



**express™**

A practical guide to transitioning  
from a second-stage protein substitute  
to express in disorders of protein metabolism



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Supporting education in the  
dietary management of rare diseases

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## 1.0 Disclaimer and important information

### **Purpose**

This practical guide is for the use of express™ in the dietary management of children and adults with disorders of protein metabolism.

### **Intended users**

This practical guide is:

- for use by healthcare professionals working with children and adults diagnosed with disorders of protein metabolism
- **not** for use by parents/caregivers of children or adults with disorders of protein metabolism or patients themselves
- for general information only and must not be used as a substitute for professional medical advice

### **Target population**

This practical guide is for use in children with diagnosed/proven disorders of protein metabolism.

### **Product information**

Express is a Food for Special Medical Purposes (FSMP). Any product information contained in this practical guide, although accurate at the time of publication, is subject to change. The most current product information may be obtained by referring to product labels and [www.vitafloweb.com](http://www.vitafloweb.com). Please refer to these sources for information regarding allergens.

Introducing and adjusting express is dependent on the individual. Practical examples are given in this guide; however, it is the responsibility of the managing health care professional to use clinical judgement to introduce and adjust express in the most appropriate way for the individual and it may not always be appropriate to use the practical guide.

### **Important notice for express**

- Use under medical supervision
- Not suitable for use as a sole source of nutrition
- Suitable from 3 years of age
- Must only be consumed by individuals with disorders of protein metabolism
- Diet must be supplemented with natural protein, water and other nutrients in prescribed quantities
- For enteral use only

### **Disclaimer**

The information contained in the practical guide is for general information purposes only and does not constitute medical advice. The practical guide is not a substitute for medical care provided by a licensed and qualified health care professional and Vitaflo does not accept any responsibility for any loss arising from reliance on information contained in this guide.

This practical guide should be read in conjunction with local, national and international guidelines and best practice. Information contained within the guide is based on the most recent scientific evidence available on the management of protein disorders as of June 2022.

This practical guide does not establish or specify particular standards of medical care for the treatment of any conditions referred to in this practical guide.

Vitaflo International Limited does not recommend or endorse any specific tests, procedures, opinions, clinicians or other information that may be included or referenced in this practical guide.

## 2.0 Introduction

Children experience many changes to their individual nutritional needs as they grow from infants to toddlers, all the way through to adulthood and the elderly years. Childhood presents periods of rapid growth, increased requirements for specific nutrients<sup>1</sup>, as well as development of behavioural and social skills associated with food and eating<sup>2</sup>.

For children with disorders of protein metabolism, to accommodate these changes, protein substitutes are designed to benefit children at different life stages. For example, micronutrient requirements change as children grow, which is reflected in the nutritional profiles of protein substitutes intended for different ages. Children change developmentally, so at weaning age, a semi-solid may be more appropriate than a liquid protein substitute. It is important that protein substitutes cater for life stages and requirements because they can provide over 75% of protein requirements<sup>3,4</sup> depending on the condition, so contribute significantly to the overall quality of a child's diet.

The change from one protein substitute to another can be an anxious time for children and their parents/caregivers, especially because we know that abnormal feeding behaviours and neophobia are prevalent in inherited metabolic disease<sup>5-8</sup>. However, it is important that the protein substitute they are taking is the best choice for them.

The express range provides a protein substitute that meets the nutritional needs of a child from 3 years old onwards, as well as accommodating social and practical needs as children grow older. This practical guide supports the transition from a second-stage protein substitute to express to help make the process of change as smooth as possible.

### 3.0 What is express?

Express is a range of powdered protein substitutes, free from the offending amino acid(s)\*. Express contains essential and non-essential amino acids, carbohydrate, vitamins, minerals and trace elements.

\*None added. Trace amounts may still be present in other ingredients (<10mg per 100g / <4mg per serving)

#### Key features of express

Available for PKU, MSUD, HCU and TYR.

