



An introduction to complementary feeding and
transitioning to a concentrated second stage protein
substitute gel™ in infants with an amino acid disorder.



Supporting education in the
dietary management of rare diseases

Important Information

This practical guide covers an introduction to complementary foods and transitioning to a concentrated low volume spoonable protein substitute (**gel**), in infants diagnosed with an amino acid disorder.

This guide should be read in conjunction with local and national protocols. It is based on best practice over the last 20 years of introducing a concentrated spoonable protein substitute to infants with an amino acid disorder from around six months of age⁽¹⁾.

In this guide the following amino acid disorders are considered: **phenylketonuria (PKU), maple syrup urine disease (MSUD), homocystinuria (HCU) and tyrosinaemia (TYR)***.

All of these amino acid disorders share the same principles of dietary management. Therefore, the management of the introduction to complementary foods and the transitioning to a concentrated protein substitute is considered in one practical guide.

For organic acidaemias see section 5.3.

The guide **does not cover** the clinical management of any these conditions. Refer to section 4.2 for the international management guideline references.

This practical guide is:

- Only to be used by healthcare professionals.
- Not for use by patients or their families/caregivers.
- For guidance only and must not be used as a substitute for professional medical advice or management.

* In this guide we have used the abbreviation TYR to represent tyrosinaemia instead of HT, as Vitaflo uses this within the names of their protein substitutes for tyrosinaemia, e.g. TYR gel.

Important notice

The **gel** range are foods for special medical purposes (FSMP) for the dietary management of various amino acid disorders - including PKU, MSUD, HCU and TYR (see appendix 5.4 for full **gel** range).

Use under medical supervision.

Not suitable for use as a sole source of nutrition.

Suitable from 6 months to 5 years of age[#].

For enteral use only.

gel must only be consumed by individuals with a proven amino acid disorder under supervision by the managing clinician or dietitian. Natural protein must be given in prescribed amounts to meet requirements.

Introducing and adjusting gel is based on the individual needs of the child. While practical examples are given in this guide, local practice may vary.

It is the responsibility of the managing healthcare professional to use their clinical judgment to introduce and adjust gel in the most appropriate way for their individual patients.

The information contained in this 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 healthcare professional and Vitaflo® International Ltd does not accept any responsibility for any loss arising from reliance on information contained in this guide.

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

[#] Although gel can be used up to 10 years of age, it is best suited to be used from 6 months to 5 years as there are more age appropriate products in the Vitaflo range for children over 5.

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Key to symbols and terms used throughout this practical guide

| Symbol | Term | Definition | Precursor amino acid free from |
|---|--|--|--------------------------------|
| | PKU | Phenylketonuria | Phenylalanine |
| | MSUD | Maple Syrup Urine Disease | Valine, Isoleucine, Leucine |
| | HCU | Homocystinuria | Methionine |
| | TYR | Tyrosinaemia type I, II, III | Tyrosine, Phenylalanine |
| | PE | Protein Equivalent - the amount of protein provided by a protein substitute. Total protein intake = PE intake from protein substitute + natural protein intake. | |
|  | | Breast milk/standard infant formula. | |
| | Precursor amino acid(s) | This refers to the amino acid(s) that can not be catabolised due to an enzyme defect in metabolic pathway (see table 1 for more detail). | |
|  | Precursor free amino acid formula for infants | Precursor free amino acid formula for infants refers to the condition-specific formula for infants free from the appropriate precursor amino acid(s) for the particular disorder being managed. | |
|  | gel | gel refers to the condition-specific gel free from the appropriate precursor amino acid(s) for the management of the particular amino acid disorder. | |
| | A measured amount of protein | <p>This refers to a measured amount of natural protein from food to provide a prescribed quantity of the relevant precursor amino acid(s).</p> <p>Systems for allocating prescribed amounts of the precursor amino acid(s) vary internationally. In the UK the amino acid is allocated in the form of a daily food exchange system, with different exchange systems for each of the 4 disorders.</p> <p>Some countries use allowance systems, where the individual counts the milligrams of the precursor amino acid(s) they consume as part of their daily allowance.</p> <p>For the simplicity of this guide, the prescription of precursor amino acid(s) is referred to as a measured amount of protein.</p> | |
|  | Very low protein foods | <p>Very low protein foods refer to foods that are very low in protein (precursor amino acid(s)) that can be incorporated into the diet without having a direct impact on target blood amino acid concentrations.</p> <p>Where an 'allowance' system is being used, every mg of precursor amino acid(s) may be counted as part of the patient's daily allowance.</p> | |



Practice on calculating measured amounts of protein and very low protein food intake may vary, adhere to local country / international guidelines.

When complementary foods and a second stage protein substitute are introduced to an infant with an amino acid disorder, feeding can become more complex for both caregivers and healthcare professionals. These changes must not only support the child's developmental and nutritional needs but also maintain satisfactory growth and optimal metabolic control. New tastes are experienced through foods, and breast milk or standard infant formula are gradually replaced by measured amounts of protein containing solids. When choosing an appropriate protein substitute, it is always important to consider the age and developmental stage of the child.

One of the challenges of introducing complementary foods and a second stage protein substitute is to find a balance between giving a variety of very low protein foods and measured amounts of protein, and maintaining sufficient intake of the precursor free protein substitute to meet the infant's requirements. Historically, protein substitutes developed for young children were powders, designed to be reconstituted as liquids. When given to infants aged from 6 months, the high volume of liquid protein substitute may reduce appetite, delaying solid food progression. Administering the second stage protein substitute as a concentrated spoonable semi-solid, encourages appetite and therefore supports introduction of solid foods⁽¹⁾.

