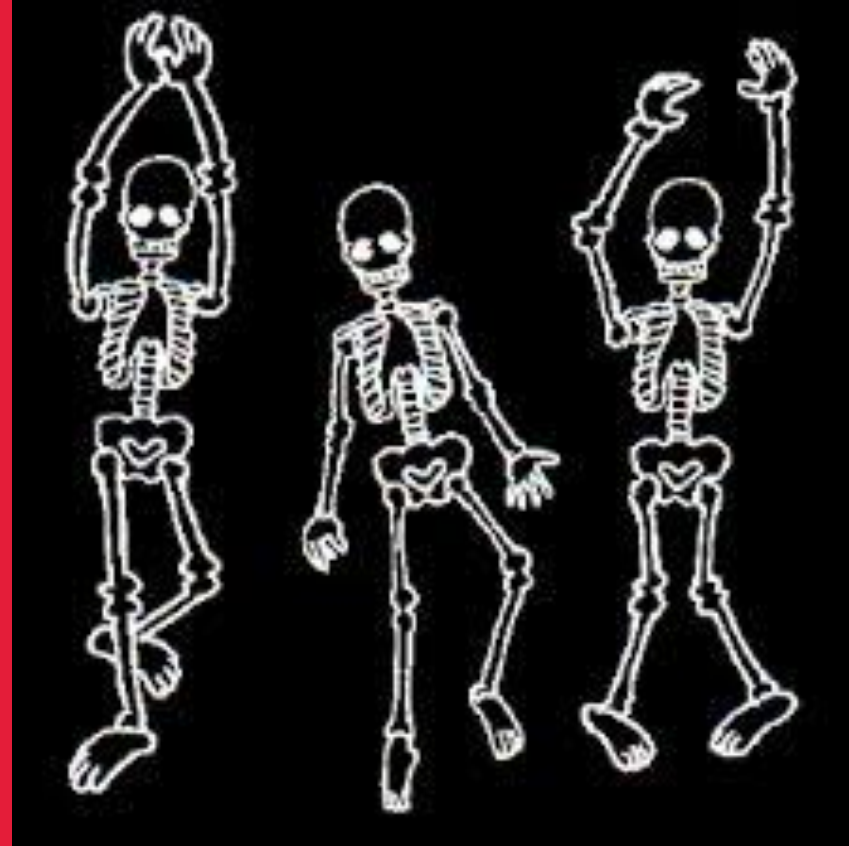


# Bones and Hormones



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**Parent Project Muscular Dystrophy**  
JOIN THE FIGHT.  
END DUCHENNE.

