

# Puberty and Growth in Duchenne MD



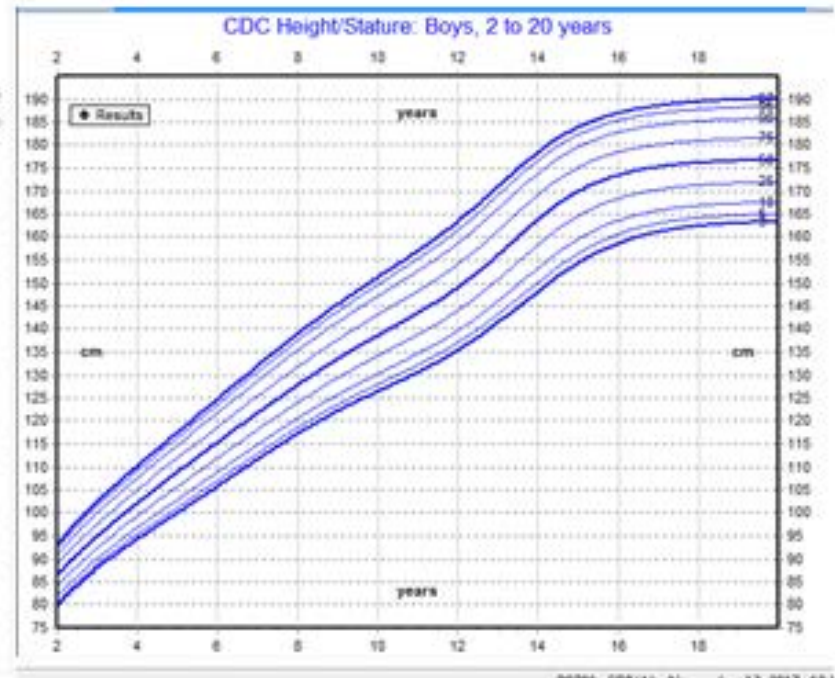
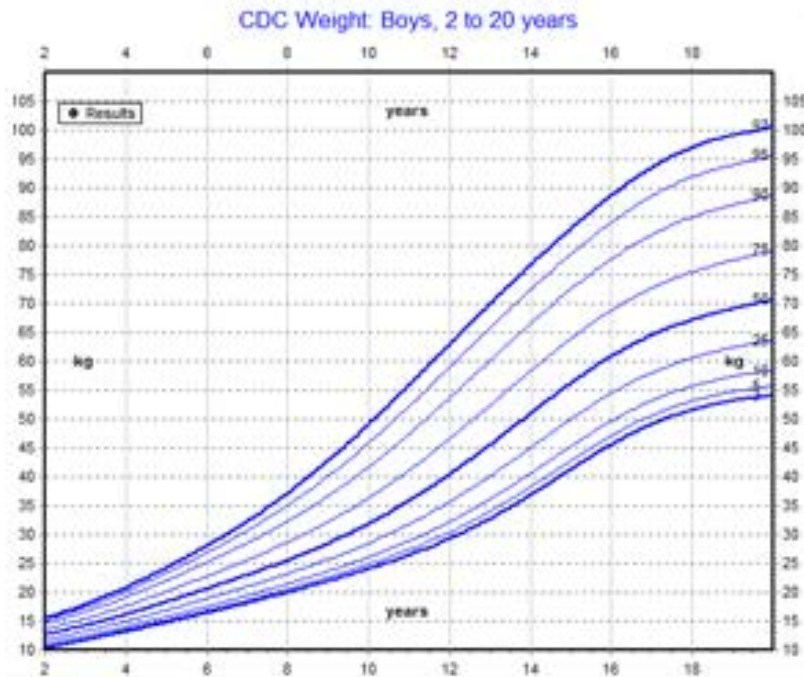
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**Parent** JOIN THE FIGHT.  
END DUCHENNE.  
**Project**  
**Muscular**  
**Dystrophy**

