Gut failure in neuro-disability: a paediatric centre experience

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Setting the scene

Many NICU survivors will grow up with a disability so profound that they are never likely to become independently mobile, to communicate effectively with others or to feed themselves.

Sullivan PB, 2008
Content

- Enteric nervous system
- Gut problems in neurologically impaired children
- Setting the scene
- Management of symptoms
Enteric Nervous System (ENS) or Intrinsic Nervous System

- 500 million neurons
- 5 times more than in the spinal cord
- Autonomous functions: coordination of reflexes
- It receives considerable innervation from the autonomic nervous system, but it can and does operate independently of the brain and the spinal cord
Enteric Nervous System

> 30 neurotransmitters: acetylcholine, dopamine and serotonin

Motor neurons → Sensory neurons

Intestinal muscles → Secretion of enzyme

Peristalsis

Gut has >90% of the body's serotonin and about 50% of the body's dopamine
Autonomic Nervous System
or Extrinsic Nervous System
Regulate unconscious actions

“Fight –or-flight”

“Rest-and-digest”
Central Nervous System disorders

Autonomic Nervous System modulates

Enteric Nervous System

Setting the scene
Main problems

• Foregut dysmotility
  High density of autonomic innervation
  1. Oral-motor impairment
  2. Oesophageal dysmotility
  3. GORD
  4. Antro-duodenal dismotility
  5. Retching
  6. Dumping Syndrome

• Constipation
Neurologically impaired children:

- **Motor Impairment**
- **Oro-motor dysfunction**
- **Communication Difficulties**
- **GROWTH FAILURE**
Prevalence and severity of feeding and nutritional problems in children with neurological impairment: Oxford Feeding Study


Comparison of type of neurological impairment

<table>
<thead>
<tr>
<th></th>
<th>Group A (n=271)</th>
<th>Group B (n=72)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total with CP</td>
<td>261</td>
<td>67</td>
</tr>
<tr>
<td>Spastic quadriplegia</td>
<td>147</td>
<td>30</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>57</td>
<td>32</td>
</tr>
<tr>
<td>Dyskinetic</td>
<td>27</td>
<td>0</td>
</tr>
<tr>
<td>Ataxic</td>
<td>18</td>
<td>3</td>
</tr>
<tr>
<td>Unclassifiable</td>
<td>12</td>
<td>2</td>
</tr>
</tbody>
</table>

*Group A*: children with feeding problems  
*Group B*: children with no feeding problems
Prevalence and severity of feeding and nutritional problems in children with neurological impairment: Oxford Feeding Study


| Table II: Feeding and nutritional problem responses in relationship to the degree of motor deficit for those in Group A |
|---|---|---|---|---|---|---|---|
| Feeding/nutritional problem | Total nr of participants | % | Severity of motor impairment | Pearson r | p value |
| Help with feeding needed | 238/268 | 89 | Mild: 27 | 85 | 126 | 188.9 | <0.001 |
| | | | Moderate: 12 | 38 | 90 | 62.68 | <0.001 |
| | | | Severe: 5 | 11 | 35 | 10.74 | <0.005 |
| Choking with food | 142/257 | 56 | Mild: 12 | 38 | 90 | 62.68 | <0.001 |
| | | | Moderate: 5 | 11 | 35 | 10.74 | <0.005 |
| | | | Severe: 5 | 11 | 35 | 10.74 | <0.005 |
| Feeding reported as stressful or unenjoyable by parent | 51/262 | 20 | Mild: 5 | 11 | 35 | 10.74 | <0.005 |
| | | | Moderate: 3 | 8 | 60 | 53.2 | <0.001 |
| Prolonged (³ 3h/d) feeding times | 71/258 | 28 | Mild: 3 | 8 | 60 | 53.2 | <0.001 |
| Parents considered child underweight | 93/240 | 38 | Mild: 6 | 25 | 62 | 26.87 | <0.001 |
| Child received caloric supplements | 23/271 | 8 | Mild: 1 | 2 | 20 | 15.64 | <0.001 |
| Gastrostomy feeding | 20/265 | 8 | Mild: 1 | 0 | 19 | 19.63 | <0.001 |
| Never had feeding and nutritional status assessed | 169/264 | 64 | Mild: 32 | 77 | 60 | 28.15 | <0.001 |
| Frequent vomiting | 55/249 | 22 | Mild: 1 | 12 | 42 | 33.3 | <0.001 |
| Bowels opened > every 3 days | 68/267 | 26 | Mild: 5 | 16 | 47 | 17.2 | >0.002 |
Impact of feeding problems on nutritional intake and growth: **Oxford Feeding Study II**

Gastrostomy for children with neurological impairment: when?

• When enteral feeding is required beyond the short-term period (> 6 w)

• When there are prolonged feeding times, inadequate weight gain AND/OR unsafe swallow

Benefits:

• Increase weight
• Improve overall health
• Decrease feeding times
• It has also demonstrated a significant, measurable improvement in the quality of life of carers

Sullivan, 2005
GOR in neurologically impaired children with gastrostomies

- It may be that **volume tolerance**, rather than acid, is the predominant problem.

- PPIs may **not be effective** in reducing symptoms due to large volumes of neutral-pH meals.

