Evidence-based Interventions for Children with Cerebral Palsy

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1. Describe evidence-based interventions for children with cerebral palsy (CP)
2. Determine how to prioritize impairments to be addressed when working with children with CP
3. Recognize recommendations for standing programs and orthopedic surgery considerations for children with CP
Medical intervention team for children with CP
http://www.cerebralpalsy.org/information/care-plan/care-team-for-managing-cp

- Obstetrics Gynecologists
- Occupational Therapist
- Ophthalmologists
- Orthopedist/Orthopedic Surgeons
- Orthotist
- Otologists
- Pediatrician
- Physical Therapists
- Psychologist and/or neuropsychologist
- Rehabilitation Medicine Specialist (Physiatrist)
- Social worker
- Speech and Language Pathologists
- Urologists

- Parents/child
- Audiologist
- Behavioral Optometrist
- Behavioral Therapist
- Counselors and Social Workers
- Dentists/Orthodontists
- Developmental Behavioral Specialists
- Developmental Pediatrician
- ENT/Otorhinolaryngologist
- Geneticists
- Neonatologists
- Neurologists
- Neuroradiologists
- Nutritionists

It takes a village!
The International Classification of Function, Disability and Health (ICF).

Jette A M PHYS THER 2006;86:726-734
Interventions for children with CP:
Where do I even begin?!

- WHEN DEVELOPING YOUR PLAN OF CARE, ALWAYS CONSIDER PARTICIPATION LEVEL FUNCTION and
- FAMILY CENTERED GOALS FOR CHILD.

“What dog is barking the loudest?”
to prioritize impairments and activity limitations to chose your interventions
- For example, is spasticity a primary impairment that needs to be addressed?
Outcomes of Treatment for Cerebral Palsy

• Optimize independent mobility as early as possible!!!
• Manage primary impairments
• Control pain
• Prevent and manage complications, associative conditions and co-mitigating factors
• Maximize independence
• Enhance social and peer interactions
• Foster self-care
• Optimize ability to communicate
• Maximize learning potential
• Provide quality of life

http://www.cerebralpalsy.org/about-cerebral-palsy/treatment
State of the Evidence Traffic Lights 2019: Systematic Review of Interventions for Preventing and Treating Children with Cerebral Palsy

Iona Novak¹ · Catherine Morgan¹ · Michael Fahey²,³ · Megan Finch-Edmondson¹ · Claire Galea⁴,⁵ · Ashleigh Hines¹ · Katherine Langdon⁵ · Maria McNamara³ · Madison CB Paton¹ · Himanshu Popat¹,⁴ · Benjamin Shore⁶ · Amanda Kamas³ · Emma Stanton¹ · Olivia P Finemore¹ · Alice Tricks¹ · Anna te Velde¹ · Leigha Dark⁷ · Natalie Morton⁶,⁹ · Nadia Badawi¹,⁴

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Abstract
Purpose of Review Cerebral palsy is the most common physical disability of childhood, but the rate is falling, and severity is lessening. We conducted a systematic overview of best available evidence (2012–2019), appraising evidence using GRADE and the Evidence Alert Traffic Light System and then aggregated the new findings with our previous 2013 findings. This article summarizes the best available evidence interventions for preventing and managing cerebral palsy in 2019.

Recent Findings Effective prevention strategies include antenatal corticosteroids, magnesium sulfate, caffeine, and neonatal hypothermia. Effective allied health interventions include acceptance and commitment therapy, action observations, bimanual training, casting, constraint-induced movement therapy, environmental enrichment, fitness training, goal-directed training, hippotherapy, home programs, literacy interventions, mobility training, oral sensorimotor, oral sensorimotor plus electrical stimulation, pressure care, stepping stones triple P, strength training, task-specific training, treadmill training, partial body weight support treadmill training, and weight-bearing. Effective medical and surgical interventions include anti-convulsants, bisphosphonates, botulinum toxin, botulinum toxin plus occupational therapy, botulinum toxin plus casting, diazepam, dentistry, hip surveillance, intrathecal baclofen, scoliosis correction, selective dorsal rhizotomy, and umbilical cord blood cell therapy.

Summary We have provided guidance about what works and what does not to inform decision-making, and highlighted areas for more research.

