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

Vitaflo in Association 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™ 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 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 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 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 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.

Contents

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

2.0 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 range is designed to support individuals in the management of their condition, aiding adherence to dietary recommendations. Express 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 range.

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)

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

Available for PKU, MSUD, HCU and TYR*

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 made to a volume preferred by your patient

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	✔			

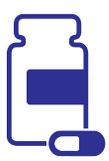
PKU express		Nutritional Information					
Nutrient	**Units**	**Per 100g (60g PE)**		**Per 15g PE**		**Per 20g PE**	
		unflavoured	**flavoured**	**unflavoured**	**flavoured**	**unflavoured**	**flavoured**
Energy	kJ	1292	1241	324	310	434	422
	kcal	304	292	76	73	102	99
Protein	g	60	60	15	15	20	20
Fat	g	0	0	0	0	0	0
of which saturates	g	0	0	0	0	0	0
Carbohydrate	g	16	13	4	3.3	5.4	4.4
of which sugars	g	0.5	1.7	0.13	0.43	0.17	0.58
Fibre	g	0	0	0	0	0	0

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

5.0 Achieving adherence with express

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 years [12-16] with significant nutritional implications.[7]

Express 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 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 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		Individuals with disorders of protein metabolism are subject to the same risks of overweight and obesity as the rest of the population.[17, 18] Express 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. Express 15 = 70-74kcal/sachet Express 20 = 95-101kcal/sachet
Flavour fatigue		A variety of flavour choices can help prevent flavour fatigue and can be mixed and matched according to individual preference or used in recipes. An unflavoured option can be used with flavour pacs or own preferred (permitted) flavours.
Dietary burden		The format of express is reported to be convenient and easy to prepare[19] reducing the burden and promoting adherence. • 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

The flexibility of express 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.

The convenience and flexibility of express 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 easier to pack away and transport. Having both options available allows flexibility and may help adherence.
Express is interchangeable with the ready-to-drink ranges cooler and air and so can be combined to suit lifestyle.

A supply of express 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 can be easily and discreetly stored for these times.

Express 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 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 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 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 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 with ice cold water may help manage nausea.

Express 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 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**

7.0 Example meal plan

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 15 made into a flexi drink with orange flavour pac	76	15	
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 15 made into a spoonable semi-solid	76	15	
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 15 unflavoured made into a flexi drink with coffee syrup	76	20	
Supper	Small tin of apricots in syrup with protein-free custard Cup of herbal tea	300		
	1 sachet of MSUD express 15 made into a spoonable semi-solid		15	
		2023	**67**	

8.0 Additional resources for express

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

9.0 References

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