Diet in CHI Type & Life Stage

Michelle Fuery

Senior Dietitian

Topics

- Dietary Goals in CHI
- Dietary Requirements by CHI

  Type
- Diet in CHI by life stage.
- Feeding Difficulties.

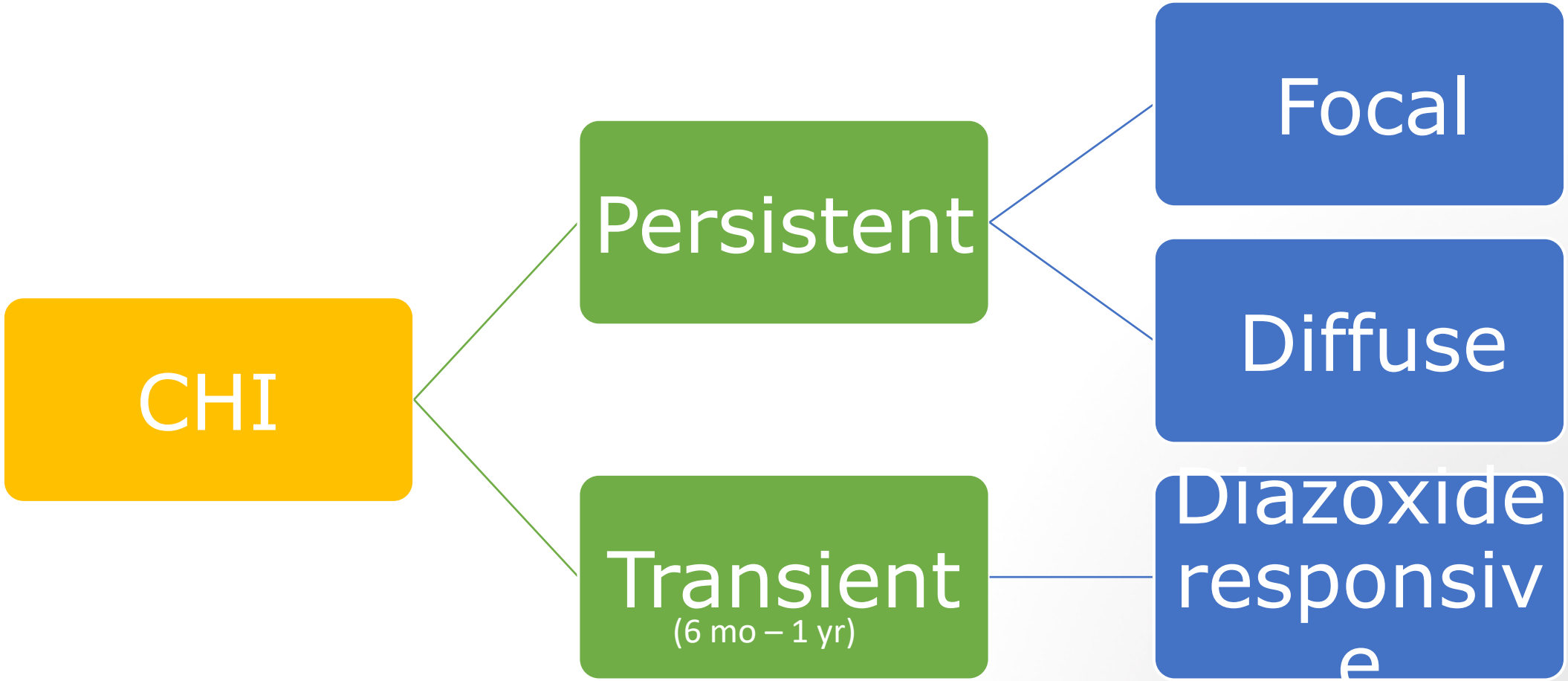
What is Congenital Hyperinsulinism?

- Excessive and unregulated secretion of insulin from beta – cells of the pancreas. The insulin does not 'turn off' normally in response to low blood glucose levels.
- Consistent high levels of insulin, switch off all alternative fuels such as ketones bodies from being made, therefore, the body cannot use ketone bodies as an alternative fuel when the body is deprived of glucose.
- This results in life- threatening hypoglycaemia and can result in long- term neurological complications without treatment.

- CHI is very rare, occurring in approx. 1 in 50,000 births.
- There are several known genetic causes of CHI.
- Abnormalities in the genes ABCC8 and KCNJ11 are the most common cause of severe CHI.

Ackermann & Palladino et al., 2015.

Types of CHI


CHI
Persistent
Focal
Diffuse
Transient
(6 mo – 1 yr)
Diazoxide responsive

Transient CHI

- Can occur in infants with no predisposing factors; the mechanisms causing this are currently unclear
- This group has been shown to be fully responsive to medical management with a medication called diazoxide, which eventually can be weaned and stopped.
- Infants with Beckwith Wiedermann syndrome, 50% have been observed to have hyperinsuliniaemic hypoglycaemia.

Persistent CHI

- The management of focal and diffuse is different.
- If accurately located and completed removed, focal disease can now be cured.
- Diffuse disease, if medically unresponsive, will require a 98% pancreatectomy

The role of the Dietitian

- Nutritional assessment and monitoring of both inpatients and outpatients with hyperinsulinism
- Optimising growth
- Modification of diet to ensure adequate carbohydrates to maintain euglycaemia
- Aiming to maintain and encourage age-appropriate feeding behaviours

Goals & Objectives of Nutritional Management

Goals:

- To attain and maintain normal nutritional status

Objectives:

- Maintain appropriate growth parameters according to WHO and CDC charts
- Maintain euglycaemia (BGLS > 4mmol/L)
- Optimise oral feeding
- Maintain and promote age- appropriate feeding behaviours

Stages of CHI

Diabetes Management

Preserve Beta cells

Preserve BGL

Medical Management – Nutritional considerations

Table 6 Medical management of HI

Medication	Mechanism of action	Typical dose	Side effects
Dextrose	Glucose source	IV 10%–50% dextrose, limit total fluid volume, use central access for >12.5% dextrose Enteral ≤10 mg/kg/min continuous ×24 hours or overnight	Volume overload
Glucagon			Nausea
Diazoxide			Salt and water retention Appetite suppression
Octreotide			Anorexia, nausea, abdominal discomfort, diarrhoea, Necrotizing enterocolitis [NEC]

Ackermann & Palladino 2015

Glucose Requirements

Age	Glucose mg/kg/min	Glucose g/ kg/hr Day	Glucose g/ kg/hr Night	CHI
Infants	8-9	0.5	0.5	15-20 mg/ kg/min
Toddler & Children	5-7	0.3-0.4	0.3-0.4	
Teens & Adults	2-4		0.2-0.25	

Anecdotally, additions of 2-6% poly joule to feeds is reasonable.
Excessive Poly Joule can cause osmotic diarrhoea, predispose to necrotising enterocolitis [NEC] or cause rebound hyperglycaemia.

