

Web Table I Formulas for Carbohydrate Metabolism Disorders

<i>Metabolic disease and dietary intervention</i>	<i>Products and age recommended</i>	<i>Company</i>	<i>Remarks (Nutrients/100 g)</i>
Galactosemia diet - Galactose free	Galactomin 17	Nutricia	514 kcal, Protein Eq: 10.3g, Fat: 27.3g, CHO: 57.1g.
	Milupa basic; Infants/toddlers	Nutricia	645 kcal, Protein: 28.3g, Fat: 58g, CHO: <0.1g
	Metanutrition GLC Infants/Children	Pristine Organics	528 kcal, Protein Eq: 14.4g, Fat: 30g, CHO: 50g
	Preigestimil; Infants	Mead Johnson	500 kcal, Protein: 14g, Fat: 28g, CHO: 51g. MCT 55%, Lactose free Extensively hydrolysed casein.
Other carbohydrate metabolism disorders diet - Free/Low carbohydrate	Nutramigen LGG; Infants	Mead Johnson	Same as preigestimil, but fat: 26g, CHO: 55 g.
	Milupa basic-ch; Infants/toddlers	Nutricia	Details under galactosemia. Also suitable for Ketogenic diet
	RCF (Ross Carbohydrate free); Infants	Abbott Healthcare	Per100mL. 81 kcal, Protein: 4g, Fat:7.2g,CHO:0.07g, Soy base, Gluten free. Ketogenic diet.
	Metanutrition CMD; Infants/Children	Pristine Organics	538 kcal, Protein Eq: 22g, Fat: 50g, No CHO
	Metanutrition GTD (Glucose transport defect); Infants/Children	Pristine Organics	720 kcal, Protein Eq: 14.4g, Fat: 70g, CHO: 8g. Ideal for Glucose transport defect (Glut 1 def)

*** All lactose free soy formulas can be used in galactosemia; **Other CMD disorders include Sucrase/ Isomaltase deficiency, Fructosemia, Glucose transport defect (Glut 1 def), Glucose-Galactose malabsorption etc. Formula for these should be individualized and chosen in consultation with a specialist; **All the above free/low carbohydrate diet contains essential/non-essential amino acids, fats, vitamins and minerals.*

