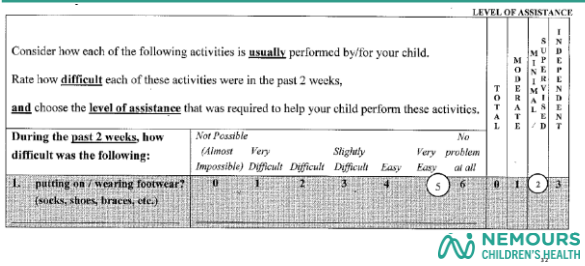
CPCHILD – Caregiver Priorities and Child Health Index of Life with Disabilities (CPCHILD)



Consider how each of the following activities is **usually** performed by/for your child. Rate how **difficult** each of these activities were in the past 2 weeks, **and** choose the **level of assistance** that was required to help your child perform these activities.	Not Possible (Almost Impossible)	Very Difficult		Difficult	Slightly Difficult		Easy	Very Easy	No problem at all	TOTAL	MODERATE	LEVEL OF ASSISTANCE: MINIMAL	SUPERVISED	INDEPENDENT
During the past 2 weeks, how difficult was the following:														
1. putting on / wearing footwear? (socks, shoes, braces, etc.)	0	1		2	3		4	(5)	6	0	1	(2)		3

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CPChild

	GMFCS IV (n=18)	GMFCS V (n=35)
Personal Care/ADLs	41.6 (13.3)	31.0 (15.2)
Positioning, Transferring & Mobility	45.5 (11.3)	28.4 (14.2)
Comfort & Emotions	81.0 (14.5)	67.9 (22.6)
Communication & Social Interaction	74.9 (22.2)	43.4 (23.7)
Health	83.7 (15.0)	57.0 (16.9)
Quality of Life	72.5 (21.8)	55.4 (24.8)

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Objectives

1. Is there a relationship between hip dysplasia and quality of life (CPCHILD score)?
2. Does quality of life (CPCHILD score) improve after hip reconstruction?

DiFazio et al, 2016

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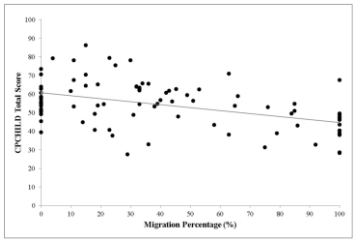
CPChild

- Worse hip, worse Quality of Life



DiFazio et al, 2016

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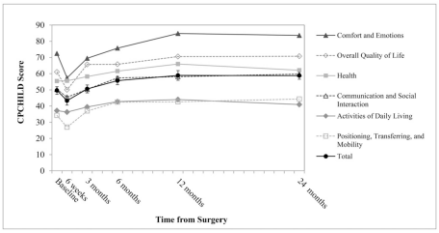
CPChild

- Hip reconstruction improves quality of life



DiFazio et al, 2016

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What did we learn?

- Hip dysplasia is important to quality of life in children with CP
- It is important to ask children and families about their quality of life to guide and assess success of treatment

DiFazio et al, 2016

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CP Child manual

- **CPCHILD™**
- **Caregiver Priorities and Child Health Index of Life with Disabilities**
- **Questionnaire**
- The CPCHILD™ questionnaire is available in the following languages. Please register for a license prior to use.

- https://lab.research.sickkids.ca/pscoreprogram/cpchild/

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Case Studies

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Thank-You

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Dystonia and cognition: Leveraging one to manage the other

Bhooma Aravamuthan, MD, DPhil

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Assistant Professor of Neurology and Pediatrics

Washington University in St. Louis

Cerebral Palsy Center, St. Louis Children's Hospital

Disclosures

- None relevant

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- Consultant for Neurocrine Biosciences
- Royalties from UpToDate
- Immediate family member on the Speaker’s Bureau for SK Life Science

Community driven dystonia research agenda

Laura Gilbert

1. Develop new treatments
2. Assess rehabilitation and psychological management approaches
3. Compare effectiveness of current treatments
4. Improve diagnosis and severity assessments
5. Assess the impact of mixed tone

Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

Community driven dystonia research agenda

Laura Gilbert

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Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

Community driven dystonia research agenda

Laura Gilbert

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5. Assess the impact of mixed tone

Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

Dystonia is:

voluntary movement triggered overflow muscle activation; arousal-dependent

Albanese et al., *Mov. Dis.* 2013; Sanger et al., *Pediatrics*, 2003

Dystonia is:

Albanese et al., *Mov. Dis.* 2007; Sanger et al., *Pediatrics, 2003;* Lin et al., *JNNP* 2014; Perides et al. *DMCN* 2020; Yuan-Kim Liow et al, *Eur. J. Pediatr. Neurol.* 2016; Monbaliu et al., *DMCN* 2017; Fehlings et al. *DMCN* 2018; Knights et al., *J. Child Neurol.* 2013; Rice et al. *DMCN* 2017; Aravamuthan et al. *DMCN* 2021

Early dystonia diagnosis: a gap

Of children with hypertonia at high risk for cerebral palsy

30%

have their tone types specified by 5 years old

Miao, Mathur, and Aravamuthan. *J Child Neurol.* 2021

Early dystonia diagnosis: a gap

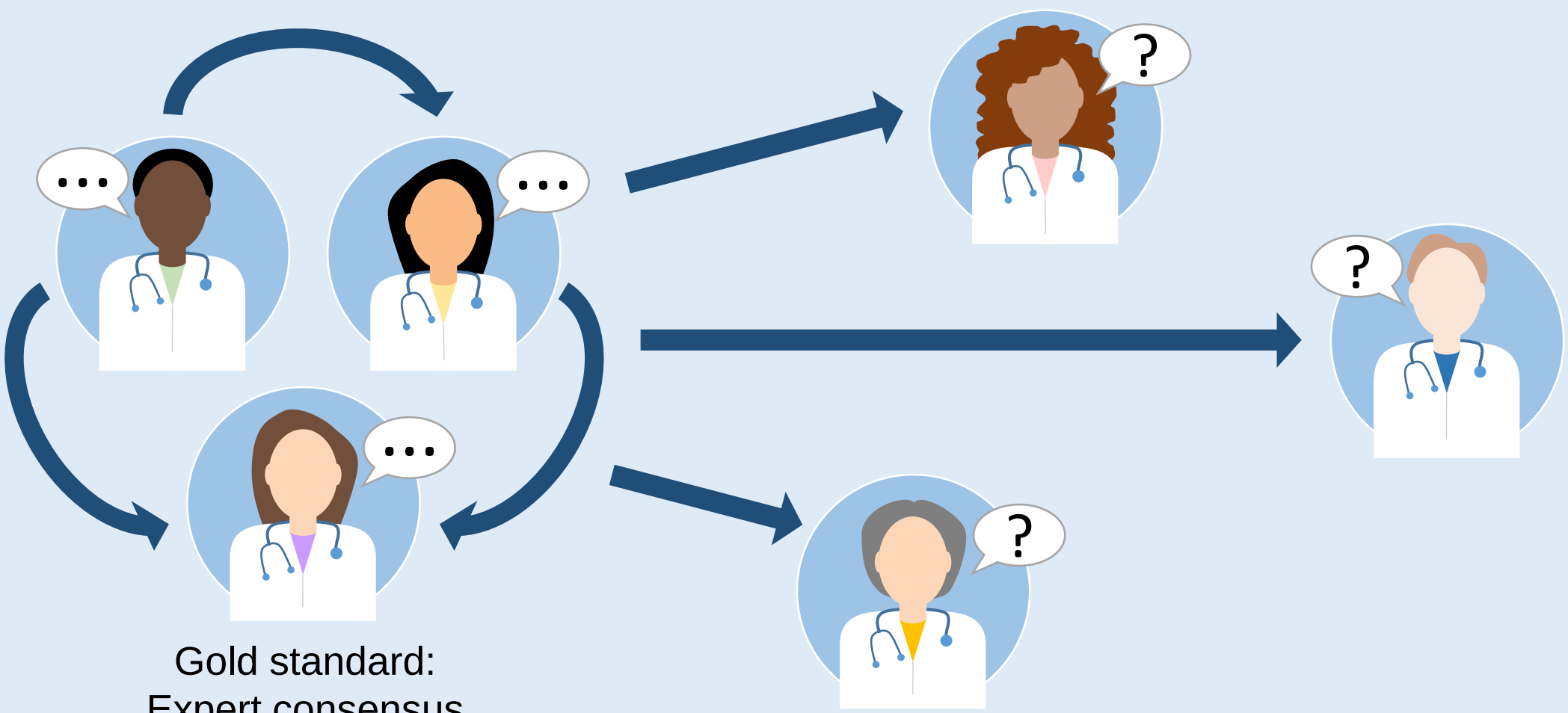
Of ambulatory children
with CP,

13%

have their leg dystonia
identified during any single
CP clinic visit

Aravamuthan et al. *Annals of the Child Neurology Society* (accepted)

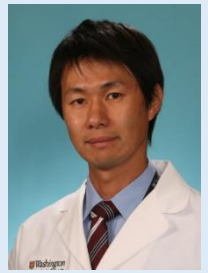
Dystonia diagnosis: status quo



Gold standard:
Expert consensus

How to experts pragmatically grade dystonia severity?


Toni Pearson


Keisuke Ueda

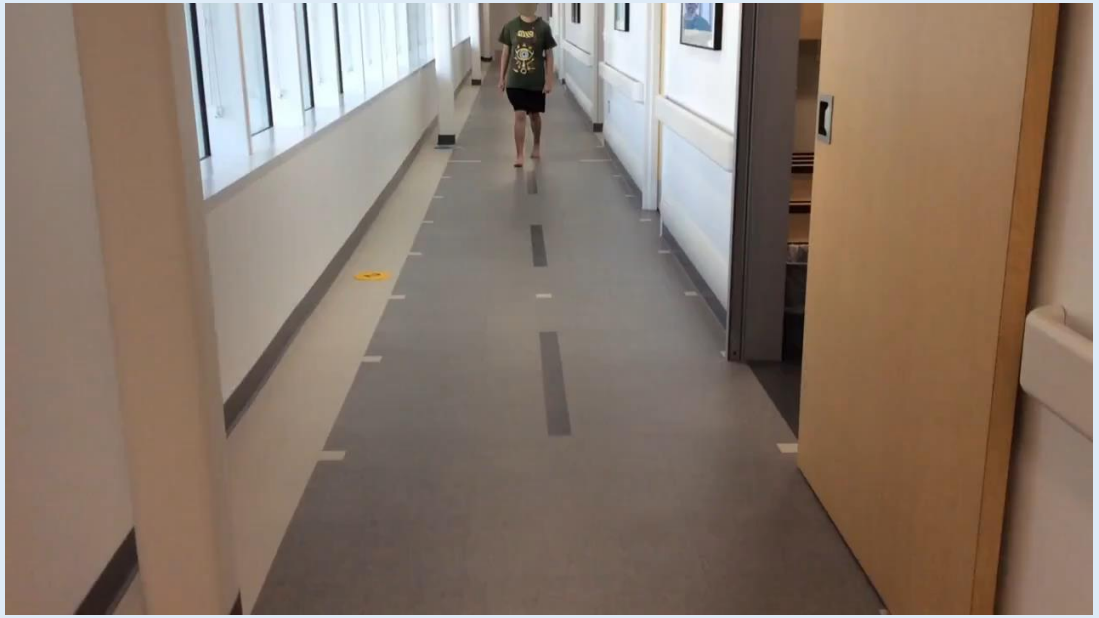

Joel Perlmutter

- Three movement disorder physicians reviewed 116 videos of people with CP (age 10-20 yo) as they walked ~15 ft in a straight line towards the camera
- All people with CP also had documented spasticity
- Grading was done with a 10 pt Likert-style scale (Global Dystonia Severity Rating Scale)
- Only graded dystonia in the lower extremities

Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

Example videos of people with CP + spasticity +/- dystonia

Video 1

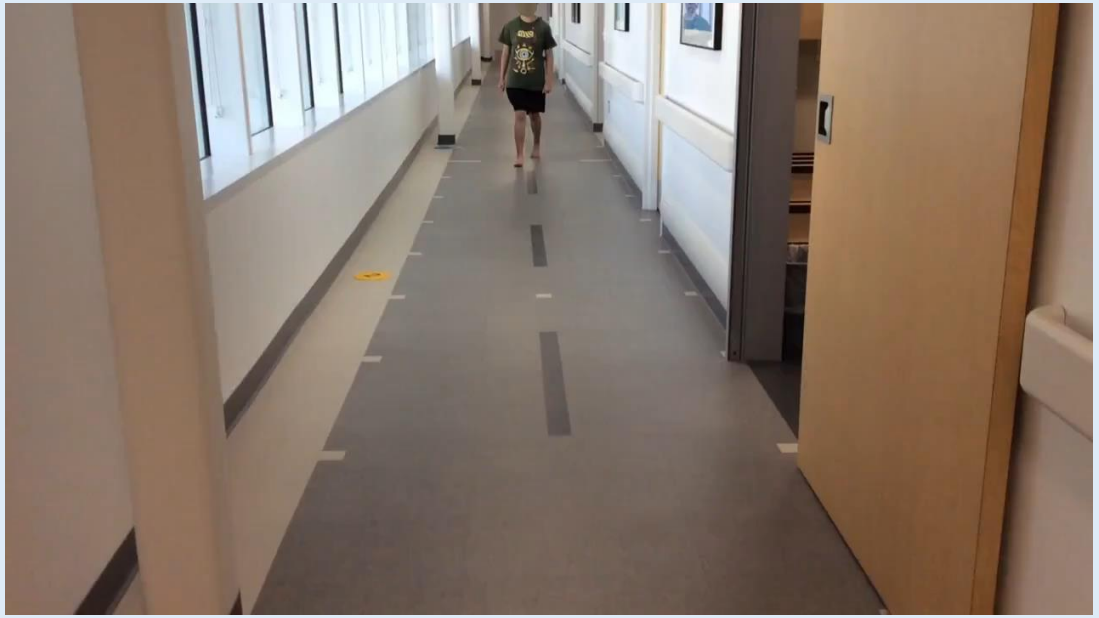


Video 2

Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

Example videos of people with CP + spasticity +/- dystonia

Avg GDRS 0



Avg GDRS 6.33

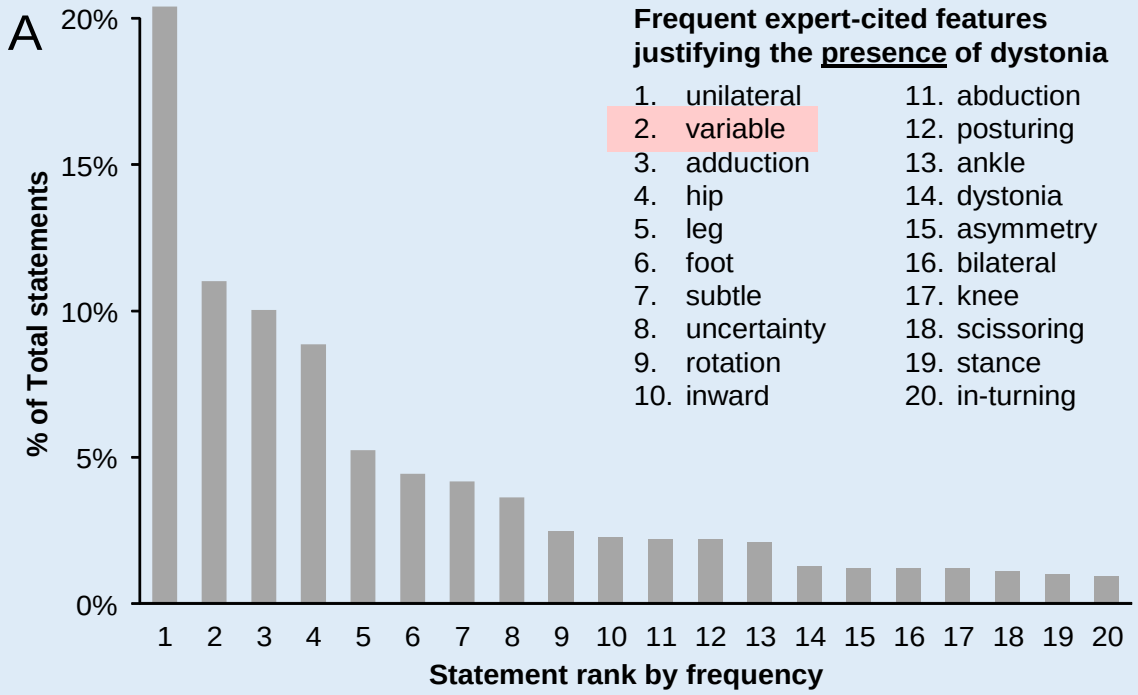
Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

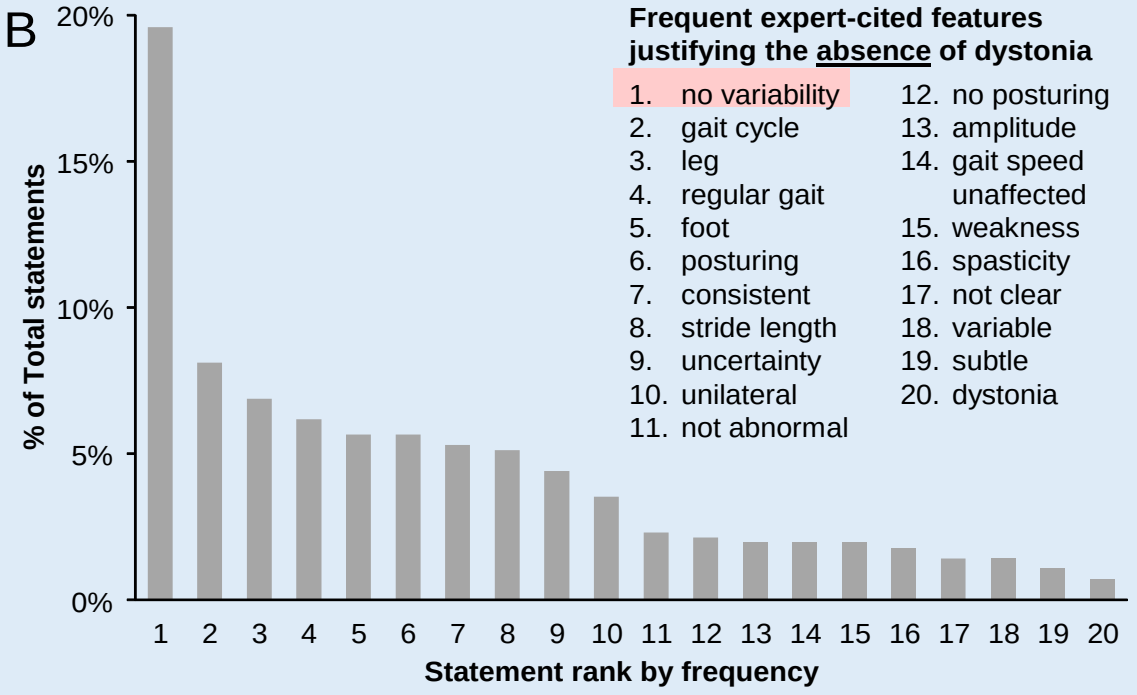
Example Grading

Video ID	Average GDRS Leg Sub-score	Justification for presence or absence dystonia	Justification for GDRS score (if dystonia present)
1	0.00	"Consistent stride length and leg/foot posture"	
2	6.33	"Intermittent variable scissoring of both legs"	"causes deviation in overall gait course - seems to limit function"

Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

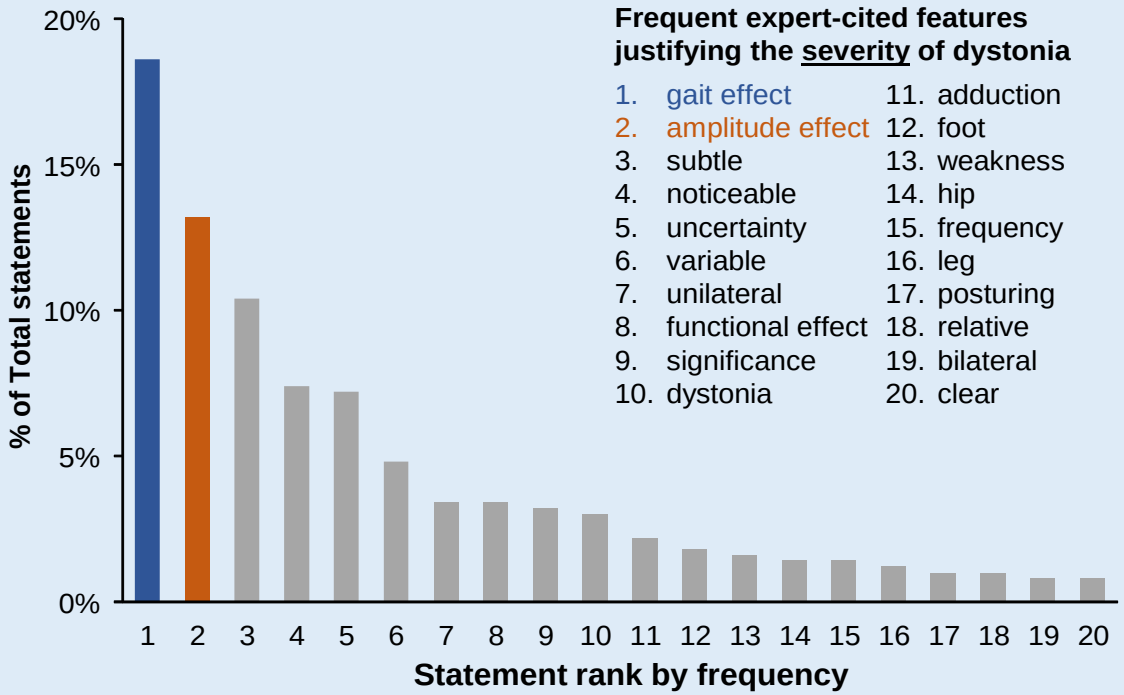
Expert-cited diagnostic features





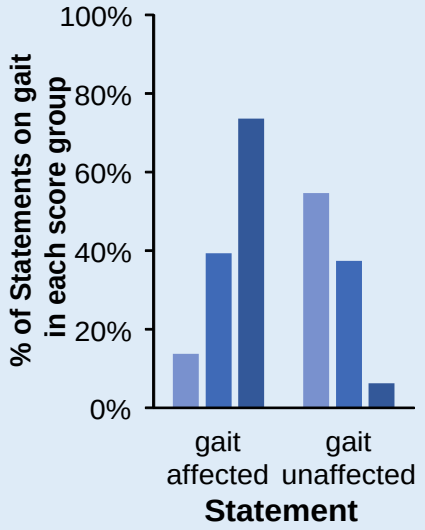
Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

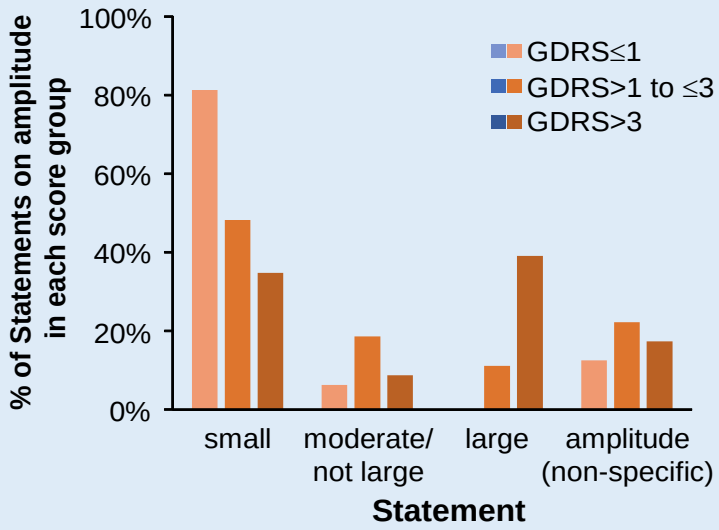
Expert-cited features of dystonia severity



Frequent expert-cited features justifying the severity of dystonia

1. gait effect
2. amplitude effect
3. subtle
4. noticeable
5. uncertainty
6. variable
7. unilateral
8. functional effect
9. significance
10. dystonia
11. adduction
12. foot
13. weakness
14. hip
15. frequency
16. leg
17. posturing
18. relative
19. bilateral
20. clear





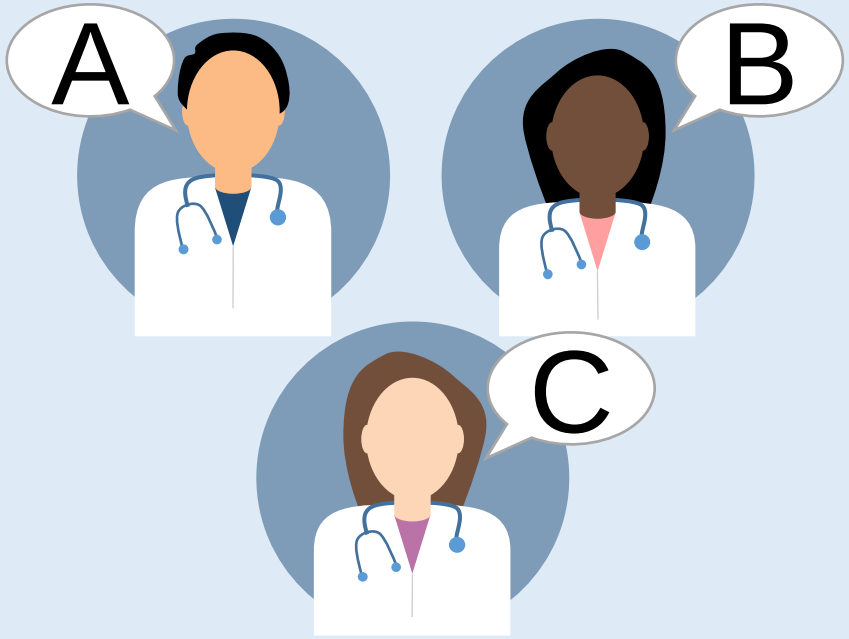
Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

How to experts pragmatically grade dystonia severity?

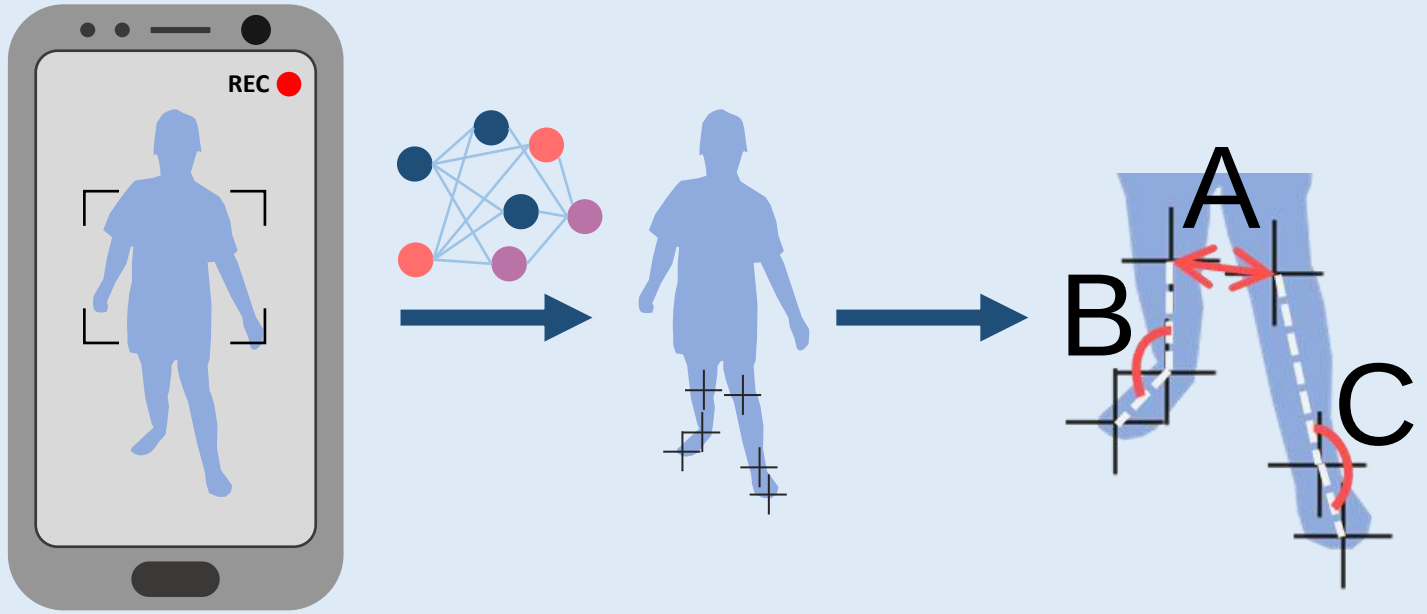
- Diagnostic: Unilateral **variable** leg and foot adduction
- Severity grading:
  - Gait effect (stability + regularity) → **function**
  - **Adduction amplitude**

Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

Dystonia diagnosis: a new way



What are the expert-cited features of dystonia?



Develop gait variable measures of these expert-cited features

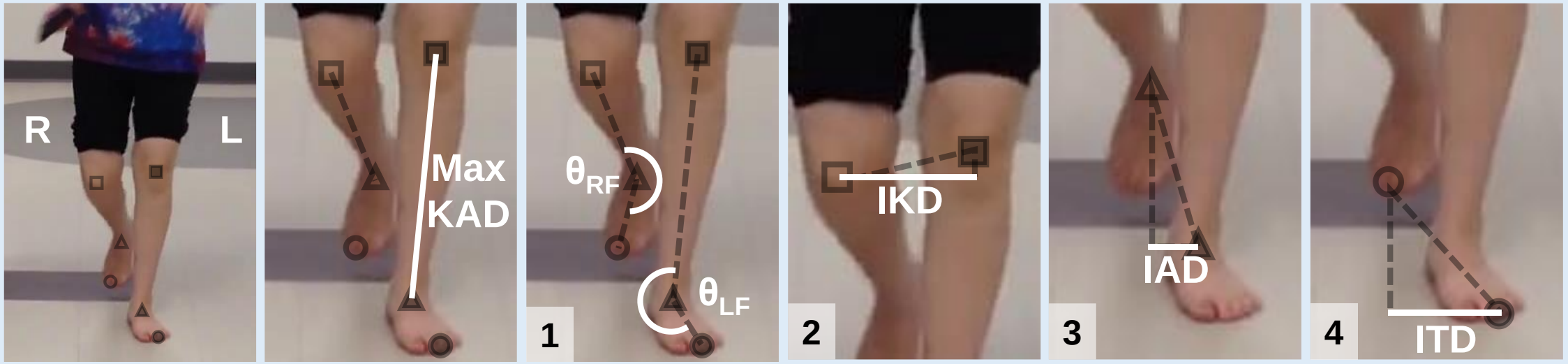
Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

Can we develop kinematic analogues of expert-cited features of dystonia severity?

- Can label knees, ankles, toes reliably on these videos
- Gait effect + variability
- Adduction amplitude

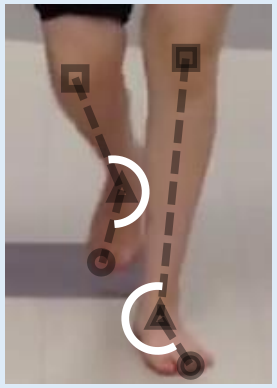
Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

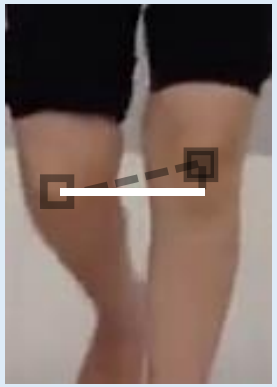
Can we develop kinematic analogues of expert-cited features of dystonia severity?

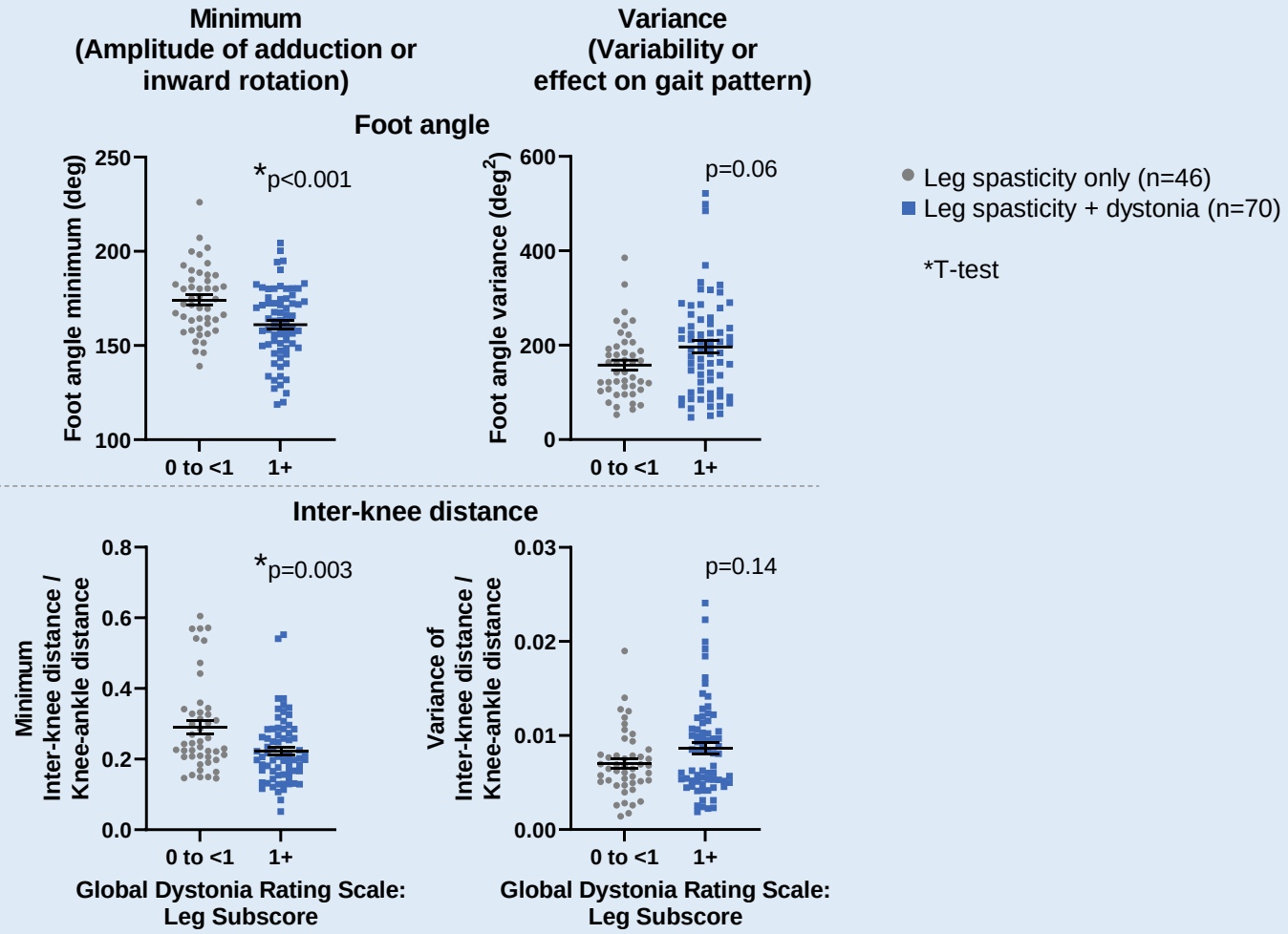


Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

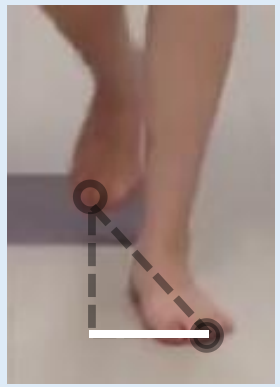
Can we develop kinematic analogues of expert-cited features of dystonia severity?