- Poly Joule, a glucose only feed is often used to meet carbohydrate needs.
- Poly Joule can be added either to expressed breast milk or formula.
- The quantity added is based on carbohydrate requirement and is often commenced while titrating IV dextrose.

Protein Requirements

- As per the standard protein requirements.
- Ensuring appropriate energy: protein ratio of 7.5-12% from 0-12 months old is important for growth given likely high carbohydrate intake displacing percentage energy from protein. Aiming for Adequate Intake.
- Hyperammonemic hyperinsulinism (HI/HA) sensitive to protein, not to be eaten in isolation.

Age	Protein	Age	Protein
0-6 months	1.43g/kg (AI)	9-13 years (Boys)	0.95g/kg
7-12 months	1.6g/kg (AI)	9-13 years (Girls)	0.87g/kg
1-3 years	1.08g/kg (RDI)	14-18 years (Boys)	0.99g/kg
4-8 years	0.91g/ kg	14-18 years (Girls)	0.77g/kg

Feeding

- Mainstay of CHI management acutely is glucose supplementation
- Long-term use of a high carbohydrate diet will contribute to obesity and related metabolic and cardiovascular complications.
- In addition to promoting excessive weight gain, this can also complicate CHI management, due to rapid fluctuations in blood glucose levels.
- It is also common for infants and children with CHI to exhibit food avoidance behaviours, which may be due to medication side effects, oral aversion secondary to frequent forced feeding at a young age, or postoperative complications.
- Such food avoidance may necessitate enteral tube feeds to promote appropriate growth and glucose control.

Protein Load Challenge

- Often used to determine if there are any underlying metabolic diagnoses to consider.

If protein to be given via food (older child)

- Only plain, cooked chicken.

If protein to be administered as liquid (younger child)

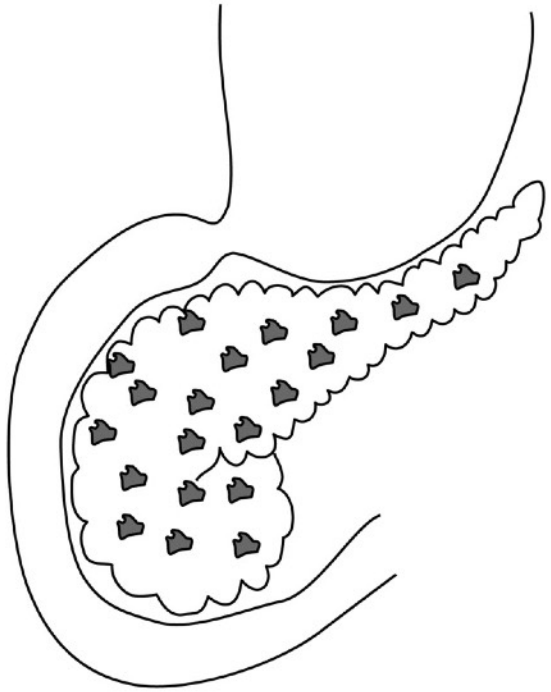
- A select Instant protein (To provide 1g/kg)

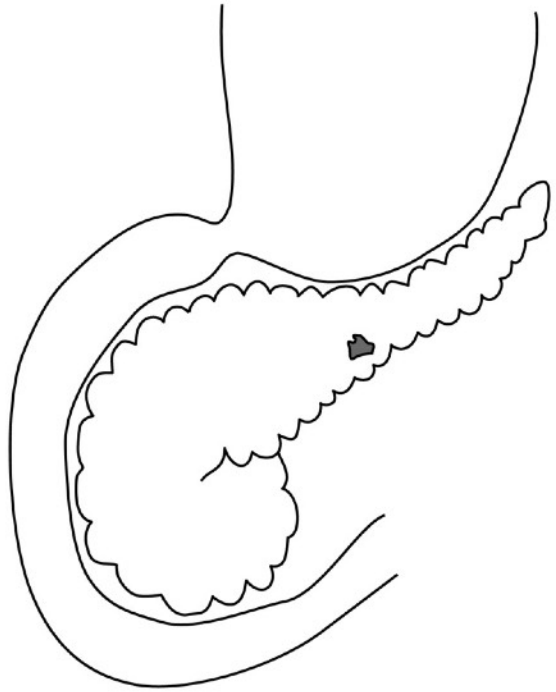
CHI due to KATP, GDH, or SCHAD mutations who consume a “normal” diet should have an oral protein tolerance test to assess for protein-sensitive hypoglycemia.

Surgical Management - Nutritional Considerations

PERT & vitABDECK, post-surgical insulin dependent diabetes mellitus

There is a risk of developing diabetes or pancreatic insufficiency with partial- pancreatic removal.

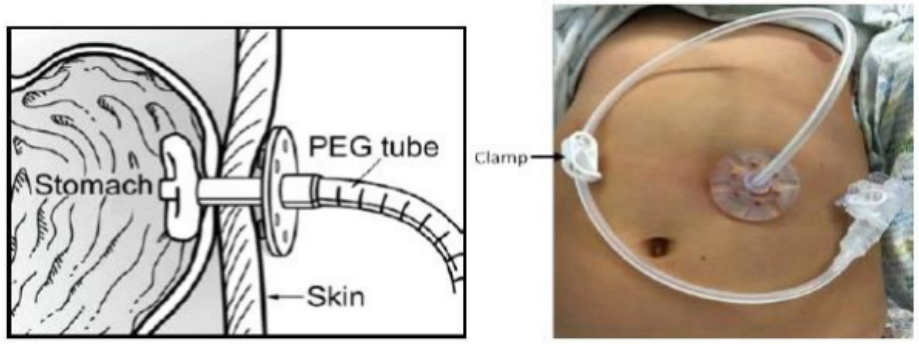




Diffuse disease:
- Entire pancreas is affected
- Associated with mutations in ABCC8/KCNJ11/GCK/GLUD1/ HNF4A/HADH and SLC16A1

Focal disease:
- A focal area of the pancreas is affected
- Associated with a paternal mutation in ABCC8 or KCNJ11 and paternal UPD encompassing 11p5.1 to 11p15.5 in the focal area

Post – Surgery



It is important to recognize that after a near-total pancreatectomy, the management can be complicated

Euglycaemia

- Diffuse HI, not requiring insulin or glucose support following pancreatectomy are still at risk of post- meal hyperglycemia and fasting hypoglycemia due to underlying beta-cell dysfunction.
- Blood glucose monitoring at home and regular follow-up are necessary.

