

# The nutritional management of Paediatric Short Bowel Syndrome

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# Disclosure

- This education event is supported by Nestle Health Science
- I have received a lecture honorarium from Nestle Health Science



# Key learning objectives

- Review of the etiology and types of SBS
- Identify factors impacting outcomes in infants and children with SBS
- Discuss the clinical management of SBS
- Understand the role of nutrition in infants and children with SBS
- Describe nutritional priorities, interventions and requirements in phases of SBS management

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Summary

# Introduction

*Paediatric short bowel syndrome (SBS) is a condition acquired or caused by congenital loss of intestinal length that results in the requirement for parenteral or enteral nutritional supplementation*

# Paediatric SBS

- Literature definition: is the need for parenteral nutrition for greater than 60 days after intestinal resection or a bowel length of less than 25% of expected<sup>1</sup>
- Incidence is 24.5 per 100,000 live births - greater in preterm infants<sup>1</sup>
- No consistent definition of Ultra SBS ~ <10 - 25cm most common in literature<sup>2</sup>
- Most common cause of paediatric intestinal failure<sup>3</sup>

1. Merritt RJ et al. J Pediatr Gastr Nutr. 2017 65:588–96

2. Höllwarth ME et al. Pediatr Surg Int. 2021 33:413–9

3. Belza C et al. Journal of Multidisciplinary Healthcare. 2020 13:9–17

# Aetiology of Paediatric SBS

- Necrotising enterocolitis (NEC)
- Intestinal atresia
- Gastroschisis
- Volvulus
- Inflammatory bowel disease
- Long-segment Hirschsprung's disease

Rarer causes include:

- Total intestinal aganglionosis
- Congenital short bowel syndrome

# Type of SBS

Jejunostomy: type I

Jejunocolic: type II

Jejunoileal: type III

Necrotising enterocolitis, Intestinal atresia

Total intestinal  
aganglionosis

Volvulus, Gastrochisis



# Important factors in Paediatric SBS

- Residual bowel length and location resected, including anastomoses<sup>1, 2</sup>
- Quality and function of remaining bowel<sup>2</sup>
- Continuity of gut and stoma placement<sup>2</sup>
- Presence or absence of ileocaecal valve (ICV)<sup>2</sup>
- Age at resection<sup>1</sup>

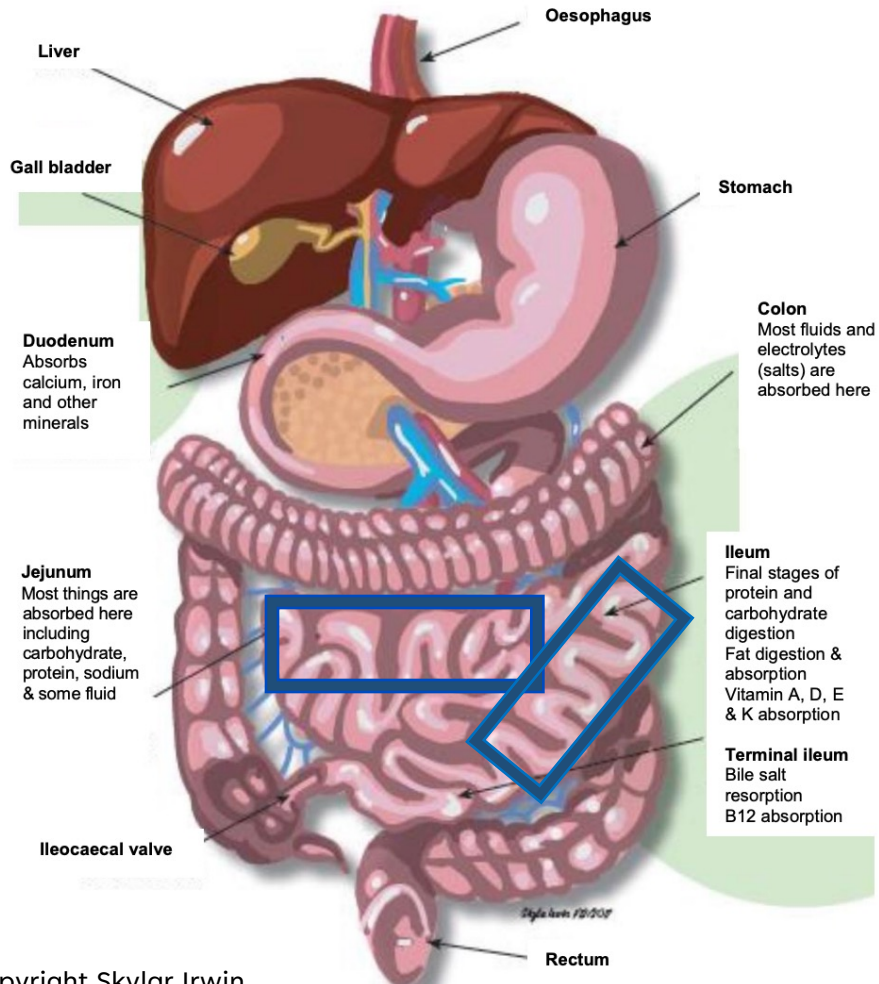
1. D'Antiga L et al. J Pediatr Gastr Nutr. 2013 56:118-26

2. NZ National Intestinal Failure and Rehabilitation Service Guidelines

<https://starship.org.nz/guidelines/browse?sp=national-intestinal-failure-and-rehabilitation-service-nz-nifrs>

Published 2021. Accessed 15 May 2021

# Consequences of intestinal resection



## Jejunual resection

- Temporary reduction in absorption of most nutrients
- No significant impact on pancreatic enzyme and bile secretion
- Modest functional adaptation through transport and enzyme activity

