Ontario

Inherited Metabolic Diseases (IMD) Program

List of Disorders, Covered Drugs, Supplements and Specialty Foods

Effective December 2019

Drugs and Devices Division
Ministry of Health

Table of Contents

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

Inquiries regarding the IMD Program should be directed to:
Drugs and Devices Division
5700 Yonge Street, 3rd Floor
Toronto ON M2M 4K5
Tel: 416-327-8109, Toll-free: 1-866-811-9893
Fax: 416-327-8123
Email: PublicDrugPrgrms.moh@ontario.ca

About the IMD Program

Funding and administration of the Inherited Metabolic Diseases (IMD) Program was transitioned to Drugs and Devices Division (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.

Notable Changes for this Edition

New products and changes are highlighted.

Drugs and Supplements

- **Taurine 2mg** tablets have been added for the treatment of mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)
- **Serine and Glycine** can now be used to treat phosphoserine aminotransferase deficiency (PSAT) and Serine Deficiency Disorder (SDD)
- **Biotin 10mg capsules and Thiamine 100mg tablets** can now be used to treat Biotin-Thiamine-Responsive Basal Ganglia Disease
- **Phelxy-Vits (multivitamins), SolCarb and PolyCal cans** for the treatment of Hereditary Fructose Intolerance (HFI)
- **Cystadrops (cysteamine)** for the treatment of patients with diagnosis of cystinosis and evidence of corneal cystine crystal deposits (CCCDs) who are under the care of an ophthalmologist.

Medical Foods, Special Low Protein Foods and Other Supplements

- 8 new medical foods have been added
- 6 medical foods will have their listed approved disorders/indication(s) expanded
- 3 new special low protein foods have been added

Numerous “housekeeping” changes to update product DIN/NPNs, remove discontinued products and update product names due to reformulations.

- Discontinued products (removed from list):
  - HCU Cooler 10 red (SFS 1076)
  - MSUD Cooler 10 red (SFS 1079)
  - TYR Cooler 10 red (SFS 1083)

Disorders Covered by the IMD Program

Amino Acid Disorders

- 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)

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 and Fat Metabolism Disorders

- 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)
- 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

Lactic Acidosis (gluconeogenesis disorders)

- Fructose-1,6-bisphosphatase deficiency
- 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

- 2-methylbutyryl-CoA dehydrogenase deficiency
- 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]

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
- Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)
- N-acetyl glutamate synthetase (NAGS) deficiency
- Ornithine transcarbamylase (OTC) deficiency

Vitamin/Cofactor Disorders

- 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-aminoadipic semialdehyde dehydrogenase deficiency) [also listed under Organic Acid Disorders]
- Vitamin E (tocopherol) deficiency

Miscellaneous Disorders

- 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)

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.

Drug/Supplement	Approved Disorder(s)	DIN/PIN/ETC.
5-hydroxytryptophan	Biopterin deficiency; Pyridoxine-dependent epilepsy (PDE)	Product must have a valid NPN or may be compounded from powder
L-tryptophan	Pyridoxine-dependent epilepsy (PDE)	Apo-Tryptophan 02248538, 02248540 Teva-Tryptophan 02240333, 02240334
Ammonul	Urea Cycle Disorders	SAP
Beta-Carotene (Lumitene, oral)	Erythropoietic Protoporphyria (EPP)	
Betaine (Cystadane)	Homocystinuria (includes homocystinuria secondary to a listed disorder)	SAP
Biotin	Biotinidase deficiency; Holocarboxylase synthetase deficiency; Propionic acidemia; Pyruvate carboxlyase deficiency Biotin-Thiamine-Responsive Basal Ganglia Disease	No DIN
Chenodeoxycholic acid	Cerebrotendinous Xanthomatosis	No DIN
Cholesterol	Smith-Lemli-Opitz syndrome (SLOS)	No DIN
Citrate C	Renal tubular acidosis, secondary to another listed disorder	HSC
Citrulline	Urea cycle disorders; Lysinuric protein intolerance; MELAS syndrome	No DIN

