

Product Information

– for professionals only –



Valin

In Short

- Valin is a Food for Special Medical Purposes (FSMP) for use in the dietary management of rare metabolic disorders, e.g. Maple Syrup Urine Disease (MSUD), Methylmalonic Aciduria (MMA) or Propionic Aciduria (PA), when supplementation with L-Valine is indicated
- L-Valine – in powder form
- suitable for tube feeding
- 100 g tin

Product Profile

Valin is highly concentrated L-Valine in powder form.

Valin is not fortified with micro-nutrients.

Administration

Valin should be taken along with other food and supplementary to the regular protein supplement – if applicable. Valin is also suitable for tube feeding.

Preparation

Valin may be mixed with the protein supplement as needed and taken with it. Stirred into liquids, such as water, (diluted) juice or milk (substitute), it should be drunk quickly. Valin may also be prepared with fruit puree and other foods permitted within the scope of the respective dietary management.

Always weigh the amount of Valin needed.
Always prepare freshly.

Function Valin is suitable for prevention or correction of disease related L-Valine deficiencies.

Indication Valin is used in the dietary management of rare metabolic disorders, e.g. Maple Syrup Urine Disease (MSUD), Methylmalonic Aciduria (MMA) or Propionic Aciduria (PA), in order to prevent or adjust inadequate serum levels of Valine, caused by the low protein diet. Adequate blood levels of Valine are particularly essential in metabolic crisis in MSUD. They stimulate protein synthesis and help to reduce serum Leucine-concentration.

Furthermore higher concentrations of Valine can help to reduce the elevated influx

of L-Leucine into the brain by competitive inhibition at the blood-brain barrier.

Skin lesions and dermatitis, which can be observed occasionally in Maple Syrup Urine Disease, Methylmalonic Aciduria or Propionic Aciduria, are often associated with Valine deficiencies. Supplementation with this amino acid may help to remove these symptoms.

Dosage The daily dosage depends on age, body weight and individual medical condition, and is, just as the right time for the daily intake, determined under medical supervision.

Valin can easily be combined with the products of the ZeroVIL- or ZeroTVMI-system.

Important Notice Must only be used under medical supervision. Not for use as a sole source of nutrition. For enteral use only. Only for people with rare metabolic disorders, e.g. MSUD, MMA or PA. Valin is not suitable for infants in the first year of life.

References:

- De Raeve et al. (1994) Acrodermatitis enteropathica-like cutaneous lesions in organic aciduria; The Journal of pediatrics 124(3):416–20. doi: 10.1016/s0022-3476(94)70364-7.
- Lane et al. (2007) Propionic academia manifesting with low isoleucine generalized exfoliative dermatosis; Pediatric dermatology 24(5):508–10. doi: 10.1111/j.1525-1470.2007.00505.x.
- Molema et al. (2019) Decreased plasma l-arginine levels in organic acidurias (MMA and PA) and decreased plasma branched-chain amino acid levels in urea cycle disorders as a potential cause of growth retardation: Options for treatment; Molecular Genetics and Metabolism 126(4):397-405. doi:

10.1016/j.ymgme.2019.02.003 . Epub 2019 Feb 25.

- van Vliet et al. (2014) Single amino acid supplementation in aminoacidopathies: a systematic review; Orphanet Journal of Rare Diseases 9:7. doi: 10.1186/1750-1172-9-7.

NUTRITION INFORMATION

Valin 100 g

Energy	kJ	1417
	kcal	333
Fat	g	0
of which saturates	g	0
Carbohydrate	g	0
of which sugars	g	0
Protein eqv.	g	83
of which L-Valine	g	100
Salt	g	0

INGREDIENTS

L-valine.

Delivery Unit	tin 100 g
Article Number	xx-001-98019
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.