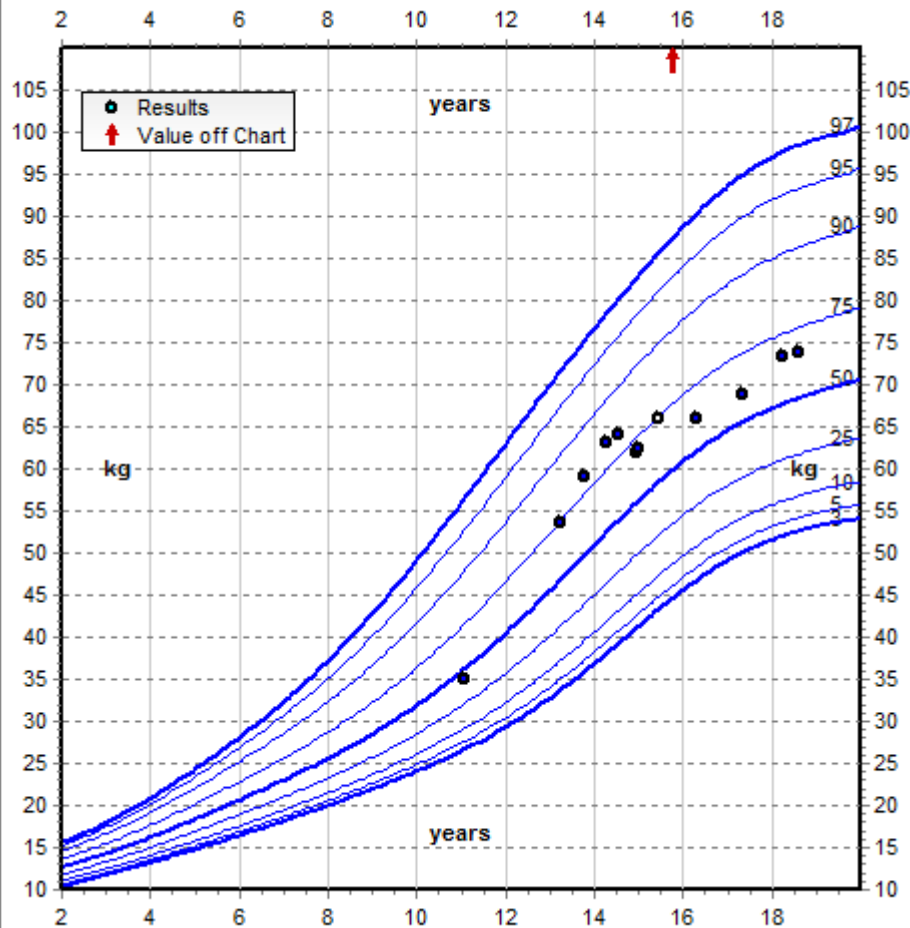
# Boys' Growth Charts

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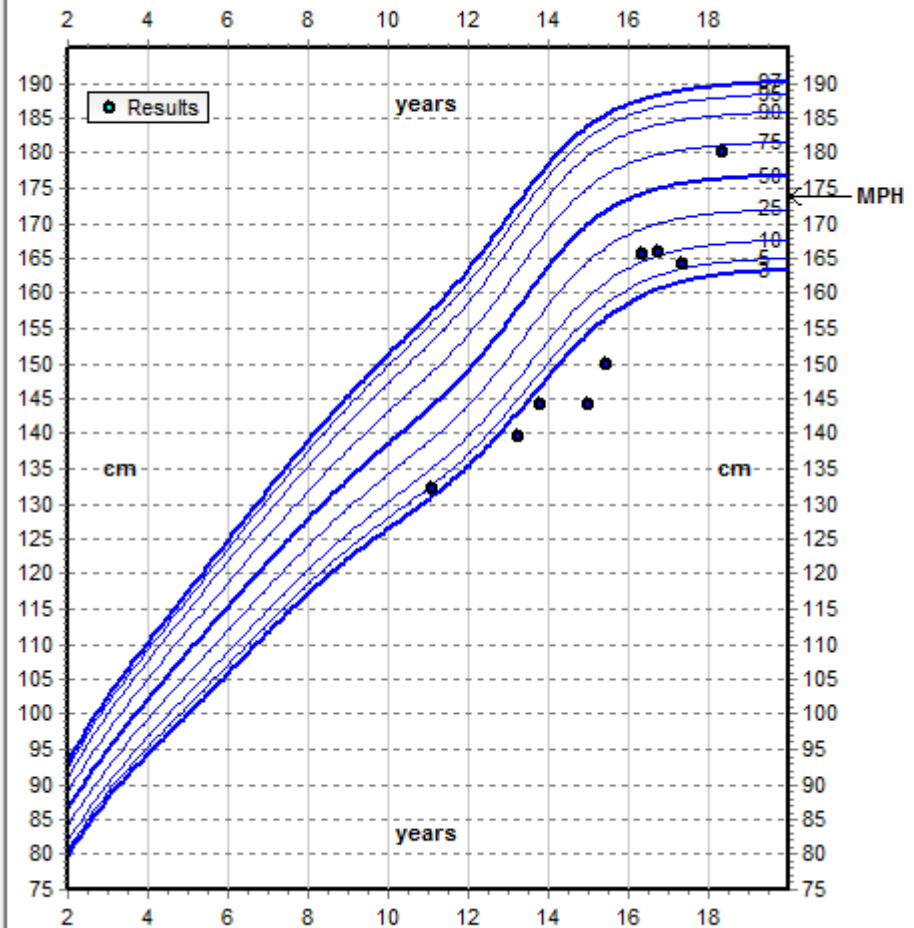


