

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

Effective January 25, 2010

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

Table of Contents

| | |
|---|----|
| Preamble..... | 3 |
| Notable Changes..... | 4 |
| Covered Disorders..... | 5 |
| Drugs and Supplements | 7 |
| Modified L-Amino Acid Mixtures (Medical Foods)..... | 9 |
| Special Low Protein Foods..... | 16 |
| Other Supplements..... | 18 |
| Infant Feeds..... | 20 |
| Complete Enteral Feeds | 20 |

Acronyms:

- DIN = Drug Identification Number
- PIN = Product Identification Number
- 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
- No PIN = Product not currently carried by The Hospital for Sick Children's Specialty Food Shop due to low volume

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Inherited Metabolic Diseases (IMD) Program

List of Disorders, Covered Drugs, Supplements and Specialty Foods

Preamble

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 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, and a pharmacist and a dietitian. The subcommittee will then make recommendations to the Executive Officer (EO) for OPDP. For some drug products, the EO may also ask the Committee to Evaluate Drugs to provide a separate review and recommendation. The EO makes the final decision on funding, taking into consideration the recommendations and public interest.

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)] to be considered under the IMD program, it is now the drug manufacturer's responsibility to seek listing on the IMD program list by filing a submission to the Ministry¹. Approval by Health Canada does not mean that it will be listed as a benefit under the IMD program. Health Canada is responsible for approving a drug for sale and marketing anywhere in Canada, but does not make any recommendations regarding funding.

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. This modernization initiative began with the May 1, 2008 version of the IMD program list.

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.

Some provisional listings may not be reflected in the published version of the IMD program list, usually when approved for individual patients. The requesting physician(s) will be notified by the

¹ Manufacturer submissions should follow the format 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](#).

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

Ministry of the final funding decision and of the details of any listing provisions as recommended by the IMD subcommittee.

Notable Changes for January 2010

1. Disorders Covered: The IMD subcommittee has recommended that a specific diagnosis be provided for all patients being registered in the IMD program. Therefore, the “*not otherwise specified*” option has been removed from most IMD categories.

Phosphoenol Pyruvate Carboxykinase (PEPCK) deficiency has been added to the Lactic Acidosis category.

The “*hypercalcemia*” listing in the Miscellaneous category has been revised to hypercalcemia ***secondary to a listed disease***. The IMD subcommittee noted that the intent of listing hypercalcemia is to make Calcilo XD available to patients with hypercalcemia associated with a listed metabolic disease. Treatment centres with patients currently registered with hypercalcemia not secondary to a listed disease should contact the ministry.

The Mitochondrial Disorders category is currently under review. All mitochondrial disorders remain provisional listings.

2. Substitutions for listed IMD drug brands/formulations: The Ministry continues to receive requests from treatment centre pharmacies to substitute alternate brands of drugs when the listed brands or DINs have been discontinued, such as thiamine and pyridoxine (vitamin B6). In addition, the transition of many over-the-counter products to natural health products under Health Canada’s Natural Health Products Regulations (amendments to the Food and Drug Act) 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. During the ongoing transition phase from DINs to NPNs, treatment centre pharmacies may wish to contact OPDP with any questions regarding coverage and billing of Drugs and Supplements under the IMD program.

