Association for Palliative Medicine
Of Great Britain and Ireland

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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.

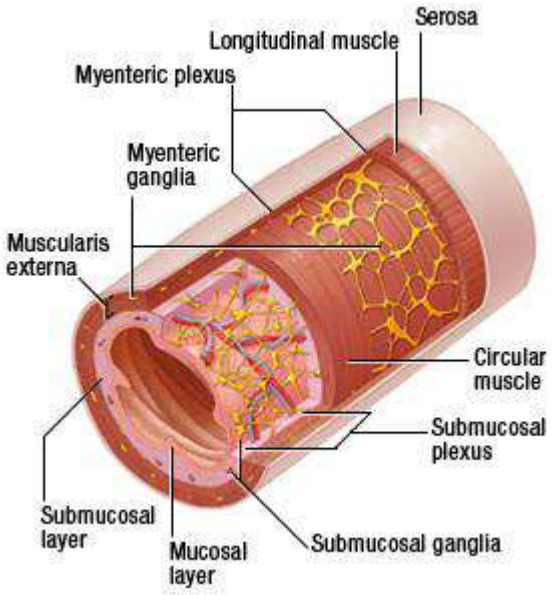
Sullivan PB, 2008

Content

- Enteric nervous system
- Gut problems in neurologically impaired children
- Setting the scene
- Management of symptoms

Enteric Nervous System (ENS)
or Intrinsic Nervous System

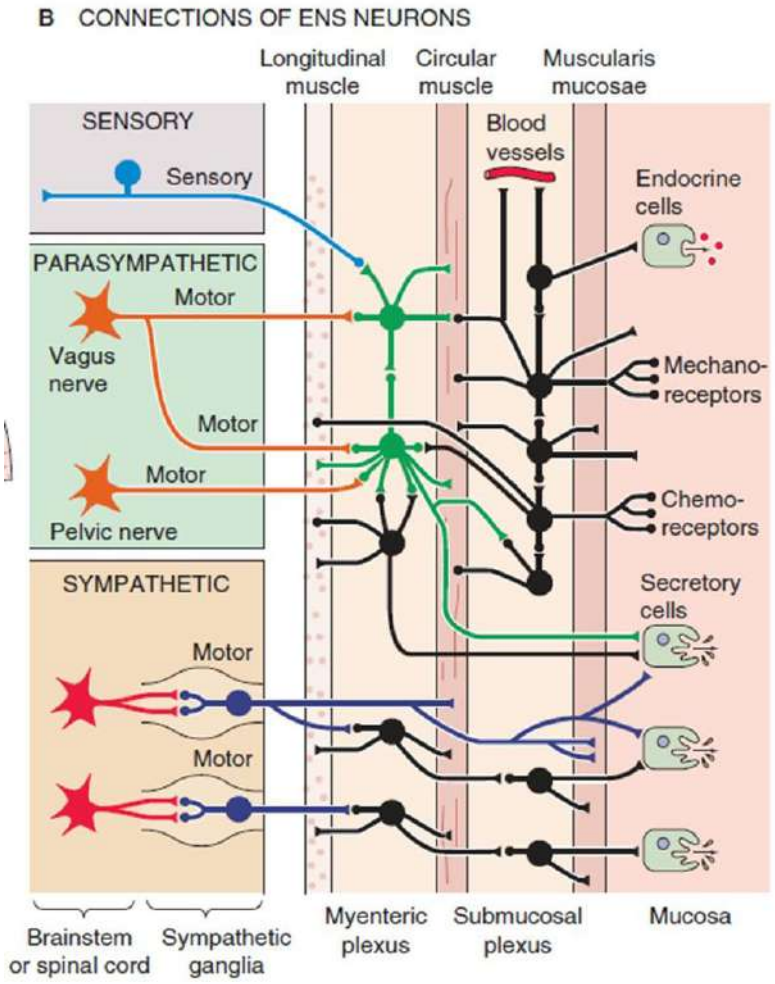
- 500 million neurons
- 5 times more than in the spinal cord