Coenzyme-Q	Mitochondrial disorders; Glutaric Aciduria II, secondary mitochondrial defects	No DIN
Copper histidine	Menkes disease	HSC
Creatine	Mitochondrial myopathies; Creatine synthesis/transport deficiency; Gyrate atrophy	SAP
Cupric Chloride	Menkes disease	
Cysteamine bitartrate (Procysbi)	Infantile Nephropathic Cystinosis **Note:** Eligibility criteria for IMD program funding (all criteria must be met): 1. For the diagnosis of infantile nephropathic cystinosis with documented cystinosin and lysosomal cysteine transporter gene mutation and; 2. For patients under the care of a physician with experience in the diagnosis of management of cystinosis; For continued funding, a yearly follow up report should be submitted to the IMD program by the prescribing physician.	02464705 02464713
Cysteamine (Cystagon or cysteamine powder for compounded products or non-viscous compounded cysteamine drops)	Cystinosis	SAP
Cysteamine bitartrate (Cystadrops)	Cystinosis **Note:** Eligibility criteria for IMD program funding (all criteria must be met): 1. For the treatment of patients with diagnosis of cystinosis and evidence of corneal cystine crystal deposits (CCCDs) who are under the care of an ophthalmologist.	02485605

Dichloroacetate	Pyruvate dehydrogenase deficiency	SAP
Glycine	Isovaleric academia; Creatine transport deficiency Serine Deficiency Disorder (SDD) 3-phosphoglycerate dehydrogenase deficiency (3PGDH) phosphoserine aminotransferase deficiency (PSAT) phosphoserine phosphatase (PSP)	No DIN
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
L-carnitine	Organic acidemias; Fatty acid oxidation defects; carnitine uptake defect, carnitine deficiency secondary to another listed disorder	02144336, 02144328, 02144344
L-isoleucine	MSUD, organic acid disorders	No DIN
L-lysine	Lysinuric protein intolerance	No DIN
L-valine	MSUD, organic acid disorders	No DIN
Lipoic Acid	Mitochondrial disorders	
Mannose	Congenital Disorders of Glycosylation	No DIN
Niacinamide	Mitochondrial cytopathies	No DIN
Nitisinone (NTBC, Orfadin)	Tyrosinemia Type I	SAP
Ornithine HCl, ornithine L-aspartate	Urea cycle disorders; Hyperammonemia-Hyperornithinemia-Homocitrullinemia (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	Product must have a valid DIN or NPN
Riboflavin	Mitochondrial disorders; Glutaric aciduria Riboflavin Transporter Deficiency	No DIN
Sapropterin (Kuvan)[1]	Biopterin deficiency	02350580
Serine	Serine Deficiency Disorder (SDD) 3-phosphoglycerate dehydrogenase deficiency (3PGDH) phosphoserine aminotransferase deficiency (PSAT) phosphoserine phosphatase (PSP)	No DIN
Sodium benzoate	Creatine synthesis/transport deficiency; Urea cycle disorders	No DIN
Sodium phenylbutyrate (Buphenyl and Pheburane)[2]	Urea cycle disorders	Buphenyl SAP Pheburane 02436663
Taurine	Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)	No DIN
Thiamine (Vitamin B1)	Mitochondrial cytopathies; Thiamine deficiency in the presence of IMD Biotin-Thiamine-Responsive Basal Ganglia Disease	Product must have a valid DIN or NPN
Tiopronin (Thiola)	Cystinuria	SAP
Vitamin A/D/E/K	Abetalipoproteinemia; Mitochondrial disorders	Product must have a valid DIN or NPN
Vitamin A	Abetalipoproteinemia	Product must have a valid DIN or NPN

[1] Sapropterin (Kuvan®) 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.

[2] The Ministry of Health is aware that the Buphenyl formulation of sodium phenylbutyrate may no longer be available in Canada. Funding for Ravicti™ is considered under the Exceptional Access Program (EAP) only.