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

DISORDERS COVERED BY THE IMD PROGRAM

| CATEGORY | DISORDER |
|---|--|
| Amino Acid Disorders | 3-phosphoglycerate dehydrogenase deficiency |
| | Alkaptonuria |
| | Gyrate Atrophy |
| | Homocystinuria |
| | Hyperphenylalaninemia, including phenylketonuria (PKU) |
| | Maple Syrup Urine Disease (MSUD) |
| | Tyrosinemia (includes Types I & II) |
| Carbohydrate Disorders | Congenital Disorder of Glycosylation type 1b (CDG-1b) |
| | Congenital Sucrase-Isomaltase deficiency (CSID) (intestinal disaccharidase deficiency) |
| | Galactosemia |
| | Glycogen Storage disorders (GSD) |
| Cholesterol Biosynthesis Disorders | Smith-Lemli-Opitz syndrome (SLOS) |
| Fatty Acid Oxidation Defects | Glutaric Aciduria II (GA II) |
| | Long Chain 3-Hydroxyacyl CoA Dehydrogenase (LCHAD)/ Trifunctional Protein Deficiency |
| | Medium Chain Acyl CoA Dehydrogenase (MCAD) deficiency |
| | Short Chain Acyl CoA Dehydrogenase (SCAD) deficiency |
| | Very Long Chain 3-Hydroxyacyl CoA Dehydrogenase (VLCAD) deficiency |
| Lactic Acidosis | Phosphoenol Pyruvate Carboxykinase (PEPCK) deficiency |
| | Pyruvate Carboxylase (PC) deficiency |
| | Pyruvate Dehydrogenase (PDH) deficiency |
| Mitochondrial Disorders | 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 |
| Organic Acid Disorders | 3-hydroxy-3-methyl-glutaric (HMG) CoA lyase deficiency |
| | 3-Methylcrotonyl-CoA carboxylase deficiency (3MCC) |
| | 3-methylglutaconic aciduria (includes Barth syndrome) |

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

| CATEGORY | DISORDER |
|------------------------------------|--|
| Organic Acid Disorders (cont'd) | β-Ketothiolase deficiency |
| | Glutaric Aciduria I (GA I) |
| | HMG CoA lyase deficiency |
| | Isovaleric acidemia |
| | Methylmalonic Acidemia (MMA) |
| | Propionic acidemia |
| Urea Cycle Disorders | Argininemia |
| | Arginosuccinic acid Lyase (AL or ASL) deficiency/ argininosuccinic aciduria (ASAuria) |
| | Carbamyl Phosphate Synthase (CPS) deficiency |
| | Citrullinemia [Arginosuccinic acid Synthetase (AS or ASS)] deficiency |
| | N-acetyl glutamate synthetase (NAGS) deficiency |
| | Ornithine Transcarbamylase (OTC) deficiency |
| Vitamin/Cofactor Disorders | Biopterin (BH4) 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 |
| | Pyridoxine (B6) dependent seizures |
| | Vitamin E (tocopherol) deficiency |
| Miscellaneous | Abetalipoproteinemia |
| | Carnitine uptake defect |
| | Cerebrotendinous xanthomatosis |
| | Creatine synthesis/transport deficiency |
| | Cystinosis |
| | Cystinuria |
| | Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) |
| | 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) |
| | Porphyrias - Erythropoietic Protoporphyrin (EPP) |

