Phenactin AA Plus 20 gram Protein Equivalent

PRODUCT INFORMATION
Phenactin AA Plus 20 gram Protein Equivalent
250 mL carton (8.5 fl. oz) Reimbursement Code: 24359-0705-03
Manufactured by Cambrooke Therapeutics, Inc. Ayer, MA 01432 www.cambrooke.com

MLT37005D1

DISPENSE BY PRESCRIPTION
Phenactin AA Plus is a phenylalanine-free medical food for the dietary management of Phenylketonuria (PKU).

DESCRIPTION

Phenactin AA Plus is a specially formulated ready to drink prescription medical food for the clinical dietary management of phenylalanine hydroxylase deficiency (phenylketonuria or PKU) and hyperphenylalanemia.

Phenactin AA Plus is to be used only under medical supervision. Phenactin AA Plus has been developed, labeled and should be administered in accordance with the Food and Drug Administration's (FDA's) regulatory definition of Medical Foods.

Congress defines "Medical Food" in the Orphan Drug Act and Amendments of 1988 as a formulation to be administered enterally (for oral or tube feeding) under the supervision of a physician and which is intended for the specific dietary management of a disease or condition for which distinctive nutritional requirements, based on recognized scientific principles, are established by medical evaluation. 21 U.S.C. 360ee(b)(3).

Phenactin AA Plus is a ready-to-drink phenylalanine-free medical food supplied in single dose, opaque 250 mL cartons, 30 cartons per case. Each carton contains 181 calories, 20 grams of protein equivalent per serving and a complete micronutrient profile.

PRIMARY INGREDIENTS

Phenactin AA Plus is a phenylalanine-free amino acid supplement that also contains carbohydrates, fats and a complete micronutrient profile. Phenactin AA Plus is indicated for individuals with phenylketonuria and hyperphenylalanemia as those with these conditions are unable to adequately breakdown phenylalanine. Phenactin AA Plus is not intended as a sole source of nutrition and must be balanced with a tolerated phenylalanine intake, typically from foods or intact protein source. It is generally recommended that a phenylalanine-free supplement in conjunction with a low protein diet be continued as life-long treatment for optimal health.[1]

Micronutrients and Macronutrients

Individuals with phenylalanine hydroxylase deficiency should limit their intake of the essential amino acid phenylalanine. Phenylalanine is found naturally in nearly all protein containing foods, including meats, dairy, legumes, grains, and to a lesser degree, vegetables and fruits. Such a severely restricted diet brings meaningful risk and challenges to receiving recommended daily intake of essential nutrients. To compensate for this, Phenactin AA Plus provides phenylalanine-free protein equivalent to help provide essential and non-essential amino acids, adequate caloric intake with a balance of carbohydrate and fats and a complete micronutrient profile. Some amount of phenylalanine is still essential and is typically consumed in a low protein diet.

Essential Fats

Phenactin AA Plus contains essential fatty acids in the amount of 1145 mg linoleic acid and 549 mg linolenic acid. Phenactin AA Plus also contains 150 mg of algae-sourced docosahexaenoic acid (DHA). Essential fatty acids cannot be produced in the body and therefore must be obtained through diet. Phenactin AA Plus contains essential fatty acids and DHA to help support brain and eye development.[2]

Digestive and Oral Health

Phenactin AA Plus contains 2 grams per serving of inulin, a pre-biotic fiber which helps to support digestive health and enhance mineral absorption.[3]

Phenactin AA Plus provides a more neutral pH compared to other ready to drink amino acid based formulas. Ready to drink amino acid formulas typically have a low or acidic pH. With frequent consumption, this may lead to oral health problems. A pH less than 5.5 may lead to dental erosion.[4] Phenactin AA Plus has a pH of 5.8 making it gentler on digestion and tooth enamel.

Bone Health

Phenactin AA Plus contains a bone health blend designed to optimize bone health. Bone health is a growing concern in individuals with inherited disorders of metabolism. Studies have found that 30-50% of those with phenylketonuria have a decreased bone mineral density[5,6,7,8,9] which may lead to osteoporosis and fractures if not detected and treated.[6,7,8] Phenactin AA Plus contains a unique blend of nutrients essential to bone health including 677 mg calcium, 15.5 mcg vitamin D, 664 mg phosphorus, 5 mg zinc, and 143 mg magnesium per serving. In addition, Phenactin AA Plus contains 40 mcg of vitamin K per serving in the forms of both K1 and K2.

