

# **Inherited Metabolic Diseases (IMD) Program**

## **List of Disorders, Covered Drugs, Supplements and Specialty Foods**

**Effective March 4, 2016**

Ontario Public Drug Programs  
Ministry of Health and Long-Term Care



**Inherited Metabolic Diseases (IMD) Program**  
**List of Disorders, Covered Drugs, Supplements and Specialty Foods**

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**Acronyms:**

DIN = Drug Identification Number

PIN = Product Identification Number

NPN = Natural Product Number

SFS SKU = The Hospital for Sick Children's Specialty Food Shop stock number

SAP = Special Access Program

No DIN = Food Supplement or Chemical, no DIN assigned

HSC = Made at The Hospital for Sick Children, no commercial product available

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## Inherited Metabolic Diseases (IMD) Program List of Disorders, Covered Drugs, Supplements and Specialty Foods

### About the IMD Program

Funding and administration of the Inherited Metabolic Diseases (IMD) program was transitioned to Ontario Public Drug Programs (OPDP) in February 2008. One goal of transitioning the IMD program to OPDP is to align it with other drug programs in Ontario. It also creates one point of access in the Ministry of Health and Long-Term Care (Ministry) for all drugs, with clear, consistent and evidence-based methods to make funding decisions.

The IMD subcommittee will evaluate requests for the addition of products and metabolic disorders to the IMD program's List of Disorders, Covered Drugs, Supplements and Specialty Foods (IMD program list). Membership of the subcommittee consists of metabolic and genetic specialists from each of the Ontario Newborn Screening Program regional treatment centres and the University Health Network, a pharmacist and a dietitian. The subcommittee will then make funding recommendations to the Executive Officer (EO), OPDP. For some drug products, the EO may also ask the Committee to Evaluate Drugs to provide a separate review and recommendation. The EO will make the final funding decision based on careful consideration of the subcommittee's recommendations, the public interest and sustainability of the Ontario Public Drug Program.

For new products to be listed under the Drugs and Supplements category on the IMD program list, requests must be submitted by a physician licensed to practice in Ontario. Completed applications will be assigned to a primary reviewer who will then submit a report to the IMD subcommittee. Both will be considered by the IMD subcommittee and their recommendations will then be forwarded to the Ministry and the EO.

For drug products approved for marketing by Health Canada [i.e., those with a drug identification number (DIN) and issued a Notice of Compliance (NOC)], it is the drug manufacturer's responsibility to follow the submission process used for products under consideration for listing on the Ontario Drug Benefit Formulary, and the requirements set out in the *Ontario Guidelines for Drug Submission & Evaluation*. This may include a submission to the Common Drug Review.

For drug products currently listed on the IMD program list, the Ministry has been working with the IMD subcommittee to specify, where appropriate, brand names, manufacturers, dosage forms and strengths, DINs, etc.

### Provisional Listings

The IMD subcommittee has recommended an "approve with provisions" category for products and disorders discussed for consideration of inclusion on the IMD product list. Products and disorders with provisional approvals will require further review to determine their final status. Provisional listings are identified in italics.

Provisional listings for individual patients may not be reflected in the published version of the IMD program list. The requesting physician(s) will be notified by the Ministry of the final funding decision and the details of any listing provisions as recommended by the IMD subcommittee.

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## Notable Changes for this Edition

New products and changes are **highlighted**.

### Disorders

- 1 new disorder has been added: Glucose transporter 1 (GLUT1) deficiency syndrome (GLUT1-DS).

### Drugs and Supplements

- Approved disorder(s) have been added to 4 drugs/ supplements.
- DIN/PIN/etc. has been clarified for **5-hydroxytryptophan**. Going forward, only 5-hydroxytryptophan products with valid Natural Product Number (NPN) or compounded from powder available in Canada will be covered.
- Alkaptonuria is removed as a funded disorder for nitisinone due to the lack of published evidence to support efficacy use in any cohort of asymptomatic or symptomatic alkaptonuria patients. The ministry also noted that there are no patients receiving nitisinone for this indication through the IMD Program. Funding remains in place for tyrosinemia type 1.

### Food products and other supplements

- 2 new medical foods for tyrosinemia have been added: Tylactin RTD 15 Original and Tylactin Restore 10 Citrus
  - The Ministry has approved the addition of these Tylactin products noting the limited number of products available on the IMD list for patients with tyrosinemia, and the neutral cost to the IMD program.
  - The Ministry has also reviewed the Glytactin series of products for phenylketonuria (PKU) and has decided to hold making a decision on coverage of these products at this time. There is significantly more choice or products for PKU patients on the current IMD list compared to tyrosinemia. In addition, the proposed price of some Glytactin products exceeds the current price of available alternatives for PKU.
- Numerous “housekeeping” changes to update product SKUs or to remove discontinued products. Discontinued products (removed from list):
  - Phlexy-10 Capsule PKU Lophlex (46995)
  - PhenylAde PheBloc LNAA Tablets (SFS1489)

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## Disorders Covered by the IMD Program

CATEGORY	DISORDER
<b>Amino Acid Disorders</b>	3-phosphoglycerate dehydrogenase deficiency
	Alkaptonuria
	Gyrate atrophy (ornithine aminotransferase deficiency)
	Homocystinuria (cystathionine beta-synthase [CBS] deficiency)
	Hyperphenylalaninemia, including phenylketonuria (PKU)
	Maple syrup urine disease (MSUD) (branched chain ketoacid dehydrogenase [BCKAD] deficiency)
	Tyrosinemia (includes Types I ,II & III)
<b>Carbohydrate Disorders</b>	Congenital disorder of glycosylation type 1b (CDG-1b)
	Congenital sucrase-isomaltase deficiency (CSID) (intestinal disaccharidase deficiency)
	Galactosemia
	Glycogen storage disorders (GSD)
<b>Cholesterol Biosynthesis Disorders</b>	Smith-Lemli-Opitz syndrome (SLOS)
<b>Fatty Acid Oxidation Defects and Fat Metabolism Disorders</b>	Abetalipoproteinemia
	Apoprotein C-II deficiency
	Carnitine palmitoyl transferase I (CPT I) deficiency
	Carnitine palmitoyl transferase II (CPT II) deficiency
	Carnitine acylcarnitine translocase (CACT) deficiency
	Carnitine uptake defect / carnitine transport defect / primary carnitine deficiency
	Chylomicron retention disease
	Glutaric aciduria Type II (GA II)