Establishing the second stage protein substitute at the right time is essential to ensure it is accepted. Introducing **gel** requires a gradual and progressive approach, increasing the amount according to individual needs. This enables the reduction of precursor free formula for infants, allowing appetite for foods. To support this process, caregivers will need education, ongoing support and practical guidance with clear instructions to help them with the challenges of complementary feeding and establishing the child onto a second stage protein substitute.

Good feeding practices for infants without an amino acid disorder should be followed when possible (see section 5.1). Differences occur with the types of first foods introduced and also the introduction of protein containing foods. To aid the acceptance of solid foods and support long term metabolic control, it is essential that this process is carefully managed⁽²⁾.

Role of gel

- **Provides a concentrated source of PE and micronutrients, which in the diet for amino acid disorders cannot be provided or are limited due to the restriction of protein rich foods.**
- **Ensures good feeding practice recommendations can be met, by reducing liquid intake and allowing appetite for solid foods to develop⁽³⁾.**
- **By helping meet infant feeding developmental milestones, gel may reduce potential feeding problems and associated anxiety for both parents/caregivers and the child.**

VitaFlo has developed this practical guide for healthcare professionals on the use and introduction of **gel** for infants at around 6 months of age for either **PKU, MSUD, HCU** or **TYR**. By following the suggested stepwise system for introducing **gel**, it should help establish a suitable framework for protein substitute administration in infants and create a sound foundation for future years.

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1.1 What is gel?

gel is a concentrated powdered protein substitute free from the appropriate precursor amino acid(s)* for the particular disorder, containing essential and non-essential amino acids, carbohydrate, vitamins, minerals and trace elements. It is intended for the dietary management of an amino acid disorder from 6 months to 5 years of age.

gel is reconstituted with a small amount of water to form a low volume smooth semi-solid paste. Introducing **gel** from around 6 months of age begins the transition from the precursor free amino acid formula for infants to the concentrated spoonable protein substitute. It is designed to help meet the infant's nutritional and developmental feeding needs; ensuring the goals of the amino acid disorders dietary management are met, aiding growth and metabolic control.

| Product Name | Precursor amino acid free from | Premeasured sachet | Protein Equivalent (PE) | Flavour(s) |
|---|---------------------------------|--------------------|-------------------------|------------------------------------|
|  PKU gel | Phenylalanine | 24g | 10g | unflavoured, orange or raspberry** |
|  MSUD gel | Valine Isoleucine Leucine | 24g | 10g | unflavoured |
|  HCU gel | Methionine | 24g | 10g | unflavoured |
|  TYR gel | Tyrosine Phenylalanine | 24g | 10g | unflavoured |

** flavoured versions suitable from 1 year of age.

| Nutritional Information - unflavoured gel | | gel per 24g |
|--|------|-------------|
| Energy | kJ | 346 |
| | kcal | 81 |
| Fat | g | 0.02 |
| of which saturates | g | 0 |
| Carbohydrate | g | 10.3 |
| of which sugars | g | 6.5 |
| Protein equivalent | g | 10.0 |
| Free from the appropriate precursor amino acids* | g | - |

gel contains a wide range of micronutrients, for full nutritional information see www.vitafloweb.com

For flavoured **gel** nutritional information see www.vitafloweb.com







* No added condition specific precursor amino acid(s). They may be present in trace amounts from other ingredients (<10mg/100g powder, <4mg/10g PE sachet).

1.2 Nutritional features of gel

Protein Equivalent (PE)

Easy to calculate the PE, supporting a staged transition from the precursor free amino acid formula for infants* to the lower volume **gel**.

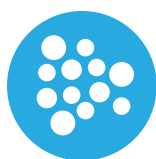


| Volume of precursor free amino acid formula for infants | Protein Equivalent | Volume of gel |
|---|--------------------|---------------|
|  100ml = 2g =  5g gel + 5ml cold water | | |
|  250ml = 5g =  ½ sachet gel (12g) + 12ml cold water | | |
|  500ml = 10g =  1 sachet gel (24g) + 24ml cold water | | |



Micronutrients

A protein-restricted diet increases the risk of micronutrient deficiencies⁽⁴⁻⁶⁾. The composition of **gel** contains a wide range of micronutrients including selenium, zinc, iron and calcium and a comprehensive range of vitamins including vitamin B12 and vitamin D important for nutrition and growth.



Long Chain Polyunsaturated Fatty Acids (LCPUFAs)

LCPUFA intake is reduced as the rich food sources are restricted in the diet for an amino acid disorder. Children receiving a protein-restricted diet have shown deficiency in docosahexaenoic acid (DHA)⁽⁷⁻⁸⁾.

keyOmega™ (VitaFlo) is a powder containing the omega 3 & 6 LCPs: DHA and AA. It is presented in convenient pre-weighed sachets and can be added in to the **gel**.

| | Sachet Size | Omega 3 LCP: DHA | Omega 6 LCP: AA |
|-----------------|-------------|------------------|-----------------|
| keyOmega | 4g | 100mg | 200mg |

