Product Information
– for professionals only –

Ornithin

In Short

- Ornithin is a Food for Special Medical Purposes (FSMP) for use in the dietary management of rare metabolic disorders, e.g. Arginase-1 Deficiency, or defects in Creatine Synthesis, e.g. Guanidinoacetate Methyltransferase (GAMT) Deficiency, when supplementation with L-Ornithine is indicated
- L-Ornithine – in powder form
- suitable for tube feeding
- 100 g tin

Product Profile

Ornithin is highly concentrated L-Ornithine in powder form.
Ornithin is not fortified with micro-nutrients.

Administration

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

Preparation

Ornithin 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. Ornithin may also be prepared with fruit puree and other foods permitted within the scope of the respective dietary management.
Always weigh the amount of Ornithin needed. Always prepare freshly.

Function Ornithin is suitable for prevention or correction of disease related L-Ornithine deficiencies.

Indication Ornithin is used in low doses in case of Arginase-1 Deficiency in addition to a protein-reduced diet. In case of Guanidinoacetate Methyltransferase (GAMT) Deficiency it is used in high doses alone or as a supplement to an arginine- or protein-reduced diet.
L-Ornithine reduces the formation and accumulation of neurotoxic guanidinoacetate derived from arginine and prevents the tubular reabsorption of arginine.

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.
Ornithin can easily be combined with the products of the plus8-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. Arginase-1 Deficiency, or defects in Creatin Synthesis, e.g. Guanidinoacetate Methyltransferase (GAMT) Deficiency. Ornithin is not suitable for infants in the first year of life.

References:

- Amayreh et al. (2014) Treatment of arginase deficiency revisited: guanidinoacetate as a therapeutic target and biomarker for therapeutic monitoring; Developmental Medicine and Child Neurololy 56(10):1021-4. doi: 10.1111/dmcn.12488.
- Khaikin et. al. (2018) Treatment outcome of twenty-two patients with guanidinoacetate methyltransferase deficiency: An international retrospective cohort study; European Journal of Paediatric Neurology 22(3):369-379. doi: 10.1016/j.ejpn.2018.02.007. Epub 2018 Feb 16.
- Stockler-Ipsirogluet al. (2014) Guanidinoacetate methyltransferase (GAMT) deficiency: Outcomes in 48 individuals and recommendations for diagnosis, treatment and monitoring; Molecular Genetics and Metabolism 111(1):16-25. doi: 10.1016/j.ymgme.2013.10.018. Epub 2013 Nov 7.

NUTRITION INFORMATION
Ornithin

		100 g
Energy	kJ	0
	kcal	0
Fat	g	0
of which saturates	g	0
Carbohydrate	g	0
of which sugars	g	0
Protein eqv.	g	0
Salt	g	0

Amino acids

L-Ornithine	g	78

INGREDIENTS
L-ornithine hydrochloride.

Delivery Unit	tin 100 g
Article Number	xx-001-98037
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.

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