Early Diagnosis of Cerebral Palsy

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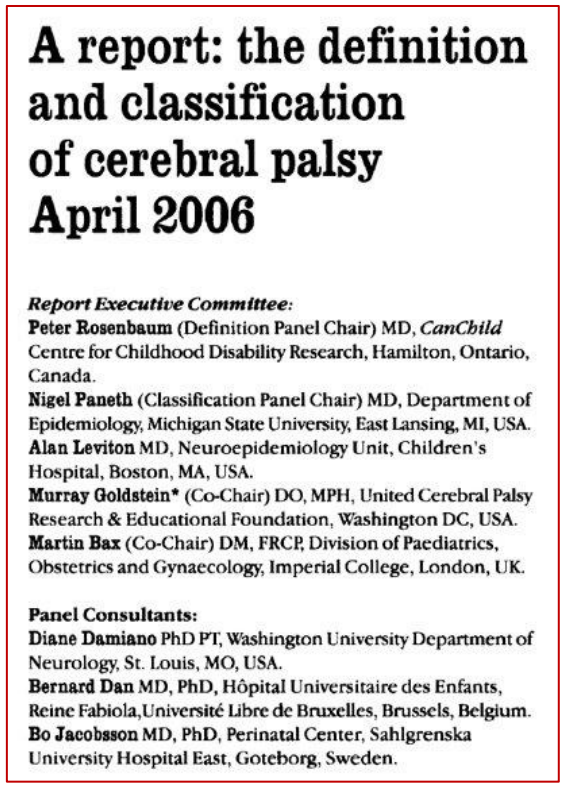
- Disclosure:

ReAlta Life Sciences

Definition of Cerebral Palsy

2006 Panel Consensus



A report: the definition and classification of cerebral palsy April 2006

Report Executive Committee:
Peter Rosenbaum (Definition Panel Chair) MD, *CanChild* Centre for Childhood Disability Research, Hamilton, Ontario, Canada.
Nigel Paneth (Classification Panel Chair) MD, Department of Epidemiology, Michigan State University, East Lansing, MI, USA.
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Martin Bax (Co-Chair) DM, FRCP, Division of Paediatrics, Obstetrics and Gynaecology, Imperial College, London, UK.

Panel Consultants:
Diane Damiano PhD PT, Washington University Department of Neurology, St. Louis, MO, USA.
Bernard Dan MD, PhD, Hôpital Universitaire des Enfants, Reine Fabiola, Université Libre de Bruxelles, Brussels, Belgium.
Bo Jacobsson MD, PhD, Perinatal Center, Sahlgrenska University Hospital East, Goteborg, Sweden.

Key features

- Definition of CP : “a group of **permanent disorders** of the development of movement and posture causing activity limitation that are attributed to **non-progressive disturbances** that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are **often accompanied by disturbances of sensation, perception, cognition, communication, and behavior, by epilepsy, and by secondary musculoskeletal problems.**“

CP Diagnosis Standardization

1. Motor symptoms due to a brain disturbance should begin before 2yo.

2. Previously acquired motor milestones should not be lost by 5yo.

3. A CP diagnosis should be provided **as early as possible** as long as motor disability without loss of motor milestones can be predicted or reported to be present at age 5.
    - Aravamuthan, et al Neurol Clin Pract, Dec 2024

Identification of CP & Prevalence

- Overall prevalence
  - Australia: 2.1/1000 live births (Smithers-Sheedy, 2016)
  - United States: 2.9/1000 live births (Durkins, 2010)
  - Pooled data : 2.1/1000 live births (Oskoui, 2013)

*Roughly half of these children were born preterm in the US study

Why focus on detection of CP before age 2?

Parent perspective

Delays in diagnosis of CP are associated with parental dissatisfaction.

Excellence in provision of care

- Early identification of CP mimics
- Implement surveillance programs
- Initiate early intervention programs to take advantage of brain plasticity
- Streamline funding and social support
- Maximize opportunities for physical and learning support

JAMA Pediatrics | Review

Early, Accurate Diagnosis and Early Intervention in Cerebral Palsy
Advances in Diagnosis and Treatment

Iona Novak, PhD; Cathy Morgan, PhD; Lars Adde, PhD; James Blackman, PhD; Roslyn N. Boyd, PhD; Janice Brunstrom-Hernandez, MD; Giovanni Cioni, MD; Diane Damiano, PhD; Johanna Darrah, PhD; Ann-Christin Eliasson, PhD; Linda S. de Vries, PhD; Christa Einspieler, PhD;

neonatal encephalopathy, infants with birth defects, and infants admitted to the NICU)

Early Detection of CP Before 5 mo CA	
3.0 Option A: The most accurate method for early detection of CP in infants with newborn-detectable risks and younger than 5 mo (CA) is to use a combination of a standardized motor assessment and neuroimaging and history taking about risk factors	Strong recommendation based on high-quality evidence of test psychometrics in newborn-detectable risk populations
Standardized motor assessment 3.1 Test: GMs to identify motor dysfunction (95%-98% predictive of CP), combined with neuroimaging	Strong recommendation based on high-quality evidence of test psychometrics in newborn-detectable risk populations
Neuroimaging 3.2 Test: MRI (before sedation is required for neuroimaging) to detect abnormal neuroanatomy in the motor areas of the brain (80%-90% predictive of CP). Note that normal neuroimaging does not automatically preclude the diagnosis of risk of CP	Strong recommendation based on high-quality evidence of test psychometrics in newborn-detectable risk populations
4.0 Option B: In contexts where the GMs assessment is not available or MRI is not safe or affordable (eg, in countries of low to middle income), early detection of CP in infants with newborn-detectable risks and younger than 5 mo (CA) is still possible and should be carried out to enable access to early intervention	Strong recommendation based on moderate-quality evidence of test psychometrics in newborn-detectable risk populations
Standardized neurological assessment 4.1 Test: HINE (scores <57 at 3 mo are 96% predictive of CP)	Strong recommendation based on moderate-quality evidence of test psychometrics in newborn-detectable risk populations
Standardized motor assessment 4.2 Test: TIMP	Conditional recommendation based on low-quality evidence of test psychometrics in at-risk populations