- Many of the symptoms/signs could be due to non-reflux causes:
  - pain
  - constipation
  - neurological issues
Nissen’s fundoplication
Surgical intervention for feeding and nutrition difficulties in cerebral palsy: a systematic review

What this paper adds:

• Few studies provide data to assess effectiveness of surgical interventions

• Gastrostomy consistently leads to weight gain and may improve other growth measures including height and skinfold thickness

• Adverse events after surgery are common

• The risk of intervention in relation to the risk of not treating is poorly understood
Many patients continue to have symptoms gastric dysrhythmias, unmasked by fundoplication.

Post- Nissen’s

- Patients with more severe reflux may have a more severe underlying motor dysfunction of the foregut
- Children with severe neurodevelopmental handicap with GORD and chronic, severe constipation

Maybe even more widespread panenteric dysfunction???
“In children with CNS disease, vomiting is related to gastric dysrhythmias and delayed gastric emptying, possibly due to activation of the emetic reflex, at least as often as to gastroesophageal reflux.

A significant proportion of these patients may thus have widespread foregut dysmotility in which gastroesophageal reflux (mainly caused by dysfunction of the lower oesophageal sphincter), gastric antral dysrhythmias (related to dysfunction of the gastric pacemaker), and delayed gastric emptying are associated.”
S., 10yrs

- Quadriplegic CP
- GORD and constipation
- Nissen’s fundoplication and PEG
- Orally fed + PEG supplements
- Thriving well
- Parents asked to go back to local teams
S., 11yrs

Re-refferred for:
reduced oral intake
poor feeding tolerance
bloatiness (air in the stomach)
discomfort
Setting the scene

• Primary condition with no gut involvement

• Successfully gastrostomy fed for many years

• Nissen’s fundoplication

• Slow deterioration

• Tolerance of clear fluid but not feeds

• Wind/gas = big problems!

• Gut irritability
What we know

- Not malabsorption
- Not mechanical obstruction
- Settles if stomach is aspirated
- Dioralyte is tolerated
“Off hand, I'd say you're suffering from an arrow through your head, but just to play it safe, I'm ordering a bunch of tests.”
• Delayed gastric emptying
• Dysmotility
Management
Management

Kolb’s Cycle of Experiential Learning

image by Karin Kirk

If you have no common sense please ask for assistance
Wind

Source:
Swallowed air >>>> fermentation

Reason for improvement on venting:
Reduction of gastric distension
Wind - management

Farrell valve bag

Gas release valve on Farrell bag. Only lets air and gas out, not liquid.

Check for: H Pylori
sugar malabsorption
Manage expectations!
Pain

Source:
*Organic*: Poor motility, pseudo-obstruction, constipation, inflammation, gallbladder or renal stones, scoliosis,…

*Functional*: visceral hyperalgesia, neurologic manifestations…

*Iatrogenic*: narcotic usage, drugs intolerance, tube-related complications…
Poor feeding tolerance

Management

• If urine output - no hurry

• Take careful history: other causes of pain or upset? recent changes? review medication and dosage?

• Intake: rate/frequency/volume

• Bowel motions: stool consistency/frequency
Medical Management

- Optimise reflux therapy (or feeding regimen!)
- Optimise colonic transit (constipation tx, probiotics, erythromycin)
- Pain control
- Consider changing drug preparations (less sucrose)
Nutritional Management

- Reintroduce feeds slowly / consider continuous feeds
- Consider change of feed (hydrolysed)
- Jejunal access
  - Double lumen gastro-jejunal tube
  - Jejunostomy

NB. Gastrostomy on free drainage

- Blended diet

- PN: Easy to start, hard to stop
Investigations

• Abdominal US or X-ray
• Contrast study
• Breath-Test or empirical trial with sucrose free diet

Buy time...
Give the gut a rest with ORS
S., 11yrs

Alginate susp: 5ml QDS
Alimemazine syroup: 15mg OD
Baclofen solution: 9mg QDS
Domperidone: 2mg/Kg TDS
Movicol: 2 sachets OD
Melatonin liquid: 6mg OD
Omperazole MUMPS : 20mg OD
Senna: 15mg PRN

Feeds provide: Approx. 450kcal/day (17kcal/kg)
Oral intake provides: Approx. 700kcal/day
Total intake (est.): 1150kcal/day (43kcal/kg 65% EAR)
1500mls fluid (oral + flushes + feed) (90% maintenance)

Stop Domperidone, alginate and PPI
Erythromycin susp: 3-4mg/kg QDS
Stimulant laxatives >>> stool softener
Is Parenteral Nutrition appropriate for palliative care?

- Risks vs benefits
- Patient’s condition and co-morbidities
- Parents’ belief and resilience
- Financial burden

Palliative PN could be considered for children with no other alternatives but....
Parenteral Nutrition

Indications

- Permanent intestinal failure (anatomical short gut or CIPO)
- Short term option when enteral feeding can’t be used (post-surgery)

Disabled children with life-limiting conditions

what if the gut does not recover?
Parenteral Nutrition

- Long hospitalisation to establish PN (access, tolerance, training)
- Infection risk
- Liver complications
- Limited mobility
- Costs
What do we do

- Consider all options (retry what has failed in the past if necessary)
- Challenge with enteral nutrition in hospital
- Discussion with palliative care team
- Sit down with parents to discuss options and set expectations

As a Team we **do not offer** HPN to severely disabled children with life-limiting conditions
So what????

- Plan ahead and **set expectations**
- Be flexible and ready to review decisions
- Consistent and persistent support
- Reassurance about expected weight gain
- Holistic approach
- Be clear about options
- Allow family to experiment/manage…but give limits!
Perseverance is not a long race; it is many short races one after the other.

Walter Elliot