Keywords Cerebral palsy · Systematic review · Traffic light system · Evidence based · GRADE
Clinical bottom line of Novak, et al SR

- CP is heterogenous group, so case-by-case consideration needed
- Hopefully we will move to using classifications (like GMFCS) when we assess outcomes of interventions for CP
- Oversimplification of traffic light model could be dangerous
- Best design for outcomes in CP are prospective cohort designs, not RCTs
- Despite controversy, this is the best article to review all the RCTs that has been published on intervention for CP
Tone Interventions

Novak, 2020
Oral medications modify the inhibitory effect on the final common pathway

- **Systemic treatment**
  - Baclofen (Lioresal)
  - Diazepam (Valium)

- **Focal treatment**
  - Botulinum toxin (Botox, Myobloc)
  - Dantrolene (Dantrium)

- **Alternative treatment**
  - CBD

Modified from Schilz, Drugs for Spasticity lecture UNM DPT, 2020
https://www.practicalpainmanagement.com/treatments/pharmacological/non-opioids/review-skeletal-muscle-relaxants-pain-management
Cannabis and spasticity
patient reports; not well studied yet

The Endocannabinoid System

Brain cells (neurons) communicate with each other by sending chemical messages. The chemicals (neurotransmitters) cross a gap between neighboring neurons before attaching to their specific receptors.

**Presynaptic**: The neuron sending a message by releasing a chemical when signaled to do so

**Postsynaptic**: The neuron receiving the message when its receptors are activated by specific chemicals (neurotransmitters)

**Neurotransmitters**: The chemical messengers that travel from one brain cell to another

**Receptors**: Activated by neurotransmitters, receptors trigger a set of events that allows a message to be passed along to other neurons

**Cannabinoids**: Natural chemicals (anandamide and 2-AG) that bind to cannabinoid receptors in the brain and the body

**THC**: The main active ingredient in marijuana; THC, also a cannabinoid, interferes with the normal functioning of the endocannabinoid system

How does THC affect behavior? It depends on where the CB receptors are in the brain.

<table>
<thead>
<tr>
<th>Brain Structure</th>
<th>Regulates</th>
<th>THC Effect on User</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amygdala</td>
<td>emotions, fear, anxiety</td>
<td>panic/paranoia</td>
</tr>
<tr>
<td>Basal Ganglia</td>
<td>planning/starting a movement</td>
<td>slowed reaction time</td>
</tr>
<tr>
<td>Brain Stem</td>
<td>information between brain and spinal column</td>
<td>anti-nausea effects</td>
</tr>
<tr>
<td>Cerebellum</td>
<td>motor coordination, balance</td>
<td>impaired coordination</td>
</tr>
<tr>
<td>Hippocampus</td>
<td>learning new information</td>
<td>impaired memory</td>
</tr>
<tr>
<td>Hypothalamus</td>
<td>eating, sexual behavior</td>
<td>increased appetite</td>
</tr>
<tr>
<td>Neocortex</td>
<td>complex thinking, feeling, and movement</td>
<td>altered thinking, judgment, and sensation</td>
</tr>
<tr>
<td>Nucleus Accumbens</td>
<td>motivation and reward</td>
<td>euphoria (feeling good)</td>
</tr>
<tr>
<td>Spinal Cord</td>
<td>transmission of information between body and brain</td>
<td>altered pain sensitivity</td>
</tr>
</tbody>
</table>

The brain structures illustrated above all contain high numbers of CB receptors

http://headsup.scholastic.com/students/the-science-of-marijuana

Modified from Schilz, Drugs for Spasticity lecture UNM DPT, 2020
Botulinum Toxin (BoNT-A) in the Management of Children with CP


• BoNT-A is effective to reduce spasticity for 3-6 months in children with CP
• This can result in ROM increases
• Muscle atrophy can occur after 12 months of injections
**Mechanism of action and side effects**

**Systemic drugs**

**Baclofen** (Lioresal)

**Diazepam** (Valium)

**Mechanism of action:**
- Decrease excitability of neurons
  - Activating presynaptic inhibition of excitatory NT release
  - Decrease glutamate release
- Activating GABA-mediated inhibition
  - Hyperpolarization (increase Cl- ions) decreasing excitability

**Adverse effects**
- Sedation
- Hypotonia (mostly baclofen)
- Confusion

**Focal drugs**

**Mechanism of action:**
- **Botox** – cleaves docking proteins preventing release of Acetylcholine at the neuromuscular junction

**Adverse effects**
- Localized myalgia (if doesn’t travel from injection site)

**Mechanism of action:**
- **Dantrolene (oral)** – blocks opening of voltage-gated Ca2+ channels on SR decreasing Ca2+ release \(\rightarrow\) decreases muscle contraction.