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

DRUGS AND SUPPLEMENTS COVERED BY THE IMD PROGRAM

| DRUGS AND SUPPLEMENTS | | |
|--|--|--|
| Drug/Supplement | Indications | DIN/PIN/etc. |
| 5-hydroxytryptophan | Biopterin (BH4) deficiency | SAP |
| Ammonul | Urea cycle disorders | SAP |
| Beta-Carotene (Lumitene, oral) | Erythropoietic Protoporphyrin (EPP) | |
| Betaine (Cystadane) | Homocystinuria | SAP |
| Biotin | Biotinidase deficiency; Holocarboxylase synthetase deficiency; Propionic acidemia; Pyruvate carboxylase deficiency | No DIN |
| Carbaglu | N-acetylglutamate synthase (NAGS) deficiency | |
| <i>Chenodeoxycholic acid</i> ³ | <i>Cerebrotendinous Xanthomatosis</i> | <i>No DIN</i> |
| <i>Cholesterol</i> ³ | <i>Cholesterol biosynthesis disorders – specific disease 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 |
| Coenzyme-Q ³ | <i>Mitochondrial disorders; Glutaric Aciduria II, secondary mitochondrial defects</i> | <i>No DIN</i> |
| Copper histidine | Menkes | HSC |
| <i>Creatine</i> ³ | <i>Mitochondrial myopathies; Creatine synthesis/transport deficiency; Gyrate atrophy</i> | SAP |
| <i>Cupric Chloride</i> ³ | <i>Menkes</i> | |
| Cystagon (cysteamine) | Cystinosis | SAP |
| Dichloroacetate | Pyruvate dehydrogenase deficiency | SAP |
| Essamino | Gyrate atrophy, Urea cycle disorders | SAP |
| Glycine | Isovaleric acidemia | No DIN |
| Hydroxycobalamin | Cobalamin defect; Methylmalonic acidemia | SAP |
| <i>Idebenone (Kirkman 45mg, 150mg only)</i> ^{2,3} | <i>Friedreich's Ataxia</i> | <i>No DIN</i> |
| L-arginine | MELAS syndrome; Urea cycle disorders; Lysinuric protein intolerance | No DIN |
| L-aspartic acid | Pyruvate carboxylase deficiency | No DIN |
| L-carnitine | Organic acidemias; Fatty acid oxidation defects; carnitine uptake defect, proven carnitine deficiency secondary to another listed disorder | DIN=02144336, DIN=02144328, DIN=02144344 |
| L-dopa | Biopterin deficiency | No DIN |
| <i>Lipoic Acid</i> ³ | <i>Mitochondrial disorders</i> | |
| L-isoleucine | MSUD, organic acid disorders | No DIN |
| L-lysine | Lysinuric protein intolerance | No DIN |

² The Ministry is aware of the recent availability of idebenone (Catena[®]) from Santhera Pharmaceuticals. This brand/formulation of idebenone is not currently reimbursed under the IMD program.

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

| DRUGS AND SUPPLEMENTS | | |
|---|---|---|
| Drug/Supplement | Indications | DIN/PIN/etc. |
| L-valine | MSUD, organic acid disorders | No DIN |
| Mannose | Congenital Disorders of Glycosylation | No DIN |
| Methionine | MTHFR Deficiency | |
| <i>Niacinamide</i> ³ | <i>Mitochondrial cytopathies</i> | <i>No DIN</i> |
| Nitisinone (NTBC, Orfadin) | Tyrosinemia; Alkaptonuria | SAP |
| Ornithine | Urea cycle disorders; HHH | No DIN |
| Phosphocysteamine | Cystinosis | SAP |
| Pyridoxine HCl | Homocystinuria; B6-dependent seizures; | DIN=00497517, DIN=00232475, DIN=00268607 |
| <i>Riboflavin</i> ³ | <i>Mitochondrial disorders</i> ; Glutaric aciduria | <i>No DIN</i> |
| Serine | 3-phosphoglycerate dehydrogenase deficiency | No DIN |
| Sucraid | Congenital Sucrase-Isomaltase deficiency | |
| Sodium benzoate | Urea cycle disorders | No DIN |
| Sodium phenylbutyrate | Urea cycle disorders | SAP |
| Tetrahydrobiopterin (BH4) (Schircks 10mg, 50mg; Kuvan will be funded if the Schircks formulation is unavailable) | Biopterin deficiency | SAP |
| <i>Thiamine (Vitamin B1)</i> ³ | <i>Mitochondrial cytopathies</i> ; Thiamine deficiency in the presence of IMD | <i>DIN=00816078, DIN=00268631 DIN= 00294853</i> |
| Tiopronin (Thiola) | Cystinuria | SAP |
| Ucephan (see Ammonul) | Urea cycle disorders | SAP |
| <i>Vitamin A, D, E, K (fat soluble multivitamin)</i> ³ | <i>Mitochondrial disorders</i> ; Abetalipoproteinemia; | |
| Vitamin A | Abetalipoproteinemia | |
| Vitamin D | Abetalipoproteinemia | |
| <i>Vitamin E</i> | Abetalipoproteinemia; Hereditary deficiency of tocopherol transport protein; <i>Mitochondrial disorders</i> ; Vitamin E deficiency | |
| <i>Vitamin K1</i> ³ | <i>Mitochondrial disorders</i> | |
| <i>Vitamin K3</i> ³ | <i>Mitochondrial disorders</i> | |

³ *Italics*: A product in *italics* is considered a provisional listing pending further review by the IMD subcommittee and 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.

