



**MSUD**

# **MSUD is a complex condition**

Vitaflo aims to simplify matters



Innovation in Nutrition

The Comprehensive MSUD Range

# At Vitaflo we appreciate the complexity of MSUD dietary management.

Vitaflo offers a comprehensive range of products for the patient with MSUD - Choosing this range helps to address the key issues in managing the dietary aspects of this condition.

## **Metabolic control - compliance**

Products which help to maintain plasma branched chain amino acid (BCAA) levels within appropriate ranges.<sup>1</sup>

## **Growth and development**

Products providing BCAA free protein (protein equivalent (PE)) which supports normal growth and development.

## **Nutritional adequacy**

A product range which is nutritionally up to date with vitamin, mineral and trace element quantities in line with Australian Nutrient Reference Values (NRVs).<sup>2</sup>

## **Illness Management**

A range of products which can be flexibly incorporated into the patient's dietary regimen to provide additional calories, BCAA free protein and single dose amino acids.

## **Convenience and Support**







Products that are simple to prepare and so time-saving **and** products which are ready to feed, so can be taken immediately.

They are also suitable for transporting outside of the home environment; making the MSUD range extremely convenient.





Vitaflo's free home delivery service in conjunction with excellent customer service and support means that MSUD product range items will be with your patient in a timely manner.



# BCAA free protein substitutes

	10g PE choices	15g PE choices	20g PE choices
Powder	<p><b>MSUD Gel</b> Suitable from 6 months to 10 years of age</p>  <p>1 sachet = 10g PE unflavoured</p>	<p><b>MSUD express 15</b> Suitable from 3 years onwards</p>  <p>1 sachet = 15g PE unflavoured</p>	<p><b>MSUD express 20</b> Suitable from 3 years onwards</p>  <p>1 sachet = 20g PE unflavoured</p>
Liquid	<p><b>MSUD cooler 10</b> Suitable from 3 years onwards</p>  <p>1 cooler = 10g PE <b>Red</b></p>	<p><b>MSUD cooler 15</b> Suitable from 3 years onwards</p>  <p>1 cooler = 15g PE <b>Red and Orange</b></p>	<p><b>MSUD cooler 20</b> Suitable from 3 years onwards</p>  <p>1 cooler = 20g PE <b>Red</b></p>

# How to use the MSUD gel and MSUD express system




Patient Age Range	Intake from products	Quantity of gel and express	%AI/RDI <sup>2</sup> of Key Nutrients
7-12 months 1-3 years	20g day <b>162kcal</b>	2 x gel 	Calcium 104-193 Phosphorus 86-144 Vitamin A 67-96 Vitamin D 140 Folic Acid 67-125 Iron 62-76 Zinc 173 Magnesium 100-106
4-8 years	30g day <b>243kcal (gel)</b> or <b>148kcal (express)</b>	3 x gel or 2 x express 15s 	Calcium 85 Phosphorus 107 Vitamin A 104 Vitamin D 132 Folic Acid 100 Iron 108 Zinc 270 Magnesium 145
9-13 years	45g day <b>222kcal</b>	3 x express 15s 	Calcium 69-90 Phosphorus 64 Vitamin A 104 Vitamin D 198 Folic Acid 100 Iron 203 Zinc 270 Magnesium 118
14-18 years 19+ years	60g day <b>296kcal (express 15)</b> <b>303kcal (express 20)</b>	3 x express 20s or 4 x express 15s 	Calcium 94-122 Phosphorus 87-109 Vitamin A 94-121 Vitamin D 90-270 Folic Acid 102 Iron 122-274 Zinc 156-313 Magnesium 91-124

**MSUD gel** combined with 30ml of water produces a smooth, semi-solid food which is suitable for spoon feeding; ideal for the infant transitioning from liquid to solid food.

**MSUD express** combined with 20ml water makes a paste which can be taken from a spoon; ideal for the older child, adolescent or adult wanting a very low volume formula.

Both **MSUD gel** and **MSUD express** can be made with 80ml water to make low volume drinks or formulae suitable for feeding enterally via a tube.

