

Phenylalanin

In Short

- Phenylalanin is a Food for Special Medical Purposes (FSMP) for use in the dietary management of rare metabolic disorders, e.g. Tyrosinemia, Alkaptonuria or FARS2 Deficiency, when supplementation with L-Phenylalanine is indicated
- suitable from birth onwards
- L-Phenylalanine – in powder form
- suitable for tube feeding
- 150 g tin

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Product Profile

Phenylalanin is highly concentrated L-Phenylalanine in powder form. Phenylalanin is not fortified with micronutrients.

Suitable from birth onwards.

only. Only for people with rare metabolic disorders, e.g. Tyrosinemia, Alkaptonuria or FARS2 Deficiency. Phenylalanin is suitable from birth onwards.

References:

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- de Laet et al. (2013) Recommendations for the management of tyrosinaemia type 1; Orphanet Journal of Rare Diseases (8):8. doi: 10.1186/1750-1172-8-8.
- Kok et al. (2021) Treatment of ARS deficiencies with specific amino acids; Genetics in Medicine 23(11):2202-2207. doi: 10.1038/s41436-021-01249-z. Epub 2021 Jun 30.
- Konolova et al. (2013) Mitochondrial aminoacyl-tRNA synthetases in human disease; Molecular Genetics and Metabolism 108(4):206-11. doi: 10.1016/j.ymgme.2013.01.010. Epub 2013 Jan 26.
- Oswald et al. (2023) Treatment of Mitochondrial Phenylalanyl-tRNA-Synthetase Deficiency (FARS2) with Oral Phenylalanine; Neuropediatrics 54(5):351-355. doi: 10.1055/a-2008-4230. Epub 2023 Jan 5.
- van 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.
- van Vliet et al. (2015) Infants with Tyrosinemia Type 1: Should phenylalanine be supplemented? Journal of Inherited Metabolic Disease reports 2015:18:117-24. doi: 10.1007/8904_2014_358. Epub 2014 Sep 26.
- Wilson et al. (2000) Phenylalanine supplementation improves the phenylalanine profile in tyrosinaemia;

Preparation

Phenylalanin 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. Phenylalanin may also be prepared with fruit puree and other foods permitted within the scope of the respective dietary management.

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

Function Phenylalanin is suitable for prevention or correction of disease related L-Phenylalanine deficiencies.

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.

Phenylalanin can easily be combined with the products of the ZeroTP-system.

Important Notice Must only be used under medical supervision. Not for use as a sole source of nutrition. For enteral use

NUTRITION INFORMATION

Phenylalanin

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-Phenylalanine	g	100
Salt	g	0

INGREDIENTS

L-phenylalanine.

Delivery Unit	tin 150 g
Article Number	xx-001-98022
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.