Bones and Hormones

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Bones – Not a steel building frame

Bones – The house that first expands then keeps remodeling
What do builders and remodelers need?

Home
• Building materials
• Workers
• Foremen
• Quality control
• Supply chain

Bone
• Calcium and Phosphorus
• Osteoblasts and Osteoclasts
• Hormones (PTH, Vitamin D)
• QC: Muscle (bone muscle unit)
• Diet (Vitamin D, Calcium in diet)
Osteoclasts and Osteoblasts

Bricks made of Calcium and Phosphorus
How do glucocorticoids affect bones?

- Weaken/Disable Osteoblasts
Where do bone breaks occur?

- **Long Bones**
  - 20-60% of boys

- **Vertebrae**
  - 30-50% of boys
2017 CDC Guidelines on Treatment of Bone Disease in Children with DMD on Corticosteroids (Preview)
CDC Guidelines on Monitoring Bone Health in Children with DMD on Corticosteroids

• Yearly:
  Diet history
  Vitamin D level (> 30 ng/dL)
  DXA Scan to assess bone density (BMD)
  (I don’t do yearly)

• Every 1-2 years:
  X-ray of Spine regardless of back pain
Measuring Bone Density/Strength

DXA: Dual Energy x-Ray Absorptiometry

- Less radiation than a airplane trip
- Must be positioned flat on back
- Lie still for short periods of time X 30 minutes
- No zippers/snaps on clothes
- Need to have done in specialized centers with software specific to children
- A Score is assigned for the whole body, spine, hip, head of femur
2017 CDC Guidelines

• If pathologic long bone fracture occurs:
  – Get DXA and treat with bisphosphonate intravenously until “clinically stable” then continue on ‘maintenance’ dose
    • Pamidronate infusion once every 3 month X 2 years
    • then twice a year
2017 CDC Guidelines

- If back pain + vertebral fracture(s) OR high grade fracture without pain:
  - Get DXA and treat with bisphosphonate intravenously until back pain improved and reshaping of vertebrae have occurred
    - Pamidronate infusion once every 3 month X 2 years
    - then twice a year
Puberty in Duchenne MD

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Typical Puberty in Males

Typical Order of Events
1. Increase in testicular size
2. Hair Development (Adrenarche)
3. Peak height velocity
4. Voice changes, facial hair

Age

Start

End

Height spurt

Testicular volume (cc)

Penis (Tanner Stage)

Pubic Hair (Tanner Stage)
Puberty in boys on corticosteroids

- **Height spurt**
  - Start
  - End

- **Testicular volume (cc)**
  - 4-6
  - 6-10
  - 10-15
  - 15-25

- **Penis (Tanner Stage)**
  - 2
  - 3
  - 4
  - 5

- **Pubic Hair (Tanner Stage)**
  - 11
  - 12
  - 13
  - 14
  - 15

Later, Longer
Delayed Puberty - Treatment

• “In an attempt to mimic normal pubertal development, testosterone replacement should be initiated at a low dose”
  • DMD Care Consideration, 2017

• When to start? After age 14 and when AM testosterone level < 75 mg/dL
• What dose? Start low, increase slowly
  – 50 mg IM monthly, increasing every 4-6 mos by 25 mg.
    Once reach 100 mg/month, switch to twice monthly
• When to stop? ???????