Novak et al. JAMA Peds 2017

DIAGNOSIS of CEREBRAL PALSY

- Clinical history
- Neurological exam
- Motor function assessments
- Neuroimaging
- Biomarkers
- Ruling out progressive disorders or other diagnoses

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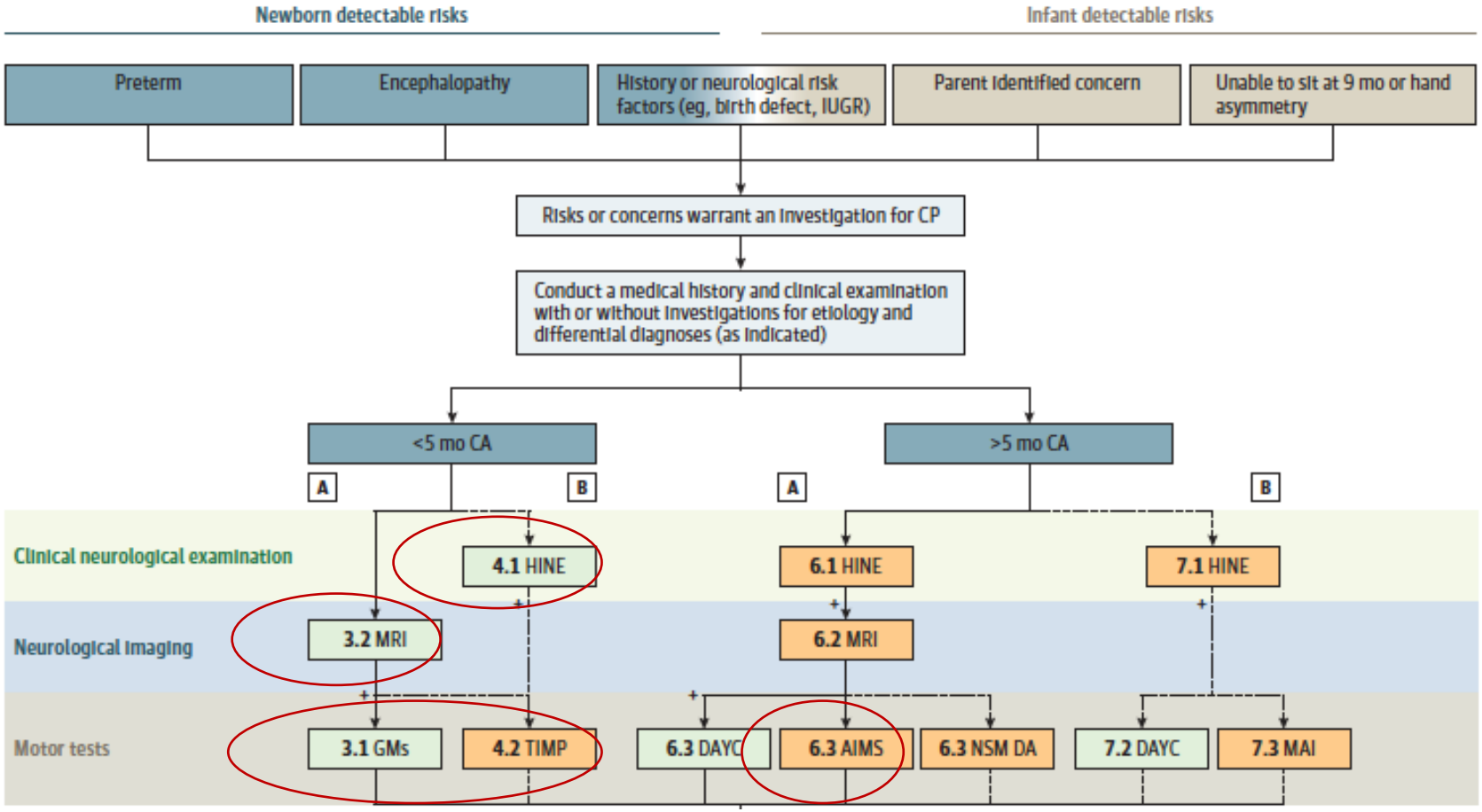
Early Detection and Intervention for CP: Guidelines

- Developed by multidisciplinary team of scientific/clinical experts and parent stakeholders
- Early recognition of CP can and should occur as early as possible so that
  1. Infants can receive diagnostic-specific early intervention and surveillance to optimize neuroplasticity and prevent complications.
  2. Parents can receive psychological and financials support, if available

Novak et al. JAMA Peds 2017

Figure. Algorithm for Early Diagnosis of Cerebral Palsy or High Risk of Cerebral Palsy



Novak et al. JAMA Peds 2017

Prechtl General Movements Assessment (GMA)

- Standardized assessment to identify infants at risk for neuromotor deficits by observing quality of general movements (GMs).
  - Body movements in variable sequence of neck, arm, trunk and leg movements.
  - Similar from early fetal life until ~48 weeks
  - Writing movements -> fidgety movements

