

Vilactin AA Plus 20 gram Protein Equivalent

PRODUCT INFORMATION

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250 mL carton (8.5 fl. oz) Reimbursement Code: 24359-0704-03

Manufactured by Cambrooke Therapeutics, Inc. Ayer, MA 01432 www.cambrooke.com

DISPENSE BY PRESCRIPTION

Vilactin AA Plus is an isoleucine, leucine, and valine-free medical food for the dietary management of Maple syrup urine disease (MSUD).

DESCRIPTION

Vilactin AA Plus is a specially formulated ready to drink prescription medical food for the clinical dietary management of Maple Syrup Urine Disease (MSUD) also called branched-chain ketoaciduria.

Vilactin AA Plus is to be used only under medical supervision. Vilactin 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).

Vilactin AA Plus is a ready-to-drink isoleucine, leucine and valine-free medical food supplied in single dose, opaque 250 mL cartons, 30 cartons per case. Each carton contains 183 calories, 20 grams of protein equivalent per serving and a complete micronutrient profile.

PRIMARY INGREDIENTS

Vilactin AA Plus is an isoleucine, leucine, and valine-free amino acid supplement that also contains carbohydrates, fats and a complete micronutrient profile. Vilactin AA Plus is indicated for those with Maple Syrup Urine Disease as those with these conditions are unable to adequately breakdown the branched chain amino acids (BCAAs): isoleucine, leucine and valine. Vilactin AA Plus is not intended as sole source nutrition and must be balanced with a tolerated natural protein intake, typically from foods or intact protein source. It is generally recommended that a branched chain amino acid-free supplement in conjunction with a low protein diet be continued as life-long treatment for optimal health unless patient undergoes liver transplant therapy.¹

Micronutrients and Macronutrients

Individuals with MSUD should limit their intake of the essential branched-chain amino acids, isoleucine, leucine, and valine. Branched chain amino acids are 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, Vilactin AA Plus provides isoleucine, leucine, and valine-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 branched chain amino acids are essential and are typically consumed with a low protein.

Essential Fats

Vilactin AA Plus contains essential fatty acids in the amount of 1145 mg linoleic acid and 549 mg linolenic acid. Vilactin 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. Vilactin AA Plus contains essential fatty acids and DHA to help support brain and eye development.^{2,3}

Digestive and Oral Health

Vilactin AA Plus contains 2 grams per serving of inulin, a pre-biotic fiber which helps to support digestive health and enhance mineral absorption.⁴

Vilactin AA Plus provides a more neutral pH compared to other ready to drink amino acid based formulas. Ready to drink amino acid based 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.⁵ Vilactin AA Plus has a pH of 5.8 making it gentler on digestion and tooth enamel.

Bone Health

Vilactin 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, another error of metabolism that requires a similar diet therapy, have a decreased bone mineral density^{6,7,8} which may lead to osteoporosis and fractures if not detected and treated. Vilactin AA Plus contains a unique blend of nutrients essential to bone health including 677 mg calcium, 620 IU vitamin D, 664 mg phosphorus, 5 mg zinc, and 143 mg magnesium per serving. In addition, Vilactin 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.⁹ In addition, Vilactin 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.⁴

Complete Ingredients

Water, amino acid blend (L-glutamine, L-alanine, L-lysine HCl, L-phenylalanine, L-tyrosine, L-threonine, L-histidine, L-proline, asparagine, L-tryptophan, L-arginine, L-serine, L-glutamic acid, glycine, L-cystine, L-methionine, taurine, L-carnitine), vitamins and minerals (calcium lactate, monosodium phosphate, monopotassium phosphate, choline bitartrate, magnesium amino acid chelate, sodium ascorbate, ascorbic acid, dl-alpha-tocopheryl acetate, iron amino acid chelate, niacinamide, zinc amino acid chelate, calcium d-pantothenate, manganese sulfate, riboflavin, B6 Pyridoxal Phosphate, thiamin HCl, copper gluconate, L-methylfolate, vitamin A palmitate, potassium iodide, sodium selenite, cholecalciferol, K1 phytonadione, K2 MK-7, sodium molybdate, cholecalciferol, chromium chloride, biotin, B12 methylcobalamin), canola oil, sugar, natural flavors (propylene glycol, water, potassium sorbate), inulin, cellulose gel and carboxymethylcellulose sodium, food starch modified, sodium hexametaphosphate, DHA algal oil, acesulfame potassium, carrageenan, sucralose. Contains corn and soy.

INGREDIENT SAFETY

The ingredients in Vilactin AA Plus are all approved food additives or Generally Recognized As Safe (GRAS) for the use in a medical food for individuals that have MSUD. 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

Vilactin AA Plus is a ready to drink isoleucine, leucine, and valine-free medical food for the dietary management of individuals under a physician's care for maple syrup urine disease (MSUD) or branched-chain ketoaciduria.

CLINICAL EXPERIENCE

Individuals with maple syrup urine disease (MSUD) are unable to adequately metabolize the branched-chain amino acids, isoleucine, leucine, and valine. As a result, isoleucine, leucine, valine, and their toxic by-products (ketoacids) build-up in the blood and urine. Protein naturally contains branched-chain amino acids, and as such, protein from the diet needs to be significantly reduced to ensure isoleucine, leucine, and valine 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 MSUD cannot have large amounts of the branched-chain amino acids, 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.^{1,10} 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 isoleucine, leucine, and valine consumed, individuals with MSUD can develop irritability, poor feeding, and lethargy, leading to encephalopathy, coma, respiratory failure, and death.¹ Initial treatment of a low protein diet and appropriate medical food, such as Vilactin AA Plus, should be started as early as possible and be continued as life-long treatment.¹

PHARMACOKINETICS

MSUD is caused by an absence or virtual absence of branched-chain alpha-keto acid dehydrogenase complex (BCKDC). BCKDC is necessary to break down excess leucine, isoleucine, and valine from food. In people without MSUD, the BCKDC breaks down excess leucine, isoleucine, and valine. Those with MSUD do not have enough of the BCKDC or its cofactors, which can cause isoleucine, leucine, and valine 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 50 - 75% of protein needs for those with MSUD depending on postprandial plasma concentrations of the BCAAs.¹ 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 MSUD consume 30-35% more protein than the Dietary Reference Intakes (DRIs).¹

Precautions and Contraindications

Vilactin AA Plus is intended to help meet nutritional requirements for patients 12 months and older with diagnosed MSUD. It is not to be used as sole source nutrition. Individuals with other inborn errors of amino acid metabolism or those without an MSUD diagnosis can experience complications, including death, if using this product due to its lack of BCAAs. Failure to consume some amount of the essential branched-chain amino acids can lead to failure to thrive, lethargy, and decreased immune function, and even death.¹

Vilactin AA Plus contains corn and soy and is not suitable for individuals with an allergy to corn and/or soy.

Adverse Reactions

Post – marketing surveillance has shown no adverse reactions.

Drug Interactions

None known.

Toxicity

None known.

SPECIAL POPULATIONS

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

Compliance to a low isoleucine, leucine, and valine diet must accompany the use of Vilactin AA Plus for all MSUD disease patients.

Compliance to a low protein diet must accompany the use of Vilactin AA Plus for all patients with maple syrup urine disease, including those considering having children or who are pregnant.^{1,9}

DOSAGE AND ADMINISTRATION

Must be administered under physician supervision.

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

HOW SUPPLIED

Vilactin 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: 24359-0704-03). Keep sealed in a cool, dry place. Refrigerate after opening. Do not freeze.

REFERENCES

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