**Adverse effects**
- Flushing, drowsiness, dysphagia, nausea

*Modified from Schilz, Drugs for Spasticity lecture UNM DPT, 2020*
Intrathecal baclofen pump

+ Low dose baclofen into spinal canal, so avoid most side effects

- Risk of infection; needs to be weaned off baclofen slowly or respiratory failure can occur
Selective Dorsal Rhizotomy (SDR)

- The neurosurgeon performs a laminectomy over the cauda equina (L1 to L3-S1, depending on the institution).
- The surgeon divides each of the dorsal roots into 3-5 rootlets and stimulates each rootlet electrically.
- By examining EMG responses from muscles in the lower extremities, the surgical team identifies the rootlets that cause spasticity.
- The abnormal rootlets are selectively cut, leaving the normal rootlets intact.
- This reduces messages from the muscle, resulting in a better balance of activities of nerve cells in the spinal cord.
SDR surgical candidate?

- Usually GMFCS levels II-IV
- Good cognitive function
- Spastic diplegia has best result
- Pure spasticity (no ataxia or dystonia)
- Good underlying strength
- Ideal candidate is child 4-7 years old, but can be older
- Fixed deformities may need to be corrected first or later
- Supportive family
- Ability to follow through with intensive therapy and HEP

SDR pre and post surgery video
• Pros
  – Decreases or eliminates spasticity
  – Can reduce risk of secondary impairments
  – Can reduce need for some orthopedic surgeries
  – Can improve function

• Cons:
  – Weakness
  – Unmasks low underlying tone, dystonia, athetosis
  – Decreased proprioception and body awareness
  – Does not take away poor selective motor control and habitual patterns already developed
  – Requires extensive rehab
Contracture & Alignment Interventions

Novak, 2020
Serial Casting

• Provides prolonged, gentle stretch: progressively increases muscle length and passive range of motion
• Each cast usually lasts 5-6 days, 1-2 days out of cast, followed by recasting at the new muscle length
• Usually for at least 3-6 weeks total with maintenance of 2-3 weeks during growth spurts
• Can have short term weakness (2 weeks on average) after casts, so strengthening is important
• Often combined with Botox in patients with UE spasticity; Gastrocnemius muscles respond best to casting without Botox due to weakness
• If R1<0 degrees DF because this is what the child is probably actively using
• The greater the difference between R1 and R2, the better the results
LE orthoses/bracing

Solid Ankle Foot Orthosis

Articulated Ankle Foot Orthosis

Floor-Reaction Ankle Foot Orthosis

For gait training, standing balance, and positioning; not for contracture management
Low load, long duration stretching

**Positioning**

- Standing on incline board
- Long sit with hip abd
- Ring sit
- Prone
- Straddle sit
- Stander with hip abduction
- Kiddy up

**Night splinting (if tolerated)**

- Ankle DF
- Hamstrings
- Hip adductors
- Biceps
- Etc.
Zero tolerance for contractures and asymmetry!