Available in 15g PE and 20g PE pre-measured sachets for PKU and 15g PE for other conditions





Added starch to replicate the texture and mouthfeel of gel™ and explore™ ranges



Available in unflavoured and flavoured options (PKU only) and compatible with flavour pacs (see table below)

Contains full micronutrient profile

Can be prepared to a low volume semi-solid or as a mini or flexi-drink

Pack Size				
	PKU	MSUD	HCU	TYR
Express 15	✓	✓	✓	✓
Express 20	✓			
Flavour Options				
Unflavoured	✓	✓	✓	✓
Orange	✓	Available in unflavoured only. However, can be used in combination with Vitaflo flavour pacs™.		
Tropical	✓			
Lemon	✓			

PKU express (Unflavoured)		Nutritional Information		
Nutrient	Units	Per 100g	Per 15g PE	Per 20g PE
Energy	kJ	1292	324	434
	kcal	304	76	102
Protein	g	60	15	20
Fat	g	0	0	0
of which saturates	g	0	0	0
Carbohydrate	g	16	4	5.4
of which sugars	g	0.5	0.13	0.17
Fibre	g	0	0	0

Other express products will have similar macronutrient profiles but will differ in their amino acid profiles.

## 4.0 Using express

### Choosing the best protein substitute for your patient.

Protein substitutes are designed to meet nutritional and practical requirements at each developmental stage of childhood as shown in the graphic below:

#### Metabolic formula for infants



These products are similar in composition to standard infant formula, without the offending amino acid(s). They are designed to meet rapid growth demands of infancy by providing all macronutrients and a micronutrient profile to meet the needs of this first life stage.

#### Second-stage protein substitute



When an infant starts to introduce complementary foods, energy provided by the protein substitute remains important, although there is a reduced emphasis when compared to formula. This allows the child to obtain more energy from foods. The micronutrient profile is designed to meet requirements of this life stage and provide nutrients that are compromised by a low protein diet. The consistency mirrors that of first complementary foods.

#### Protein substitute for early and late childhood



As children progress from toddlers, the protein substitute provides a more concentrated source of amino acids, designed to meet nutritional requirements when the solid diet is fully established. It can be used throughout childhood, adolescence, and adulthood.

**Why change from a second-stage protein substitute to express?**

Children often become very used to their protein substitute and it can be difficult to persuade them, and their families, to make the change - especially if they feel that their current protein substitute meets all their needs. The information below describes why it is advantageous to transition to the most appropriate protein substitute, and how this is beneficial to the long-term nutritional management of your patient.

As children grow from toddlers to school children, their needs change nutritionally and developmentally and preferences and lifestyle factors develop. This means that their second-stage protein substitute gradually fails to meet their needs, and transition to a more age-appropriate protein substitute, such as express, is indicated.

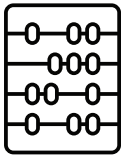
**Changing needs in early childhood which indicate transition from a second-stage protein substitute to a more age-appropriate protein substitute:**



**Nutritional**

**Energy:** Once children have established complementary feeding, they may need a protein substitute that provides fewer calories to allow energy to come from the diet instead. Express is lower in energy than second stage protein substitutes so can be used for this purpose.

**Micronutrients:** Requirements for particular micronutrients change throughout childhood with some being more important at different life stages. Whereas second stage protein substitutes are designed for infants and young children, express has been specifically formulated so that when given at the recommended intake for meeting age specific protein requirements, it also provides age specific recommended intakes of micronutrients (where intakes from the foods permitted in a low protein diet, are likely to be low).



**Developmental**

Concentrated second-stage protein substitutes such as the gel and explore ranges are designed to meet developmental needs of complementary feeding with particular consideration to **volume** and **consistency**.

**Volume:** As protein, and hence protein equivalent requirements increase, a protein substitute more concentrated in amino acids and micronutrients is beneficial. Express prepared to a semi-solid, is beneficial to children who are used to their second-stage protein substitute in this format and do not want to change. However, as children develop and grow, express can be prepared to larger volumes if children wish to take their protein substitute as a drink.

**Consistency:** Express has a similar mouthfeel when prepared as a semi solid, to the gel and explore ranges. This is designed to help children transition from their semi-solid second-stage protein substitute more easily.

