



Formulas for Metabolic Conditions

Children with metabolic conditions should have their nutrition managed by a team of specialists including a dietician familiar with metabolic conditions. The following is a list of metabolic formulas. Due to frequently changing product information, we advise checking the manufacturer's website as well. The Medical Home Portal does not endorse a specific brand.

Name	Ages	Use	Manufacturer
Amino Acid Metabolic Disorders/Protein Free Diet			
PFD Toddler	toddlers	Amino acid metabolic disorders/those requiring a protein-free diet. Protein and amino acid free product. Vanilla powder.	Mead Johnson (Enfamil US)
PFD 2	children & adults	Amino acid metabolic disorders/those requiring a protein-free diet. Protein and amino acid free product. Vanilla powder.	Mead Johnson (Enfamil US)
Pro-Phree	infants & toddlers	Protein restriction or those who require extra calories, minerals, and vitamins. Protein-free. Unflavored powder.	Abbott
Glutaric acidemia type 1			
GA	all ages	Glutaric acidemia type 1. Free of the essential amino acids lysine and tryptophan. Vanilla powder.	Mead Johnson (Enfamil US)
GA-1 Anamix Early Years	infants	Glutaric acidemia type I. Lysine-free and low tryptophan free. Contains prebiotics. Unflavored powder.	Nutricia
GA Gel	1-10 yrs	Glutaric acidemia type I. Lysine-free and low tryptophan free. Unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
GA Express 15	3 yrs & older	Glutaric acidemia type I. Lysine-free and low tryptophan free. Unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
GlutarAde Essential GA-1	children & adults	Glutaric acidemia type I. Lysine-free and low tryptophan. Unflavored powder.	Nutricia
GlutarAde GA-1 Amino Acid Blend	infants	Glutaric acidemia type I. Lysine-free and low tryptophan protein supplement. Unflavored powder.	Nutricia
GlutarAde GA-1 Junior	children & adults	Glutaric acidemia type I. Lysine-free and low tryptophan. Unflavored powder.	Nutricia
Glutarex-1	infants & toddlers	Glutaric aciduria type I. Lysine- and tryptophan-free. Unflavored powder.	Abbott

Name	Ages	Use	Manufacturer
Glutarex-2	children & adults	Glutaric aciduria type I. Lysine- and tryptophan-free. Unflavored powder.	Abbott
Homocystinuria or Hypermethioninemia			
HCU Anamix Early Years	infants	Homocystinuria or hypermethioninemia. Methionine free. Contains prebiotics. Unflavored powder.	Nutricia
HCU Anamix Next	1 yr & older	Homocystinuria or hypermethioninemia. Methionine free. Mildly flavored powder.	Nutricia
HCU Cooler	3 yrs & older	Homocystinuria or hypermethioninemia. Methionine-free. "Red and orange" flavored ready to feed. Supplemental nutrition.	Vita-Flo-USA (Nestle)
HCU Express 15	3 yrs & older	Homocystinuria or hypermethioninemia. Methionine-free. Unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
HCU Gel	1-10 yrs	Homocystinuria or hypermethioninemia. Methionine-free. Unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
HCU LoPhlex LQ	4 yrs & older	Homocystinuria (HCU). Methionine free. Fruit-based. Mixed berry blast ready to feed.	Nutricia
HCU Xmet Maxamum	>8 yrs	Homocystinuria or hypermethioninemia. Methionine free. Does not contain fats. Supplemental nutrition. Unflavored powder.	Nutricia
HCY 1	infants & toddlers	Homocystinuria. Free of the essential amino acid methionine. Vanilla powder.	Mead Johnson (Enfamil US)
HCY 2	children & adults	Homocystinuria. Free of the essential amino acid methionine. Vanilla powder.	Mead Johnson (Enfamil US)
Hominex-1	infants & toddlers	Homocystinuria or hypermethioninemia. Methionine-free. Unflavored powder.	Abbott
Hominex-2	children & adults	Homocystinuria or hypermethioninemia. Methionine-free. Unflavored powder.	Abbott
Leucine metabolism disorders including isovaleric acidemia			
IVA Anamix Early Years	infants	Isovaleric acidemia and other proven disorders of leucine metabolism. Leucine-free. Contains prebiotics. Unflavored powder.	Nutricia
IVA Anamix Next	1 yr & older	Isovaleric acidemia and other proven disorders of leucine metabolism. Leucine-free. Mildly flavored powder.	Nutricia
I-Valex-1	infants & toddlers	Leucine catabolism disorder. Leucine-free. Unflavored powder.	Abbott

