

# Management of GI Issues in Duchenne

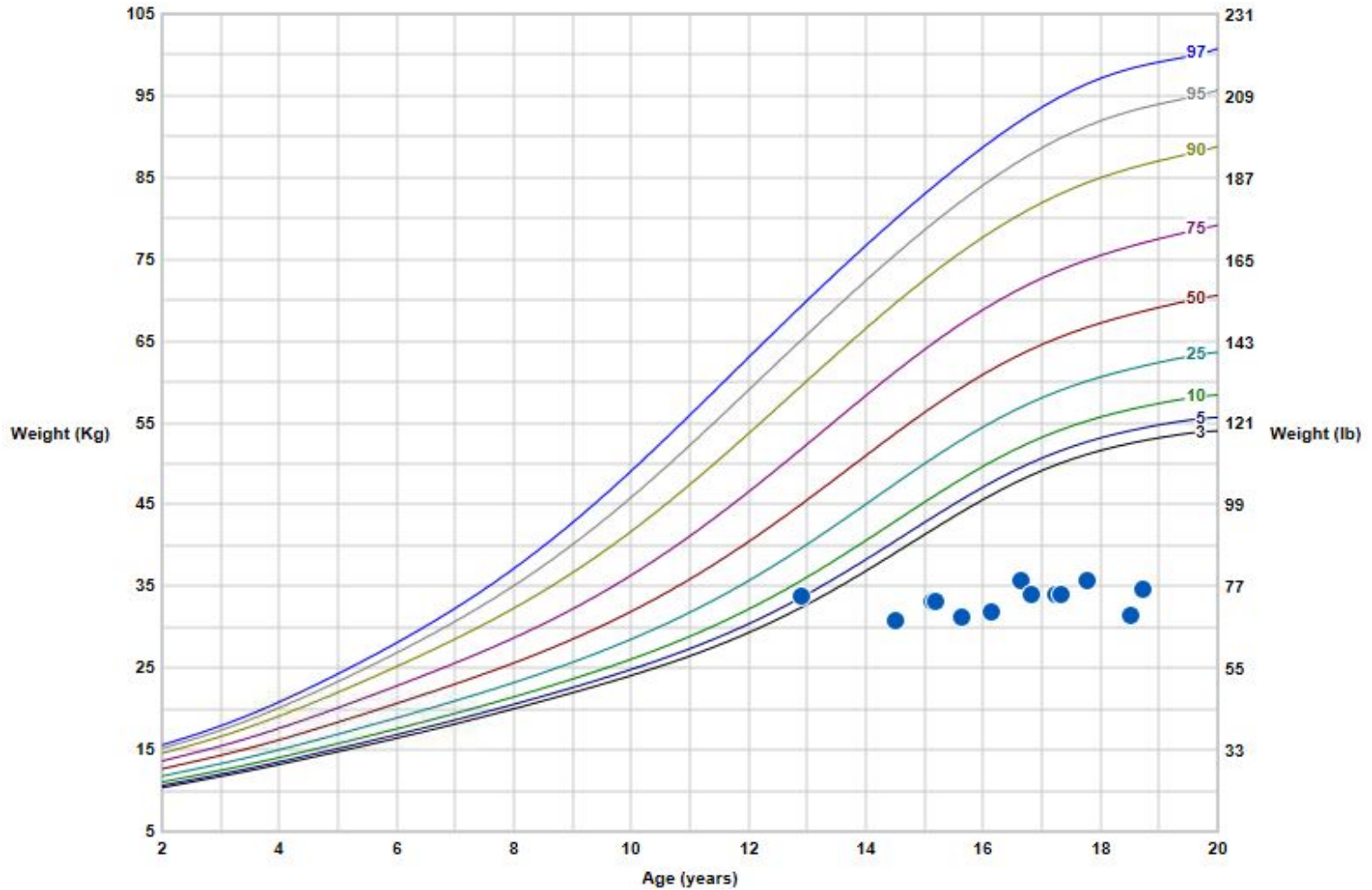
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**Parent** JOIN THE FIGHT.  
**Project** END DUCHENNE.  
**Muscular**  
**Dystrophy**

# Objectives

- Understand Need to Evaluate Nutrition Status Every Visit
- Learn Factors That Affect Eating
- Time for Surgical G-tube

# Growth Chart



# What Impairs Eating?

- Difficulty Feeding Self
- Impaired Swallowing
- Altered Gastric Function
- Constipation