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CATEGORY	DISORDER
Fatty Acid Oxidation Defects and Fat Metabolism Disorders (cont'd)	Homozygous hypobetalipoproteinemia
	Lipoprotein lipase (LPL) deficiency
	Long chain 3-hydroxyacyl CoA dehydrogenase (LCHAD)/ Trifunctional protein deficiency
	Medium chain acyl CoA dehydrogenase (MCAD) deficiency
	Medium/short chain hydroxyacyl CoA dehydrogenase (M/SCHAD) deficiency
	Short chain acyl CoA dehydrogenase (SCAD) deficiency
	Very long chain 3-hydroxyacyl CoA dehydrogenase (VLCAD) deficiency
<b>Lactic Acidosis (gluconeogenesis disorders)</b>	Fructose-1,6-bisphosphatase deficiency
	Phosphoenol pyruvate carboxykinase (PEPCK) deficiency
	Pyruvate carboxylase (PC) deficiency
	Pyruvate dehydrogenase (PDH) deficiency
<b>Mitochondrial Disorders</b>	Primary mitochondrial disorder not otherwise specified
	Coenzyme Q10 deficiency
	Complex 1 deficiency
	Friedreich's ataxia
	Leber's hereditary optic neuropathy (LHON)
	Leigh's disease
	Mitochondrial encephalopathy, Lactic acidosis, Stroke-like episodes (MELAS) syndrome
	Mitochondrial myopathy
<b>Organic Acid Disorders</b>	2-methylbutyryl-CoA dehydrogenase deficiency

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CATEGORY	DISORDER
Organic Acid Disorders (cont'd)	2-methyl-3-hydroxybutyryl-CoA dehydrogenase (MHBD) deficiency / short branched chain hydroxyacyl CoA dehydrogenase (SBCHAD) deficiency
	3-hydroxyisobutyric aciduria
	3-hydroxy-3-methyl-glutaric (HMG) CoA lyase deficiency
	3-methylcrotonyl-CoA carboxylase deficiency (3MCC)
	3-methylglutaconic aciduria (includes Barth syndrome)
	β-Ketothiolase deficiency
	Glutaric aciduria Type I (GA I)
	HMG CoA lyase deficiency
	Isobutyryl-CoA dehydrogenase deficiency
	Isovaleric acidemia
	Malonic aciduria
	Methylmalonic acidemia (MMA)
	Propionic acidemia
Pyridoxine (B6)-dependent seizures (alpha-aminoadipic semialdehyde dehydrogenase deficiency) [also listed under Vitamin/Cofactor Disorders]	
<b>Urea Cycle Disorders</b>	Argininemia
	Arginosuccinic acid lyase (AL or ASL) deficiency/ argininosuccinic aciduria (ASAciduria)
	Carbamyl phosphate synthase (CPS) deficiency
	Citrullinemia [arginosuccinic acid synthetase (AS or ASS)] deficiency
	Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)
	N-acetyl glutamate synthetase (NAGS) deficiency
	Ornithine transcarbamylase (OTC) deficiency

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CATEGORY	DISORDER
<b>Vitamin/Cofactor Disorders</b>	Biopterin deficiency (includes dihydropteridine reductase [DHPR] deficiency)
	Cobalamin (B12) defect (includes Cbl C,D,G)
	Cobalamin (B12) transport deficiency, includes transcobalamin II (TC II) deficiency
	Hereditary deficiency of tocopherol transport protein
	Methylene tetrahydrofolate reductase (MTHFR) deficiency
	Multiple carboxylase deficiency (MCD) – Biotinidase deficiency
	Multiple carboxylase deficiency (MCD) – Holocarboxylase synthetase deficiency
	Pyridoxal-5-phosphate dependent epilepsy
	Pyridox(amine) 50-phosphate oxidase (PNPO) deficiency
	Pyridoxine (B6)-dependent seizures (alpha-aminoacidic semialdehyde dehydrogenase deficiency) [also listed under Organic Acid Disorders]
	Vitamin E (tocopherol) deficiency
<b>Miscellaneous Disorders</b>	Acute intermittent porphyria
	Cerebrotendinous xanthomatosis
	Creatine synthesis/transport deficiency
	Cystinosis
	Cystinuria
	Erythropoietic protoporphyria (EPP)
	Glucose transporter 1 (GLUT1) deficiency syndrome (GLUT1-DS)
	Hypercalcemia secondary to a listed disease (please specify)
	Lysinuric protein intolerance (LPI)
	Menkes disease
	Renal tubular acidosis (RTA) secondary to a listed disease (please specify on registration form)



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## Drugs and Supplements

**Note:** The Ministry continues to receive requests to substitute alternate brands of drugs when the listed brands or DINs have been discontinued. In addition, the transition of many over-the-counter products to natural health products under Health Canada's Natural Health Products Regulations has resulted in many DINs being changed to a **Natural Product Number (NPN)**. The Ministry will consider reasonable substitutions if there is little or no difference in cost. Treatment centre pharmacies can contact OPDP with any questions regarding coverage and billing of Drugs and Supplements under the IMD program.

A product in *italics* is considered a provisional listing pending further review by the MOHLTC. Depending on the outcome of this review, the MOHLTC may decide not to fund the product(s) or the listed indication(s) in the future.

