

# MSUD *gel*™

A leucine, isoleucine and valine free drink or gel for use in the dietary management of Maple Syrup Urine Disease (MSUD).

For children of 12 months to 10 years of age.

## PRESENTATION

MSUD gel is a leucine, isoleucine and valine free powder containing a balanced mixture of the other essential and non-essential amino acids, carbohydrates, minerals, trace elements and vitamins.

MSUD gel is available unflavoured. Separate FlavourPac sachets are available in orange, lemon, raspberry and blackcurrant.

## INDICATIONS

MSUD gel is for use in the dietary management of proven Maple Syrup Urine Disease in children from 12 months to 10 years of age. MSUD gel contains the necessary amino acids, minerals, trace elements and vitamins to meet the needs of this age group.

Nutritional Information							
Typical composition of the dry powder		100g	20g sachet	Typical composition of the dry powder			
		100g	20g sachet	100g	20g sachet		
Energy							
	kJ	1428	286	Vitamin C	mg	45	9
	kcal	342	68	Vitamin K	µg	25	5
Protein equivalent	g	42	8.4	Thiamin	mg	0.8	0.16
Total amino acids	g	50	10	Riboflavin	mg	1.0	0.2
Carbohydrate	g	43	8.6	Niacin	mg	13.5	2.7
of which sugar	g	28	5.6	Vitamin B <sub>6</sub>	mg	1.1	0.2
Total fat	g	<0.5	<0.1	Folic acid	µg	130	26
				Vitamin B <sub>12</sub>	µg	2	0.4
				Biotin	µg	25	5
				Pantothenic acid	mg	3.75	0.75
<b>Minerals</b>							
Sodium	mg	380	76	<b>Amino acids</b>			
	mmol	16.3	3.26	L-Alanine	g	5.1	1.02
Potassium	mg	936	187	L-Arginine	g	4.83	0.97
	mmol	24.3	4.8	L-Aspartic acid	g	6.12	1.22
Chloride	mg	582	117	L-Cystine	g	1.45	0.29
	mmol	16.3	4.1	L-Glutamine	g	5.5	1.1
Calcium	mg	1085	217	Glycine	g	4.05	0.81
Phosphorus	mg	827	165	L-Histidine	g	1.74	0.35
Magnesium	mg	168	34	L-Isoleucine	g	0.00	0.00
				L-Leucine	g	0.00	0.00
<b>Trace Elements</b>							
Iron	mg	10	2	L-Lysine	g	3.64	0.73
Copper	mg	0.7	0.14	L-Methionine	g	1.04	0.21
Zinc	mg	10	2	L-Proline	g	4.28	0.86
Manganese	mg	1.7	0.34	L-Phenylalanine	g	2.72	0.54
Iodine	µg	120	24	L-Serine	g	2.89	0.58
Molybdenum	µg	50	10	L-Threonine	g	2.61	0.52
Selenium	µg	30	6	L-Tryptophan	g	1.45	0.29
Chromium	µg	70	14	L-Tyrosine	g	2.89	0.58
				L-Valine	g	0.00	0.00
<b>Vitamins</b>							
Vitamin A	µg	600	120	L-Carnitine	mg	0.12	0.02
Vitamin D	µg	12	2.4	Taurine	mg	0.11	0.02
Vitamin E	mg	9	1.8				

## DOSAGE & ADMINISTRATION

Dosage of MSUD gel to be determined by the clinician or dietitian.

1. The daily protein requirement is given as MSUD gel (100g is equivalent to 42g of protein).
2. Additional protein is provided from natural protein sources in medically prescribed quantities to provide Leucine, Isoleucine and Valine in controlled amounts which is essential for normal development.
3. Foods high in carbohydrate and fat but low in protein should be provided (as an energy source) to supply the remainder of the calories.
4. A supplement of Isoleucine and Valine is given if required (if plasma concentrations fall too low).

## PREPARATION GUIDELINES

MSUD gel can be taken as a gel or as a low volume drink. Empty the contents of a sachet into the beaker provided, add a FlavourPac sachet if required and add cold water. 30ml for a gel or half way up (100ml) for a drink. Secure the lid and shake for 10 seconds. For the gel leave for 2 mins, for a drink serve immediately.

Water or permitted drinks should be taken after the MSUD gel.

This product should be freshly prepared and when possible used immediately. However, any unused portions should be refrigerated and used within 24 hours - re-shake before use.

## IMPORTANT NOTICE

Must only be given to patients with proven Maple Syrup Urine Disease under strict medical supervision.

Not recommended as a sole source of nutrition.

Contains a source of phenylalanine.

Not for intravenous use.

Not recommended for infants under 12 months of age.

## STORAGE

Store in a cool, dry place.

## PACK SIZE

MSUD gel is provided in packs of 30 x 20g sachets.



Innovation in Nutrition

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