# Representative Growth of Boys with DMD non ambulatory – no steroids

CDC Weight: Boys, 2 to 20 years



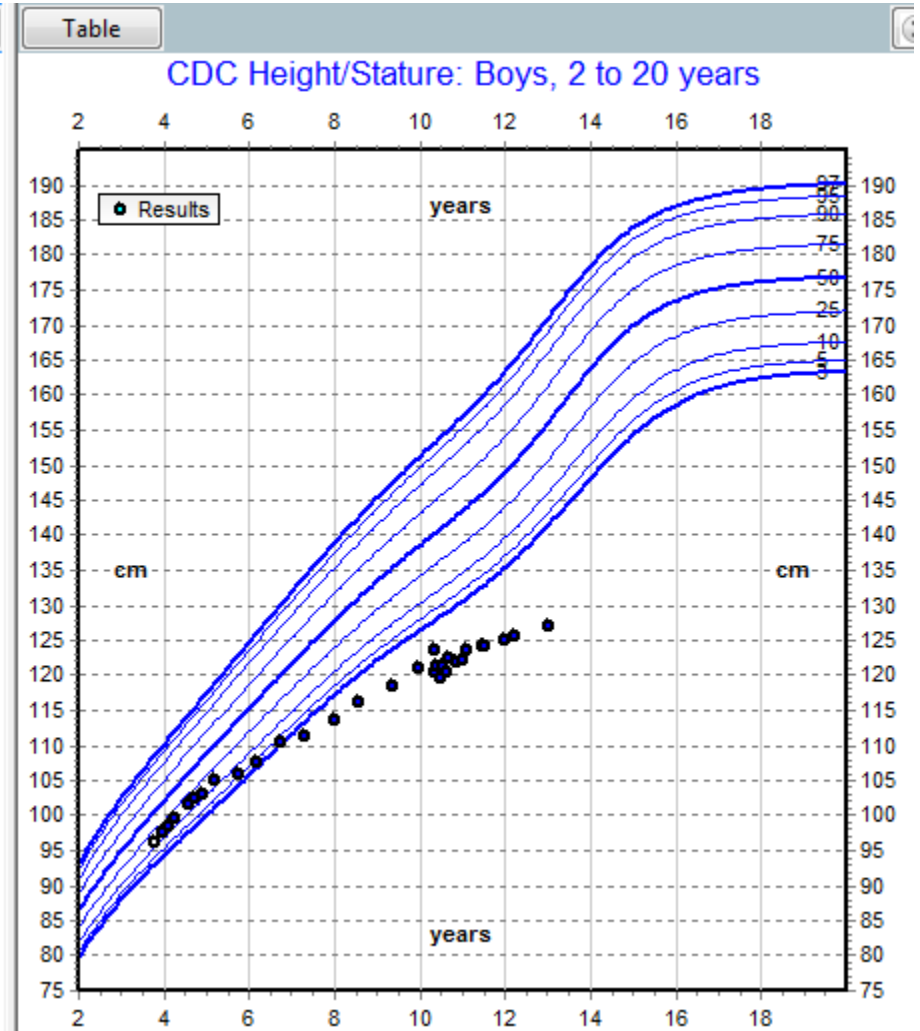
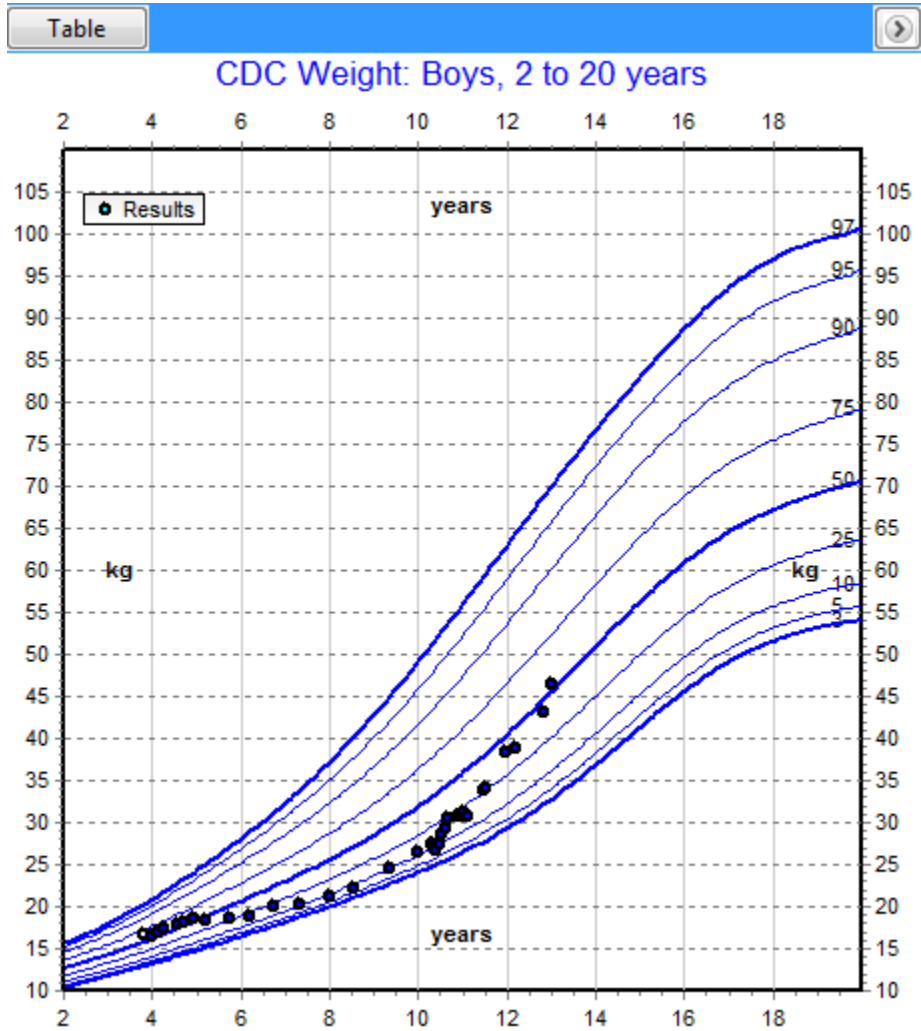
CDC Height/Stature: Boys, 2 to 20 years



