



# Nutritional Supplements In Hutchinson-Gilford Progeria Syndrome

## Information for Families and Caregivers From The Progeria Research Foundation

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## **Our Goal**

The Progeria Research Foundation hopes to improve health care of children with Hutchinson-Gilford Progeria Syndrome (HGPS) by providing medical and health recommendations to their families, physicians and caretakers. Nutrition is a very difficult daily aspect of HGPS, because these children often have extremely poor appetites. Improved caloric intake may result in better skin and nail health, improved energy level and mood, and therefore improved quality of life. We have been asked by several families with HGPS children to provide recommendations for improved nutrition and calorie intake.

Some children with HGPS have high cholesterol or increases in other blood lipids, and all children with HGPS develop heart disease. Most of the nutritional supplements described here provide balanced nutrition, including fats and carbohydrates. There is currently no way to know whether a low-fat diet has any effect on the progression of the heart disease in HGPS. Therefore, we ask that you consult with your nutritionist or doctor if you have concerns about whether your child should be on a low-fat diet.

This nutritional information is presented in the following three parts:

1. Convenient summary table with information about where to obtain products
2. Detailed description of dietary supplements of value for children with trouble getting enough calories
3. Tasty Tips for how to make these supplements into foods your children will enjoy.

We hope that this nutritional recommendation information provides you with helpful information for use in current and future decisions about nutrition for children with Progeria.

## **Disclaimer**

This document may contain references to products or services not available in all countries.

Although we hope that the recommendations we provide are helpful to children with Progeria, The Progeria Research Foundation, Inc. makes no representations or warranties of any kind with respect to the products or publications in this document, either express or implied.

Each individual is different and will experience different results when following the recommendations contained in this document. We cannot guarantee positive results for any individual using any of the products or following any of the recommendations mentioned in this document.

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# Nutritional Supplements Information

We realize that some of these products may not be available to you where you live. However you can use the information in this document as a guideline to help you find similar products made by other manufacturers. In the following pages you will find information\* about these nutritional supplements:

<b>Product</b>	<b>Manufacturer</b>	<b>Recommended Age for Use**</b>	<b>Lactose Tolerance and Nutritional Status</b>	<b>Where Available (Not an exhaustive list, please check for local availability)</b>
PEDIASURE®	Abbott Laboratories (Ross Products Division)	1-10 years old	Lactose Free, Nutritionally Complete	Australia, Canada, Italy (as powder called Rinforza), Mexico, Netherlands, Puerto Rico, UK and USA
KINDERCAL®	Bristol Myers Squibb (Mead Johnson Nutritionals)	1-10 years old	Lactose Free, Nutritionally Complete	Argentina (powder), Mexico (powder), Puerto Rico, USA, and Venezuela (powder).
SCANDISHAKE®	Axcan Scandipharm	5+ years old	Comes in Lactose Free and Non-Lactose Free, Not Nutritionally Complete	Argentina, Brazil, Canada, Chile, Puerto Rico, UK and USA.
ENSURE®	Abbott Laboratories (Ross Products Division)	10+ years old	Lactose Free, Nutritionally Complete	Austria, Australia, Canada, France, Germany, Italy, Mexico, Netherlands (powdered), Puerto Rico, UK (powdered) and USA.
BOOST®	Bristol Myers Squibb (Mead Johnson Nutritionals)	10+ years old	Lactose Free, Nutritionally Complete	Canada, Puerto Rico, and USA
ENLIVE! ®	Abbott Laboratories (Ross Products Division)	All ages	Lactose Free, Not Nutritionally Complete	Australia, Germany, Puerto Rico and USA

\*This information was obtained from official company Internet sites and sales representatives. It is not designed to replace a doctor's care. To help determine the specific nutritional needs of your child, please consult your child's pediatrician.

\*\*The nutritional supplements described in this document are for use in children over the age of 1 year (please see specific age ranges listed). Methods exist for supplementing an infant's (0-11 months) diet, but require the

individualized care provided by a pediatrician and/or pediatric nutritionist.



Cost: (Prices will vary)	\$49.00 (case of 24 x 8 oz. cans)
Availability:	Australia Canada Italy (as powder called Rinforza) Mexico Netherlands Puerto Rico UK USA

PEDIASURE® is designed for children 1 to 10 years of age. Doctors and nutritionists recommend this kind of product for children who have a hard time eating enough regular food to support their nutritional needs. PEDIASURE® can be used as a snack in addition to a regular balanced diet or as the sole source of nutrition during times when a child is unable to eat regular food. For your child's specific nutritional needs please consult your physician.