**MOHLTC Inherited Metabolic Diseases (IMD) Program
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MEDICAL FOODS COVERED BY THE IMD PROGRAM

| MODIFIED L-AMINO ACID MIXTURES ("MEDICAL FOODS") | | | |
|---|---|-------------------------|----------------|
| Product | Indications | Company | PIN/SKU |
| Acerflex | MSUD | Nutricia (SHS) | 47020 |
| Camino pro MSUD Drink, Fruit Punch | MSUD | Cambrooke Foods | 61002 |
| Camino pro MSUD Drink, Pina Colada | MSUD | Cambrooke Foods | 61001 |
| Camino pro MSUD Sorbet Stix, Lemon Ice | MSUD | Cambrooke Foods | 62001 |
| Camino pro MSUD Sorbet Stix, Very Berry | MSUD | Cambrooke Foods | 62002 |
| Camino pro PKU Drink, Fruit Punch | PKU, Biopterin (BH4) Deficiency | Cambrooke Foods | 31002 |
| Camino pro PKU Drink, Pina Colada | PKU, Biopterin (BH4) Deficiency | Cambrooke Foods | 31001 |
| Camino pro PKU Sorbet Stix, Lemon Ice | PKU, Biopterin (BH4) Deficiency | Cambrooke Foods | 32001 |
| Camino pro PKU Sorbet Stix, Drink, Very Berry | PKU, Biopterin (BH4) Deficiency | Cambrooke Foods | 32002 |
| Complex MSUD Amino Acid Bars | MSUD | Applied Nutrition | 47021 |
| Complex MSUD Amino Acid Blend | MSUD | Applied Nutrition | 47031 |
| Complex MSUD Vanilla Flavoured Drink Mix | MSUD | Applied Nutrition | 47024 |
| Complex Essential MSD Drink Mix – Vanilla | MSUD | Applied Nutrition | 59720 |
| Complex MSUD Amino Acid Blend | MSUD | Applied Nutrition | 47031 |
| Cyclinex–1 | Urea cycle disorders, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | Abbott Nutrition (Ross) | 47012 |
| Cyclinex–2 | Urea cycle disorders, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | Abbott Nutrition (Ross) | 47013 |
| Essential AA Mix | Urea cycle disorders, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH), Gyrate Atrophy | Nutricia (SHS) | 47002 |
| EAA Supplement | Urea Cycle Disorders, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH), Gyrate Atrophy | Vitaflo | 47052 |
| GA1 Gel, Unflavoured | Glutaric aciduria Type I | Vitaflo | 18888 |
| Glutarex–1 | Glutaric aciduria Type I, Type II | Abbott Nutrition (Ross) | 46998 |
| Glutarex–2 | Glutaric aciduria Type I, Type II | Abbott Nutrition (Ross) | 46999 |
| HCU Cooler | Homocystinuria | Vitaflo | 18889 |
| HCU express | Homocystinuria | Vitaflo | 1809 |

