early diagnosis of cerebral palsy

physical medicine and rehabilitation

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objectives

- Recognize how to diagnose cerebral palsy
- Identify the assessment tools used to aid in early diagnosis
- Describe early treatment options for cerebral palsy

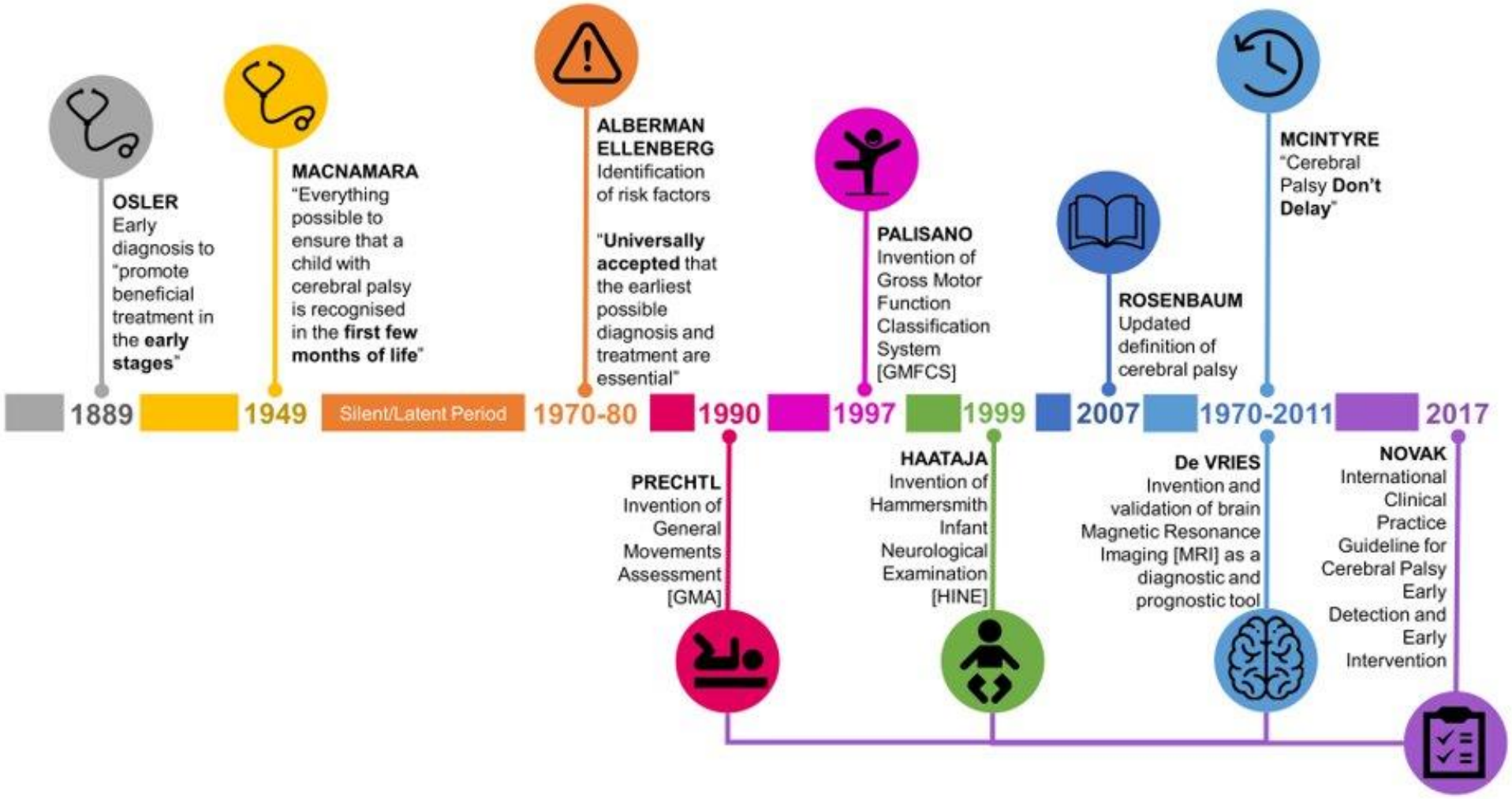
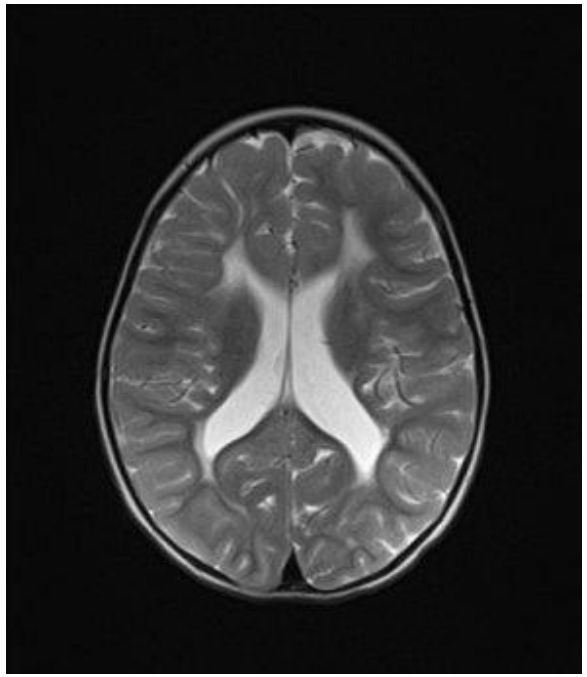