Name	Ages	Use	Manufacturer
I-Valex-2	children & adults	Leucine catabolism disorder. Leucine-free. Unflavored powder.	Abbott
LMD	all ages	Leucine metabolism disorders, including isovaleric acidemia. Free of the essential amino acid leucine. Vanilla powder.	Mead Johnson (Enfamil US)
XLeu Maxamum	>8 yrs	Isovaleric acidemia and other proven disorders of leucine metabolism. Leucine-free. Unflavored powder.	Nutricia
Maple syrup urine disease (MSUD)			
BCAD 1	infants & toddlers	Maple syrup urine disease (MSUD). Isoleucine-, leucine- and valine-free. Vanilla powder.	Mead Johnson (Enfamil US)
BCAD 2	children & adults	Maple syrup urine disease (MSUD). Isoleucine-, leucine- and valine-free. Vanilla powder.	Mead Johnson (Enfamil US)
Complex MSD Amino Acid Mix	children & adults	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Contains proteins, no fat or sugars. Supplemental nutrition. Unflavored powder.	Nutricia
Complex Essential MSD	all ages	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Vanilla powder.	Nutricia
Complex MSD Junior	children & adults	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Unflavored powder.	Nutricia
Isoleucine 500 and 1000	1 yr & older	Maple syrup urine disease (MSUD). Unflavored powder packets of isoleucine. Supplemental nutrition.	Vita-Flo-USA (Nestle)
Ketonex-1	infants & toddlers	Maple syrup urine disease (MSUD). Isoleucine-, leucine- and valine-free. Unflavored powder.	Abbott
Ketonex-2	children & adults	Maple syrup urine disease (MSUD). Isoleucine-, leucine- and valine-free. Unflavored powder.	Abbott
MSUD Anamix Early Years	infants	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Contains prebiotics. Unflavored powder.	Nutricia
MSUD Cooler	3 yrs & older	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. "Red and orange" flavored ready to feed. Supplemental nutrition.	Vita-Flo-USA (Nestle)

Name	Ages	Use	Manufacturer
MSUD Express 15 and Express 20	3 yrs & older	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
MSUD Gel	1-10 yrs	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
MSUD LoPhlex LQ	4 yrs & older	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Fruit-juice based. Mixed berry blast ready to feed.	Nutricia
MSUD Maxamum	>8 yrs	Maple syrup urine disease (MSUD). Free of isoleucine, leucine, and valine. Fat free. Supplemental nutrition. Orange powder.	Nutricia
Phenylketonuria (PKU)			
Phenex-1	infants & toddlers	PKU. Phenylalanine-free. Unflavored powder.	Abbott
PhenylAde Essential Drink Mix	children & adults	PKU. Phenylalanine-free. Orange-crème, vanilla, chocolate, strawberry, and unflavored powder.	Nutricia
PhenylAde GMP Drink Mix and Mix-Ins	1 yr & older	Phenylketonuria (PKU). Low phenylalanine protein drink mix. Vanilla and original flavored powder.	Nutricia
PhenylAde MTE	children & adults	PKU. Phenylalanine-free. Added minerals. Unflavored powder.	Nutricia
PhenylAde PheBLOC	1 yr & older	Phenylketonuria (PKU). Phenylalanine-free, contains remaining amino acids. Supplemental nutrition. Unflavored powder and tablets.	Nutricia
PhenylAde 40 Drink Mix	children & adults	PKU. Phenylalanine-free, contains more protein than essential drink mix. Citrus and unflavored powder.	Nutricia
PhenylAde 60 Drink Mix	children & adults	PKU. Phenylalanine-free, contains more protein than essential drink mix. Vanilla and unflavored powder.	Nutricia
Phenyl-Free 1	infants & toddlers	Phenylketonuria (PKU). Free of the essential amino acid phenylalanine. Vanilla powder.	Mead Johnson (Enfamil US)
Phenyl-Free 2	children & adults	Phenylketonuria (PKU). Free of the essential amino acid phenylalanine. Vanilla powder.	Mead Johnson (Enfamil US)