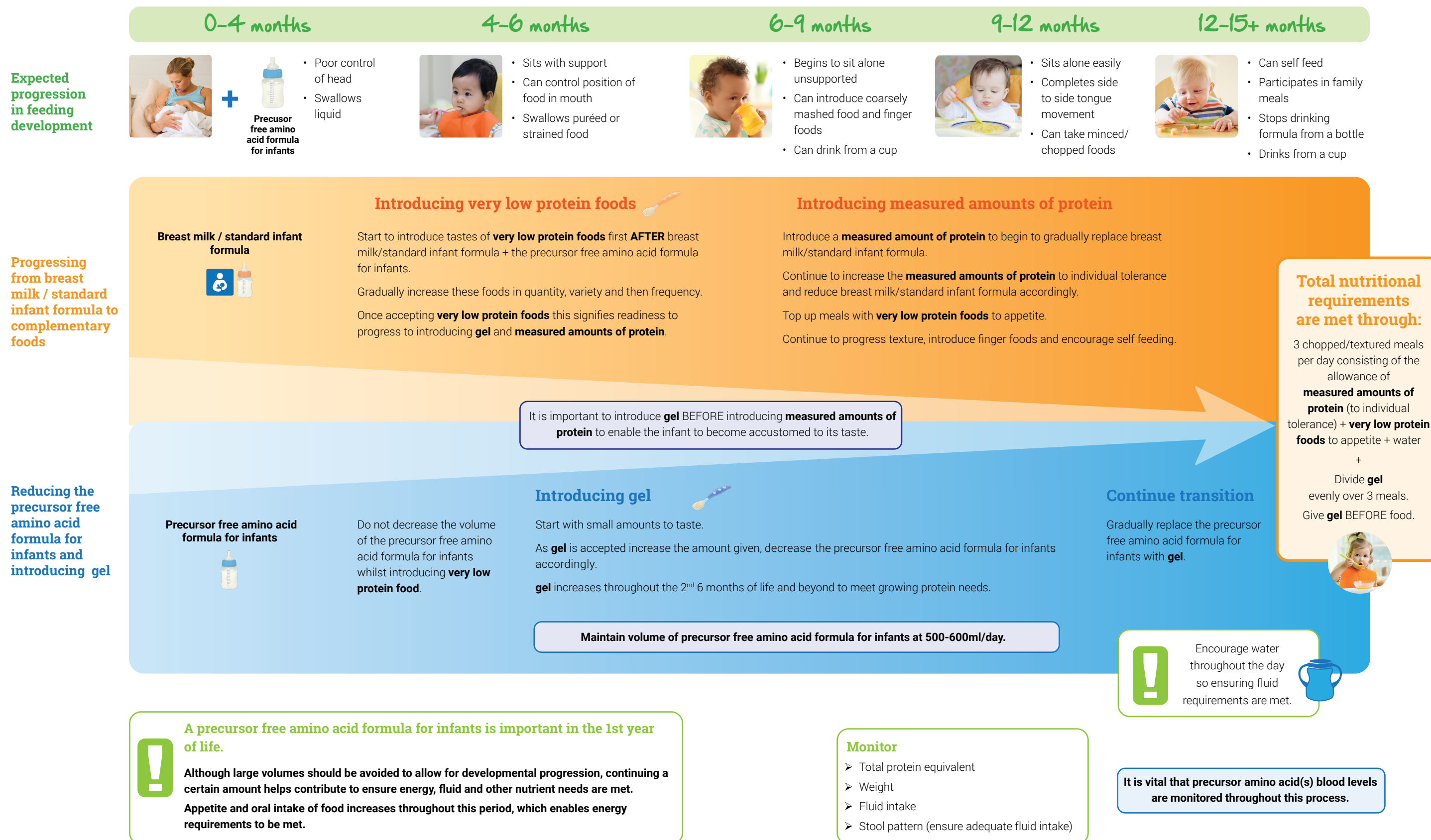
The recommended daily amount of keyOmega should be advised by a health care professional; however 1 sachet daily will meet the EFSA recommendations⁸ for DHA in children aged from 7 to 24 months.

* Based on a typical precursor free amino acid formula for infants.

2.1 Overview of the introduction to complementary feeding and gel

The following chart shows an overview of the stepwise practical introduction to complementary feeding, both **very low protein foods** and **measured amounts of protein** from food + **gel**. Details of each step follows in section 2.2.