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| MODIFIED L-AMINO ACID MIXTURES ("MEDICAL FOODS") | | | |
|---|---|-------------------------|----------------|
| Product | Indications | Company | PIN/SKU |
| HCU gel | Homocystinuria | Vitaflo | 1808 |
| Hominex – 1 | Homocystinuria | Abbott Nutrition (Ross) | 46986 |
| Hominex – 2 | Homocystinuria | Abbott Nutrition (Ross) | 46979 |
| Ketonex – 1 | MSUD | Abbott Nutrition (Ross) | 47007 |
| Ketonex – 2 | MSUD | Abbott Nutrition (Ross) | 47009 |
| Lophlex Berry | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1248 |
| Lophlex Orange | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47000 |
| MMA/PA express | Methylmalonic Acidemia (MMA), Propionic Acidemia (PA) | Vitaflo | 47054 |
| MMA/PA gel | Methylmalonic Acidemia (MMA), Propionic Acidemia (PA) | Vitaflo | 47053 |
| MSUD Analog | MSUD | Nutricia (SHS) | 47035 |
| MSUD express cooler | MSUD | Vitaflo | 1822 |
| MSUD express (unflavoured sachets) | MSUD | Vitaflo | 1807 |
| MSUD Gel | MSUD | Vitaflo | 1806 |
| MSUD Maxamaid | MSUD | Nutricia (SHS) | 78964 |
| MSUD Maxamum | MSUD | Nutricia (SHS) | 47022 |
| Periflex Infant | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 11400 |
| Periflex Junior Unflavored | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47061 |
| Periflex Junior Orange | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47062 |
| Periflex Junior Chocolate | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47063 |
| Periflex Advance Unflavored | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47064 |
| Periflex Advance Orange | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47065 |
| Periflex Advance Chocolate | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47066 |
| Phenex–1 | PKU, Biopterin (BH4) Deficiency | Abbott Nutrition (Ross) | 47005 |
| Phenex–2 | PKU, Biopterin (BH4) Deficiency | Abbott Nutrition (Ross) | 47003 |
| Phenex–2 Vanilla | PKU, Biopterin (BH4) Deficiency | Abbott Nutrition (Ross) | 47028 |
| PhenylAde 40 Citrus Flavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 1319 |
| Phenylade 40 Unflavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 1320 |
| PhenylAde 60 – Vanilla Flavoured Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47088 |
| PhenylAde Chocolate Flavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47044 |
| PhenylAde Orange Flavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47023 |
| PhenylAde Strawberry Flavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47025 |
| Phenylade Vanilla Flavored Drink Mix | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47017 |
| PhenylAde Essential Drink Mix- Chocolate (can) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 9501 |
| PhenylAde Essential Drink Mix- Orange (can) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 9503 |

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

| MODIFIED L-AMINO ACID MIXTURES ("MEDICAL FOODS") | | | |
|---|---------------------------------|-------------------|----------------|
| Product | Indications | Company | PIN/SKU |
| PhenylAde Essential Drink Mix- Strawberry (can) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 9504 |
| PhenylAde Essential Drink Mix- Vanilla (cans) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 9502 |
| PhenylAde Essential Drink Mix- Chocolate (pouch) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 95014 |
| PhenylAde Essential Drink Mix- Orange (pouch) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 95034 |
| PhenylAde Essential Drink Mix- Strawberry (pouch) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 95044 |
| PhenylAde Essential Drink Mix- Vanilla (pouch) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 95024 |
| PhenylAde Amino Acid Bars Chocolate Flavored | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 470181 |
| PhenylAde Amino Acid Bars Chocolate Flavored | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 470182 |
| PhenylAde Amino Acid Bars White Chocolate Flavored | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47019 |
| PhenylAde MTE Amino Acid Blend Unflavored | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 47015 |
| PhenylAde MTE Amino Acid Blend (in pouches) | PKU, Biopterin (BH4) Deficiency | Applied Nutrition | 1315 |
| Phenyl-Free 1 | PKU, Biopterin (BH4) Deficiency | Mead Johnson | 46988 |
| Phenyl-Free 2 | PKU, Biopterin (BH4) Deficiency | Mead Johnson | 47029 |
| Phenyl-Free 2HP | PKU, Biopterin (BH4) Deficiency | Mead Johnson | 47030 |
| Phlexy-10 add-ins | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47067 |
| Phlexy-10 Capsules | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46995 |
| Phlexy-10 Drink Mix – Blackcurrant/Apple | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46996 |
| Phlexy-10 Drink Mix – Tropical Surprise | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46997 |
| PKU cooler 10 orange | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47055 |
| PKU cooler 10 purple | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47056 |
| PKU cooler 15 orange | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47049 |
| PKU cooler 15 purple | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47051 |
| PKU cooler 20 orange | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47057 |
| PKU cooler 20 purple | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47058 |
| PKU Cooler 10 – White | PKU, Biopterin (BH4) Deficiency | Vitaflo | 98745 |
| PKU Cooler 15 – White | PKU, Biopterin (BH4) Deficiency | Vitaflo | 98756 |
| PKU Cooler 20 – White | PKU, Biopterin (BH4) Deficiency | Vitaflo | 98766 |
| PKU express orange | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1804 |
| PKU express lemon | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1805 |
| PKU express tropical | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1818 |
| PKU express unflavored | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1803 |
| PKU gel orange | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1801 |
| PKU gel raspberry | PKU, Biopterin (BH4) Deficiency | Vitaflo | 47045 |