## Ileal resection

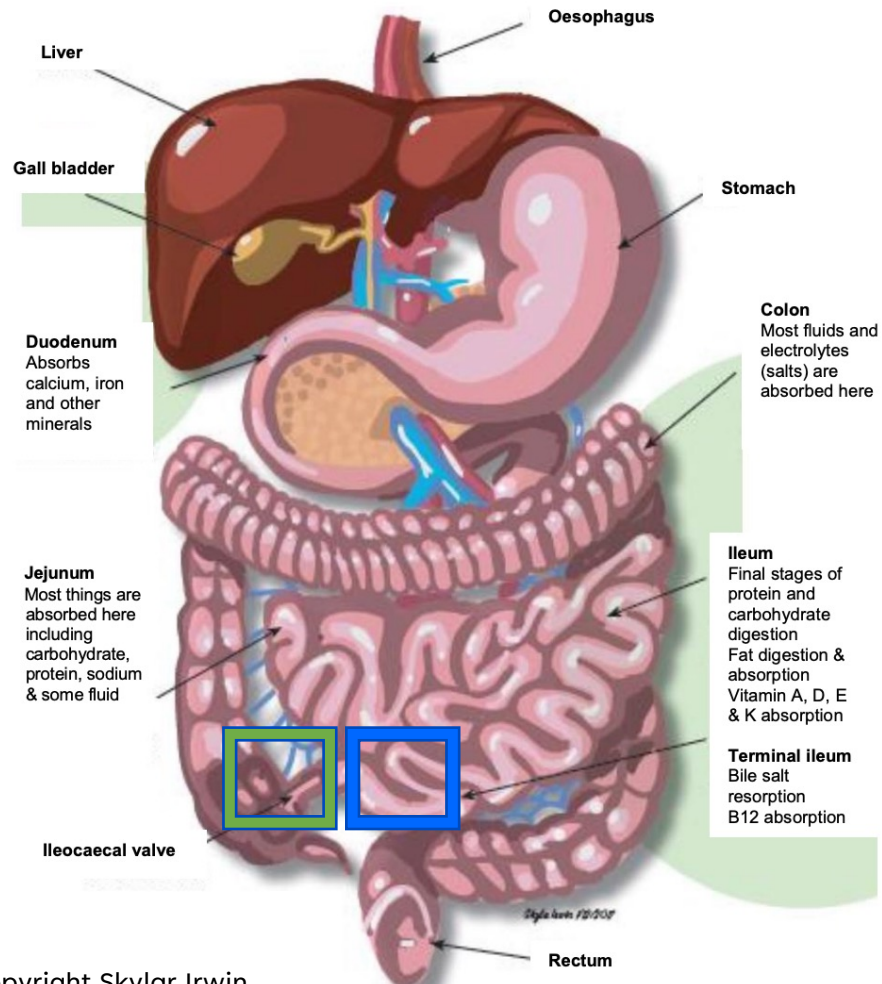
- Reduction in trophic gut hormone production ie. Glucagon-Like Peptide-2
- Reduced fluid absorption and increase in watery diarrhoea
- Loss of “ileal break”

NZ National Intestinal Failure and Rehabilitation Service Guidelines

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# Consequences of intestinal resection



## Absence of Terminal Ileum

- Require B12 supplementation
- Disruption of enterohepatic circulation = bile acid deficiency
  - Malabsorption of fats and fat-soluble vitamins
  - Increased oxalate absorption (oxalate nephrolithiasis)
  - Colonic secretomotor diarrhoea due to the presence of bile salts

## Loss of ileocaecal valve

- Loss of barrier and regulation of ileal and colonic fluids
- Increased risk of rapid transit time and small bowel bacterial overgrowth

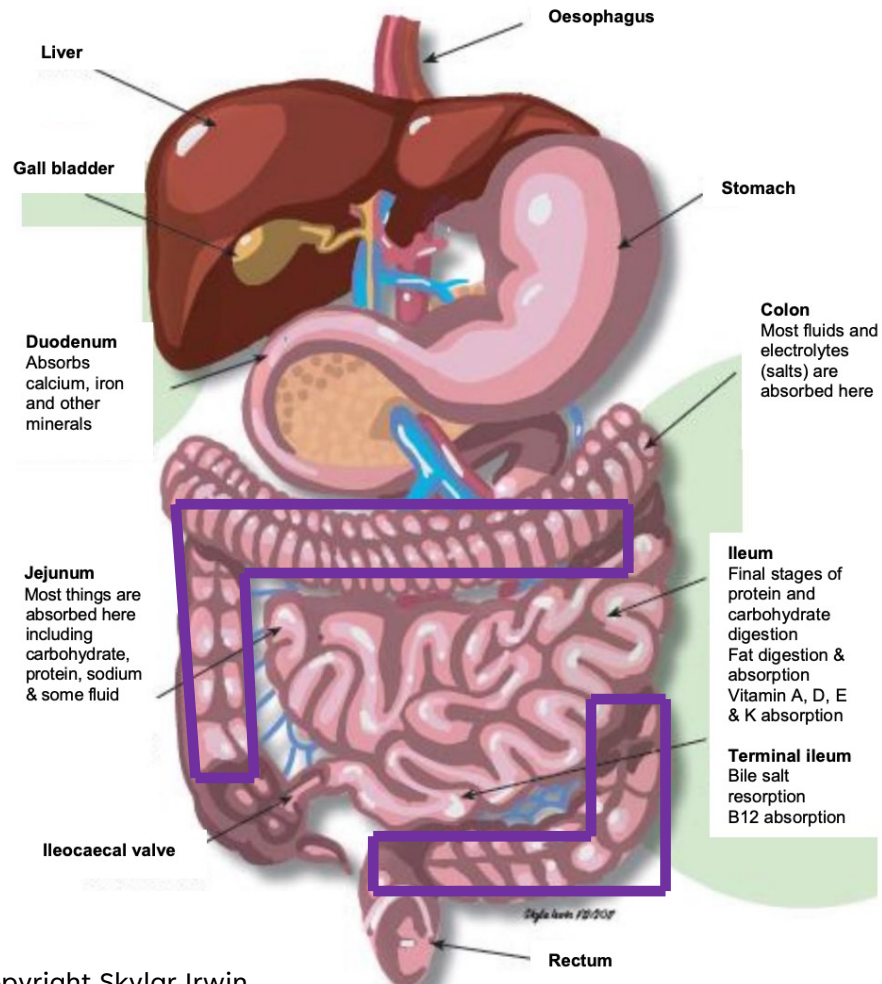
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# Consequences of intestinal resection



## Loss of colon

- Reduced water, electrolyte and short chain fatty acids reabsorption
- Loss of Peptide YY production

## Presence of colon in continuity

- Influences functional adaptation of the jejunum

NZ National Intestinal Failure and Rehabilitation Service Guidelines

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# Symptoms of Paediatric SBS

