PRISTINE

Nutrition for Inborn Errors of Metabolism

Inborn errors of metabolism are rare genetic (inherited) disorders in which the body cannot properly turn food into energy. The disorders are usually caused by defects in specific proteins (enzymes) that help break down (metabolize) parts of food.

A food product that is not broken down into energy can build up in the body and cause a wide range of symptoms. Several inborn errors of metabolism cause developmental delays or other medical problems if they are not controlled.



S.N.	IEM	Description	Age group	Protein (%)	Product Name
		Amino	acidopathies		
	Phenyl- ketonuria	Phenylalanine free dief	Infant & toddlers	15	Metanutrition PKU 1
1			Children & adults	60	Metanutrition PKU 2
			Children & adults	85	Metanutrition PKU 3
/	Maple Syrup Urine Disorder	Jrine Isoleucine, Leucine and Valine free diet	Infant & toddlers	15	Metanutrition MSUD 1
2			Children & adults	48	Metanutrition MSUD 2
3	Tyrosin <mark>emi</mark> a	Tyrosine and Phenylalanine free diet	Infant & toddlers	15	Metanutrition TYROS 1
			Children & adults	48	Metanutrition TYROS 2
4	Homo- cystinuria	Methionine free diet	Infant & toddlers	15	Metanutrition HCY 1
			Children & adults	48	Metanutrition HCY 2

S.N.	IEM	Description	Age group	Protein (%)	Product Name
		Oı	rg <mark>ani</mark> c Acidemias		
	Isovaleric Acidemia, 3-Methylcrotonyl-CoA		Infant & toddlers	20	Metanutrition ISVA 1
5	Carboxylase deficiency, 3-Methyl glutaconyl-CoA Hydratase deficiency	Leucine free diet	Children & adults	48	Metanutrition ISVA 2
6	Methylmalonic Acidemia /Propionic Acide <mark>mia</mark>	mia /Propionic Methionine, Threonine and Valine	Infant & toddlers	15	Metanutrition MMA/PA 1
			Children & adults	48	Metanutrition MMA/PA 2
7	Glutaric Acidemia	Lysine and Tryptophan free diet	Inf <mark>ant &</mark> toddlers	15	Metanutrition GA 1
	Type I		Children & adults	48	Metnautrition GA 2

S.N.	IEM	Description	Age group	Protein (%)	Product Name	
	Urea Cycle Disorders					
	Argininemia, Arginino <mark>succini</mark> c		Infant & toddlers	15	Metanutrition UCD 1	
8	Aciduria, Carbamoylphosphate Synthetase Deficiency, Citrullinemia	Non essential amino acid free diet	Children & adults	48	Metanutrition UCD 2	
/			Lipid Disorders			
9	3-Hydroxy Long chain Acyl-CoA dehydrogenase deficiency	LCHAD deficiency	Infant & toddlers	15	Metanutrition LCHAD	
10	Defects in the intraluminal hydrolysis of fat: defective mucosal fat absorption, defective lymphatic transport of fat	Milk protein based powder with medium chain triglycerides (MCT) for children and adults	Infant & toddlers	15	Metanutrition LD	

SN	IEM	Description	Age group	Protein (%)	Product Name
		Carbohydra	te Metabolism Disorders		
11	Galactosemia	Galactose free formula	Infant & toddlers	15	Metanutrition GLC
12	Disorders of carbohydrate metabolism, sucrase/isomaltase deficiency, Dissacharidase deficiencies, Fructose Intolerance	Protein hydrolysate formula base powder with iron for use with added carbohydrate.	Infants & toddlers	30	Metanutrition CMD
13	Glucose transport defect	Low carbohydrate, sucrose, fructose, sugar free formula	Infants & toddlers	15	Metanutrition GTD

S.N.	IEM	Description	Age group	Protein (%)	Product Name
		Ger	neral Formulation		
1	Amino acid metabolic disorders	Amino acid and protein free diet powder	Infant & toddlers	Nil	Metanutrition AAMD 1
1			Children & adults	Nil	Metanutrition AAMD 2
2	Low Protein Supplement	Amino acid free diet	Infant & toddlers	3	Metanutrition LPS
3	Severe allergy due to multiple food protein i <mark>nto</mark> lerance	Amino acid based hypoallergenic formula	Infant & toddlers	15	Metanutrition MFPI
4	Severe allergy due to intact milk protein and soy protein	For cow's milk allergy hypoallergenic formula	Infant & toddlers	15	Metanutrition SA MP/SP



Nutrition for Inborn Errors of Metabolism

with Individual Ingredients

S.N	Individual Ingredient
1	Betaine
2	Carnitine
3	Sodium Benzoate
4	Isoleucine
5	Valine
6	Arginine
7	Citrulline
8	L-Methionine
9	Sialic Acid



Organic Authenticity & Standards



Certification from NPOP
(National standards for organic products and processes)



ISO

(International Organization For Standardization)
A standard that ensures that the product is safe,
reliable and of good quality.



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Control Union Certifications: 803369



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