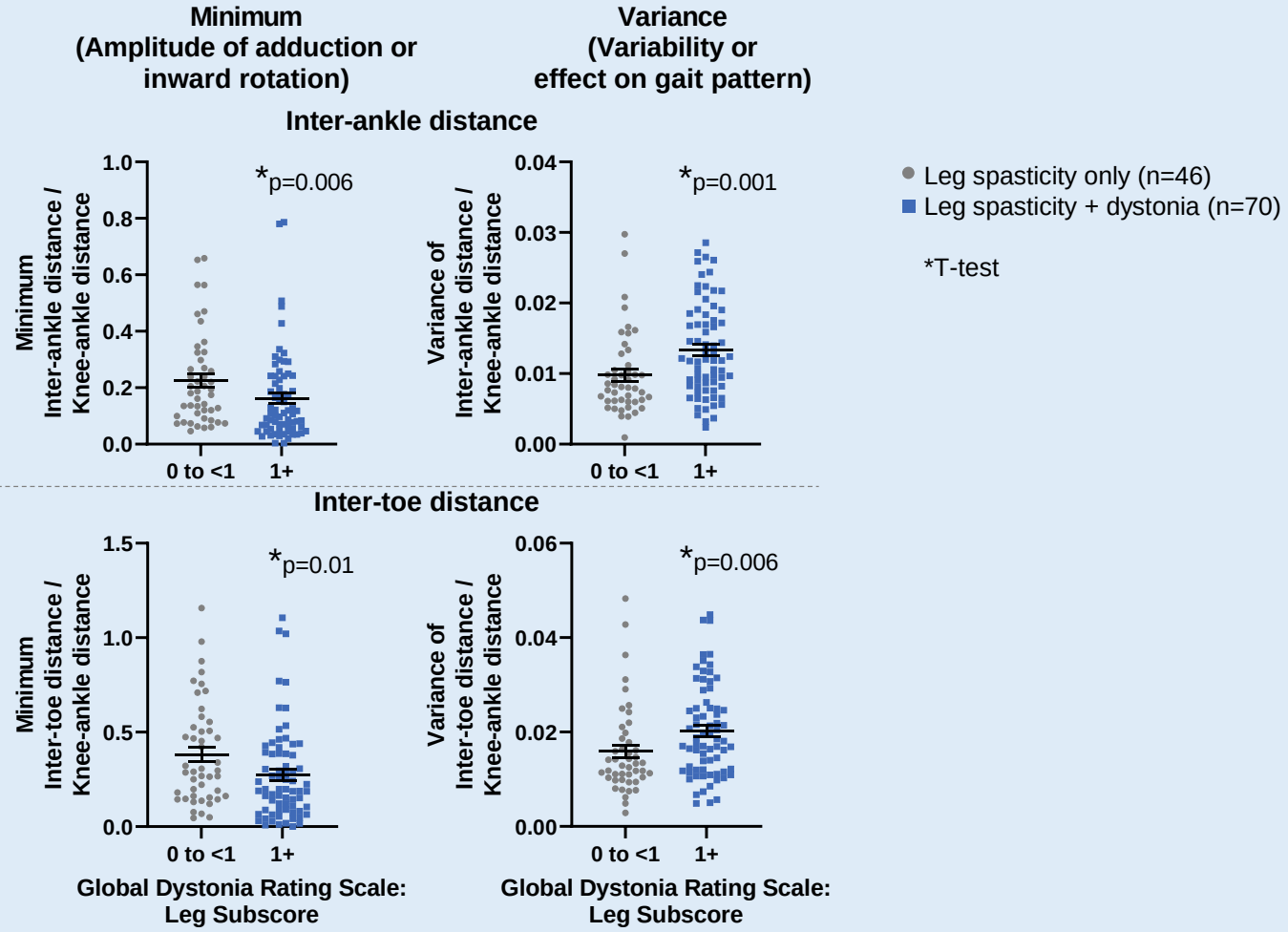




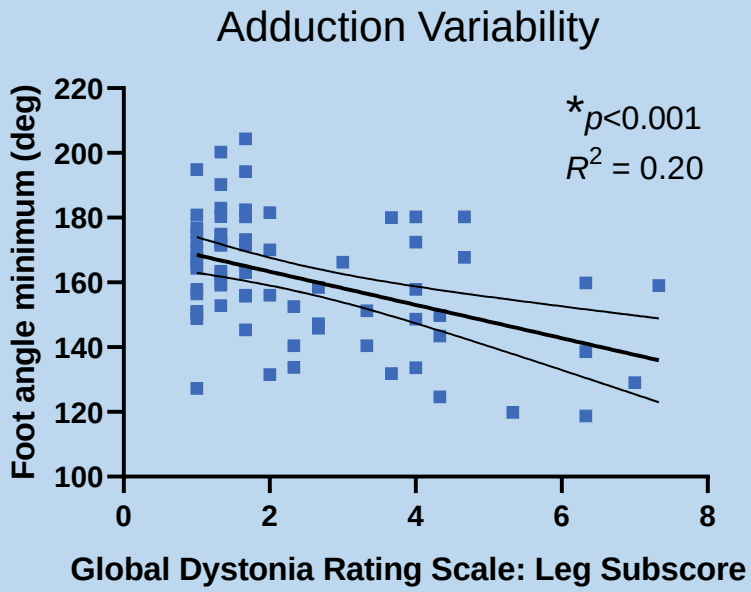


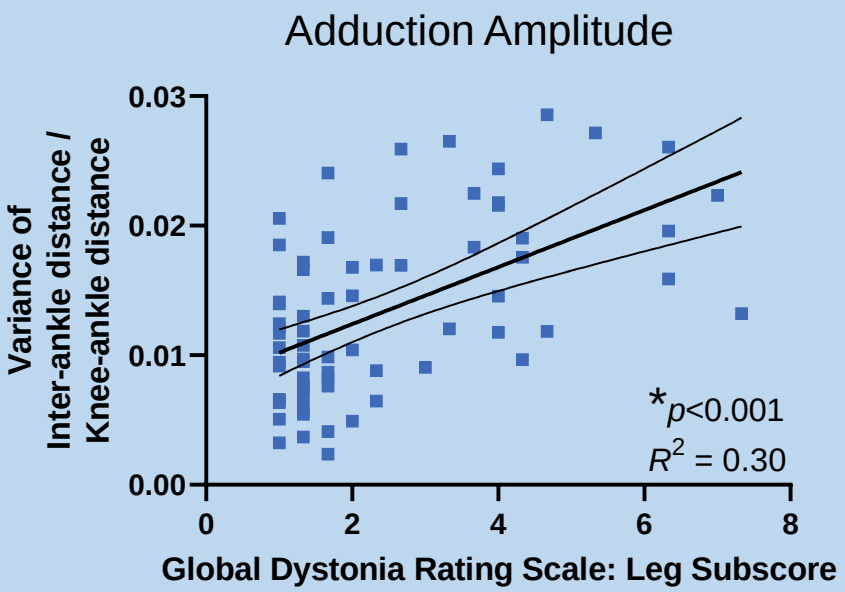
Can we develop kinematic analogues of expert-cited features of dystonia severity?

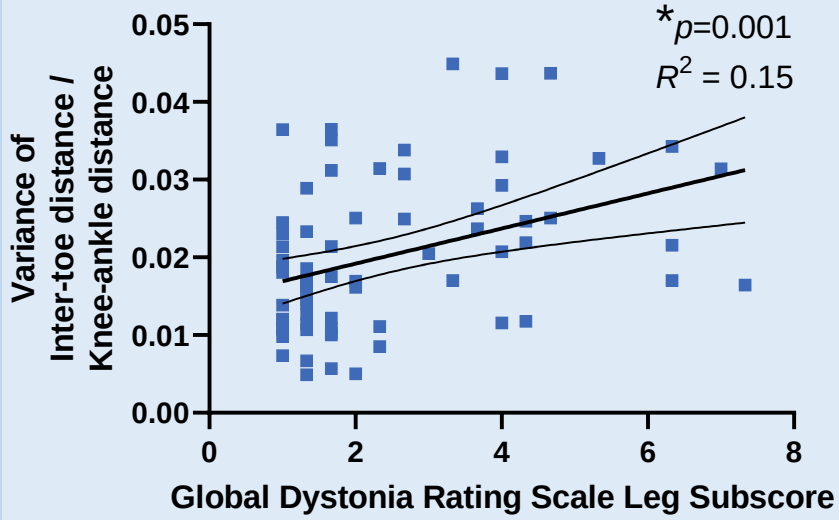




Can we develop quantitative analogues of expert-cited features of dystonia severity?



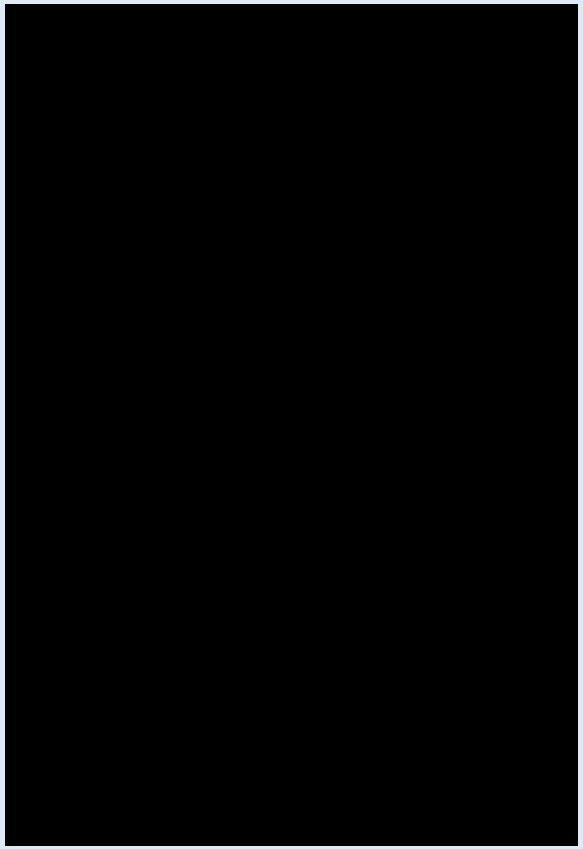




*Multiple linear regression, n=70

Aravamuthan et al., *Dev Med Child Neurol.* 2021; *Dev Med Child Neurol.* 2023

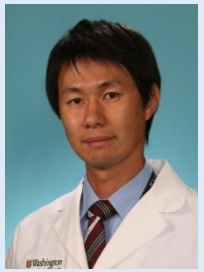
Upper extremity features of dystonia



Laura Gilbert

Sushma Gandham

Toni Pearson



Keisuke Ueda

Gilbert....Aravamuthan. *Neurology: Clinical Practice* 2023

Upper extremity features of dystonia


Laura Gilbert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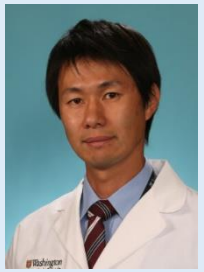
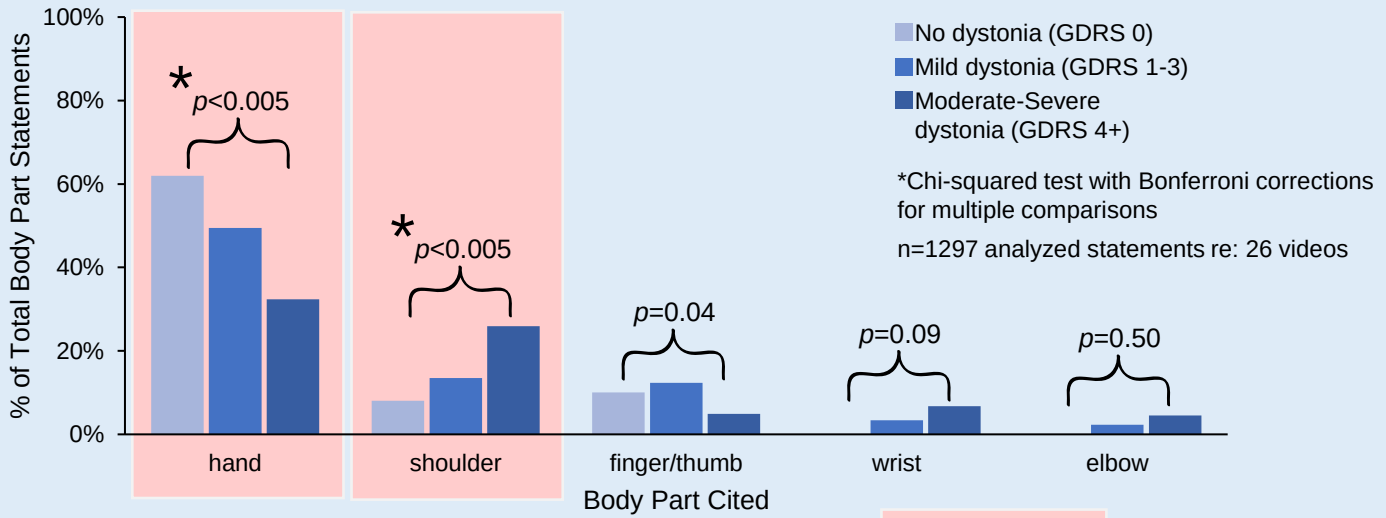
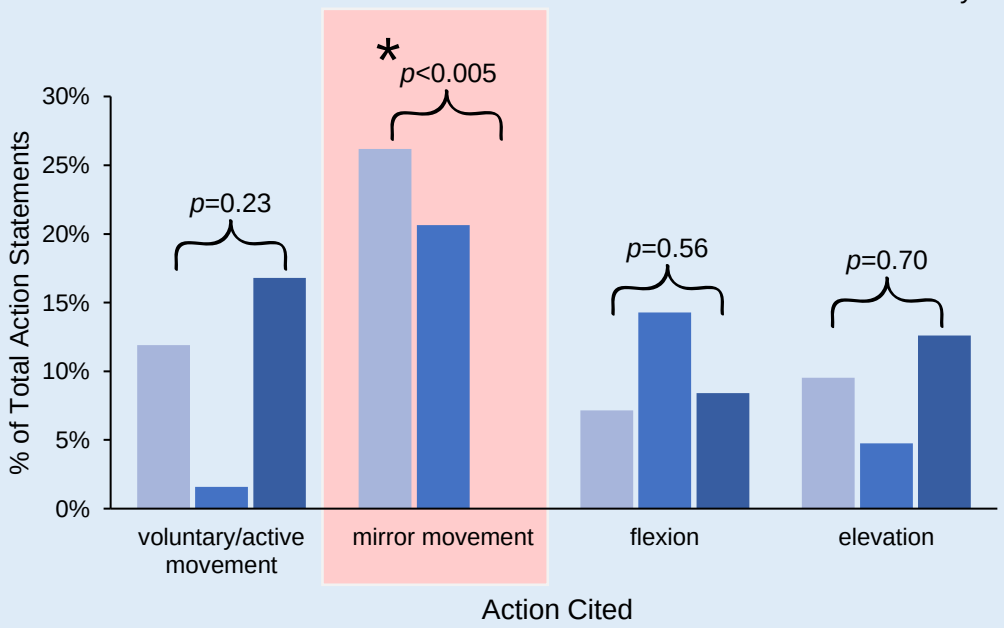
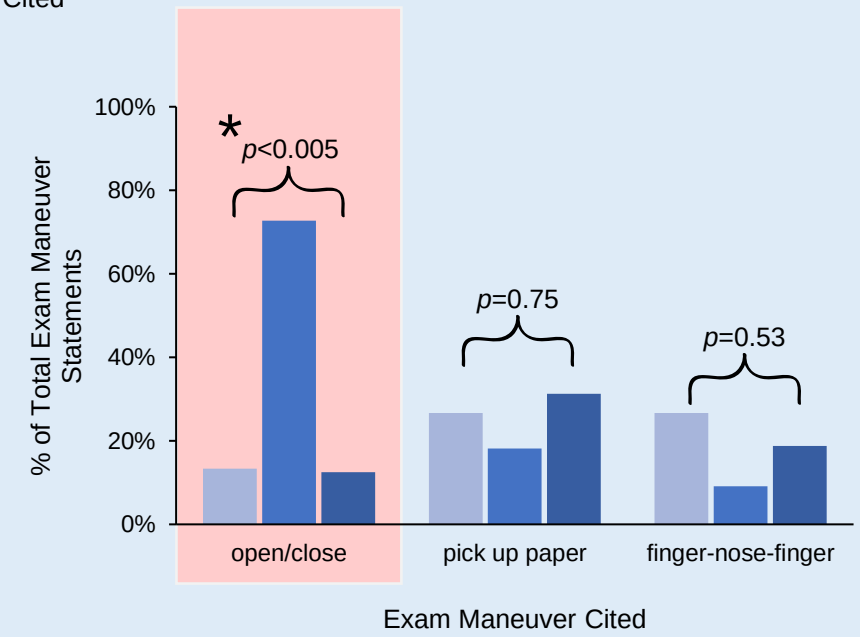

Keisuke Ueda





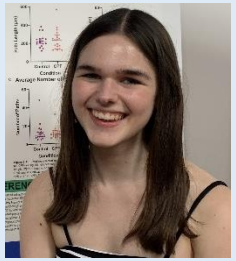


Gilbert....Aravamuthan. *Neurology: Clinical Practice* 2023

Looking at overflow leg adduction as a sign of dystonia

Gemperli....Aravamuthan. *bioRxiv* 2023, *In submission*

Dystonia diagnosis: community expertise


Alyssa Rust


Fayza Jaleel

1. When you try to move one part of your body, do you move another part of your body without meaning to?
2. During activities when someone handles a part of your body, do you move a different part of your body without meaning to?

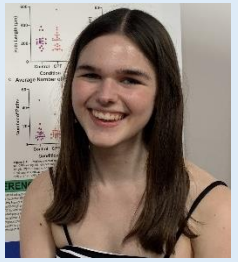
YES to both?

92%

had dystonia (PPV)

Jaleel, Rust....Aravamuthan. *Annals Clin Trans Neurol* 2023

Dystonia diagnosis: community expertise


Alyssa Rust


Fayza Jaleel

1. When you try to move one part of your body, do you move another part of your body without meaning to?
2. During activities when someone handles a part of your body, do you move a different part of your body without meaning to?

NO to both?

77%

did <u>NOT</u> have dystonia (NPV)

Jaleel, Rust....Aravamuthan. *Annals Clin Trans Neurol* 2023

Community driven dystonia research agenda

Laura Gilbert

1.
2. management
3. ents

- **Lower extremities:** Variable leg adduction
- **Upper extremities:** Moves from distal (hand) to proximal (shoulder) with increasing severity
- **Exam trigger:** Hand open/close
- **History:** Ask about tactile and voluntary movement triggers

4. Improve diagnosis and severity assessments

5. Assess the impact of mixed tone

Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

Community driven dystonia research agenda

Laura Gilbert

1. Develop new treatments
2. Assess rehabilitation and psychological management approaches
3. Compare effectiveness of current treatments
4. Improve diagnosis and severity assessments
5. Assess the impact of mixed tone

Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

Dystonia

CANNOT be managed without first managing its **triggers**

Dystonia triggers

- Pain
- Poor sleep
- Heightened mood
  - Excitement
  - Anxiety

Dystonia triggers – Data from St. Louis

- Pain
- Poor sleep
- Heightened mood
  - Excitement
  - Anxiety

69%

have dystonia

17%

have dystonia as their predominant tone

Dystonia triggers – Data from St. Louis

- Pain – **34%**
- Poor sleep – **28%**
- Heightened mood
  - Excitement
  - Anxiety – **26%**