Name	Ages	Use	Manufacturer
Phenyl-Free 2 HP	children & adults	Phenylketonuria (PKU). Free of the essential amino acid phenylalanine. Contains higher protein content. Vanilla powder.	Mead Johnson (Enfamil US)
PKU Air	3 yrs & older	PKU. Free of the essential amino acid phenylalanine. Green (citrus), gold (coffee), and yellow (mango) flavored with variable grams of protein, low calorie ready to feed. Supplemental nutrition.	Vita-Flo-USA (Nestle)
PKU Cooler	3 yrs & older	PKU. Free of the essential amino acid phenylalanine. "Red, orange, purple, and white flavored" with variable grams of protein, ready to feed. Supplemental nutrition.	Vita-Flo-USA (Nestle)
PKU Express 15 and Express 20	3 yrs & older	PKU. Free of the essential amino acid phenylalanine. Lemon, orange, tropical, or unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
PKU Gel	1-10 yrs	PKU. Free of the essential amino acid phenylalanine. Raspberry, orange, or unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
PKU Lophlex	9 yrs & older	PKU. Phenylalanine-free. Supplemental nutrition. Berry or orange powder mixes into paste or drink.	Nutricia
PKU Lophlex LQ	4 yrs & older	PKU. Phenylalanine-free. Orange, tropical, and berry ready to feed.	Nutricia
PKU Periflex Advanced	older children & adults	PKU. Phenylalanine-free. Orange and unflavored powder.	Nutricia
PKU Periflex Early Years	infants	PKU. Phenylalanine-free, contains prebiotics. Unflavored powder.	Nutricia
PKU Periflex Jr. Plus	children	PKU. Phenylalanine-free. Berry, vanilla, orange, and unflavored powder.	Nutricia
PKU Periflex LQ	children & adults	PKU. Phenylalanine-free. Berry-crème and orange-crème ready to feed.	Nutricia
PKU Sphere	4 yrs & older	PKU. Low in essential amino acid phenylalanine. Contains glycomacropeptide for palatability. Red berry and vanilla flavored ready to feed with variable g of Phe.	Vita-Flo-USA (Nestle)

Name	Ages	Use	Manufacturer
PKU Trio	1 & older	PKU. Free of the essential amino acid phenylalanine Vanilla and unflavored medical food powder.	Vita-Flo-USA (Nestle)
XPhe Maxamum	older children & adults	PKU. Phenylalanine-free. Contains protein and carbohydrate, no fats. Supplemental nutrition. Orange and unflavored powder.	Nutricia
Propionic or Methylmalonic Acidemia			
Methionine 100	1 yr & older	Propionic or methylmalonic acidemia. Unflavored powder packets of methionine. Supplemental nutrition.	Vita-Flo-USA (Nestle)
MMA/PA Gel	1-10 yrs	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free. Unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
MMA/PA Express 15	3 yrs & older	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free. Unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
MMA/PA Cooler	3 yrs & older	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free. "Red" flavored ready to feed.	Vita-Flo-USA (Nestle)
MMP/PA Anamix Early Years	infants	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free and isoleucine- low. Contains prebiotics. Unflavored powder.	Nutricia
MMP/PA Anamix Next	1 yr & older	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free and isoleucine- low. Mildly flavored powder.	Nutricia
OA 1	infants & toddlers	Propionic or methylmalonic acidemia. Isoleucine, methionine-, threonine-, and valine-free. Vanilla powder.	Mead Johnson (Enfamil US)
OA 2	children & adults	Propionic or methylmalonic acidemia. Isoleucine, methionine-, threonine-, and valine-free. Vanilla powder.	Mead Johnson (Enfamil US)
Propimex-1	infants & toddlers	Propionic or methylmalonic acidemia. Methionine- and valine-free; low in isoleucine and threonine. Unflavored powder.	Abbott

Name	Ages	Use	Manufacturer
Propimex-2	children & adults	Propionic or methylmalonic acidemia. Methionine- and valine-free; low in isoleucine and threonine. Unflavored powder.	Abbott
XMTVI Maxamum	>8 yrs	Propionic or methylmalonic acidemia. Methionine-, threonine-, valine-free and isoleucine-low. Fat free. Supplemental nutrition. Orange powder.	Nutricia
Tyrosinemia			
Phenylalanine50	1 yr & older	Tyrosinemia. Unflavored powder packets of phenylalanine. Supplemental nutrition.	Vita-Flo-USA (Nestle)
TYR Anamix Early Years	infants	Tyrosinemia. Phenylalanine- and tyrosine-free. Contains prebiotics. Unflavored powder.	Nutricia
TYR Anamix Next	infants	Tyrosinemia. Phenylalanine- and tyrosine-free. Mildly flavored powder.	Nutricia
TYR Cooler	3 yrs & older	Tyrosinemia. Iron fortified, free of the essential amino acids phenylalanine and tyrosine. "Red and orange" flavored ready to feed. Supplemental nutrition.	Vita-Flo-USA (Nestle)
TYR Express 15 and Express 20	3 yrs & older	Tyrosinemia. Iron fortified, free of the essential amino acids phenylalanine and tyrosine. Unflavored powder mixes into paste or drink. Supplemental nutrition.	Vita-Flo-USA (Nestle)
TYR Gel	1-10 yrs	Tyrosinemia. Iron fortified, free of the essential amino acids phenylalanine and tyrosine. Unflavored powder mixes into paste or drink.	Vita-Flo-USA (Nestle)
Tyrex-1	infants & toddlers	Tyrosinemia types I, II or III. Phenylalanine- and tyrosine-free. Unflavored powder.	Abbott
Tyrex-2	children & adults	Tyrosinemia types I, II or III. Phenylalanine- and tyrosine-free. Unflavored powder.	Abbott
TYROS 1	infants & toddlers	Tyrosinemia. Iron fortified, free of the essential amino acids phenylalanine and tyrosine. Vanilla powder.	Mead Johnson (Enfamil US)
TYROS 2	children & adults	Tyrosinemia. Iron fortified, free of the essential amino acids phenylalanine and tyrosine. Vanilla powder.	Mead Johnson (Enfamil US)
Urea Cycle Disorders/Waste Nitrogen Disorders			