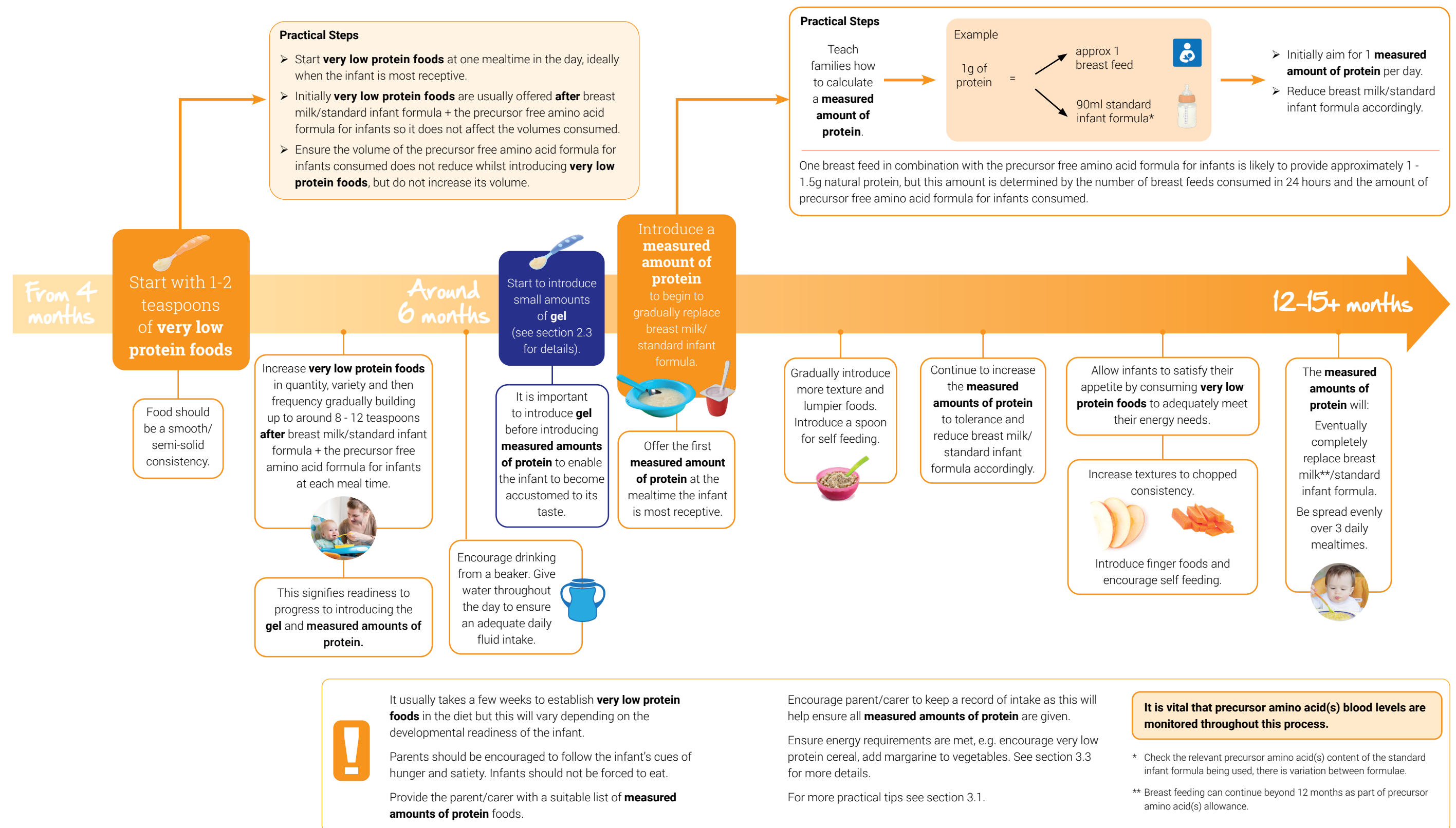
This is a guide only and every infant will progress at different rates.



2.2 Stepwise introduction to complementary feeding

The following chart shows the stepwise practical introduction to complementary feeding, both **very low protein food** and **measured amounts of protein**. In an amino acid disorder introducing complementary food from 17-26 weeks of age can be advantageous, as early exposure to food is likely to lead to acceptance of a wider range of **very low protein foods**, **measured amounts of protein** and **gel**, whilst maintaining blood levels of the precursor amino acid(s) within treatment recommendations⁽²⁾.

When introducing food, it is best to give **very low protein foods** first as they do not adversely affect blood levels. This allows the infant to experience different tastes without affecting metabolic control. This **promotes a positive feeding experience** as there is less pressure to consume a required amount of food. Once accepting **very low protein foods** this signifies readiness to progress to introducing **gel** and **measured amounts of protein** (exchanges).



2.3 Stepwise introduction to gel

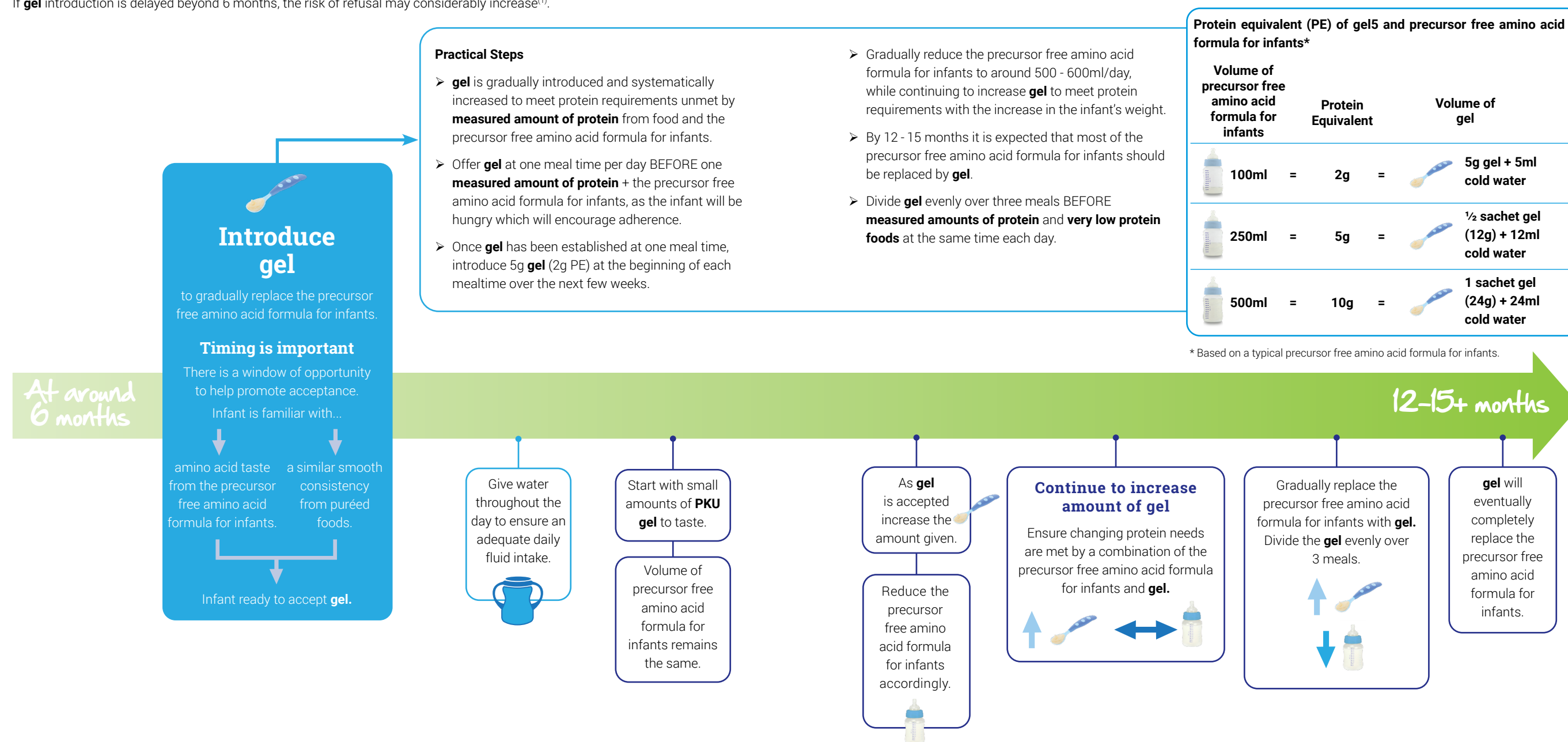
The following chart shows the stepwise introduction to gel.

