Orthopaedic Issues in Adults with CP: If I Knew Then, What I Know Now

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Epidemiology

- 87-93% of children born with CP survive into adulthood (Nielson 2002)
- 85% of cohort alive at 20 survived to age 50 (Hemming 2006)
- Exact number of individuals in US unknown, estimate 700,000 to 1 million
- Number increasing due to increased survival of low-birth-weight infants and increased longevity of adults
Essential to assume that ALL patients will outlive us.
Curmudgeon’s Perspective

- Paradigm shift in how disability is viewed. We have moved from a largely medical to a social model.
- That is great for many disorders, but not cerebral palsy.
- Pain from contractures, fractures, subluxations/dislocations, sores, etc must be a health delivery priority if individuals with cerebral palsy are to live long and live well.
Orthopaedic Core Concepts:

- Our first responsibility is to prevent pain
- Bones, muscles, and joints are the most important parts of the body
- Move it, or lose it
#1 I would ensure that every child with cerebral palsy had a complete musculoskeletal exam annually

AND that non-ambulating children had a hip xray
Rationale
Hip subluxation is highly correlated with limited hip abduction
#2 I would only allow wheelchairs to be used for transportation
Cartilage nutrition depends on joint motion – individuals who sit all day are starving their cartilage.
#3 I would encourage parents and caregivers to learn about Bone Health
Bone is Unique

- Structure & protection
- Major storage form of calcium in body
- Only organ able to heal w/o scarring

www.dechildrens.com
Strong bones are not a right

- Key elements of peak bone mass
  - Weight bearing exercise
  - Nutrition
  - Genetics
  - Ethnicity

Normal vs. Osteoporotic
PEAK BONE MASS

Gender

Bone Mass (Grams of Calcium)

Fracture Threshold
Dominant vs Non-dominant Arm
Calcium PLUS Exercise

Placebo
Calcium supplement

Fine motor
- Specker & Binkley, J Bone & Min Research 2003

Gross motor
Measuring Bone Density
Interpreting DXA’s

- **T score** refers to how the patient compares to a cohort of healthy young females.
- **Z score** compares the patient to individuals of their same age and sex.
Prevalence of reduced bone mass in children and adults with spastic quadriplegia

King W et al. Dev Med Child Neurol 2003

Reviewed 48 patients 5-48 years (median 15)

Lumbar spine Z score: -2.37 ± 0.21

58% had z-score: < -2

39% had history of fracture

Those with history of fracture had significantly lower z score

with history of fracture: -2.82 ±0.29

without history of fracture: -2.11±0.26

Age and Vitamin D level not significant
Bone Density and Metabolism in Children and Adolescents With Moderate to Severe Cerebral Palsy  
Henderson et al  
*Pediatrics* 2002

117 subjects  2-19 yrs old (mean 9.7)

-Osteopenia in femur of 77\% of population based cohort
  -Older than 9 years old: prevalence of 86\% (19 out of 22)
  -Of the 3 who did not have osteopenia—2 were capable of assisted ambulation
-\( BMD \) severely diminished in distal femur  
\( z\)-score -3.5 ± 0.2
Bone Density and Metabolism in Children and Adolescents With Moderate to Severe Cerebral Palsy Henderson et al

Pediatrics 2002 continued

15% had already fractured (of those, 38% multiple fx)
Fractures occurred in 28% of children older than 10 yrs

BMD $z$ score correlated strongly with Gross Motor Function Level

- 96% of level 5 children had osteopenia
- 43% of level 3 children had osteopenia
Is this a push for standing frames?

Well, yes and no
A randomized controlled trial of standing programme on bone mineral density in non-ambulant children with CP
Caulton et al Arch Dis Child. 2004

26 children with CP; 14M, 12F; age 4.3-10.8 yrs

Intervention group increased their standing duration by 50% for the academic school year

Results: 6% increase in vertebral BMD
no BMD increase in tibia—authors conclude standing program does not decrease risk of long bone fracture
Implications for children and adults with CP

Fractures in Patients with Cerebral Palsy

<table>
<thead>
<tr>
<th>Bone Group</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Axial skeleton</td>
<td>4%</td>
</tr>
<tr>
<td>Skull</td>
<td>1%</td>
</tr>
<tr>
<td>Vertebrae</td>
<td>2%</td>
</tr>
<tr>
<td>Pelvis</td>
<td>1%</td>
</tr>
<tr>
<td>Upper Limb</td>
<td>14%</td>
</tr>
<tr>
<td>Clavicle</td>
<td>1%</td>
</tr>
<tr>
<td>Humerus</td>
<td>8%</td>
</tr>
<tr>
<td>Radius</td>
<td>2%</td>
</tr>
<tr>
<td>Hand</td>
<td>3%</td>
</tr>
<tr>
<td>Lower Limb</td>
<td>82%</td>
</tr>
<tr>
<td>Femur</td>
<td>48%</td>
</tr>
<tr>
<td>Proximal metaphysis</td>
<td>26%</td>
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<tr>
<td>Shaft</td>
<td>24%</td>
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<tr>
<td>Distal metaphysis</td>
<td>50%</td>
</tr>
<tr>
<td>Tibia</td>
<td>27%</td>
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<tr>
<td>Proximal metaphysis</td>
<td>46%</td>
</tr>
<tr>
<td>Shaft</td>
<td>36%</td>
</tr>
<tr>
<td>Distal metaphysis</td>
<td>18%</td>
</tr>
<tr>
<td>Foot</td>
<td>7%</td>
</tr>
</tbody>
</table>

Around the knee 36%
Will these individuals do better?
Low Magnitude Mechanical Stimuli are Anabolic to Bone
So, what about nutrition?

Bone density and fracture studies are inconclusive about the role of low vitamin D in bone health in children with CP.
What do I know? What do I see?

A Treatise of the Rickets: Being a Diseal common to Children.

Wherin (among many other things) is shewed,
1. The Essence
2. The Causes
3. The Signs
4. The Remedies

Published in Latin by Francis Glisson, George Bate, and Abraham Regemont; Doctors in Physick, and Fellows of the College of Physicians at London.

Translated into English by Phil. Armin.

London: Printed by Peter Cole, at the sign of the Printing-Prest in Cornhill, near the Royal Exchange, 1651.

www.dechildrens.com
Clinical example
#4 I would monitor spines more carefully

Because of its musculoskeletal origin, Scoliosis in CP patients can progress even after skeletal maturity is reached. Curves over 50 degrees progress 1 degree a year (Bleck et al 1984). Non-ambulatory ambulatory individuals are more likely to develop scoliosis than ambulatory individuals.
#5 I would insist that stretching and a fitness program are as important as English!
Racing Toward Immortality
(Or at Least Your 150th Birthday)

The spectacular — and scary — promise of embryonic-cell research.
By Stephen S. Hall