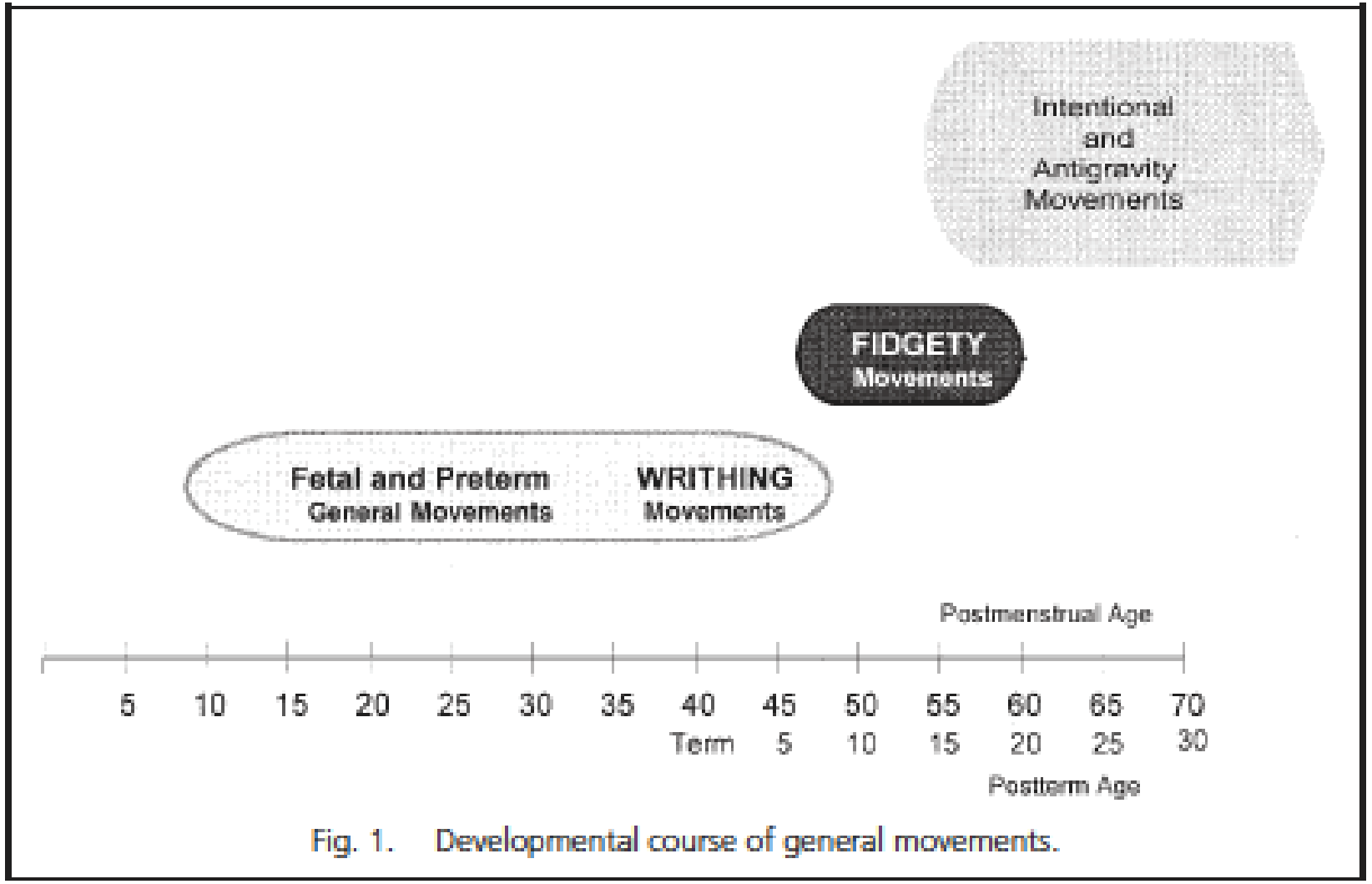
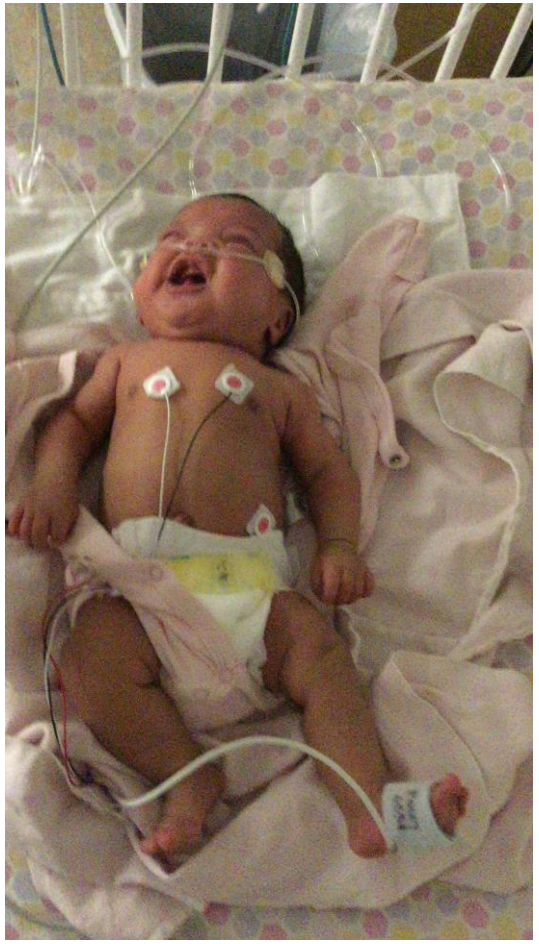


Fig. 1. Developmental course of general movements.

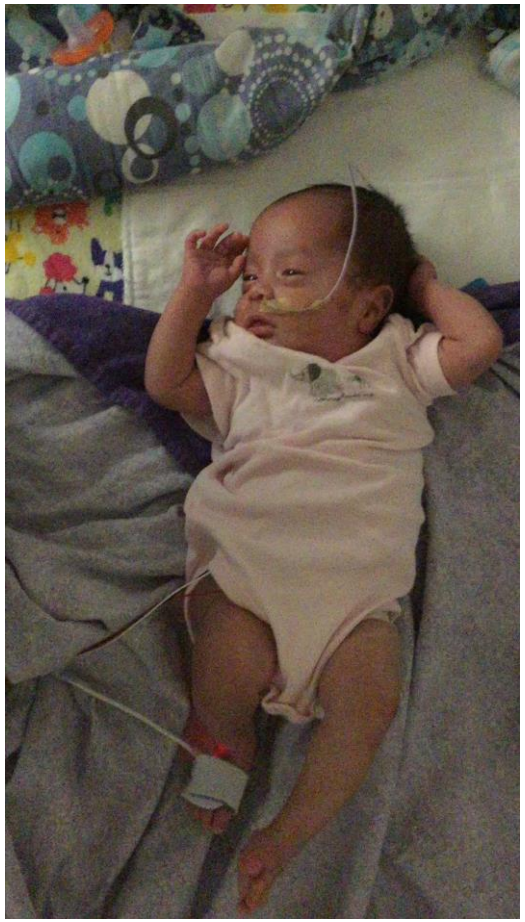
Einspieler et al. MRDD Research Reviews. 2005

