



expressTM
plus⁺

A practical guide to the use of express plus+ in older children,
teenagers and adults



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With You

Supporting education in the
dietary management of rare diseases

1.0 Disclaimer and important information

Purpose

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

Intended users

This practical guide is:

- for use by healthcare professionals working with older children, teenagers and adults diagnosed with disorders of protein metabolism
- **not** for use by parents/caregivers of older children, teenagers and 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 older children, teenagers and adults with diagnosed/proven disorders of protein metabolism.

Product information

Express plus+ 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. Please refer to these sources for information regarding allergens.

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

Important notice for express plus+

- 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 healthcare 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 for the dietary management of disorders of protein metabolism. Information contained within the guide is based on the most recent scientific evidence available on the management of disorders of protein metabolism as of August 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.

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Collaborators

This practical guide was written by Vitaflo dietitians in collaboration with Louise Robertson, Specialist Dietitian in Inherited Metabolic Disorders, University Hospitals Birmingham NHS Foundation Trust.

2.0 Abbreviations

DHA

docosahexaenoic acid

GA1

glutaric aciduria type 1

HCU

homocystinuria

MMA

methylmalonic acidaemia

MSUD

maple syrup urine disease

PE

protein equivalent

PKU

phenylketonuria

PA

propionic acidaemia

3.0 Introduction

Management of disorders of protein metabolism has progressed over recent years to give individuals a better quality of life and improve long-term health outcomes. However, the restrictions that management entails continue to pose a significant burden on the lives of individuals living with metabolic disorders.^{1,2} Medical management has developed over recent years, for example the use of nitisinone (NTBC) in hereditary tyrosinaemia and betaine in homocystinuria (HCU). However, dietary management remains the cornerstone and protein substitutes are a major component of care plans.³⁻⁶ Adherence to these dietary recommendations is a constant challenge for individuals with IEM and their health care professionals, with consequential decreased metabolic control linked to poorer clinical and social outcomes.⁷⁻¹¹

The express plus+ range is designed to support individuals in the management of their condition, aiding adherence to dietary recommendations. Express plus+ is available for phenylketonuria (PKU), maple syrup urine disease (MSUD), tyrosinaemia, HCU, glutaric aciduria type 1 (GA1), methylmalonic acidaemia (MMA) and Propionic Acidaemia (PA). This practical guide provides information to support its use in these conditions and help achieve the most out of the express plus+ range.

4.0 What is express plus?

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

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

Main features of express plus+

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

Available for PKU, MSUD, HCU, TYR, GA and MMA/PA

Contains DHA (200mg/30g PE)









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**

* Unflavoured varieties are compatible with flavour pacs available in blackcurrant, tropical, orange and raspberry

** express plus+ made to a volume preferred by your patient

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






| PKU express plus* | | Nutritional Information | | | | | |
|--------------------|-------|-------------------------|-----------|-------------|-----------|-------------|-----------|
| Nutrient | Units | Per 100g (60g PE) | | Per 15g PE | | Per 20g PE | |
| | | unflavoured | flavoured | unflavoured | flavoured | unflavoured | flavoured |
| Energy | kJ | 1339 | 1297 | 335 | 325 | 455 | 434 |
| | kcal | 316 | 306 | 79 | 77 | 107 | 102 |
| Protein | g | 60 | 60 | 15 | 15 | 20 | 20 |
| Fat | g | 2.2 | 2.2 | 0.55 | 0.55 | 0.75 | 0.75 |
| of which saturates | g | 0.7 | 0.7 | 0.18 | 0.18 | 0.24 | 0.24 |
| DHA | mg | 400 | 400 | 100 | 100 | 136 | 136 |
| Carbohydrate | g | 14 | 11.5 | 3.5 | 2.9 | 4.8 | 3.9 |
| of which sugars | g | 1.2 | 2.9 | 0.30 | 0.73 | 0.41 | 0.99 |
| Fibre | g | 0 | 0 | 0 | 0 | 0 | 0 |

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

5.0 Achieving adherence with express plus+

Management guidelines across most disorders of protein metabolism recommend a diet for life incorporating the use of a protein substitute.³⁻⁶ In practice, maintaining dietary restriction and adherence to protein substitutes becomes more difficult beyond the childhood and adolescent years¹²⁻¹⁶ with significant nutritional implications.⁷

Express plus+ is designed to address some of the barriers that individuals with disorders of protein metabolism face, which can make adherence challenging.

| Barrier to adherence | | |
|---|---|--|
| Large volumes to drink |  | Express plus+ can be prepared as a spoonable semi-solid, mini drink or flexi drink in order to provide a volume to suit the individual. Some individuals prefer a small, quick-to-drink volume whereas others may prefer a longer drink. |
| Medicalisation of the diet |  | The non-medical name and packaging of express plus+ can help normalise the diet and aid adherence. This may benefit individuals who may be embarrassed or self-conscious about their diet as the packaging is discreet. |
| Protein substitute adds energy to the diet |  | <p>Individuals with disorders of protein metabolism are subject to the same risks of overweight and obesity as the rest of the population.^{17,18}</p> <p>Express plus+ is lower in energy, compared to many other protein substitutes, to allow energy to come from food rather than the protein substitute and help prevent excessive energy intake.</p> <p>Express plus+ 15 = 70-74kcal/sachet Express plus+ 20 = 95-101kcal/sachet</p> |
| Flavour fatigue |  | <p>A variety of flavour choices can help prevent flavour fatigue and can be mixed and matched according to individual preference or used in recipes.</p> <p>An unflavoured option can be used with flavour pacs or own preferred (permitted) flavours.</p> |
| Dietary burden |  | <p>The format of express plus+ is reported to be convenient and easy to prepare¹⁹ reducing the burden and promoting adherence.</p> <ul style="list-style-type: none"> • Pre-measured sachet removes the need to weigh out powder, is discreet and easy to store and transport • Two pack sizes (15g PE and 20g PE) accommodate individual protein requirements and can reduce the number of sachets required. |

