SPASTICITY MANAGEMENT FOR ADULTS WITH CEREBRAL PALSY

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Disclosures

No financial disclosures

PI Medtronic ISPR Registry
Co-PI Gablofen study
Objectives

- Epidemiology
- Spasticity
- Functional Changes
- Oral Medications
- Injection Therapy
- Orthopedics
- ITB therapy
- SDR
- Conclusions
Epidemiology

- CP most common major motor disorder of childhood
- Survival in adults with CP quite good
- California Data
  - CP not severe: 98.2% age 4-14 survived 20 yrs (1983-2002)
  - CP severe: 85% survived that period of time
  - Improvement in survival over time, mortality (most impaired children) improved an average rate of 3.4% per year
- Relatively high rates of survival with mild CP, increasingly better survival rates for those with severe impairments, population growing

“Spasticity is a motor disorder characterized by a velocity dependent increase in tonic stretch reflexes, with exaggerated tendon jerks resulting from hyperexcitability of the stretch reflex, as one component of the upper motor neuron syndrome” Lance 1980

- Resistance to stretch increases with increasing speed and varies with the direction of the joint movement
- Rapid rise in resistance to stretch above a threshold speed or joint angle

Sanger et al, Classification and Definition of Disorders Causing Hypertonia in Childhood, Pediatrics 2003
Why is spasticity important?

- Clinically diagnosed and treated
- Musculoskeletal and neurologic exam
  - Tone, reflexes, strength, coordination
- Spasticity → significant disability

- ADLs
- Seating
- Comfort
- Contracture
- Loss of ROM
- Negative impact on function
- Bone deformity
- Pain
- Skin
- Hygiene
- Ability to provide cares
Spasticity

Effect of comprehensive care and spasticity management on ADL self-performance and care delivery

- 205 pts intellectual disability (ID), 72 spasticity interfered w/ care or function and at least 1 ADL
- Care goal
- 54 met criteria, 38 consented, 20 completed (14 CP)
- Baseline evaluation (MAS, ROM, timed and videotaped care task)
- Spasticity treatment initiated, f/u after optimization
- Outcome measure: comparison of ease of videotaped care
- Results: significant improvement across all outcome measures. Comprehensive spasticity management – meaningful improvement in ADL care for pts with ID

PD Charles, MD et al, Spasticity Treatment Facilitates Direct Care Delivery for Adults with Profound Intellectual Disability. Movement Disorders 2010; 25 (4):466-73
Functional Changes

- CP reduced muscle volumes in LE
- To maintain function must be above threshold value
- Sarcopenia - loss of muscle mass in developing adult
- Muscle deficits CP, with decline of muscle properties associated with aging, contribute to early loss of mobility
- Value of strengthening – improve muscle reserve in short-term and maintain muscle mass above critical threshold in the long-term

Adam Shortland PhD, Muscle deficits in cerebral palsy and early loss of mobility: can we learn something from our elders? Developmental Medicine and Child Neurology 2009,51(suppl.4):59-63
McMaster group, developed GMFM

Only large longitudinal study of motor function

Subset 229 from original study, additional longitudinal study GMFM to age 21

Most severe disability (GMFCS) curves estimated declines in function into adulthood

Motor decline – increased body size, decreased activity and changes in spinal alignment

Traditional Step-Ladder Approach to Management of Spasticity

- Neurosurgical Procedures
- Orthopedic Procedures
- Neurolysis/Chemodenervation
- Oral Medications
- Rehabilitation Therapy
- Remove noxious stimuli
Oral Medications

- Oral medications alone inadequate
- Side effects (SE) may negatively impact function and health
- SE – dose minimized, in conjunction with other treatments i.e. Injections.
- Multimodal approach
- Outcomes optimized by combining interventions
- Systemic medication – generalized spasticity, passive ease of care

GABAergic
- Benzodiazepine
- Baclofen
- Alpha-2 adrenergic agonists
  - Clonidine
  - Tizanidine

Dantrolene

GABA analogs
- Pregablin
- Gabapentin
Injection Therapy

Distribution

- Focal
- Functional group of joints
- Limb
- Multi-segmental – spastic hemiparesis

Goals

- Improved ROM, reduction of co-contraction
- Improved local function
- Ease of cares/comfort
- Prevent secondary complications: contractures, skin breakdown