DRUGS AND SUPPLEMENTS		
Drug/Supplement	Approved Disorder(s)	DIN/PIN/ETC.
5-hydroxytryptophan	Biopterin deficiency; <b>Pyridoxine-dependent epilepsy (PDE)</b>	<b>Product must have a valid NPN or may be compounded from powder</b>
Ammonul	Urea Cycle Disorders	SAP
Beta-Carotene (Lumitene, oral)	Erythropoietic Protoporphyrin (EPP)	
Betaine (Cystadane)	Homocystinuria (includes homocystinuria secondary to a listed disorder)	SAP
Biotin	Biotinidase deficiency; Holocarboxylase synthetase deficiency; Propionic acidemia; Pyruvate carboxylase deficiency	No DIN
<i>Chenodeoxycholic acid</i>	<i>Cerebrotendinous Xanthomatosis</i>	<i>No DIN</i>
<i>Cholesterol</i>	<i>Smith-Lemli-Opitz syndrome (SLOS)</i>	<i>No DIN</i>
Citrate C	Renal tubular acidosis, secondary to another listed disorder	HSC
Citrulline	Urea cycle disorders; Lysinuric protein intolerance	No DIN
<i>Coenzyme-Q</i>	<i>Mitochondrial disorders; Glutaric Aciduria II, secondary mitochondrial defects</i>	<i>No DIN</i>

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DRUGS AND SUPPLEMENTS		
Drug/Supplement	Approved Disorder(s)	DIN/PIN/ETC.
Copper histidine	Menkes disease	HSC
<i>Creatine</i>	<i>Mitochondrial myopathies; Creatine synthesis/transport deficiency; Gyrate atrophy</i>	SAP
<i>Cupric Chloride</i>	<i>Menkes disease</i>	
Cysteamine (Cystagon or cysteamine powder for compounded products)	Cystinosis	SAP
Dichloroacetate	Pyruvate dehydrogenase deficiency	SAP
Glycine	Isovaleric academia; Creatine transport deficiency	No DIN
Hemin (Normosang)	Acute intermittent porphyria <b>Note:</b> Eligibility criteria for IMD program funding (all criteria must be met): <ol style="list-style-type: none"> <li>1. <i>Diagnosis of acute intermittent porphyria should be confirmed by biochemical and molecular testing, and;</i></li> <li>2. <i>Hemin (Normosang) infusions should only be given under supervision and be prescribed by hematologists, gastroenterologists or metabolic specialists, and;</i></li> <li>3. <i>For continued funding, yearly follow up report should be submitted to the IMD program by the prescribing physician.</i></li> </ol>	SAP
Hydroxycobalamin	Cobalamin defect; Methylmalonic acidemia	SAP
L-arginine	Lysinuric protein intolerance; MELAS syndrome; Pyridoxine (B6)-dependent seizures; Urea cycle disorders; Creatine transport deficiency	No DIN
L-aspartic acid	Pyruvate carboxylase deficiency	No DIN

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DRUGS AND SUPPLEMENTS		
Drug/Supplement	Approved Disorder(s)	DIN/PIN/ETC.
L-carnitine	Organic acidemias; Fatty acid oxidation defects; carnitine uptake defect, carnitine deficiency secondary to another listed disorder	DIN=02144336, DIN=02144328, DIN=02144344
L-isoleucine	MSUD, organic acid disorders	No DIN
L-lysine	Lysinuric protein intolerance	No DIN
L-valine	MSUD, organic acid disorders	No DIN
<i>Lipoic Acid</i>	<i>Mitochondrial disorders</i>	
Mannose	Congenital Disorders of Glycosylation	No DIN
<i>Niacinamide</i>	<i>Mitochondrial cytopathies</i>	<i>No DIN</i>
Nitisinone (NTBC, Orfadin)	Tyrosinemia Type I	SAP
Ornithine HCl, ornithine L-aspartate	Urea cycle disorders; HHH; Creatine synthesis/transport deficiency	No DIN
Phosphocysteamine	Cystinosis	SAP
Pyridoxal-5-phosphate	Pyridoxal-5-phosphate dependent epilepsy; Pyridox(amine) 50-phosphate oxidase (PNPO) deficiency	Product must have a valid NPN
Pyridoxine HCl	Homocystinuria (includes homocystinuria secondary to a listed disorder); Pyridoxine (B6)-dependent seizures	DIN 00497517 NPN 00232475 NPN 00268607
<i>Riboflavin</i>	<i>Mitochondrial disorders; Glutaric aciduria</i>	<i>No DIN</i>
Sapropterin (Kuvan) <sup>1</sup>	Biopterin deficiency	DIN 02350580
Serine	3-phosphoglycerate dehydrogenase deficiency	No DIN
Sodium benzoate	Creatine synthesis/transport deficiency; Urea cycle disorders	No DIN

<sup>1</sup> Sapropterin (Kuvan<sup>®</sup>) is not reimbursed under the IMD program for the treatment of phenylketonuria (PKU). Funding for PKU is considered under the Exceptional Access Program (EAP) only. Under an agreement with the manufacturer, the maximum reimbursable price for Kuvan under the IMD program is \$33.00 per 100mg tablet.

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DRUGS AND SUPPLEMENTS		
Drug/Supplement	Approved Disorder(s)	DIN/PIN/ETC.
<i>Sodium phenylbutyrate</i> <sup>2</sup>	<i>Urea cycle disorders</i>	
<i>Thiamine (Vitamin B1)</i>	<i>Mitochondrial cytopathies; Thiamine deficiency in the presence of IMD</i>	<i>DIN 00816078 NPN 00268631 NPN 00294853</i>
<i>Tiopronin (Thiola)</i>	<i>Cystinuria</i>	<i>SAP</i>
<i>Vitamin A/D/E/K</i>	<i>Abetalipoproteinemia; Mitochondrial disorders</i>	Product must have a valid DIN or NPN
Vitamin A	Abetalipoproteinemia	Product must have a valid DIN or NPN
Vitamin B50 Complex	Mitochondrial disorders (for patients requiring multiple B vitamins and where the cost of vitamin B50 complex does not exceed the cost of the individual vitamins).	Product must have a valid NPN
Vitamin D	Abetalipoproteinemia	Product must have a valid DIN or NPN
<i>Vitamin E</i>	<i>Abetalipoproteinemia; Hereditary deficiency of tocopherol transport protein; Mitochondrial disorders; Vitamin E deficiency</i>	Product must have a valid DIN or NPN
<i>Vitamin K1</i>	<i>Mitochondrial disorders</i>	Product must have a valid DIN or NPN
<i>Vitamin K3</i>	<i>Mitochondrial disorders</i>	Product must have a valid DIN or NPN

<sup>2</sup> The Ministry of Health and Long-Term Care is aware that the Buphenyl formulation of sodium phenylbutyrate may no longer be available in Canada. The IMD program will provide funding for Pheburane<sup>®</sup> on an interim basis pending a further review to determine the final listing status.

