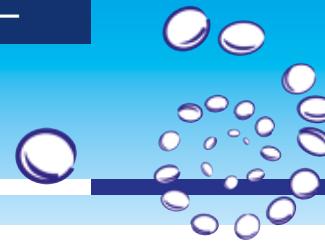


## Isoleucin<sup>100</sup> minis



### In Short

- Isoleucin<sup>100</sup> minis are 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-Isoleucine is indicated
- highly concentrated, pressed and coated L-Isoleucine – in tablet form
- easy to swallow – easy application
- neutral in smell and taste
- 100 mg (0,8 mmol) L-Isoleucine per tablet – each 100 g tin at 500 tablets

### Product Profile

Isoleucin<sup>100</sup> minis are highly concentrated L-Isoleucine in tablet form – pressed and coated.

Isoleucin<sup>100</sup> minis allow a tasteless intake of this L-amino acid.

Due to their dosage form they offer a convenient alternative, flexible and easy to take and simple to measure out.

Isoleucin<sup>100</sup> minis are not fortified with micronutrients.

**Indication** Isoleucin<sup>100</sup> minis are used in the dietary management of rare metabolic disorders, e.g. Maple Syrup Urine Disease, Methylmalonic Aciduria or Propionic Aciduria, in order to prevent or adjust inadequate serum levels of Isoleucine, caused by the low protein diet. Adequate blood levels of Isoleucine are particularly essential in metabolic crises in MSUD. They stimulate protein synthesis and help to reduce serum Leucine-concentration.

Furthermore higher concentrations of L-Isoleucine can help to reduce the elevated influx of 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 L-Isoleucine 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.

Isoleucin<sup>100</sup> minis can easily be combined with the products of the ZeroVIL-or ZeroTVM-system and with Valin<sup>100</sup> minis when indicated.

**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. Isoleucin<sup>100</sup> minis are 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 acidemia 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.

**Function** Isoleucin<sup>100</sup> minis are suitable for prevention or correction of disease related L-Isoleucine deficiencies.

NUTRITION INFORMATION			
Isoleucin <sup>100</sup> minis			
	100 g	0,20 g (1 tablet)	
<b>Energy</b>	kJ 1365	3	
	kcal 323	0,6	
<b>Fat</b>	g 1	<0,1	
of which saturates	g 1	<0,1	
<b>Carbohydrate</b>	g 29	0,06	
of which sugars	g 1,8	<0,1	
<b>Fibre</b>	g 17	0,03	
<b>Protein eqv.</b>	g 42	0,08	
of which L-Isoleucine	g 50	0,1	
	mmol 0,8		
<b>Salt</b>	g 0	0	

## INGREDIENTS

L-isoleucine, maltodextrin, bulking agent: microcrystalline cellulose, anti-caking agents: calcium silicate & magnesium stearate & magnesium carbonate & calcium phosphate, glazing agents: E 464 & E 463.

Delivery Unit	tin 500 tablets = 100 g
Article Number	xx-001-31799
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.