Figure from te Velde, et al (2019)

definition and diagnosis

definition of cerebral palsy

- "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"
  - 1) Motor disorder
  - 2) Non-progressive
  - 3) Due to injury/change to the developing brain



https://radiopaedia.org/cases/periventricular-leukomalacia

diagnosis

- Diagnosis is made clinically
  - Will use various tools and elements of history and physical to see if the child fits with the diagnosis
    - History: gestational age, complications with pregnancy or delivery, hypoxic event, etc
    - Physical: Evidence of motor disorder: Spasticity, ataxia, dyskinesia
    - Imaging: History of IVH, MRI with evidence of prior injury
    - Assessment tools: HINE/HNNE, GMA

risk factors for cerebral palsy

- #1 risk factor is prematurity
- Pregnancy risk factors: infection, multiple gestation, substance abuse, pre-eclampsia, IUGR, Rh incompatibility
- Peri-birth factors: Low birth weight, jaundice, difficult delivery, breech presentation, hypoxic event, hypoglycemia
- Neonatal and infant factors: stroke, TBI, infection, surgical complication

Importance of early diagnosis

- Diagnosis has typically been made between 12 and 24 months in developed countries
  - Can be closer to 5 years of age in developing countries
- Goal to get age of diagnosis under 12 months
  - Get children earlier access to therapies and interventions
  - Psychological support for parents