# How to use the MSUD cooler system

Patient Age Range	Intake from products	Quantity of cooler	%AI/RDI <sup>2</sup> of Key Nutrients																
3 years 4-8 years	30g day <b>195kcal</b>	3 x cooler 10s 	<table border="0"> <tr><td>Calcium</td><td>86-120</td></tr> <tr><td>Phosphorus</td><td>107-116</td></tr> <tr><td>Vitamin A</td><td>98-131</td></tr> <tr><td>Vitamin D</td><td>300</td></tr> <tr><td>Folic Acid</td><td>75-100</td></tr> <tr><td>Iron</td><td>111-123</td></tr> <tr><td>Zinc</td><td>210-280</td></tr> <tr><td>Magnesium</td><td>127-206</td></tr> </table>	Calcium	86-120	Phosphorus	107-116	Vitamin A	98-131	Vitamin D	300	Folic Acid	75-100	Iron	111-123	Zinc	210-280	Magnesium	127-206
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9-13 years	45g day <b>291kcal</b>	3 x cooler 15s 	<table border="0"> <tr><td>Calcium</td><td>69-90</td></tr> <tr><td>Phosphorus</td><td>64</td></tr> <tr><td>Vitamin A</td><td>97</td></tr> <tr><td>Vitamin D</td><td>450</td></tr> <tr><td>Folic Acid</td><td>75</td></tr> <tr><td>Iron</td><td>206</td></tr> <tr><td>Zinc</td><td>210</td></tr> <tr><td>Magnesium</td><td>102</td></tr> </table>	Calcium	69-90	Phosphorus	64	Vitamin A	97	Vitamin D	450	Folic Acid	75	Iron	206	Zinc	210	Magnesium	102
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**Allergen Declaration:** Contains fish (tuna oil), soya (soya lecithin).

**All 3 pack sizes have the same nutritional content per 100ml.** The combination which best meets protein requirements and maintains optimal MSUD control can be chosen.

**Protein intake can be easily modified.** As the patient's protein requirement increases they may change to a larger pack size or take extra of the same pack size. Stress that

often occurs when a patient must change to a protein substitute with an unfamiliar taste, may lessen.

**Micronutrient and protein content in cooler are closely linked.** Meeting an individual's protein requirement<sup>1</sup> solely from cooler ensures key micronutrient needs are greatly satisfied.

# Benefits of the MSUD range

## Nutritionally up-to-date

- Provides > 64% RDI<sup>2</sup> for calcium and phosphorus (see tables on previous<sup>2</sup> pages).
- Content of Vitamin A & D, Folic Acid and Iron are in line with current Australian NRVs.<sup>2</sup>

## MSUD cooler system and DHA

- MSUD cooler contains long-chain polyunsaturated fatty acids such as preformed DHA.
- Patients with MSUD have been found to have significantly lower plasma concentrations of DHA compared to controls.<sup>1</sup>
- Supplementation with DHA results in higher DHA status without adverse reactions in patients with an inborn error of metabolism (IEM).<sup>4</sup>
- Patients need not be concerned about taking additional DHA supplementation or about an excessive Vitamin A intake.
- The DHA in cooler assists in meeting suggested daily targets (SDTs) for reducing chronic disease risk in the general population.<sup>3</sup>

## Accuracy and Convenience

- **MSUD gel** and **MSUD express** are available as premeasured sachets and so exact amounts are ensured every time.
- **MSUD cooler** is ready to feed and so is easy to transport outside of the home and no preparation is required.
- **MSUD gel** and **MSUD express** can deliver concentrated amounts of BCAA free protein in a very low volume. Consuming lower total volumes of formula may increase compliance for patients with an IEM.<sup>5,6</sup>

## Flexibility with Vitaflo's Modular Options

- **Prozero** can be combined with **MSUD gel** and **MSUD express** to provide additional calories in a palatable form.
- **MSUD amino5** is ideal for delivering additional amounts of BCAA free protein.
- **Basecal** can be incorporated into the MSUD dietary regimen to deliver protein free calories while also providing vitamins, minerals, trace elements and essential fatty acids.
- Single dose amino acids allow for immediate changes to MSUD dietary regimens.



# BCAA free protein supplementation

**MSUD amino5** is a powdered amino acid supplement containing a complete range of amino acids without the BCAAs.

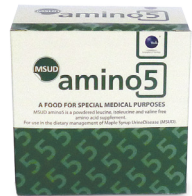
It contains no carbohydrate, fat or vitamins and minerals and is available in pre-measured sachets. It is ideal for use in sick day regimens to deliver additional BCAA free protein without significantly increasing the caloric intake.