# Ability to Feed Self

- Decreased Use of Arms/Hands
- Fed by Others
- Calorie Count
  - Recall
  - Prospective

# Problem Swallowing / Dysphagia

- Oral Pharyngeal Phase
  - Physiological Changes
  - Reduced Endurance
- Esophageal Phase
  - Feel Food Getting Stuck
  - Chest Pain

# Oral Pharyngeal Evaluation

- Validated Survey: Sydney Swallow Questionnaire (SSQ)
  - Archer SK, Garrod R, Hart N, Miller S. Dysphagia in Duchenne muscular dystrophy assessed by validated questionnaire. *Int J Lang Commun Disord*. 2013;**48**(2):240–6.
- Speech Therapy Evaluation
- Video Swallow Study (V.S.S)
- Fiberoptic Endoscopic Evaluation of Swallowing (F.E.E.S.)
- G-tube Evaluation if Abnormal

# Esophageal Dysphagia Treatment

- Start on Acid Reflux Medication
- Proton Pump Inhibitor
  - Omeprazole
  - Lansoprazole
  - Pantoprazole
  - Esomeprazole



# No Response to PPI

- UGI
- Consider Eosinophilic Esophagitis
  - Any Allergies
- GI Referral
  - Upper endoscopy
  - Empiric therapy with swallowed steroids
  - Evaluate Gastric Function

# Impaired Gastric Function

- Reports of Gastroparesis
  - Bensen ES et al. Acute gastric dilatation in Duchenne muscular dystrophy: a case report and review of the literature. Archives of physical medicine and rehabilitation. 1996;77:512-4
- Progressive Impaired Gastric Function
  - Borrelli O, et al. Evolution of gastric electrical features and gastric emptying in children with Duchenne and Becker muscular dystrophy. Am J Gastroenterol. 2005;100:695-702.

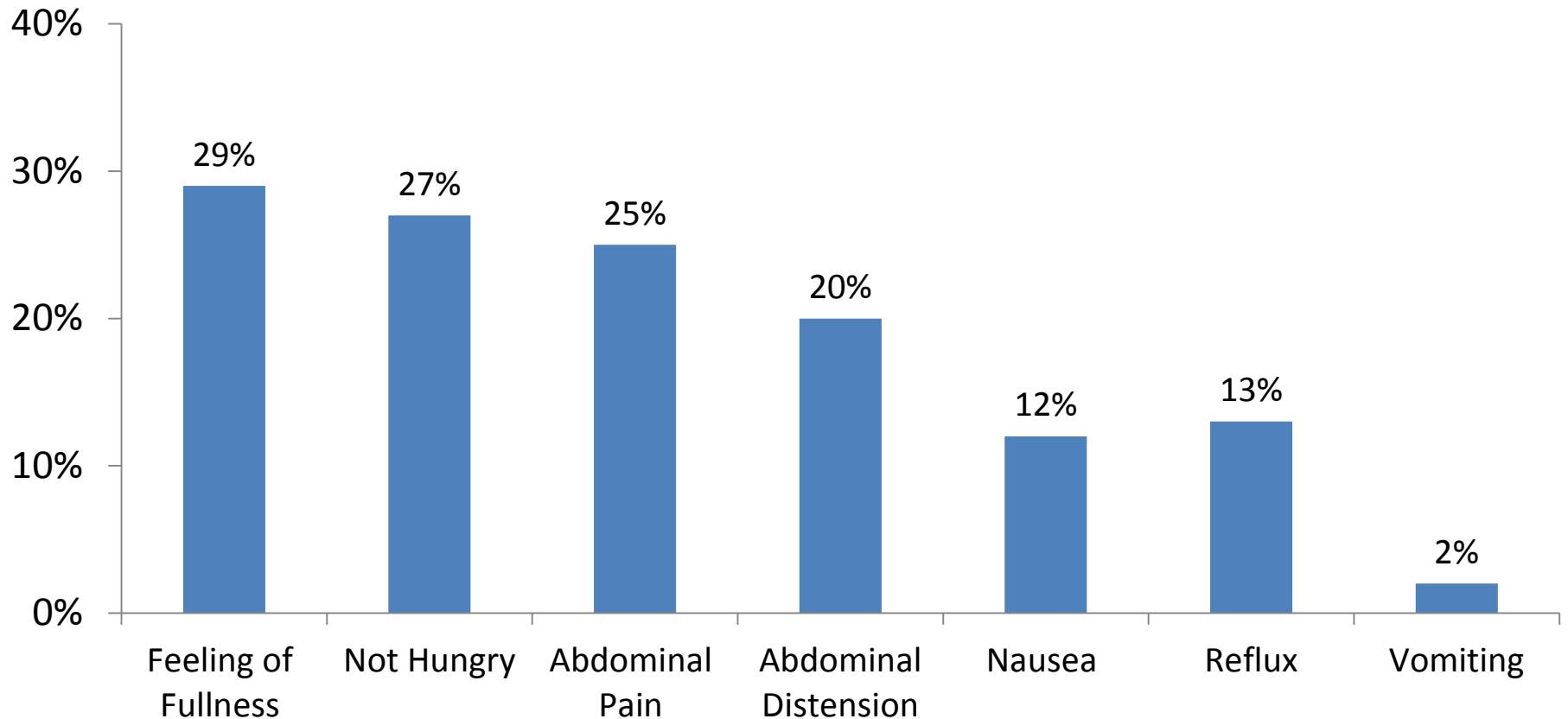
# Treatment of Reflux / Impaired Gastric Function