69%

have dystonia

17%

have dystonia as their predominant tone

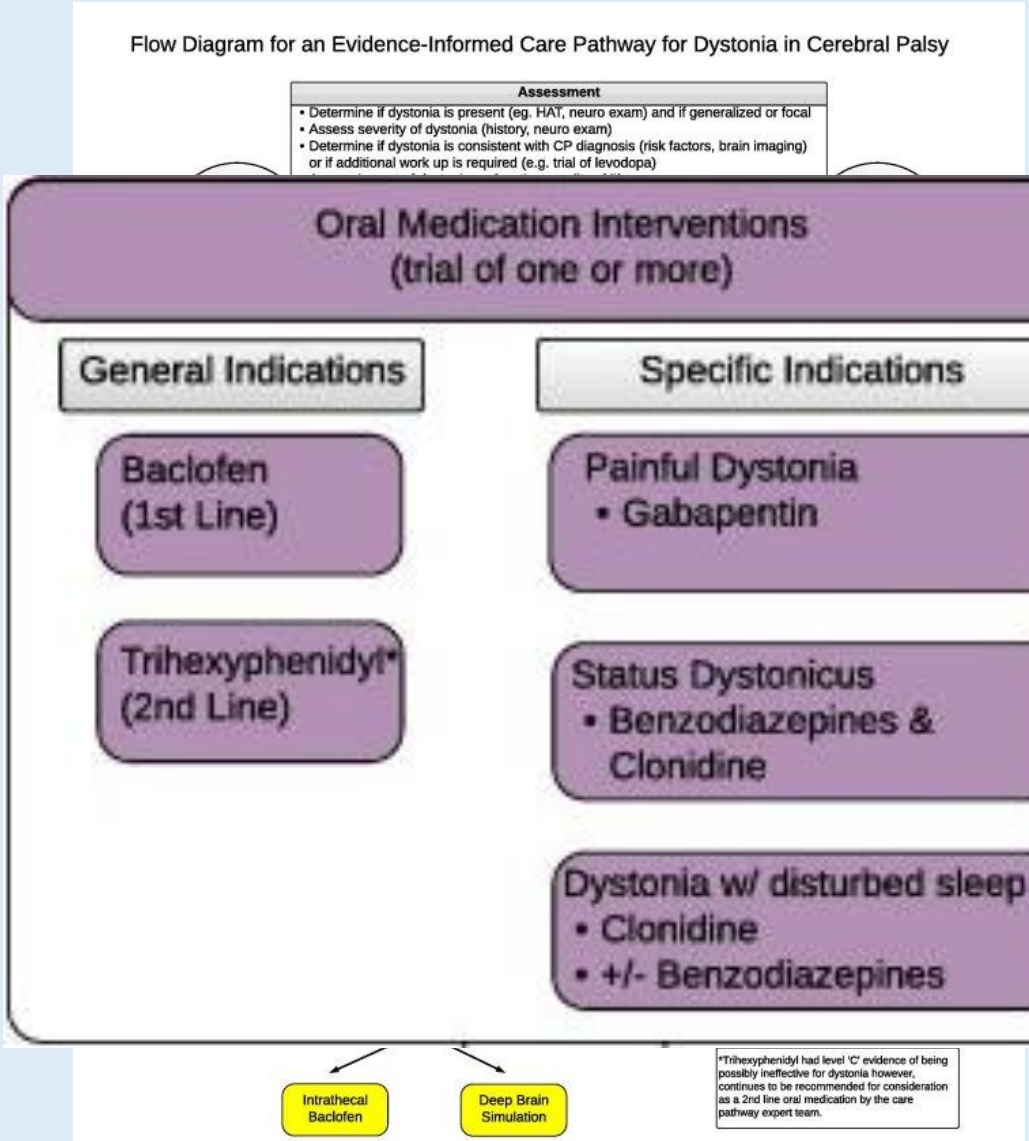
Dystonia

CANNOT be managed without first managing its **triggers**

Present in ~25-30%

Dystonia triggers

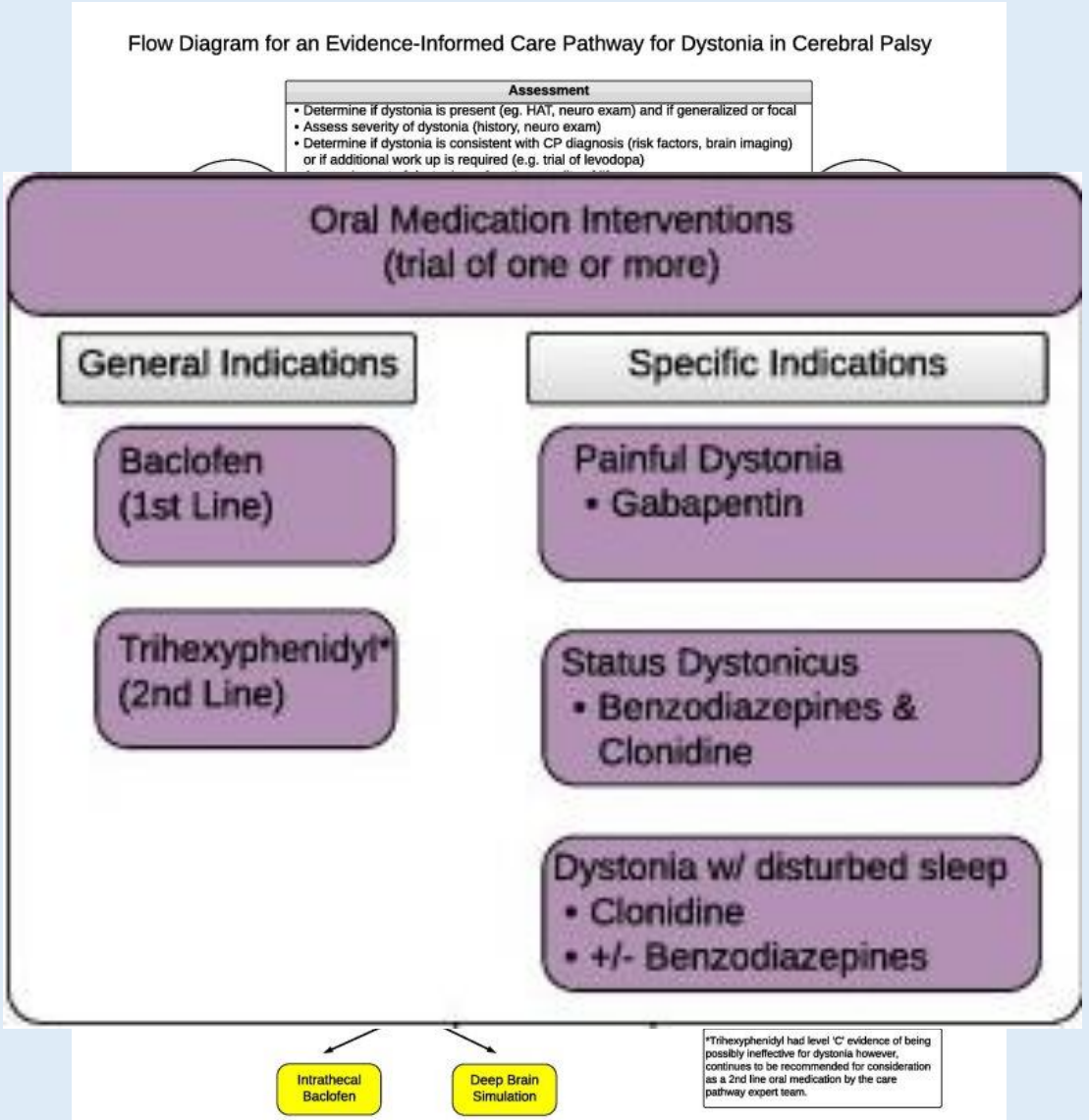
- Pain ✓
- Poor sleep ✓
- Heightened mood
  - Excitement
  - Anxiety ?



https://www.aacpdm.org/publications/care-pathways/dystonia-in-cerebral-palsy

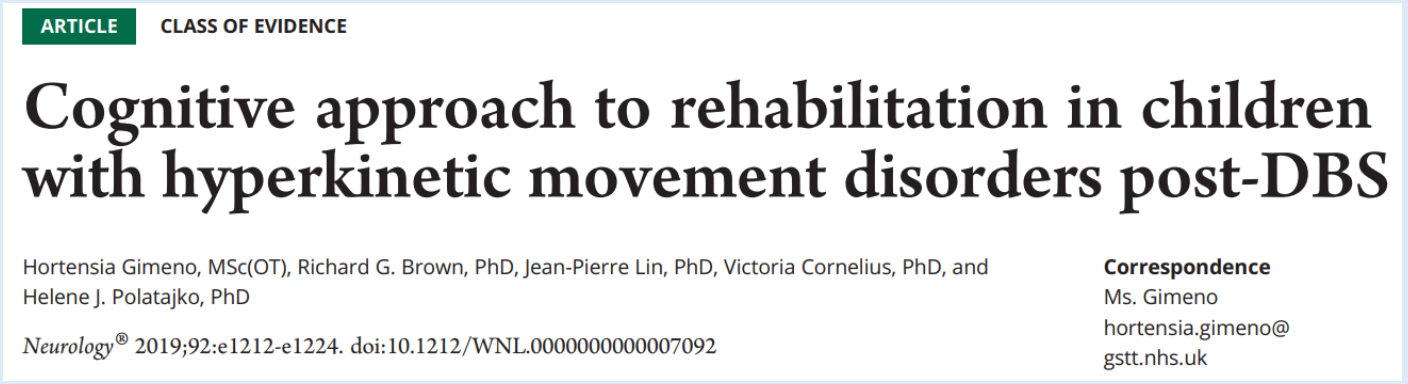
Dystonia triggers

- Pain ✓
- Poor sleep ✓
- Heightened mood
  - Excitement
  - Anxiety – SSRI/SNRI
  - Aggression or Mood Lability
    - Antipsychotic



https://www.aacpdm.org/publications/care-pathways/dystonia-in-cerebral-palsy

CO-OP



ARTICLE CLASS OF EVIDENCE

Cognitive approach to rehabilitation in children with hyperkinetic movement disorders post-DBS

Hortensia Gimeno, MSc(OT), Richard G. Brown, PhD, Jean-Pierre Lin, PhD, Victoria Cornelius, PhD, and Helene J. Polatajko, PhD

Correspondence
Ms. Gimeno
hortensia.gimeno@gstt.nhs.uk

Neurology® 2019;92:e1212-e1224. doi:10.1212/WNL.0000000000007092

- Cognitive Orientation to daily Occupational Performance
  - Set a focused goal
  - Iteratively apply **single, feasible, and individualized** changes to achieve goal
- Example Goals:
  - Eating ice cream
  - Tying a tie

CO-OP

ARTICLE CLASS OF EVIDENCE

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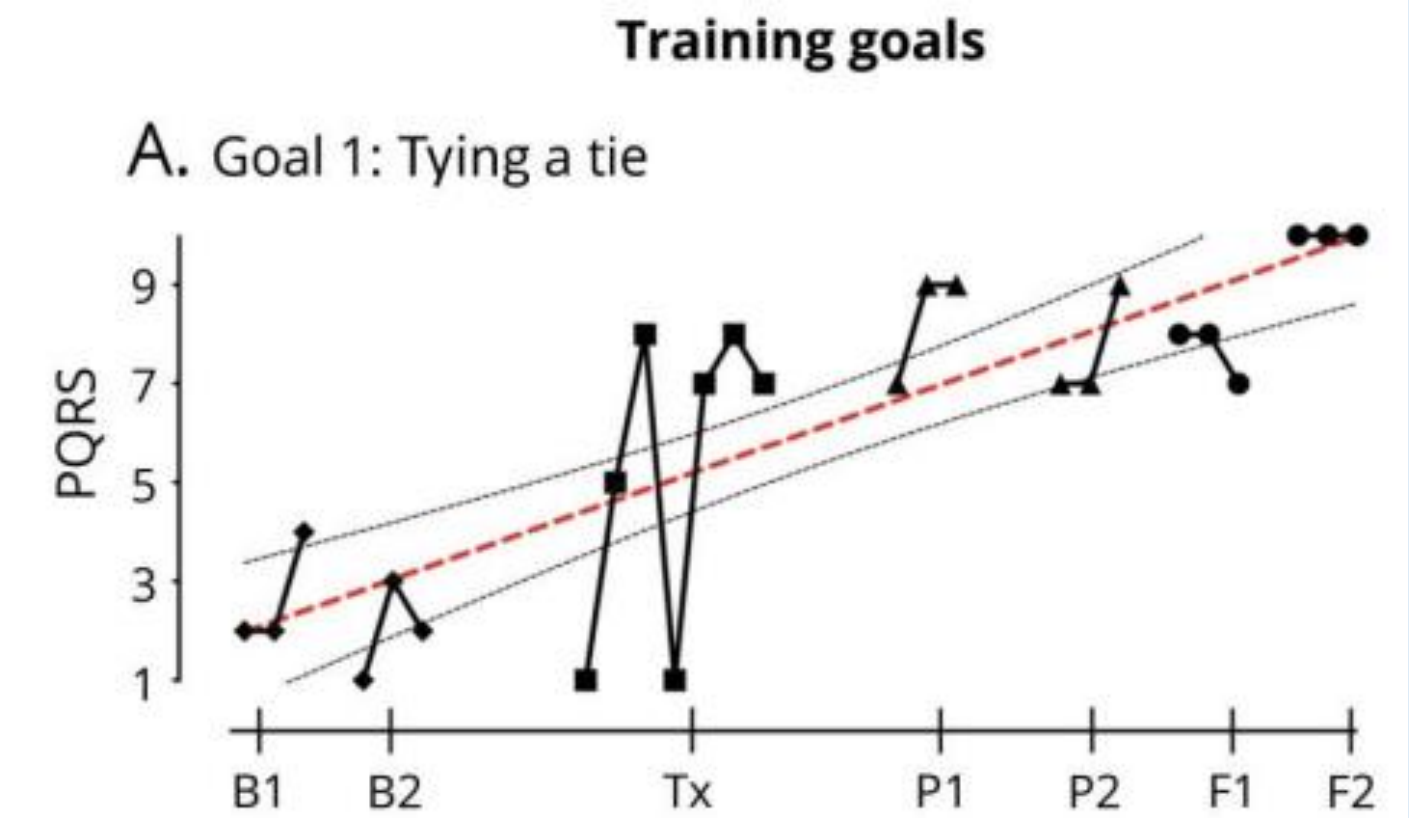

COPM
Canadian Occupational Performance Measure

D-FIS

DEVELOPMENTAL MEDICINE & CHILD NEUROLOGY ORIGINAL ARTICLE

The Dyskinetic Cerebral Palsy Functional Impact Scale: development and validation of a new tool

KIRSTY STEWART[1,2,3] | JENNIFER LEWIS[1] | MARGARET WALLEN[3] | NATASHA BEAR[4] | ADRIENNE HARVEY[5]

D-FIS Parent Report – SAMPLE COPY

SITTING: ability to sit

0	**No impact**: able to sit independently without support with good balance
1	**Mild impact**: dyskinesia has some impact on sitting ability and balance
2	**Moderate impact**: dyskinesia interferes with ability to sit independently, minimal postural supports are required for sitting and balance
3	**Severe impact**: dyskinesia interferes with sitting ability and balance, full postural supports are required
4	**Extreme impact**: dyskinesia prevents ability to sit even with maximal support
NA	*Sitting is difficult but not due to dyskinesia*

STANDING: ability to stand

0	**No impact**: able to stand independently with good balance
1	**Mild impact**: dyskinesia has some impact on standing and balance
2	**Moderate impact**: dyskinesia interferes with ability to stand independently, minimal supports are required for balance

1 = Not a priority
2 = Small priority
3 = Medium priority
4 = Highest priority

What priority is sitting ability?

1 2 3 4

What priority is standing ability?

Community driven dystonia research agenda

Laura Gilbert

1. Develop new treatments
2. Assess rehabilitation and psychological management approaches
3. C[illegible]nts
4. I[illegible]nts
5. Assess the impact of mixed tone

- **Assess for triggers** – treat triggers first (refer as needed)
- **Set focused goals** – address single goals at a time

Cerebral Palsy Research Network

Gilbert...Aravamuthan, *Neurology.* 2022

ACKNOWLEDGEMENTS

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-Toni Pearson
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-Hanyang (Ben) Miao
-Sarah Smith
-Sushma Gandham
-Kat Gemperli
-Keerthana Chintalapati
-Laura Gilbert
-Alyssa Smith
-Nathan Suh
-Victoria Zhang
-Frances Avila-Soto
-Arohi Saxena
-Gazelle Zerafati-Jahromi
-Esra Pehlivan

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-Amit Mathur

University of Texas Southwestern

-Bill Dauer
-Sam Pappas

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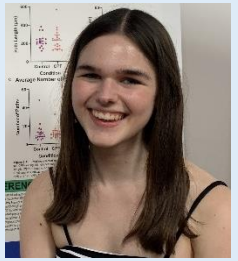
5K12NS098482-02
WashU Dept. of Neurology
McDonnell Center
SLCH Foundation

aravamuthanb@wustl.edu @drbhooma sites.wustl.edu/aravamuthanlab

Dystonia diagnosis: community expertise – GMFCS I-III


Alyssa Rust


Fayza Jaleel

YES to both?

88%

had dystonia (PPV)

NO to both?

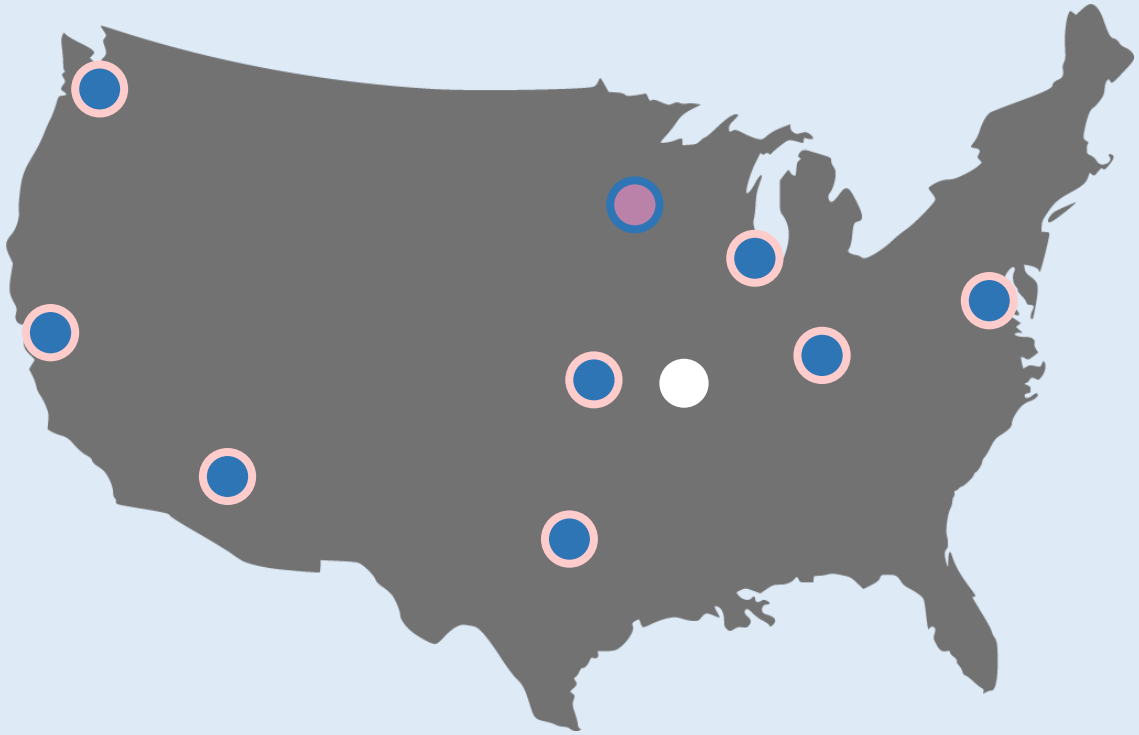
90%

did NOT have dystonia (NPV)

Jaleel, Rust....Aravamuthan. *Annals Clin Trans Neurol* 2023

Looking at overflow leg adduction as a sign of dystonia

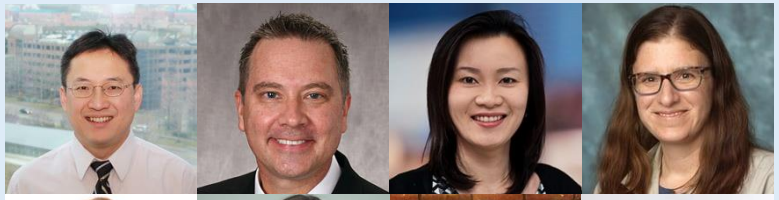
Keerthana Chintalapati



Tim Feyma Tom Novachek

Gillette Children's Specialty Healthcare

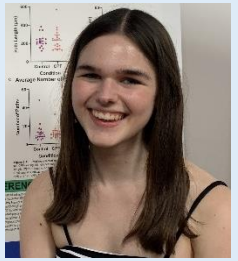
Steve Wu | Michael Kruer | Dararat 'Pam' Mingbunjerdsuk | Joanna Blackburn



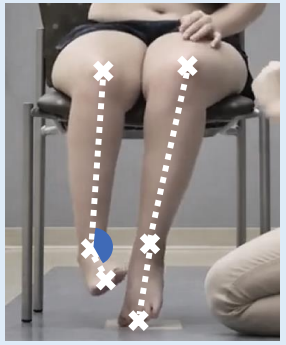
Rose Gelineau-Morel | Laura Tochen | Jeff Waugh | Jen O'Malley

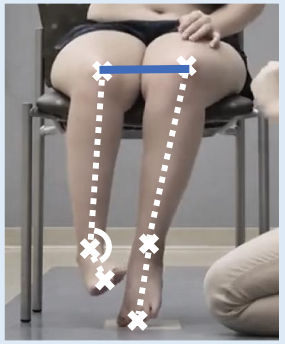
Gemperli....Aravamuthan. *bioRxiv* 2023, *In submission*