Web Table II FORMULAS FOR PROTEIN/AMINO ACID METABOLISM DISORDERS

Metabolic disease/ Dietary intervention	Products/ Age recommended	Company	Remarks (Nutrients/100 g)
Urea cycle disorders (UCD)/ Diet: Low in Non-essential amino acids. High in essential AA	Milupa UCD- 1 (Mixt) Anamix; Infant UCD (Form); below 1 y	Nutricia	Milupa UCD 1: 280 kcal, Protein Eq: 50 g, Fat: 0 g, CHO: 19.9 g (After 1 y: Milupa UCD 2 - Prima Mixt)
	Cyclinex-1; Infants/toddlers	Abbott	Cyclinex 1 (Per 100 ml): 77 kcal Protein : 1 g, Fat: 4 g, CHO: 9 g (Children/adults: Cyclinex -2)
	Metanutrition UCD-1; below 3 y	Pristine Organics	483 kcal, Protein Eq: 12.5 g, Fat: 25 g, CHO: 52 g (>3 y: Metanutrition UCD-2)
	UCD Trio; above 1 y	Nestle (Vitafo)	393 kcal, Protein Eq: 15 g, Fat: 14.3 g, CHO: 51 g.
Phenylketonuria (PKU)/ Diet – Phenylalanine free	Milupa PKU-1 (Mixture) Anamix Infant PKU (Form) Below 1 y	Nutricia Intl	Milupa PKU-1: 302 kcal, Protein: 50 g, Fat: 0 g, CHO: 25.6 g (After 1 y: Milupa PKU 2 - Prima Mixt)
	Phenex 1; Infants/toddlers	Abbott Healthcare	Phenex 1 (Per 100 mL): 72 kcal, Protein Eq: 2 g, Fat: 3 g, CHO: 8 g. (After 3 y: Phenex 2)
	Phenyl-Free-1; For infants	Mead Johnson	500 kcal, Protein: 16.2 g, Fat: 26 g, CHO: 51 g. Iron fortified.
	Metanutrition PKU-1; below 3 y	Pristine Organics	483 kcal, Protein: 12.5 g, Fat: 25 g, CHO: 52 g (Above 3 y: Metanutrition PKU-2/3)
	PKU Gel; 6 mo to 10 y	Nestle (Vitafo)	339 kcal, Protein Eq: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 405 kcal, Protein: 30 g, Fat: 14.3 g, CHO: 38.9 g. Contains milk and soya. 297 kcal, Protein: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya.
Tyrosinemia (TYR) Diet – Phenylalanine and Tyrosine free	Milupa TYR-1 (Mixture) Anamix Infant TYR (Form); Below 1 y	Nutricia	Milupa TYR -1: 302 kcal, Protein: 50 g, Fat: 0 g, CHO: 25.6 g (Above 1 y: Milupa TYR 2 - Prima Mixt)
	Tyrex- 1; Infants/toddlers	Abbott	Tyrex 1 (Per 100 mL): 72 kcal, Protein: 2.25g, Fat: 3.25g, CHO: 7.95g, has L-carnitine and taurine (Children/adults: Tyrex-2)
	Tyros 1; for infants Mead Johnson		500 kcal, Protein: 16.7g, Fat: 26g, CHO: 51g; Iron fortified
	Metanutrition Tyros-1; below 3 y	Pristine Organics	483 kcal, Protein: 12.5g, Fat: 25g, CHO: 52g (Above 3 y: Metanutrition Tyros-2)
	TYR Gel; 6 mo to 10 y TYR Express; above 3 y	Nestle (Vitafo)	TYR gel : 339 kcal, Protein: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 297 kcal, Protein: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya.