Other co-morbidities: seizures, altered cognition

Orthopedic

Primary neurologic disorder that secondarily affects the musculoskeletal system

- Musculotendinous/ tendon lengthenings
- Tendon transfers
- Osteotomies
- Arthrodesis

- Lever-arm dysfunction:
  malrotation of the bony anatomy results in functional weakness

- Equinas foot deformity
- Contractures
- Patella alta
- Hip dysplasia
- Degenerative arthritis
- Spondylolysis
- Cervical stenosis

Kevin P Murphy, MD. Cerebral palsy lifetime care – four musculoskeletal conditions. Developmental Medicine and Child Neurology 2009, 51 (Suppl. 4): 30-37
ITB Therapy

ITB treatment outcomes in Adults with CP

- Review of ITB for adult spasticity (spinal or cerebral origin) – increased independence, mobility, self-care abilities, improved sleep, decreased bladder hyper-reflexia and decreased pain
- QOL: reduced muscle tone without sedation, ease of caregivers, improved ease of positioning, less pain/increased and improved transfers

Plasma vs CSF drug levels

Oral 60 mg/day
baclofen ng/ml

IT-Chronic 200 mcg/day
baclofen ng/ml

ITB Therapy

Cost

- Comparison of simulated continued conventional medical management protocol versus ITB therapy
- Cost projections over 30 yrs
- ITB was less costly than conventional protocol over baseline implantation cycle
- Cost in the month of implantation and following year, cumulatively 26K more than conventional
- ITB financial breakeven occurs between 2\textsuperscript{nd} and 3\textsuperscript{rd} year post implant
- Lifetime analysis savings for ITB 8k per patient per year
- Reductions inpatient admissions, physicians office visits and outpatient PT

Selective Dorsal Rhizotomy

- Descriptive study: to assess outcomes in adults with CP who underwent SDR as children
- Survey, older adolescents/adults with CP, SDR 1986-2000
- Subjective impressions and medical interventions post-SDR
- function, pain, education, living situation, subjective health status and satisfaction with life

**TABLE 3: Main outcome variables according to GMFCS levels**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>GMFCS Levels I–II</th>
<th>GMFCS Level III</th>
<th>GMFCS Levels IV–V</th>
</tr>
</thead>
<tbody>
<tr>
<td>no. of patients</td>
<td>22</td>
<td>20</td>
<td>46</td>
</tr>
<tr>
<td>history of orthopedic procedure after SDR (% patients)</td>
<td>77</td>
<td>75</td>
<td>72</td>
</tr>
<tr>
<td>perception of health (% patients)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>excellent</td>
<td>18</td>
<td>40</td>
<td>24</td>
</tr>
<tr>
<td>very good</td>
<td>45</td>
<td>30</td>
<td>26</td>
</tr>
<tr>
<td>good</td>
<td>32</td>
<td>20</td>
<td>41</td>
</tr>
<tr>
<td>fair</td>
<td>0</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>poor</td>
<td>5</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>recent pain (% patients)</td>
<td>64*</td>
<td>25</td>
<td>44</td>
</tr>
<tr>
<td>chronic back pain (% patients)</td>
<td>77†</td>
<td>25</td>
<td>50</td>
</tr>
<tr>
<td>mean SWLS scores‡</td>
<td>27.3 ± 5.9</td>
<td>27.9 ± 6.0</td>
<td>24.6 ± 8.3</td>
</tr>
</tbody>
</table>

* p = 0.03.
† p < 0.01.
‡ Not significant.
TABLE 4: Perceptions of the rhizotomy among 88 patients who underwent the procedure as children

<table>
<thead>
<tr>
<th>Question</th>
<th>% Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>How did the rhizotomy affect your QOL?</td>
<td></td>
</tr>
<tr>
<td>increased</td>
<td>65</td>
</tr>
<tr>
<td>decreased</td>
<td>8</td>
</tr>
<tr>
<td>no change</td>
<td>5</td>
</tr>
<tr>
<td>not sure</td>
<td>22</td>
</tr>
<tr>
<td>Would you recommend the rhizotomy to others?</td>
<td></td>
</tr>
<tr>
<td>yes</td>
<td>65</td>
</tr>
<tr>
<td>no</td>
<td>4</td>
</tr>
<tr>
<td>not sure</td>
<td>31</td>
</tr>
</tbody>
</table>