Around 6 months - introduction to gel

It is important to introduce **gel** before introducing the **measured amounts of protein (exchanges)** to enable the infant to become accustomed to its taste.

If **gel** introduction is delayed beyond 6 months, the risk of refusal may considerably increase⁽¹⁾.

Throughout this process calculate total protein goals and consider natural protein from food. The remaining protein requirements are met by a combination of **gel** and the precursor free amino acid formula for infants, until fully established on **gel**.



A precursor free amino acid formula for infants is important in the 1st year of life.

Although large volumes should be avoided to allow for developmental progression, continuing a certain amount helps contribute to ensure energy, fluid and other nutrient needs are met.

Appetite and oral intake of food increases throughout this period, which enables energy requirements to be met.



Introduce **gel** at a convenient meal time when the parent/carer is not rushed and the infant is most receptive.

Every infant will progress at different rates, don't hesitate with steps to increase **gel**. Increase **gel** as quickly as the infant accepts and according to protein needs.





For more practical tips see section 3.1.

It is vital that precursor amino acid(s) blood levels are monitored throughout this process.

2.4

Guide for meeting the increasing protein needs of an infant with an amino acid disorder

The following table shows a worked example of how the infant's protein needs are met. It is **only a guide** and every infant will progress at different rates, therefore it may take longer to completely replace the precursor free amino acid formula for infants with **gel**.

| Weight | 6kg | 7kg | 8kg | 9kg | 10kg | 11kg | 12kg |
|---|---|--------------------------|--------------------------|---------------------------|----------------------------|----------------------------|----------------------------|
| Approximate age | Around 6 months | | | | | | 24 months |
| Precursor free amino acid formula for infants  | 700ml 14g PE | 600ml 12g PE | 600ml 12g PE | 600ml 12g PE | 400ml 8g PE | 200ml 4g PE | - |
| Gel  | Start with small amounts of gel to taste. | 4g gel x 3 = 5g PE | 7g gel x 3 = 8g PE | 9g gel x 3 = 11g PE | 15g gel x 3 = 18g PE | 21g gel x 3 = 25g PE | 27g gel x 3 = 32g PE |
| This infant tolerates 4g of natural protein*. Breast milk/ standard infant formula   → Measured amounts of protein | 4g protein | 4g protein | 4g protein | 4g protein | 4g protein | 4g protein | 4g protein |
| Very low protein food | Appetite and oral intake of food increases throughout this period, which enables energy requirements to be met. | | | | | | |
| Total protein intake/day | 18g | 21g | 24g | 27g | 30g | 33g | 36g |



The protein requirements in this example are calculated using the UK guideline of 3g total protein/kg/day^(10, 11), for infants and children 0 - 2 years old with PKU. In the absence of disorder specific research this guidance for protein intake is used for other amino acid disorders. Local recommendations and policies may differ on protein requirements for these infants and children and should be checked and adhered to where appropriate.

* Protein tolerance will vary between individuals and between the different conditions.













2.5 Sample meal plan - 10 month old boy with phenylketonuria

In the table below, a typical meal plan for meeting protein and energy requirements is shown for a child with phenylketonuria :

| | |
|---------------------------------------|---|
| Weight of infant | 10kg |
| Protein requirements | 30g/day (3g/kg/day^(10, 11)) |
| Natural protein tolerance | 4g |
| Amino acid formula for infants | 550ml/day |
| xgel | 3 x ½ sachets |



**Remember
to give a drink
of water at each
meal time.**

| | | | Protein/PE (g) | kcal |
|---------------------|---|---|--------------------------------|-------------------------------------|
| On waking | 200ml precursor free amino acid formula for infants  | | 4 | 134 |
| Breakfast | ½ sachet gel  | | 5 | 40.5 |
| | 1 slice of low protein toast, butter and apricot jam  | | | } 123 |
| | 20g yoghurt [†] + 6 raspberries  | 1 | | |
| | 150ml precursor free amino acid formula for infants  | 3 | | 102 |
| Midday | ½ sachet gel  | | 5 | 40.5 |
| | 60g sweet potatoes 40g baked beans 5g butter  | 2 | | } 169 |
| | ½ sachet low protein custard 40g chopped banana  | | | |
| | | | | |
| Evening meal | ½ sachet gel  | | 5 | 40.5 |
| | 20g low protein pasta 25g of spinach Tomatoes /onions  | 1 | | } 159 |
| | Chopped melon  | | | |
| | | | | |
| Bedtime | 200ml precursor free amino acid formula for infants  | | 4 | 122 |
| | Total | | 30g/day (3g/kg/day) | 934kcal* (93kcal/kg/day) |

* Calories estimated using manufacturer's labelling, food portion sizes⁽¹²⁾ and food composition tables⁽¹³⁾.