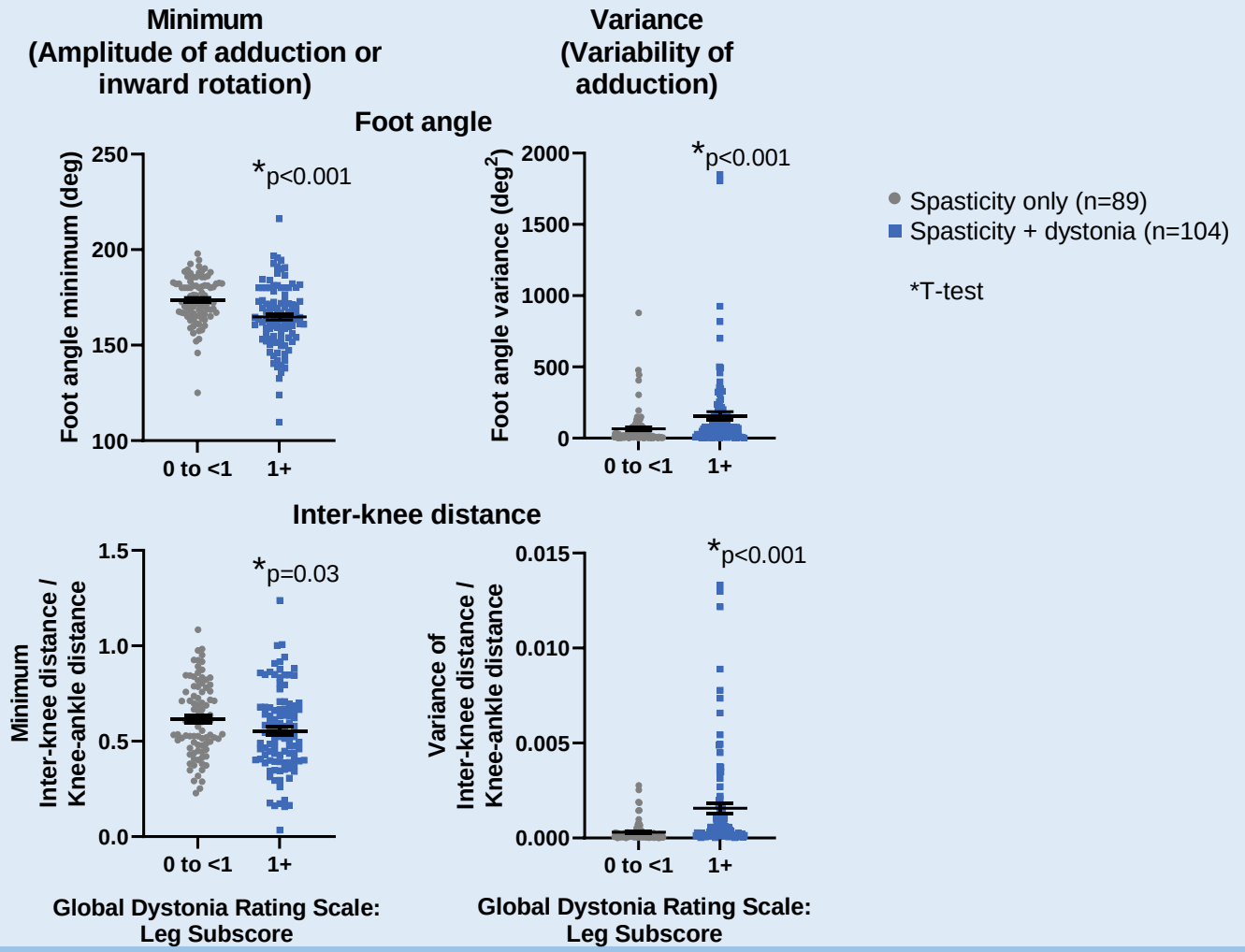
Looking at overflow leg adduction as a sign of dystonia


Alyssa Rust


Nathan Suh

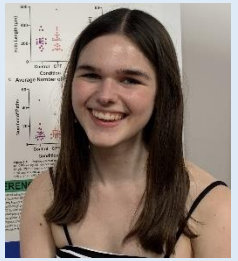




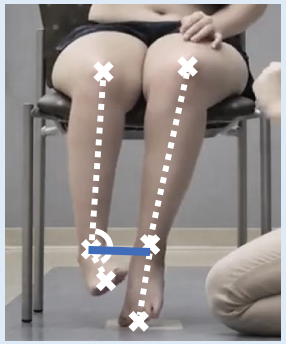


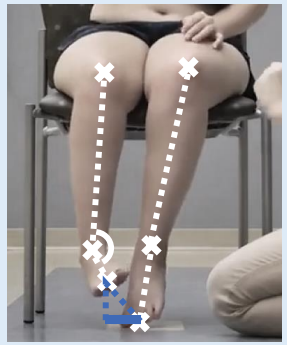
Gemperli....Aravamuthan. *bioRxiv* 2023, *In submission*

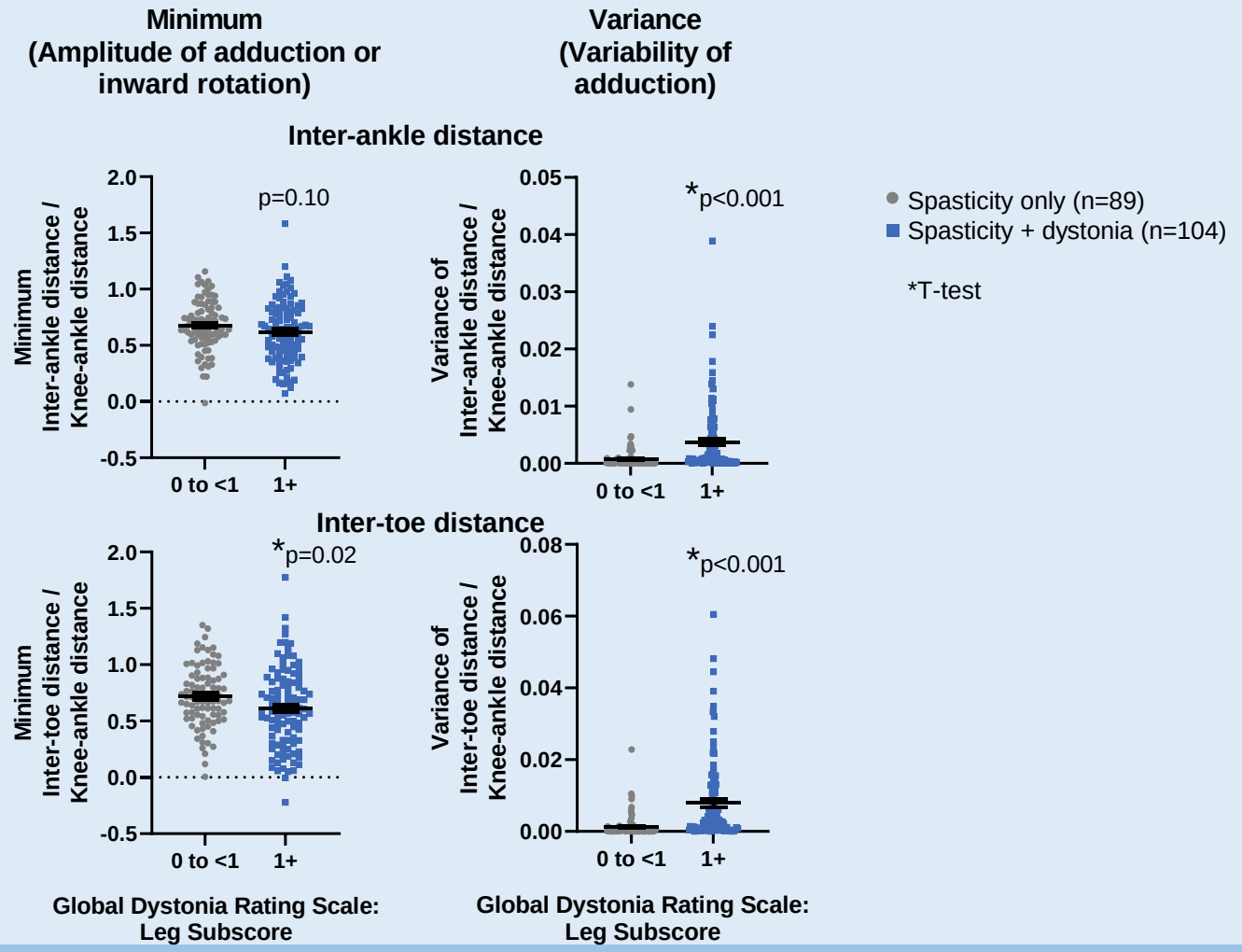
Looking at overflow leg adduction as a sign of dystonia


Alyssa Rust


Nathan Suh

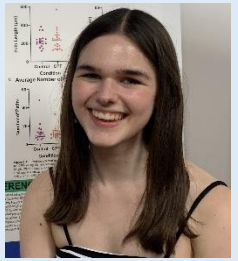




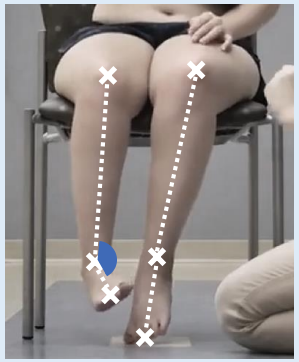


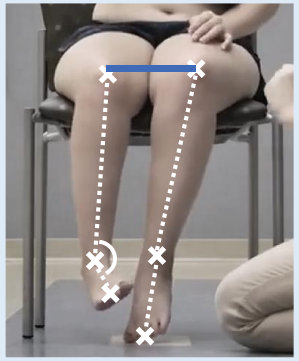
Gemperli....Aravamuthan. *bioRxiv* 2023, *In submission*

Looking at overflow leg adduction as a sign of dystonia


Alyssa Rust


Nathan Suh

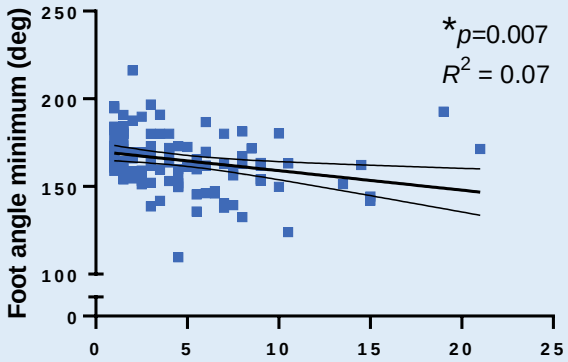


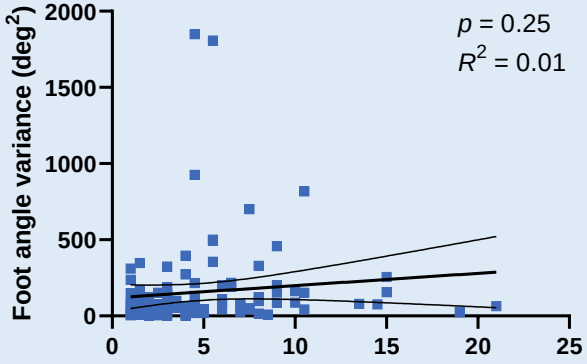


Minimum (Amplitude of adduction or inward rotation)

Variance (Variability of adduction)

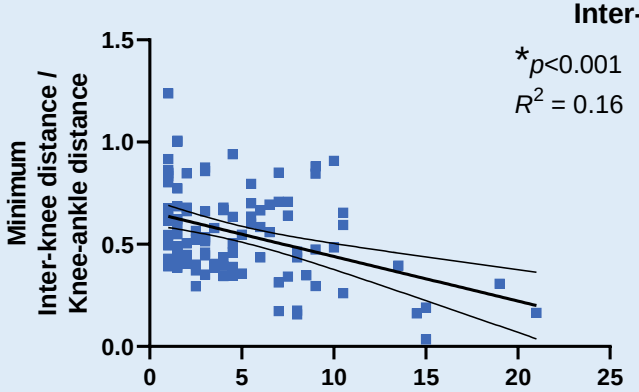
Foot angle

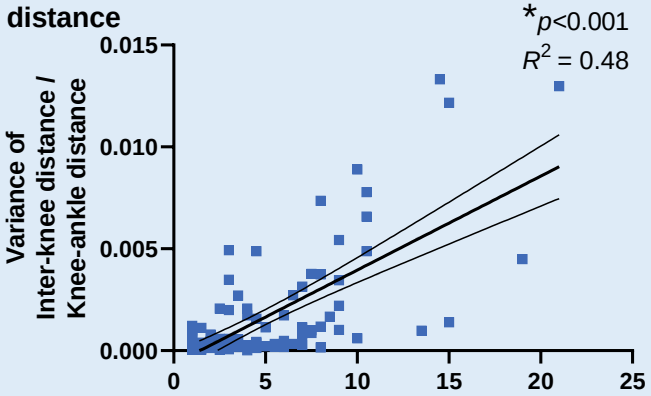




*Multiple linear regression (n=104)

Inter-knee distance





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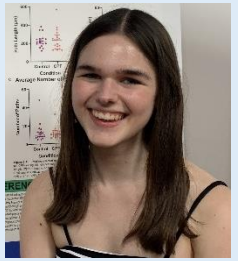
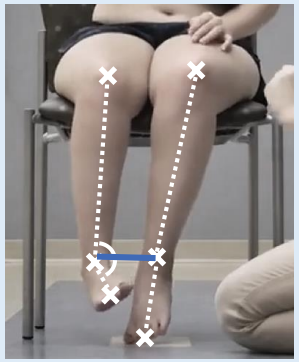
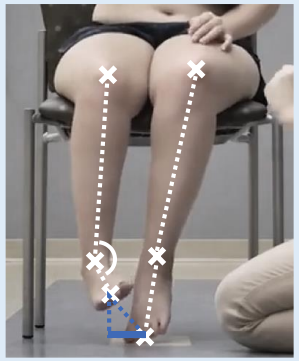

Alyssa Rust


Nathan Suh

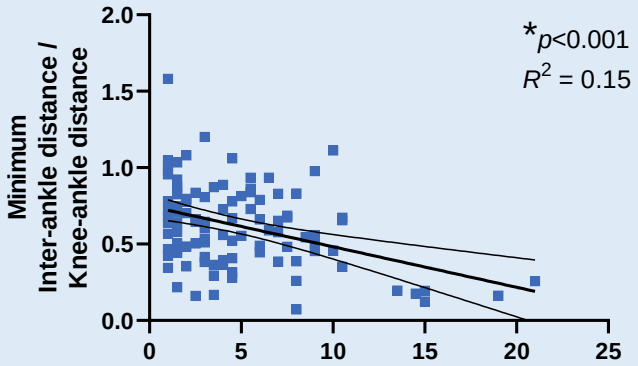


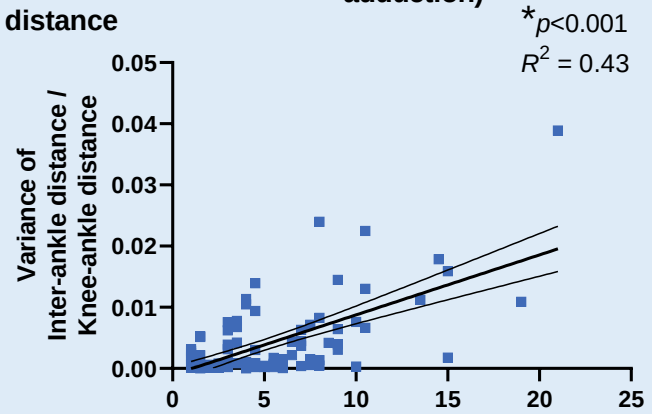


Minimum (Amplitude of adduction or inward rotation)

Variance (Variability of adduction)

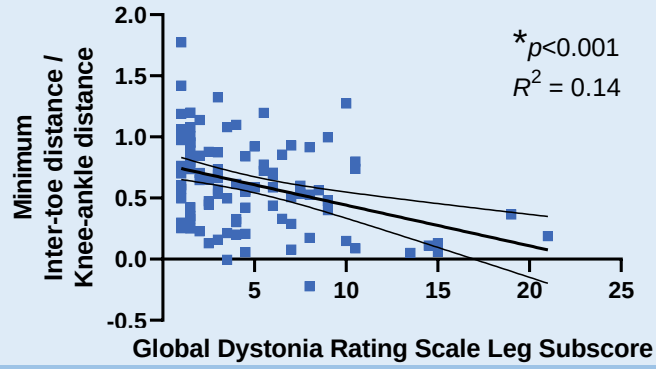
Inter-ankle distance

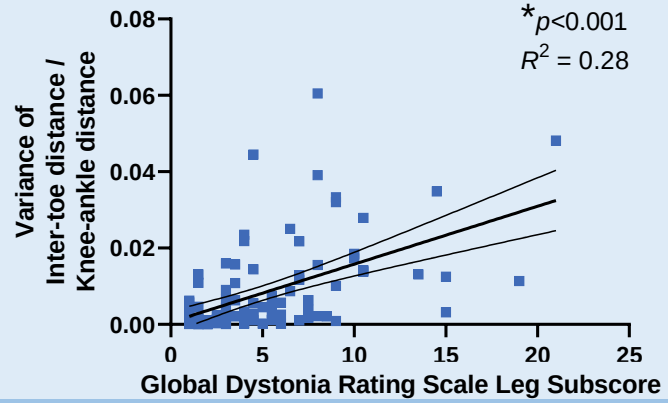




*Multiple linear regression (n=104)

Inter-toe distance





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NEMOURS HEALTH SYSTEM CEREBRAL PALSY CONFERENCE FOR PEDIATRIC THERAPISTS

Neuro-Orthopedic Care for Conditions that are 'Cerebral Palsy Like': Key Similarities and Differences

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October 23, 2023

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Cerebral palsy

- CP most common cause physical disability in children
- Static encephalopathy but progressive MSK pathology
- Equinus and hip displacement most common
- Variable phenotype

Used with permission

Occurs in 2-2.5 per 1000 live births (Stanley et al. 2000)

100 per 1000 live births in extreme prematurity (Platt 2007; Robertson 2007)

2

CP: Formal Definition

"a group of permanent disorders of the development of movement and posture, causing activity limitation…attributed to *non-progressive* disturbances …in the *developing fetal or infant brain*.".

"…often accompanied by disturbances of sensation, perception, cognition, communication and behaviour…epilepsy and…secondary musculoskeletal problems".