# How to measure height in a nonambulatory child?

- Arm span?
- Forearm (Ulnar) length?
- Tibia length?
- Segmentally measured (joint to joint)?
  
- No method has been validated in nonambulatory children
- Pick a method and stick to it! (I like arm span)

# Representative Growth of Boys with DMD, ambulatory , on steroids



# Why is growth attenuated and obesity so common?

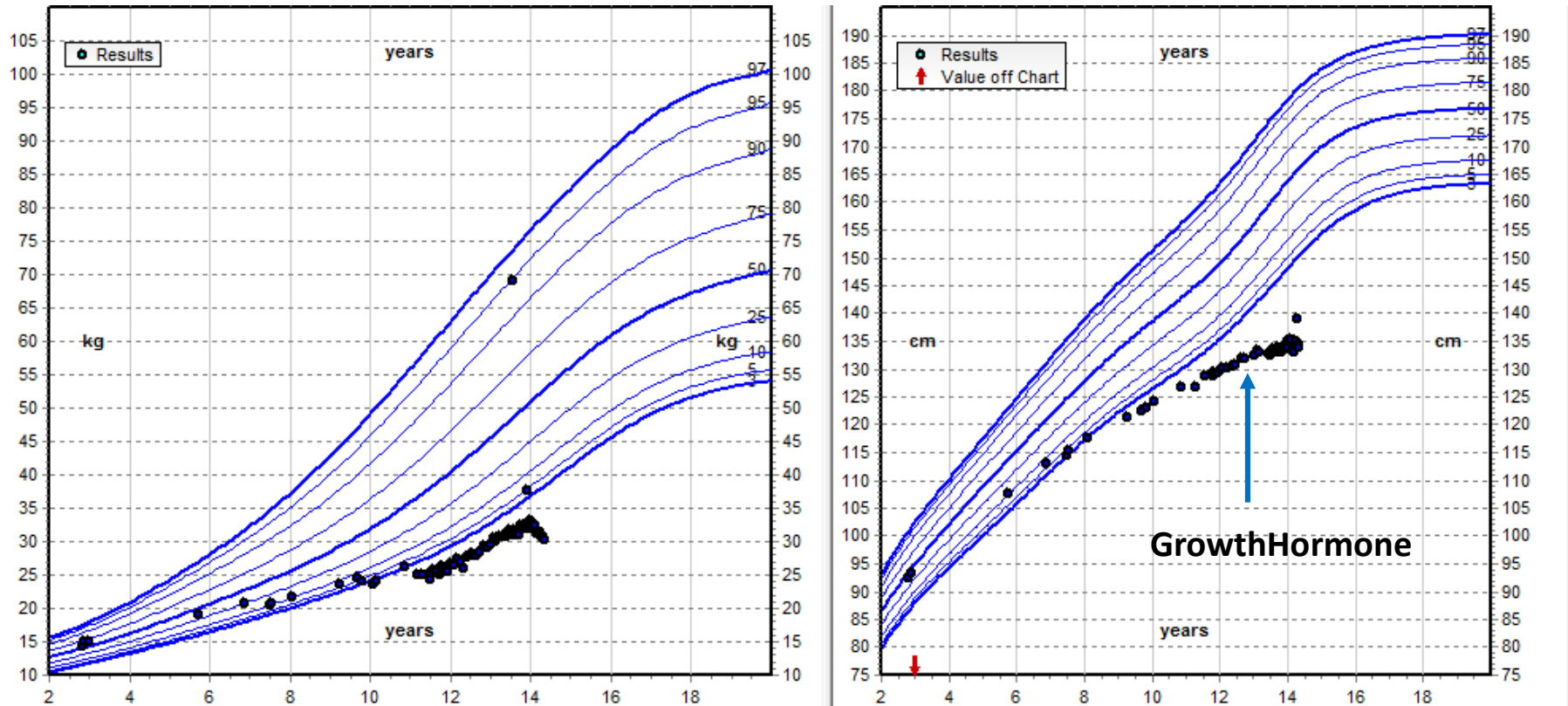
- Corticosteroids (Prednisone, Deflazacort)



- Lower metabolic rate and calorie use leading to weight gain/obesity

# Does Growth Hormone Help?

“Until more evidence is available, the routine use of rhGH to treat DMD-related growth failure is not recommended. Instead, the decision to treat with rhGH should be based on a thorough discussion about the potential risks and benefits of the therapy”



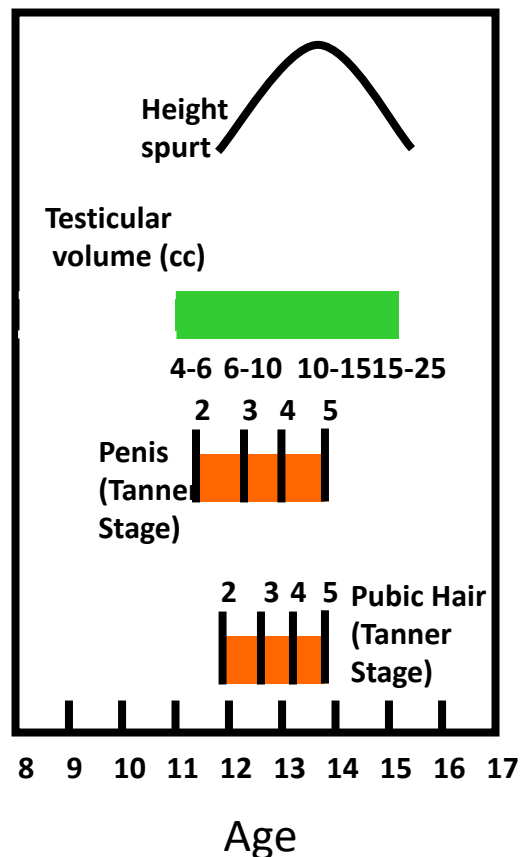
# What about growth hormone and muscle strength?