## Nutritional information (per sachet)

Energy	kcal	20	Leucine	g	0
Protein equivalent	g	5.0	Lysine	g	0.49
Alanine	g	0.68	Methionine	g	0.14
Arginine	g	0.65	Phenylalanine	g	0.36
Aspartic Acid	g	0.82	Proline	g	0.57
Cystine	g	0.19	Serine	g	0.39
Glutamine	g	0	Threonine	g	0.35
Glycine	g	0.54	Tryptophan	g	0.19
Histidine	g	0.23	Tyrosine	g	0.39
Isoleucine	g	0	Valine	g	0

## MSUD amino5 as a premeasured sachet -

- Pre-measured sachets deliver exact amounts every time
- Can be concentrated and so ideal for sick day regimens
- Reduced wastage and hygienic as only opened as required.
- Suitable for use from 3 years of age.



## Single dose amino acids

Supplementation with isoleucine and valine is crucial for optimal dietary management of MSUD<sup>7</sup> as indicated by individual plasma amino acid levels.

Vitaflor provides each of these as single amino acids in 2 amounts:

**Isoleucine50** 50mg of L-Isoleucine in a 4g sachet.

**Isoleucine1000** 1000mg of L-Isoleucine in a 4g sachet.

**Valine50** 50mg of L-Valine in a 4g sachet.

**Valine1000** 1000mg of L-Valine in a 4g sachet.

**MSUD amino5** and each of the **single dose amino acids** can be mixed in with usual protein substitute, added to modular feeds, incorporated into other permitted food/drink or taken via enteral feeding tubes.

# Increasing Calories



or



+



MSUD gel or MSUD express with Prozero provides:

- **Palatable, protein substitutes** - ideal for patients with MSUD - more calories, for minimal volume, can be delivered than when prepared with water alone.
- **Adaptable, protein substitutes** - the calorie content and taste can be tailored to suit the needs of the patient - simply adjust the amount of Prozero added.

The amount of Prozero mixed with either MSUD gel or MSUD express can be tailored to the individual's calorie requirements.





## Younger Children

Minimum amounts of Prozero we recommend adding to **MSUD gel** are presented in the table below:

		MSUD gel as a High Calorie gel	MSUD gel as a High Calorie Drink
MSUD gel		1 sachet	1 sachet
Prozero	ml	30	125
Approx. Final Volume	ml	50	150
Energy	kcal/kJ	<b>101/429</b>	<b>165/694</b>
PE	g	10	10

1 sachet MSUD gel = 24g = **81kcal/346kJ**

**Prozero Allergen Declaration:** Contains milk (milk fat and sugar)

## Older Children and Adults

Minimum amounts of Prozero we recommend adding to **MSUD express** are presented in the table below:

		MSUD express15 as a High Calorie Paste	MSUD express15 as a High Calorie Drink	MSUD express20 as a High Calorie Paste	MSUD express20 as a High Calorie Drink
MSUD express15		1 sachet	1 sachet	nil	nil
MSUD express20		nil	nil	1 sachet	1 sachet
Prozero	ml	20	125	20	125
Approx. Final Volume	ml	45	150	55	150
Energy	kcal/kJ	<b>87/371</b>	<b>158/663</b>	<b>114/485</b>	<b>185/777</b>
PE	g	15	15	20	20

1 sachet MSUD express15 = 25g = **74kcal/315kJ**

1 sachet MSUD express20 = 34g = **101kcal/429kJ**





**Prozero Allergen Declaration:** Contains milk (milk fat and sugar)

# Illness Management

Vitaflo understands that for patients with MSUD and their families, times of inter-current infection, surgery and injury can be very stressful.

Intake has usually decreased and an “illness” regimen is immediately required.

Several convenient, simple to prepare options for delivery of additional calories have been provided:

Younger Children	
<p><b>MSUD gel + Prozero</b></p> <p>1 ×  + 1 × </p> <p>1 sachet + 1 x 250ml carton</p> <p><b>10gPE and 248kcal</b></p>	<p><b>MSUD gel + Prozero</b></p> <p>4 ×  + 1 × </p> <p>4 sachets MSUD gel + 1 x 1 Litre carton</p> <p><b>40gPE and 990kcal</b></p>

**Prozero Allergen Declaration:** Contains milk (milk fat and sugar)

Vitaflo also recognises that during times of illness, adjustments to recommended amounts of isoleucine and valine are often necessary and may occur frequently.

**Isoleucine50, Isoleucine1000, Valine50** and **Valine1000** provide flexibility to illness regimens:

## Convenient

- Can be directly taken from sachet and mixed with the permitted feed of choice
- Isoleucine and valine are both available in sachets containing 50mg or 1000mg so ideal for matching to patient requirements

## Accuracy

- Pre-measured sachets deliver exact amounts every time.
- Single dose amino acids are soluble in water and so make ready solutions; precise amounts of amino acid can be titrated and incorporated into the dietary regimen as prescribed.