Hypoglycaemia

- If postoperative hypoglycemia persists once the patient is on full enteral feeds, alternatives to IV GIR such as octreotide and/or enteral dextrose via PEG should be considered/ poly Joule overnight, EBM, formula

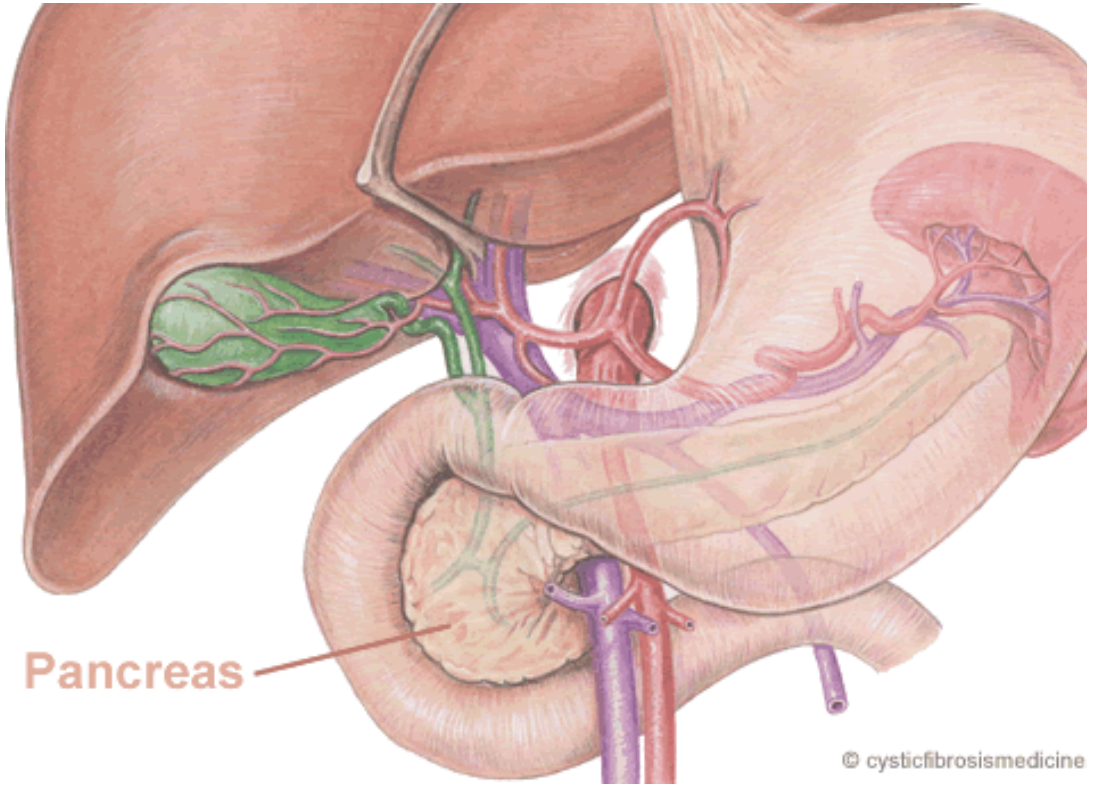
Hyperglycaemia

- Patients with pre- meal hyperglycemia after pancreatectomy will require subcutaneous insulin therapy.

Banerjee et al., Orphanet Journal of Rare Diseases. 2022;17:61.
Gastrostomies booklet - All you need to know. (2).pdf (rch.org.au)

Pancreas

- Two main functions:-
- Secretes digestive enzymes
- Secretes insulin

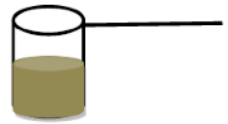


Pancreatic Insufficiency

Pancreatic exocrine insufficiency is observed in 50%–72% of patients with diffuse CHI who underwent near-total pancreatectomy

- ABDECK
- Creon, pancreatic enzymes replacement

Enzymes: Creon Micro



½ scoop with each breastfeed/formula feed

Vitamins: ABDECK

½ capsule daily

Insufficient digestive enzymes from pancreas

- Causes malabsorption of fat
- The infant is hungry, feeds frequently, fails to gain weight adequately, loose smelly bowel motions
- Malabsorption of fat soluble vitamins

Nutrition Information

Servings per package – 16
Serving size – 30g (2/3 cup)

	Per serve	Per 100g
Energy	**432kJ**	**1441kJ**
Protein	2.8g	9.3g
Fat		
Total	0.4g	1.2g
Saturated	0.1g	0.3g
Carbohydrate		
Total	18.9g	62.9g
Sugars	3.5g	11.8g
Fibre	6.4g	21.2g
Sodium	65mg	215mg

Ingredients: Cereals (76%) (wheat, oatbran, barley), psyllium husk (11%), sugar, rice, malt extract, honey, salt, vitamins.

Total Fat ▶
Generally choose foods with less than **10g per 100g.**
For milk, yogurt and icecream, choose less than **2g per 100g.**
For cheese, choose less than **15g per 100g.**

Saturated Fat ▶
Aim for the lowest, per 100g.
Less than 3g per 100g is best.