IN THE NICU: WRITHING MOVEMENTS

38.6 wks - NW



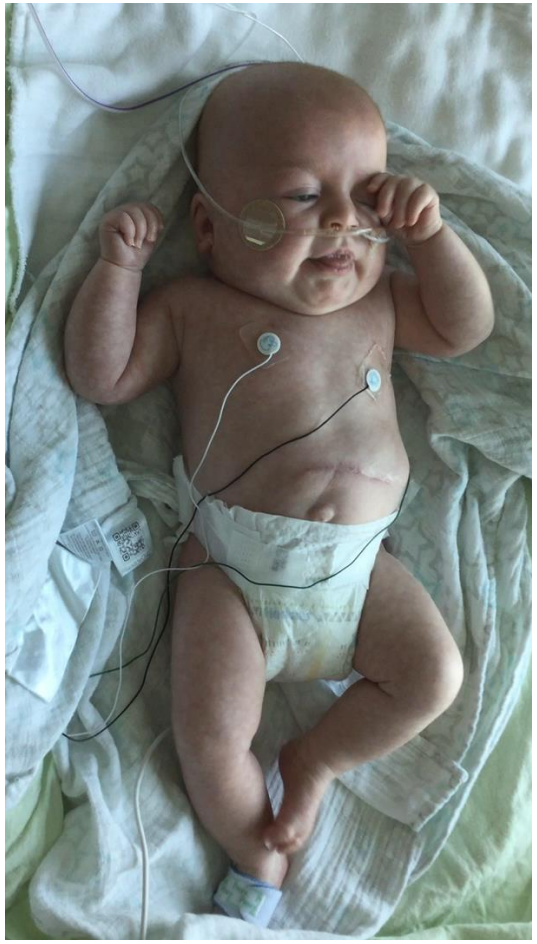
39.6 WKS - CS



IN THE CLINIC: FIDGETY MOVEMENTS

14 wks - Fidgety

13 WKS – ABSENT FIDGETY



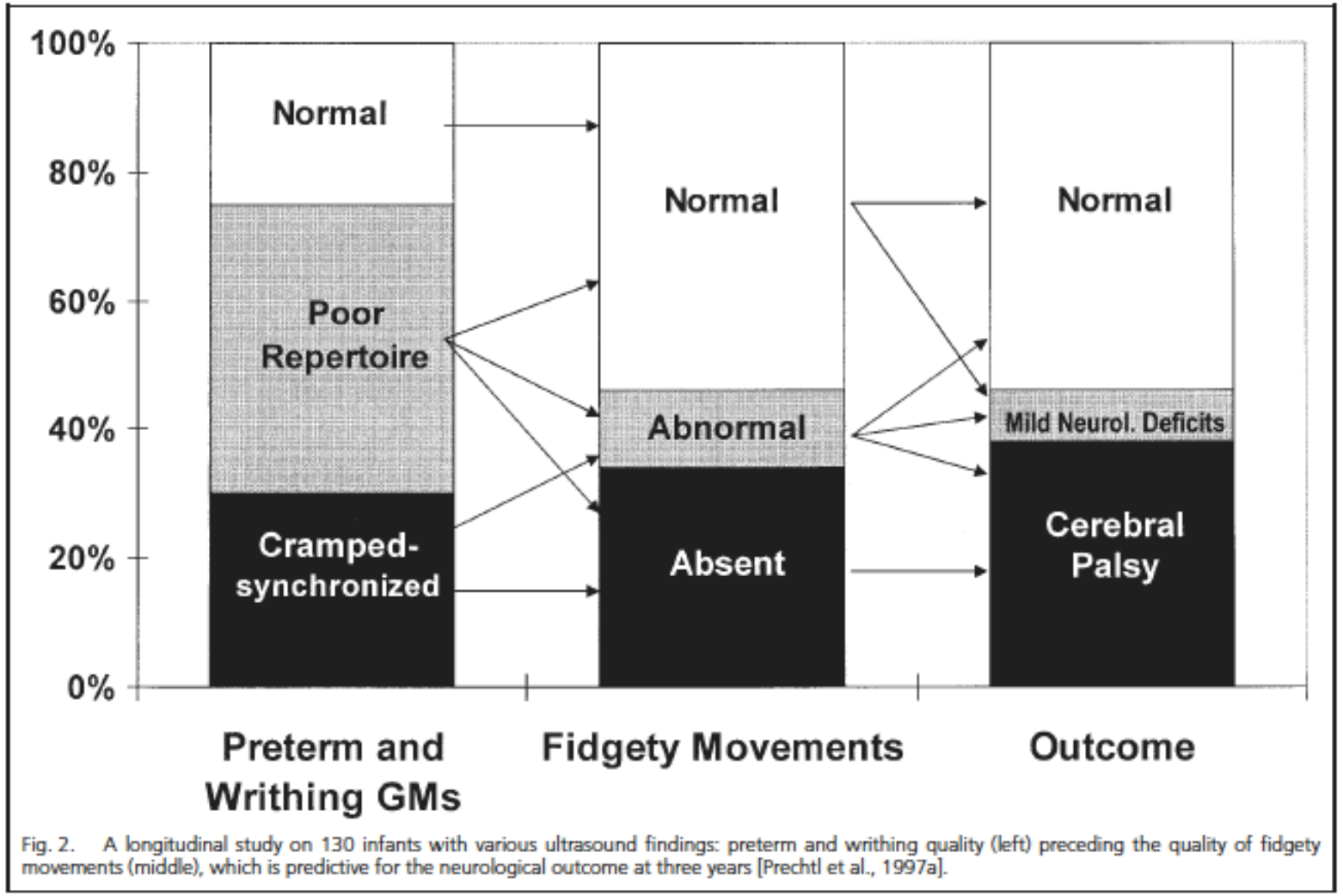


Fig. 2. A longitudinal study on 130 infants with various ultrasound findings: preterm and writhing quality (left) preceding the quality of fidgety movements (middle), which is predictive for the neurological outcome at three years [Prechtl et al., 1997a].

