Consumer Information

Glykogea Kid

In Short

- Glykogea Kid is a powdered formula and tube feeding, rich in fat and protein, low in carbohydrate, nutritionally complete Food for Special Medical Purposes (FSMP)
- for use in the dietary management of Glycogen Storage Diseases, e.g. Glycogenosis Type III, VI and IX
- source of protein: biological high valued whey protein isolate
- high-molecular source of carbohydrate – amylopectin starch from rice
- with micronutrients in age-adapted, adequate amounts
- suitable from 1 year of age
- in 400 g tin

Product Profile

Glykogea Kid is a nutritionally complete formula and tube feeding, rich in protein and fat and low in carbohydrate, for use in the dietary management of Glycogen Storage Diseases. Suitable from 1 year of age.

In Glykogea Kid, the nutrient ratio of carbohydrates:fat:protein is 26:51:22 energy%, considering current scientific knowledge of the dietary management of GSD/GSD Type III.

As a protein source, Glykogea Kid contains biologically highly valued whey protein isolate. The recommendations for the optimal amount of protein range between 2 g/kg body weight/day (adults) and 3 g/kg body weight/day (children) or 20 to 30 percent of energy (1).

The high-molecular carbohydrate component in Glykogea Kid consists of waxy rice starch, almost exclusively of amylopectin. The source of the soluble fibre is polydextrose from partially hydrolyzed maize starch.

The fat component in Glykogea Kid consists of 50 % easily digestible MCT-lipids and 50 % LCT-lipids with an optimal ratio of ω-6- and ω-3 fatty acids.

Glykogea Kid is free from soy, egg and gluten-containing cereals and free from lactose and fructose.

Glykogea Kid is supplemented with vitamins, minerals and trace elements in age-adapted, adequate amounts by the Reference Values for Nutrient Intake from the nutrition societies from Germany, Austria and Switzerland (D-A-CH).

Good to know

According to current knowledge, a diet high in protein and fat but low in carbohydrates is favored for both children and adults with glycogen storage disease type III (1).

Due to an intact process of gluconeogenesis, protein serves as an alternative source for endogenous glucose synthesis (2). Increasing protein intake may improve muscle function by promoting muscle protein synthesis. In addition, it prevents the breakdown of endogenous muscle protein in metabolic situations where glucose is needed. The replacement of carbohydrates with protein leads to a reduction in the accumulation of glycogen and its abnormally structured intermediates in the liver as well (2, 3).

Case studies show the improvement of skeletal and cardiac myopathies with the use of a high-protein diet and the avoidance of excessive carbohydrate intake (4). Similarly, increased exercise tolerance and muscle strength, as well as improved growth, has been observed in children (3). For this reason, a high-protein diet is recommended for children with GSD IIIa with existing myopathy and growth retardation (2).

Monosaccharides should be avoided in favor of complex carbohydrates to prevent excessive glycogen storage and a rise in lactate. To keep blood glucose levels constant, starch is used in the dietary management. Increased fat content and a higher concentration of ketone bodies led to an improvement in hypertrophic cardiomyopathy (5).

Similarly, the use of MCT fats, a modified Atkins diet, and a high-protein diet has been shown to be effective in the dietary management of various forms of GSD, particularly in the case of children with GSD type III (6, 7).

Important Notice: Must only be used under medical supervision. Do not use in severe disorders of absorption and digestion. Only for people from 1 year of age with Glycogen Storage Diseases, e.g. Glycogenosis Type III, VI and IX. Suitable for exclusive or supplementary feeding. For enteral use only.

Dosage and Usage

Glykogea Kid can be used supplementary or according to energy requirements. The daily dosage depends on age, body weight and individual metabolic condition and should be given in several single portions distributed during the day. If applicable begin with low, then gradually increasing doses.

The individual dosage is calculated on the basis of the daily energy requirements according to the Reference Values for Nutrient Intake from the nutrition societies from Germany, Austria and Switzerland (D-A-CH) and should be re-examined and adjusted regularly under medical control.

Glykogea Kid is mainly used as a tube feeding, but can also be prepared as a formula (drink). Glykogea Kid dissolves easily in cold and warm water.

Always prepare freshly. Do not reuse leftovers!

Preparation

Measure or rather weigh out amount of powder according to the required energy density. Preferably use a shaker for preparation, so that the powder dissolves optimally. See pictograms.

- **Formula (Drink)**

Stir required amount of powder into water with favoured temperature and dissolve.

- **Tube Feeding**

Stir required amount of powder into boiled water, cooled down to approx. 40 °C, and dissolve lump-free. Glykogea Kid can be applied using standard tubes.

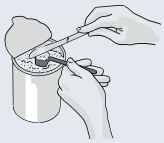
Preparation with Shaker

Clean shaker before use and boil it out.

Boil water and let cool down to approx. 40 °C. Pour water into the shaker.

Loosen the powder with the dosing aid.

Take powder loosely with the dosing aid.



Strip off excess powder with a knife back.

Determine the tare weight of the shaker. Carefully empty the dosing aid into the shaker. Weigh out and thus verify the amount of powder.

Close shaker and shake.