barriers to early diagnosis

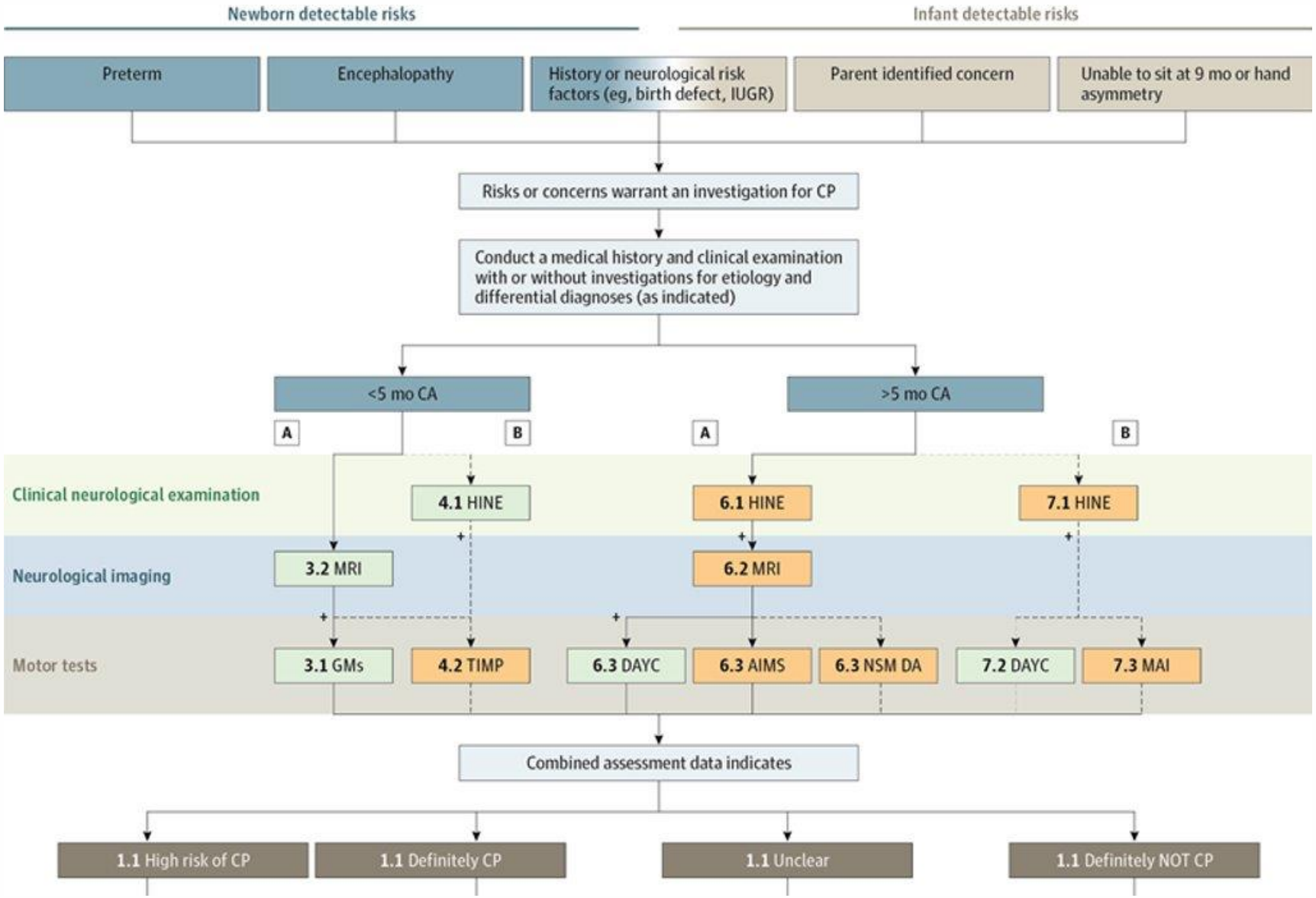
- No definitive test
- Time needed to rule out alternate diagnoses
- Hesitancy to give a diagnosis that may be incorrect
- Difficulty to assess movement in very young infants
    - Children with milder CP may reach most or all milestones on time

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; Michael Fahey, PhD; Darcy Fehlings, PhD; Donna M. Ferriero, MD; Linda Fetters, PhD; Simona Fiori, PhD; Hans Forssberg, PhD; Andrew M. Gordon, PhD; Susan Greaves, PhD; Andrea Guzzetta, PhD; Mijna Hadders-Algra, PhD; Regina Harbourne, PhD; Angelina Kakooza-Mwesige, PhD; Petra Karlsson, PhD; Lena Krumlinde-Sundholm, PhD; Beatrice Latal, MD; Alison Loughran-Fowlds, PhD; Nathalie Maitre, PhD; Sarah McIntyre, PhD; Garey Noritz, MD; Lindsay Pennington, PhD; Domenico M. Romeo, PhD; Roberta Shepherd, PhD; Alicia J. Spittle, PhD; Marelle Thornton, DipEd; Jane Valentine, MRCP; Karen Walker, PhD; Robert White, MBA; Nadia Badawi, PhD