- NOT for children with galactosemia
- LACTOSE FREE (except when powder is added to milk)

This product is made from water, sugar (table sugar, sugars derived from corn), milk proteins, and vegetable oils with added vitamins and minerals, some thickening agents, emulsifiers and natural and artificial flavors. It also contains "medium chain triglycerides" which are fats that are more easily absorbed than those found in regular vegetable oils.

Nutritional Information (Per 8 fl oz; 237 ml)	
Flavors: (availability may vary from country to country)	Vanilla Banana Cream Strawberry Chocolate
Nutrients:	
Energy:	237 Calories
Fat:	11.8 grams (This product is NOT low-fat.)
Protein:	7.1 grams
Carbohydrates:	26 grams
Ingredients:	
<p>Vanilla: (U)-D Water, sugar (sucrose), maltodextrin (corn), sodium caseinate, high-oleic safflower oil, soy oil, fractionated coconut oil (medium-chain triglycerides), whey protein concentrate; Less than 0.5% of: calcium phosphate tribasic, natural and artificial flavor, potassium citrate, magnesium chloride, cellulose gel, potassium phosphate dibasic, potassium chloride, soy lecithin, mono- and diglycerides, choline chloride, carrageenan, ascorbic acid, cellulose gum, m-inositol, taurine, ferrous sulfate, zinc sulfate, sodium chloride, niacinamide, alpha-tocopheryl acetate, L-carnitine, calcium pantothenate, thiamine chloride hydrochloride, pyridoxine hydrochloride, riboflavin, manganese sulfate, cupric sulfate, vitamin A palmitate, folic acid, biotin, potassium iodide, sodium selenate, sodium molybdate, phylloquinone, vitamin D3 and cyanocobalamin.</p>	



Cost: (Prices will vary)	\$7.99 (pack of 4 x 8 oz. cans)
Availability:	Argentina (powder) Mexico (powder) Puerto Rico USA Venezuela (powder)

KINDERCAL® is designed for children 1-10 years of age. This product is relatively new to the market and therefore has more limited availability than Pediasure®. Like Pediasure®, KINDERCAL® is a nutritionally complete supplement designed for children who have difficulty eating enough regular food to meet their nutritional needs. It can be used as a snack in addition to a regular balanced diet or as the sole source of nutrition during times when a child is unable to eat regular food. For your child's specific nutritional needs please consult your physician.

- NOT for children with galactosemia
- LACTOSE FREE (except when powder is added to milk)

This product is made from water, sugar (table sugar and corn syrup), milk proteins, and vegetable oils with added vitamins and minerals, some thickening agents, emulsifiers and natural and artificial flavors. It also contains "medium chain triglycerides" which are fats that are more easily absorbed than those found in regular vegetable oils.

Nutritional Information (Per 8 fl oz; 236 ml)	
Flavors: (availability may vary from country to country)	Vanilla Chocolate
Nutrients:	
Energy:	250 Calories
Fat:	10.5 grams (This product is NOT low-fat.)
Protein:	7.1 grams
Carbohydrates:	32 grams
Ingredients:	
Water, maltodextrin, sugar, milk protein concentrate, vegetable oil (canola, high oleic sunflower, MCT, corn oils), and less than 1% artificial flavor, potassium citrate, magnesium chloride, thiamin hydrochloride, riboflavin, biotin, vitamin B12, taurine, niacinamide, calcium pantothenate, vitamin B6 hydrochloride, folic acid, inositol, potassium iodide, sodium ascorbate, beta-carotene, potassium phosphate, calcium phosphate, salt, choline chloride, soy lecithin, carrageenan, carnitine, vitamin A palmitate, vitamin D3, vitamin E acetate, ferrous sulfate, zinc sulfate, cupric sulfate, sodium selenite, chromic chloride, sodium molybdate, manganese sulfate.	



<http://www.axcanscandipharm.com/products/scandishake.html>

Cost: (Prices will vary)	\$7.75 per box (4 x 8oz servings) \$41.82 (24 serving box) \$63.37 (Case of 9 boxes)
Availability:	Argentina Brazil Canada Chile Puerto Rico UK USA

SCANDISHAKE® is the most calorie dense nutritional supplement available. Mixed with 8 ounces of whole milk, a single serving of SCANDISHAKE® contains approximately 600 calories! Doctors often recommend it to people who have extreme difficulty keeping weight on due to poor nutrient absorption or poor appetite. The manufacturer does not recommend this product be given to children younger than 5 years old. This product should be used ONLY as a supplement to regular balanced meals as it DOES NOT contain all the vitamins and minerals your child will need in a day. To determine if a product like this is right for your child, please consult your physician.