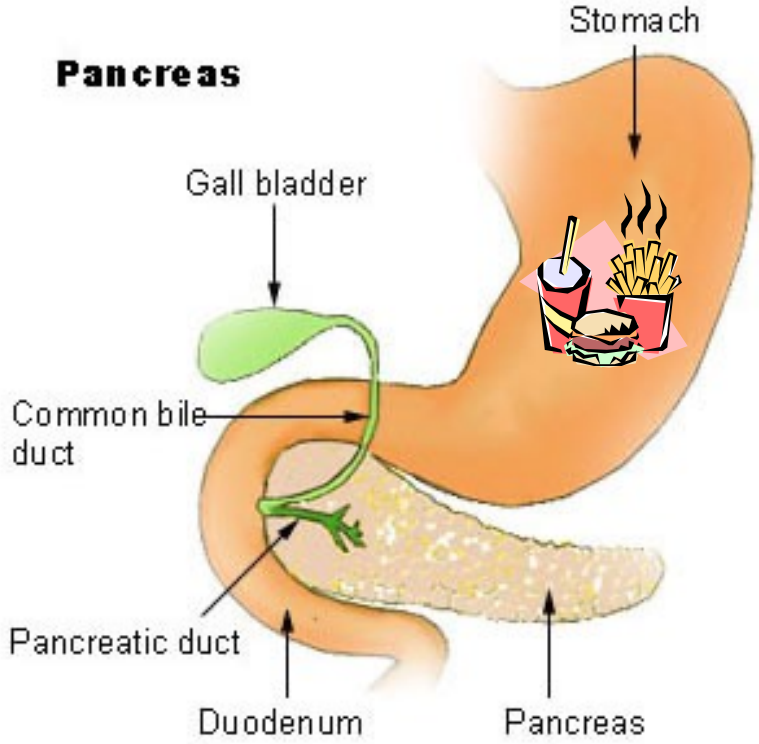
Other names for ingredients high in saturated fat: Animal fat/oil, beef fat, butter, chocolate, milk solids, coconut, coconut oil/milk/cream, copha, cream, ghee, dripping, lard, suet, palm oil, sour cream, vegetable shortening.

Fibre ▶
Not all labels include fibre.
Choose breads and cereals with **3g or more per serve**

Replacement pancreatic enzymes

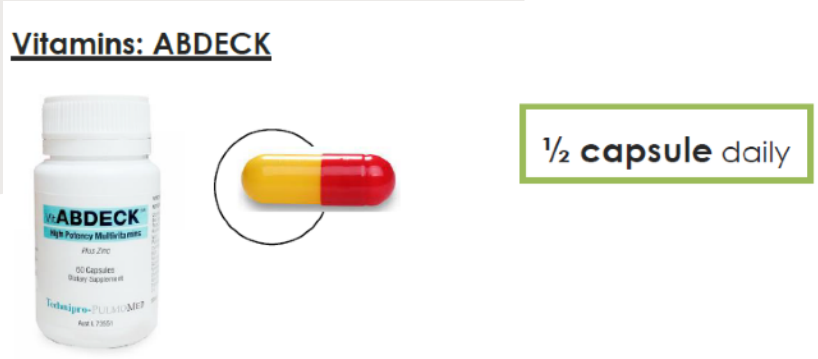
Children with fat malabsorption are given replacement enzymes to improve digestion.

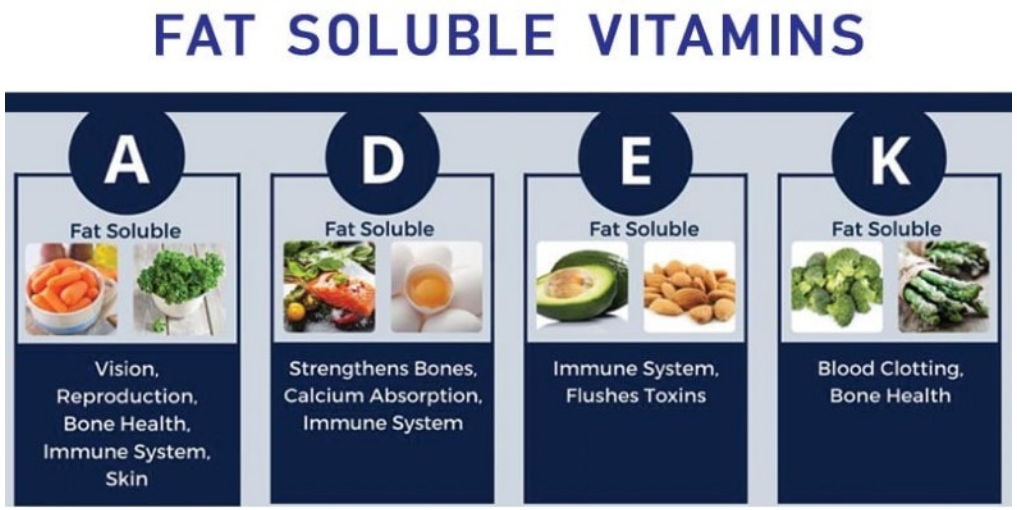
- Take enzymes before a meal so they are already in the small intestine, or duodenum, ready to digest food
- Solid foods may be added when your baby is ready, usually at 4-6 months of age. Single-grain infant rice cereal is a good first choice.
- Gradually add a variety of strained fruits, vegetables and meats. Plain strained meats contain more protein than combination dinners.
- Soft or pureed table foods may be introduced into your baby's diet at about 6-8 months of age.



Vitamins

- The vitamins in our diet are either:
  - water soluble (e.g. Vit C) or fat soluble
- Fat soluble vitamins: Vits A,D,E,K
- When malabsorption of fat is present, fat soluble vitamins are poorly absorbed
  - we give a vitamin supplement - Vit ABDEK – to counter-act this





Stages of CHI

Diabetes Management

Preserve Beta cells

Preserve BGL

Preservation of Beta- cell function

LOW GLYCAEMIC INDEX (CARBS)

HEALTHY FAT

LEAN PROTEINS

MOVEMENT

MONITORING BGL

Beltrand, et al., 2012. Diabetes Care 35: 2.

Preservation of Beta- cell function

Reduce the workload of the pancreas.

What are carbs?

Carbs give you energy for physical activity and help you grow.

How are carbs used by my body?