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

| MODIFIED L-AMINO ACID MIXTURES ("MEDICAL FOODS") | | | |
|--|---|-------------------------|----------------|
| Product | Indications | Company | PIN/SKU |
| PKU gel unflavored | PKU, Biopterin (BH4) Deficiency | Vitaflo | 1800 |
| Propimex-1 | Methylmalonic acidemia, Propionic acidemia | Abbott Nutrition (Ross) | 47008 |
| Propimex-2 | Methylmalonic acidemia, Propionic acidemia | Abbott Nutrition (Ross) | 47010 |
| Tyrex-1 | Tyrosinemia Type I | Abbott Nutrition (Ross) | 47006 |
| Tyrex-2 | Tyrosinemia Type I | Abbott Nutrition (Ross) | 47016 |
| TYR Cooler | Tyrosinemia Type I | Vitaflo | 67896 |
| TYR express | Tyrosinemia | Vitaflo | 1811 |
| TYR gel | Tyrosinemia | Vitaflo | 46899 |
| I-Valex-1 | Isovaleric acidemia | Abbott Nutrition (Ross) | 46993 |
| I-Valex-2 | Isovaleric acidemia | Abbott Nutrition (Ross) | 46992 |
| XLeu Analog | Isovaleric acidemia | Nutricia (SHS) | 47040 |
| XLeu Maxamaid | Isovaleric acidemia | Nutricia (SHS) | 78965 |
| XLeu Maxamum | Isovaleric acidemia | Nutricia (SHS) | 78966 |
| XLys, XTrp Analog | Glutaric aciduria Type I, Type II | Nutricia (SHS) | 47037 |
| XLys, XTrp Maxamaid | Glutaric aciduria Type I, Type II | Nutricia (SHS) | 47050 |
| XLys, XTrp Maxamum | Glutaric aciduria Type I, Type II | Nutricia (SHS) | 78967 |
| XMet Analog | Homocystinuria | Nutricia (SHS) | 47036 |
| XMet Maxamaid | Homocystinuria | Nutricia (SHS) | 78968 |
| XMet Maxamum | Homocystinuria | Nutricia (SHS) | 46987 |
| XMTVI Analog | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia (SHS) | 78969 |
| XMTVI Maxamaid | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia (SHS) | 78970 |
| XMTVI Maxamum | Methylmalonic acidemia (MMA), Propionic acidemia (PA) | Nutricia (SHS) | 78971 |
| XPhe Analog | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 47034 |
| XPhe Maxamaid Artificially Orange Flavored Powder | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46985 |
| XPhe Maxamaid Unflavored Powder | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46977 |
| XPhe Maxamaid Artificially Strawberry Flavored Powder | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1252 |
| XPhe Maxamum Artificially Orange Flavored Powder | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46984 |
| XPhe Maxamum Unflavored Powder | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 46989 |
| XPhe Maxamum Unflavored Powder Sachets | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1251 |
| XPhe Maxamum Artificially Orange Flavored Powder Sachets | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1250 |
| XPhe Maxamum drink Orange | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1255 |