Einspieler et al. MRDD Research Reivews. 2005

SYSTEMATIC REVIEW OF PREDICTIVE VALUE OF GMA

- Fidgety period: sensitivity 97%(93-99) specificity 89% (83-93).
- Writhing period: sensitivity 93% (86-96) specificity 59% (CI 45-71)
- CS had the best specificity: 97% (74-100)

Cramped synchronized movements in term and absent fidgety movements at 3-4 months predicts CP

Kwong 2018

Hammersmith Infant Neurological Examination (HINE)

- Standardized and scoreable clinical neurological examination for infants 2-24 months
- 26 items assessing cranial nerve function, posture, quality/quantity of movements, tone, reflexes and reactions
- Each item scored individually (0, 1, 2 or 3), with a global score adding the scores of all individual items (range: 0-78)

Scoring of the HINE

HAMMERSMITH INFANT NEUROLOGICAL EXAMINATION (v 07.07.17)

Name **Date of birth**

Gestational age **Date of examination**

Chronological age / Corrected age **Head circumference**

SUMMARY OF EXAMINATION
Global score (max 78)
Number of asymmetries
Behavioural score (not part of the optimality score)

Cranial nerve function	score	(max 15)
Posture	score	(max 18)
Movements	score	(max 6)
Tone	score	(max 24)
Reflexes and reactions	score	(max 15)

COMMENTS

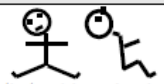
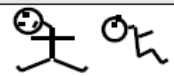
(Throughout the exam, if a response is not optimal but not poor enough to score 1, give a score of 2)

NEUROLOGICAL EXAMINATION

ASSESSMENT OF CRANIAL NERVE FUNCTION

	score 3	2	score 1	score 0	score	Asymmetry / Comments
Facial appearance (at rest and when crying or stimulated)	Smiles or reacts to stimuli by closing eyes and grimacing		Closes eyes but not tightly, poor facial expression	Expressionless, does not react to stimuli		
Eye movements	Normal conjugate eye movements		**Intermittent** Deviation of eyes or abnormal movements	**Continuous** Deviation of eyes or abnormal movements		
Visual response Test ability to follow a black/white target	Follows the target in a complete arc		Follows target in an incomplete or asymmetrical arc	Does not follow the target		
Auditory response Test the response to a rattle	Reacts to stimuli from both sides		Doubtful reaction to stimuli or asymmetry of response	No response		
Sucking/swallowing Watch infant suck on breast or bottle. If older, ask about feeding, assoc. cough, excessive dribbling	Good suck and swallowing		Poor suck and/or swallow	No sucking reflex, no swallowing		

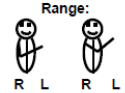
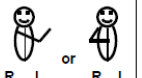
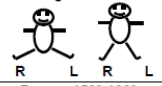
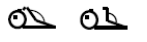
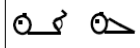
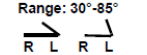
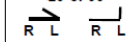
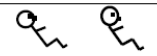
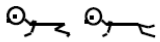
ASSESSMENT OF POSTURE (note any asymmetries)

	score 3	score 2	score 1	score 0	sc	Asymmetry / comments
Head in sitting	 Straight; in midline		 Slightly to side *or* backward *or* forward	 Markedly to side *or* backward *or* forward		
Trunk in sitting	 Straight		 Slightly curved or bent to side	 Very rounded; rocketing back; bent sideway		
Arms at rest	In a neutral position, central straight or slightly bent		**Slight** internal rotation *or* external rotation **Intermittent** dystonic posture	**Marked** internal rotation *or* external rotation or dystonic posture hemiplegic posture		
Hands	Hands open		**Intermittent** adducted thumb *or* fisting	**Persistent** adducted thumb *or* fisting		
Legs in sitting	Able to sit with a straight back and legs straight or slightly bent (long sitting) 		Sit with straight back but knees bent at 15-20 ° 	Unable to sit straight unless knees markedly bent (no long sitting) 		
in supine and in standing	Legs in neutral position straight *or* slightly bent	**Slight** internal rotation or external rotation	Internal rotation *or* external rotation at the hips	**Marked** internal rotation *or* external rotation *or* fixed extension or flexion or contractures at hips and knees		
Feet in supine and in standing	Central in neutral position Toes straight midway between flexion and extension		**Slight** internal rotation *or* external rotation **Intermittent** Tendency to stand on tiptoes or toes up or curling under	**Marked** internal rotation *or* external rotation at the ankle **Persistent** Tendency to stand on tiptoes *or* toes up or curling under		

ASSESSMENT OF MOVEMENTS

	Score 3	Score 2	Score 1	Score 0	score	Asymmetry / comments
Quantity Watch infant lying in supine	Normal		Excessive or sluggish	Minimal or none		
Quality Observe infant's spontaneous voluntary motor activity during the course of the assessment	Free, alternating, and smooth		Jerky Slight tremor	• Cramped & synchronous • Extensor spasms • Athetoid • Ataxic • Very tremulous • Myoclonic spasm • Dystonic movement		

