Managing the Musculoskeletal Impairments in Children with Cerebral Palsy
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Future Options for Managing Complex Multi-system Disabilities
Freeman Miller

CP and CP like Conditions
Impairments:
Musculoskeletal and motor control
Behavior
Cognitive

Goals for Childhood Management
Maximize function at maturity
Defined by the outcome measures
ICF from World Health Organization
Underlying Assumption

At skeletal maturity
Body function/structure will remain stable
Activity and Participation may decrease due to early aging

What is the evidence that changes in adults remain stable?

Recall of 101 adults

All have gait full gait evaluations as teenagers
They are now 25-45 year old (30 mean)
mostly GMFCS I-III
13.3 year follow up

Result - 101 evaluations

Body function
Activity

Result - 101 evaluations

PROMIS outcomes

Unpublished results
Teenage to Mid Adulthood
For GMFCS I-III
Motor function remains stable
Pain, depression, and participation
Are close to age matched peers
Consistent with other published reports


Adolescent function is maintained in adults
(GMFCS I - III)

Conclusion
Our goal for Childhood Management
Should be Maximize Adolescent Function
Physical function - Gait activity
Participation
Psychologic
Social

Childhood Orthopedic Care
Improve Activity, Participation, and QoL

GMFCS I - III -
maximize mobility function

GMFCS IV -V
Prevent hip dislocation (hip health)
Manage spinal deformity
Comfort care -
Tone and Contracture Management

Management Goals
Short term outcome
Is there treatment benefit 3,6,12 months
Does this benefit QoF of child or family?
Control groups are needed to understand treatment effect.

Little good objective data.

Loss of walking ability:
- 5 y/o
- 10 y/o
- 13 y/o

This is the natural history of diplegia falling into a crouch.
- Poor social and family structure.
- No follow up.
- Loss of function is preventable.

Natural history:
- Tripletic, intelligent, family involved, good PT.
- 4 y/o
- 5 y/o
- 11 y/o
- 17 y/o

Planovalgus:
- 3 y/o
- 6 y/o
- 9 y/o
Natural History
Predicting expected outcome with GMFCS

Rocket Science
Can we hit the target of ideal CP treatment?

GMFCS Has Variability
Cause large group overlaps

CP Predictions - GMFCS
Childhood Motor Development - Forward based prediction modeling based large current and historical data.

Natural History spasticity
Gastroc-soleus spasticity Ashworth Scale

Natural History
Strength per Weight

Muscle Length
Muscle length does not grow as much as bone length
Causes - “muscle contracture”

CP Muscle Natural History Relative to Growth
Tone - decreases
Strength - relative to weight decreases
Length - relative to height decreases

Adolescent growth - weak, less spastic, and short muscles
Evidence for Gait Analysis
Assess pathology and prescribe treatment

Does gait analysis improve outcome?
YES when the recommendations are followed


Melbourne Study

1. Gait
2. Gross Motor Function
3. Quality of Life
4. Activities and Participation

Only one study with a control group

Does Orthopedic Surgery Improve Outcome

Inclusion:
Spastic Diplegia
7-12 years
GMFCS Levels II and III
require SEMLS

Exclusion:
hip subluxation (MP>20%)
SDR within 12 months
IT Baclofen
Botulinum toxin A within 6 months

Results

<table>
<thead>
<tr>
<th>Baseline Measure</th>
<th>SEMLS</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. Children</td>
<td>11</td>
<td>8</td>
</tr>
<tr>
<td>Mean Age (SD)</td>
<td>9.5 (1.4)</td>
<td>9.9 (1.6)</td>
</tr>
<tr>
<td>GMFCS II</td>
<td>7</td>
<td>7</td>
</tr>
<tr>
<td>GMFCS III</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>GGI (SD)</td>
<td>370 (225)</td>
<td>355 (183)</td>
</tr>
<tr>
<td>GMFM (SD)</td>
<td>65.3 (11.1)</td>
<td>70.3 (11.3)</td>
</tr>
<tr>
<td>Gait Pattern - True Equinus</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>- Jump</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>- Apparent Equinus</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>- Crouch</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>- Asymmetric</td>
<td>0</td>
<td>1</td>
</tr>
</tbody>
</table>
Outcome


