Objective:

1. Identify effective methods for the practical application of concepts related to improving the delivery of services for persons with developmental disabilities

2. Identify advances in clinical assessment and management of selected healthcare issues related to persons with developmental disabilities

Notes:
Bone Health In Cerebral Palsy Patients

Introduction

Overview

• Definitions
• Bone metabolism
• Bone Density
• Fracture Risk
• Low BMD Treatment
Definitions

• Cerebral Palsy
  – STATIC encephalopathy to the immature developing brain that may be due to anoxic or hypoxic brain injury
  – Diagnosis made OVER TIME
• Bone Mineral Density (BMD)
  – Measures amount of Ca/Phos in area of bone
  – Helps determine fracture risk

Normal Bone Metabolism
Bone Structure/Anatomy

Abnormal Bone

Section of a normal vertebral body with dense and well calcified trabeculae (left) and osteoporotic vertebral with resorption of trabeculae which weakens the mechanical properties of the vertebral body.
Abnormal Bone Metabolism

• Osteopenia 2° deficiency of bone tissue development
• Determined by GMFCS level

Osteoporosis in Children

• Defined by International Society of Clinical Densitometry
• Not based on BMD alone
• Current Definition:
  – BMD Z-score < -2.0 for age, gender, body size
  • Significant h/o fracture:
    • 2 upper extremity fractures
    • 2 vertebral compression fractures
    • Single lower extremity fracture

Bone Density

• DXA scan gold standard
• Comparison Values determined in CP pts
• Different than in adults
  – Lumbar spine (same)
  – Distal femur
    • Due to associated hip & knee contractures
  – Total body
DXA Scan

- Gold Standard for determining BMD
- Values differ between adult and pediatric pts
- Adults- T-scores
  - SD from mean of HEALTHY ref population
- Peds- Z-scores
  - SD from mean of AGE and SEX matched controls
  - Every SD drop equals 10-20% in BMD

DXA values

- Performed differently in CP pts due to joint contractures
- Lateral Distal Femur used
  - Due to spine/hip implants, accommodate for joint contractures
  - Common area of fx and low BMD
  - Scan pt in comfortable side-lying position
- 3 areas of distal femur evaluated: metaphysis, metadiaphyseal, cortical bone regions

DXA Scan CP Pts
Bone Density in CP

- 86% pts > 9 yoa had osteopenia of distal femur
- Fx risk DOUBLES w/ every standard deviation \( \downarrow \) BMD
- Wren et al:
  - CP pts all levels low bone density in tibias
  - Spine deficits greater in more involved children
  - Even higher functioning pts (GMFCS I/II) have \( \downarrow \) lower extremity bone accrual

Bone Density in CP

- Henderson et al (2005)-
  - Large bone health study of CP pts
  - Longitudinal study CP quads- decreased BMD associated w/ poor nutritional status
  - Osteopenia is manifestation of growth failure
  - Pts w/ lowest BMD: spastic quad, \( \downarrow \) mobility, poor nutrition
  - Non-ambulatory significantly lower BMD than ambulators
- Need long term studies to determine how peds CP pts will fair as adults
### Low Bone Density Factors

- Poor Nutrition
- Non-ambulatory
- Post-op Immobilization
- Anti-Seizure medications (Dilantin)

### Poor Nutrition

- Poor nutrition factors:
  - PO motor feeding difficulties
  - Food/Volume intolerances
  - GERD
  - Lack of sunlight exposure
  - Pharmacological

### Anti-Seizure Meds

- Phenytoin, Phenobarbitol, Carbamazepine
  - CYP 450 Enzyme induction
  - Increased clearance/catabolism of Vit D into non-active metabolites
  - 2° hyperparathyroidism and bone turnover
- Valproic acid
  - 10% reduction BMD
  - May cause renal dysfunction w/ Ca & Phos loss
  - May also directly activate osteoblasts/osteoclasts to increase bone turnover
- Carbamezapine
  - Induces bone turnover 2° osteoclast activation
Fracture Risk Factors

- Osteopenia/Thin bone cortices
- Stiff joints/contractures
- Poor balance/falls
- Seizures
- Previous Fx
- Poor nutrition/gastrostomy tube

Fracture Risk

- Prevalence Fxs CP pts: 5 to 60%
- Most common region: distal femur
- 66% in non-ambulatory pts
- Usually minimal trauma involved
- 2 to 6x ↑ fx risk in pts w/ CP and SZS d/o
- 4% per year fx incidence in moderate to severe CP pts