- 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



Motor neurons ⇩ Intestinal muscles ⇩ Peristalsis

Sensory neurons ⇩ Secretion of enzyme

> 30 neurotransmitters: acetylcholine, dopamine and serotonin

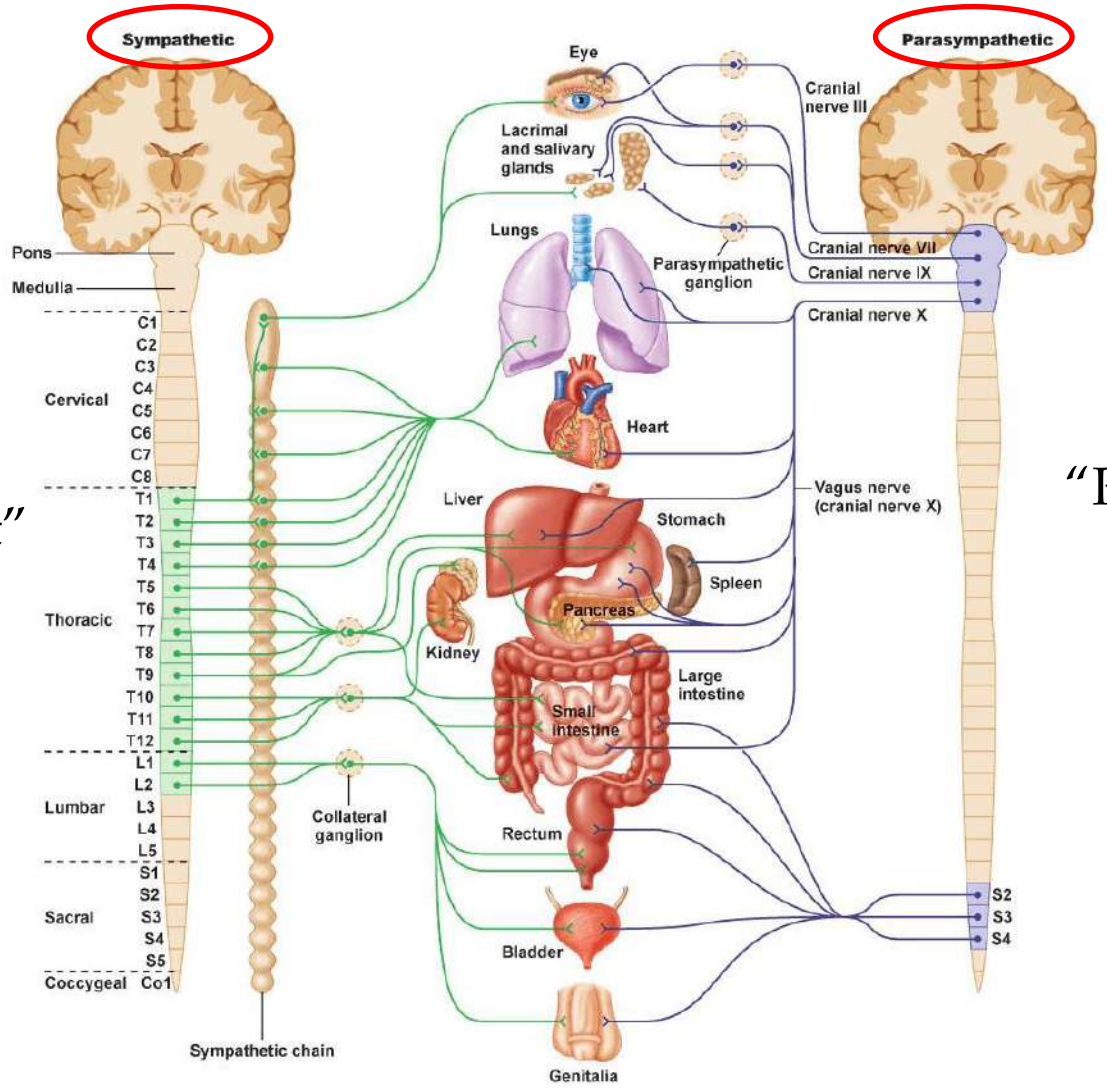
Gut has >90% of the body's serotonin and about 50% of the body's dopamine

Autonomic Nervous System

or Extrinsic Nervous System

Regulate unconscious actions

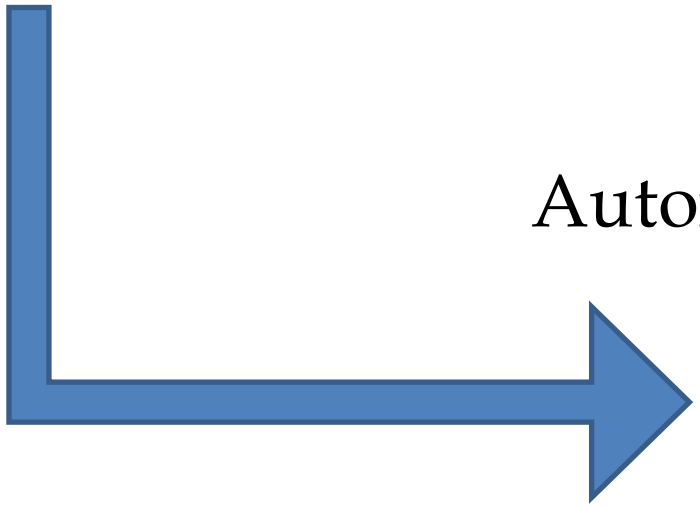
"Fight –or-flight"