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
Vitamin E	Abetalipoproteinemia; Hereditary deficiency of tocopherol transport protein; Mitochondrial disorders; Vitamin E deficiency	Product must have a valid DIN or NPN
Vitamin K1	Mitochondrial disorders	Product must have a valid DIN or NPN
Vitamin K3	Mitochondrial disorders	Product must have a valid DIN or NPN

Medical Foods

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
BCAD 1	MSUD	Mead Johnson	SFS1071
Complex MSUD Amino Acid Blend	MSUD	Nutricia	47031
Complex Essential MSD Drink Mix – Vanilla	MSUD	Nutricia	59720
Cyclinex–1	Urea cycle disorders, Gyrate atrophy, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) Creatine synthesis/transport deficiency	Abbott Nutrition	47012
Cyclinex–2	Urea cycle disorders, Gyrate atrophy, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) Creatine synthesis/transport deficiency	Abbott Nutrition	47013
Essential Amino Acid Mix	Urea cycle disorders, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH) Gyrate Atrophy, Creatine synthesis/transport deficiency	Nutricia	SFS1556
EAA Supplement	Urea cycle disorders, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Gyrate Atrophy, Creatine synthesis/transport deficiency	Vitaflo	47052
GA1 Express 15, Unflavoured	Glutaric aciduria Type I, Pyridoxine (B6) dependent seizures	Vitaflo	SFS1075
GA1 Gel 10, 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
GlutarAde Junior Drink Mix	Glutaric Aciduria Type I, Pyridoxine dependent (B6) seizures	Nutricia	
Glytactin RTD 10, Original	PKU	Cambrooke	TBC

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
Glytactin RTD 10, Chocolate	PKU	Cambrooke	TBC
Glytactin RTD 15, Original	PKU	Cambrooke	SFS1802
Glytactin RTD 15, Chocolate	PKU	Cambrooke	SFS1801
Glytactin BetterMilk 15, Original	PKU	Cambrooke	SFS1809
Glytactin BetterMilk 15, Orange Crème	PKU	Cambrooke	SFS1810
Glytactin BetterMilk 15, Strawberry Crème	PKU	Cambrooke	SFS1811
Glytactic BetterMilk Lite	PKU	Cambrooke	SFS2392
Glytactin Complete 10, Fruit Frenzy	PKU	Cambrooke	SFS1816
Glytactin Complete 10, Peanut Butter	PKU	Cambrooke	SFS1817
Glytactin Complete 15 Fruit Frenzy	PKU	Cambrooke	SFS1818
Glytactin Complete 15 Peanut Butter	PKU	Cambrooke	SFS1819
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 Anamix Next	Homocystinuria	Nutricia	
HCU Cooler 15 orange	Homocystinuria	Vitaflo	18889
HCU Cooler 15 red	Homocystinuria	Vitaflo	SFS0538
HCU Cooler 20 red	Homocystinuria	Vitaflo	SFS1077
HCU express 15, Unflavoured	Homocystinuria	Vitaflo	1809
HCU Express 20	Homocystinuria	Vitaflo	SFS1078

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
HCU 10 gel, Unflavoured	Homocystinuria	Vitaflo	1808
HCU Lophlex LQ Mixed Berry Blast	Homocystinuria	Nutricia	SFS1086
Homactin AA Plus	Homocystinuria	Cambrooke	
Hominex – 1	Homocystinuria	Abbott Nutrition	46986
Hominex – 2	Homocystinuria	Abbott Nutrition	46979
Isovactin AA Plus	Isovaleric acidemia	Cambrooke	
IVA Anamix Next	Isovaleric acidemia	Nutricia	
IVA Cooler 15 red	Isovaleric acidemia	Vitaflo	SFS1315
Ketocal	Glucose transport 1 deficiency syndrome (GLUT 1-DS), Pyruvate dehydrogenase (PDH) deficiency)	Nutricia	SFS3575
Ketocal LQ4:1 Vanilla	Glucose transport 1 deficiency syndrome (GLUT 1-DS), Pyruvate dehydrogenase (PDH) deficiency)	Nutricia	SFS0879
Ketocal LQ4:1 Unflavoured	Glucose transport 1 deficiency syndrome (GLUT 1-DS), Pyruvate dehydrogenase (PDH) deficiency)	Nutricia	SFS3576
Ketonex – 1	MSUD	Abbott Nutrition	47007
Ketonex – 2	MSUD	Abbott Nutrition	47009
MMaPa Anamix Next	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Nutricia	
MMA/PA Cooler 15 red	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	SFS1314
MMA/PA express 15 Unflavoured	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	47054
MMA/PA gel 10 Unflavoured	Methylmalonic Acidemia, Propionic Acidemia	Vitaflo	47053
MSUD Cooler 15 orange	MSUD	Vitaflo	1822

