Management of GI Issues in Duchenne

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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)

• 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

• Progressive Impaired Gastric Function
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)

- Feeling of Fullness: 29%
- Not Hungry: 27%
- Abdominal Pain: 25%
- Abdominal Distension: 20%
- Nausea: 12%
- Reflux: 13%
- Vomiting: 2%
Constipation

• Under Reported and Under Treated

  – Almost half of 120 surveyed met criteria

• Unrecognized
Criteria for Diagnosing Constipation

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

<table>
<thead>
<tr>
<th>Bristol stool chart</th>
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<tbody>
<tr>
<td><strong>Type 1</strong></td>
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<tr>
<td><strong>Type 2</strong></td>
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<tr>
<td><strong>Type 3</strong></td>
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<td><strong>Type 4</strong></td>
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<td><strong>Type 5</strong></td>
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<td><strong>Type 6</strong></td>
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<tr>
<td><strong>Type 7</strong></td>
</tr>
</tbody>
</table>

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

• Continence
  – “Stool Accidents?”
  – “Smears in Underwear?”
Criteria for Diagnosing Constipation

• Consistency

• Size

• Effort/Pain

• Continence

• Frequency (≥ 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

• Evidence against Colonic Dysmotility
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