- PPI
- Erythromycin
- Bethanocol
- Buspirone

# Constipation

- Overlooked Cause for Reducing Appetite
- Common Cause of Other GI Symptoms

# PPMD Online GI Survey (N=338)



# Constipation

- Under Reported and Under Treated
  - Kraus et al. Constipation in Duchenne Muscular Dystrophy: Prevalence, Diagnosis, and Treatment. J Pediatr 2016;171:183-8
  - Almost half of 120 surveyed met criteria
- Unrecognized










R<sup>M</sup>C

STRETCH

# Criteria for Diagnosing Constipation

- Consistency
  - “Hard Stool or Little Balls or Lumpy Bumpy?”

**Bristol stool chart**

|        |   |  |
|--------|---|--|
| Type 1 |    | Separate hard lumps, like nuts<br>(hard to pass)   |
| Type 2 |    | Sausage-shaped but lumpy                           |
| Type 3 |    | Like a sausage but with cracks on<br>its surface   |
| Type 4 |    | Like a sausage or snake, smooth<br>and soft        |
| Type 5 |   | Soft blobs with clear-cut edges<br>(passed easily) |
| Type 6 |  | Fluffy pieces with ragged edges, a<br>mushy stool  |
| Type 7 |  | Watery, no solid pieces,<br><b>Entirely liquid</b> |

Constipated



# Criteria for Diagnosing Constipation

- Consistency
- Size
  - “Ever pass anything that amazes you?”
  - “How often clog toilet?”

# Criteria for Diagnosing Constipation

- Consistency
- Size
- Effort/Pain
  - “Hurt when stool comes out?”
  - “Strain while holding breath/face change color?”
  - “How long to pass stool/stays in bathroom?”

# Criteria for Diagnosing Constipation

- Consistency
- Size
- Effort/Pain
- Contenance
  - “Stool Accidents?”
  - “Smears in Underwear?”

# Criteria for Diagnosing Constipation

- Consistency
- Size
- Effort/Pain
- Contenance
- Frequency ( $\geq 4$  days)