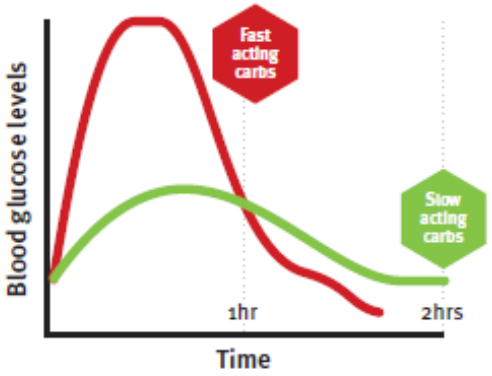
When carbs are eaten, they break down into glucose and enter the bloodstream.

Some carbs, like lollies, break down fast (fast-acting carbs and cause high BGL's.).

Carbs like wholegrain bread break down slowly (slow-acting carbs).

Over the day, it is better to eat carbs that break down slowly to help keep your blood glucose levels within target.

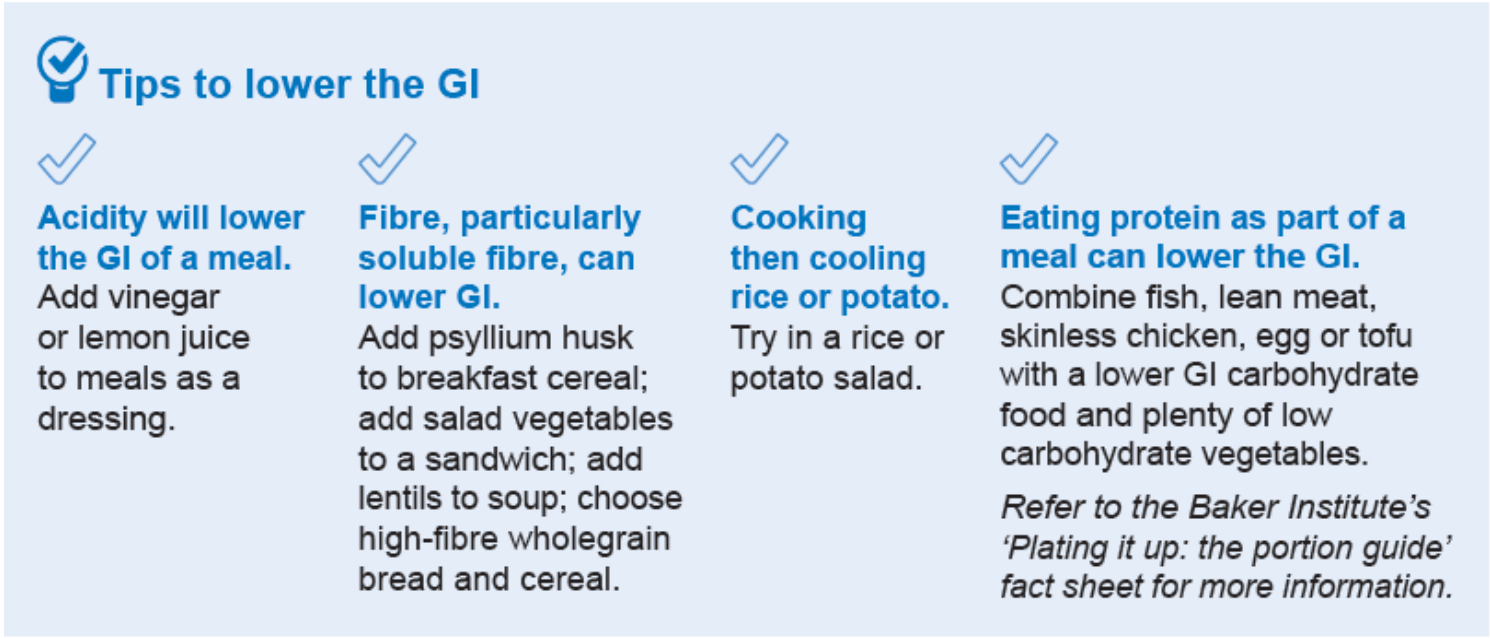
Slow vs fast acting carbs



Diabetes Won't Stop Me. 2022.
The Royal Children's Hospital, Melbourne, 2018.

Preservation of Beta- cell function

Reduce the workload of the pancreas.



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Preservation of Beta- cell function

Reduce the workload of the pancreas.

Limit these carb foods

Each food item shown contains 15 grams of carbohydrates, which is 1 carb portion.

- Biscuits
- Cakes, muffins, donuts, pastries
- Sugar, honey, golden syrup
- Jam, hazelnut spread
- Chocolates, lollies
- 2-minute noodles
- Regular soft drinks, cordial
- Fruit juice
- Flavoured milks
- Potato chips, corn chips
- Ice-cream, ice-blocks
- Custard, jelly

Rethink that sugary drink

What's the fuss about sugary drinks?

Sweet drinks like soft drinks, cordial, energy drinks, sports drinks, fruit juice and flavoured milky drinks are high in sugar and will raise your blood glucose levels quickly.

Diabetes Won't Stop Me. 2022.

Preservation of Beta-cell function: Dietary Strategies

- Lowering the GI of your diet will help prevent and manage overweight and obesity.
- Swapping high GI foods for low GI choices in the same food group or category at main meals and for snacks.
- The foods you choose should also be lower in saturated fat, moderate in sodium and higher in fibre.
- Serve size is also important, as eating too much of any kind of food, even healthy choices, may make you put on weight.

Diabetes Won’t Stop Me. 2022.

Drink water

Drink lots of water

Your body needs water to:

- Keeps our body hydrated
- Keeps our kidneys happy
- Help our joints to move well
- Help to digest our food and go to the toilet
- Help with weight loss

Tips to drink more water

- Freeze a water bottle so it stays cold during the day
- Carry a water bottle with you everywhere
- Set a daily goal
- Set some reminders or alarms on your phone
- Use a water bottle that shows how much you drink
- Have a glass of water at each meal time

How much water do I need to drink?

1 – 3 year old	4 – 8 year old	9 – 13 year old	14 – 18 year old
1L (4 cups)	1.25L (5 cups)	1.5L (6 cups)	1.75L (7 cups)

Diabetes Won’t Stop Me. 2022.

Preservation of Beta- cell function

Reduce the workload of the pancreas.