Vitamin K1 is activated to K2 in the body and plays a role as co-factor in the carboxylation of osteocalcin. Lower levels of carboxylated osteocalcin are associated with increased fracture risk.[10] In addition, Phenactin AA Plus contains 2 grams per serving of inulin, a soluble prebiotic fiber. Inulin consumed at 3 grams per day has been shown to aid in calcium absorption.[3]

Complete Ingredients

Water, amino acid blend (L-leucine, L-tyrosine, L-lysine HCl, L-valine, L-aspartic, L-proline, L-threonine, L-isoleucine, L-serine, L-arginine, L-histidine, L-cystine, L-glycine, Lmethionine, L-tryptophan, L-alanine, taurine, L-carnitine), vitamins and minerals (calcium lactate, monosodium phosphate, monopotassium phosphate, choline bitartrate, magnesium phosphate, sodium ascorbate, ascorbic acid, dl-alpha-tocopheryl acetate, ferrous fumarate, niacinamide, zinc sulfate, calcium d-pantothenate, manganese sulfate, riboflavin, B6 pyridoxine HCl, thiamin HCl, copper gluconate, folic acid, vitamin A palmitate, potassium iodide, sodium selenite, K1 phytonadione, K2 MK-7, sodium molybdate, cholecalciferol, chromium chloride, biotin, B12 cyanocobalamin), canola oil, sugar, natural flavors (propylene glycol, water, maltodextrin, modified food starch), inulin, cellulose gel and carboxymethylcellulose sodium, food starch modified, sodium hexametaphosphate, DHA algal oil, acesulfame potassium, carrageenan, sucralose.

INGREDIENT SAFETY

The ingredients in Phenactin AA Plus are all approved food additives or Generally Recognized As Safe (GRAS) for the use in a medical food for individuals that have PKU. GRAS is the statutory safety standard of the U.S. Food and Drug Administration (FDA). The standard for an ingredient to achieve GRAS status for a certain use requires technical demonstration of non-toxicity and safety, general recognition of safety through widespread usage and agreement by experts in the field.

MEDICAL FOOD STATUS

INDICATIONS FOR USE

Phenactin AA Plus is a ready to drink phenylalanine-free medical food for the dietary management of individuals under a physician's care for phenylalanine hydroxylase deficiency (phenylketonuria) or hyperphenylalaninemia.

CLINICAL EXPERIENCE

Individuals with phenylketonuria (PKU) are unable to adequately metabolize the essential amino acid, phenylalanine. As a result, phenylalanine and its by-products can accumulate in the blood. Protein naturally contains phenylalanine, and as such, protein from the diet needs to be significantly reduced to ensure phenylalanine consumption is limited. Protein, however, is essential for the body and is vital in the maintenance of body tissue, including development and repair. Even though individuals with phenylketonuria cannot have large amounts of phenylalanine, they still require other amino acids to help maintain healthy muscle and tissue. The major source of dietary protein for those with an amino acid metabolism disorder consists of mixtures of synthetic amino acid formulas devoid of the offending amino acid(s) and a small amount of dietary protein mostly from fruits and vegetables or alternative intact protein source.[11] The use of synthetic amino acid formulas and low protein diet is the standard of care for those with amino acid metabolism disorders.[1] Without control of phenylalanine consumed, individuals with phenylalanine hydroxylase deficiency can develop significant intellectual disability.[12] Initial treatment of a low protein diet and appropriate medical food, such as Phenactin AA Plus, should be started as early as possible and be continued as life-long treatment.[13]

PHARMACOKINETICS

Phenylketonuria is caused by an absence or virtual absence of phenylalanine hydroxylase (PAH) enzyme activity. PAH is necessary to break down excess phenylalanine from food. In people without PKU, the PAH enzyme breaks down excess phenylalanine. Those with PKU do not have enough of the PAH enzyme or its cofactor, which can cause phenylalanine to build up in the blood and brain to toxic levels, affecting brain development and function. With the use of a synthetic amino acid formula, patients are able to consume adequate protein equivalent with the remaining amino acids. Amino acid formulas typically provide 85% of protein needs for those with PKU.[14] Since amino acids are already broken down to their simplest form, they may not be absorbed as efficiently as intact protein. It is recommended that those with PKU consume 20-40% more protein than the Dietary Reference Intakes (DRIs.).[1,15,16]

Precautions and Contraindications

Phenactin AA Plus is intended to help meet nutritional requirements for patients 12 months and older with diagnosed phenylketonuria. It is not to be used as sole source nutrition. Individuals with other inborn errors of amino acid metabolism or those without a phenylketonuria diagnosis can experience complications if using this product due to its lack of phenylalanine which contributes to mood regulation, alertness, dopamine transmission, learning and memory. Failure to consume some amount of the essential amino acid phenylalanine can lead to death.[1,13]

Adverse Reactions

Post – marketing surveillance has shown no adverse reactions.