- A LACTOSE FREE version is available which is designed to be added to soy-milk or lactose-reduced milk.

This product is made from corn syrup, sugar, vegetable oils and some milk proteins. It also contains some preservatives, emulsifiers and natural and artificial flavors. Medium chain triglycerides, which are easily absorbed fats, have also been added.

Nutritional Information (When mixed with 8 fl oz whole milk; 236 ml)	
Flavors: (availability may vary from country to country)	Vanilla Strawberry Chocolate
Nutrients:	
Energy:	600 Calories
Fat:	30 grams (This product is NOT low-fat.)
Protein:	12 grams
Carbohydrates:	69 grams
Ingredients:	
Maltodextrin, partially hydrogenated vegetable oil (contains one or more of the following: coconut or soybean oils), sugar, nonfat milk, corn syrup solids, sodium caseinate, lecithin, medium chain triglycerides, natural and artificial flavors, dipotassium phosphate, and monoglycerides.	



<http://www.ensure.com/>

Cost: (Prices will vary)	\$33.00 (case of 24 x 8 oz. cans)	
Availability: (Note: Formulations in each country may vary slightly.)	Austria Australia Canada France Germany Italy	Mexico Netherlands (powdered) Puerto Rico UK (powdered)  USA

ENSURE® is complete, balanced nutrition recommended for people older than 10 who have a hard time eating enough regular food to support their nutritional needs. ENSURE® can be used as a snack to supplement a regular balanced diet or as a sole source of nutrition during times when a person is unable to eat regular food. For your child's specific nutritional needs please consult your physician.

- LACTOSE FREE

This product is made from water, sugar (table sugar and corn syrup), milk proteins, and vegetable oils with added vitamins and minerals, some thickening agents, emulsifiers and natural and artificial flavors.

Nutritional Information (Per 8 fl oz; 237 ml)		
Flavors: (availability may vary from country to country)	Vanilla Chocolate Strawberry Butter Pecan Coffee (available through Ensure Home Delivery only)	
Nutrients:	Liquid:	Powdered:
Energy:	250 Calories	250 Calories
Fat:	6 grams	8.8 grams
Protein:	8.8 grams	8.8 grams
Carbohydrates:	40 grams	34 grams
Ingredients:		
Vanilla: -D Water, sugar (sucrose), corn syrup, maltodextrin (corn), calcium caseinate, high-oleic safflower oil, canola oil, soy protein isolate, whey protein concentrate, corn oil, calcium phosphate tribasic, potassium citrate, magnesium phosphate dibasic, natural and artificial flavor, soy lecithin, sodium citrate, magnesium chloride, salt (sodium chloride), carrageenan, choline chloride, potassium chloride, ascorbic acid, ferrous sulfate, alpha-tocopheryl acetate, zinc sulfate, niacinamide, calcium pantothenate, manganese sulfate, cupric sulfate, vitamin A palmitate, thiamine chloride hydrochloride, pyridoxine hydrochloride, riboflavin, folic acid, chromium chloride, biotin, sodium molybdate, sodium selenate, potassium iodide, phylloquinone, vitamin D3 and cyanocobalamin.		





<http://www.meadjohnson.com/products/cons-adult-med/2boost.html>

Cost: (Prices will vary)	\$33.00 (pack of 24 x 8 oz. cans)
Availability:	Canada Puerto Rico USA

BOOST® is a newer product and less widely available than its competition Ensure®, however its formulation is comparable. This product can be given to children 10 years and older as a snack or supplement to a regular balanced diet or as a sole source of nutrition during times when a child has difficulty eating regular food. For your child's specific nutritional needs please consult your physician.

- LACTOSE FREE

This product is made from water, sugar (table sugar and corn syrup), milk proteins, and vegetable oils with added vitamins and minerals, some thickening agents, emulsifiers and natural and artificial flavors.