### Lower Limb

<table>
<thead>
<tr>
<th>GMFCS I-III</th>
<th>Red</th>
<th>Yellow</th>
<th>Green</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hip Abduction</td>
<td>≤30°</td>
<td>&gt;30°</td>
<td>&lt;40°</td>
</tr>
<tr>
<td>Knee Popliteal angle</td>
<td>≤130°</td>
<td>&gt;130°</td>
<td>&lt;140°</td>
</tr>
<tr>
<td>Knee Extension</td>
<td>≤-10°</td>
<td>&gt;-10°</td>
<td>&lt;0°</td>
</tr>
<tr>
<td>Ankle Dorsiflexion (flexed knee)</td>
<td>≤10°</td>
<td>&gt;10°</td>
<td>&lt;20°</td>
</tr>
<tr>
<td>Ankle Dorsiflexion (extended knee)</td>
<td>≤0°</td>
<td>&gt;0°</td>
<td>&lt;10°</td>
</tr>
<tr>
<td>Hip Internal rotation</td>
<td>≤30°</td>
<td>&gt;30°</td>
<td>&lt;40°</td>
</tr>
<tr>
<td>Hip External rotation</td>
<td>≤30°</td>
<td>&gt;30°</td>
<td>&lt;40°</td>
</tr>
<tr>
<td>Elys’ test</td>
<td>≤100°</td>
<td>&gt;100°</td>
<td>&lt;120°</td>
</tr>
<tr>
<td>Hip Extension</td>
<td>≤0°</td>
<td>&gt;0°</td>
<td>≥0°</td>
</tr>
</tbody>
</table>

### GMFCS IV – V

<table>
<thead>
<tr>
<th>GMFCS IV – V</th>
<th>Red</th>
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<th>Green</th>
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<td>&lt;30°</td>
</tr>
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<td>&lt;40°</td>
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<td>Hip External rotation</td>
<td>≤30°</td>
<td>&gt;30°</td>
<td>&lt;40°</td>
</tr>
<tr>
<td>Elys’ test</td>
<td>≤90°</td>
<td>&gt;90°</td>
<td>&lt;110°</td>
</tr>
<tr>
<td>Hip Extension</td>
<td>≤-10°</td>
<td>&gt;-10°</td>
<td>&lt;0°</td>
</tr>
</tbody>
</table>

Work to keep neutral hip ext!

*Bousquet (2018)*
Hip Health

Surveillance

– Regular hip surveillance (xray) every 6 months
– [www.cpup.se](http://www.cpup.se)

“to prevent the occurrence of hip dislocation and severe deformities by means of a continuous and standardized surveillance, if necessary combined with treatment at an early stage and thereby optimize the functional ability and quality-of-life of those with CP”

Early (by 9 mos) and daily positioning in hip abduction for those with spasticity or significant weakness at risk for subluxation or dislocation

– Sitting
– Lying
– Standing
– Hippotherapy (on a horse)


CARE PATHWAYS

HIP SURVEILLANCE IN CEREBRAL PALSY

Authors (AACPDm Hip Surveillance Care Pathway Team): M O’Donnell (team lead), T Mayson (project manager and clinical examination sub-group leader), S Miller (radiology sub-group leader), R Cairns, K Graham, S Love, F Miller, K Mulpuri, U Narayanan, H Read, B Shore, K Stannage, P Thomason, J Vargus-Adams, L Wiggins, R Willoughby, M Wynter
Hippotherapy

- Core strengthening
- Postural control/balance
- Hip health
- Pelvic mobility
- FUN!
Hip surgery

Definitions

- Subluxation 30°/33° (new) 40° (old) to 99% uncovered
- Dislocation 100% “out”

Once 40% of femoral head is uncovered, it is the point of “no return” and surgery is necessary
Early correction is best to prevent secondary impairments
Surgery

A variety of surgical procedures can be used to help correct static and dynamic defects:

– lengthen, transfer, move attachments, or divide tendons
– relocate joints
– fuse joints
– rotate bones
– transect specific nerves
– correct bony abnormalities
– release contracted skin
Orthopedic Surgery Goals:

– To correct or prevent deformity or contractures
– To improve muscle balance across a joint
– To stabilize a joint
– To decrease energy requirements, especially during walking

• Multiple events vs. single event, multilevel surgery (SEMLS)
• Delay the operation for as long as possible, preferably until child is at least 6 yrs old, except correction for hip subluxation
Mobility Interventions

Novak, 2020
Strength and power are directly related to function in people with CP.

- Isokinetic
  - Biodex dynamometer
- Isotonic
  - Free weights
  - Weight machines
  - Dynamometer
- Closed chain
  - *Total Gym (weight bar)
  - Leg Press
  - Weighted squats
- Functional activities
  - Sit to stand, etc.

