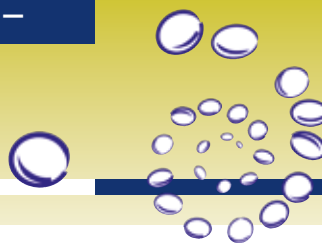


Product Information

– for professionals only –



Glycin⁵⁰⁰ minis



In Short

- Glycin⁵⁰⁰ minis are a Food for Special Medical Purposes (FSMP) for use in the dietary management of rare metabolic disorders, e.g. Isovaleric Acidemia (IVA), Serine Deficiency Disorders or Creatine Transporter Deficiency (CRTR-D), when supplementation with Glycine is indicated
- highly concentrated, pressed and coated Glycine – in tablet form
- easy to swallow – easy application
- neutral in smell and taste
- 500 mg (6,7 mmol) Glycine per tablet – 158 g tin at 250 tablets

Product Profile

Glycin⁵⁰⁰ minis are highly concentrated Glycine in tablet form – pressed and coated. Glycin⁵⁰⁰ 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.

Glycin⁵⁰⁰ minis are not fortified with micro-nutrients.

Administration

Glycin⁵⁰⁰ minis should always be taken with a sufficient quantity of liquid. They should be taken along with other food and supplementary to the regular protein supplement – if applicable.

Intake

The tablets should be swallowed unchewed as whole tablets. They should always be taken with a sufficient quantity of liquid. The tablets are not intended to be dissolved in water or other drinks prior to intake and they are not meant to dissolve in the mouth. We recommend to take the tablets in an upright position. The box must be closed tightly after use, also with the inner lid.

Note: The tablets are covered with a tasteless coating. This makes it easy to swallow them. However the layer is very thin. Thus, if the tablets were kept in the mouth for too long, this layer would dissolve and release the typical taste of an amino acid. This is why we recommend the administration above.

Function Glycin⁵⁰⁰ minis are suitable for prevention or correction of disease related Glycine deficiencies.

Indication Glycin⁵⁰⁰ minis are used for the dietary management of rare metabolic disorders, e.g. Isovaleric Acidemia (IVA), Serine Deficiency Disorders or Creatine Transporter Deficiency (CRTR-D), when supplementation with glycine is required. In CRTR-D, combined L-arginine and glycine supplementation therapy is used to alleviate epilepsy. (1)

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.

Glycin⁵⁰⁰ minis can easily be combined with the products of the ZeroLeu-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. IVA, Serine Deficiency Disorders or CRTR-D. Glycin⁵⁰⁰ minis are not suitable for infants in the first year of life.

(1) References:

- <https://www.ncbi.nlm.nih.gov/books/NBK3794/>
- Cecil KM, Salomons GS, Ball WS Jr, Wong B, Chuck G, Verhoeven NM, Jakobs C, DeGrauw TJ. Irreversible brain creatine deficiency with elevated serum and urine creatine: a creatine transporter defect? *Ann Neurol*. 2001;49:401–4
- Mercimek-Mahmutoglu S, Connolly MB, Poskitt KJ, Horvath GA, Lowry N, Salomons GS, Casey B, Sinclair G, Davis C, Jakobs C, Stockler-Ipsiroglu S. Treatment of intractable epilepsy in a female with SLC6A8 deficiency. *Mol Genet Metab*. 2010a;101:409–12.
- van de Kamp JM, Mancini GM, Salomons GS. X-linked creatine transporter deficiency: clinical

aspects and pathophysiology. *J Inherit Metab Dis*. 2014;37:715–33.

- Valayannopoulos V, Boddaert N, Chabli A, Barbier V, Desguerre I, Philippe A, Afejar A, Mazzuca M, Cheillan D, Munnich A, de Keyser Y, Jakobs C, Salomons GS, de Lonlay P. Treatment by oral creatine, L-arginine and L-glycine in six severely affected patients with creatine transporter defect. *J Inherit Metab Dis*. 2012;35:151–7.
- van de Kamp JM, Pouwels PJ, Aarsen FK, Ten Hoopen LW, Knol DL, de Klerk JB, de Coo IF, Huijman JG, Jakobs C, van der Knaap MS, Salomons GS, Mancini GM. Long-term follow-up and treatment in nine boys with X-linked creatine transporter defect. *J Inherit Metab Dis*. 2012;35:141–9.

NUTRITION INFORMATIONGlycin⁵⁰⁰ minis

100 g

0,63 g
(1 tablet)

| | | | |
|-------------------------|----|--------------------|------|
| Energy | kJ | 1300 | 8 |
| | | kcal | 308 |
| Fat | g | 1 | <0,1 |
| | | of which saturates | <0,1 |
| Carbohydrate | g | 0 | 0 |
| | | of which sugars | 0 |
| Fibre | g | 17 | 0,1 |
| | | Protein eqv. | 0,4 |
| of which Glycine | g | 80 | 0,5 |
| | | mmol | 6,7 |
| Salt | g | 0 | 0 |

INGREDIENTS

Glycine, bulking agents: microcrystalline cellulose & silicium dioxide, anti-caking agents: calcium silicate & magnesium stearate & magnesium carbonate & calcium phosphate, glazing agents: E 464 & E 463.

| | |
|----------------|-----------------------------|
| Delivery Unit | tin 250 tablets = 158 g |
| Article Number | xx-001-31813 |
| Delivery to | Pharmacies, clinics |
| Storage | Store in a cool, dry place. |