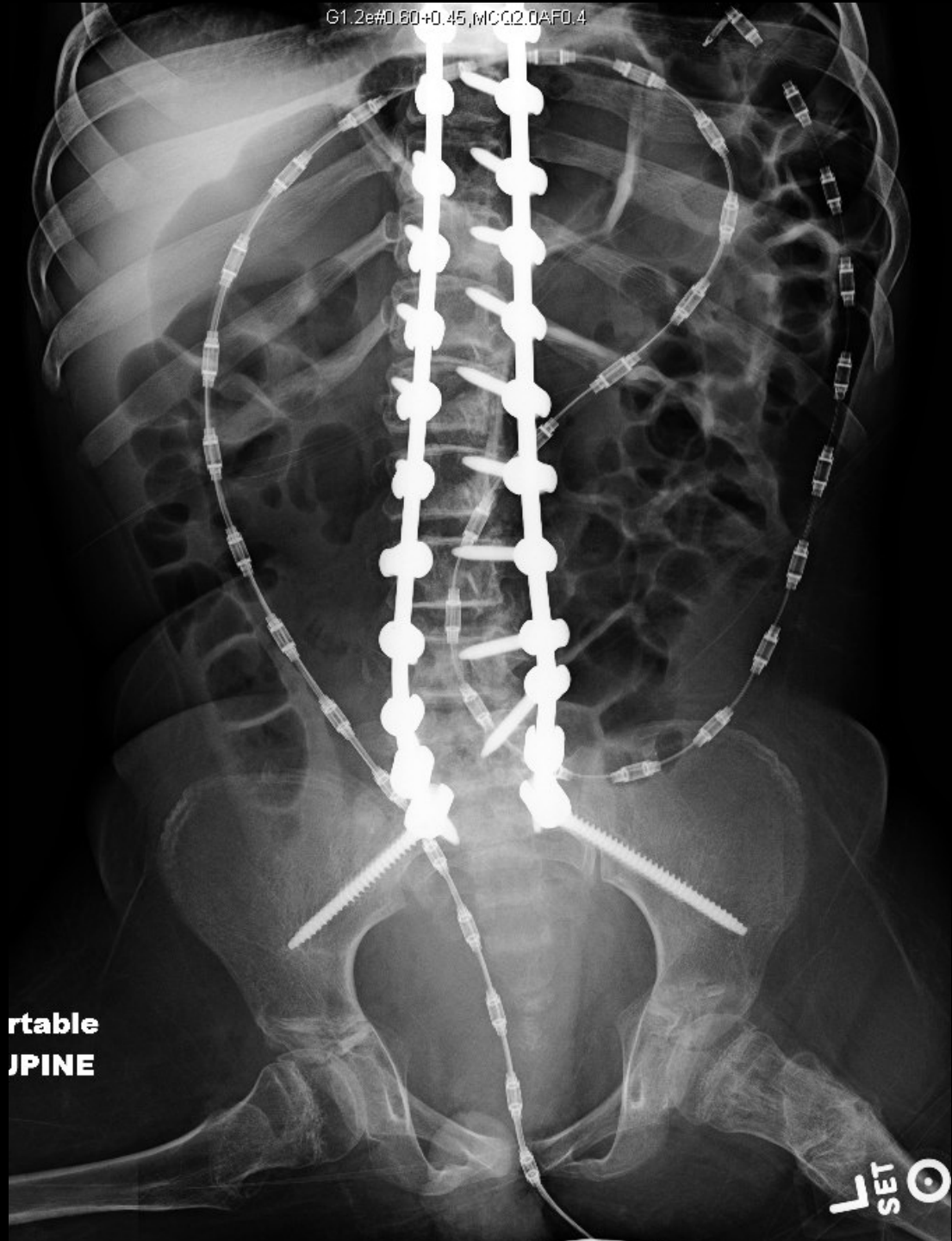
# Treatment of Constipation

- Standard Therapies
  - Polyethylene Glycol 3350
  - Lactulose
  - Colace
- Abdominal x-ray (Identify Severe Constipation)
- GI Referral
  - If no improvement
  - Severe Constipation

# Colonic Motility in DMD?

- **Evidence for Colonic Dysmotility**
  - Leon SH, et al. Chronic intestinal pseudoobstruction as a complication of Duchenne's muscular dystrophy. *Gastroenterology*. 1986;90:455-9.
  - Gottrand F, et al. Segmental colonic transit time in Duchenne muscular dystrophy. *Arch Dis Child*. 1991;66:1262.
  - Lo Cascio CM, et al. Gastrointestinal Dysfunction in Patients with Duchenne Muscular Dystrophy. *PLoS One*. 2016;11:e016377
- **Evidence against Colonic Dysmotility**
  - Kraus D, Wong B, Hu S, Kaul A. Gut Transit in Duchenne Muscular Dystrophy Is Not Impaired: A Study Utilizing Wireless Motility Capsules. *J Pediatr*. 2017

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# Colonic Dysmotility in DMD?