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Web Table II continued

Metabolic disease/ Dietary intervention	Products/ Age recommended	Company	Remarks (Nutrients/100 gm)
Homocystinuria (HCU)/ Diet – Methionine free	Milupa HOM-1 Mixt Anamix infant HCU Form; below 1 y Hominex-1; below 3 y	Nutricia Abbott Healthcare	Milupa HOM-1: 302 kcal, Protein: 50 g, Fat: 0 g, CHO: 25.6 g (Above 1 y: Milupa HOM 2-prima Mixt) Hominex-1 (Per 100 mL): 72 kcal, Protein: 2 g, Fat: 3 g, CHO: 8 g, (Above 3 y: Hominex-2)
	Metanutrition HCY-1; Below 3 y	Pristine Organics	483 kcal, Protein Eq: 12.5g, Fat: 25 g, CHO: 52 g (Above 3 y: Metanutrition HCY 2)
	HCU Gel; Above 3 y 6 mo to 10 y: HCU Express	Nestle (Vitafo)	339 kcal, Protein Eq: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 297 kcal, Protein: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya.
Maple syrup urine disease (MSUD)/ Diet – Leucine, Isoleucine and Valine free	Milupa MSUD-1 Mixt Anamix Infant MSUD Form; below 1 y Ketonex-1 Below 3 y	Nutricia Abbott	Milupa MSUD-1: 302 kcal, Protein: 50 g, Fat: 0 g, CHO: 25.6 g (Above 1 y: Milupa MSUD-2 prima Mixt) Ketonex-1 (Per 100ml): 72 kcal, Protein: 2 g, Fat: 3 g, CHO: 8 g (above 3 y: Ketonex-2)
	BCAD-1, below 1 y Metanutrition MSUD-1 below 3 y	Mead Johnson Pristine Organics	500 kcal, Protein: 16.2 g, Fat: 26 g, CHO: 51 g. Iron fortified 483 kcal, Protein: 12.5 g, Fat: 25 g, CHO: 52 g (above 3 y Metanutrition MSUD-2)
	MSUD Gel; 6 mo to 10 yrs MSUD Express; above 3 y	Nestle (Vitafo)	339 kcal, Protein: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 297 kcal, Protein Eq: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya.
Methylmalonic acidemia and Propionic acidemia (MMA/PA)/ Diet – Methionine and Valine - Free and Isoleucine and Threonine -Low/free	Milupa OS-1 Mixt Anamix Infant MMA/PPA; below 1 y OA 1; below 1 y Metanutrition MMA/PA-1; below 3 y	Nutricia Mead Johnson Pristine Organics	Milupa OS-1: 286 kcal, Protein Eq: 50 g, Fat: 0 g, CHO: 21.5 g (above 1 y: Milupa OS 2 - Prima Mixt)
	MMA/PA Gel; 6 mo to 10 y MMA/PAExpress above 3 y	Nestle (Vitafo)	500 kcal, Protein: 15.7 g, Fat: 26 g, CHO: 51 g 483 kcal, Protein: 12.5 g, Fat: 25 g, CHO: 52 g (above 3 y: Metanutrition MMA/PA-2)
Isovaleric acidemia (IVA)/ Diet – Leucine free	Milupa LEU-1 Mixt. Anamix infant IVA Form; below 1 y I-Valex-1; below 3 y	Nutricia Abbott	339 kcal, Protein Eq: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 297 kcal, Protein Eq: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya
	Metanutrition IVA-1; below 3 y	Pristine Organics	Milupa LEU-1: 286 kcal, Protein : 50 g, Fat: 0 g, CHO: 21.5 g (Above 1 y: Milupa LEU 2-prima Mixt) I-Valex-1 (Per 100 mL): 72 kcal, Protein: 2 g, Fat: 3 g, CHO: 8 g, (above 3 y: I-Valex-2)
			483 kcal, Protein: 12.5 g, Fat: 25 g, CHO: 52 g (Above 3 y: Metanutrition IVA-2)

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Web Table II continued

<i>Metabolic disease/ Dietary intervention</i>	<i>Products/ Age recommended</i>	<i>Company</i>	<i>Remarks (Nutrients/100 gm)</i>
Glutaric acidemia Type I (GA I)	Milupa GA-1 Mixt Anamix infant GA I Form; below 1 y	Nutricia	Milupa GA-1: 290 kcal, Protein: 50 g, Fat: 0 g, CHO: 22.4 g (above 1 y; Milupa GA2-prima Mixt)
Diet – Lysine free and Tryptophan - Low/free	Glutarex-1; below 3 y Metanutrition GA-1; below 3 y	Abbott Healthcare Pristine Organics	Glutarex (Per 100 mL): 72 kcal, Protein: 2 g, Fat: 3 g, CHO: 8 g, (above 3 y; Glutarex-2) 483 kcal Protein: 12.5 g, Fat: 25g, CHO: 52 g. (Above 3 y; Metanutrition GA-2)
Hyperlysinemia/ Diet – Lysine free	GA 1 Gel6; mo to 10 y GA 1 Express; above 3 y Milupa LYS-1 Mixt; below 1 y	Nestle (Vitaflo) Nutricia	339 kcal, Protein: 41.7 g, Fat: 0.05 g, CHO: 42.9 g 297 kcal, Protein: 60 g, Fat: 0.2 g, CHO: 13.7 g. Contains soya 290 kcal, Protein : 50g, Fat: 0 g, CHO: 22.4 g May use in pyridoxine dependent epilepsy in infants (Above 1 y: Milupa LYS 2-prima Mixt)
Non-ketotic hyper glycinemia (Glycine encephalopathy);/ Diet – Protein free, dietary intervention limited role	Milupa Basic – p; Infants Metanutrition HLP; Any age Pro-Phree; Infants/toddlers	Nutricia Pristine Organics Abbott Healthcare	536 kcal Protein Eq: 0g, Fat: 32 g, CHO: 62 g 535 kcal, Protein: 0 g, Fat: 25 g, CHO: 70 g Per 100 mL: 77 kcal, Protein Eq: 0 g, Fat: 4 g, CHO: 10 g.
*Protein and Aminoacid free formulas	Milupa Basic-p; infants Pro-Phree; infants/toddlers PFD; toddler young children Metanutrition AAMD-1; below 3 y	Nutricia Abbott Healthcare Mead Johnson Pristine Organics	536 kcal Protein Eq: 0 g Fat: 32 g, CHO: 62 g Per 100 mL: 77 kcal, Protein: 0 g, Fat: 4 g, CHO: 10 g. 530 kcal, Protein: 0 g, Fat: 32 g, CHO: 60 g. Metanutrition AAMD-1: 385 kcal, Protein: 0 g, Fat: 5 g, CHO: 85 g (above 3 y Metanutrition AAMD-2)