ASSESSMENT OF TONE

	Score 3	Score 2	Score 1	Score 0	sc	Asym/Co
Scarf sign Take the infant's hand and pull the arm across the chest until there is resistance. Note the position of the elbow in relation to the midline.	**Range:** 					
Passive shoulder elevation Lift arm up alongside infant's head. Note resistance at shoulder and elbow.	Resistance overcomeable 	Resistance difficult to overcome 	No resistance 	Resistance, not overcomeable 		
Pronation/supination Steady the upper arm while pronating and supinating forearm, note resistance	Full pronation and supination, no resistance		Resistance to full pronation / supination overcomeable	Full pronation and supination not possible, marked resistance		
Hip adductors With both the infant's legs extended, abduct them as far as possible. The angle formed by the legs is noted.	**Range: 150-80°** 	**150-160°** 	**>170°** 	**<80°** 		
Popliteal angle Keeping the infant's bottom on the bed, flex both hips onto the abdomen, then extend the knees until there is resistance. Note the angle between upper and lower leg.	**Range: 150°-100°** 	**150-160°** 	**~90° or > 170°** 	**<80°** 		
Ankle dorsiflexion With knee extended, dorsiflex the ankle. Note the angle between foot and leg.	**Range: 30°-85°** 	**20-30°** 	**<20°or 90°** 	**> 90°** 		
Pull to sit Pull infant to sit by the wrists. (support head if necessary)						
Ventral suspension Hold infant horizontally around trunk in ventral suspension; note position of back, limbs and head.						

- Key data elements
  - Global Score
  - # of asymmetries

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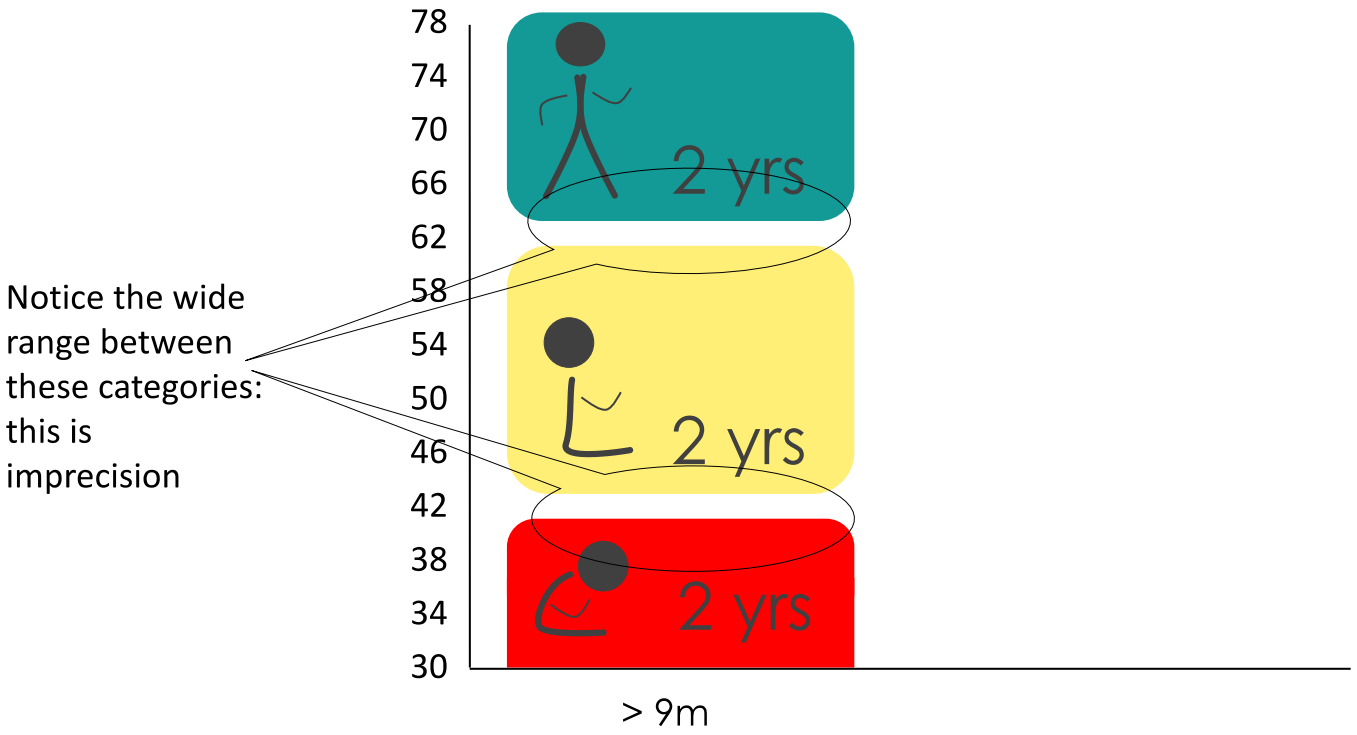
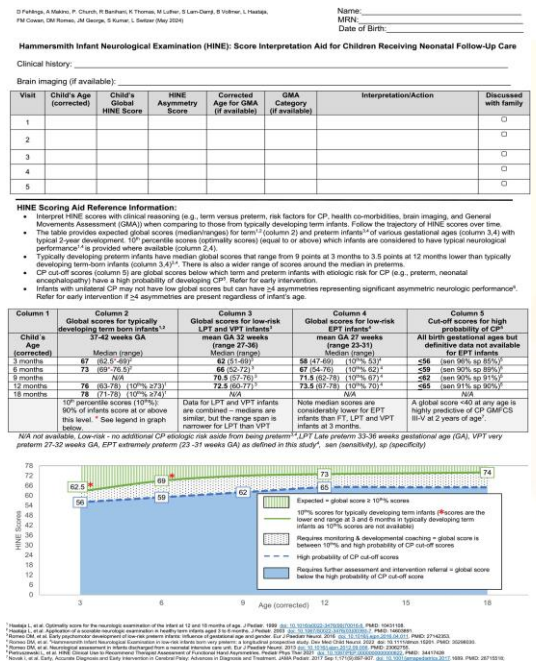
Predicting motor function at 2 yrs in infants with brain insults



