The management of major gastrointestinal findings in CDG

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Major gastrointestinal findings in CDG

- Feeding problems
- Gastroesophageal reflux
- Failure to thrive
- Protein loosing enteropathy
- Liver disease, fibrosis / cirrhosis
Feeding problems I
The generals

Baby falls asleep soon after starting feeds
  ? Insufficient sucking ➔ check positioning, exchange tits
  ➔ review by clinician

Baby has lots of gas / stomach discomfort
  ? Lactose intolerance ➔ switch to soy formula

Baby cries for hours after meal
  ? Colics ➔ Rocking chair, use cradle, baby massage
Baby vomits large amounts of feeds after meal
  ? Pyloric stenosis  ➔ x-ray

Baby seems to have slow weight gain
  ? Problems to absorb, digest feeds  ➔ blood, urine, stool tests

Baby seems to have little interest in food
  ? Developmental problem  ➔ examination by clinician
Feeding problems Management

- Adjustment of feeding routine
- Increasing calorie intake
- Exchange of milk feeds / food
- Exclusion diet
unhappy unsettled baby spitting up after feeds

and / or

only churning, gurgling, crying, wimping

Gastroesophageal reflux

Common problem!
Gastroesophageal reflux – Management I

Smaller, more frequent feedings
Holding baby in upright position for 20min after feedings

Small holes in tits

Thickening feed

Breastfeeding mums to avoid foods aggravating reflux:
role of thumb no-go for acidic, fatty or gassy
Gaviscon:
mixes with gastric content, stabilizes and thickens them

Ranitidine:
reducing stomach acid, helping existing inflammation in the esophagus

Surgical:
Failure to thrive

= inappropriate weight gain

• Inadequate nutrient intake

• Inadequate nutrition absorption or increased losses

• Increased requirements or ineffective utilization
= inappropriate weight gain

• Inadequate nutrient intake

• Inadequate nutrition absorption or increased losses

• Increased requirements or ineffective utilization
Failure to thrive

= inappropriate weight gain

• Inadequate nutrient intake
  feeding difficulties
  inadequate appetite
  inability to take in appropriate amounts
Failure to thrive

= inappropriate weight gain

• Inadequate nutrient intake

• Inadequate nutrition absorption or increased losses

• Increased requirements or ineffective utilization
Failure to thrive

= inappropriate weight gain

- Inadequate nutrition absorption or increased losses

  malabsorption
  gut disease
  abnormal gut
Failure to thrive

= inappropriate weight gain

• Inadequate nutrient intake

• Inadequate nutrition absorption or increased losses

• Increased requirements or ineffective utilization
Failure to thrive

= inappropriate weight gain

- Increased requirements or ineffective utilization

heart disease
hormonal imbalances
metabolic syndrome
Failure to thrive Management

Depending on the reason for inappropriate weight gain

- increasing daily calorie intake
- increasing caloric density of feeds
- spreading out feeds
- continuous vs bolus feeds
Gastrointestinal findings in CDG

• Protein loosing enteropathy
  = protein loss in gut

particularly in CDG-PMI (-Ib)–
but also seen in CDG-PMM (-Ia), CDG-Alg6 (Ic),
CDG-Alg8 (Ih) ...
Gastrointestinal findings in CDG

- Increased liver enzymes: often only intermitted

- Liver fibrosis / cirrhosis: scaring process, initially reversible
Major gastrointestinal findings in specific CDG subtypes

<table>
<thead>
<tr>
<th>CDG type</th>
<th>PMM-CDG (Ia)</th>
<th>PMI –CDG (Ib)</th>
<th>Alg8 - CDG (Ih)</th>
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<tbody>
<tr>
<td>Feeding difficulties</td>
<td>++ - +++</td>
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<tr>
<td>Failure to thrive</td>
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<td>Liver abnormalities</td>
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The team player

We are all together

- Nurses
- Pediatrician
- Specialist
- Feeding clinics
- Biochemist
- Radiologist
- Surgeon
- Parents
  Family
- The team player