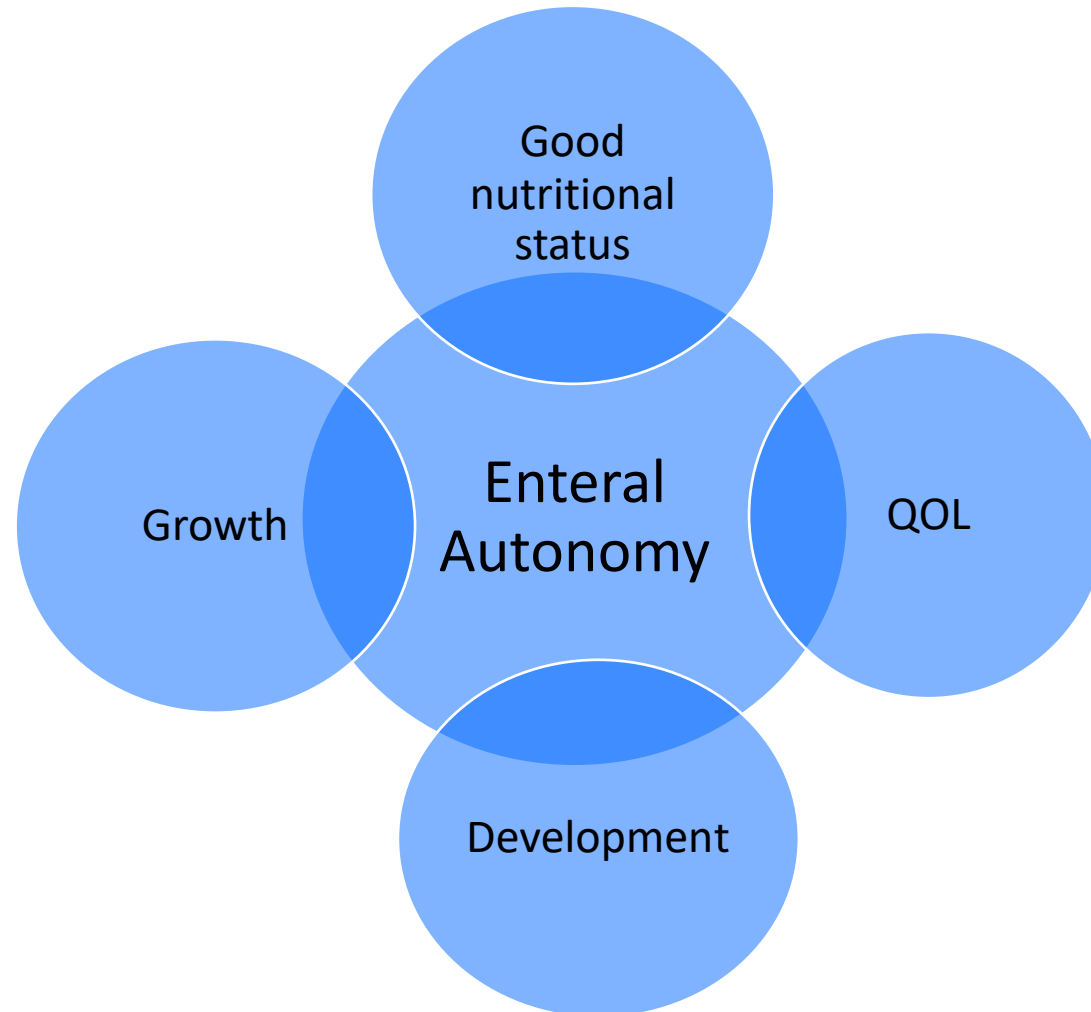
- Poor digestion and absorption
- Diarrhoea
- Abdominal distension
- Vomiting
- Nausea
- Gastric hypersecretion
- Dehydration
- Poor growth
- Fatigue

# Clinical management

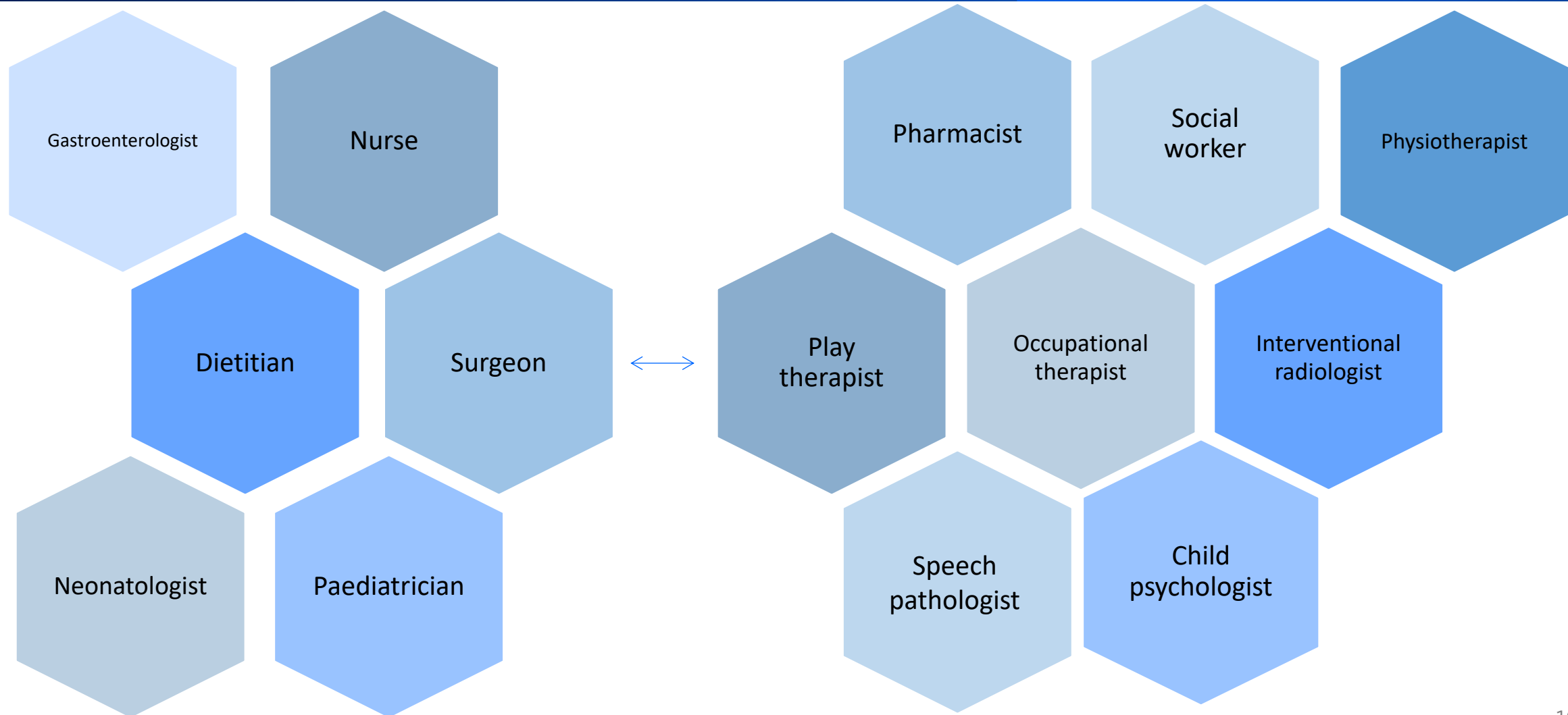
# Aims of SBS management

Ultimate goal is  
enteral autonomy

BUT...