(Rosenbaum et al, DMCN 2007)

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3

Cerebral palsy is not a diagnosis

"…refers not to a specific disease entity, but rather to a group of conditions with variable severity that has certain developmental features in common." (Graham, Rosenbaum et al, Nature Reviews 2016)

NIH describes "brain damage" causing CP to include:

- Periventricular leukomalacia (premature, diplegia)
- Intracranial hemorrhage (hemiplegia, quadriplegia)
- Hypoxic ischemic encephalopathy (quadriplegia)
- Cerebral dysgenesis (genetic causes)

(NIH: National Institute of Neuro Disorders & Stroke)
https://www.ninds.nih.gov/health-information/disorders/cerebral-palsy

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4

Patterns of Involvement

Motor Type		Topography	
Spastic	85%	Hemiplegia	30%
Mixed	6.5%	Diplegia	24%
Dyskinetic	1.5%	Quadriplegia	32%
Hypotonic	3%		
Ataxic	3%		

(Howard, Soo et al, J Paediatr Child Health 2005)

CP "look-alikes" often have hypertonia and bilateral involvement

5

Mimics of Cerebral palsy: similar characteristics

- **Spasticity**
- **Muscle contractures**
- **Gait abnormalities**
- **Hip displacement**
- **Scoliosis**
- **Comorbidities:**
  - epilepsy, pulmonary

- MECP2 disorders (Rett/MECP2 Duplication)
- Hereditary spastic paraplegia
- Angelman's Syndrome
- PURA syndrome
- Glutaric acidemia Type 1 (Amish)

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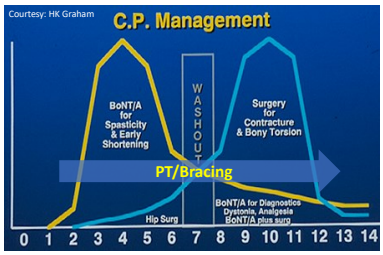
Treatment continuum for CP: Does it apply to mimics?



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Surgical principles in cerebral palsy

- Sort out spasticity
- Know your patient (strength, SMC)
- Minimise soft tissue surgery
- Maximise Bony (Lever Arm) Surgery
- Reduce displaced hips
- Balanced spine/level pelvis

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Principles of hip reconstruction

- Before femoral head/acetabulum too deformed
- Varus derotational osteotomy (VDRO) to reduce the hip
- NSA target lower with increasing GMFCS
  - GMFCS I-III: 120 °
  - GMFCS IV: 110° (some walking ability)
  - GMFCS V: 100°
- Acetabuloplasty for stability
- If adductor contractures, soft tissues first
- Femoral derotation to 10° (from 40° typically)

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Patient/Family Goals of Scoliosis Surgery

- Decreased caregiver burden
- Functional seating
- Decrease Pain
- Improve quality of life
- Minimize complications

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Hereditary spastic paraplegia: diplegia mimic

- Corticospinal/dorsal spinal cord axonal atrophy
- 0.1-10/100,000, triggered various times: infant to adult
- Manifestations: often mistaken for CP diplegia
  - Bilat lower limb spasticity
  - Muscle weakness
  - Gait abnormalities

Family History of CP should prompt investigations for HSP

- Many inheritance patterns, >80 genes involved (SPAST)
- Pure and complicated types
  - Pure most commonly seen by MSK practitioners
  - Complicated = ++comorbidities (epilepsy, dementia, etc.)

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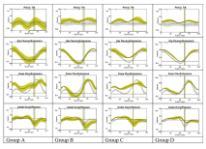
Orthopedic Aspects of HSP

- Progressive deterioration of corticospinal tracts
  - Variable rate of progression/age at onset (birth to early teens)
- Spastic diplegia is different (improving function)
- Gait abnormalities
  - Jump, Crouch, stance knee hyperextension, scissoring
  - Contractures of Achilles, hamstrings, adductors common
  - Typically symmetrical
- Foot deformities (planovalgus, cavus, equinus)
- Hip displacement
  - Prevalence unknown, progression unknown



(Nichols et al, Gait & Posture 2007)

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HSP Treatment: Many Options, Little Evidence

- **Physical Therapy**
- **Orthotics**
- **Spasticity Management**
  - Oral medication
  - BoNT-A
  - Intrathecal baclofen (ITB) pump
  - Selective dorsal rhizotomy
- **Orthopedic surgery**
  - Multilevel surgery (MLS) for gait correction
  - Preventative/reconstructive for hip displacement



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Hereditary Spastic Paraplegia: Orthopedic Surgery

- **Sparse, expert opinion, case reports**
- **Very slow progression typical**
  - "…results of surgical lengthenings should be expected to last…" *
- **<u>Most common ortho procedures:</u> hamstrings, heel cords/calf, hip adductors**
- **Improvement in gait expected**
  - knee extension during stance, scissoring, and equinus
- **Planovalgus feet also common**
  - May require bony fusions
  - Facilitates bracing for foot positioning
- **Spasticity typically persists but surgical benefits often maintained**
- **May be resistant to surgery: heterogeneous phenotype**

*(Dennis & Green, JPO 1988)

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Case: HSP with knee hyperextension and symptomatic spasticity

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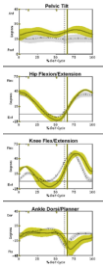
HSP: 10yoM, no prior surgery, no BoNT-A: KNEE HYPEREXTENSION

- Coronal plane ok
- Rectus spasticity on Duncan-Ely Test
- BUT ankle equinus and forward lean on crutches



Rectus femoris transfer performed
- To decrease knee hyperextension?

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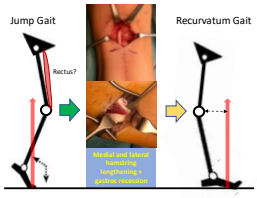
Knee Recuvatum in CP: Rectus can have a role



IN CP: HAMSTRINGS LENGTHENED + GRF ANTERIOR TO KNEE
UNOPPOSED RECTUS SPASTICITY
PLANTARFLEXION-KNEE EXTENSION COUPLE
= KNEE HYPEREXTENSION

- **Overactive plantarflexion-knee extension couple**
  - Early stance = tight plantarflexors, FF WB
  - Late stance = foot flat, 2nd ankle rocker arrested
- **Extensor spasticity/patterning (rectus femoris)**
- **Forward lean in HSP exacerbates**
  - GRF ++ in front of knee

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HSP: 11yoM, postop rectus transfer: any better?

Pre-op | **1 yr Post-op rectus femoris transfer**

Definite improvement in knee hyperextension: will it last?

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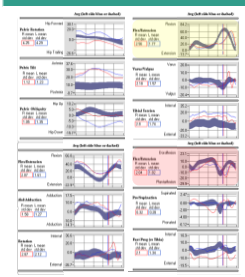
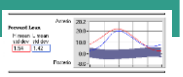
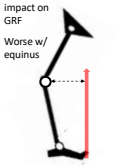
HSP: 11yoM, postop rectus transfer





Forward lean impact on GRF

Worse w/ equinus



- Improved hyperextension
- Equinus with toe drag
- Mild scissoring
- Internal tibial torsion

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HSP: 15yoM, knee hyperextension OK but ++Spasticity

ITB pump planned for lower limb spasticity

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HSP: 18yo, 2 yrs post ITB pump implantation

Pre-op ITB

2 years Post-op ITB

Is the gait any better? Subjectively yes

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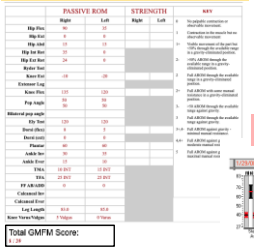
HSP: 23yo, Where are we now?

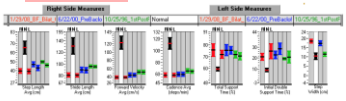
Physical Exam: Static Contractures OK (except knee)



	Right	Left
Iliopsoas	3	4
Glut Max	2+	4
Adductor	3	3
Rectus Femoris	1+	2
Rectus Catch Angle		
Vastus Medialis	3	4
Hamstring	3	4
Post Tib	3	3
Ant Tib	3	3
Gastroc	3	3/4
Clonus	CONT	CONT
Knee Jerk	4	4
Ankle Jerk	2+	2+
Babinski	UP	UP
Proximal Muscle Tone	3	4
Distal Muscle Tone	3	3

MAS: Spasticity persists despite ITB

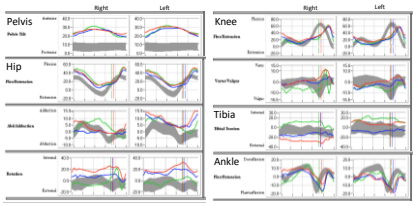
Temporospatial: worsened over time



22

HSP: 23yo, Where are we now?

Kinematics:
Increased internal femoral rotation/external tibial rotation over time
Sagittal plane unchanged despite ITB and rectus transfer



Sobering experience for spasticity management in HSP

Persistence despite aggressive treatment (ITB) over long term

Torsional abnormalities worsen regardless (like CP)

Condition Labels
- 1/29/08_BF_Bilat_Crutches
- 6/22/00_PreBaclofen
- 10/25/96_1stPostRectus

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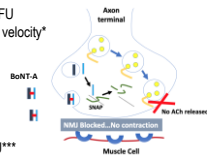
Hereditary Spastic Paraplegia: Spasticity Managment

- **Botulinum toxin**
  - Uncontrolled studies to date, very low quality, short term FU
  - BoNT-A + PT (18w) reduced spasticity and improved gait velocity*
- **Intrathecal Baclofen**
  - Reduced spasticity and improved walking score**
  - Low complication rate
- **Selective dorsal rhizotomy**
  - Systematic Review: case series' only, 6 and 12 month FU***
  - Significant reduction in spasticity scores, low complication rates
  - Minimal orthopedic outcomes reported



*(de Neit et al, J Rehabil Med 2015), **(Margetis et al, Clin Neurol Neurosurg 2014)
***(Bellofatto et al, Front Neurol 2019)

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Case: HSP with equinus, hip displacement

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HSP: 3yoM, 05/2018, DOB 03/2015, Pre-ITB pump (09/2018)

Mother and grandfather with HSP
Severe spasticity lower limbs, upper limbs normal
Gait video pre-op ITB pump insertion

- Stiff, flexed knees
- Internal foot progression, R>L
- Uses arms to propel with Kaye walker

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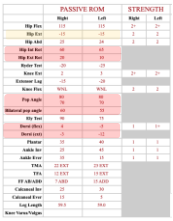
HSP: 7yo, 04/2022, Post ITB pump, Equinus gait/knee hyperextension

Concerns: feet cross over each other, foot turns in
Previous surgery: ITB pump insertion 09/2018

GMFM-D = 19/39 FMS 3/3/1

- Left equinovarus foot
- Right planovalgus foot
- Knee hyperextension (equinus + forward trunk lean)



	PASSIVE ROM		STRENGTH	
	Right	Left	Right	Left
Hip Flex	110	115	2+	2+
Hip Ext	-15	-15	2	2
Hip Abd	25	24	2	2
Hip Int Rot	60	65		
Hip Ext Rot	50	50		
Hip Ext Rot	-20	-19		
Knee Ext	2	3	2+	2+
Extension Lag	+15	+20		
Knee Flex	WNL	WNL	2	2
Pop Angle	80	80		
Bilateral pop angle	60	55		
Hip Test	90	75		
Dorsi (flex)	8	5	3	3+
Dorsi (ext)	-5	-12		
Plantar	35	40	3	3
Ankle Inv	25	45	3	3
Ankle Ever	10	15	3	3
TMA	22 EXT	29 EXT		
TFA	12 EXT	15 EXT		
FF ADAIR	7 ADD	15 ADD		
Calcaneal Inv	25	30		
Calcaneal Ever	15	5		
Leg Length	59.5	59.0		
Knee Varus/Valgus				

- Hamstrings
- Gastrocs
- Internal femoral torsion

MAS

	Right	Left
Hip Flexors	1	1
Hip Abd	1	1
Adductor	2	2
Hamstrings		
Rectus Femoris	1+	1+
Iliotibial Band	1+	1+
Iliopsoas	1	1
Gastroc	1+	1+
Soleus	1+	1+
Tibialis Anterior	1+	1+
Tibialis Posterior	1+	1+
Peroneal Muscle Group		
Distal Muscle Group		

Spasticity improved but still present

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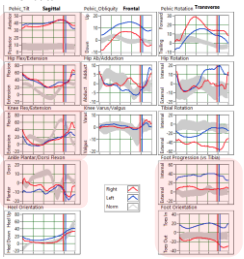
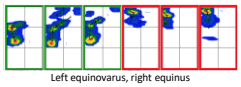
HSP: 7yo, 04/2022, DOB 03/2015, Equinus gait/knee hyperextension

Kinematics



Pedoborography



Left equinovarus, right equinus

Metabolic Cost

Derived Measure Summary				Pred. and Index based on K5 Data	
	Actual	Pred	Index	Low Ind	Hi Ind
O2 Cost	0.494	0.258	1.295	0.773	1.572
VO2 (Rest)	6.986	7.323	-0.094	-1.895	2.018
VO2 (Walk)	20.573	16.569	0.761	-1.457	2.308

NEMOURS CHILDREN'S HEALTH

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HSP: 7yo, 04/2022, DOB 03/2015, Equinus gait/knee hyperextension

Gait Lab Interpretation

Issues Impacting Gait Function : 7 year old boy with HSP who has increasing planovalgus on the right and equinovarus on the left making his brace wear uncomfortable. His preferred gait pattern is with. swing through gait although he can reciprocate. He also has limited hip abduction and almost no knee flexion in swing. Gait is relatively efficient and demonstrates good conditioning.

Surgical Recommendations: MLS

Diagnostic	
	Recommend post operative gait analysis in one year to document outcome
Notes:	He has not had an evaluation of his hip xrays which he needs for final surgical planning
Surgical	
	Bilateral gastrocnemius recession to improve premature heel rise and high early plantar flexion moment.
Notes:	due to his age he likely would benefit from soft tissue lengthenings of his plantarflexors, maybe the tibialis posterior on the left and possibly the peroneals on the right just to allow him to tolerate his solid AFOs.

- Still some hip displacement + adductor spasticity (MAS=2)
  - Adductor/iliopsoas lengthenings added

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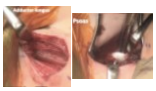
HSP: 7yo, 05/2023, DOB 03/2015, 10 mos post soft tissue MLS

Soft Tissue MLS, 07/22:
Bilat gastroc recession
Left tibialis posterior recession
Left plantar fascia release
Bilat hip adductor longus releases
Bilat iliopsoas recessions

- Left tibialis posterior recession for hindfoot varus
- Left plantar fascia release for midfoot cavus



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Case: HSP with crouch/lever arm dysfunction

NEMOURS CHILDREN'S HEALTH

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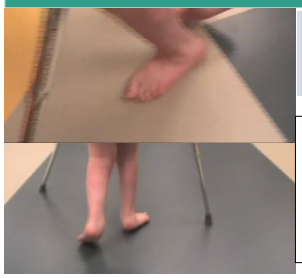
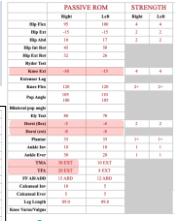
HSP: 16yo, Crouch, lever arm dysfunction, no prior surgery



- Two siblings with HSP
- Ambulates with solid AFOs
- Forearm crutches, W/C for distance
- (FMS 3/3/1)

MAS



NEMOURS CHILDREN'S HEALTH

GMFM-D =13/39

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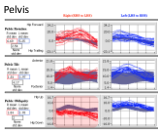
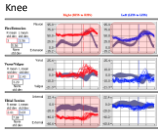
HSP: 16yo, Crouch, lever arm dysfunction

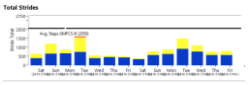
Pelvis



Knee



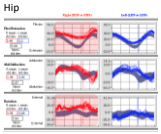
Total Strides



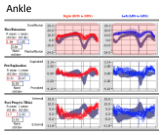
Walking activity: well below TD, even for GMFCS III

Severe lever arm dysfunction and crouch

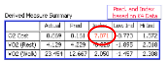
Hip



Ankle



Metabolic Cost: Severe energy expenditure



NEMOURS CHILDREN'S HEALTH

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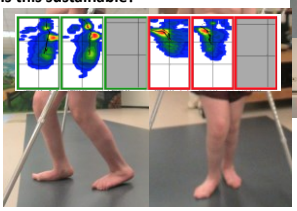
HSP: 16yo, Crouch, lever arm dysfunction

Flexed knees with capsular contracture
Internal femoral torsion, external tibial torsion
Severe planovalgus feet, hallux valgus

Is this sustainable?



In CP diplegia:
Femur DROs (DFEO)
Tibial DROs
Foot reconstruction

Is this appropriate for HSP?

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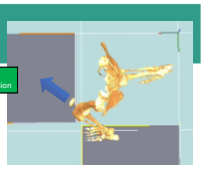
Lever Arm Dysfunction

Planovalgus foot, poor lever arm



"Improvement of crouch can be achieved by correction of foot deformity alone, with no additional knee surgical intervention, just by improving lever arm function."
(Kadhim & Miller, Gait Posture 2014)

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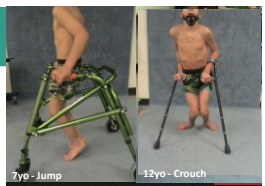
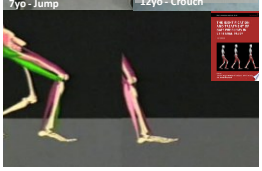
Effects of Crouch Gait

- **Flexed knee, excessive equinus**
  - GRF behind the knee, increased knee extension moments
- **Lack of endurance, high energy expediture**
  - Measured by O_2 cost in the gait lab
- **Anterior knee pain, traction at inferior pole patella**
  - Not reliably improved with surgery
- **Crouch is part of natural history in bilateral CP**
  - Not only iatrogenic

Alfred I. duPont Hospital for Children (Kanashvili, Miller et al, Gait Posture 2021)





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HSP: 16yo, Surgical prescription, Crouch, lever arm dysfunction

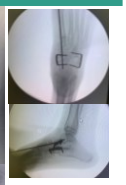
Gait Lab Interpretation

Issues Impacting Gait Function - 13 year old boy with a complex form of FSP who is being evaluated for possible correction of his gait. He is functioning at a very high level for the severity of his disability. He has a very high energy cost of walking with severe hamstring contractures, knee flexion contractures worse on the right and severe planovalgus foot worse on the right.

Muscle Tone - Also the quantity is likely an important support and stability mechanism for his gait, he would be a poor candidate for tone reduction.	
Behavior and Cognitive Function:	No problem
Motor Control and Static Balance:	Moderate impairment limiting gait function
Muscle Strength:	Weakness the major impairment
Movement Disorder:	None
Muscle Tone:	Spasticity associated impairment to gait
Stance Stability:	Bilateral Planovalgus feet
	Bilateral High knee flexion stiffness crouch

Surgical Recommendations: MLS

Surgical	
	Bilateral Hamstring lengthening medial and lateral to improve knee extension at foot contact and midstance.
	Bilateral Knee capsulotomy to decrease fixed knee flexion contracture.
	Bilateral Gastrocnemius recession to improve premature heel rise and high early plantar flexion moment.
	Bilateral Planovalgus correction to improve stance stability and moment arm malalignments.
	Bilateral Ilopsoas correction to improve push off power comfortable shoe wear.
Notes:	One option for the right side would be to do an extension and derotation osteotomy of the right distal femur, this would allow correction of the torsion and knee flexion, and tibial derotation, however he is functioning at a relatively high cost of walking and he may have problems recovering from this level of surgery. Because of this the above suggestion is made which will not directly improve the malalignment but should lead to improvement in walking.