Name	Ages	Use	Manufacturer
Arginine 500 & 2000	1 yr & older	Inborn errors of metabolism. Arginine amino acid supplement. Unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
Citrulline 200 & 1000	1 yr & older	Inborn errors of metabolism. Citrulline amino acid supplement. Unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
Creatine 500	1 yr & older	Inborn errors of metabolism. Creatine amino acid supplement. Unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
Cyclinex-1	infants & toddlers	Urea cycle disorder, gyrate atrophy or HHH syndrome. Nonessential amino acid-free. Unflavored powder.	Abbott
Cyclinex-2	children & adults	Urea cycle disorder, gyrate atrophy or HHH syndrome. Nonessential amino acid-free. Unflavored powder.	Abbott
EAA Supplement	3 yrs & older	Urea cycle disorders (UCD). Essential amino acid supplement. Unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
UCD Anamix Junior	>1 yr	Urea cycle disorders (UCD); hyperammonemia, hyperornithinemia, and homocitrullinemia (HHH) syndrome; and gyrate atrophy. Contains essential amino acids to help maintain a positive nitrogen balance. Vanilla and unflavored powder.	Nutricia
UCD Trio	1 yr & older	Urea cycle disorders (UCD). Unflavored powder.	VitaFlo (Nestle)
WND 1	infants & toddlers	Waste nitrogen disorders/inborn errors of the urea cycle. Free of nonessential amino acids. Vanilla powder.	Mead Johnson (Enfamil US)
WND 2	children & adults	Waste nitrogen disorders/inborn errors of the urea cycle. Free of nonessential amino acids. Vanilla powder.	Mead Johnson (Enfamil US)
Errors of Fat Metabolism			
BetaQuik	3 yrs & older	Conditions requiring a source of medium-chain triglycerides (MCT). Contains MCT oil. Supplemental nutrition. Unflavored ready to feed, 225 mL bottle.	VitaFlo (Nestle)
Carbzero	3 yrs & older	Conditions requiring a source of long chain triglycerides (LCT). Contains LCT oil. Supplemental nutrition. Unflavored ready to feed, 225 mL bottle.	Nestle

Name	Ages	Use	Manufacturer
Enfaport	infants	Long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency or chylothorax. Contains medium chain triglycerides (MCT) oils and high protein. Unflavored ready to feed liquid.	Mead Johnson (Enfamil US)
LipiStart	>1 yr	Long-chain fatty acid oxidation disorders, fat malabsorption and disorders requiring a high medium-chain triglycerides (MCT) and low long-chain triglycerides (LCT) diet. Unflavored powder.	VitaFlo (Nestle)
Liquigen	all ages	Long-chain fatty acid oxidation disorders and conditions managed with ketogenic diet, chylothorax, fat malabsorption, short bowel syndrome, and other conditions where medium chain triglycerides (MCTs) are part of dietary management. Contains MCTs. Supplemental nutrition. Unflavored emulsion.	Nutricia
MCTProCal	1 yr & older	Long-chain fatty acid oxidation disorders, fat malabsorption, and other disorders requiring a high MCT, low long-chain triglyceride (LCT) diet. MCT oil in unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
Monogen	>1 yr	Long chain fatty acid oxidation disorders, hyperlipoproteinemia type 1, chylothorax, intestinal lymphangiectasia, intractable malabsorption with steatorrhea, post-operative feeding in short bowel syndrome, other lipid and lymphatic disorders where a low fat, high MCT diet is indicated. Low in long-chain fats and high in medium chain triglycerides (MCT). Unflavored powder.	Nutricia
Errors of Carbohydrate Metabolism			
Glycosade	5 yrs & older	Glycogen Storage Disease where a long acting starch is indicated. Slow release corn starch carbohydrate source. Unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
3232 A	infants & children	Disaccharidase deficiency or other disorders of carbohydrate metabolism. Contains protein hydrolysate formula base that is to be used with added carbohydrate. Contains milk products, but formulated to be hypoallergenic. Unflavored powder for supplemental feeding.	Mead Johnson (Enfamil US)
Low Calcium Diet (Williams Syndrome)			