5 year follow-up
Shows gait function maintained

Post-op SEMLS 1
19 Y/o


Conclusion Relative to Natural Evolution

Moderate evidence - Gait analysis directed orthopedic surgery improves gait impairment from childhood to adolescence
Poor to No Evidence that it improves

Pre-op SEMLS 2


Stiff knee gait
Crouch Gait
Equinus


Conclusion Relative to Natural Evolution

Moderate evidence - Gait analysis directed orthopedic surgery improves gait impairment from childhood to adolescence
Poor to No Evidence that it improves

Stiff knee gait
Crouch Gait
Equinus


Conclusion Relative to Natural Evolution

Moderate evidence - Gait analysis directed orthopedic surgery improves gait impairment from childhood to adolescence
Poor to No Evidence that it improves
Does Spasticity Management Improve Gait Impairment?

Rhizotomy - one long-term study with a comparable control group showed no benefit over orthopedic only

Botulinum Toxin - no evidence of any long-term benefit

Intrathecal Baclofen - no evidence


No Evidence of long-term adult functional stability

GMFCS IV-V

Hip Health - Natural History is defined
Spinal Deformity - Natural History is well known

CP Hip Subluxation & Dislocation Risk by Age

Age at which subluxation or dislocation occurs

% Total

Age 2 3 4 5 6 7 8 9 10 12 14 16 18

CP Hip Subluxation Risk by GMFCS

Risk of developing MP>30% or MP>33%

1082 patients

Data summary from four publications
CP Hip Subluxation
Rate of progression by GMFCS

Soo et al., JBJS 2006, AND Hagglund, JBJS 2005

Natural History

1.5Y 2.5Y 5Y 10Y 13Y 15Y

Hip Treatment Protocols
Muscle surgery in young Children

duPont Outcome - 67% Good+Fair (<40%) 33% Poor+Failure (>40%) At Maturity


Screening and Surveillance Programs

AACPDM - release a consensus program
Australian
Scottish
Swedish

Australian Outcome Reports
Good outcomes -
GMFCS II - 94%, GMFCS III - 49%, GMFCS IV - 27%, GMFCS V - 14%

Graham K. Adductor Surgery to Prevent Hip Displacement in Children with Cerebral Palsy. The Predictive Role of the GMFCS. JBJS Am 2012. 94: 326-34

Outcome difference likely due to surgical dose *
Hip Treatment Protocols

**Hip Bone Reconstruction** - highly successful
Agreed world wide standard of care
Hips remain reduced and near normal

No evidence that it improves participation
Evidence that it improves Caregiver outcome


Other treatment options

**Orthotics and PT**

No data with controls

**Hip Treatment Protocols**

**Botulinum toxin**

Good double blinded trial showing no clinical benefit


Natural history Scoliosis

Surgical instrumentation and fusion

When scoliosis curve 60 - 90 degrees
Outcome - Improved Quality of Life
Spinal surgery has the best result in patient reported outcome

Spasticity Management in GMFCS IV - V

Intrathecal Baclofen - current standard
- Requires continuous follow up
- Significant complication rate
- Multiple reports of high caregiver satisfaction


Conclusion
- Lack good data on long-term natural history
- Based on limited data, surgical correction of deformities in GMFCS I-III improves and maintains gait function long-term
- There is no data suggesting spasticity management improves function long-term
- Preventing hip dislocation is effective
- Correction of scoliosis improves life quality

What is the Current State?
- There are 100s of short term outcome studies
- How do we consider short term data with long-term?
  - Meaning of 3 or 12 month results in a 2yr?
  - Botulinum - classic example
  - Double Blinded research evidence of reduced spasticity and gait effect at 3 month
  - Longer outcome studies show permanent harm to the muscle due to fibrosis
  - Neither effect has shown a long-term outcome impact on function (no data)

What is needed?
- A strong focus to develop Patient Registries
  - Where short term studies are entered and subsequently followed up long-term
  - That have specific data fields relevant to treatment given
  - Include data on social history, cognitive and behavioral function.
  - A commitment to follow up through the patient’s complete growth into adulthood

Merci