Power Training instructions: "Lift/Push as fast as possible. Lower slow and controlled over 2-3s"
<table>
<thead>
<tr>
<th></th>
<th>Intensity</th>
<th>Volume</th>
<th>Speed</th>
<th>Frequency</th>
<th>Rest</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle Strength</td>
<td>&gt;85% of 1RM</td>
<td>Build to 3 sets of 6-10</td>
<td>Slow to moderate controlled</td>
<td>2–3 x/wk (non-consecutive)</td>
<td>1-2 min between sets; 24 hrs btw sessions</td>
<td>8-20 weeks</td>
</tr>
<tr>
<td>(High resistance)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Power</td>
<td>40-85% of 1RM</td>
<td>Build to 6 sets of 5-6</td>
<td>Concentric part “as fast as possible”</td>
<td>2–3 x/wk (non-consecutive)</td>
<td>1-2 min between sets; 24 hrs btw sessions</td>
<td>8-20 weeks</td>
</tr>
<tr>
<td>(High resistance &amp; High speed)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bone Mass &amp; Structure</td>
<td>High ground reaction force</td>
<td>50-100</td>
<td>High strain</td>
<td>3-6 x/wk (non-consecutive)</td>
<td>1-10 sec btw reps; 4-8+ hrs btw sessions</td>
<td>9-12 months (min 3 months)</td>
</tr>
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</tr>
</tbody>
</table>

Optimal Training Parameters - Summary
Hippotherapy

- Core strengthening
- Postural control/balance
- Hip health
- Pelvic mobility
- FUN!
Electrical Stimulation

1. Neuromuscular Electrical Stimulation (NMES)
   - Surface electrical stimulation to muscles for the purpose of strengthening or training a muscle.
   - Typically higher intensity, shorter duration

   a. Functional Electrical Stimulation (FES)
      Surface or implanted electrical stimulation to muscles or nerves to perform a motor activity. Originally use of NMES for an orthotic, but now means stimulation for functional purposes

   b. Task Specific Electrical Stimulation (TASES) - term used by Judy Carmick to differentiate it from ES. Functional if timing is correct for task, but is still beneficial for ROM and muscle strengthening. NMES used with a remote triggering switch.

   c. Lateral Electrical Stimulation (LESS) - used for scoliosis
Tips:
- Always try it on yourself first!
- Comfort is essential
- Find the motor point with the electrode in your hand
- ABC: Active Black Cathode
E-stim settings for peds

- Usually biphasic wave forms
- 35-50 pps have been used and studied in pediatrics (40-50 usually too high for peds)
- Twitch contraction 1-10 pps
- Tetanic Contraction
  - Non fatigue = 15 to 40 pps
- Recommended pulse duration: 300 – 400 µs
- If too short needs amplitude to be high and maybe uncomfortable.
- Stimulation only occurs under electrodes, so use 2 electrodes on larger muscles
- TRY ON YOURSELF FIRST

- Have child just get used to electrodes first, then do sensory level only, work up to tetanic contraction once child is comfortable; FOLLOW THE CHILD’S LEAD
Bone health

Mechanical loading is needed during critical periods of bone growth

Ideally you need weight bearing with active muscle tension pulling with different forces in varying directions!
Gait training
Mobility training, including gait training

What matters in gait training is intensity!
Treadmill and overground training are both effective.

Adapted ride cars and early mobility

If a child is not sitting independently by 2 years old, the child will most likely not learn to walk independently
Provide independent mobility early!

*Pediatric Physical Therapy*
2012; 24: 149-154
Modified Ride-on Toy Cars for Early Power Mobility

Prone scooterboards help babies learn to propel themselves when it is developmentally appropriate

http://www.udel.edu/gobabygo/
http://nationswell.com/babiesdrivingracecars/
http://newsok.com/article/3610368
Neurodevelopmental Treatment (NDT)=Facilitation
Minimal handling for maximal active movement
(not passive type!)