Fig. 1. Bar graph demonstrating GMFCS level versus effects on QOL. Most respondents who noted a decreased QOL or no change were at GMFCS Levels IV and V. Light gray bar indicates improved QOL; medium gray bar, decreased QOL or no change; and dark gray bar, not sure. # = number.
### TABLE 5: Summary of pain data

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Any Pain Recently?</th>
<th>Chronic Back Pain?</th>
</tr>
</thead>
<tbody>
<tr>
<td>positive response (% of patients)</td>
<td>44</td>
<td>51</td>
</tr>
<tr>
<td>average pain score (NRS 0–10)</td>
<td>5.5 ± 2.8</td>
<td>5.9 ± 2.8</td>
</tr>
<tr>
<td>average pain range</td>
<td>2–10</td>
<td>1–10</td>
</tr>
<tr>
<td>% patients w/ pain mild (1–3)</td>
<td>31</td>
<td>20</td>
</tr>
<tr>
<td>moderate (4–7)</td>
<td>41</td>
<td>52</td>
</tr>
<tr>
<td>severe (8–9)</td>
<td>11</td>
<td>13</td>
</tr>
<tr>
<td>very severe (10)</td>
<td>17</td>
<td>15</td>
</tr>
<tr>
<td>satisfaction w/ life score</td>
<td></td>
<td></td>
</tr>
<tr>
<td>pain</td>
<td>24.0†</td>
<td>24.3‡</td>
</tr>
<tr>
<td>no pain</td>
<td>27.7</td>
<td>28.2</td>
</tr>
</tbody>
</table>

* The past week.
† p = 0.02.
‡ p = 0.01.

### TABLE 2: Summary of health outcomes and interventions in 88 patients with CP

<table>
<thead>
<tr>
<th>Parameter</th>
<th>% Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>perception of health</td>
<td></td>
</tr>
<tr>
<td>excellent</td>
<td>26</td>
</tr>
<tr>
<td>very good</td>
<td>32</td>
</tr>
<tr>
<td>good</td>
<td>34</td>
</tr>
<tr>
<td>fair</td>
<td>6</td>
</tr>
<tr>
<td>poor</td>
<td>2</td>
</tr>
<tr>
<td>presence of scoliosis</td>
<td>41</td>
</tr>
<tr>
<td>scoliosis intervention</td>
<td>10*</td>
</tr>
<tr>
<td>orthopedic surgery</td>
<td></td>
</tr>
<tr>
<td>hip surgery</td>
<td>36</td>
</tr>
<tr>
<td>tendon lengthening surgery</td>
<td>63</td>
</tr>
<tr>
<td>adductors</td>
<td>20</td>
</tr>
<tr>
<td>hamstrings</td>
<td>38</td>
</tr>
<tr>
<td>heel cords</td>
<td>35</td>
</tr>
<tr>
<td>other†</td>
<td>&lt;5</td>
</tr>
<tr>
<td>baclofen pump</td>
<td>15‡</td>
</tr>
<tr>
<td>injections (botulinum toxin &amp;/or phenol)</td>
<td>53</td>
</tr>
<tr>
<td>current oral spasticity medication</td>
<td>38</td>
</tr>
</tbody>
</table>

* Represents 25% of scoliosis patients—6 surgeries, 3 orthotic and seating only.
† Iliopsoas, quadriceps, upper extremity, toes and feet.
‡ One-third of these have had the pump removed.

Conclusions

- The number of individuals living with CP is increasing.
- Adults with CP often develop musculoskeletal and neurologic symptoms, such as severe pain, chronic fatigue and premature decline in mobility and function as they age. This negatively impacts independence, activity participation and QoL.
- Spasticity can interfere with functional abilities, contribute to the development of contractures, bony deformities, pain and impair seating/positioning and ease of cares.
- Classic model of delivering spasticity management in a linear hierarchical approaches has evolved to a more modern approach of choosing singular or multiple interventions.
- 70% of individuals with CP have spasticity. Comprehensive spasticity management helps prevent or improve symptoms associated with the secondary complications of spasticity.

Adam Shortland PhD, Muscle deficits in cerebral palsy and early loss of mobility: can we learn something from our elders? Developmental Medicine and Child Neurology 2009, 51(suppl. 4):59-63

Sanger et al, Classification and Definition of Disorders Causing Hypertonia in Childhood, Pediatrics 2003

Allison Brashear, Elie Elovic, Spasticity Diagnosis and Management, 2010, Ch 1.1 Why is spasticity important?

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