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## Medical Foods

MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)			
Product	Disorder(s)	Mfr	PIN/ SFS SKU
BCAD 1	MSUD	Mead Johnson	SFS1071
Camino pro MSUD Drink, Fruit Punch	MSUD	Cambrooke Foods	61002
Camino pro MSUD Drink, Pina Colada	MSUD	Cambrooke Foods	61001
Camino pro PKU Drink, Fruit Punch	PKU, Biopterin deficiency	Cambrooke Foods	31002
Camino pro PKU Drink, Pina Colada	PKU, Biopterin deficiency	Cambrooke Foods	31001
Complex MSUD Amino Acid Bars	MSUD	Nutricia	47021
Complex MSUD Amino Acid Blend	MSUD	Nutricia	47031
Complex MSUD Vanilla Flavoured Drink Mix	MSUD	Nutricia	47024
Complex Essential MSD Drink Mix – Vanilla	MSUD	Nutricia	59720
Complex MSUD Amino Acid Blend	MSUD	Nutricia	47031
Cyclinex–1	Urea cycle disorders, Creatine synthesis/transport deficiency	Abbott Nutrition	47012
Cyclinex–2	Urea cycle disorders, Creatine synthesis/transport deficiency	Abbott Nutrition	47013
Essential Amino Acid Mix	Urea cycle disorders, Gyrate Atrophy, Creatine synthesis/transport deficiency	Nutricia	SFS1556
EAA Supplement	Urea cycle disorders, Gyrate Atrophy, Creatine synthesis/transport deficiency	Vitaflo	47052

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<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
GA1 Express 15	Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures	Vitaflo	SFS1075
GA1 Gel, Unflavoured	Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures	Vitaflo	18888
GlutarAde GA-1 Amino Acid Blend	Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures	Nutricia	SFS1074
Glutarade Essential GA-1 Drink Mix	Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures	Nutricia	SFS1310
Glutarex–1	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Abbott Nutrition	46998
Glutarex–2	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Abbott Nutrition	46999
HCU Cooler 10 red	Homocystinuria	Vitaflo	SFS1076
HCU Cooler 15 orange	Homocystinuria	Vitaflo	18889
HCU Cooler 15 red	Homocystinuria	Vitaflo	SFS0538
HCU Cooler 20 red	Homocystinuria	Vitaflo	SFS1077
HCU express	Homocystinuria	Vitaflo	1809
HCU Express 20	Homocystinuria	Vitaflo	SFS1078
HCU gel	Homocystinuria	Vitaflo	1808
HCU Lophlex LQ Mixed Berry Blast	Homocystinuria	Nutricia	SFS1086
Hominex – 1	Homocystinuria	Abbott Nutrition	46986
Hominex – 2	Homocystinuria	Abbott Nutrition	46979
IVA Cooler 15 red	Isovaleric acidemia	Vitaflo	SFS1315
Ketonex – 1	MSUD	Abbott Nutrition	47007

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<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
Ketonex – 2	MSUD	Abbott Nutrition	47009
MMA/PA Cooler 15 red	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	SFS1314
MMA/PA express	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	47054
MMA/PA gel	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	47053
MSUD Analog	MSUD	Nutricia	47035
MSUD Cooler 10 red	MSUD	Vitaflo	SFS1079
MSUD Cooler 15 orange	MSUD	Vitaflo	1822
MSUD Cooler 15 red	MSUD	Vitaflo	SFS0536
MSUD Cooler 20 red	MSUD	Vitaflo	SFS1080
MSUD Express (unflavoured sachet)	MSUD	Vitaflo	1807
MSUD Express 20	MSUD	Vitaflo	SFS1081
MSUD Gel	MSUD	Vitaflo	1806
MSUD Lophlex LQ Mixed Berry Blast	MSUD	Nutricia	SFS1087
MSUD Maxamaid	MSUD	Nutricia	78964
MSUD Maxamum	MSUD	Nutricia	47022
Periflex Advance Orange	PKU, Biopterin deficiency	Nutricia	47065
Periflex Advance Unflavoured	PKU, Biopterin deficiency	Nutricia	47064
Periflex Infant	PKU, Biopterin deficiency	Nutricia	11400
Periflex Junior Orange	PKU, Biopterin deficiency	Nutricia	47062

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<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
PKU Periflex Junior Plus, Orange	PKU, Biopterin deficiency	Nutricia	SFS1805
Periflex Junior Plus Plain	PKU, Biopterin deficiency	Nutricia	SFS1488
Periflex LQ Berry	PKU, Biopterin deficiency	Nutricia	1256
Periflex LQ Orange	PKU, Biopterin deficiency	Nutricia	1255
Phenex–1	PKU, Biopterin deficiency	Abbott Nutrition	47005
Phenex–2	PKU, Biopterin deficiency	Abbott Nutrition	47003
Phenex–2 Vanilla	PKU, Biopterin deficiency	Abbott Nutrition	47028
PhenylAde 40 Citrus Flavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	1319
Phenylade 40 Unflavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	1320
PhenylAde 60 – Unflavoured Drink Mix (can)	PKU, Biopterin deficiency	Nutricia	SFS0454
PhenylAde 60 – Unflavoured Drink Mix (pouch)	PKU, Biopterin deficiency	Nutricia	SFS0531
PhenylAde 60 – Vanilla Flavoured Drink Mix (can)	PKU, Biopterin deficiency	Nutricia	47088
PhenylAde 60 – Vanilla Flavoured Drink Mix (pouch)	PKU, Biopterin deficiency	Nutricia	SFS0532
PhenylAde Chocolate Flavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	47044