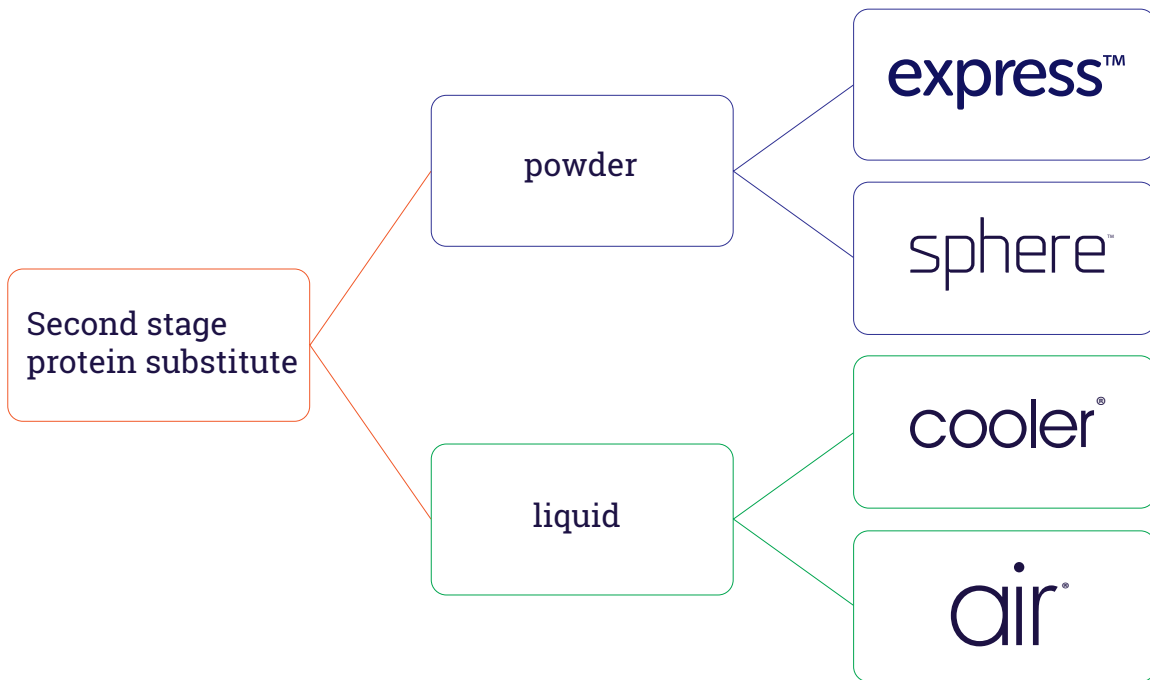


**Preferences and lifestyle**

As children start school and develop their social lives, preferences and lifestyle factors start to influence suitability of their protein substitute. Their second stage protein substitute may be too small a pack size, packaged to appeal to younger children or lack flexibility with volume. To help compliance, express is designed to fit in with their lifestyle more easily as they become older.

## 5.0 The process of transition

An ideal time to transition is when a child is school aged, and the change can be part of this natural change and engrained in the developmental process. Beyond this age, transition tends to become more difficult to achieve and children become less receptive. There is a choice of protein substitutes available to make the process easier and every child is different, having their own preferences. The express range is just one option. The graphic below shows these options\*.



\*Not all products available for all conditions

Progressing from a second-stage protein substitute to a powdered protein substitute such as express can make the change easier, especially for those children who struggle to transition to a ready to drink liquid. Express can be prepared as a semi-solid similar to a second-stage protein substitute, and then increased in volume to a mini drink and subsequently a flexi drink, according to preference.



### spoonable semi-solid

1. Empty the contents of the express sachet into the shaker.
2. Add 10-20ml or 2-4 teaspoons of water.
3. Stir well for 10 seconds and eat with a spoon.



### mini drink

1. Empty the contents of the express sachet into the shaker.
2. Add 80-100ml of water.
3. Secure lid and shake well for 10 seconds.