- No trials have been performed in boys with DMD
- In children, no data on GH effect on muscle strength
- Cost: 30000-50000/year



# Delayed Puberty

## Typical Puberty in Males

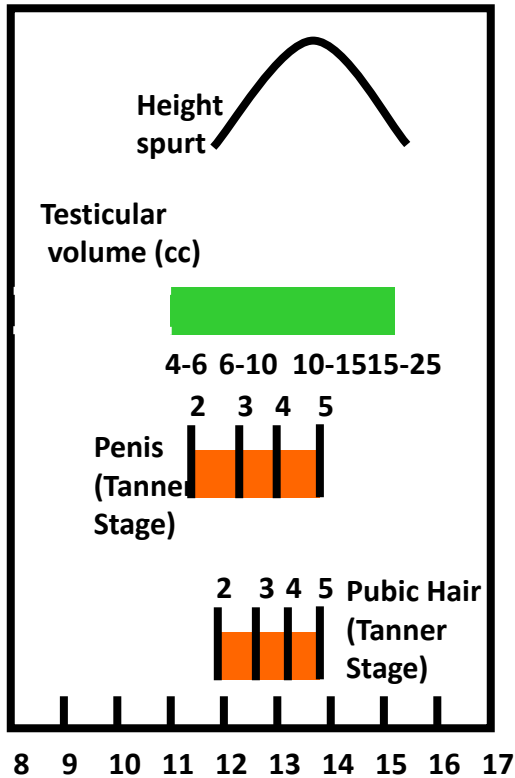


### Typical Order of Events

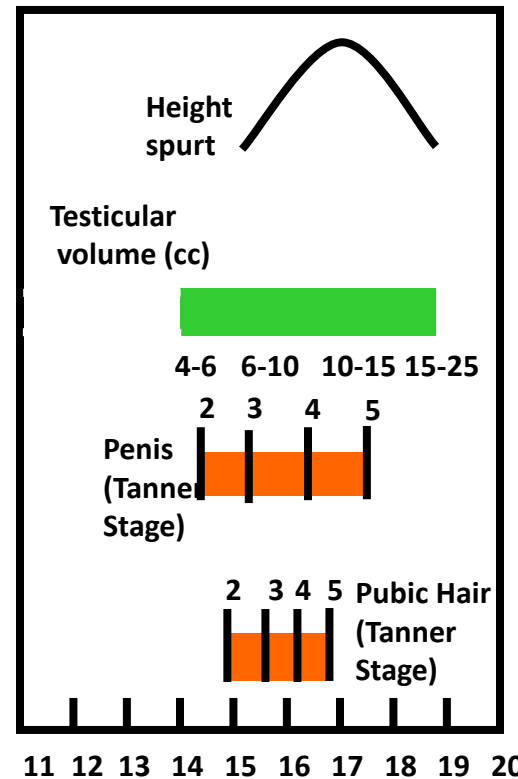
1. Increase in testicular size
2. Hair Development (Adrenarche)
3. Peak height velocity
4. Voice changes, facial hair