"Rest-and-digest"

Setting the scene

Central Nervous System disorders



Autonomic Nervous System

modulates

Enteric Nervous System

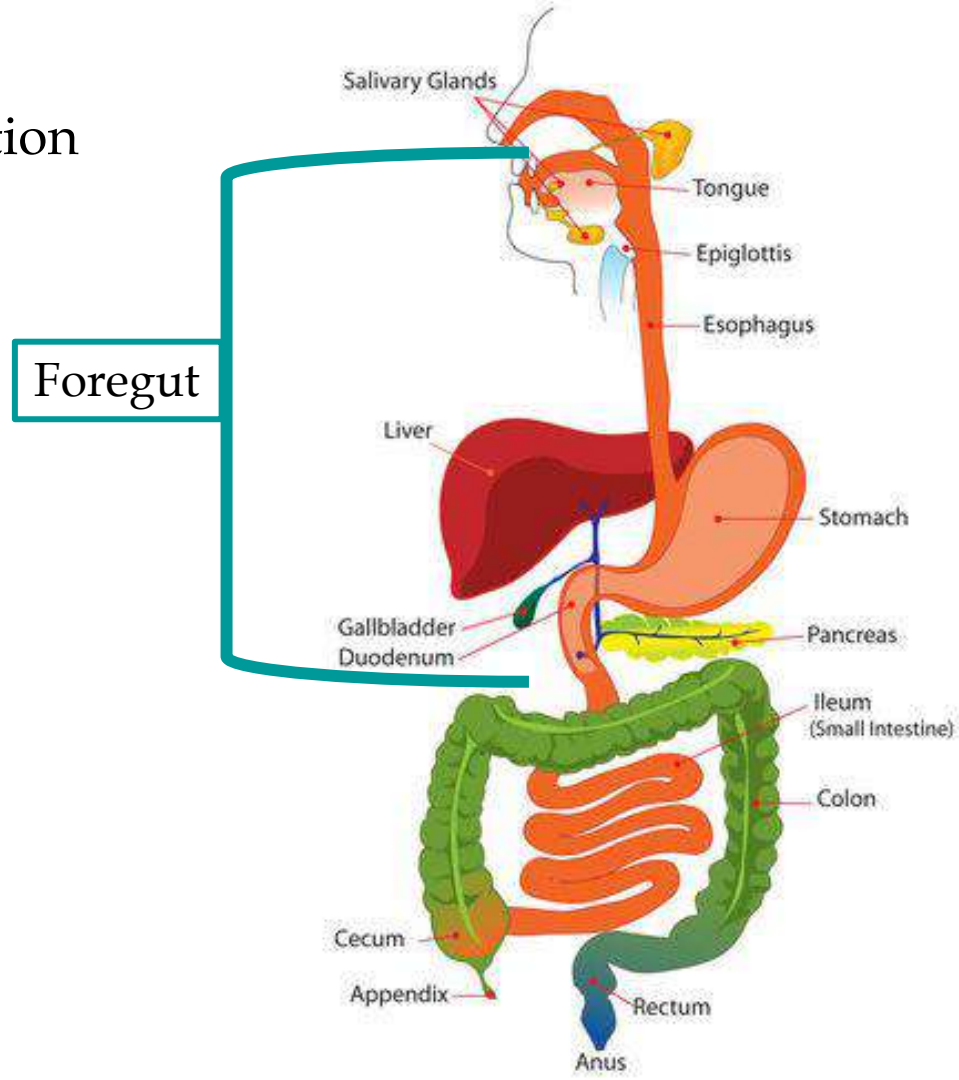
Main problems

- Foregut dysmotility

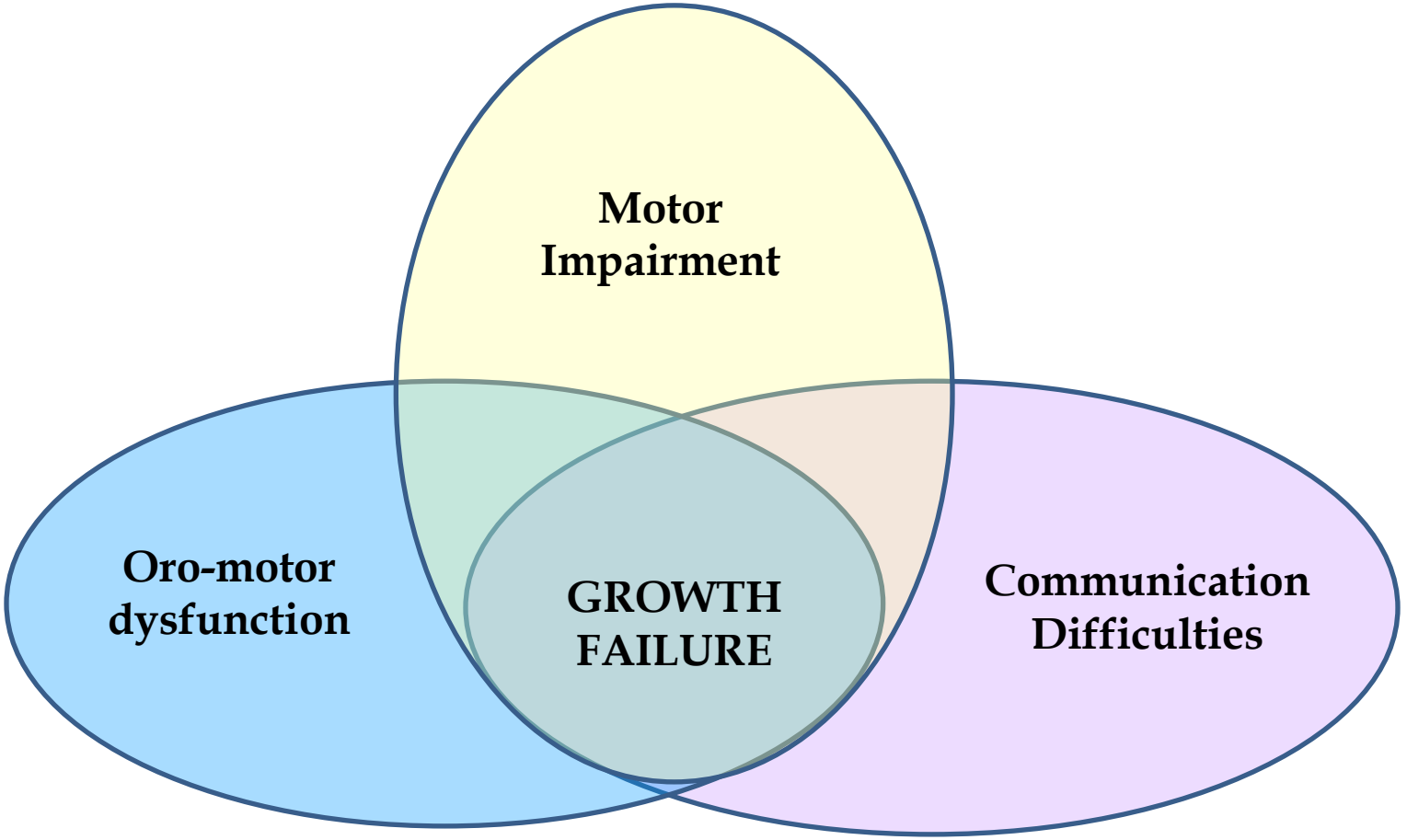
High density of autonomic innervation

1. Oral-motor impairment
2. Oesophageal dysmotility
3. GORD
4. Antro-duodenal dismotility
5. Retching
6. Dumping Syndrome

- Constipation



Neurologically impaired children:



Prevalence and severity of feeding and nutritional problems in children with neurological impairment: **Oxford Feeding Study**

PB Sullivan et al. *Developmental Medicine & Child Neurology 2000, 42: 674–680*

Comparison of type of neurological impairment

	Group A (n=271)	***Group B (n=72)***
Total with CP	261	67
Spastic quadriplegia	147	30
Hemiplegia	57	32
Dyskinetic	27	0
Ataxic	18	3
Unclassifiable	12	2

Group A: children with feeding problems
Group B: children with no feeding problems

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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*
			Mild	*Moderate*	*Severe*		
Help with feeding needed	238/268	89	27	85	126	188.9	<0.001
Choking with food	142/257	56	12	38	90	62.68	<0.001
Feeding reported as stressful or unenjoyable by parent	51/262	20	5	11	35	10.74	< 0.005
Prolonged (³ 3h/d)feeding times	71/258	28	3	8	60	53.2	<0.001
Parents considered child underweight	93/240	38	6	25	62	26.87	<0.001
Child received caloric supplements	23/271	8	1	2	20	15.64	<0.001
Gastrostomy feeding	20/265	8	1	0	19	19.63	<0.001
Never had feeding and nutritional status assessed	169/264	64	32	77	60	28.15	<0.001
Frequent vomiting	55/249	22	1	12	42	33.3	<0.001
Bowels opened > every 3 days	68/267	26	5	16	47	17.2	>0.002

Impact of feeding problems on nutritional intake and growth: **Oxford Feeding Study II**