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<b>MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)</b>			
<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
PhenylAde Orange Flavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	47023
PhenylAde Strawberry Flavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	47025
Phenylade Vanilla Flavoured Drink Mix	PKU, Biopterin deficiency	Nutricia	47017
PhenylAde Essential Drink Mix- Chocolate (can)	PKU, Biopterin deficiency	Nutricia	9501
PhenylAde Essential Drink Mix- Orange (can)	PKU, Biopterin deficiency	Nutricia	9503
PhenylAde Essential Drink Mix- Strawberry (can)	PKU, Biopterin deficiency	Nutricia	9504
PhenylAde Essential Drink Mix- Vanilla (can)	PKU, Biopterin deficiency	Nutricia	9502
PhenylAde Essential Drink Mix- Chocolate (pouch)	PKU, Biopterin deficiency	Nutricia	95014
PhenylAde Essential Drink Mix- Orange (pouch)	PKU, Biopterin deficiency	Nutricia	95034
PhenylAde Essential Drink Mix- Strawberry (pouch)	PKU, Biopterin deficiency	Nutricia	95044
PhenylAde Essential Drink Mix- Vanilla (pouch)	PKU, Biopterin deficiency	Nutricia	95024
PhenylAde Amino Acid Bars Chocolate Flavoured	PKU, Biopterin deficiency	Nutricia	470181

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<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
PhenylAde Amino Acid Bars Chocolate Flavoured	PKU, Biopterin deficiency	Nutricia	470182
PhenylAde Amino Acid Bars White Chocolate Flavoured	PKU, Biopterin deficiency	Nutricia	47019
PhenylAde MTE Amino Acid Blend Unflavoured	PKU, Biopterin deficiency	Nutricia	47015
PhenylAde MTE Amino Acid Blend (pouch)	PKU, Biopterin deficiency	Nutricia	1315

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MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)			
Product	Disorder(s)	Mfr	PIN/ SFS SKU
PhenylAde PheBloc LNAA Pouches	<p>PKU</p> <p><b>Note:</b> Eligibility criteria for IMD program funding (all criteria must be met):</p> <ol style="list-style-type: none"> <li>1. <i>The patient will have untreated/late-diagnosed PKU <u>OR</u> early diagnosed PKU with elevated blood phenylalanine concentrations beyond his/her ability to correct with the phenylalanine-restricted diet;<u>AND</u></i></li> <li>2. <i>The patient will have blood phenylalanine concentrations which are chronically above his/her treatment goal;<u>AND</u></i></li> <li>3. <i>The patient will be 13 years of age or over;<u>AND</u></i></li> <li>4. <i>The patient will not be pregnant or planning a pregnancy;<u>AND</u></i></li> <li>5. <i>The patient will not also be treated with sapropterin dihydrochloride (Kuvan);<u>AND</u></i></li> <li>6. <i>It is recommended that the patient use PheBloc (divided evenly between meals) to provide between 0.25-0.5 grams protein equivalent/kg/day (use adjusted body weight for overweight &amp; obese individuals);<u>AND</u></i></li> <li>7. <i>It is recommended that the patient supplement the intake of protein from PheBloc with natural protein foods (and a “medical food” if necessary), to achieve a total protein intake of no less than 1 gram protein/kg/day (use adjusted body weight for overweight &amp; obese individuals); <u>AND</u></i></li> <li>8. <i>The patient will use PheBloc only under the direction of a metabolic geneticist/metabolic dietitian.</i></li> </ol> <p><i>Continued use of PheBloc is approved only for patients who are assessed by the metabolic geneticist to have had an improvement in signs and/or symptoms associated with elevated blood phenylalanine concentrations.</i></p>	Nutricia	SFS1487 (pouches),

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MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)			
Product	Disorder(s)	Mfr	PIN/ SFS SKU
Phenyl-Free 1	PKU, Biopterin deficiency	Mead Johnson	46988
Phenyl-Free 2	PKU, Biopterin deficiency	Mead Johnson	47029
Phenyl-Free 2HP	PKU, Biopterin deficiency	Mead Johnson	47030
Phlexy-10 Drink Mix – Blackcurrant/Apple	PKU, Biopterin deficiency	Nutricia	46996
Phlexy-10 Drink Mix – Tropical Surprise	PKU, Biopterin deficiency	Nutricia	1231
PKU Air 15 green	PKU, Biopterin deficiency	Vitaflo	SFS1311
PKU Air 20 gold	PKU, Biopterin deficiency	Vitaflo	SFS1313
PKU Air 20 green	PKU, Biopterin deficiency	Vitaflo	SFS1312
PKU Cooler 10 orange	PKU, Biopterin deficiency	Vitaflo	47055
PKU Cooler 10 purple	PKU, Biopterin deficiency	Vitaflo	47056
PKU Cooler 10 red	PKU, Biopterin deficiency	Vitaflo	SFS0533
PKU Cooler 15 orange	PKU, Biopterin deficiency	Vitaflo	47049
PKU Cooler 15 purple	PKU, Biopterin deficiency	Vitaflo	47051
PKU Cooler 15 red	PKU, Biopterin deficiency	Vitaflo	SFS0534
PKU Cooler 20 orange	PKU, Biopterin deficiency	Vitaflo	47057
PKU Cooler 20 purple	PKU, Biopterin deficiency	Vitaflo	47058
PKU Cooler 20 red	PKU, Biopterin deficiency	Vitaflo	SFS0535
PKU Cooler 10 – White	PKU, Biopterin deficiency	Vitaflo	98745
PKU Cooler 15 – White	PKU, Biopterin deficiency	Vitaflo	98756
PKU Cooler 20 – White	PKU, Biopterin deficiency	Vitaflo	98766

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<b>MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)</b>			
<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
PKU Express lemon	PKU, Biopterin deficiency	Vitaflo	1805
PKU Express orange	PKU, Biopterin deficiency	Vitaflo	1804
PKU Express tropical	PKU, Biopterin deficiency	Vitaflo	1818
PKU Express unflavoured	PKU, Biopterin deficiency	Vitaflo	1803
PKU Express 20 lemon	PKU, Biopterin deficiency	Vitaflo	SFS1820
PKU Express 20 orange	PKU, Biopterin deficiency	Vitaflo	SFS1821
PKU Express 20 tropical	PKU, Biopterin deficiency	Vitaflo	SFS1822
PKU Express 20 unflavoured	PKU, Biopterin deficiency	Vitaflo	SFS1082
PKU gel orange	PKU, Biopterin deficiency	Vitaflo	1801
PKU gel raspberry	PKU, Biopterin deficiency	Vitaflo	47045
PKU gel unflavoured	PKU, Biopterin deficiency	Vitaflo	1800
PKU Lophlex Berry	PKU, Biopterin deficiency	Nutricia	1248
PKU Lophlex LQ Juicy Orange	PKU, Biopterin deficiency	Nutricia	SFS0835
PKU Lophlex LQ Mixed Berry Blast	PKU, Biopterin deficiency	Nutricia	SFS0834
PKU Lophlex LQ Tropical	PKU, Biopterin deficiency	Nutricia	SFS0452
PKU Lophlex Orange	PKU, Biopterin deficiency	Nutricia	47000
Propimex-1	Methylmalonic acidemia, Propionic acidemia	Abbott Nutrition	47008
Propimex-2	Methylmalonic acidemia, Propionic acidemia	Abbott Nutrition	47010