Double arthrodesis
1st MTP fusion

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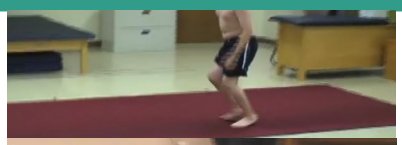
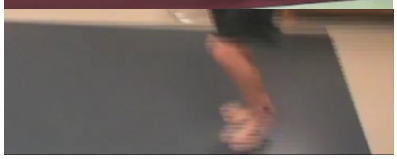
HSP: 14yo, crouch, MLS: Soft tissue + foot reconstruction

Preop



Postop



Surgical prescription from gait analysis

- Bilat Iliopsoas lengthening
- Bilat hamstring lengthening
- Bilat posterior knee capsulotomy
- Bilat gastroc lengthening
- Bilat foot reconstruction
- Bilat hallux valgus repair

- Improved knee alignment
- Reduced crouch
- AFOs would help increase PF-KE couple

NEMOURS CHILDREN'S HEALTH

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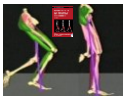
Ortho surgery in HSP: focus on prerequisites of gait

1. **Stability in stance** → Reconstruction/soft tissue for foot deformity
2. **Foot clearance during swing** → Correct equinus, stiff knee (rectus)
3. **Pre-positioning of the foot during swing** → Correct equinus/HF varus, hamstrings
4. **Adequate step length** → Correct knee flexion, initial contact problems
5. **Energy conservation** → Correct crouch, lever arm dysfunction



INSUFFICIENT EVIDENCE
Needs more studies
Not the same as spastic diplegia
BUT
Biomechanics still reigns

NEMOURS CHILDREN'S HEALTH

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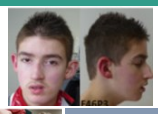
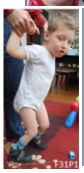
MECP2 disorders: diplegia/quadriplegia mimic

- **Rett Syndrome, 1:9000**
- **MECP2 Duplication Syndrome, 1:100,000**
- **Autistic features**
- **Developmental regression**
- **Ataxic gait ➡ non-ambulatory (variable)**
- **Abnormal tone**
- **Seizures** (disease severity)

(Miguet et al, J Med Genet 2018)





MECP2-DS more severe phenotype

Lack of "wild type" cells = X-chromosome deactivators

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MECP2 Disorders: Rett and MDS

- **MECP2 gene required for neuronal health**
  - X-chromosome, Xq28 locus
  - Dysregulation of DNA methylation/epigenetics
  - Rett Syndrome = mostly females, *de novo*
- **MECP2 dose: delicate balance***
  - **Rett** = loss of MECP2 gene function (MECP2 underexpression)
  - **MECP2-DS** = gain of MECP2 gene function (MECP2 overexpression)
- **Scoliosis most studied**
  - Prevalence from population-based studies
  - Genetic severity a risk factor

MECP2-DS

MECP2-RETT

*

(Loder et al, JPO 1989)
(Downs et al, Spine PA 2016)
(Amir et al, Nat Genet 1999)

*https://mecp2.wordpress.com/2012/04/26/xq28-mecp2-duplication-and-translocation/

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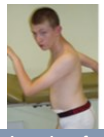
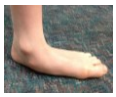
MECP2: Orthopedic Aspects

- **Spasticity but not pyramidal**
  - Infantile hypotonia ➡ hypertonia
  - Lower limbs > upper limbs
  - Contractures: ankles, knees, hips, trunk
  - BoNT-A suggested (no evidence)
- **Gait abnormalities**
  - Ataxia, regression over time
  - **Stability in Stance** important
  - Hyperlordosis
  - Crouch gait common
  - Planovalgus feet





Treat based on functional impairment rather than kinematics alone

Gait deterioration risk MECP2-DS > Rett

(Giudice-Narin et al, 2019; Lobardi et al, 2015)

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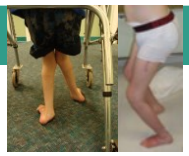
MECP2: Foot Deformities



- **Plantigrade foot important**
  - Stable platform for walking
  - Lever arm function
  - Brace tolerance
  - Standing and transfer function
  - Wheelchair footplate accommodation
  - Surgery typically based on CP principles

(Loder et al, JPO 1989)

- **Foot deformities thought prevalent in MECP2 disorders**
  - Previously no dedicated reports
  - Risk factors unknown
  - Variable treatment

NEMOURS CHILDREN'S HEALTH

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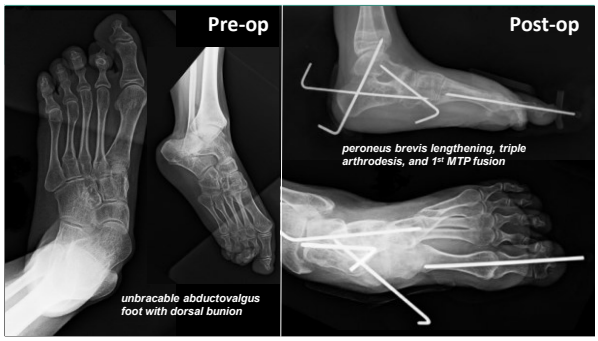

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MECP2 foot: Nemours experience

- **Seven of 56 (13%) developed foot deformities, requiring surgical management**
  - Most commonly equinus or equinovarus (**71%**)
  - Calcaneovalgus (**29%**)
- **Most common surgical procedures**
  - Achilles tendon lengthening
  - Triple arthrodesis
  - *No reported complications*
- **Most common indication**
  - Brace intolerance (standing, walking)
- **Mean age at surgery:** 15.9 (11.4-20.1) years

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Risk Factors for Foot Surgery in MECP2 Disorders

Risk Factors	Foot Deformities Requiring Surgical Treatment		P Value
	No	Yes	
Scoliosis (>40° and/or surgery)	**22 (45%)**	**6 (86%)**	**0.04***
Hip displacement (MP>30%)	**15 (31%)**	**5 (71%)**	**0.04***
Hip Surgery	**4 (8%)**	**4 (57%)**	**0.001***
Non-ambulatory	29 (66%)	6 (86%)	0.3
Genetically Severe	18 (50%)	1 (20%)	0.2
Seizures (Yes)	42 (86%)	7 (100%)	0.3
Comorbidities (Yes)	13 (26%)	4 (57%)	0.1

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MECP2: Hip displacement

- **Hip displacement thought prevalent**
  - Few reports
  - Risk factors unknown
  - Variable treatment
  - Typically based on CP principles
- **RCH Melbourne: Tay at al, 2010**
  - Only previous study on MECP2 hips (Rett)
  - 31 patients
  - **48% with MP>30%**
    - Suggested risk factors:
      - scoliosis & ambulatory status
  - No statistical analysis
  - Most non-walkers (77%)

11 yo M with MECP2 Duplication Syndrome

(Tay et al, Dev Med Child Neurol 2010)

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MECP2 hip: Nemours experience

- **High incidence in MECP2 disorders**
  - 55 patients with MECP2-related Dx
    - **MECP2-DS**: 2 patients
  - 36% overall prevalence hip displacement
  - Onset 7.7 years old
    - Peak progression 10% MP/year: 9.2 years
  - Correlated to gene severity, seizures, amb status, scoliosis
- **Case: MECP2-DS with hip displacement**
  - Standing transfers, **left hip pain**
  - No adductor contractures
  - Coxa valga
  - Acetabular dysplasia (capacious)

INDICATIONS FOR HIP SURGERY:

- Unclear
- Spastic motor type often
- Extrapolate from CP Rx

This child presented with hip pain!

(Kanashvili et al, JPO 2021)

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Risk Factors for hip displacement

Risk Factor	MP<30% N(%)	MP>30% N(%)	p-value
Non-walkers	18(50)	17(85)	0.034*
Genetically More Severe	11(31)	8 (40)	0.233
Clinically-relevant Scoliosis	12(33)	16(80)	0.001*
Presence of Seizures	29(81)	20(100)	0.040*
Two of more comorbidities	7(19)	9(45)	0.489

Given the age range for the onset of hip displacement in the current study, we suggest at least an annual AP pelvis x-ray from age 4 years

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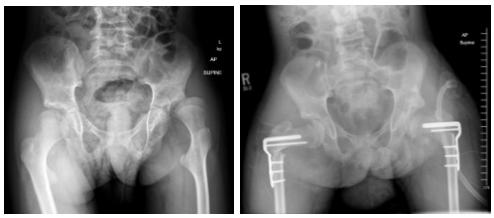
Hip reconstruction: B VDRO & L Dega



11yoM MECP2 duplication Syndrome

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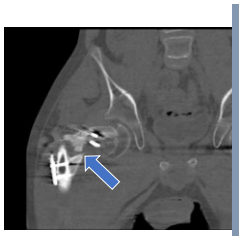
Infected non-union R VDRO: 5 mos post-op



18yoF, Rett Syndrome

- Pre-op nutritional optimization
- Cardiopulmonary assessment
- Preop ESR, Urine, CXR

Higher risk of infection

- Antibiotic beads at index procedure?
- Longer post-op Abx?
- Immunoglobulin?
- Incisional wound vac?

- Make parents aware of risk

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MECP2-DS: Scoliosis

- **Scoliosis very common (w/ kyphosis)**
  - 15-50% prevalence
  - No surgical reports in literature
  - Brace suggested " when necessary"
    - Ambulatory child
    - Seating support in non-ambulatory (soft TLSO)
- **Treat per neuromuscular scoliosis principles**
  - Progressive curve > 50°
  - Seating intolerance
  - Include pelvis if >15° pelvic obliquity
    - OR non-ambulatory

(Giudice-Narin et al, 2019; Miguet et al, 2015)

11 yo M MECP2-DS

Need regular spine surveillance – clinical/radiographic

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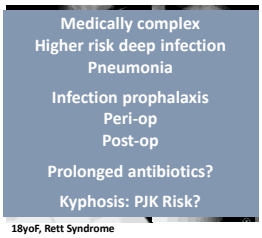
Scoliosis correction: progressive curve

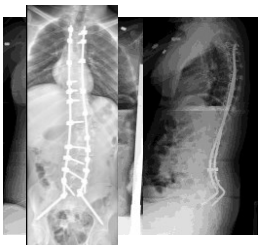
Medically complex
Higher risk deep infection
Pneumonia

Infection prophalaxis
Peri-op
Post-op

Prolonged antibiotics?

Kyphosis: PJK Risk?


18yoF, Rett Syndrome


"Balanced spine over a level pelvis"

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MECP2 Disorders: Key questions

- **Natural history of hip displacement**
  - Does surgery prevent OA/pain?
- **Surgery for gait disorders**
  - When? What? How much?
  - Flexed gait compensatory for lack of balance?
  - Does Sx prolong ambulatory potential?
- **Treatment of spasticity**
- **Natural history of scoliosis**
  - Effect on respiratory function
  - Predictors of post-op outcomes

Unlike scoliosis, hip displacement not linked to genetic severity

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Neuro-orthopedic Dx similar to CP

- Beware of HSP (family Hx)
- Spasticity treatment not predictable
- Higher risk of postop complications
- Stick to the principles for nonop/op Rx
- Reduce displaced hips
- Balanced spine/level pelvis

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Alfred I. duPont Hospital for Children

THANK YOU

NEMOURS CHILDREN'S HEALTH

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Equipment Updates:
New Developments in Seating and Positioning

Liz Koczur, MPT, PCS, CBIS
Denise Peischl, BSBME
October 23, 2023

NEMOURS CHILDREN'S HEALTH

1

Equipment

- We will review the following equipment with an emphasis on the newest or latest systems
  - Specialty Beds
    - Why's / When's / What's
  - Specialty Carseats
    - What are my options?
  - Scooters/Go-Chairs
  - Power Chair – interface technologies
    - LUCI – collision avoidance technology
    - Eye Gaze – drive control

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Beds

- Hospital Bed
  - Standard bed upon discharge from Hospital
    - Offers Head-Foot elevation; may not be electric
    - Offers High Low elevation
  - When is this bed recommended?
    - Recommended by case management recommended after surgeries
    - + Hi Lo/Head foot elevation
    - + bed rails
    - - no specialty mattress
    - - entrapment risk

3

Wound Care Bed

- Staging helps determine bed options
- Clinitron and the Emerg bed by Ethos

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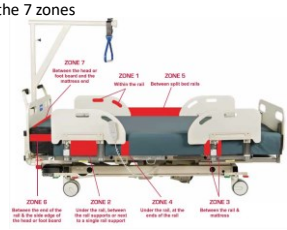
Specialty Beds

- Why are they recommended
  - Designed to address the 7 zones of entrapment



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Specialty Beds

- Sleepsafe Bed
  - **SleepSafe® - LOW BED**
  - **SleepSafe® II - MEDIUM BED**
  - **SleepSafer® - TALL BED**
  - **SleepSafe® - EXTENSION BED**
  - **SleepSafe® - BASIC BED**

NEMOURS CHILDREN'S HEALTH

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Specialty Beds

- Sleepsafe Bed
  - Who would benefit from a Sleepsafe Bed?
    - Needs specific transfer height for safety of caregiver dependent transfers
    - Needs fully enclosed to prevent elopement
    - Needs head/foot elevation for respiratory function or GI function




NEMOURS CHILDREN'S HEALTH

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Specialty Beds

- Beds by George – The Haven Bed
  - Mesh enclosed or panel enclosed
  - No high/low or head foot elevation
  - Heavy duty Mesh available
  - Low transfer height for independent users
  - Good Ventilation
  - Low transfer height

Mesh is attached to a tubular aluminum frame that is behind the "wood"

NEMOURS CHILDREN'S HEALTH

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Specialty Beds

- Beds by George – The Dream Series Beds
  - Wooden panel beds
  - Standard vs. High side panels

NEMOURS CHILDREN'S HEALTH

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Specialty Beds

- Kaesserbetten Bed
  - Panel enclosed

NEMOURS CHILDREN'S HEALTH

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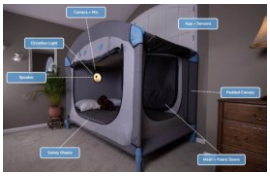
Specialty Beds

- CUBBY bed
  - Soft enclosed bed



- **Safety and sensory Canopy**
- **Circadian Light**
- **Speaker**
- **Monitor/camera**
- **Safety sheets (zip and lock to canopy wall.**
- **Motion & Sound Detection Alerts; Smoke & Carbon Monoxide Alarm Alerts; Temperature & Humidity Sensors**

NEMOURS CHILDREN'S HEALTH

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Specialty Beds

- Safety Sleeper bed
  - Soft enclosed bed – soft fabric
  - Can be used on floor or on top of bed
  - Can travel with you

NEMOURS CHILDREN'S HEALTH

12

Carseats

- Special needs carseats were originally designed for the 100+ lbs individual
- Can provide positioning
- Provides 5-point harness for older child
- Carseats are heavy (~30 lbs) and are difficult if you have to transfer the seat between vehicles

NEMOURS CHILDREN'S HEALTH

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Equipment

SPIRIT Car Seat

www.inspiredbydrive.com

- Only specialty car seat with positioners
- Min weight 25lbs to max weight of 130 lbs
- Max height: 66"
- Seat depths: 12 or 16" (with extender)

Roosevelt

www.merrittcarseat.com

- Min weight 35 lbs to max weight of 115 lbs
- Min height 33.5" to 62"
- Heights up to 5'6" - min seat depth of 12"
- Seat depths: 12.5" or 15.5 or 17" (with extender)

NEMOURS CHILDREN'S HEALTH

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Carseats

- Roosevelt options that aid in escape proof

Buckle Guard

Chest Clip Guard

NEMOURS CHILDREN'S HEALTH

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Carseats

- Carrot3 Car Seat (www.etac.com) Convaid
  - Min user weight 30 lbs
  - Max user weight 108 lbs
  - User height: 37" – 60"
  - Seat depth adjustments: 9.8" - 22.3"
  - Hip width 11.5"
  - Product weight ~20 lbs
  - One of the few you can use a transport tray

- Carrot3 Booster (www.Inspiredbydrive.com)
  - Min user weight 79lbs
  - Max user weight 165 lbs
  - Seat depths 17"
  - Hip width 16.5"
  - Back ht. (max): 33" (top of head)

NEMOURS CHILDREN'S HEALTH

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Carseats

The Churchill by Merritt Manufacturing

- 44 – 175 lbs
- 44" – 72" tall
- Seat depths of 15" or 18"
- Hip width 17"

www.merrittcarseat.com/churchillcarseat

NEMOURS CHILDREN'S HEALTH

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Power options

- Scooters/GO Chair vs. Power Wheelchair
  - When do you consider Scooter?
  - Why would you consider Scooters?

NEMOURS CHILDREN'S HEALTH

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Power options

- Scooter – Go Chair
  - Portable/disassemble for easy transport
  - Base: 4 wheels/ joystick drive control

Go Chair® Features:

- 300 lbs. weight capacity
- Up to 3.8 mph maximum speed
- 25.5" turning radius
- 8.7 miles per charge at 300 lbs., 13.2 miles per charge at 200 lbs.
- Height and width adjustable armrests
- Number one for portability
- Convenient under seat storage keeps your personal items close at hand
- Feather-touch disassembly easily disassembles into 5 lightweight pieces for transport and storage
- Your choice of six colors
- Heaviest piece is only 36 lbs.
- 18" x 17" or 20" x 18", 5-point swivel seat with latch release
- 18 amp batteries
- Large footplate
- 60° lap belt
- K0899 Power Mobility Device, not coded by DME PDAC or does not meet critieria

NEMOURS CHILDREN'S HEALTH

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Power options

- Go Go Elite Traveller
  - Portable/disassemble for easy transport
  - Max user weight 300lbs

NEMOURS CHILDREN'S HEALTH

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Eye Gaze

- Ability Drive from TOLT Technologies
  - Alternative wheelchair drive control
  - Uses a modified tablet computer and eye tracking camera to create a virtual joystick
  - User looks at appropriate graphic on the screen to move in 8 directions
  - When user looks away, chair will stop
  - https://www.tolt.tech/

NEMOURS CHILDREN'S HEALTH

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LUCI Collision Avoidance System

- LUCI is an accessory for power wheelchairs, designed to give riders a safer and more inclusive experience. It is an attachable hardware/software product which uses cloud and sensor-fusion technologies to provide security, stability, and connectivity for power wheelchairs

NEMOURS CHILDREN'S HEALTH

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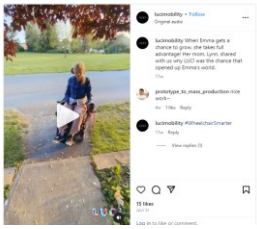
LUCI Collision Avoidance System

- Our 1st LUCI user!



NEMOURS CHILDREN'S HEALTH

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Power Assist

- Power add-on systems or Power Assist systems
  - Systems that offer power assistance for manual wheelchairs
  - Adds on to manual wheelchair
  - Improves function/lessens fatigue
  - Consider when entering middle school/high school

NEMOURS CHILDREN'S HEALTH

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Power Assist

Power assist / Power add on system

- attaches to the wheelchair axle
- wearable devices (Bluetooth) or wired controller (Switch) signal the motor to start, accelerate, and stop.
- Maneuvering and stopping the wheelchair is still guided by the user's hands on the handrims.