PB Sullivan et al. *Developmental Medicine & Child Neurology 2002, 44: 461–467*

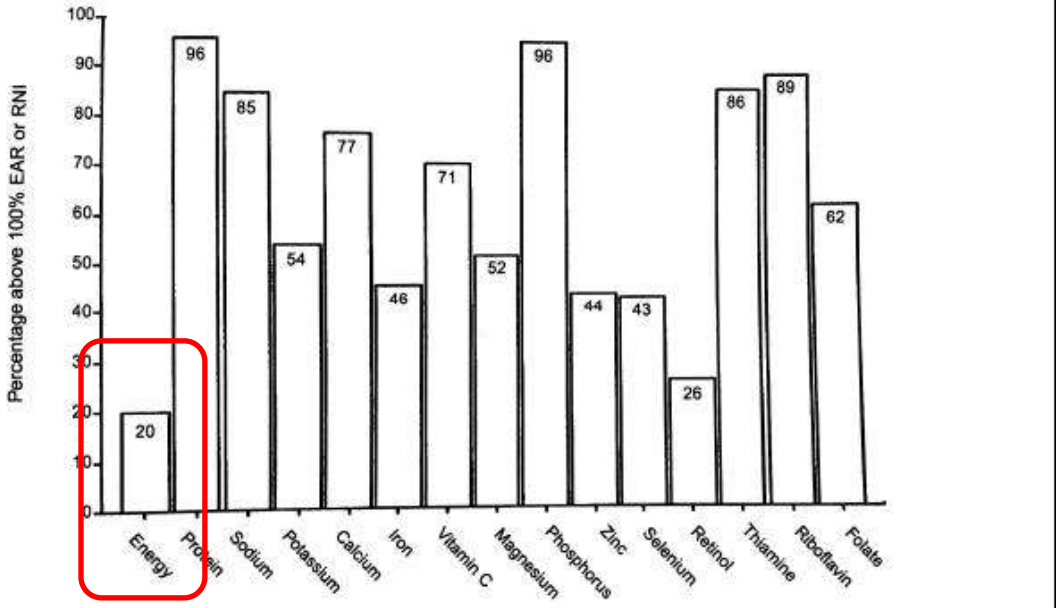


Figure 1: Dietary intakes of children visited at home. Estimated average requirement (EAR) Reference nutrient intake (RNI)

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:

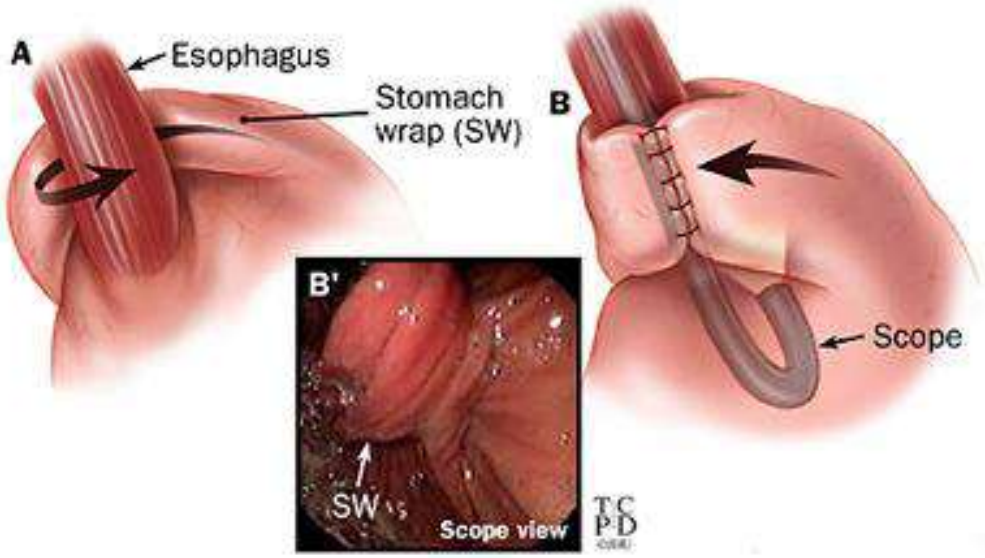
- Increase weight
- Improve overall health
- Decrease feeding times
- It has also demonstrated a significant, measurable improvement in the quality of life of carers

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

Ferluga et al. *Developmental Medicine & Child Neurology 2014, 56: 31–43*

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

Post- Nissen's

Many patients continue to have symptoms

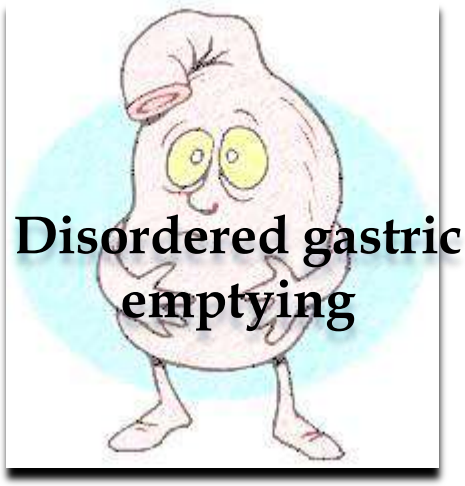
gastric dysrhythmias, unmasked by fundoplication.