• Handling, guiding, manual assistance, assisting with
  alignment, external feedback for initial motor learning,
tactile cues...
  – Whatever you call it or approach you use, you assist your
    patient with components of movement (like weight
    shifting), balance, stability, transitions, motor control,
    and functional movement
Perception-Action Approach

- Uses current motor control theories
  - Minimized guidance
  - Internal feedback for long term motor learning
  - Allows patient to safely explore movement within their environment
  - Trial and error learning
Postural control and balance training
Standing programs

• Daily standing with hip abduction (15-30 degrees each leg)
  – 60 minutes a day, according to research
  – Can be two 30 minute sessions
  – Reduces leg spasticity, increases bone mineral density, and improves range of motion of hip adductors, hamstrings, gastrocnemius muscles

Whole Body Vibration

- Bone density
- Strengthening
- Proprioception

Susan Hastings, PT, DPT, PCS (2016)
Physical Activity/Fitness

• Most children with CP are less active than their peers, have poorer cardiorespiratory fitness, & higher oxygen costs

• Physical activity in childhood can establish lifestyle habits for adulthood
  – exercise programs for children with CP can increase muscle strength and aerobic capacity without causing adverse effects
FITT recommendations for children and adolescents

<table>
<thead>
<tr>
<th>Frequency</th>
<th>Intensity</th>
<th>Time</th>
<th>Type</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aerobic</strong></td>
<td>daily</td>
<td>Moderate to vigorous</td>
<td>Running, fast walking, swimming, dancing, cycling, etc.</td>
</tr>
<tr>
<td><strong>Muscle</strong></td>
<td>3 or more days/wk</td>
<td>part of 60 min/day</td>
<td>Can be unstructured active play or structured (lifting weights, etc.)</td>
</tr>
<tr>
<td><strong>Bone</strong></td>
<td>3 or more days/wk</td>
<td>part of 60 min/day</td>
<td>Running, jumping, impact sports, resistance training</td>
</tr>
</tbody>
</table>

Adaptive sports and leisure in the community

Sense of belonging + purpose + fun = great quality of life and health
Constraint vs. bimanual training

• For children with unilateral weakness, constraining the uninvolved upper extremity during periods of intense practice shows improvements that are sustained if the family continues to practice regularly.

• Bimanual training (using both UEs together) has the same outcome, but it is the regular practice that is the key.

Mirror Therapy

- Activation of mirror neurons can reduce neglect of hemiplegic limb and improve hand function

Biofeedback

- Can improve motor performance and motor learning


UE splinting

- Resting splints and stretch splints for night
- Dynamic splints need to improve function for day use
Strapping, fabrifoam, dynamic trunk orthoses, and compression suits

goals:
– biomechanical alignment
– improve patient posture and function
– enhance sensory input, especially proprioception

Lateral hip rotation strapping with abdominal assist for hip & femur. Maria, age 4 years, diplegic CP

http://theratogs.com/intoeing-out-toeing/

Work best if a child just needs minimal assist for posture or movement
Self care and function
Interventions

Novak, 2020
Intensity of practice requires parent and caregiver education

- Goal-directed training in natural environments
- Training and implementation by parents, caregivers, teachers, aides, etc.
  - Ask these people how and when implementation would work for them each day
  - Focus on what will give you the most reward for the least effort in the priority areas
  - Most families can handle only 1-3 things a day
Adaptive equipment/positioning

http://www.rifton.com/products
Assistive technology
Parent Outcomes

Interventions

Novak, 2020
Quality of Life Research

Parent questionnaires of adolescents with CP:

• Over time, parents’ hopes for a cure were transformed into hopes for their child’s happiness.

• The parents’ and children’s ability to make choices about their preferences translated to highest “happiness” scores.

Movement Early in Life **Required** for Normal Spinal Cord Development

If extremities don’t move normally early in life during the postnatal refinement period (weeks 3 to 7 for cats), for whatever reason, there is permanent spinal cord loss of corticospinal tract synapses. Early intervention is critical!

Good rehabilitation outcomes for kids with CP incorporate these principles:

• Intensity matters! Daily parent/child home programs of effective interventions are a must.
• Saliency matters! It must be important to the patient/family!
• Intervention at critical periods of development is essential! Early intervention (ei) is most effective!
• “Use it or lose it”
• Children should practice and discover in real world environments
• Independent initiation of movement is required to develop spatial cognitive awareness.
• Learning happens when training is task-oriented.
Any Questions

mbarkocy@salud.unm.edu