Newborn detectable risks
Infant detectable risks
Preterm
Encephalopathy
History or neurological risk factors (eg, birth defect, IUGR)
Parent identified concern
Unable to sit at 9 mo or hand asymmetry
Risks or concerns warrant an investigation for CP
Conduct a medical history and clinical examination with or without investigations for etiology and differential diagnoses (as indicated)
<5 mo CA
>5 mo CA
A
B
A
B
Clinical neurological examination
4.1 HINE
6.1 HINE
7.1 HINE
Neurological imaging
3.2 MRI
6.2 MRI
Motor tests
3.1 GMs
4.2 TIMP
6.3 DAYC
6.3 AIMS
6.3 NSM DA
7.2 DAYC
7.3 MAI
Combined assessment data indicates
1.1 High risk of CP
1.1 Definitely CP
1.1 Unclear
1.1 Definitely NOT CP

recommendations from the "early, accurate diagnosis" review article

- <5 months – MRI, General Movements Assessment or Hammersmith Infant Neurologic Exam is most predictive of CP
- >5 months – MRI, Hammersmith Infant Neurologic Exam, standardized motor assessment (DAY-C, AIMS) is most predictive of CP

general movements assessment

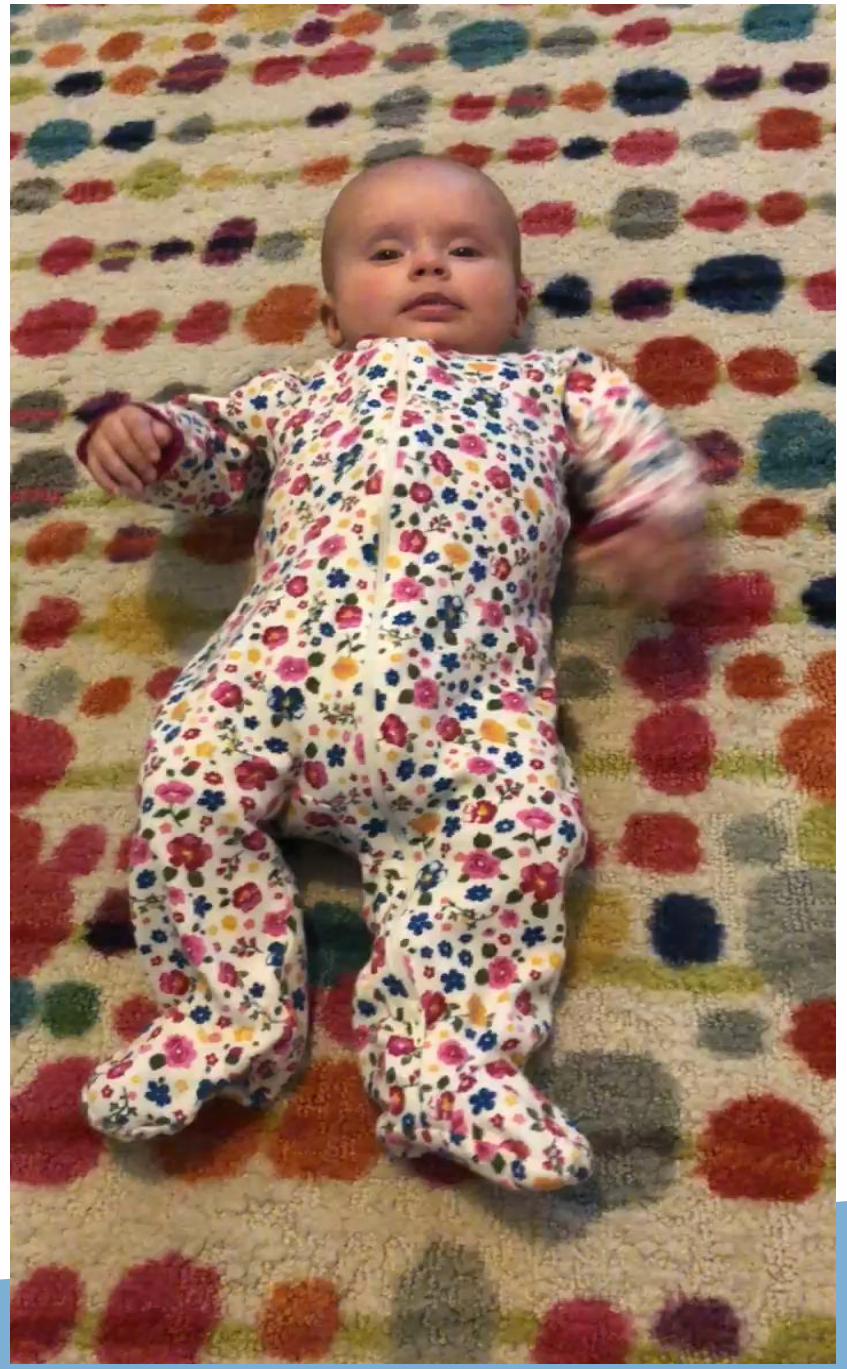
- Looks at the patterns of the general, spontaneous movements that all babies are expected to make
- Can be completed from birth up to 20 weeks of age
  - Writhing phase (up to 6-9 weeks)
  - Fidgety phase (up to 20 weeks)
- Baby is videotaped, lightly dressed, while laying down and without any external stimuli