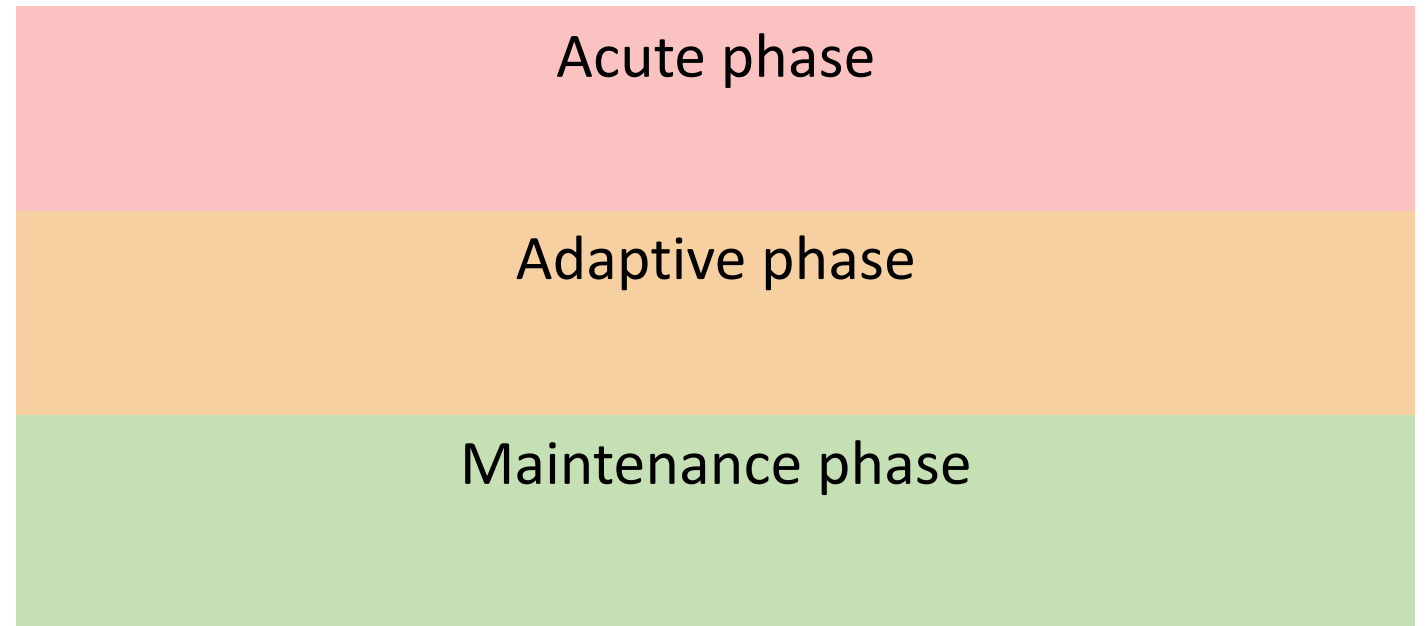


# It will take a village!





# Phases of SBS management



# Acute phase

Focus of phase: restore and maintain fluid, electrolyte and acid-base balance

- *Medical:*
  - Symptom management to achieve stability
- *Surgical:*
  - Stoma – plan for reconnection, distal feeding
- *Nutritional:*
  - Parenteral nutrition (PN) provides full nutrition
  - Start trophic enteral nutrition using an oral rehydration solution

# Adaptive phase

Focus of phase: Intestinal adaptation

## **Structural changes**

- Enterocyte numbers and intestinal weight increase
- Villous lengthening and crypt hyperplasia
- Angiogenesis
- Dilatation and lengthening of intestinal lumen

## **Functional changes**

- Increased brush boarder enzyme activity
- Increased cellular transporter activity
- Slower transit time

# Adaptive phase

- *Medical:*
  - Management of symptoms of SBS and complications of PN
  - Use of hormone therapy ie. Glucagon-Like Peptide-2 analogue
  - Ensure growth and development is acceptable
- *Surgical:*
  - Interventions to achieve enteral autonomy such as tapering, restoring bowel continuity and lengthening procedures ie LILT or STEP
- *Nutritional:*
  - Wean PN, advance enteral and oral nutrition to achieve enteral autonomy
  - Meet nutritional requirements and prevent deficiency
- *MDT...*

# Maintenance phase

Focus of phase: Maintenance of enteral autonomy with consideration to any ongoing symptoms, growth, development and QOL

○ *Medical:*

- Monitoring of growth, development and biochemical data
- Treatment of new and long term issues / comorbidities
- Maintenance of therapeutic options ie. supplements, medications

○ *Nutritional:*

- Oral and/or enteral nutrition to meet nutritional requirements
- Tailor plans and dietary advice to the patient

# Nutritional management

# Role of nutrition

## Priorities by phase of Paediatric SBS

### Acute phase

- Achieve stability – fluid and electrolytes

### Adaptive phase

- Promote and stimulate intestinal adaptation
  - *'Exposure' of the lumen to nutrients*
  - *Stimulate trophic hormones and secretion production*
  - *Create a nutrient 'workload'*

### Maintenance phase

- Maintain enteral autonomy



# Acute phase - nutritional management

## Assessment

### *Anthropometry*

- Weight, length/height, head circumference, MUAC, malnutrition status

### *Biochemistry*

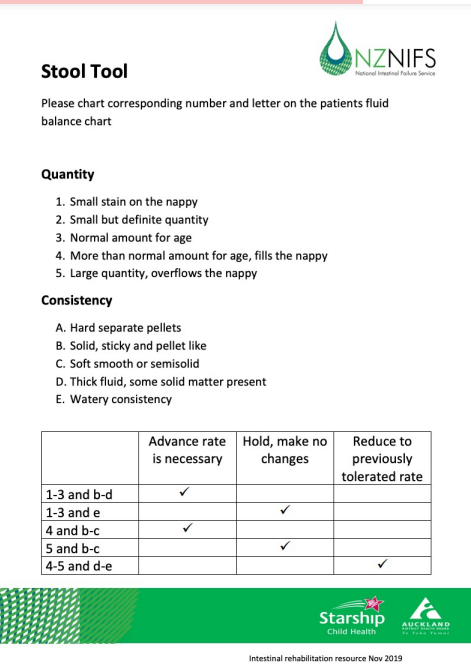
- Blood, urine and stool

### *Clinical*

- Anatomy, remnant bowel length and quality, stoma and surgical plan
- Symptoms ~ vomiting, diarrhoea/stool transit
- Fluid balance
- Access: CVAD, feeding tubes
- Medications, IV fluids and infusions

### *Diet/Nutrition plan*

- Current nutrition intervention, allergy history, refeeding syndrome risk



**Stool Tool**

Please chart corresponding number and letter on the patients fluid balance chart

**Quantity**

1. Small stain on the nappy
2. Small but definite quantity
3. Normal amount for age
4. More than normal amount for age, fills the nappy
5. Large quantity, overflows the nappy

**Consistency**

- A. Hard separate pellets
- B. Solid, sticky and pellet like
- C. Soft smooth or semisolid
- D. Thick fluid, some solid matter present
- E. Watery consistency

	Advance rate is necessary	Hold, make no changes	Reduce to previously tolerated rate
1-3 and b-d	✓		
1-3 and e		✓	
4 and b-c	✓		
5 and b-c		✓	
4-5 and d-e			✓

Starship Child Health AUCKLAND  
Intestinal rehabilitation resource Nov 2019



# Acute phase – nutritional management

## *Estimating nutritional requirements*

- Energy, protein, fluid, electrolytes, trace elements, vitamin and minerals requirement as per the 2018 ESPGHAN guidelines<sup>1</sup>
- PN solutions available