† Check manufacturer's label.



If infant is still breast feeding, count as part of protein allowance.

Protein tolerance will vary between individuals and between the different conditions.

3.1 Practical points for parents/caregivers for the successful introduction to gel and complementary foods

It is important to share practical advice with parents/caregivers to aid the successful introduction of **gel** and complementary foods. Setting the right foundations and promoting good habits with a consistent approach from the beginning will support success in the long term.

Introducing gel

Be organised - encourage parent/caregiver to be prepared, e.g. **gel** powder pre-measured in a small bowl, baby bib, feeding spoon and water. A small 5ml syringe would be useful for measuring small amounts of water.

Introducing gel - offer the infant **gel** at a time of day when most receptive/interested and a convenient meal time when the parent/carer is not rushed.

Routine - give **gel** at the same time and in the same way each day. Give at the start of each meal or half an hour before to ensure it is taken. Be consistent.

Ensure all of the gel is finished - emphasise the importance of taking the prescribed amount.

Same consistency - if more than one person administers **gel**, ensure that they make it to the same consistency each time. If this varies widely the infant may reject due to unfamiliarity.

Take it slowly and go at the infant's pace - give lots of praise - smiling and encouragement will help the infant accept **gel**. **Persevere** with consistent timings and encouragement.

Offer water - to ensure an adequate daily fluid intake.

Be patient - it is common for any infant to spit out or refuse foods at times. It is no different with **gel**, this is normal infant behaviour. Encourage parents to persevere and keep calm and continue with positive feeding techniques.

During teething - making **gel** up with ice cold water may help with sore gums.

Introducing complementary foods

Encourage the use of **homemade foods** & provide information on suitable commercially available **very low protein foods**.

Provide families with lots of **practical advice** regarding cooking skills and ideas.

Encourage batch cooking, this is especially useful in the early stages when portion sizes are small.

Repeat exposure - remember it can take ≥ 8 times of offering the same food for an infant to become accustomed to the taste and accept it - **continue to offer foods even if at first refused**^(14, 15).

Encourage **eating with the family** and share some low protein meals/foods.

Establish **regular meal times**, but **avoid prolonged feeding**, keep to 20 - 30 minutes per meal.

Encourage parent/carer to **keep a record of intake** as this will help ensure all **measured amounts of protein** food are given.

Provide information for other caregivers

Provide any new caregiver with an explanation of the particular amino acid disorder.

Provide the caregiver with clear written instructions of the feeding plan and the necessary feeding equipment.

Emphasise the importance of ensuring the child finishes all of their **gel** at each meal.

3.2 Additional information

Advice on illness

In PKU, HCU and TYR a formal Emergency Regimen (ER) is not normally necessary during illness as acute metabolic decompensation does not occur.



MSUD

It is essential to start an ER at the first sign of illness. This is **not covered** in this guide. It is vital to adhere to international/local country guidelines⁽¹⁶⁾.

For PKU, HCU and TYR

It is normal for an infant to refuse or have difficulties taking their usual amount of **gel** when ill or teething. This may be an anxious time for parents/caregivers and so it is helpful to stay in regular contact with the family to give advice and reassurance and to assess how the infant is progressing. Sometimes parents/caregivers feel that switching protein substitute may help if the child is refusing their protein substitute. This should be avoided and the underlying issues of the illness or teething addressed.

It is very important that an infant continues to take their **gel** during illness to help maintain metabolic control. However it may be necessary to recommend that the infant has the same daily dose, but administered in smaller more frequent amounts throughout the day, e.g. divide into five amounts per day rather than three. More time may be required to administer **gel**.

Continue to offer gel throughout the day even when an infant refuses or is unwell. Giving an infant a 'day off' from their gel will adversely affect their metabolic control and give the wrong message to the infant and family. Stopping gel, even for 24 hours, may create difficulties with its reintroduction.

Extra fluids in the day may be necessary, particularly if the infant has vomiting or diarrhoea.

Parents should be given written advice on how to manage illness and should seek medical advice early.

Feeding problems in toddlers

As with many toddlers, feeding problems are common in young children with amino acid disorders^(17, 18). Although children with an amino acid disorder may be fussy for the same reasons as other children, some issues may be unique to children with specific amino acid disorders, and place them at higher risk of developing feeding problems.

Strategies to minimise or improve feeding problems in an infant with an amino acid disorder

- Good and consistent feeding routines are required from the commencement of complementary feeding and the introduction of **gel**⁽¹⁾.
- Parents/caregivers/families should eat together with the infant to promote a positive environment and social interaction where possible.
- Offer a suitable, healthy and varied low protein diet, but avoid offering too many food choices at one meal.
- Allow infants to satisfy their appetite by consuming **very low protein foods** to adequately meet but not exceed their energy needs. As these foods are **very low protein foods**, they will not adversely affect metabolic control.
- Limit excessive snacking.
- Encourage positive mealtime behaviours e.g. eat at the table together, allow toddler to help set the table.
- The infant should be seated during meals and distractions minimised.
- Encourage self-feeding at the appropriate stage.
- Give **gel** in three equal doses before foods, at the same times each day.