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MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)			
Product	Disorder(s)	Mfr	PIN/ SFS SKU
Tylactin RTD 15, Original	Tyrosinemia	Cambrooke	SFS1804
Tylactin Restore 10, Citrus	Tyrosinemia	Cambrooke	SFS1803
Tyrex-1	Tyrosinemia	Abbott Nutrition	47006
Tyrex-2	Tyrosinemia	Abbott Nutrition	47016
TYROS 1	Tyrosinemia	Mead Johnson	SFS1072
TYR Cooler 10 red	Tyrosinemia	Vitaflo	SFS1083
TYR Cooler 15 orange	Tyrosinemia	Vitaflo	67896
TYR Cooler 15 red	Tyrosinemia	Vitaflo	SFS0537
TYR Cooler 20 red	Tyrosinemia	Vitaflo	SFS1084
TYR Express	Tyrosinemia	Vitaflo	1811
TYR Express 20	Tyrosinemia	Vitaflo	SFS1085
TYR gel	Tyrosinemia	Vitaflo	46899
TYR Lophlex LQ Mixed Berry Blast	Tyrosinemia	Nutricia	SFS1088
UCD Anamix Junior, unflavoured	Creatine synthesis/transport deficiency, HHH, Gyrate atrophy, Urea cycle disorders	Nutricia	SFS1089
UCD Anamix Junior, vanilla	Creatine synthesis/transport deficiency, HHH, Gyrate atrophy, Urea cycle disorders	Nutricia	SFS1090
I-Valex-1	Isovaleric acidemia	Abbott Nutrition	46993
I-Valex-2	Isovaleric acidemia	Abbott Nutrition	46992
XLeu Analog	Isovaleric acidemia	Nutricia	47040
XLeu Maxamaid	Isovaleric acidemia	Nutricia	78965

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<b>MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)</b>			
<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
XLeu Maxamum	Isovaleric acidemia	Nutricia	78966
XLys, XTrp Analog	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Nutricia	47037
XLys, XTrp Maxamaid	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Nutricia	47050
XLys, XTrp Maxamum	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Nutricia	78967
XMet Analog	Homocystinuria	Nutricia	47036
XMet Maxamaid	Homocystinuria	Nutricia	78968
XMet Maxamum	Homocystinuria	Nutricia	46987
XMTVI Analog	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Nutricia	78969
XMTVI Maxamaid	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Nutricia	78970
XMTVI Maxamum	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Nutricia	78971
XPhe Maxamaid Artificially Orange Flavoured Powder	PKU, Biopterin deficiency	Nutricia	46985
XPhe Maxamaid Unflavoured Powder	PKU, Biopterin deficiency	Nutricia	46977
XPhe Maxamum Artificially Orange Flavoured Powder (can)	PKU, Biopterin deficiency	Nutricia	46984
XPhe Maxamum Artificially Orange Flavoured Powder (sachet)	PKU, Biopterin deficiency	Nutricia	1250
XPhe Maxamum Unflavoured Powder (can)	PKU, Biopterin deficiency	Nutricia	46989

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<b>MODIFIED L-AMINO ACID MIXTURES (MEDICAL FOODS)</b>			
<b>Product</b>	<b>Disorder(s)</b>	<b>Mfr</b>	<b>PIN/ SFS SKU</b>
XPhe Maxamum Unflavoured Powder (sachet)	PKU, Biopterin deficiency	Nutricia	1251
XPhe, XTyr Analog	Tyrosinemia	Nutricia	47038
XPhe, XTyr Maxamaid Artificially Orange Flavoured Powder	Tyrosinemia	Nutricia	46990
XPhen, XTYR Maxamum	Tyrosinemia	Nutricia	1253
XPTM Analog	Tyrosinemia	Nutricia	47039



**Inherited Metabolic Diseases (IMD) Program**  
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## Special Low Protein Foods

<b>SPECIAL LOW PROTEIN FOODS</b>	
<b>Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency</b>	
Product	PIN/SFS SKU
Applied Nutrition Homestyle Blueberry Muffin Mix	38126
Applied Nutrition Cinnamon Chip Flavour Muffin Mix	30400
Applied Nutrition Homestyle Sugar Cookie Mix	38127
Applied Nutrition Homestyle Yellow Cake Mix	38128
Aproten Crispbread Rusks 260g	31128
Aproten Crackertoast 250g	31130
Aproten Farina/Pizza Mix	31172
Aproten Low protein Bread	0255
Aproten Pasta: ditalini 500g	311702
Aproten Pasta: fusilli 500g	311703
Aproten Pasta: spaghetti 500g	311706
Aproten Pasta: penne 500g	311704
Aproten Pasta: anellini 500g	311701
Aproten Pasta: rigatoni 500g	311705
Aproten Pasta: fettucine 250g	311707
Aproten Pasta: tagliatelle 250g	311708
Aproten Biscotto 180g	31171
Aproten Rice (Chicchi) 500g	12163
Cambrooke All-Purpose Baking Mix (2 lbs)	SFS1355
Cambrooke Artisan Buns	401010
Cambrooke Bagels plain	SFS0444
Cambrooke Brooklyn Dog Buns	40616
Cambrooke Focaccia Sticks - Italian (8 sticks)	40112