Patient age	Kcal/kg/day	
	Acute/critical illness	Stable/recovery
Term neonates	45 - 60	60 - 85
Infants 4 - 10 kg	45 - 60	60 - 85
Children 1 - 20 kg	40 - 55	55 - 75
Children 20 - 30 kg	40 - 55	55 - 75
Children 30 - 60 kg	30 - 40	40 - 65
Young people > 60kg	20 - 30	25 - 50
Adult	20 - 35	25 - 50

Table used with permission from: NZ National Intestinal Failure and Rehabilitation Service  
<https://starship.org.nz/guidelines/browse?sp=national-intestinal-failure-and-rehabilitation-service-nz-nifrs>  
Published 2021. Accessed 15 May 2022

1. ESPGHAN/ESPEN/ESPR/CSPEN guidelines on pediatric parenteral nutrition. Clinical Nutrition, 2018.

# Acute phase - nutritional management

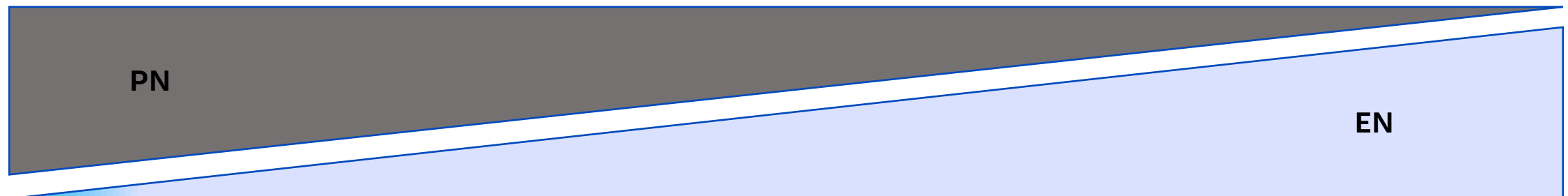
## Intervention

- Collaborate with medical team regarding fluid and electrolyte management
- Full PN with consideration to:
  - Fluid requirement dependent on underlying symptoms and fluid balance
  - Mixed lipid emulsion use such as SMOFLipid™
  - Use of Omegaven™ with Intestinal Failure Associated Liver Disease (IFALD)
  - 24 hour infusion
  - Cycling PN if appropriate
  - Do not overfeed
- Start trophic enteral feeds via tube
  - Advocate use of the gut with the surgeons – likely first trial with ORS

# Acute phase - nutritional management

## Monitoring

- Weight change
- Biochemistry – daily bloods with PN changes
- Symptoms: input and output from bowel/stoma, abdominal distension
- Changes to plan or clinical status, surgical complications, medications, IV fluid replacement plan





# Adaptive phase – nutritional management

## Intervention

*“Enteral nutrition in patients with SBS is an art, aimed to compose the nutritional therapy in a sophisticated way to stimulate optimally the remaining intestinal parts in a given patient”*

- Wean PN when total enteral energy meets 20% of requirements
- Reduce PN duration according to age and enteral nutrition
- Enteral nutrition should be advanced slowly in a stepwise fashion according to symptoms via gastric or jejunal feeding

# Adaptive phase – nutritional management

## Intervention

- Start at standard strength and advance concentration as tolerated
- Start small intermittent breast or bottle feeds to develop oral feeding skills
- Incorporate day feeds with continuous nocturnal feeds
- Change one thing at a time
- Maintain growth, prevent deficiency and aim for good quality of life

*WHAT CAN I USE TO FEED.....*

# Adaptive phase – nutrition options

## REMEMBER

The key to progress is 'exposure' of the lumen to nutrients and creating a nutrient 'workload'

### **Breastmilk**

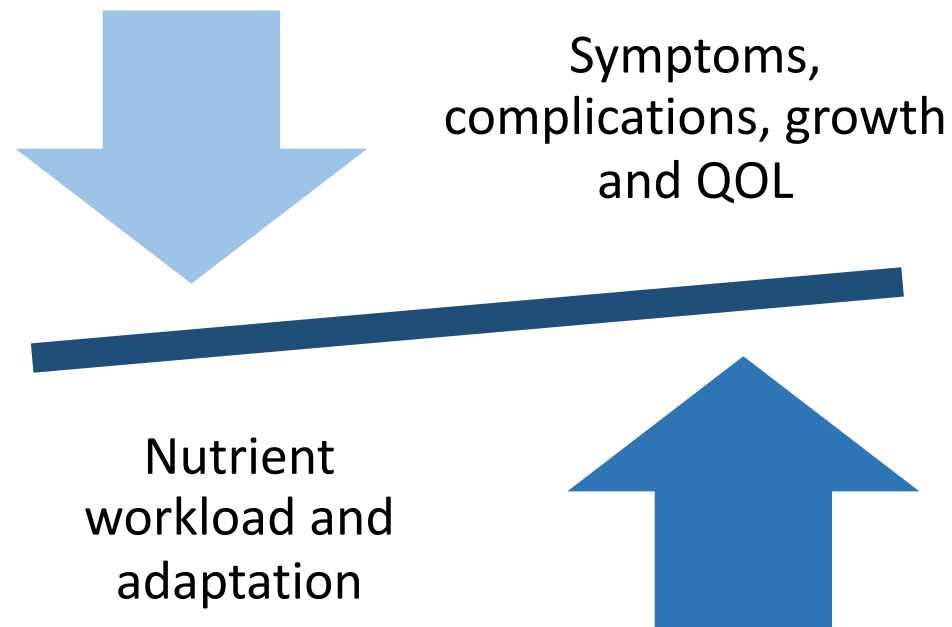
- Use if available and the infant tolerates this without significant worsening symptoms
- Contains whole protein and LCT
- Protective factors such as mother's microbiota, immunoglobulin A, nucleotides, epidermal growth factor, growth hormone
- Human milk lactase and lipase