6.0 Using express plus+

The flexibility of express plus+ makes it a versatile protein substitute that supports many lifestyles and circumstances. The information below gives some guidance on how you can achieve the most from express plus+



The convenience and flexibility of express plus+ makes it a good choice for individuals with a disorder of protein metabolism returning to diet.

For individuals introducing a protein substitute for the first time, or following a break from management, express plus+ can be prepared according to tolerance.

Preparing to a larger volume as a mini or flexi drink initially, may help acceptability and tolerance by diluting both taste and osmolality.

The volume can then be gradually decreased, if the individual prefers a smaller drink or semi-solid.

Trial different flavours and recipes to establish preference. Sample packs are available from your Vitaflo representative.



Individuals who prefer a ready-to-drink protein substitute as their usual choice may find them bulky to travel with. Similarly, more traditional style protein substitutes in tins can be inconvenient to transport.

On days out or when traveling, patients may find express plus+ easier to pack away and transport. Having both options available allows flexibility and may help adherence. Express plus+ is interchangeable with the ready-to-drink ranges cooler and air and so can be combined to suit lifestyle.



A supply of express plus+ can be useful to keep at key places such as a college locker, work drawer or partner's / family member's home. In the event of an individual forgetting to take their protein substitute with them, there is an easily stored supply in case of emergency. Even if the individual usually has a ready-to-drink protein substitute, express plus+ can be easily and discreetly stored for these times.



Express plus+ can be used for individuals who prefer their protein substitute to remain discreet. Small sachets providing an easily consumed low volume may help individuals feel less self-conscious when taking their protein-substitute.



For individuals who prefer to take their protein substitute incorporated into drink recipes, express plus+ is ideal. For further details, please refer to your local Vitaflo representative.



Pregnancy presents additional challenges to taking a protein substitute such as nausea and vomiting.²⁰ The flexibility of express plus+ can help individuals who may be struggling with the effects of pregnancy.

Volume: some women may find smaller volumes help with nausea, whereas others may find a more dilute preparation helpful. Express plus+ can be prepared to suit the needs of the individual which may differ at different times of the day and different stages of pregnancy.

Mouthfeel: express plus+ is designed to have a smooth mouthfeel which can be helpful for individuals suffering from nausea.

Unflavoured option: a neutral flavour may benefit women suffering from nausea and can also be flavoured with tolerated (permitted) flavours.

Temperature: making express plus+ with ice cold water may help manage nausea.

Express plus+ is interchangeable with the cooler and PKU air range for micronutrients per gram of PE, so can be swapped completely or partially very easily. To aid tolerance, swapping in one express plus+ in at a time can help.

Note that there are differences in energy between products so the diet may need to be adjusted and metabolic control monitored. For up-to-date nutritional information for all products visit www.nestlehealthscience.co.uk/vitaflo



| | |
|--|---------------------------|
| Condition: | Maple Syrup Urine Disease |
| Age: | 42 years |
| Total protein | 67g |
| Natural protein | 7 x 50mg exchanges |
| Protein equivalent requirement: | 60g |

| | Intake | Energy (kcal) | PE (g) | Exchanges |
|----------------------------|---|---------------|--------|-----------|
| Breakfast | 20g wheat biscuit with protein-free milk alternative (2 exchanges) Bowl of exchange-free fruit salad Cup of tea with protein-free milk alternative | 300 | | 2 |
| | 1 sachet of MSUD express plus+ 20 made into a flexi drink with orange flavour pac | 107 | 20 | |
| Mid-morning | 4 low protein bread sticks Homemade tomato salsa dip Glass of permitted flavoured water | 150 | | |
| Lunch | 1 low protein bread roll with butter 30g baked beans (2 exchanges) 2 slices of protein-free cheese Glass of water | 335 | | 2 |
| | 1 sachet of MSUD express plus+ 20 made into a spoonable semi-solid | 107 | 20 | |
| Mid-afternoon snack | Small packet of olives Cup of coffee with protein-free milk alternative | 80 | | |
| Evening meal | Sweet potato wedges with olive oil and pepper 15g peas, 40g French beans (1 exchange) 3 low protein sausages 35g dairy-free rice pudding with honey and prunes (2 exchanges) | 630 | | 3 |
| | 1 sachet of MSUD express plus+ 20 unflavoured made into a flexi drink with coffee syrup | 107 | 20 | |
| Supper | Small tin of apricots in syrup with protein-free custard Cup of herbal tea | 230 | | |
| | | 2046 | | 67 |

For further details, please refer to your local Vitaflo representative.



Data cards



Nutrition comparison table



Case studies



Recipe booklet



Preparation videos



HCP information booklets



Patient information leaflets

Notes

Notes

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Vitaflo International Ltd,
Suite 1.11, South Harrington Building,
182 Sefton Street, Brunswick Business Park,
Liverpool L3 4BQ, UK

+44 (0)151 709 9020

www.vitafloweb.com

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