### flexi drink

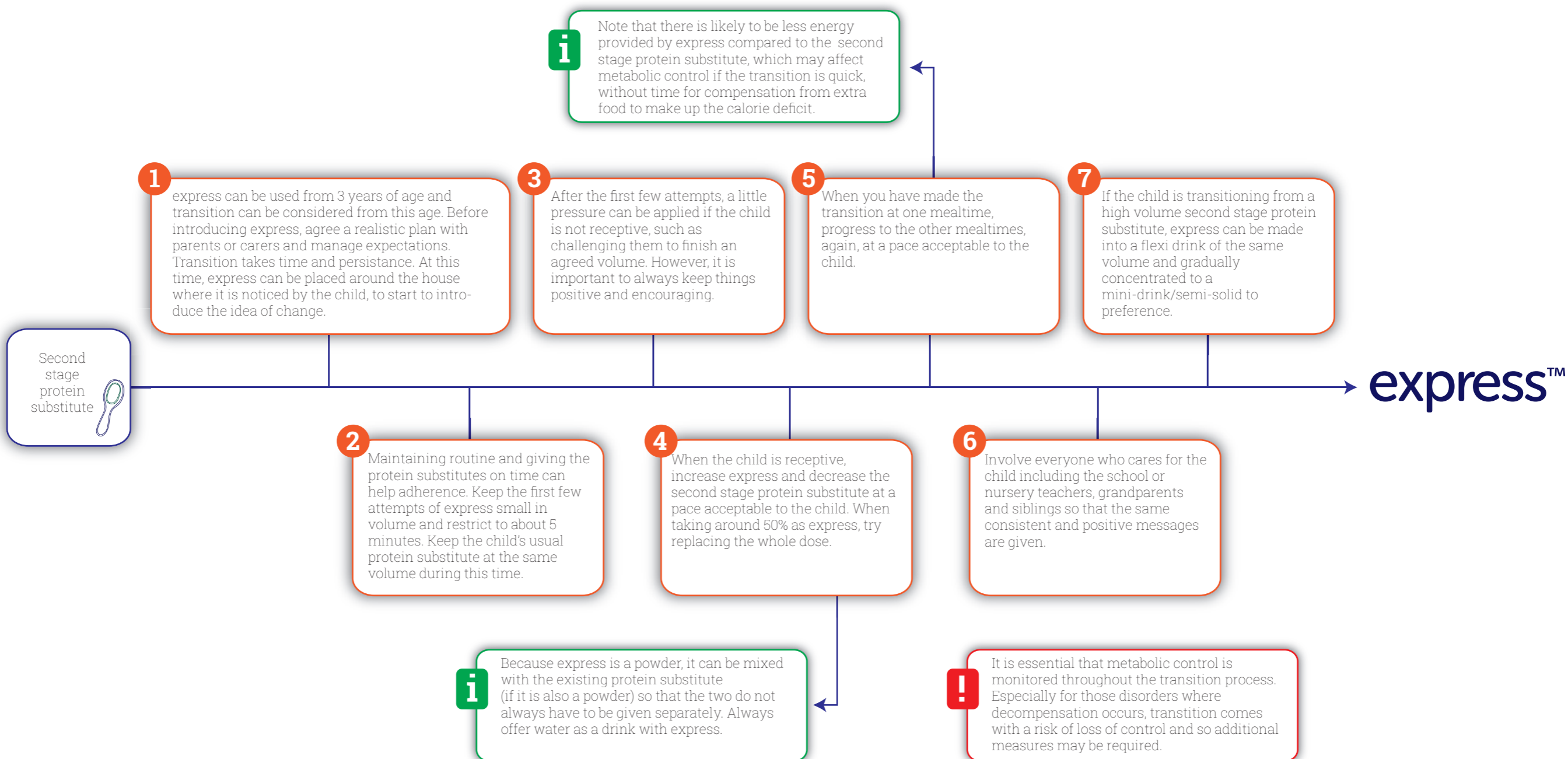
1. Empty the contents of the express sachet into the shaker.
2. Add as much water as you like!
3. Secure lid and shake well for 10 seconds.

Liquid ready-to-drink protein substitutes are often preferred by older children due to convenience, packaging and resemblance to commercial drinks<sup>9</sup>. Express is interchangeable with the cooler, air and sphere ranges and so transition to these ranges is easy and can be combined. Please refer to individual data cards for macronutrient and micronutrient details.



### Making the transition

The graphic below shows the stages of transition when moving from a second stage protein substitute to express. Transition will take various lengths of time for different children. It is normal for a child to need lots of encouragement, supervision and reassurance throughout the process and perseverance is needed by all carers for the child.