Open shaker and fill content into the tube bag. Feed by tube.

References:

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(3) Derks TG, Smit GP (2015) Dietary management in glycogen storage disease type III: what is the evidence? J Inherit Metab Dis. 2015 May;38(3):545-50. doi: 10.1007/s10545-014-9756-x. Epub 2014 Aug 28.

(4) Sentner CP, Caliskan K, Vletter WB, Smit GP.(2012) Heart Failure Due to Severe Hypertrophic Cardiomyopathy Reversed by Low Calorie, High Protein Dietary Adjustments in a Glycogen Storage Disease Type IIIa Patient. JIMD Rep. 2012; 5:13-6. doi: 10.1007/8904_2011_111. Epub 2011 Dec 13.

(5) Valayannopoulos V, Bajolle F, Arnoux JB, Dubois S, Sannier N, Baussan C, Petit F, Labrune P, Rabier D, Ottolenghi C, Vassault A, Broissand C, Bonnet D, de Lonlay P. (2011) Successful treatment of severe cardiomyopathy in glycogen storage disease type III With D,L-3-hydroxybutyrate, ketogenic and high-protein diet. Pediatr Res. 2011 Dec;70(6):638-41. doi: 10.1203/PDR.0b013e318232154f.

(6)Rossi A, Hoogeveen I, Bastek V, de Boe F, Montanari C, Meyer U, Maiorana A, Bordugo A, Dianin A, Campana C, Rigoldi M. (2020), Dietary lipids in glycogen storage disease type III: A systematic literature study, case studies, and future recommendations, Journal of Inherited Metabolic Disease, Volume 43, Issue 4 p. 770-777, https://doi.org/10.1002/jimd.12224

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NUTRITION INFORMATION

Glykogea Kid		100 g powder	30 g powder	per 100 kcal
Energy	kJ	2113	634	418
	kcal	506	152	100
Fat, of which	g	29	9	5
saturates	g	16	5	3
mono-unsaturates	g	7	2	1,4
polyunsaturates	g	5	1,5	1
Carbohydrate, of which	g	33	10	7
sugars, of which	g	0	0	0
Lactose	g	0	0	0
Fructose	g	0	0	0
Fibre	g	2,6	0,8	0,5
Protein	g	28	8	5
Salt	g	0,6	0,2	0,11
Vitamins				
Vitamin A	µg	312	94	58
Vitamin D3	µg	8	2,4	1,5
Vitamin E	mg	3	1	0,6
Vitamin K1	µg	18	6	3,4
Vitamin C	mg	32	9	6
Thiamin (Vit. B1)	mg	0,3	0,1	0,06
Riboflavin (Vit. B2)	mg	0,4	0,13	0,08
Niacin	mg	4	1	0,7
Vitamin B6	mg	0,1	0,03	0,02
Folic acid	µg	105	32	20
Vitamin B12	µg	2	0,6	0,4
Biotin	µg	3,5	1	0,7
Pantothenic acid	mg	2	0,6	0,4
Minerals				
Sodium	mg	230	69	46
	mmol	10	3	2
Potassium	mg	525	158	98
	mmol	13	4	2,5
Chloride	mg	233	70	43
	mmol	7	2	1,2
Calcium	mg	315	94	59
	mmol	8	2,4	1,5
Phosphorus	mg	237	71	44
	mmol	8	2,3	1,4
Magnesium	mg	42	13	8
	mmol	1,7	0,5	0,3

		100 g powder	30 g powder	per 100 kcal
Trace elements				
Iron	mg	3	0,9	0,6
Zinc	mg	1,6	0,5	0,3
Copper	mg	0,26	0,08	0,05
Manganese	mg	0,5	0,16	0,1
Fluoride	mg	0,3	0,09	0,06
Selenium	µg	11	3	2
Chromium	µg	7	2	1,3
Molybdenum	µg	21	6	4
Iodine	µg	53	16	10

Standard dilution
30 g Glykogea Kid + 100 ml drinking water
= 120 ml ready-to-drink feed

The level dosing aid equals approx. 11 g Glykogea Kid.
We recommend the use of a scale for weighing out the required amount of Glykogea Kid.

Osmolality

Glykogea Kid (g)	Drinking water (ml)	Osmolality (mosmol/kg)
30	100	128

INGREDIENTS

Rice starch, ***whey protein***, vegetable oils (MCT-oil, rape seed oil, corn oil, palm oil), dextrin, calcium phosphate, L-lysine acetate, L-tryptophan, L-isoleucine, maltodextrin, vitamin B2, vitamin B1, potassium chloride, magnesium carbonate, ferrous sulphate, sodium chloride, vitamin C, zinc sulphate, vitamin E, sodium fluoride, vitamin D, niacin, manganese sulphate, pantothenic acid, cupric sulphate, vitamin B6, vitamin A, folic acid, chromium (III) chloride, sodium selenite, potassium iodide, sodium molybdate, vitamin K, biotin, vitamin B12.

Delivery Unit	tin 1 x 400 g
Article Number	xx-001-51001
Delivery to	Pharmacies, clinics
Storage	Store in a cool, dry place.

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