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

| MODIFIED L-AMINO ACID MIXTURES ("MEDICAL FOODS") | | | |
|---|---------------------------------|----------------|----------------|
| Product | Indications | Company | PIN/SKU |
| XPhe Maxamum drink Forest Berries | PKU, Biopterin (BH4) Deficiency | Nutricia (SHS) | 1256 |
| XPhe, XTyr Analog | Tyrosinemia Type I and II | Nutricia (SHS) | 47038 |
| XPhe, XTyr Maxamaid Artificially Orange Flavored Powder | Tyrosinemia Type I and II | Nutricia (SHS) | 46990 |
| XPhen, XTYR Maxamum | Tyrosinemia Type II | Nutricia (SHS) | 1253 |
| XPTM Analog | Tyrosinemia Type I | Nutricia (SHS) | 47039 |

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

SPECIAL LOW PROTEIN FOODS COVERED BY THE IMD PROGRAM

| Special Low Protein Foods | |
|--|----------------|
| <i>Indications: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type 2 (GA2), Bioppterin (BH4) Deficiency and Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)</i> | |
| Product | PIN/SKU |
| Applied Nutrition Crackels | 1326 |
| Applied Nutrition Homestyle Blueberry Muffin Mix | 38126 |
| Applied Nutrition Homestyle Sugar Cookie Mix | 38127 |
| Applied Nutrition Homestyle Yellow Cake Mix | 38128 |
| Aproten Crispbread Rusks 260g | 31128 |
| Aproten Crackertoast 250g | 31130 |
| 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 (6lbs) | 402010 |
| Cambrooke Artisan Buns | 401010 |
| Cambrooke Focaccia Sticks - Italian (8 sticks) | 40112 |
| 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 - Tweeks | 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 (6lbs) | 402020 |
| Cambrooke Short Grain Japanese Rice (1kg) | 40407 |
| Cambrooke Tuscan Pizza Shells (4 shells) | 40115 |
| 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 Southwest Burger Mix | 0035 |

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

| Special Low Protein Foods | |
|--|----------------|
| <i>Indications: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type 2 (GA2), Bioppterin (BH4) Deficiency and Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)</i> | |
| Product | PIN/SKU |
| 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 |
| Juvela Low Protein Mix (Nutricia) | 77444 |
| Kingsmill Wheat Starch 2 kg (5 lbs) | 31193 |
| Kingsmill Egg Replacer 350 g | 31197 |
| Kingsmill Vacuum Packed Unimix Bread 550g | 31219 |
| Kingsmill Gel Dessert: Cherry 320g | 311981 |
| Kingsmill Gel Dessert: Orange 320g | 311982 |
| Kingsmill Unimix All Purpose Baking Mix | 31118 |
| Loprofin Pasta: vermicelli 250g | 12610 |
| Loprofin Pasta: fusilli 500g | 126113 |
| Loprofin Pasta: spaghetti 500g | 126112 |
| Loprofin Pasta: penne 500g | 126111 |
| Loprofin Pasta: animal pasta 500g | 114155 |
| Loprofin Crackers 150g | 12650 |
| Loprofin L/P Baking Mix 500g | 38125 |
| Loprofin Breakfast Cereal 375g | 12612 |
| Milupa Ip Bar | 30911 |
| Milupa Ip Drink | 12653 |
| 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 |
| Taste Connections – Low Protein Bread Mix | 450 |
| Taste Connections – Low Protein Multi-Baking Mix | 451 |
| Taste Connections – Low Protein Versa Mix | 4522 |
| WelPlan Baking Mix 400g | 31235 |