- Small, frequent low GI carbohydrate meals are often required to maintain glycaemia
- Dietary education regarding carbohydrate serves and low GI food choices (as per children with type 1 diabetes) is given to ensure appropriate dietary quality and consistency of carbohydrate intake.
- Complex carbohydrates in the form of uncooked cornstarch are regularly required intermittently throughout the day and or night to maintain glycaemia.
- It is not advised to use uncooked cornstarch in children under the age of two due to immature pancreatic amylase reducing its efficacy and gut upset.
- In those > 2 years of age, effectiveness is variable.

Beltrand, et al., 2012. Diabetes Care 35: 2.

Foods to choose

- Choose leaner cuts of meat
- Cut any fat off before cooking
- Use spray oil when frying
- Cook from fresh, rather than buying ta
  or premade meals
- Choose reduced-fat, low-fat or no-fat n
  yoghurt and cheese.
- Oily fish (Fresh, frozen or canned)

Cut back on bad fats

Eating foods low in bad fats (saturated and trans-fats) can help keep your cholesterol down.

Try to swap these bad fats with foods that have good fats.

Foods with lots of bad fats

- Fatty meats
- Full-fat dairy products
- Butter
- Coconut oil
- Palm oil
- Deep-fried takeaway foods
- Pies, biscuits, buns, pastries, cakes

Preservation of Beta- cell function: Healthy Fats & Lean Protein.

- The foods you choose should also be lower in saturated fat, moderate in sodium and higher in fibre.
- Fear of hypo and assoc. risk can cause stress for the family and can lead to disordered eating in children or overfeeding, which, together with limited physical activity, may result in obesity.
- Many children become overweight in early life, although this may improve over time.

Diabetes Won’t Stop Me. 2022.

Preservation of Beta-cell function: Healthy Weight

- CHI is difficult to manage nutritionally due to the high carbohydrate requirement causing issues with weight gain and feeding.
- Aggravating factors such as obesity lead to a risk of developing dysglycaemia.
- Obesity from frequent feeding may negatively impact on the child's body image.

Diabetes Won't Stop Me. 2022.
Banerjee et al., Orphanet Journal of Rare Diseases. 2022;17:61.

Stages of CHI

Diabetes Management

Preserve Beta cells

Preserve BGL

Diabetes Management: hyperglycaemia

INSULIN

CARBOHYDRATE COUNTING

LOW GLYCAEMIC INDEX

HEALTHY FOOD

ACTIVITY

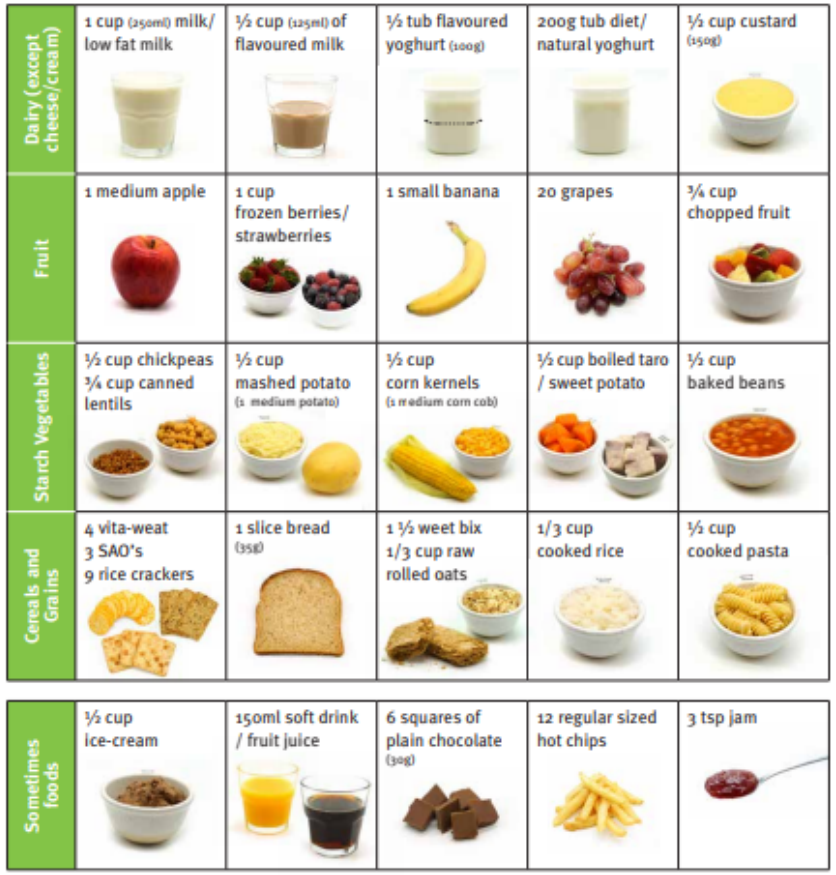
MONITOR BGL

Banerjee et al., Orphanet Journal of Rare Diseases. 2022;17:61.

Carbohydrate Counting

What is a carb portion?

1 carb portion = 15g carbs
Measure and weigh your food.
Check labels as brands vary.



Carb Counting

Carb counting is a way to estimate the amount of carbs in your food. This can help you better manage your blood glucose levels. Together with your dietitian, you will learn how to carb count.

Diabetes Won't Stop Me. 2022.

Diabetes Management

Quality Carbohydrates

How are carbs used by my body?

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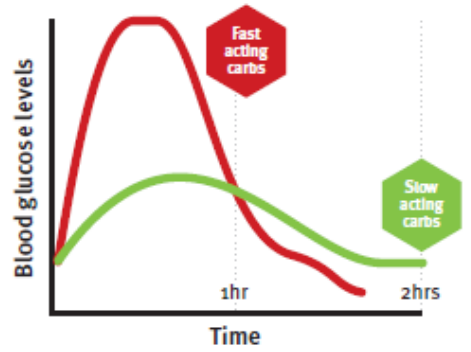