Name	Ages	Use	Manufacturer
Calcilo XD	Infants	Hypercalcemia including Williams syndrome. Low-calcium, vitamin D-free, lactose-free. 20 kcal/oz. Unflavored powder.	Abbott
Modular Diet (Supplemental Components)			
Duocal	all ages	When a high energy, low fluid, low electrolytes diet is indicated, including protein restricted diets, disorders of protein and amino acid metabolism, malabsorptive states, modular diets, and catabolic states (e.g., burns, trauma, post-operative stress). A high-calorie, protein-free nutritional supplement to add to foods and beverages including infant formula. Not a complete formula. Unflavored powder.	Nutricia
Essential Amino Acid Mix	1 yr and older	Dietary management of urea cycle disorders and conditions in which a nutritionally complete feed is not suitable or a modular approach is required. Contains essential amino acids. Supplemental nutrition. Unflavored powder.	Nutricia
Liquigen	all ages	Long-chain fatty acid oxidation disorders and conditions managed with ketogenic diet, chylothorax, fat malabsorption, short bowel syndrome, and other conditions where medium chain triglycerides (MCTs) are part of dietary management. Contains MCTs. Supplemental nutrition. Unflavored emulsion.	Nutricia
MCTProCal	1 yr & older	Long-chain fatty acid oxidation disorders, fat malabsorption, and other disorders requiring a high MCT, low long-chain triglyceride (LCT) diet. MCT oil in unflavored powder. Supplemental nutrition.	VitaFlo (Nestle)
Milupa LP Drink Mix	all ages	Inherited metabolic disorders, renal or liver failure requiring a low protein diet. Suitable for a wheat starch-, soy-, egg-, and peanut-free diet. Low protein, and contains fats and carbohydrates. Unflavored powder.	Nutricia
Monogen	>1 yr	Long chain fatty acid oxidation disorders, hyperlipoproteinemia type 1, chylothorax, intestinal lymphangiectasia, intractable malabsorption with steatorrhea, post-operative feeding in short bowel syndrome, other lipid and lymphatic disorders where a low fat, high MCT diet is indicated. Low in long-chain fats and high in medium chain triglycerides (MCT). Unflavored powder.	Nutricia

Name	Ages	Use	Manufacturer
Polycal	all ages	Conditions where a high energy, low fluid or modular diet is indicated, and for the dietary management of energy needs in patients with inborn errors of metabolism. Contains powdered maltodextrin and is high calorie, but protein- and fat-free. Supplemental nutrition. Unflavored powder.	Nutricia
Portagen	children & adults	Defective fat hydrolysis (decreased pancreatic lipase, decreased bile salts); defective mucosal fat absorption (decreased mucosal permeability, decreased absorptive surface); and/or defective lymphatic transport of fat (i.e., intestinal lymphatic obstruction). Supplemental nutrition. Unflavored powder.	Mead Johnson (Enfamil US)
ProViMin	All ages	abetalipoproteinemia; cholestasis; chylothorax; fatty acid oxidation defects; glutaric aciduria type II; hyperlipoproteinemia type I (fasting chylomicronemia); hypobetalipoproteinemia; lymphangiectasis, intestinal malabsorption of carbohydrate and/or fat; supplement for any patient who requires increased protein, minerals, and vitamins; X-linked adrenoleukodystrophy. Protein-based, and virtually carbohydrate- and fat-free to which the amount and type of carbohydrate and fat can be added. Unflavored powder.	Abbott
Single Amino Acids –Amino Acid Powder	1 yr & older	Dietary management when a modular approach is required. Contains single amino acids. Supplemental nutrition. Unflavored powder.	VitaFlo (Nestle)
Single Amino Acids – Pure Amino Acid Powder	all ages	Dietary management when a modular approach is required. Contains essential amino acids. Supplemental nutrition. Unflavored powder.	Nutricia

- **Source:** Information compiled from manufacturer websites ([Mead Johnson/Enfamil](#), [Nutricia](#), and [Abbott](#)).