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

OTHER SUPPLEMENTS COVERED BY THE IMD PROGRAM

| OTHER SUPPLEMENTS | | |
|------------------------------------|---|----------------|
| Product | Indications | PIN/SKU |
| Complete Amino Acid Mix (Nutricia) | Carbohydrate disorders, Lactic acidosis, where a modular approach is required | 47001 |
| Duocal | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 46982 |
| Flavour Pacs– Orange | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 1813 |
| Flavour Pacs –Lemon | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 1812 |
| Flavour Pacs –Raspberry | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 1814 |
| Flavour Pacs–Blackcurrant | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 1815 |
| Flavour Pacs- Tropical flavour | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH) | 1819 |
| MCT Pro-Cal | Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders | 22990 |
| MCT Oil | Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders | 39409 |
| Microlipid | Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders | 74410 |
| NanoVM 1-3 | Amino acid disorders, Carbohydrate disorders, Fatty Acid Oxidation Defects, Lactic Acidosis, Mitochondrial Disorders, Organic acid disorders, Urea cycle disorders, Biopterin (BH4) Deficiency, Hyperammonemia-Hyperonithinemia-Homocitrullinemia (HHH), Amish Microcephaly | 96320 |

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

| OTHER SUPPLEMENTS | | |
|---|--|--------------------------------|
| Product | Indications | PIN/SKU |
| NanoVM 4-8 | Amino acid disorders, Carbohydrate disorders, Fatty Acid Oxidation Defects, Lactic Acidosis, Mitochondrial Disorders, Organic acid disorders, Urea cycle disorders, Biopterin (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Amish Microcephaly | 96321 |
| Phlexy-Vits | Amino acid disorders, Carbohydrate disorders, Fatty Acid Oxidation Defects, Lactic Acidosis, Mitochondrial Disorders, Organic acid disorders, Urea cycle disorders, Biopterin (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Amish Microcephaly | 46994 |
| Polycal | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects, Lactic acidosis, Biopterin (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 79644 |
| Polycose | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate disorders, Mitochondrial disorders, Fatty acid oxidation defects, Lactic acidosis, Biopterin (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | Powder: 1405 Liquid: no PIN |
| Pro-Phree | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 47011 |
| Protifar | Carbohydrate disorders | 79645 |
| ProViMin Powder | 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 (BH4) Deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) | 31301 |
| Resource Beneprotein Instant Protein Powder | Carbohydrate disorders | 99557 |

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

| INFANT FEEDS COVERED BY THE IMD PROGRAM | | |
|--|--|----------------|
| Product | Indications | PIN/SKU |
| Calcilo XD | Hypercalcemia secondary to a listed disease | 91595 |
| Monogen | Fatty Acid Oxidation Defects | 47060 |
| Lipistart | Fatty Acid Oxidation Defects | 47068 |
| Portagen Powder | Mitochondrial disorders, Fatty Acid Oxidation Defects | 39581 |
| Pregestimil A+ Powder | Amino acid disorders, Organic acid disorders, Urea cycle disorders, Carbohydrate Disorders | 000045 |
| Ross Carbohydrate Free | Lactic acidosis | 1585 |

| COMPLETE ENTERAL FEEDS COVERED BY THE IMD PROGRAM | | |
|--|---------------------|----------------|
| <p>Effective January 1, 2009, reimbursement of complete enteral feeds under the IMD program was changed from specific disease-based criteria to the clinical criteria described below, applicable to all persons registered under the IMD program.</p> <p>Eligibility criteria:</p> <p>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:</p> <ul style="list-style-type: none"> • 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) <p>Exclusion criteria:</p> <p>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:</p> <ul style="list-style-type: none"> • voluntary meal replacement • convenience • food allergies • body building <p>(If the following products are the sole source of nutrition, coverage is available for eligible persons under the Ontario Drug Benefit Program)</p> | | |
| Product | Manufacturer | PIN/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 |

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

| COMPLETE ENTERAL FEEDS | | |
|---|---------------------|------------|
| Product | Manufacturer | PIN |
| 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 |
| 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 Just for Kids 1.5 Cal with Fibre | Nestle Nutrition | 99538 |
| Suplena | Abbott Nutrition | 1441 |
| Tolerex | Nestle Nutrition | 50524 |
| Vital HN | Abbott Nutrition | 1521 |
| Vivonex Pediatric | Nestle Nutrition | 50544 |
| Vivonex Plus | Nestle Nutrition | 50545 |