# Adaptive phase – nutrition options

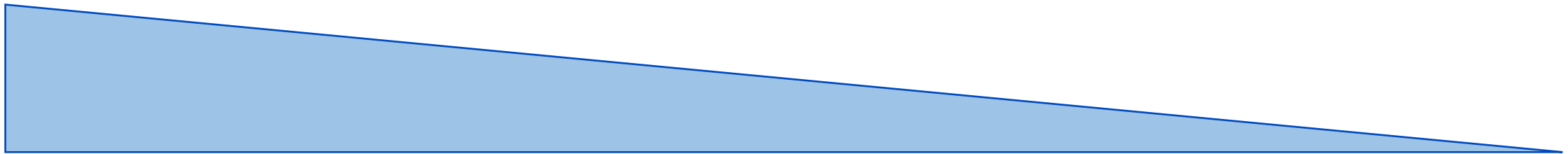
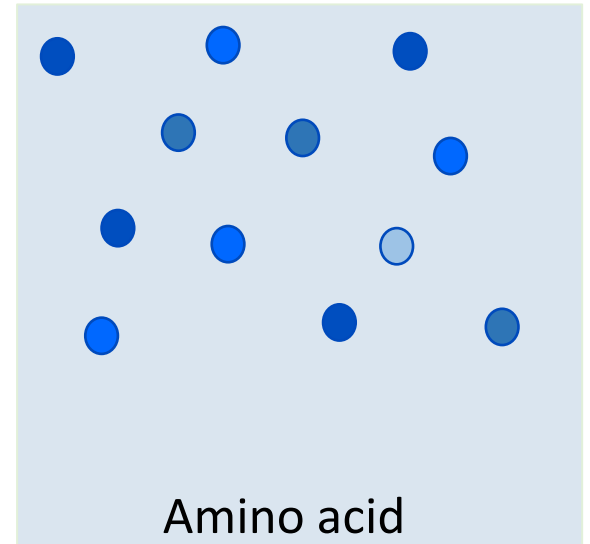
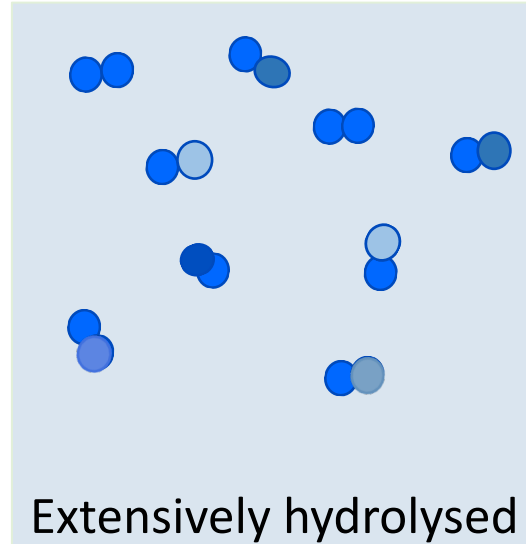
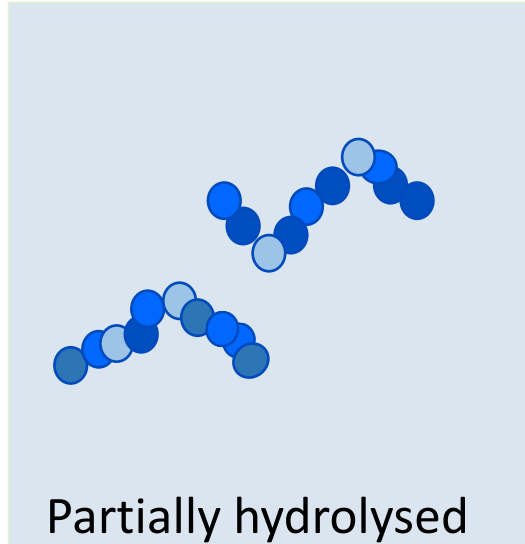
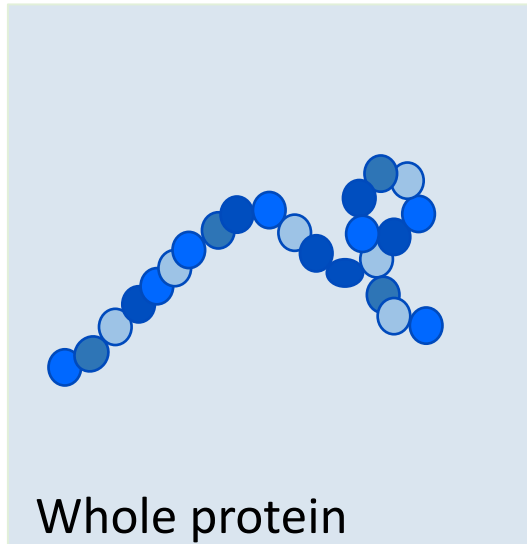
## Formula

- Know the composition of macronutrients in formulas available, every formula has a place and be flexible





# Nutrient 'workload' - protein



Highest workload  
Most trophic

Lowest workload  
Less trophic

# Nutrient 'workload' - lipid

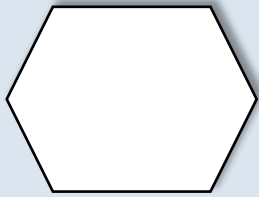
## **Long chain triglycerides**

- Low osmolar load, higher 'workload'
- Source of essential fatty acids
- Malabsorption where there is loss of ileum and IFALD

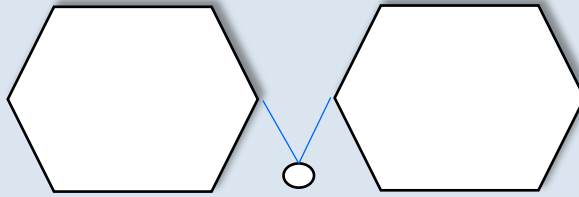
## **Medium chain triglycerides**

- Higher osmolar load
- Water soluble
- Improved absorption in SBS where there is loss of ileum and IFALD