Fracture Morbidity

- Most fractures occur in lower extremity (i.e., distal femur)
- CP pts have ↑ fx risk than nl children, especially from 13-15 yoa (Maruyama)
- Cause pain, decubiti, further bone loss, difficulty w/c positioning/transport
- High cost for med care: $10,000 per pt, $150 million in U.S. per annum (Apkon)
  - Loss of school time for pt, loss of work time for caregiver
Screening

- Lab levels
  - Serum albumin
  - Calcium
  - Phosphorus
  - Alk Phos
  - 25-OH Vit D
- DXA scan
  - Baseline for all GMFCS IV and V pts upon fracturing (Apkon & Kecskemethy)
- All pts who sustain lower extremity fracture

Osteopenia Tx

- Physical Therapy
- Standers
- Low amplitude mechanical loading (vibration)
- Optimize nutrition
- Calcium + Vit D supplementation
- Bisphosphanates

Physical Activity/Therapy

- Weight bearing and resistance exercise important for development of bone mass
- Bones adjust strengthen in proportion to amount of stress placed on them
- Chad et al-
  - 8 months of weight bearing physical activity significantly ↑ total proximal femur and femoral neck BMC
  - Control patients (no weight bearing) ↓ 6% study period
- Little current evidence evaluating effect of PT on CP pts and BMD
Standing Frames

- Anti-gravity
- Theoretically, similar to resistant exercising, especially with dynamic standers

Caulton et al-
- 26 non-ambulatory CP pts, upright/semi-prone standers
- Avg 48 min/day x 5 days/week x 5 months
- Increased BMD of spine, not tibia

Katz et al-
- 12 non-ambulatory CP pts, upright stander
- 2 hrs/day, 5 days/week, 6 months
- RMD femur and calcaneous
  when compliant w/ standing program

Vibration Therapy

- Low-amplitude, high frequency vibration
- Usually incorporated as vibrating platform in standing frame

Ward et al-
- Children w/ limited mobility, RCT on vibrating platform vs sham platform and DXA assessment
- BMD ↑ 6.3% in tibia BMD and 5.5% spine BMD
Nutrition

- Optimize nutritional parameters
- Regular assessment by dietician
- Consider G-tube placement for supplemental nutrition
  - Arrowsmith et al-
    - G-tube supplementation ↑ total body protein and muscle mass, NOT ↑ BMD but did not ↓ over time either

Vit D Metabolism

Vit D/Ca

- Shaw et al did not find vit D status to coincide w/ magnitude of osteopenia
- Jekovec-Vrhovsec et al-
  - 23 CP quad pts, spine BMD, 9 mos supplementation Vit D and Ca
  - Pts w/ supplementation had significant ↑ BMD over those w/o
- Bischof et al-
  - Vit D supplementation ↑ fx rate in children w/ severe CP and rickets
- Henderson et al (2002)-
  - 9% ↑ BMD in mod to severe affected CP pts when given Ca and Vit D supplementation
Vit D/Ca

- AAP intake recommendations:
  - 400 IU/d if exclusively and partially breast-fed infant
  - 400 IU/D if non-breast fed and < 1000 mL/d of Vit D fortified milk for all children > 1 year old
  - 800 mg/day Calcium for children < 9 years
  - 1200-1500 mg/day Calcium for children 9-18 years

Bisphosphonates

- Henderson et al-
  - RCT w/ placebo arm w/ IV pamidronate
  - 89% increase in distal femur BMD over 18 months in quadriplegic CP
  - No pt in bisphosphonate group had fx (3 in control did)
- Grissom et al-
  - OI and CP pts, IV pamidronate, DXA scans
  - BMD both groups >4.0, all 3 femur regions improved in BMD
  - 38% reduction fx rate
- Allington et al-
  - Cyclic IV pamidronate 18 non-ambulatory CP pts
  - ↑ BMD, ↓ pain w/ manipulation, no add’l fxs 12 months after infusions
Bisphosphonates

- Iwasaki et al-
  - When treated with both Vit D and risedronate, BMD ↑ more than with Vit D alone
- Paksu et al-
  - 36 pts, CP quad
  - Weekly PO alendronate x 12 months
  - After 1 year, ↑ serum Ca and Phos, ↑ BMD and z-score
- Unal et al-
  - PO alendronate more cost effective than IV pamidronate to ↑ BMD

Treatment

Conclusions

- Bone health important in overall care of CP pts
- More study required regarding outcomes of various bone health modalities in CP pts
- Screening is important, especially after fracture
- Multiple tx options for ↓ BMD available, combo of all should be utilized to maintain bone health in CP population
References


Thank You