

Alanin

In Short

- Alanin is a Food for Special Medical Purposes (FSMP) for use in the dietary management of rare metabolic disorders, e.g. Glycogen Storage Disease (GSD) Type II = Pompe Disease, when supplementation with L-Alanine is indicated
- L-Alanine – in powder form
- suitable for tube feeding
- 250 g tin

Product Profile

Alanin is highly concentrated L-Alanine in powder form.

Alanin is not fortified with micro-nutrients.

Administration

Alanin should always be taken along with other foods. Alanin is also suitable for tube feeding.

Preparation

Alanin stirred into liquids, such as water, (diluted) juice or milk, should be drunk quickly. Alanin may also be prepared with fruit puree and other foods.

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

Function Alanin is suitable for prevention or correction of disease related L-Alanine deficiencies.

Indication Alanin is used for the dietary management of rare metabolic disorders, e.g. Glycogen Storage Disease (GSD) Type II = Pompe Disease. L-alanine reduces muscle protein breakdown and resting energy expenditure.

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.

In a case study of GSD II with infantile onset, significant improvements were observed with the administration of 2 g L-alanine/kg bw/day.

In a small study with GSD II with adult onset, the administration of 140 mg L-alanine/kg bw/day showed clearly positive effects on resting energy expendi-

ture and reduced protein degradation. In a child undergoing enzyme replacement therapy, the same effects were achieved with a dosage of 500 – 600 mg/kg bw/day.

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. GSD II. Alanin is not suitable for infants in the first year of life.

References:

- Bodamer et al. (2020) The effects of L-alanine supplementation in late-onset glycogen storage disease type II; Neurology 55(5):710-2. doi: 10.1212/WNL.55.5.710.
- Bodamer et al. (2002) L-Alanine Supplementation in Late Infantile Glycogen Storage Disease; Pediatric Neurology 27(2):145-6. doi: 10.1016/s0887-8994(02)00413-7.
- Rovelli et al. (2022) L-alanine supplementation in Pompe disease (IOPD): a potential therapeutic implementation for patients on ERT? A case report; Italian Journal of Pediatrics 48(1):48. doi: 10.1186/s13052-022-01249-y.

NUTRITION INFORMATION		
Alanin		
		100 g
Energy	kJ	1417
	kcal	333
Fat	g	0
of which saturates	g	0
Carbohydrate	g	0
of which sugars	g	0
Protein equiv.	g	83
of which L-Alanine	g	100
Salt	g	0

INGREDIENTS

L-alanine.

Delivery Unit	tin 250 g
Article Number	xx-001-98028
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.



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