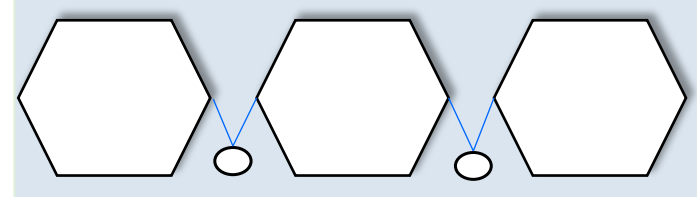
# Nutrient 'workload' - carbohydrate



Monosaccharides



Disaccharides

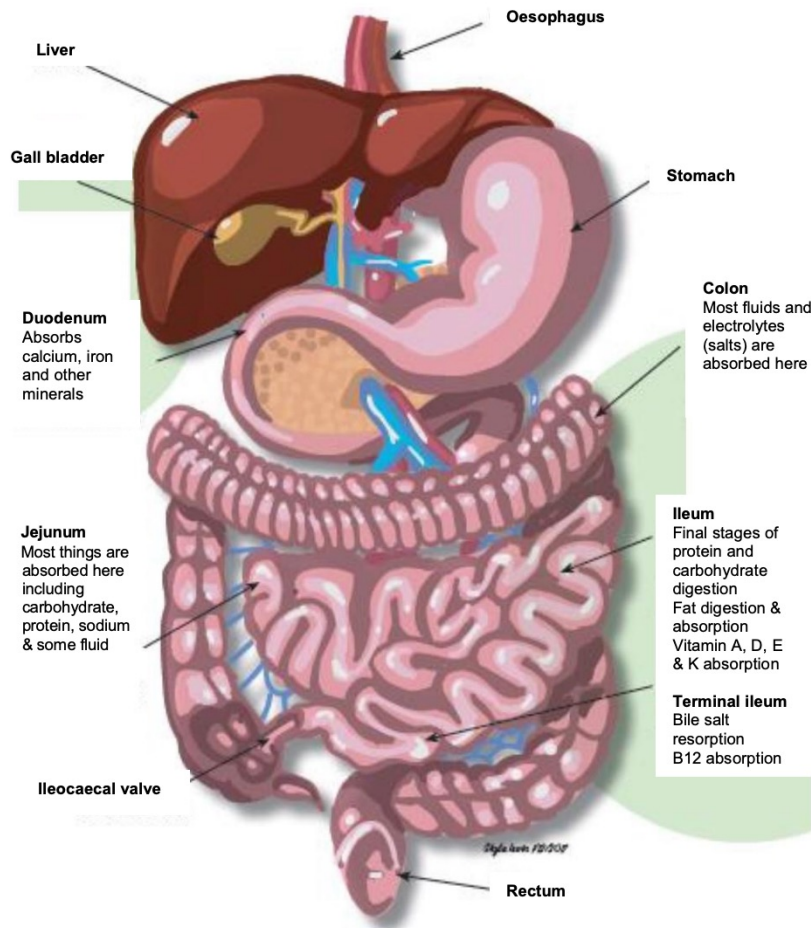


Polysaccharides

Lowest workload  
Less trophic

Highest workload  
More trophic

# Adaptive phase – nutrition options



SBS Scenario	First option	Second option
Loss of jejunum	Whole protein LCT	Hydrolysed Low lactose
Loss of ileum	Whole protein	Hydrolysed
Loss of terminal ileum	LCT	MCT and LCT
Loss of colon	Whole protein LCT	Hydrolysed LCT
Colon not in continuity	Whole protein LCT	Hydrolysed LCT
Ultra SBS	Breastmilk LCT	Amino acid LCT
IFALD	LCT and MCT	

SBS: Short bowel syndrome, LCT: long chain triglyceride, MCT: Medium chain triglyceride, IFALD: Intestinal Failure Associated Liver Disease

# Adaptive phase – nutrition options

## Solids and food

- Offer solids from 4 – 6 months when infant is developmentally ready
- Timely introduction stimulates oral motor activity and reduces risk of oral aversion
- Infants with hypersensitivity and aversive feeding behaviour should be referred to a speech pathologist early
  
- Start with small amounts
- What solids should be tried.....
  
- Use of blenderised foods....

# Adaptive phase – other tools

## **Fibre**

- Important where children have an intact colon
- Soluble fibre ie. pectin or guar gum can:
  - Slow small bowel transit
  - Improve stool consistency
  - Aid water and sodium absorption

## **Prebiotics**

- Fermented by bacteria in the colon to short chain fatty acids (acetic, propionic and butyric acid)
- Stimulate intestinal adaptation by release of GLP-2

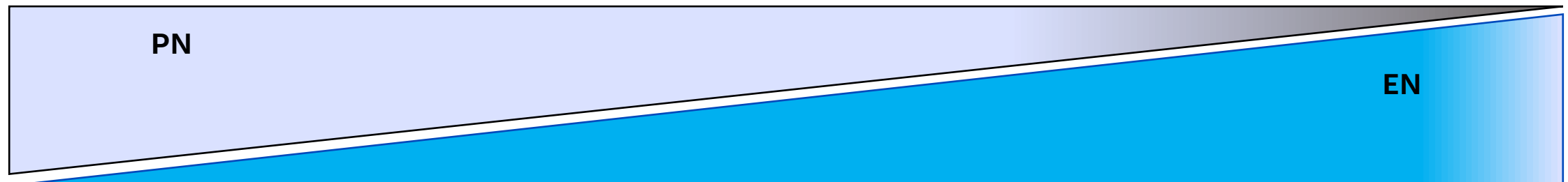
## **Probiotics**

- Bifidobacterium breve, Lactobacillus casei are used most commonly in treating SBBO in combination with cycling antibiotics
- Evidence lacking in human studies

# Adaptive phase - nutritional management

## Monitoring

- Growth rate
- Bloods, stool and urine
- Symptoms: stool output/transit, nausea, pain and abdominal distension
- Medical / surgical complications and considerations
- Medication change
- Feeding progress, development, activity level
- Team and family goals





# Maintenance phase – nutritional management

## Regular assessment and monitoring

- Growth
- Biochemistry
- Symptom changes
- Medication changes
- Enteral and oral nutrition
- Vitamin and mineral supplements
- Estimated requirements - age and stage
- Development, activity and lifestyle



# Maintenance phase – nutritional management

## Intervention

- Oral and/or enteral nutrition diet individualised to patients needs and age
- High energy diet = complex CHO, LCT, adequate protein
  - Loss of ileum: malabsorption of fat, micronutrients
  - Loss of colon: loss energy potential from short chain fatty acids
  - Hydrolysed protein and MCT may be still be required
  - Oral rehydration solutions
- Oral nutritional supplements can be an important part in paediatric SBS

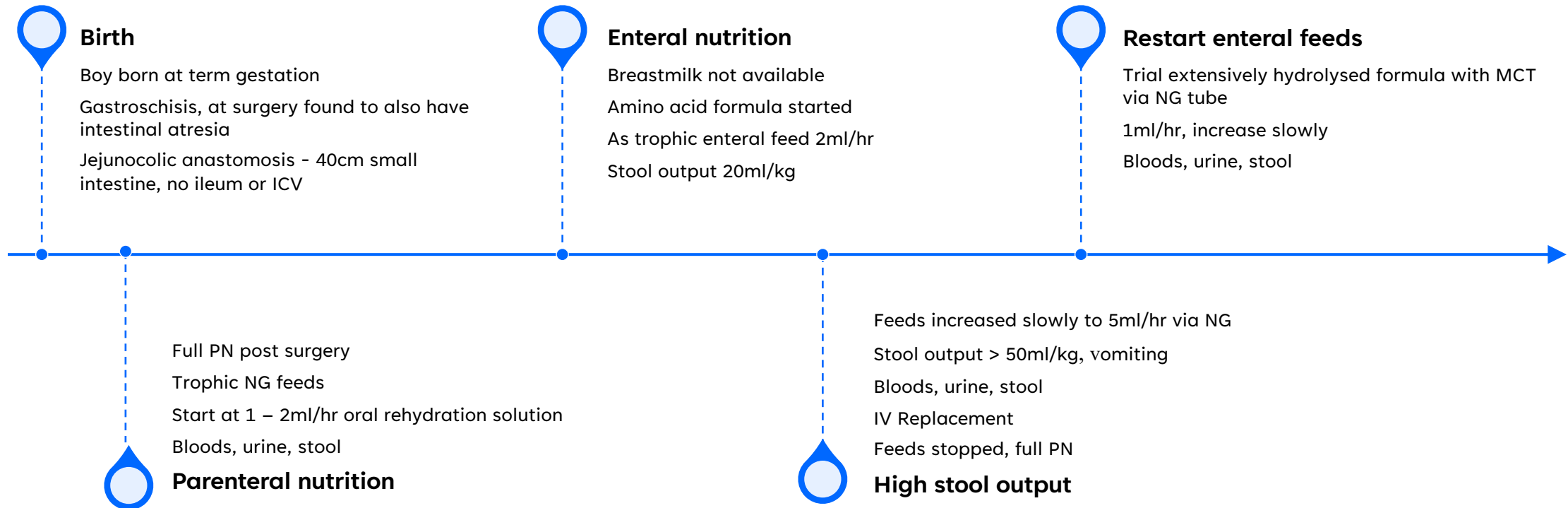
# Maintenance phase – nutritional management