# 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

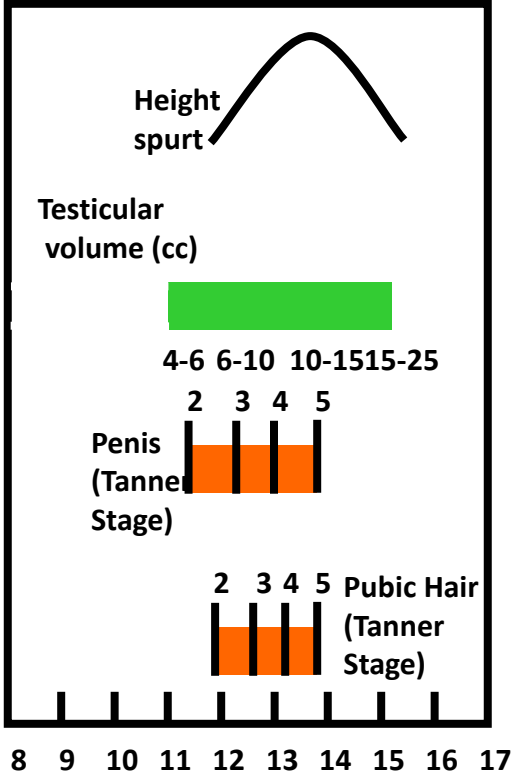
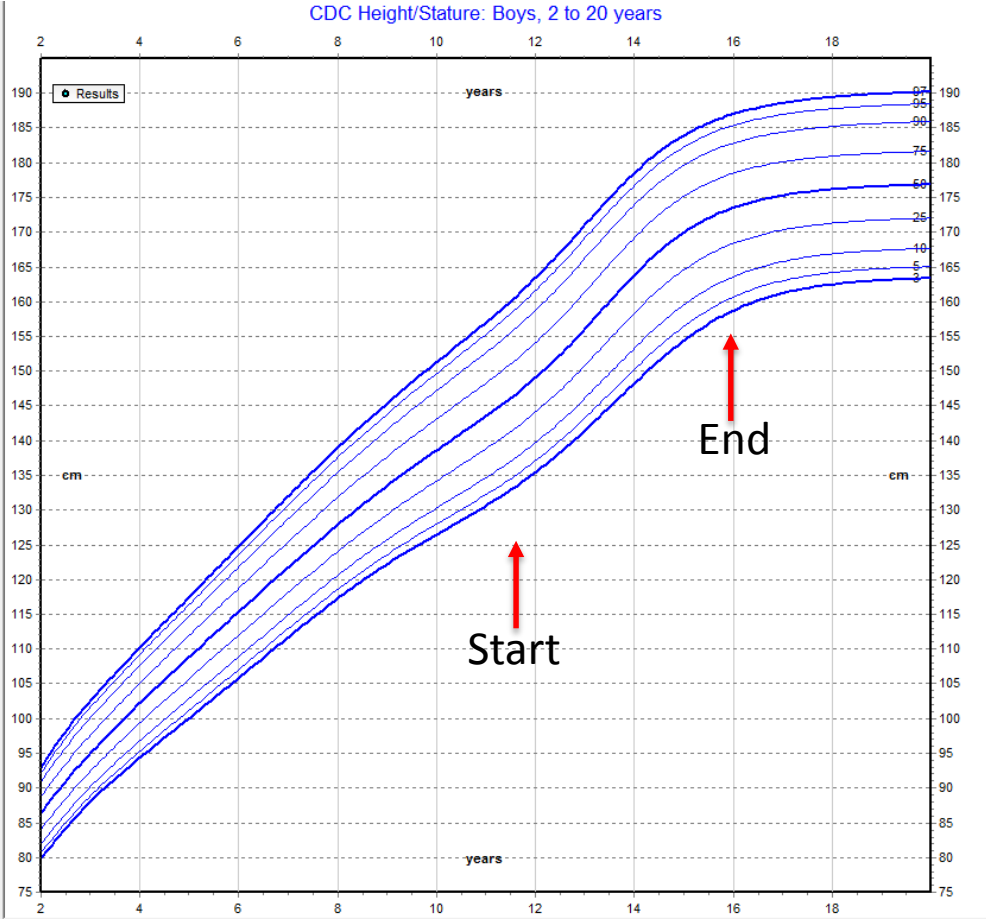