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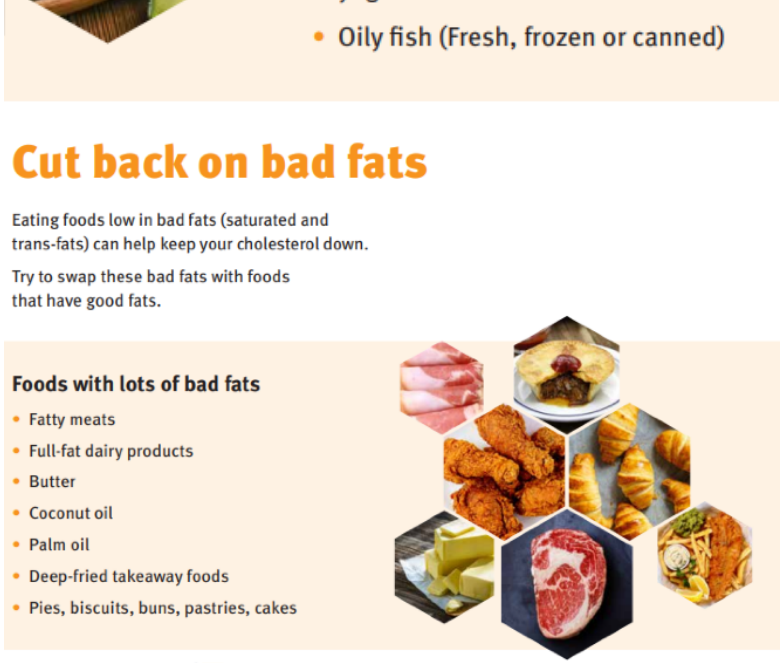

Diabetes Won't Stop Me. 2022.

Preservation of Beta- cell function: Healthy Fats & Lean Protein.

- To promote healthy weight management

Preservation of Beta- cell function: Healthy Fats & Lean Protein.

- To promote healthy weight management
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- The foods you choose should also be lower in saturated fat, moderate in sodium and higher in fibre.
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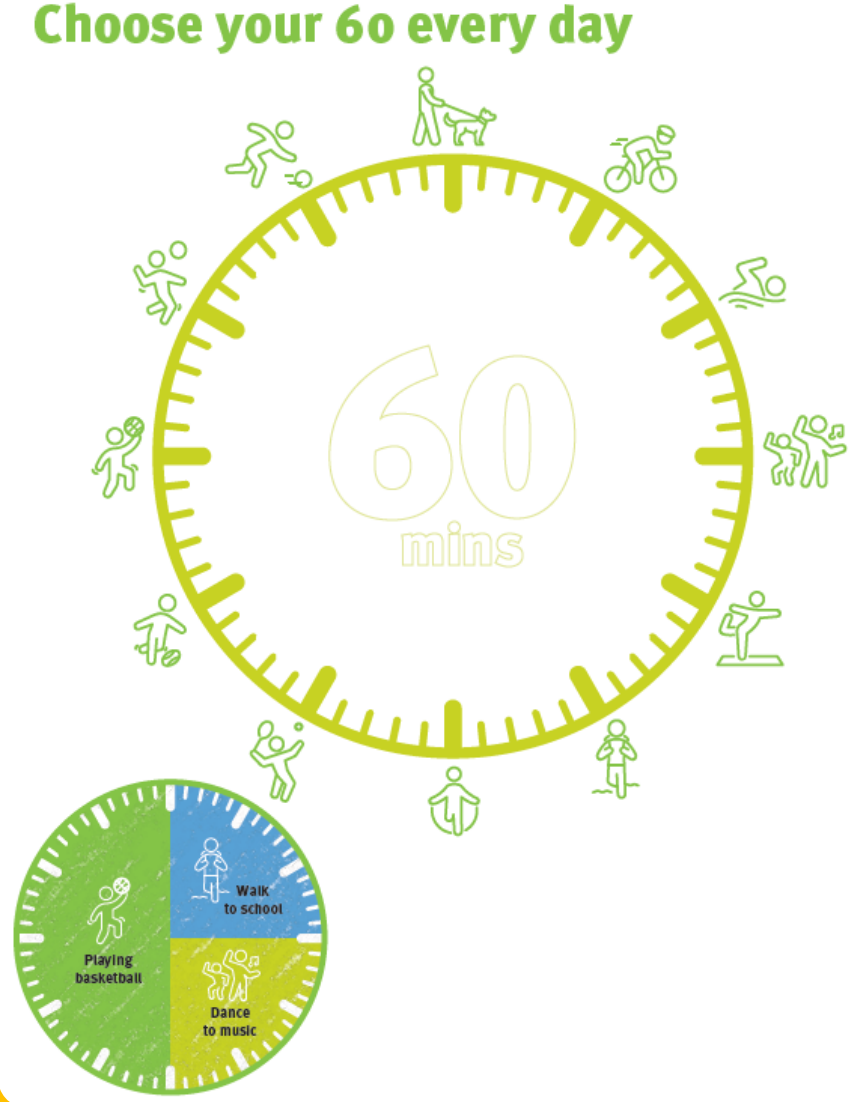


Movement

- To promote healthy weight management
- Ensure hypo treatment available.

Diabetes Won't Stop Me. 2022.

Feeding Difficulties

The feeding problems most frequently reported were

- Poor appetite (40%)
- Refusal to eat (39%)

More than 26% of families reported:

- gastroesophageal reflux, gagging, problems with texture, vomiting and slow eating.
- Poses a significant burden for families, requiring careful meal planning.