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
MSUD Cooler 15 red	MSUD	Vitaflo	SFS0536
MSUD Cooler 20 red	MSUD	Vitaflo	SFS1080
MSUD Express 15 (unflavoured sachet)	MSUD	Vitaflo	1807
MSUD Express 20 Unflavoured	MSUD	Vitaflo	SFS1081
MSUD Gel 10 Unflavoured	MSUD	Vitaflo	1806
MSUD Lophlex LQ Mixed Berry Blast	MSUD	Nutricia	SFS1087
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
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

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
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 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- Unflavoured (can)	PKU, Biopterin deficiency	Nutricia	
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

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
PhenylAde Essential Drink Mix- Unflavoured (pouch)	PKU, Biopterin deficiency	Nutricia	
Phenylade GMP	PKU, Biopterin deficiency	Nutricia	
Phenylade GMP Mix In	PKU, Biopterin deficiency	Nutricia	
PhenylAde MTE Amino Acid Blend Unflavoured	PKU, Biopterin deficiency	Nutricia	47015
PhenylAde MTE Amino Acid Blend (pouch)	PKU, Biopterin deficiency	Nutricia	1315

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
PhenylAde PheBloc LNAA Pouches	PKU **Note:** Eligibility criteria for IMD program funding (all criteria must be met): 1. The patient will have untreated/late-diagnosed PKU OR early diagnosed PKU with elevated blood phenylalanine concentrations beyond his/her ability to correct with the phenylalanine-restricted diet;AND 2. The patient will have blood phenylalanine concentrations which are chronically above his/her treatment goal;AND 3. The patient will be 13 years of age or over; AND 4. The patient will not be pregnant or planning a pregnancy;AND 5. The patient will not also be treated with sapropterin dihydrochloride (Kuvan);AND 6. 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 & obese individuals); AND 7. 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 & obese individuals); AND 8. The patient will use PheBloc only under the direction of a metabolic geneticist/metabolic dietitian. 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.	Nutricia	SFS1487 (pouches),
Phenyl-Free 1	PKU, Biopterin deficiency	Mead Johnson	46988
Phenyl-Free 2	PKU, Biopterin deficiency	Mead Johnson	47029

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
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 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
PKU Express 15 lemon	PKU, Biopterin deficiency	Vitaflo	1805
PKU Express 15 orange	PKU, Biopterin deficiency	Vitaflo	1804
PKU Express 15 tropical	PKU, Biopterin deficiency	Vitaflo	1818
PKU Express 15 unflavoured	PKU, Biopterin deficiency	Vitaflo	1803
PKU Express 20 lemon	PKU, Biopterin deficiency	Vitaflo	SFS1820

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
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 10 orange	PKU, Biopterin deficiency	Vitaflo	1801
PKU gel 10 raspberry	PKU, Biopterin deficiency	Vitaflo	47045
PKU gel 10 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
PKU Minis	PKU, Biopterin Deficiency	MetaX	SFS3581
PKU Sphere 20 – Red Berry	PKU, Biopterin deficiency	Vitaflo	
PKU Sphere 20 – Vanilla	PKU, Biopterin deficiency	Vitaflo	
Plus8 Minis	Urea cycle disorders,Gyrate atrophy, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Creatine synthesis/transport deficiency	MetaX	SFS3582
Promactin AA Plus	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Cambrooke	
Propimex–1	Methylmalonic acidemia, Propionic acidemia	Abbott Nutrition	47008
Propimex–2	Methylmalonic acidemia, Propionic acidemia	Abbott Nutrition	47010