**Inherited Metabolic Diseases (IMD) Program**  
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<b>SPECIAL LOW PROTEIN FOODS</b>	
<b>Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency</b>	
<b>Product</b>	<b>PIN/SFS SKU</b>
Cambrooke Foods – American Cheese Slices	40311
Cambrooke Foods – Camburger buns	40124
Cambrooke Foods – Camburgers	2901
Cambrooke Foods – Cheese Ravioli	40413
Cambrooke Foods – Pasta Elbows	40455
Cambrooke Foods – Pasta, Portabella Spinach Ravioli	40116
Cambrooke Foods – Swiss Cheese Slices	40312
Cambrooke Foods - Tweekz	40902
Cambrooke Foods –Tomato Tortilla Wraps	2119
Cambrooke Foods –Tortilla Wraps	2118
Cambrooke Homestyle White Bread (1 loaf)	40110
Cambrooke Imitation Mozzarella Shredded Cheese	40315
Cambrooke Macaroni & Cheese	40406
Cambrooke MixQuick Multi-Purpose Batter (2 lbs)	SFS 1354
Cambrooke Pita Pockets	SFS0226
Cambrooke Short Grain Japanese Rice (1kg)	40407
Cambrooke Tuscan Pizza Shells (4 shells)	40115
Cambrooke Veggie Meatballs	40911
Country Sunrise Chicken Flavoured Pattie/Nugget Mix	SFS0233
Country Sunrise Imitation Peanut Butter and Honey Spread	0165
Country Sunrise Instant Mashed Potatoes	0065
Country Sunrise Mushroom Burger Mix	0034
Country Sunrise Scrambled Egg/Omelet Mix	0105
Country Sunrise Soft Tortillas	SFS0695

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<b>SPECIAL LOW PROTEIN FOODS</b>	
<b>Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency</b>	
<b>Product</b>	<b>PIN/SFS SKU</b>
Country Sunrise Southwest Burger Mix	0035
Country Sunrise Vegetable Hot Dog Mix	38346
Dairy-Free milk packets	1517
D.S. (Dietary Specialties) Elbows 250g	2293
D.S. Spanish Rice	1021
D.S. OHZ in Sauce	1022
D.S. Imitation Peanut Butter 16oz	2303
D.S. Imitation Rice 500g	2295
D.S. Imitation Macaroni & Cheese 6oz	2306
D.S. Lasagna 100g	2543
D.S. Tri-coloured Shells 250g	2290
D.S. Porridge 500g	2292
D.S. Herb & Garlic Ziti 250g	2291
D.S. Bread Machine Baking Mix 1.8kg	2294
George Washington Seasoning: brown 31g	141702
George Washington Seasoning: golden 31g	141701
Homestyle Fudge Brownie Mix	SFS0230
Juvela Low Protein Mix (Nutricia)	77444
Kingsmill Egg Replacer 350 g	31197
Kingsmill Vacuum Packed Unimix Bread 550g	31219
Kingsmill Unimix All Purpose Baking Mix	31118
La Tiara Taco Shells	SFS0079
Loprofin Pasta: animal pasta 500g	114155
Loprofin Pasta: fusilli 500g	126113

**Inherited Metabolic Diseases (IMD) Program**  
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<b>SPECIAL LOW PROTEIN FOODS</b>	
<b>Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency</b>	
<b>Product</b>	<b>PIN/SFS SKU</b>
Loprofin Pasta: lasagna (Nutricia)	SFS0515
Loprofin Pasta: penne 500g	126111
Loprofin Pasta: spaghetti 500g	126112
Loprofin Pasta: vermicelli 250g	12610
Loprofin Crackers 150g	12650
Loprofin L/P Baking Mix 500g	38125
Loprofin Breakfast Cereal 375g	12612
Loprofin Chocolate Cake Mix	SFS0211
Loprofin Rice	SFS0116
Milupa Ip Bar	30911
Milupa Ip Drink	12652
Milupa Ip Dry Cereal Chocolate Ringlets	30947
Milupa Ip Dry Cereal Flakes	30940
Milupa Ip Fruity Cereal Mix – Apple Banana	65991
Milupa Ip Fruity Cereal Mix – Pear	65992
PKU Perspectives – Sandwichmate Imitation Cheese Slices	SFS0871
PKU Perspectives – Shredmate Imitation Shredded Mozzarella Cheese	SFS1091
Taste Connections – Low Protein Bread Mix	450
Taste Connections – Low Protein Multi-Baking Mix	451
Taste Connections – Low Protein Versa Mix	4522
Walden Farms Peanut Spread	SFS0080
Wel-Plan Baking Mix 400g	31235

**Inherited Metabolic Diseases (IMD) Program**  
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## Other Supplements

OTHER SUPPLEMENTS		
Product	Disorder(s)	PIN/ SFS SKU
Complete Amino Acid Mix (Nutricia)	Carbohydrate disorders, Lactic acidosis, where a modular approach is required	SFS1557
Duocal	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	46982
Flavour Pacs– Orange	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	1813
Flavour Pacs – Lemon	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	1812
Flavour Pacs – Raspberry	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	1814
Flavour Pacs– Blackcurrant	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	1815
Flavour Pacs- Tropical flavour	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	1819

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<b>OTHER SUPPLEMENTS</b>		
<b>Product</b>	<b>Disorder(s)</b>	<b>PIN/ SFS SKU</b>
<p><b>Glycosade</b> (high-amylopectin-containing cornstarch)</p>	<p>Glycogen storage disease (GSD)</p> <p><b>Note:</b> Eligibility criteria for IMD program funding (all criteria must be met:</p> <ol style="list-style-type: none"> <li>1. <i>The patient will have a diagnosis of glycogen storage disease type 1a/b or type 3 which requires cornstarch feedings overnight for metabolic control; AND</i></li> <li>2. <i>The patient will be ≥24 months of age; AND</i></li> <li>3. <i>The patient will use Glycosade, only under the direction of a metabolic geneticist/metabolic dietitian; AND</i></li> <li>4. <i>The patient will use Glycosade to maintain blood sugars overnight. Regular (i.e. unmodified) cornstarch will continue to be the standard treatment during the day.</i></li> </ol> <p><b>EXCEPTION:</b> <i>If all attempts with conventional therapy (regular cornstarch &amp; diet) during the day fail to achieve metabolic control in a patient, the metabolic geneticist may wish to consider a trial of Glycosade during the day.</i></p> <p><i>Continued use of Glycosade is approved only for patients who demonstrate improved metabolic control.</i></p> <p><b>Glucose transporter 1 deficiency syndrome (GLUT1-DS)* with the following eligibility criteria:</b></p> <ul style="list-style-type: none"> <li>o <i>The patient has confirmed diagnosis of GLUT1-DS; and</i></li> <li>o <i>The patient will use Glycosade to maintain blood sugars overnight along with low glycemic index diet during the day; and</i></li> </ul> <p><b>Patient has demonstrated failure to comply with ketogenic diet.</b></p>	<p>SFS0624</p>
<p>Liquigen</p>	<p>Abetalipoproteinemia, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic Acidosis, Mitochondrial disorders</p>	<p>SFS1490</p>
<p>MCT Pro-Cal</p>	<p>Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders</p>	<p>22990</p>
<p>MCT Oil</p>	<p>Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders</p>	<p>39409</p>