GMA evidence

- 2019 study by Støen et al looked at 405 NICU patients with risk factors for CP and performed the GMA at age 10-15 weeks post term age and followed them clinically to assess for diagnosis of CP (at least 18-24 months)
  - 23.7% had an abnormal GMA; 21.5% had abnormal head imaging
- 10.4% received a diagnosis of CP
  - For those with absent fidgety movements - 48.3% were diagnosed with CP
  - Highest accuracy of CP prediction combined presence of absent fidgety movements and abnormal imaging (95.3% accurate, with 61.9% sensitivity and 99.2% specificity)

general movements assessment examples

general movements assessment examples



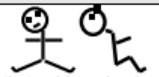
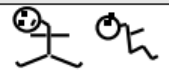
dayton children's

- Y

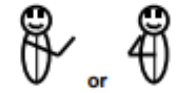
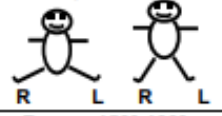
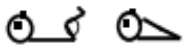
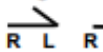
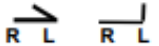
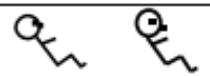
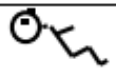
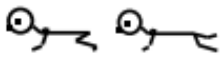
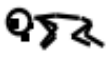
hammersmith infant neurologic exam (HINE)

- Can be used on infants 2-24 months of age.
- Includes a scored neurologic exam + evaluation of developmental and behavioral milestones
  - Neurologic exam includes 26 items divided into 5 domains
  - Lower score = abnormal

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 in supine and in standing	Able to sit with a straight back and legs straight or slightly bent (long sitting)  Legs in neutral position straight *or* slightly bent	**Slight** internal rotation or external rotation	Sit with straight back but knees bent at 15-20 °  Internal rotation *or* external rotation at the hips	Unable to sit straight unless knees markedly bent (no long sitting)  **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 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:**  R L R L		 R L	 or R L R L		
Passive shoulder elevation Lift arm up alongside infant's head. Note resistance at shoulder and elbow.	Resistance overcomeable  R L	Resistance difficult to overcome R L	No resistance  R L	Resistance, not overcomeable  R L		
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°**  R L R L	**150-160°**  R L	**>170°**  R L	**<80°**  R L		
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°**  R L R L	**150-160°**  R L	**~90° or > 170°**  R L R L	**<80°**  R L		
Ankle dorsiflexion With knee extended, dorsiflex the ankle. Note the angle between foot and leg.	**Range: 30°-85°**  R L R L	**20-30°**  R L	**<20°or 90°**  R L R L	**> 90°**  R L		
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.						