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# Puberty

## Typical Puberty in Males

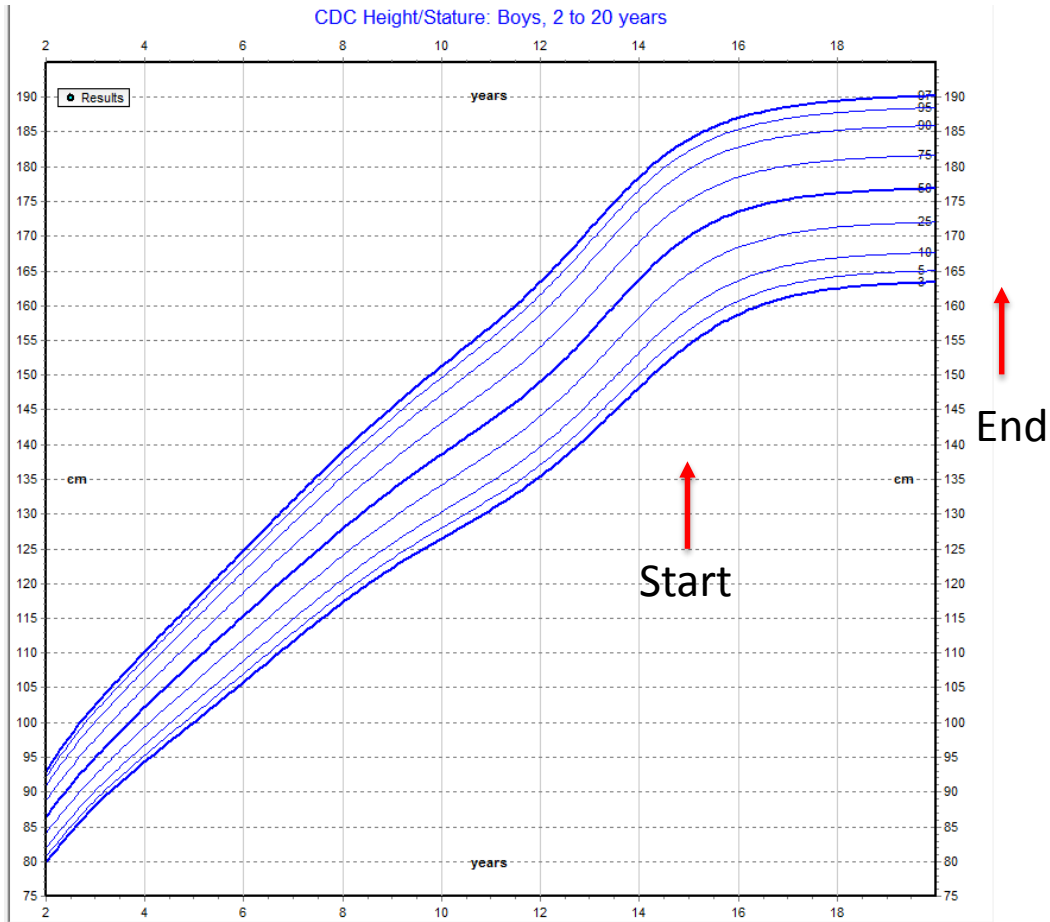


### Typical Order of Events

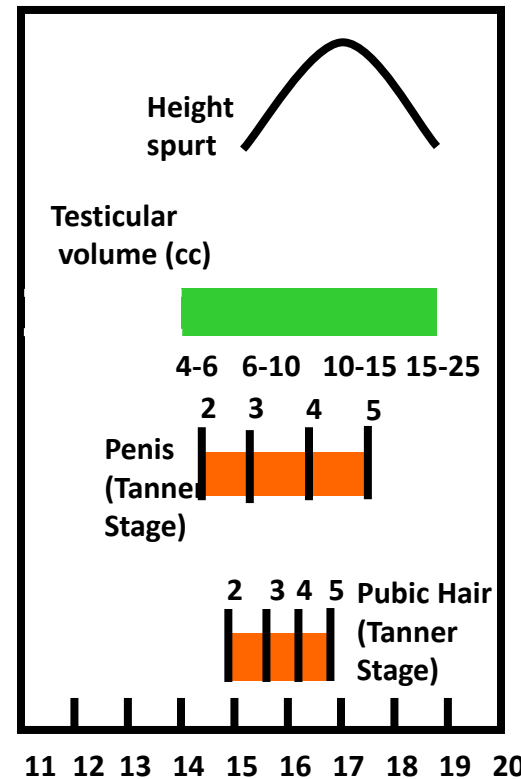
1. Increase in testicular size
2. Hair Development (Adrenarche)
3. Peak height velocity
4. Voice changes, facial hair

Age

# Puberty in boys on corticosteroids



## Puberty in DMD Males



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? ???????