## Managing fussy eating and food neophobia

Fussy eating and food neophobia can affect all children, but research in inherited metabolic disorders shows high prevalence<sup>7</sup>. Research specifically in children with PKU shows higher incidences of both food neophobia<sup>5,6</sup> and fussy eating behaviours<sup>8</sup>. Tonon et al reported that these behaviours persist into adulthood in individuals with PKU at a higher rate than controls<sup>10</sup>. It is common therefore, for children of all ages to resist trying something new. This may happen when introducing a new protein substitute. Some strategies for managing food neophobia in children with disorders of protein metabolism are shown below with tips for taking their protein substitute.



**Food neophobia** – avoidance and rejection of new foods with an unwillingness to try. It is derived from a survival mechanism to avoid poisonous foods and other items.<sup>11,12</sup>

**Fussy/picky eating** – rejection of a wide variety of familiar foods which can be based on flavour, look and texture and may have been previously accepted.

Why are children with metabolic conditions at more risk of fussy eating and food neophobia?	Strategies for easier transitioning to, and maintaining adherence to express
<p>Breastfeeding is reduced in volume and often longevity in infants with a disorder of protein metabolism, reducing introduction of new flavours through breastmilk. This may increase the risk of fussy eating later on.</p> <p>A restricted diet can adversely affect eating patterns and feeding development.</p> <p>Gastrointestinal problems associated with metabolic conditions can impact on a child's relationship with food.</p> <p>Parents of PKU children, compared to controls, report more stress and anxiety related to feeding<sup>13</sup> which can impact negatively on child feeding behaviour. Emotional stress is also reported in parents of MSUD children<sup>14</sup> as well as PKU.</p> <p>Children with a metabolic condition may be given meals separately to other family members or friends<sup>8</sup>, reducing the opportunity for social feeding development.</p>	<p>Encourage parents/carers to: Treat all protein substitutes with the same importance as a medicine<sup>3</sup>.</p> <p>Supervise the child until the protein substitute is completely finished<sup>3</sup>.</p> <p>Give before meals to avoid complaints of being full.</p> <p>Avoid giving when the child is tired and uncooperative.</p> <p>Avoid mixing express with foods – allow the child to enjoy food.</p> <p>Be firm, but encouraging – star charts and rewards may help with this.</p> <p>Keep to the minimum time possible (less than 5 minutes is ideal) – timer games may help with this.</p> <p>Maintain routine, consistency<sup>3</sup> and expectations of the child.</p> <p>Involve nursery and schools where children may be more receptive to new foods than with their family.</p>

## 7.0 Example meal plan



<b>Condition:</b>	Hereditary tyrosinaemia type 1
<b>Age:</b>	9 years
<b>Total protein</b>	55g/day (2g/kg) requirement:
<b>Natural protein allowance:</b>	10g/day
<b>Protein equivalent requirement:</b>	45g/day

	Intake	Energy (kcal)	Protein/PE (g)
<b>Breakfast</b>	30g permitted breakfast cereal (2g protein exchange) 200ml protein-free milk alternative	270	2
	1 sachet TYR express 15 made into a flexi drink	76	15
<b>Mid-morning snack</b>	Melon and peach slices 1 rice cake (1g protein exchange)	100	1
<b>Lunch</b>	1 small low protein bread roll with butter Low protein cheese Packet of crisps (1g protein exchange) Cereal bar (1g protein exchange)	550	2
	1 sachet TYR express 15 made into a spoonable semi-solid	76	15
<b>Mid-afternoon snack</b>	1 large strawberry jelly pot (gelatine and aspartame free)	120	-
<b>Evening meal</b>	Medium jacket potato with butter (3g protein exchange) 15g peas (1g protein exchange) 50g baby carrots 1 low protein veggie sausage (1g protein exchange)	380	5
	1 sachet TYR express 15 made to a flexi drink and flavoured with chocolate syrup	76	15
<b>Supper</b>	1 small packet of raisins 200mls protein-free milk alternative	180	-
		<b>1828</b>	<b>55</b>

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**Notes**

Notes



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