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OTHER SUPPLEMENTS		
Product	Disorder(s)	PIN/ SFS SKU
Microlipid	Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders	74410
PFD 1	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	SFS0539
Phlexy-Vits	Amino acid disorders, Carbohydrate disorders, Fatty Acid Oxidation Defects, Lactic Acidosis, Mitochondrial Disorders, Organic acid disorders, Urea cycle disorders, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Amish Microcephaly	46994
Polycal	Abetalipoproteinemia, Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	SFS1654
Pro-Phree	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	47011
Protifar	Carbohydrate disorders	79645
ProViMin Powder	Abetalipoproteinemia, Carbohydrate disorders	47026
Quick Thick, LP/GF	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate Disorders, Mitochondrial disorders, Fatty acid oxidation defects, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	31301
Resource Beneprotein Instant Protein Powder	Carbohydrate disorders	99557

**Inherited Metabolic Diseases (IMD) Program**  
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<b>OTHER SUPPLEMENTS</b>		
Product	Disorder(s)	PIN/ SFS SKU
SolCarb	Abetalipoproteinemia, Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic acidosis, Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	SFS1316
Vitapro	Carbohydrate disorders	SFS0550



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## Infant Feeds

Product	Disorder(s)	PIN/ SFS SKU
Calcilo XD	Hypercalcemia secondary to a listed disease	91595
Monogen	Fatty Acid Oxidation Defects	47060
Lipistart	Fatty Acid Oxidation Defects	47068
Nutramigen A+	Amino Acid Disorders, Biopterin deficiency, Creatine synthesis/transport deficiency, Glutaric Aciduria II (GA II), Organic Acid Disorders, Pyridoxine (B6) dependent seizures, Urea Cycle Disorders	SFS000046
Portagen Powder	Fatty Acid Oxidation Defects, Mitochondrial disorders	39581
Pregestimil A+ Powder	Amino acid disorders, Carbohydrate Disorders, Organic acid disorders, Urea cycle disorders	000045
Ross Carbohydrate Free	Lactic acidosis	1585

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## Complete Enteral Feeds

### Eligibility criteria for enteral feeds:

Nutrition products will be reimbursed under the IMD program for registered persons when prescribed by a practitioner and when one of the following criteria is met:

- the product is used as part of a treatment plan in addition to medical foods (i.e., modified L-amino acid mixtures) or special low protein foods; OR
- the product is used as part of a treatment plan without medical foods (i.e., modified L-amino acid mixtures) or special low protein foods, and is the primary source of nutrition (e.g., greater than 90% of caloric intake); OR
- the product is used as part of a treatment plan for prevention of hypoglycemia in glycogen storage disorders (continuous and/or bolus feeds)

### Exclusion criteria for enteral feeds:

A nutrition product will not be reimbursed under the IMD program if the patient qualifies for reimbursement under the Ontario Drug Benefit (ODB) program or if it is intended for one of the following uses:

- voluntary meal replacement
- convenience
- food allergies
- body building

**If the following products are the sole source of nutrition, coverage may be available for eligible persons under the Ontario Drug Benefit Program**

COMPLETE ENTERAL FEEDS		
Product	Manufacturer	PIN/SFS SKU
Boost 1.0 Standard Chocolate	Nestle Nutrition	995151
Boost 1.0 Standard Strawberry	Nestle Nutrition	995153
Boost 1.0 Standard Vanilla	Nestle Nutrition	995152
Boost 1.5 Plus Calories Chocolate	Nestle Nutrition	995201
Boost 1.5 Plus Calories Strawberry	Nestle Nutrition	995202
Boost 1.5 Plus Calories Vanilla	Nestle Nutrition	995203
Ensure Chocolate	Abbott Nutrition	914251
Ensure Strawberry	Abbott Nutrition	914252
Ensure Vanilla	Abbott Nutrition	914253
Ensure Plus Chocolate	Abbott Nutrition	914351

**Inherited Metabolic Diseases (IMD) Program**  
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<b>COMPLETE ENTERAL FEEDS</b>		
Product	Manufacturer	PIN/SFS SKU
Ensure Plus Strawberry	Abbott Nutrition	914352
Ensure Plus Vanilla	Abbott Nutrition	914353
Isosource HN	Nestle Nutrition	99458
Isosource HN with Fibre	Nestle Nutrition	99459
Jevity 1	Abbott Nutrition	1499
Nutren Junior	Nestle Nutrition	111912
Nutren Junior Fibre with Prebio	Nestle Nutrition	111911
Osmolite 1	Abbott Nutrition	1497
Pediasure Vanilla	Abbott Nutrition	91423
Pediasure Chocolate	Abbott Nutrition	914231
Pediasure Strawberry	Abbott Nutrition	914232
Pediasure Vanilla with Fibre	Abbott Nutrition	91424
Pediasure Plus Vanilla with Fibre	Abbott Nutrition	91422
Peptamen Junior Vanilla	Nestle Nutrition	11120
Peptamen Unflavoured	Nestle Nutrition	11100
Resource Kids Essential 1.5	Nestle Nutrition	99538
Suplena	Abbott Nutrition	1441
Tolerex	Nestle Nutrition	50524
Vital HN	Abbott Nutrition	1521
Vivonex Pediatric	Nestle Nutrition	SFS0212
Vivonex Plus	Nestle Nutrition	50545