Drug Interactions

None known.

Toxicity

None known.

SPECIAL POPULATIONS

Indicated for phenylketonuria patients over 12 months of age. Always check with the monitoring physician for proper dosage recommendations.

Compliance to a low phenylalanine diet must accompany the use of Phenactin AA Plus for all phenylketonuria patients. This is especially important for those considering having children or who are pregnant due to the potential birth defects with high phenylalanine levels.

DOSAGE AND ADMINISTRATION

Phenactin AA Plus is a medical food to be administered, enterally by mouth or tube, under the supervision of a physician.

Recommended daily requirements vary with age, weight and activity levels. Follow recommendation of the medical practitioner to determine the best amount of Phenactin AA Plus to be used each day.

HOW SUPPLIED

Phenactin AA Plus provides 20 grams Protein Equivalent per serving and is supplied in 250 mL (8.5 fl. oz.) cartons.

The cartons are packaged 30 per case (reimbursement code: 24359-0705-03). Keep sealed in a cool, dry place. Refrigerate after opening. Do not freeze.

REFERENCES

1 Singh, R.H., Rohr, F., et al. Recommendations for the nutritional management of phenylalanine hydroxylase deficiency. Gen Med 16; 2 (2014)121-131.

2 Singh M. Essential fatty acids, DHA and human brain. Indian J Pediatr. 2005 Mar;72(3):239-42.

3 Scholz-Ahrens KE, Schrezenmeir J. Inulin, oligofructose and mineral metabolism - experimental data and mechanism. Br J Nutr. 2002 May;87 Suppl 2:S179-86.

4 Touger-Decker, R., van Loveren, C. Sugars and dental caries. The American Journal of Clinical Nutrition. 78, 8815-8925.

5 Allen JR, Humphries IRJ, Walters D., et al. Decreased Bone Mineral Density in Children with Phenylketonuria. Am J Clin Nutrition 1994: Vol 59: 419-22.

6 Koura, H.M., Abdallah Ismail N., Kamel, A.F., Ahmed, A.M., Saad Hussein, A., Effat, L.K. A long-term study of bone mineral density in patients with phenylketonuria under diet therapy, Arch. Med. Sci. 7 (2011) 493–500.

7 Miras, A. Boveda, M.D., et al. Risk factors for developing mineral bone disease in phenylketonuria patients. Mol. Gen. Met. 108 (2013) 149-154.

8 Pérez Dueñas, B., Cambra, F.J., Vilaseca, M.A., Lambruschini, N., Campistol, J., Camacho, J.A. New approach to osteopenia in phenylketonuric patients, Acta Paediatr. 91 (2002) 899–904.

9 Modan-Moses, D., Vered, I., Schwartz, G., Anikster, Y., Abraham, S., Segev, R., Efrati, O., Peak bone mass in patients with phenylketonuria, J. Inherit. Metab. Dis. 30 (2007) 202–208.

10 Marieke, J. eta al. (2008). Vitamin K status is associated with childhood bone mineral content. British Journal of Nutrition, 100, 852-858.

11 MacLeod, E.L. and Ney, D.M. Nutritional Management of Phenylketonuria Ann Nestle Eng Jun (2010); 68 (2): 58-69.

12 Mitchell JJ. Phenylalanine hydroxylase deficiency. In: Pagon RA, Adam MP, Bird TD, et al. (eds). GeneReviews™ [Internet]. University of Washington, Seattle: Seattle, WA, 2000:1993-2013.

13 Vockley J, Andersson H, Antshel KM, et al. (2014)Phenylalanine hydroxylase deficiency diagnosis and management guideline. Genet Med; 16, 2: 188-200.

14 University of Washington PKU Clinic Management Guidelines. 2013. http://depts.washington.edu/pku/resources/mgmtguidelines.html.pdf. Accessed 8/10/15.

15 MacDonald, A., Rocha, J.C., van Rijn, M., & Feillet, F. (2011). Nutrition in phenylketonuria. Mol. Genet. Metab., 104 Suppl, S10-8.

16 Genetic Metabolic Dietitian International: PKU Nutrition Management Guidelines. https://southeastgenetics.org/ngp/guidelines.php/85/nr/114/detailed/PKU%20Nutrition%20Guidelines/Version%201.10/#sd. Accessed 8/10/15.