HINE evidence

- Romeo et al (2013) evaluated 1,541 Italian children discharged from a level II-III NICU until age 2, and obtained HINE scores at 3, 6, 9, and 12 months corrected gestational age
- Found scores <56 at 3 months (sen 96%, sp 85%) and < 65 (sen 91%, sp 90%) at 12 months had high probability of developing CP

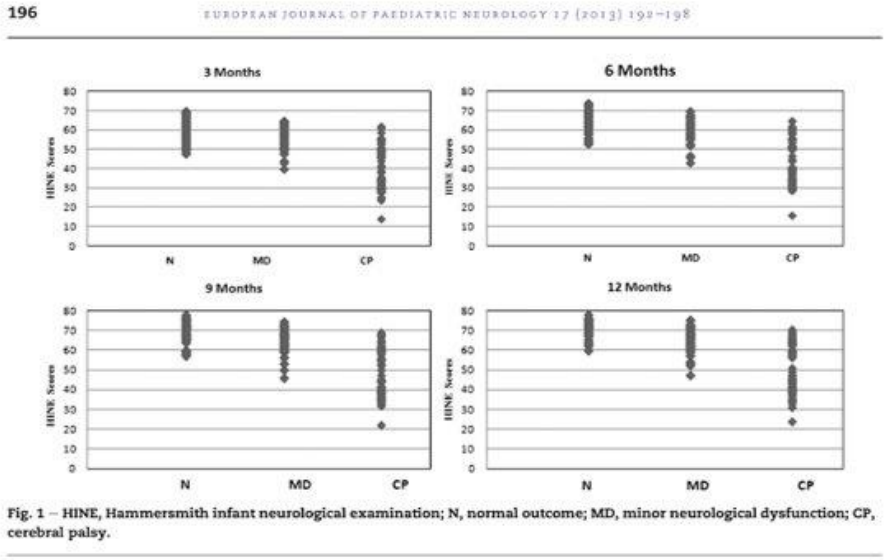


Fig. 1 – HINE, Hammersmith infant neurological examination; N, normal outcome; MD, minor neurological dysfunction; CP, cerebral palsy.

Interim diagnosis

- It is recommended to use an interim diagnosis in children who do not meet the criteria for a CP diagnosis at time of assessment but there is a high index of suspicion
- “High risk of cerebral palsy”
- Can still refer to cerebral palsy specific therapies and interventions

Implementation of guidelines

- NICU follow up clinics at 5 different US institutions implemented testing into their clinics based on the early detection algorithm
  - For high-risk infants with abnormal GMA in the NICU, would have 3-4 month clinic visit that included the HINE, GMA, and a motor assessment
  - Those deemed high risk for CP would follow up again in 3 months for repeat HINE, imaging review, and diagnosis if clinically appropriate
- Age of CP diagnosis decreased from 19.5 months to 9.5 months

when to refer?

- Delays or other concerns present in high risk infants
- Abnormal neurologic exam findings
  - Low tone, high tone
  - Early hand preference or other asymmetries
  - Inability to sit by 9 months
  - Inability to bear weight through the plantar surface of the feet