- 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???

Vomiting and Gastroesophageal Motor Activity in Children with Disorders of the Central Nervous System

A Ravelli, PJ Milla. *JPGN Vol26(1), Jan 1998;56-63*



gas bloat and persistent retching

"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

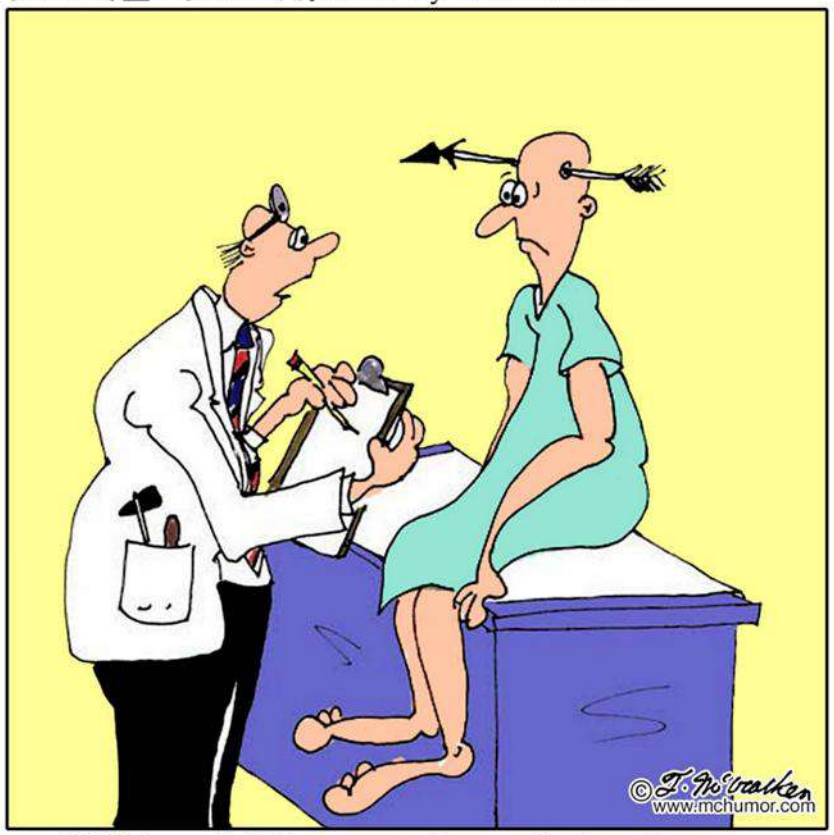
Setting the scene

- 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

MCHUMOR.COM by T. McCracken

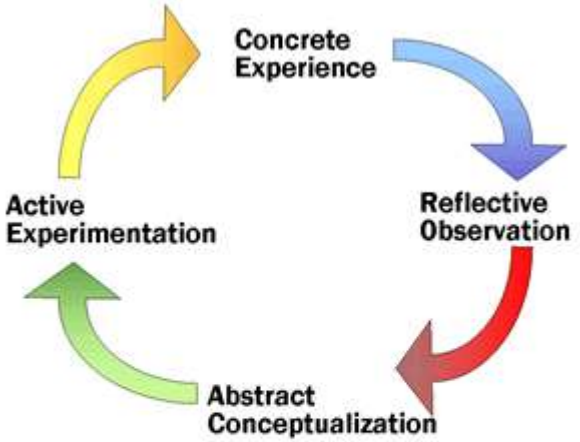


"Off hand, I'd say you're suffering from an arrow through your head, but just to play it safe, I'm ordering a bunch of tests."