HINE Scoring Aid



Name:
MRN:
Date of Birth:

Hammersmith Infant Neurological Examination (HINE): Score Interpretation Aid for Children Receiving Neonatal Follow-Up Care

Clinical history:

Brain imaging (if available):

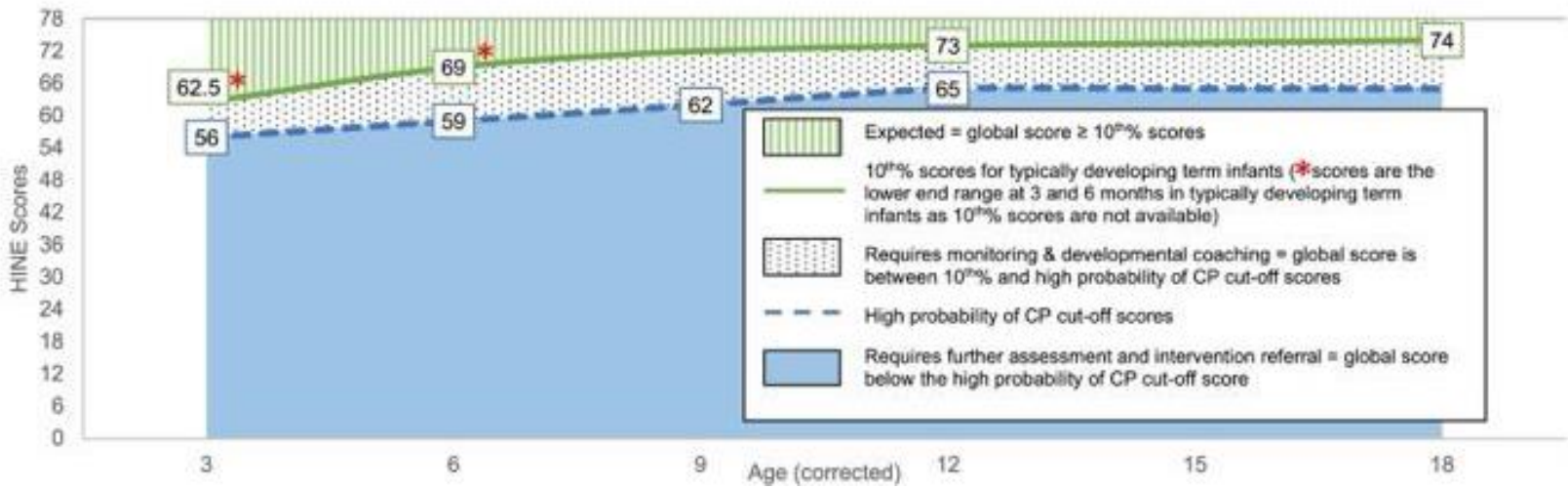
Visit	Child's Age (corrected)	Child's Global HINE Score	HINE Asymmetry Score	Corrected Age for GMA (if available)	GMA Category (if available)	Interpretation/Action	Discussed with family
1							☐
2							☐
3							☐
4							☐
5							☐

HINE Scoring Aid Reference Information:

- Interpret HINE scores with clinical reasoning (e.g., term versus preterm, risk factors for CP, health co-morbidities, brain imaging, and General Movements Assessment (GMA)) when comparing to those from typically developing term infants. Follow the trajectory of HINE scores over time.
- The table provides expected global scores (median/ranges) for term[1,2] (column 2) and preterm infants[3,4] of various gestational ages (column 3,4) with typical 2-year development. 10th percentile scores (optimality scores) (equal to or above) which infants are considered to have typical neurological performance[1,4] is provided where available (column 2,4).
- Typically developing preterm infants have median global scores that range from 9 points at 3 months to 3.5 points at 12 months lower than typically developing term-born infants (column 3,4)[3,4]. There is also a wider range of scores around the median in preterms.
- CP cut-off scores (column 5) are global scores below which term and preterm infants with etiologic risk for CP (e.g., preterm, neonatal encephalopathy) have a high probability of developing CP[5]. Refer for early intervention.
- Infants with unilateral CP may not have low global scores but can have ≥4 asymmetries representing significant asymmetric neurologic performance[6]. Refer for early intervention if ≥4 asymmetries are present regardless of infant's age.

Column 1	Column 2 Global scores for typically developing term born infants[1,2]	Column 3 Global scores for low-risk LPT and VPT infants[3]	Column 4 Global scores for low-risk EPT infants[4]	Column 5 Cut-off scores for high probability of CP[5]
Child's Age (corrected)	37-42 weeks GA Median (range)	mean GA 32 weeks (range 27-36) Median (range)	mean GA 27 weeks (range 23-31) Median (range)	All birth gestational ages but definitive data not available for EPT infants
3 months	67 (62.5*-69)[2]	62 (51-69)[3]	58 (47-69) (10th% 53)[4]	≤56 (sen 96% sp 85%)[5]
6 months	73 (69*-76.5)[2]	66 (52-72)[3]	67 (54-76) (10th% 62)[4]	≤59 (sen 90% sp 89%)[5]
9 months	N/A	70.5 (57-76)[3]	71.5 (62-78) (10th% 67)[4]	≤62 (sen 90% sp 91%)[5]
12 months	76 (63-78) (10th% ≥73)[1]	72.5 (60-77)[3]	73.5 (67-78) (10th% 70)[4]	≤65 (sen 91% sp 90%)[5]
18 months	78 (71-78) (10th% ≥74)[1]	N/A	N/A	N/A
	10th percentile scores (10th%): 90% of infants score at or above this level. * See legend in graph below.	Data for LPT and VPT infants are combined – medians are similar, but the range span is narrower for LPT than VPT	Note median scores are considerably lower for EPT infants than FT, LPT and VPT infants at 3 months.	A global score <40 at any age is highly predictive of CP GMFCS III-V at 2 years of age[7].