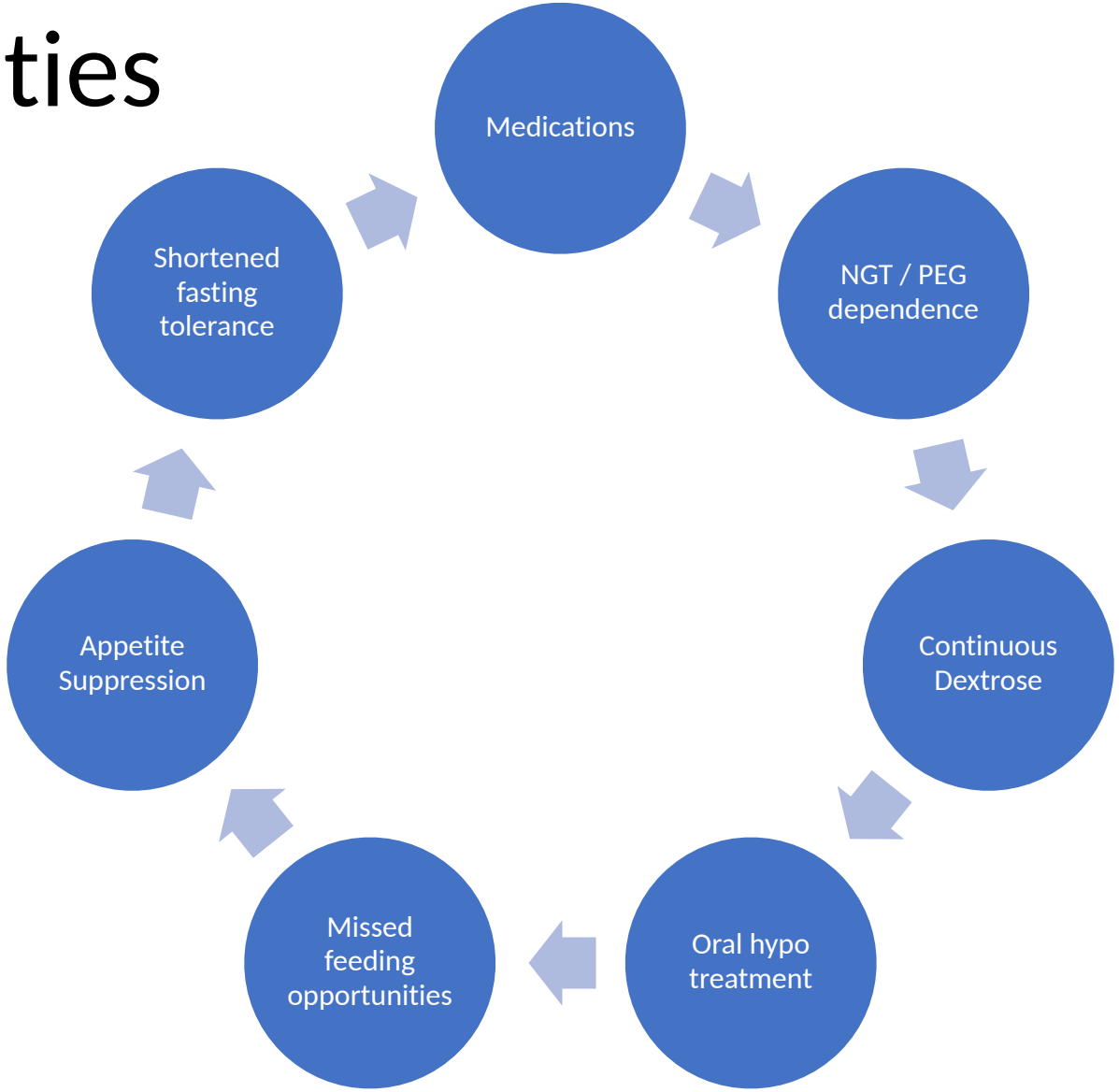
Banerjee et al. Orphanet Journal of Rare Disease. 2022;17:61.

Feeding difficulties

As many as 60%

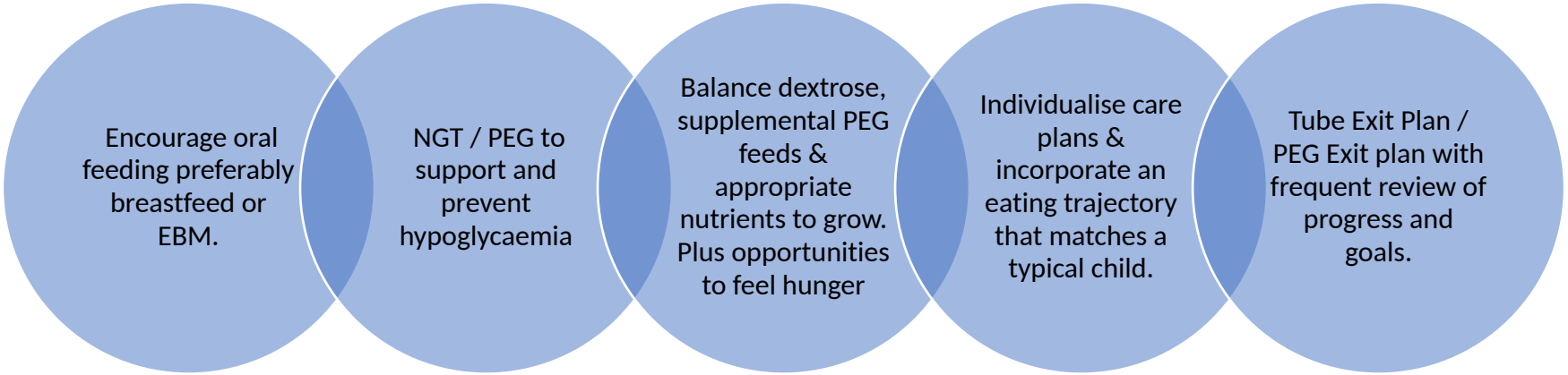
In children whose HI can be controlled without surgery, dextrose may be part of their management plan, along with medication and dietary changes.

Despite best efforts, some children with HI do have an aversion to eating by mouth. 'It all depends on the child and there are varying degrees of aversion.



Children's Hospital of Philadelphia [CHOP] Jul 20, 2022.

What are strategies to help with feeding?



Focus on normal, age and developmentally appropriate eating and nutrition.

Nutrition needs to change for a CHI baby just like it would for a typical infant.

They need to start cereals at 6 months and then solids foods. This can be later if the infant is born pre- term.

Children's Hospital of Philadelphia [CHOP] Jul 20, 2022.

Feeding Therapies

- Can help children learn to chew and swallow, how to use a spoon, how to get used to soft foods and different textures.
- Coach parents to encourage the kids on dextrose and /or tube feedings to maximise their oral intake with healthy foods.
- Feeds through the G- tube will ensure the child receives proper nutrition and calories for healthy growth until they can consume enough food by mouth.
- They may need to use the feeding tube only at night or they may need feeds more frequently.
- While the exact age varies, by 4 years old many children are eating the typical three meals and two snacks a day and no longer need to use the G-tube for nutrition, although some children may still receive dextrose via the G tube.
- Child can wear a small backpack that holds their dextrose, as well as formula sometimes.

Children's Hospital of Philadelphia [CHOP] Jul 20, 2022.

Thank you

- To the families, bubs and children and medical teams that I learn from everyday.
- Please continue to discuss your goals for your children with CHI with your multidisciplinary teams.

- Thank you for inviting me to talk today.