3.3 Meeting energy requirements

When an infant transitions on to a solid diet, energy from breast milk or standard infant formula must be replaced by food. In a child with an amino acid disorder, the transition from an precursor free amino acid formula for infants to a second stage protein substitute such as **gel** creates an energy deficit to allow development of appetite for solid foods.

There has been some concern that the low energy density of a concentrated second stage protein substitute might not compensate for the energy content of the precursor free amino acid formula for infants and this might impact on growth and weight gain.

In an observational study⁽¹⁾ it was found that despite the lower energy content of the complementary feeding protein substitute compared to the precursor free amino acid formula for infants, no infant was reported to have growth issues. It was assumed that an increase in solid food intake compensated for the energy discrepancy. This demonstrated that this type of protein substitute can be introduced without adverse effects on appetite or growth.

A later study looking at the growth, protein and energy intake in children with PKU taking a low energy, concentrated weaning protein substitute in the first 2 years of life showed that using the low volume protein substitute met phe-free protein requirements, facilitated transition to solid foods and supported normal growth⁽¹⁹⁾.

It is important to make sure energy demands are met in this transitional period and beyond to facilitate growth and development.

General tips to increase energy intake if needed (section 3.4 for further details):

- Fortify lower energy foods, such as vegetables and fruit, with extra energy, e.g. adding fat
- Offer more energy dense foods rather than increasing portion sizes
- Avoid drinks 1 hour before meals and offer after a meal so the child does not fill themselves up with fluid
- Have mealtimes when the infant is not too tired to encourage cooperation and appetite



gel can be made up with protein free milk alternatives, such as ProZero, instead of water to add extra calories. Make up gel in the usual way but add the milk alternative instead of water. ProZero is a liquid blend of carbohydrate and fat designed as a protein free alternative to milk. It can be used in the dietary management of inborn errors of metabolism from 6 months of age.

3.4 Tips on how to increase energy in the diet

The following methods can be used to increase the energy content of a low protein diet:

Food sources

Low protein

Sweet potato
Butternut squash
Cassava
Low protein pasta/rice/flour

Protein free

Gelatine free jellies
Fruit purée
Honey (after 1 year of age)

TIPS

These foods are easily incorporated into puddings or savoury dishes:

- Cassava to make a low protein crumble
- Low protein flours can be used to make cakes/pancakes and dough for bread sticks
- Sweet potato and butternut squash in mild curry sauces

TIPS

Sweet and sugary foods are easily incorporated into puddings or given as snacks:

- Add to fruit
- Add to low protein pancakes

Remember to advise to eat sugary foods within a meal and ensure dental hygiene.

Low protein

Mayonnaise*
Avocado
Low protein cheese*

Protein free

Cooking oils
Margarine
Butter
Butter spreads
Protein free/low protein alternatives to milk*

TIPS

These foods are easily incorporated into puddings or savoury dishes:

- Mix cream* with low protein fruit to make fruit 'ice creams'
- Add suitable coconut milk* to low protein vegetable sauces
- Mix cream/mayonnaise* with avocado to make a spread for low protein toast

TIPS

Fats are easily incorporated into savoury meals:

- Lightly fry/roast low protein vegetables
- Mix into sweet potatoes/butternut squash
- Add into low protein tomato based sauces or mild curry sauces



Some of these foods may be counted as part of the protein allowance, depending on the amino acid disorder adhere to local country guidelines.

* Check manufacturer's label for suitability.

4.1 General references for the practical guide

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4.2 International management guideline references

General reference

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5.0 Appendices

5.1 Complementary feeding an infant without an amino acid disorder





5.2 References for complementary feeding an infant without an amino acid disorder

5.3 Organic acidaemias and international management guideline references

5.4 gel range

5.1 Complementary feeding an infant without an amino acid disorder

This summary represents a guide to infant development and common practices for the introduction of complementary foods. Individual infants develop and progress at different rates. Advice should be tailored to the individual infant and nutritional requirements assessed on an individual basis. Complementary feeding practice varies internationally and this should be considered.