*CHO: Carbohydrate; AA: Aminoacids; Gel (Vitaflo, Nestle) - Concentrated powdered protein which when mixed up with water is easily made to a smooth, semisolid consistency. Available as pre-measured sachet, 10 gm Protein Eq per sachet. Suitable from 6 months to 10 years of age. Available for PKU, TYR, MSUD, HCU, MMA/PA and GA I; Trio (Vitaflo, Nestle) – Powdered protein substitute. Contains milk and soya. Suitable from 1 year of age. Available for UCD and PKU; Express (Vitaflo, Nestle) – Powdered protein substitute. Available as pre-weighed sachets, 15 gm protein equivalent. Contains soya. Suitable from 3 years of age. Available for PKU, TYR, MSUD, HCU, MMA/PA and GA I; Gel, Trio and Express contain essential and non-essential aminoacids (but excluding the offending aminoacids), carbohydrate, vitamins, minerals and trace element; Mixtures (Mixt) should be taken mixed with calculated amount of food or drink. Formula (Form) can be used as a supplementary feed upto 3 yrs; *Recommended as emergency regimen in sick patients with suspected Amino acidemias, Organic acidurias, Urea cycle disorders.*

Web Table III Formulas for Lipid Metabolism Disorders

<i>Diseases/diet intervention</i>	<i>Products</i>	<i>Company</i>	<i>Remarks (Nutrients/100 g)</i>
Lipid Metabolism disorders (LMD)/ diet: Low fat	Milupa Basic-f; Infants/toddlers	Nutricia	374 kcal, Protein: 14 g, Fat: <0.5 g, CHO: 79 g.
	Monogen; Infants/ children	Nutricia	420 kcal, Protein: 12.5 g (Whey based), Fat: 11 g, (MCT 84%), CHO: 68 g.
	ProViMin; Infants/ children	Abbott	Per 100 mL: 62.6 kcal, Protein: 14.6 g (Casein based), Fat: 0.3 g, CHO: 0.4 g.
	Metanutrition LD (Lipid disorders); Infants/children	Pristine Organics	462 kcal, Protein Eq: 12.5 g, Fat: 20 g, CHO: 58 g. MCT 80%.
	Metanutrition LCHAD (Long chain hydroxyacyl-CoA dehydrogenase); Infants/children	Pristine Organics	520 kcal, Protein: 12.5 g, Fat: 30 g, CHO: 50 g. Long chain triglycerides (LCT) free. Indicated in LCHAD deficiency

*** LMD include Fatty acid oxidation disorder, Severe cholestais, Intestinal lymphangiectasia, Abeta/Hypobetalipoproteinemia, Chylothorax, Malabsorption and Maldigestion of fats. Formula for these should be individualized and chosen in consultation with a specialist.*