# Puberty in boys vs boys on corticosteroids

## Puberty in Males

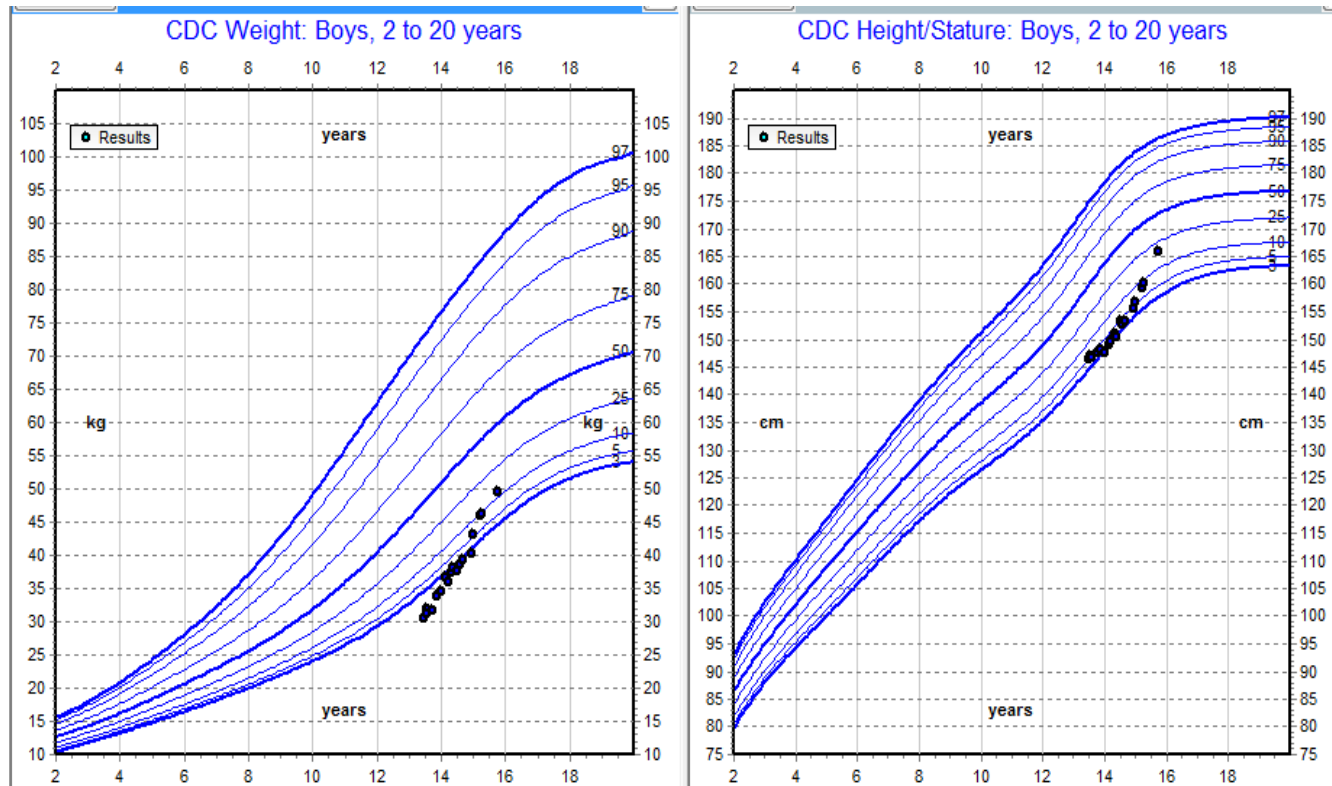


## Puberty in DMD Males

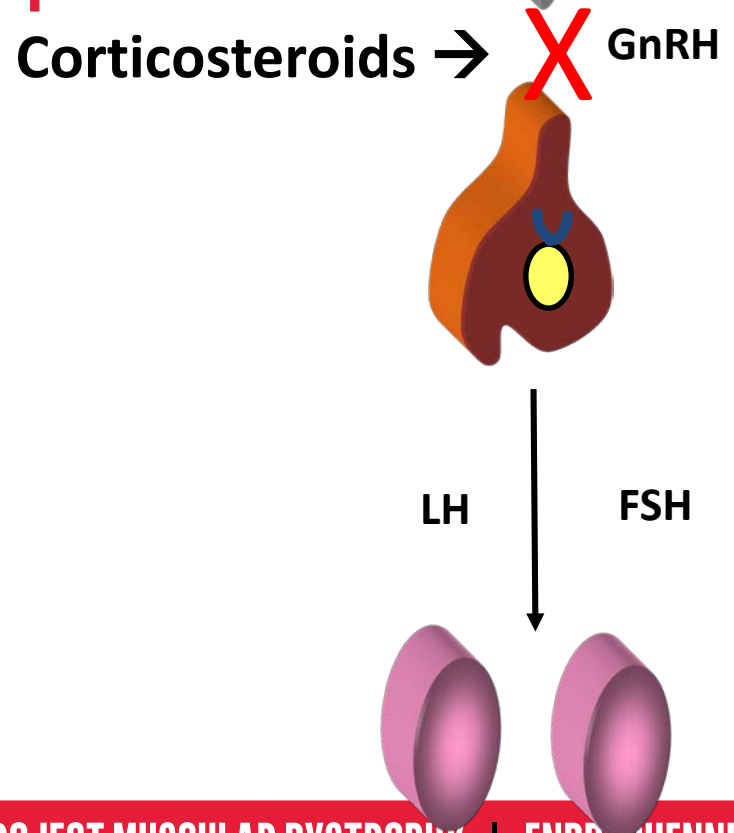
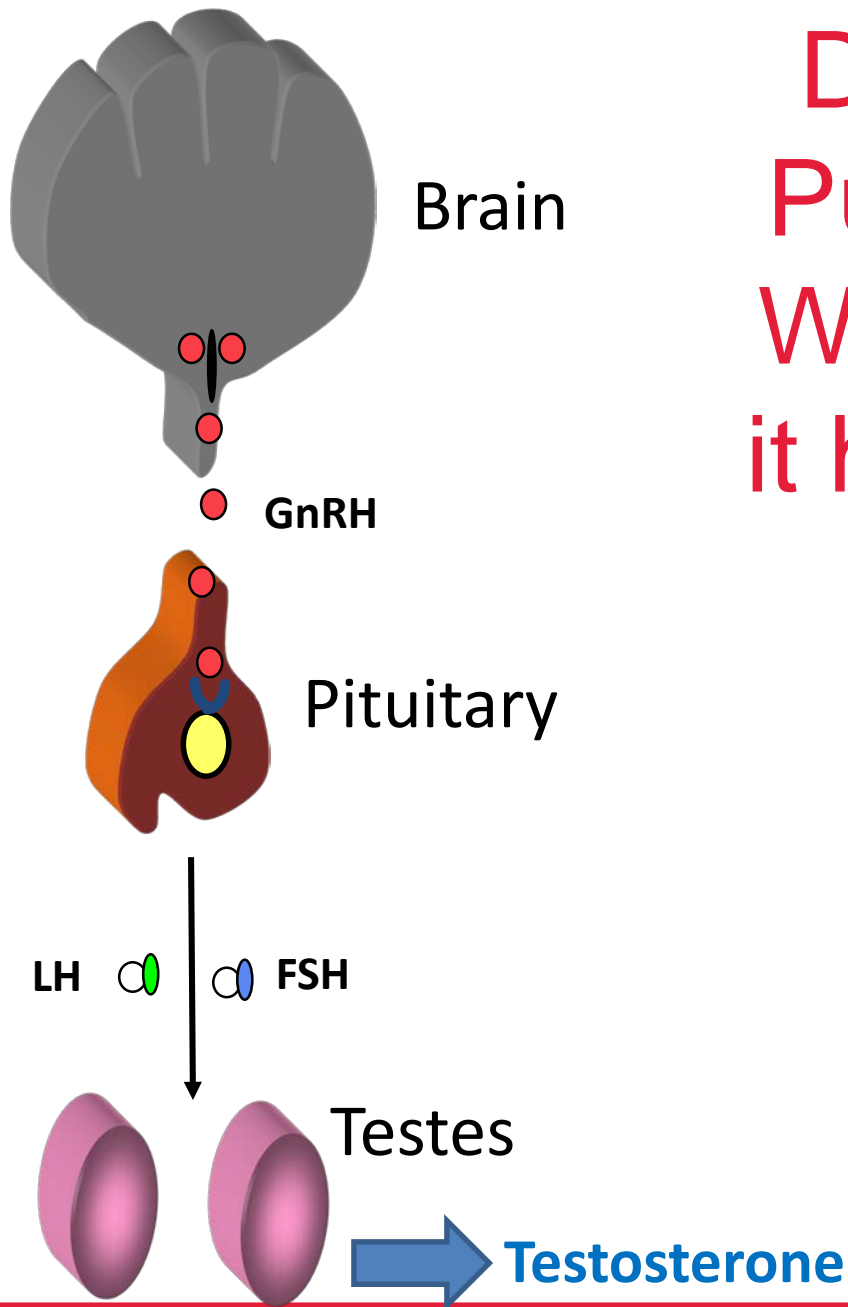


Later,  
Longer

# Delayed Puberty – Growth Chart (ambulatory boy without DMD)



# Delayed Puberty – Why does it happen?



# Evaluation for Puberty

- Pubic Hair may not indicate puberty!
- First sign is testicular enlargement to 4 mL or greater
  - Must feel comfortable assessing testicle size
  - Must have time/patient willingness to maneuver wheelchair and position in chair for exam
- Early puberty is a night-time process!
  - Only morning levels (< 10AM) of testosterone are helpful
  - Use sensitive testosterone (by LC/MS assay)

# 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? ???????

# Benefits of Testosterone

- Positive effects on bone mineral density
- Positive effects on muscle mass/strength
- Positive effects on psyche
  
- Drawbacks: Monthly injections, barriers to obtain testosterone.

# Questions?