| Developmental stage | | Recommendations and practice | |
|--|---|---|---|
| Stage 1 | | | |
| Start - Introduction of complementary foods begins not before 4 months but no later than 6 months ^(1 - 4) | | | |
|  | <p>Infants begin to sit with assistance and develop some head control.</p> <p>Infants begin to mouth, hold and gel objects and may show interest in food.</p> <p>Infants may open their mouth in anticipation of a spoon.</p> | <p>Homemade or commercial complementary food</p> <p>Purée texture foods are introduced once per day when the infant is most receptive and progresses to 2-3 times per day according to acceptance.</p> <p>Complementary foods are offered before a milk feed.</p> | <p>Breast feed/formula</p> <p>Milk feeds remain the same volume (around 120-150ml/kg/day) and complementary foods are experiential rather than nutritional at this stage.</p> <p>National policy should be consulted regarding recommendations for vitamin and mineral supplementation.</p> |
| Stage 2 | | | |
| Progress - Progression of texture and variety of complementary foods | | | |
|  | <p>The gag reflex to lumpy foods begins to decline.</p> <p>Infants develop skills to clear a spoon with their top lip.</p> <p>Mouth movements become more coordinated and chewing develops.</p> | <p>Homemade or commercial complementary food</p> <p>Frequency of complementary foods progresses to 3 small meals per day, offered before milk feeds.</p> <p>Food consistency can progress according to developmental stage from puréed to lumpy and mashed textures⁽¹⁾.</p> | <p>Breast feed/formula</p> <p>A cup or beaker can be introduced from 6 months old.</p> <p>As the quantity of complementary foods increases, the volume of milk feeds reduce according to appetite but should remain at least 500-600mls/day⁽⁷⁾.</p> <p>Water is offered at mealtimes.</p> |
| Stage 3 | | | |
| Establish - Further progression of texture and variety of complementary foods | | | |
|  | <p>The presence of teeth allows infants to bite harder foods.</p> <p>Motor control develops to enable self-feeding from a spoon.</p> | <p>Homemade or commercial complementary food</p> <p>Infants progress to chopped foods, harder finger foods and foods that require more biting and chewing.</p> <p>Snacks are introduced between meals⁽³⁾.</p> | <p>Breast feed/formula</p> <p>Milk feeds gradually reduce to 2-3 times per day.</p> <p>Water is offered at mealtimes.</p> |
| Stage 4 | | | |
| Beyond the first year - Joining in family meals and self feeding | | | |
|  | <p>Infants start to recognise foods.</p> <p>Chewing is not fully mature but infants can cope with most textures.</p> | <p>Family meals</p> <p>Meals based on foods prepared for the rest of the family/caregivers are offered and social engagement at mealtimes is always encouraged.</p> | <p>Breast feed/cow's milk/follow-on formula</p> <p>Continue breast feeds according to mother's/infant's wishes.</p> <p>If formula fed, pasteurized whole cow's milk can replace formula if the complementary feeding diet is adequate to meet nutritional requirements⁽³⁾ or follow-on formula can be used.</p> <p>Water is offered to meet fluid requirements.</p> |

Total nutritional requirements met by 3 chopped meals per day, snacks & drinks from a cup. Protein is achieved from a variety of food sources.

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5.3 Organic acidaemias

Several countries worldwide undertake expanded new-born screening for some of the organic acidaemias (OAA). Early detection by screening and pre-symptomatic initiation of treatment can greatly improve the outcome.

gel products are also available for the organic acidaemias (i.e.) Methylmalonic Acidaemia (MMA), Propionic Acidaemia (PA) and Glutaric Aciduria Type 1 (GA-I).

For those infants that are able to be introduced to complementary foods and a second stage protein substitute start at the usual time of around 6 months of age. Proceed in a similar method for the management of PKU, MSUD, HCU, TYR that are described in more detail in this guide. The basic principles in the guide to introduce complementary food and the appropriate **gel** can be used, making appropriate adjustments to any calculations or meal plan examples that are discussed.

Although there are similarities between the amino acid disorders and the OAA, there are also differences. The main difference, in relation to protein substitutes, is that, where used, the quantities prescribed tend to be smaller as there is more tolerance of natural protein in OAA.

Some of these patients however never progress onto solids or take very small amounts because of feeding difficulties and so will not be weaned in this way.

To avoid potential confusion, this guide has not focused on the OAA.

For further info on the OAA see references below.

General references for organic acidaemias

Dixon M, MacDonald A, White F, Stafford J. 17. Disorders of Amino Acid Metabolism, Organic Acidaemias and Urea Cycle Disorders. In: Shaw V (ed.). Clinical Paediatric Dietetics. 4th edition. Oxford, UK: Wiley Blackwell; 2015. p.381-525.

MMA/PA

Baumgartner MR, Hörster F, Dionisi-Vici C, Haliloglu G, Karall D, Chapman KA, *et al.* Proposed guidelines for the diagnosis and management of methylmalonic and propionic acidemia. Orphanet Journal of Rare Diseases. 2014;9:130-66.

IVA













Dionisi-Vici C, Deodato F, Röschinger W, Rhead W, Wilcken B. 'Classical' organic acidurias, propionic aciduria, methylmalonic aciduria and isovaleric aciduria: Long-term outcome and effects of expanded newborn screening using tandem mass spectrometry. Journal of Inherited Metabolic Disease. 2006;29(2-3):383-9.

GA Type 1

Boy N, Mühlhausen C, Maier EM, Heringer J, Assmann B, Burgard P, *et al.* Proposed recommendations for diagnosing and managing individuals with glutaric aciduria type I: second revision. Journal of Inherited Metabolic Disease. 2017;40(1):75-101.t

5.4 gel Range

gel is available for the following conditions, suitable from 6 months:

| | Product / Condition | Precursor amino acid(s) free from | Flavour | Sachet Size | Protein Equivalent (PE) |
|---|---|---|--|-------------|-------------------------|
|  |  PKU gel Phenylketonuria (PKU) | Phenylalanine | unflavoured orange or raspberry* | 24g | 10g |
|  |  MSUD gel Maple Syrup Urine Disease (MSUD) | Valine Isoleucine Leucine | unflavoured | 24g | 10g |
|  |  HCU gel Homocystinuria (HCU) | Methionine | unflavoured | 24g | 10g |
|  |  TYR gel Tyrosinaemia (TYR) | Tyrosine Phenylalanine | unflavoured | 24g | 10g |
|  |  MMA/PA gel Methylmalonic Acidaemia/Propionic Acidaemia (MMA/PA) | Methionine Threonine Valine Low Isoleucine | unflavoured | 24g | 10g |
|  |  GA gel Glutaric Aciduria Type 1 (GA-I) | Lysine Low Tryptophan | unflavoured | 24g | 10g |

It is essential that the correct **gel** product is prescribed for the particular condition. To make this easier the **gel** range has been given specific colours according to condition.

* Suitable from 1 year of age.



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