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
Tylactin Build 20	Tyrosinemia	Cambrooke	SFS3583
Tylactin RTD 15, Original	Tyrosinemia	Cambrooke	SFS1804
Tylactin Restore 10, Citrus	Tyrosinemia	Cambrooke	SFS1803
Tyr Anamix Next	Tyrosinemia	Nutricia	
Tyr Sphere 20 Berry	Tyrosinemia	Nestle	SFS3578
Tyrex–1	Tyrosinemia	Abbott Nutrition	47006
Tyrex–2	Tyrosinemia	Abbott Nutrition	47016
TYROS 1	Tyrosinemia	Mead Johnson	SFS1072
TYR Cooler 15 orange	Tyrosinemia	Vitaflo	67896
TYR Cooler 15 red	Tyrosinemia	Vitaflo	SFS0537
TYR Cooler 20 red	Tyrosinemia	Vitaflo	SFS1084
TYR Express 15	Tyrosinemia	Vitaflo	1811
TYR Express 20	Tyrosinemia	Vitaflo	SFS1085
TYR gel 10 Unflavoured	Tyrosinemia	Vitaflo	46899
TYR Lophlex LQ Mixed Berry Blast	Tyrosinemia	Nutricia	SFS1088
UCD Anamix Junior, unflavoured	Creatine synthesis/transport deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Gyrate atrophy, Urea cycle disorders	Nutricia	SFS1089
UCD Anamix Junior, vanilla	Creatine synthesis/transport deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH), Gyrate atrophy, Urea cycle disorders	Nutricia	SFS1090
I-Valex–1	Isovaleric acidemia	Abbott Nutrition	46993

Medical Foods Product	Disorder(s)	Mfr	PIN/ SFS SKU
I-Valex–2	Isovaleric acidemia	Abbott Nutrition	46992
Vilactin AA Plus	MSUD	Cambrooke	
XLeu Maxamum	Isovaleric acidemia	Nutricia	78966
XLys, XTrp Analog	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Nutricia	47037
XLys, XTrp Maxamum	Glutaric aciduria Type I, Type II, Pyridoxine (B6) dependent seizures	Nutricia	78967
XMet Analog	Homocystinuria	Nutricia	47036
XMet Maxamum	Homocystinuria	Nutricia	46987
XMTVI Maxamum	Methylmalonic acidemia (MMA), Propionic acidemia (PA)	Nutricia	78971
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
XPhe Maxamum Unflavoured Powder (sachet	PKU, Biopterin deficiency	Nutricia	1251
XPhe, XTyr Analog	Tyrosinemia	Nutricia	47038
XPhen, TYR Maxamum	Tyrosinemia	Nutricia	SFS 2433

Special Low Protein Foods

Approved Disorders: Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II, Biopterin deficiency, Creatine synthesis/transport deficiency

Special Low Protein Foods Product	PIN/SFS SKU
Aproten Crispbread Rusks 260g	31128
Aproten Crackertoast 250g	31130
Aproten Farina/Pizza Mix	31172
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: rigatini 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 Bread	401010
Cambrooke Bagels plain	SFS0444
Cambrooke Brooklyn Dog Buns	40616
Cambrooke Creamy Hot Cereal Cinnamon	SFS2888
Cambrooke Creamy Hot Cereal Vanilla	SFS3577
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 Solo Elbows	40455
Cambrooke Foods – Pasta, Portabella Spinach Ravioli	40116

Special Low Protein Foods Product	PIN/SFS SKU
Cambrooke Foods – Swiss Cheese Slices	40312
Cambrooke Foods - Tweekz	40902
Cambrooke Foods –Tortilla Wraps	2118
Cambrooke Homestyle White Bread (1 loaf)	40110
Cambrooke Imitation Mozzarella Shredded Cheese	40315
Cambrooke Macaroni & Cheese Duets	40406
Cambrooke MixQuick Multi-Purpose Batter (2 lbs)	SFS 1354
Cambrooke Pita Pockets	SFS0226
Cambrooke Short Grain Japanese Rice (1kg)	40407
Cambrooke Tuscan Pizza Crusts (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 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. Porridge 500g	2292
D.S. Bread Machine Baking Mix 1.8kg	2294