early intervention

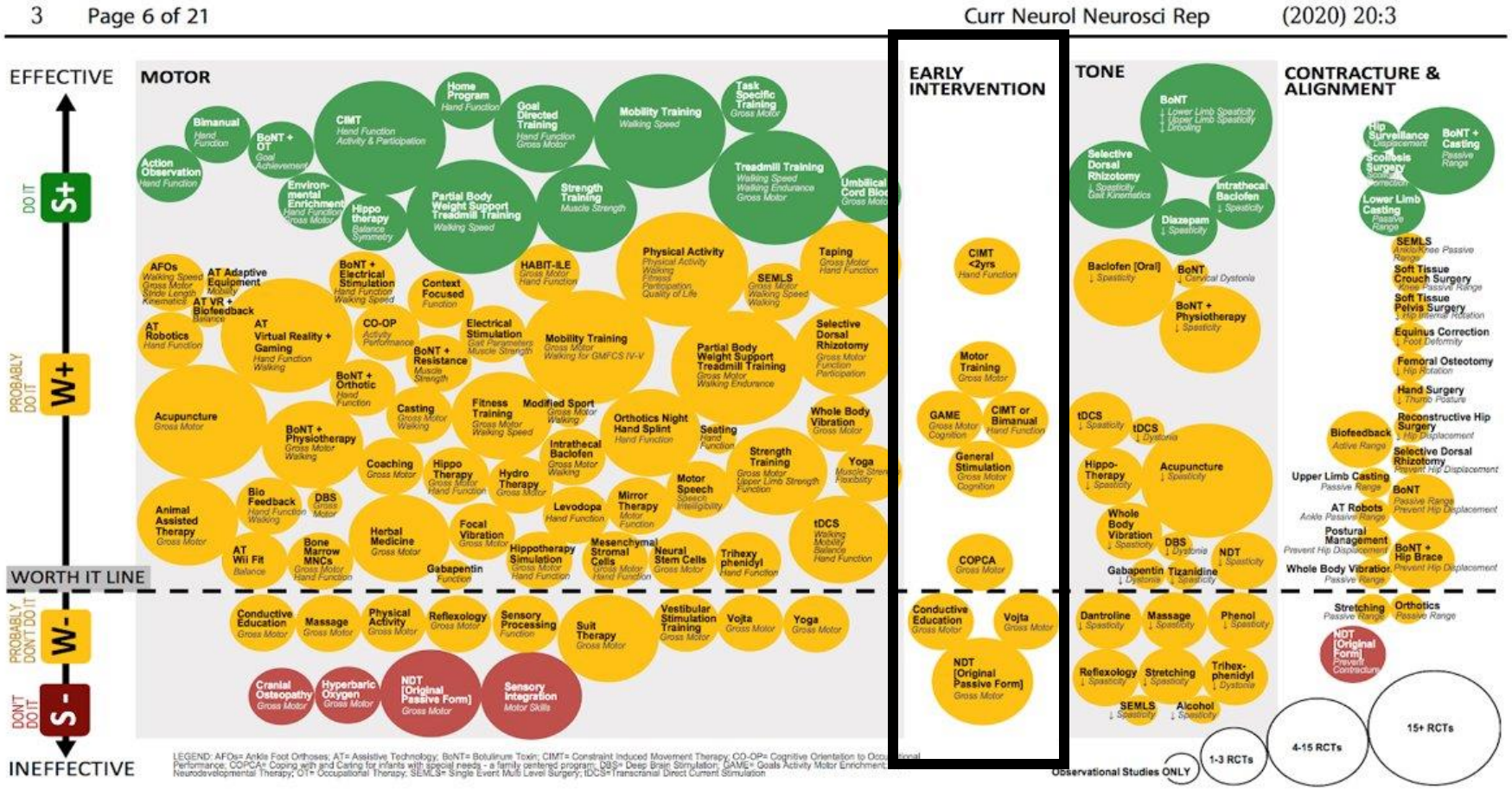


Figure from Novak, et al (2020)

dayton children's

general management

- Therapies (PT, OT, speech, and feeding therapy)
- Control spasticity and agitation
  - Medications
  - Rage of motion and stretching program
  - Splinting or bracing

Photo credit: OSF Healthcare

baby-CIMT

- Limited evidence looking at Constraint Induced Movement Therapy (CIMT) in infants and young children
  - There are some clinical trials in progress
- One smaller study compared 8 infants with CIMT intervention with 13 infants with baby massage; all infants were between age 3 to 8 months with hand asymmetry
  - CIMT group had improved use of the affected hand

GAME (Goals, Activity, Motor Enrichment)

- Morgan et al in 2016 performed a single blind randomized control trial of GAME (Goals - Activity - Motor Enrichment) Intervention vs standard care to 30 high risk infants from ~4-5 months until 12 months of age
  - Measured outcomes at 16 weeks post enrollment and 12 months of age
  - Found improved motor outcomes (Peabody) for GAME group at 12 months

Small Step Program

- Intervention delivered in the home by parents focusing on mobility, hand use, and communication
- Infants aged 4-9 months, 19 in Small Step Program, 20 in standard care
  - 35 week intervention
- Main outcome Peabody Developmental Motor Scales
  - No group difference at end of treatment
  - Did note that baseline prior to intervention mattered less for those in Small Step program
- 10 from each group ultimately given a formal diagnosis of CP

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dayton children's

Questions?