- Delayed gastric emptying
- Dysmotility

Management

Management



Kolb's Cycle of Experiential Learning

image by Karin Kirk

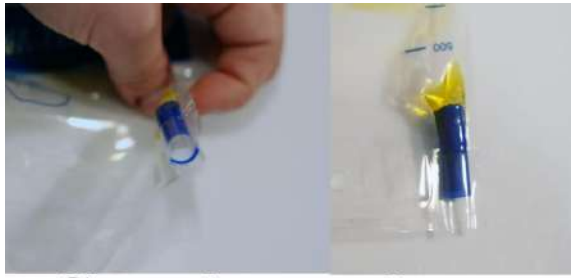
Wind

Source:
Swallowed air >>>> fermentation

Reason for improvement on venting:
Reduction of gastric distension

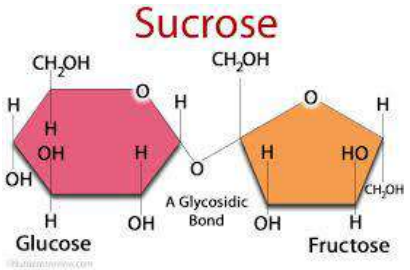
Wind - management

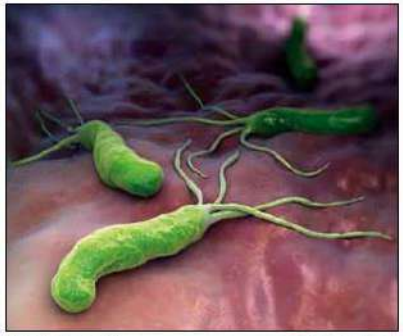
Farrell valve bag



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

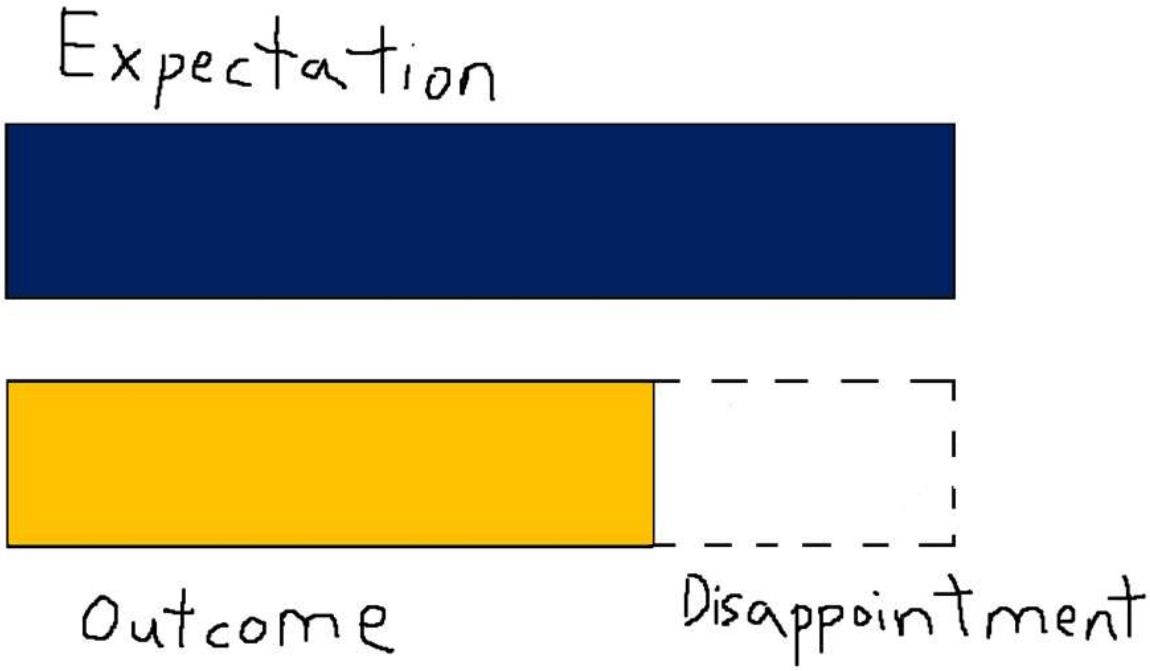
Check for: H Pylori
sugar malabsorption





Helicobacter pylori ©iStock.com/iLexx

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
NUTRITION
NONFUNCTIONAL
CATHETER
SPOILAGE
STASIS
EMBOLISM
VITAMINS
PREMIXED
ADMINISTERED
SOLUTION
GLUCOSE
PERITONITIS
WEIGHT
SEPARATELY
ENTERIC
ADMIXTURE
EXTENDED
COMMON
EXPANDER
STIMULATION
ACCESS
ACIDS
HEPARIN
CHOLECYSTITIS
INDIVIDUALLY
DEXTROSE
TOTAL
INSERTION
STANDARDIZED
MEMBRANES
ADDED
PARENTERAL
POTASSIUM
ADMINISTRATION
BILIARY
INCLUDE
FORMATION
PERIPHERALLY
SEPSIS
FEEDING
HYPERGLYCEMIA
LIVER
SOLUBLE
DISPENSE
COMPONENT
INTRAVENOUS
INTRAVENOUSLY
VENOUS
BOWEL
GASTROINTESTINAL
CHOLECYSTOKININ
DISORDER
MALNUTRITION
METABOLISED
BODY
ELECTROLYTES
CHOLESTATIC
ROUTES
PERIODIC
HYPOKALEMIA
CLOT
ORAL
ABSORPTIVE
COMATOSE
INDIVIDUAL
SYNDROME
SHORT
BILE
EMULSIFIER
TRANSAMINASES
COMPLICATION
FATTY
LIPID
WEEKS
PREVENT
PATHOGENESIS
VALVE
SLUDGE
DIGESTIVE
EXOGENOUS
EMULSIONS
ULCERATIVE
TRACT
MORTALITY
COMPLETE
ATTACHMENT
LONG
AMINO
ENDOGENOUS
DISUSE
LINE
PATIENT
FISTULA
MINIMISED
CANCER