Nutritional Information (Per 8 fl oz; 236 ml)	
Flavors: (availability may vary from country to country)	Vanilla Chocolate Butter Pecan, Chocolate Mocha Strawberry Chocolate Malt
Nutrients:	
Energy:	240 Calories
Fat:	4 grams
Protein:	10 grams
Carbohydrates:	41 grams
Ingredients:	
(Vanilla flavor) water, corn syrup solids, sugar, milk protein concentrate, canola oil, high oleic sunflower oil, corn oil, and less than 1% of the following: soy lecithin, carrageenan, salt, artificial flavor, vitamins (vitamin A palmitate, beta-carotene, sodium ascorbate, vitamin D3, vitamin E acetate, thiamin hydrochloride, riboflavin, niacinamide, vitamin B6 hydrochloride, folic acid, vitamin B12, biotin, calcium pantothenate, vitamin K1) and minerals (calcium carbonate, ferrous sulfate, potassium iodide, magnesium phosphate, zinc sulfate, cupric sulfate, potassium citrate, potassium chloride, sodium selenite, chromic chloride, sodium molybdate, manganese sulfate).	



<http://www.ensure.com/OurProducts/Enlive.asp>

Cost: (Prices will vary.)	\$37.00 (case of 27 x 8.1 fl. oz. boxes)
Availability: (This list is not complete. Please check if available in your country.)	Australia Germany Puerto Rico USA

ENLIVE! is a high-calorie, fat-free alternative to sweeter-tasting creamy supplements. Although it cannot be used as the sole source of nutrition, it is a great way to increase the nutrient intake of a child who is on a fat restricted diet. ENLIVE! is great as a juice substitute. Please consult your physician to determine if a product like this is right for your child.

- FAT FREE
- LACTOSE FREE

This product is made from water, sugar (table sugar and corn syrup), and milk proteins with added vitamins, minerals, and natural and artificial colors and flavors.

Nutritional Information (Per 8.1 fl oz; 240 ml)	
Flavors: (availability may vary from country to country)	Apple Peach
Nutrients:	
Energy:	300 Calories
Fat:	0 grams (This product is FAT FREE!)
Protein:	10 grams
Carbohydrates:	65 grams
Ingredients:	
<p>Apple: -D Water, maltodextrin (corn), sugar (sucrose), whey protein isolate, hydrochloric acid, citric acid, malic acid, natural and artificial flavor, ascorbic acid, artificial (caramel) color, zinc sulfate, alpha-tocopheryl acetate, ferrous sulfate, niacinamide, manganese sulfate, calcium pantothenate, copper sulfate, vitamin A palmitate, thiamine chloride hydrochloride, pyridoxine hydrochloride, riboflavin, folic acid, chromium chloride, sodium molybdate, biotin, potassium iodide, sodium selenite, phylloquinone, vitamin D3 and cyanocobalamin.</p>	

# TASTY TIPS!

## 1. SERVE COLD and COVERED!

Due to the fact that these supplements contain a lot of added vitamins and minerals, they taste better than they smell. If you are serving the supplement to your child as a beverage, make sure it is cold. Serve it from the can with a straw or put it in a bottle or a cup with a cover.

## 2. BE CREATIVE!

We have included some product literature that contains recipes for alternative ways of using the supplements in foods that your child may prefer.

- Use vanilla flavored products as a substitute for milk in baked products.
- Add fruit, crushed ice and place in the blender to make a "smoothie"

## 3. VANILLA

Unless you know that your child has a preference for a particular flavor of a supplement, buying vanilla is recommended. It is the best flavor to use in recipes and flavored syrups or fruit can be added for flavor variety.

## 4. POWDERED PRODUCTS

When mixing the powdered supplements with liquid to make a beverage, be sure to let it sit in the refrigerator for a while to let the powder completely hydrate.

If you adding a powdered supplement in the dry state to food, do so after the food has been cooked.

## OTHER PRODUCTS THAT MAY BE AVAILABLE TO YOU

Product	Manufacturer	Description
ReSource® Benefiber	Novartis	Soluble fiber supplement. Can be added to any food to increase fiber content. May help reduce constipation.
ReSource® Standard	Novartis	Similar to Ensure and Boost. Complete nutritional supplement.
ReSource® Just for Kids	Novartis	Similar to Pediasure and Kindercal. Complete nutritional supplement for ages 1-10.
ReSource® Fruit Beverage	Novartis	Similar to Enlive!. Fat-free juice drink containing vitamins, mineral and protein.
Nutren	Nestle	Similar to Ensure and Boost. Complete nutritional supplement.
Nutren Jr.	Nestle	Similar to Pediasure and Kindercal. Complete nutritional supplement for ages 1-10.
Nu-Basics	Nestle	Similar to Enlive!. Fat-free juice drink containing vitamins, mineral and protein.