N/A not available, Low-risk - no additional CP etiologic risk aside from being preterm[3,4],LPT Late preterm 33-36 weeks gestational age (GA), VPT very preterm 27-32 weeks GA, EPT extremely preterm (23 -31 weeks GA) as defined in this study[4], sen (sensitivity), sp (specificity)

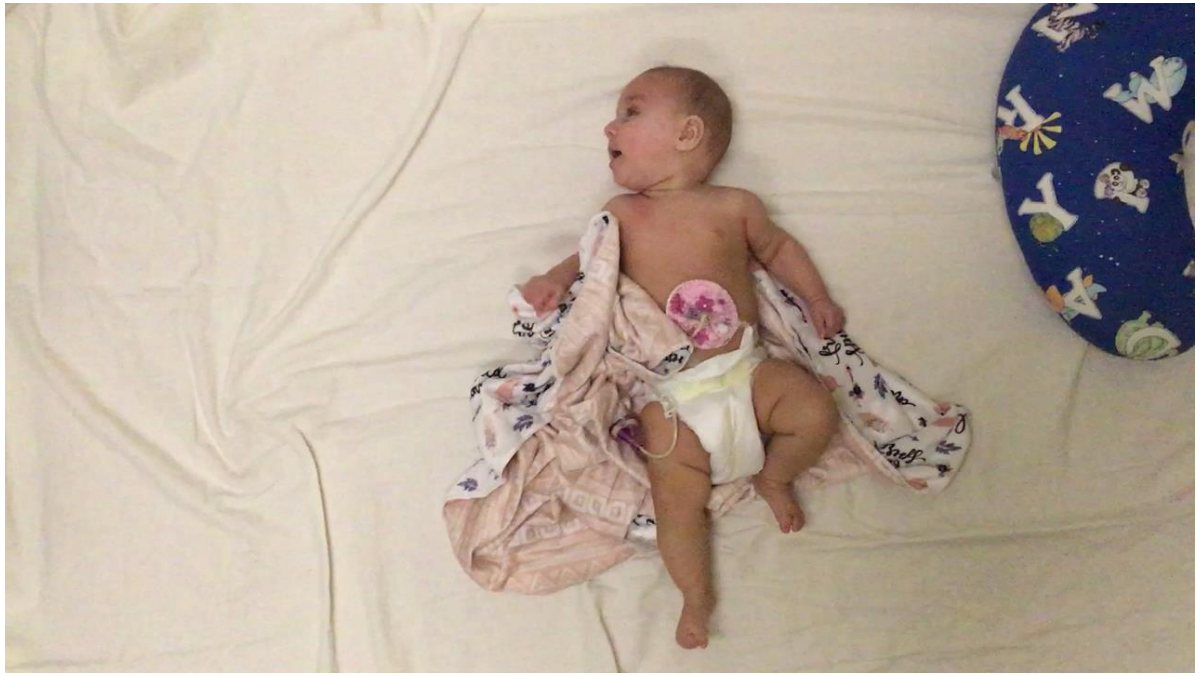


Fehlings et al. Develop Med Child Neuro 2024

Motor Assessment Tools

Test	Age Range	Description	Sens/Spec	Time frame/cost
Test of Infant Motor Performance (2002)	Preterm (>34 weeks) to 4 months adj age	Early identification of infants with neuromotor dysfunction	Sens 83-96% Spec 78 – 65%	30 minutes
Alberta Infant Motor Scales (1994)	0 – 18 months	Useful in high risk infants for screening gross motor function	Sens 74 – 86% Spec 81 – 93%	20 minutes
Peabody Developmental Motor Scale 2nd ed. (2000)	Birth – 6 years	Measures of gross and fine motor skills, has subtests, gives age equivalent	At 4 mo: sens 25%, spec 73% At 8 mo: sens 86% spec 46 – 60%	20 -30 minutes for each subtest
Bayley Scales of Infant Development: (PDI)	1 – 42 months	Frequently used by NICU follow-up programs and outcome studies		15-20 minutes

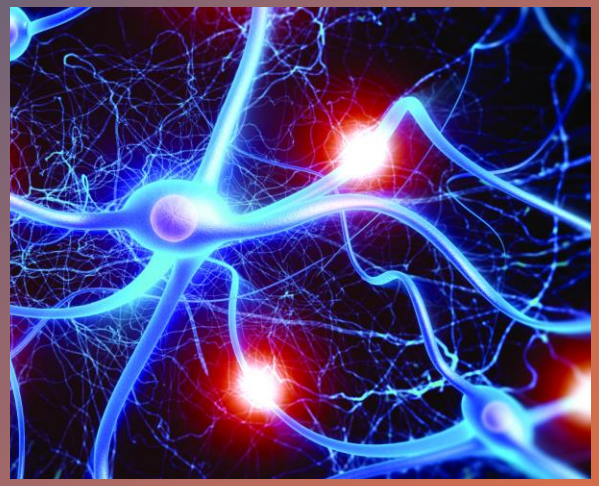
Case



- Born at 26w4d
- ELBW, CLD
- Left grade III IVH with periventricular hemorrhagic infarction, cerebellar hemorrhage

Case

- 16 weeks AGA
  - GMA: Absent Fidgety
  - HINE: 37.5, No asymmetries
  - TIMP: <5%ile, Age equivalency 2-3 weeks
  - Dx: High risk for CP, recommended MRI
  - Brain MRI consistent with injury
- Therapy:
  - 1x/week private feeding therapy with OT
  - 1x/week EI with OT. Visits difficult to schedule and family report that EI provider had told them she did not qualify for PT.



- At 6mos AGA
  - HINE : 36
  - Dx: Quadriplegic CP,
- Therapy Recommendations:
  - Vision therapy through EI
  - Physical therapy through EI or privately
  - Home exercises:
    - Head control
    - Demonstrated exercises in prone
    - Demonstrated assisted rolling supine to prone.
    - Demonstrated and discussed positioning during feeding

References

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