NEMOURS CHILDREN'S HEALTH

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Power Assist

- Smart Drive
  - Speed: 5.5 mph
  - Weight: 13.2 lbs
  - Range: 12.3 miles
  - Wired or Bluetooth connection
  - www.permobilus.com

NEMOURS CHILDREN'S HEALTH

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Power Assist

- SMOOV
  - Speed: 6 mph
  - Weight: 16 lbs
  - Range: 16 miles
  - www.smoov.com

NEMOURS CHILDREN'S HEALTH

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Power Assist

- Emotion
  - Speed: 5.3
  - Weight of each wheel: 17 lbs
  - Range: 15.5 miles
  - www.alber-usa.com/us/products

CHILDREN'S HEALTH

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Thank You

Wheelchair Seating and Mobility Clinic
Scheduling: 302-651-5850

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CP Conference for Pediatric Therapists: Case Study Session 2023

- Wade Shrader, MD
- Jason Howard, MD
- Maura McManus, MD
- Laura Owens, MD
- Bhooma Aravamuthan, MD
- Amy Bailes, PhD, PT
- Paul Enlow, PhD
- Nancy Lennon, DPT, MS, PT
- Chris Church, MPT
- Liz Koczur, MPT
- Faithe Kalisperis, PT, DPT
- Brittany Virgil, PT, DPT
- Kathleen Miller-Skomorucha, OTR/L
- Jessica Dunn, OTR/L
- Denise Peischl, BSBME

NEMOURS CHILDREN'S HEALTH

1

Objectives

- Interdisciplinary discussion and review of patient cases
- Improve audience's understanding of the medical and therapy continuum of care of a child with CP and CP-like conditions
- Improve the audience's understanding of therapy dosing and interdisciplinary team decision making regarding inpatient and outpatient therapy for a child with CP and CP-like conditions
- Discuss challenges, mood, behavioral or cognitive, that may interfere or change the course of therapy.

NEMOURS CHILDREN'S HEALTH

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3

Case 1: Brief History / Etiology

- 6 year old boy (at time of surgery)
- Medical History:
  - PVL
  - Esotropia, delayed visual maturity
  - 30 weeks gestation
    - NICU 26 days
      - Mild respiratory distress, temperature instability and hyperbilirubinemia

NEMOURS CHILDREN'S HEALTH

4

Case 1: Brief History / Etiology

- GMFCS Level IV
  - Primary mode of mobility - crawling
- Movement Disorder
  - Spasticity
  - Dystonia

NEMOURS CHILDREN'S HEALTH

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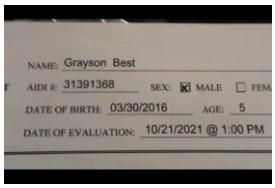
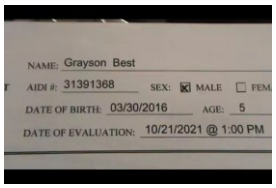
Case 1: Dystonia Video Age 5





NEMOURS CHILDREN'S HEALTH

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Case 1: Pre-op Exam

	PASSIVE ROM	
	Right	Left
Hip Abd	15	10
Knee Ext	-17	-7
Pop Angle	75 60	80 70
Dorsi (flex)	0	-10
Dorsi (ext)	-10	-20

GMFCS:	IV (Requires physical assistance or specialized devices for walking.) --Requires phys specialized devices for walking.
GMFCS:	IV – Requires physical assistance or specialized devices for walking.
FMS (5m):	C – Crawling –
FMS (50m):	1 – Uses wheelchair –
FMS (500m):	1 – Uses wheelchair –
MACS:	III – Handles objects with difficulty; needs help to prepare and/or modify activities. –
CFCS:	III – Effective Sender AND Effective Receiver with familiar partners –

TONE

	Right	Left
Iliopsoas	2	2
Glut Max	0	0
Adductor	3-4	3-4
Rectus Femoris	3	3
Rectus Catch Angle		
Vastus Medialis	1+	1+
Hamstring	2	3
Post Tib	3	3
Ant Tib	2-3	2-3
Gastroc	3	3-4

Modified Ashworth Scale	
0	Hypotonic
1	No increase in tone
1+	Slight increase in tone manifested by a catch and release or by minimal resistance at the end of range of motion
2	Slight increase in muscle tone manifested by a catch followed by minimal resistance throughout the remainder (less than half) of the range of motion
3	More marked increase in muscle tone through most of the range of motion, but affected part easily moved
3+	Considerable increase in muscle tone, passive movement difficult
4	Affected part rigid

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Case 1: Pre-op Exam

HAT Diagnosis

Diagnosis	UER	LER	UEL	LEL
Dystonia	Yes	Yes	Yes	Yes
Spasticity	Yes	Yes	Yes	Yes
Rigidity	No	No	No	No
Mixed Tone	Yes	Yes	Yes	Yes

Barry-Albright Dystonia Scale

Score Eyes	1
Score Mouth	2
Score Neck	3
Score Trunk	2
Score Left UE	3
Score Right UE	3
Score Left LE	3
Score Right LE	3
Total Score	20

8

Case 1: Pre-op X-Rays

- Orthopedic Exam/Imaging:

9

Case 1: Therapy Issues

- Equipment:
  - Power wheelchair
  - Bath chair
  - Adaptive stroller
  - Gait trainer
  - Stander

NEMOURS CHILDREN'S HEALTH

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Case 1: Surgical Management

- Surgical Procedures:
  - Proximal femoral growth modulation
  - Adductor tenotomy
  - Hip flexors tenotomy
  - Proximal HS release

NEMOURS CHILDREN'S HEALTH

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Case 1: PT Plan of Care: / Rehab Management

- Physical Therapy:
  - Acute PT in hospital
  - Outpatient PT after discharge
    - Returned to satellite location
    - Plan of care details:
      - 4 x per week for 2-3 weeks
      - 2 x per week for a total of 12 weeks

NEMOURS CHILDREN'S HEALTH

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Case 1: PT Plan of Care: / Rehab Management


NEMOURS CHILDREN'S HEALTH

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Case 1 Outcomes / Future Planning / Summary

Drs McManus / Shrader?

NEMOURS CHILDREN'S HEALTH

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Case 2: Brief History / Etiology

- 12 year old girl (at time of surgery)
- Established care at Nemours in 2022
  - Previously followed at Children's Specialized in NJ
- Birth History
  - 31 week preemie
  - 3 weeks in NICU
  - IVH Level 1

NEMOURS CHILDREN'S HEALTH

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Case 2: Brief History / Etiology

- Ambulation History
  - Began walking at 18 months with walker
  - Walked independently at age 4
- Surgical History
  - No orthopedic surgery
  - Several rounds of botox injections to hamstrings and gastrocs

NEMOURS CHILDREN'S HEALTH

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Case 2 Problem List

- GMFCS Level II
- Mari's Reported Problem List:
  - Fatigues easily
  - Back pain
  - Right knee pain
  - Frequent falling
  - Walking with bent knees
  - Resistant to walking with assistive device
    - Peers

NEMOURS CHILDREN'S HEALTH

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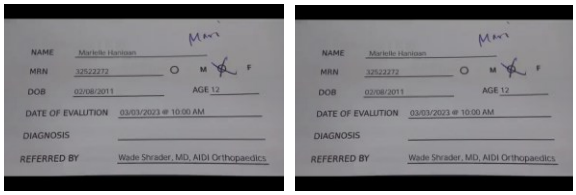
Case 2 Preop Gait Video



NEMOURS CHILDREN'S HEALTH

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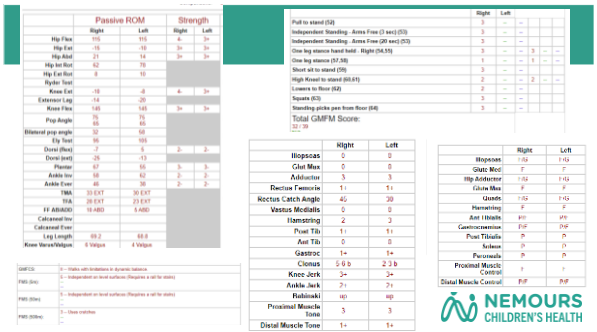

NEMOURS CHILDREN'S HEALTH

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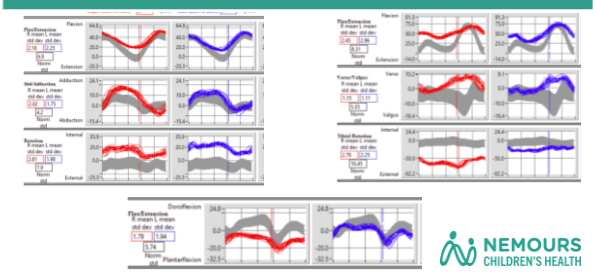
Case 2 Preop Gait Kinematics



NEMOURS CHILDREN'S HEALTH

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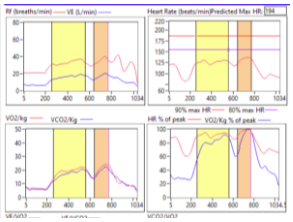
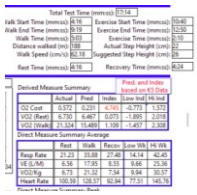
Case 2 Preop Gait Energy Cost





NEMOURS CHILDREN'S HEALTH

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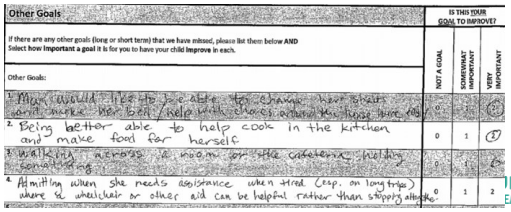
Case 2 Preop Physical Exam / X-Rays / Gait

- Preoperative Outcome Measures:
  - Goal – Gait Outcomes Assessment List – PRO Questionnaire



Other Goals	IS THIS YOUR GOAL TO IMPROVE?		
If there are any other goals (long or short term) that we have missed, please list them below AND Select how **important a goal** it is for you to have your child **improve** in each.	NOT A GOAL	SOMEWHAT IMPORTANT	VERY IMPORTANT
Other Goals:			
1. High would like to be able to change her shirt and make her bed / help with chores around the house more (4)	0		
2. Being better able to help cook in the kitchen and make food for herself	0	1	(2)
3. Walking across the room for the cafeteria / holding something	0	1	(2)
4. Admitting when she needs assistance, when tired (esp. on long trips) where a wheelchair or other aid can be helpful rather than stopping altogether.	0	1	2

RS EALTH

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Case 2 Preop X-Rays

- X-rays/Imaging:

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehabilitation

- Surgical Procedures: 5/9/2023
  - Bilateral DFEO
  - Right gastroc recession
  - Right tibial osteotomy
- Plan of care:
  - Application of long leg casts after surgery
  - Home with 3-4 weeks, WBAT
    - Casts off 5/30/23
      - Application of SLCs
  - Evaluation for inpatient rehabilitation
    - ELOS 2-8 weeks
    - Transition to outpatient PT when indicated
      - Intensive frequency recommended for at least 6 months post-op

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehabilitation

- Post-operative Imaging:

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehabilitation

- Post-operative Imaging:
- Note:
  - High anxiety obtaining post-op imaging
    - 4 people for transfer onto table
    - Doffing of hinged knee braces took over an hour

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehabilitation

- Inpatient Rehabilitation
  - Significant anxiety component during early recovery
    - Anxiety limited knee flexion ROM and initial strengthening > Pain
  - Extended time until tolerated not being fully extended in wheelchair
    - Even with braces still locked in extension
  - Limited her gait speed
    - Extremely "cautious "

First time out of her wheelchair without knee fully in extension and proud!

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehab

- Inpatient Rehabilitation:
  - Anxiety was decreased by completing goal-oriented tasks in therapy
    - Baking was used frequently
      - Calmed her anxiety
      - Reward system

NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehab

- Inpatient Rehabilitation Discharge: 7/27/23
- Transition to intensive outpatient therapies
  - CORP program
    - PT and OT 5 days a week recommended
      - Insurance visit limits

NEMOURS CHILDREN'S HEALTH

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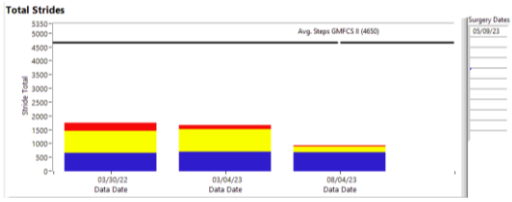
Case 2 Plan of Care: Surgical / Medical / Rehab



NEMOURS CHILDREN'S HEALTH

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Case 2 Plan of Care: Surgical / Medical / Rehab

- Equipment:
  - Manual wheelchair
    - CAT 5
      - Swing away front end
      - Desk length arm rests

NEMOURS CHILDREN'S HEALTH

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Case 2 Outcomes / Future Planning

- Physical Therapy Outcomes
  - Improved 5 rep sit-to-stand test
    - Not within GMFCS Level Norms at Discharge
  - Improved 6MWT
    - Not within GMFCS Level Norms at Discharge
- Future Planning: PT
  - Continued intensive outpatient PT recommended
    - Outpatient 3x/week (Insurance limitations)
    - School PT frequency increased
    - Hippotherapy
    - Aquatic therapy
  - Follow up 10/31/23:
    - Gait lab
    - Orthopedics

NEMOURS CHILDREN'S HEALTH

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Case 3: Brief History / Etiology

- 19 year old male (at time of surgery)
- Birth History
  - Triplet pregnancy, born at 25 weeks gestation
  - NICU
    - IVH
    - Ventilator

NEMOURS CHILDREN'S HEALTH

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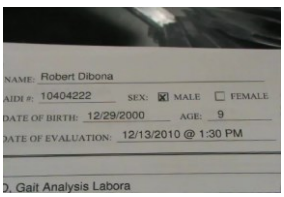
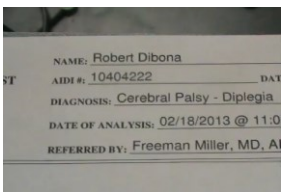
Case 3: Brief History / Etiology





NEMOURS CHILDREN'S HEALTH

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Case 3: Brief History / Etiology

- Surgical History
  - 2011 SEMLS
    - Left femoral derotation osteotomy on the left
    - Calcaneal lengthening
    - Hamstring lengthening
    - Correction of planovalgus foot deformity
    - Left rectus transfer
    - Left split tibialis anterior transfer
  - 2015
    - Baclofen Pump
    - Soft tissue (Hamstring and left UE)

NEMOURS CHILDREN'S HEALTH

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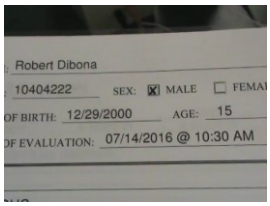
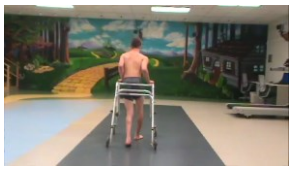
Case 3: Brief History / Etiology





NEMOURS CHILDREN'S HEALTH

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Case 3 Problem List (age 19)

- Anxiety
- Dystonia

- Robert's reported problem list:
  - Flat feet
  - Foot turns out
  - Frequent falling, Toes drag and catch on floor
  - Knees rubbing
  - Shoes wearing out quickly
  - Walking with bent knees

NEMOURS CHILDREN'S HEALTH

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Case 3 Preop Physical Exam

	PASSIVE ROM						STRENGTH	
	Right			Left			Right	Left
Hip Flex	110	110	120	115	115	120	3+	3+
Hip Ext	0	-15	0	0	-15	0	3+	3+
Hip Abd	35	30	30	32	28	32	3+	3+
Hip Int Rot	65	65	70	80	60	75		
Hip Ext Rot	35	20	20	-30	-5	15		
Ryder Test								
Knee Ext	-9	-15	-14	-20	-24	-8	3+	3+
Extensor Lag								
Knee Flex	140	WNL	WNL	140	WNL	WNL	3-	3+
Pop Angle	65	60 55	58 55	75	80 75	70 65		
Bilateral pop angle								
Ely Test	45	90	120	45	90	100		
Dorsi (flex)	15	15	16	20	22	10	3+	3
Dorsi (ext)	0	10	11	5	15	5		
Plantar	45	25	35	50	30	35	2-	3
Ankle Int		20	40		32	30	3	3
Ankle Ext		20	27		30	20	3	3
TMA		35 EXT	20 EXT	20 EXT	50 EXT	30 EXT	25 EXT	
TFA	5 EXT		10 EXT	10 EXT	50 EXT	25 EXT	5 EXT	
FF ABADD	5 ABD		0	10 ABD	25 ABD	5 ABD	15 ADD	
Calcaneal Int	20	15	25	10	5	12		
Calcaneal Ext	10	10	12	10	8	10		
Leg Length	78.2	78.0	83.0	77.4	81.0	82.8		

	Right			Left		
Iliopsoas	1+	0	0	1+	0	0
Glut Max	1-3	1+	0	1-3	0	0
Adductor	2	1	1	3	1	1
Rectus Femoris	1-4	1	1+	1-4	0	1
Rectus Catch Angle						
Vastus Medialis	1-4	1+	1+	1-4	0	1
Hamstring	3	2	1	3	1	1+
Post Tib	1+	1	0	0	0	1+
Ant Tib	1+	1	0	0	0	0
Gastroc	1+	1+	1+	1+	0	2
Clonus	2-3 b	0	-	2-3 b	0	-

NEMOURS CHILDREN'S HEALTH

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Case 3 Preop Physical Exam

Total GMFM Score:

MOBILITY SCALES

NEMOURS CHILDREN'S HEALTH

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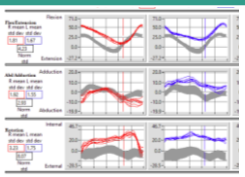
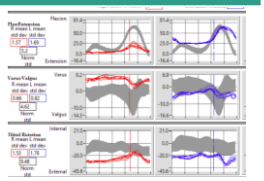
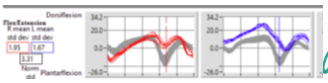
Case 3 Gait Kinematics







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Case 3 Preop Physical Exam / X-Rays / Gait

- Imaging/X-rays:

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Case 3 Preop Physical Exam / X-Rays / Gait

- Imaging/X-rays:

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Surgical Procedures: 8/25/2020
  - Bilateral distal femoral extension and derotation osteotomies
  - Right femoral shortening osteotomy
  - Right rectus release
  - Bilateral tibial derotation osteotomies
  - Left foot lateral column lengthening
  - Right MTP fusion to correct hallux valgus

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Post-op Imaging:

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Post-op Imaging:

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Post-op Imaging:

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Post-op Imaging:


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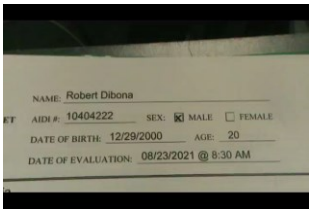
Case 3: Post-Op Gait



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Case 3 Plan of Care: Surgical / Medical / Rehab

- Rehabilitation Plan of care:
  - Application of long leg casts after surgery
  - Home with 3-4 weeks, WBAT
  - Evaluation for inpatient rehabilitation
    - ELOS 2-8 weeks
    - Transition to outpatient PT when indicated
      - Intensive frequency recommended for at least 6 months post-op
- Bumps in the Road:
  - Increased dystonia post-operative
    - Wounds on toes
      - Offloading boot on right LE
    - Decreased weight bearing on right LE
    - Increased inpatient length of stay
  - Baclofen pump failed early on during CORP outpatient therapy course
    - Large set back functionally
    - Increased outpatient intensive therapy course

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Case 3 Plan of Care: Surgical / Medical / Rehab

- Videos - dystonia limiting function:


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Case 3 Plan of Care: Surgical / Medical / Rehab

- Pictures of wounds from dystonic toe movements:


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Case 3: Summary / Outcomes

- Non-union right femur
  - Two surgical repairs
- Great functional outcomes
  - Improved PT outcome measures
    - 6MWT
    - 5 Rep Sit to Stand
  - Pain free
  - Improved functional mobility and independence
    - Working two jobs
    - Pet Owner
    - Went to Disney! Life Goal!

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Case 3 Outcomes / Future Planning

- Long journey but very happy outcome ☺
- Photo from October 2023

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Thank-You

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