## Older Children and Adults

<p><b>MSUD express20 + Prozero</b></p> <p>1 ×  + 2 × </p> <p>1 sachet + 2 x 250ml cartons</p>	<p><b>MSUD express20 + Prozero</b></p> <p>4 ×  + 2 × </p> <p>4 x sachets + 2 x 1 Litre cartons</p>
<p><b>20g PE and 435kcal</b></p>	<p><b>80g PE and 1736kcal</b></p>
<p><b>MSUD amino5 + basecal200*</b></p> <p>2 ×  + 1 × </p> <p>2 sachets + 1 sachet</p>	<p><b>MSUD amino5 + basecal200</b></p> <p>16 ×  + 8 × </p> <p>16 sachets + 8 sachets</p>
<p><b>10gPE and 240kcal</b></p>	<p><b>80g PE and 1920kcal</b></p>

### Allergen Declaration.

**Basecal200-** Contains fish (tuna oil), milk (milk protein), soya (soya lecithin).

**Prozero-** Contains milk (milk fat and sugar).

\*At standard dilution basecal200 provides 1kcal/ml.

basecal200 can be made more concentrated if required.

For basecal200 product literature and samples, contact VitaFlo to arrange.

All of VitaFlo's increased calorie options are suitable to take orally or via an enteral feeding tube.

Flavourpac sachets are available in 5 different flavour options - **blackcurrant**, **orange**, **raspberry** and **tropical**. They mix well with MSUD gel, MSUD express and Prozero and are suitable to use from 3 years onwards.



## References

1. Frazier D, Allgeier C, Homer C et al. Nutrition management guideline for maple syrup urine disease: An evidence- and consensus-based approach. Mol Genet Metab. 2014; 112(3): 210-217.
2. NHMRC. Nutrient Reference Values for Australia and New Zealand. Commonwealth of Australia, 2006
3. Chronic Disease/Macronutrient Balance/n-3 and n-6 fatty acids in 3 above
4. Fekete K and Desci T. Long-Chain Polyunsaturated Fatty Acids in Inborn Errors of Metabolism. Nutrients. 2010; 2(9): 965-974.
5. MacDonald A, Lilburn M, Cochrane B, et al. A new, low-volume, protein substitute for teenagers and adults with phenylketonuria. J Inheret Metab Dis. 2004; 27: 127-135.
6. Gokmen-Ozel H, Macdonald A, Daly A, Hall K, Chakrapani A. Long term efficiency of "Ready to Drink" protein substitute in PKU. Mol Genet Metab. 2009; 98: Abstract 170.

# Prescription Information

## MSUD gel, MSUD express, MSUD cooler and MSUD amino5

### PBS Listed

Restricted benefit for patients with proven maple syrup urine disease.

### Maximum Quantity

MSUD gel 30 x 24g x 4 with 5 repeats.  
MSUD express15 30 x 25g x 4 with 5 repeats.  
MSUD express20 30 x 34g x 4 with 5 repeats.  
MSUD cooler10 30 x 87ml x 4 with 5 repeats.  
MSUD cooler15 30 x 130ml x 4 with 5 repeats.  
MSUD cooler20 30 x 174ml x 4 with 5 repeats.  
MSUD amino5 30 x 6g x 12 with 5 repeats.

### Flavour options

**MSUD gel** and **MSUD express** are available unflavoured. Flavour pacs can be provided free of charge.

**Cooler10** and **cooler20** are available in **red**. **Cooler15** is available in **orange** and **red**.

## Valine50 Valine1000 Isoleucine50 Isoleucine1000

### PBS Listed

Restricted benefit for patients with proven maple syrup urine disease.

### Maximum Quantity

Isoleucine50 30 x 4g x 4 with 5 repeats.  
Isoleucine1000 30 x 4g x 4 with 5 repeats.  
Valine50 30 x 4g x 4 with 5 repeats.  
Valine1000 30 x 4g x 4 with 5 repeats.

### Important

The products featured on this page are foods for special medical purposes and must be used under medical supervision. MSUD gel and Prozero are suitable from 6 months onwards, MSUD express, cooler, amino5 and basecal200 are suitable from 3 years onwards. No product is suitable as a sole source of nutrition. For enteral use only.

**For samples of any featured product  
contact Vitaflo to order.**



### Vitaflo's free home pharmacy service.

Call 1800 230 889 or email [v2u@vitaflo.com.au](mailto:v2u@vitaflo.com.au) to register for this convenient and time saving delivery service.

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Innovation in Nutrition

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