Special Low Protein Foods Product	PIN/SFS SKU
Eggz	1370
George Washington Seasoning: brown 31g	141702
George Washington Seasoning: golden 31g	141701
Kingsmill Egg Replacer 350 g	31197
La Tiara Taco Shells	SFS0079
Loprofin Pasta: animal pasta 500g	114155
Loprofin Pasta: fusilli 500g	126113
Loprofin Pasta: lasagna (Nutricia)	SFS0515
Loprofin Pasta: penne 500g	126111
Loprofin Pasta: spaghetti 500g	126112
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 lp Drink	12652
Milupa lp Dry Cereal Chocolate Ringlets	30947
Milupa lp Dry Cereal Flakes	30940
Milupa lp Fruity Cereal Mix – Apple Banana	65991
Milupa lp Fruity Cereal Mix – Pear	65992
Pea-Not Butter	SFS1968
Promin Pastameal (Porridge)	NFDC Code 936
Shake ‘N’ Cheese	40309
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

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
Glycosade (high-amylopectin-containing cornstarch)	Glycogen storage disease (GSD) **Note:** Eligibility criteria for IMD program funding (all criteria must be met: 1. The patient will have a diagnosis of glycogen storage disease type 0, 1a/b, type 3, 6 or type 9 which requires cornstarch feedings overnight for metabolic control; AND 2. The patient will be ≥24 months of age; AND 3. The patient will use Glycosade, only under the direction of a metabolic geneticist/metabolic dietitian; AND 4. 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. **EXCEPTION:** If all attempts with conventional therapy (regular cornstarch & 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. Continued use of Glycosade is approved only for patients who demonstrate improved metabolic control. Glucose transporter 1 deficiency syndrome (GLUT1-DS)* with the following eligibility criteria: o The patient has confirmed diagnosis of GLUT1-DS; and o The patient will use Glycosade to maintain blood sugars overnight along with low glycemic index diet during the day; and Patient has demonstrated failure to comply with ketogenic diet.	SFS0624
KetoVie Chocolate	Glucose transporter 1 deficiency syndrome (GLUT1-DS), Pyruvate dehydrogenase (PDH) deficiency	50203
KetoVie Vanilla	Glucose transporter 1 deficiency syndrome (GLUT1-DS), Pyruvate dehydrogenase (PDH) deficiency	50103

Other Supplements Product	Disorder(s)	PIN/ SFS SKU
Liquigen	Abetalipoproteinemia, Fatty acid oxidation defects and Fat Metabolism Disorders, Lactic Acidosis, Mitochondrial disorders	SFS1490
MCT Pro-Cal	Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders	22990
MCT Oil	Abetalipoproteinemia, Fatty acid oxidation defects, Lactic Acidosis, Mitochondrial disorders, Glucose transport 1 deficiency syndrome (GLUT 1-DS), Pyruvate dehydrogenase (PDH) deficiency	39409
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 Hereditary Fructose Intolerance (HFI)	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) Hereditary Fructose Intolerance (HFI)	SFS1654
Pro-Phree	Amino acid disorders, Organic acid disorders, Urea cycle disorders, Glutaric Aciduria Type II (GA2), Biopterin deficiency, Hyperammonemia-Hyperornithinemia-Homocitrullinemia (HHH)	47011
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

Other Supplements Product	Disorder(s)	PIN/ SFS SKU
Resource Beneprotein Instant Protein Powder	Carbohydrate disorders, Organic acid disorders, Glucose transporter 1 deficiency syndrome (GLUT1-DS), Pyruvate dehydrogenase (PDH) deficiency	99557
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) Hereditary Fructose Intolerance (HFI)	SFS1316

Infant Feeds

Infant Feeds Product	Disorder(s)	PIN/ SFS SKU
Calcilo XD	Hypercalcemia secondary to a listed disease	91595
Monogen DHA & ARA	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

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 <u>sole</u> 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 1.5 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 1.2	Nestle Nutrition	99458
Isosource Fibre 1.2	Nestle Nutrition	99459
Jevity 1	Abbott Nutrition	1499
Nutren Junior	Nestle Nutrition	111912

Complete Enteral Feeds Product	Manufacturer	PIN/SFS SKU
Nutren Junior Fibre with Prebio	Nestle Nutrition	111911
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
Vivonex Pediatric	Nestle Nutrition	SFS0212
Vivonex Plus	Nestle Nutrition	50545