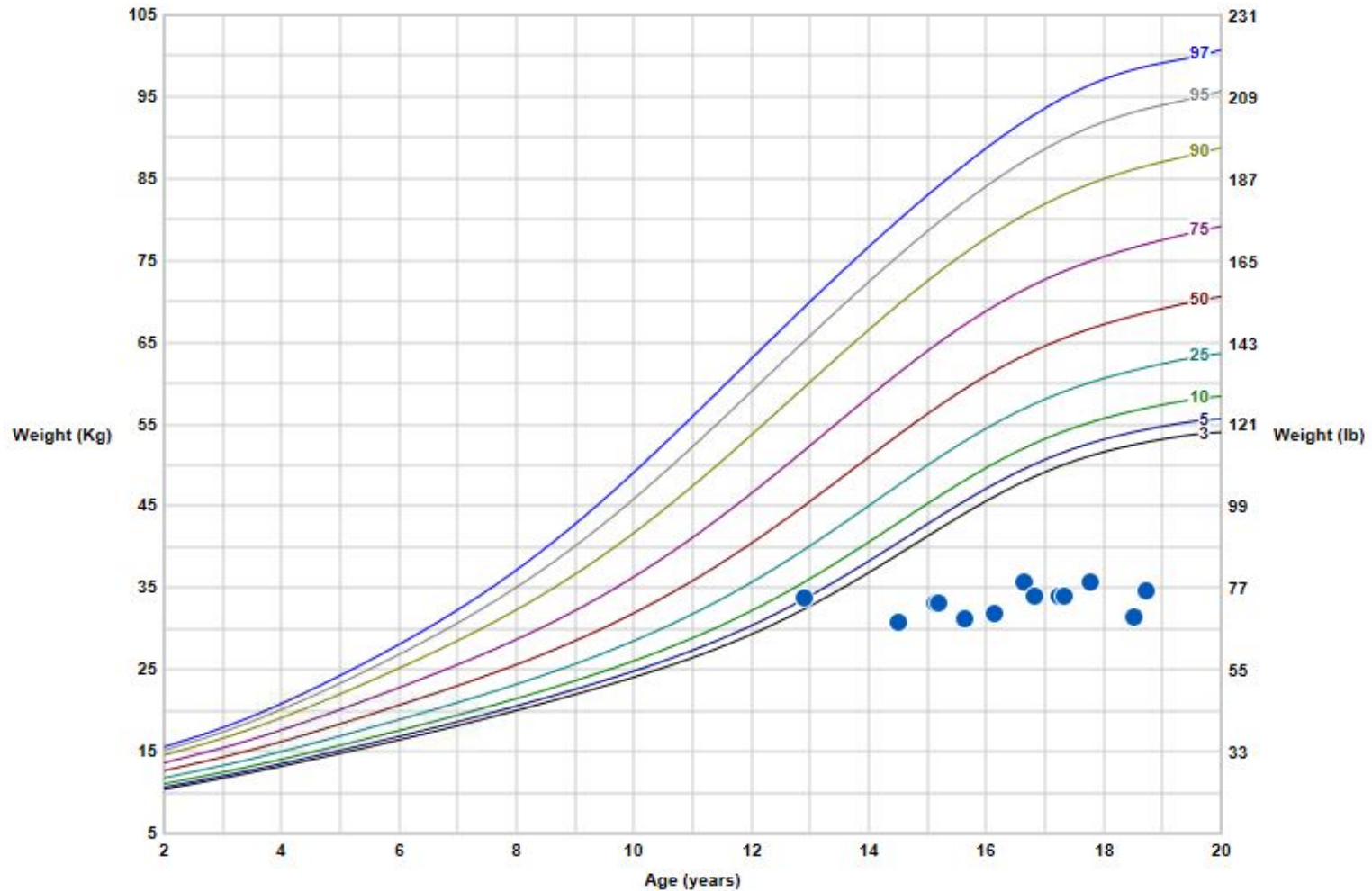
- Probably Depends on Gene Variability
  - Difference in Mutations
  - Other Genetic Variability
- Medical Therapy Same Regardless
- Motility Studies
  - If fail medical therapy
  - Clarify Surgical Options



# Treatment of Constipation

- Reluctance: Fear of BM in public
- Explain complications
  - Impaction
  - Impairs respiratory effort
  - Affects sleep
  - Loss of appetite
  - Loss of function (complete incontinence)

# Effective Treatment of Constipation Can Increase Appetite



# Indications for Tube Feeds

- Inadequate Caloric Intake
- Oral Pharyngeal Dysphagia
- Malnutrition

# Work-Up for G-tube

- Evaluate Swallow Function
- Gastric Emptying Study
- UGI (evaluate anatomy)
- If possible, trial of NG tube feeds

# Reluctance

- Quality of Life Concerns
- Unspoken Issues
  - Feel like failure (Parents)
  - Already feel different
  - Fear of surgeries
- “Do not want to give up eating”

# Benefit of Surgical G-tube

- Improve Nutritional Status
- Maintain Fluid / Hydration (even when sick)
- Improve Quality of Life
  - Increased function
  - Improved cognitive function

# Summary

- Evaluate Nutrition
- Evaluate for Factors Impair Eating
  - Constipation
- Consider G-tube Early