## Intervention

- Vitamin and mineral supplementation
  - Loss of ileum: vitamin B12, fat soluble vitamins, magnesium, calcium, zinc, copper, selenium
  - Loss of colon: sodium
- Limit simple sugars
- Encourage fibre, especially soluble fibre where there is a intact colon
- Restriction of oxalate with loss of ileum and intact colon
- Adequate fluid for age and stage
- Regular meal pattern with small frequent meals, chew food well

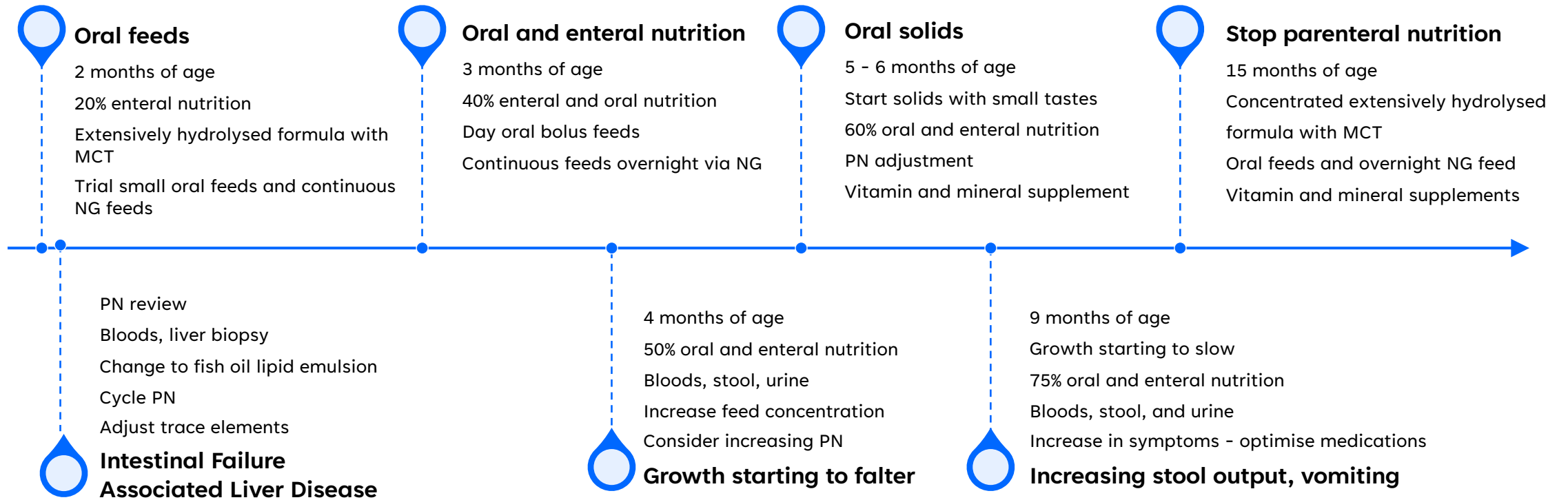
# **Theory to Practice: Bailey**

# Acute phase



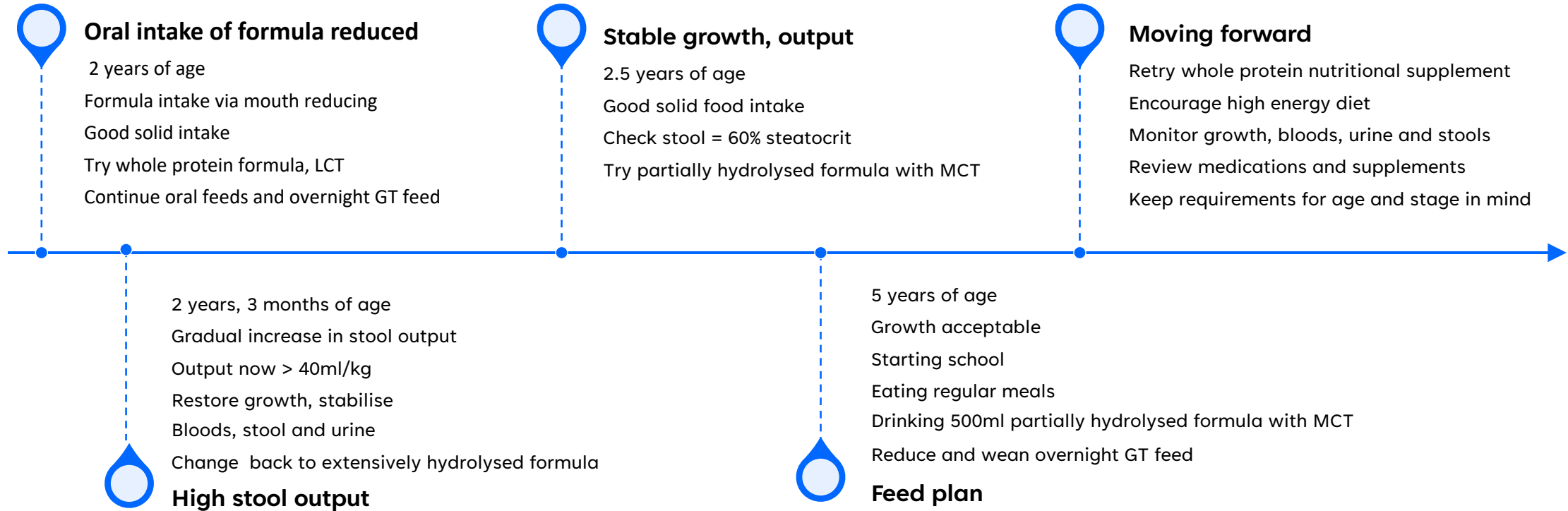
ICV: ileocaecal valve, PN: parenteral nutrition, NG: nasogastric, MCT: medium chain triglyceride

# Adaptive phase



PN: parenteral nutrition, NG: nasogastric, MCT: medium chain triglyceride

# Maintenance phase



GT: gastrostomy, LCT: long chain triglyceride, MCT: medium chain triglyceride

# Summary

- Paediatric SBS is a condition that requires intensive MDT management
- Nutrition is key in all phases of SBS management
- Interventions are tailored to the infant or child
- No hard and fast rules
- Work does not end with enteral autonomy
- Ensure your goals are aligned with your patients goals in all phases



**Thank you**