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

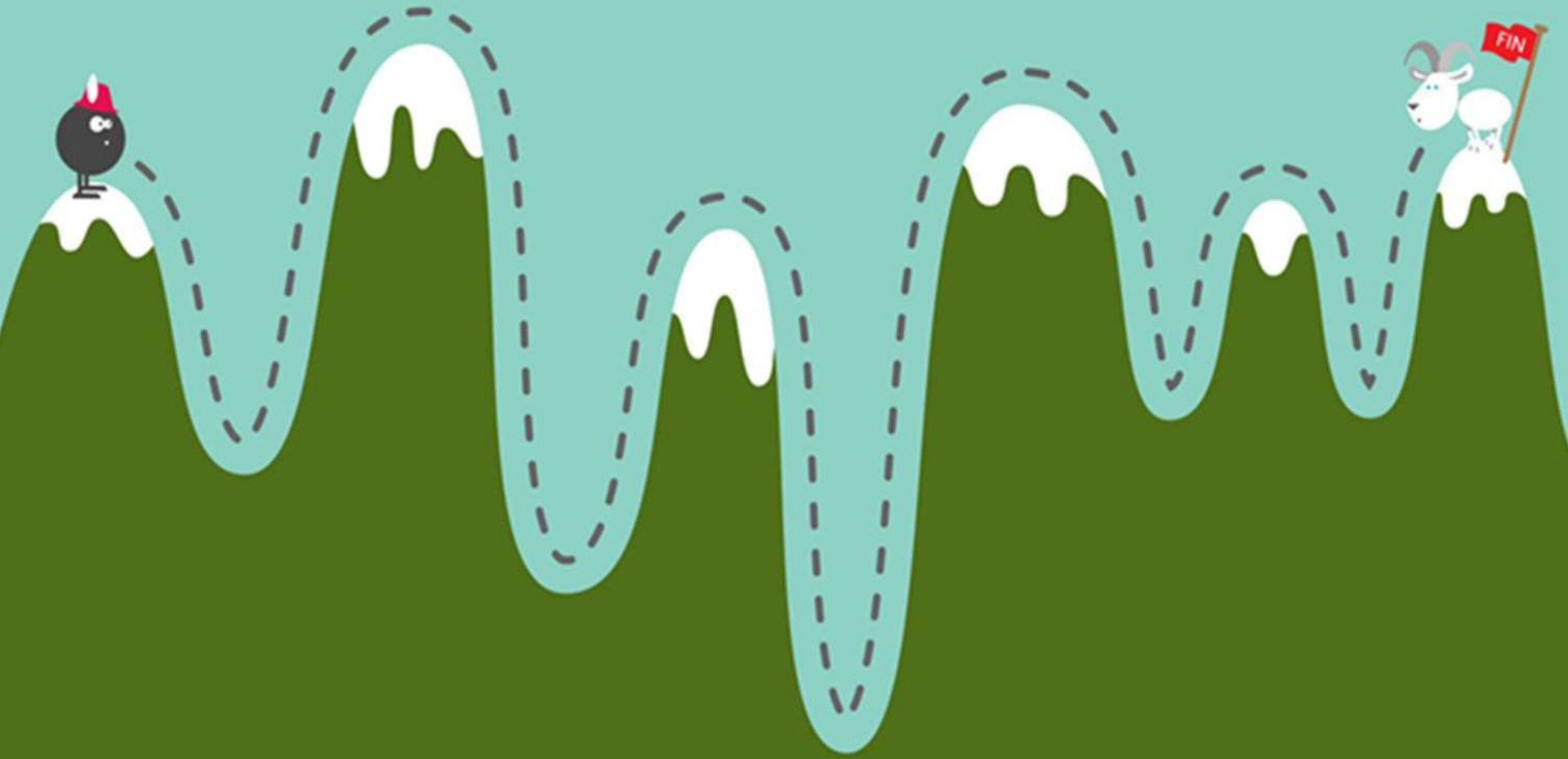
- Consider all options (retry what has failed in the past if necessary)
- Challenge with enteral nutrition in hospital
- Discussion with palliative care team
- Sit down with parents to discuss options and set expectations

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