Dietary Interventions for Rare Metabolic Disorders – Now Available in India!

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The human body is a miracle machine, and metabolism is the most critical process that converts food to energy which in turn serves as fuel to stay healthy and fight diseases. When a baby is born with a rare metabolic disorder, this essential process of digestion is hampered, leading to multiple health complications, and sometimes even death. As pediatricians, our role is to ensure infants and children live healthy and happy. While we focus on larger health issues like malnutrition and immunization in the public health narrative, we miss out on a small segment – babies who are diagnosed with rare metabolic disorders.

Nearly 74000 babies are born every day in India, of which approximately 74 are born with Inborn Errors of Metabolism (IEM). This translates to more than 27000 infants annually who need specialized nutrition and medical attention. While the individual incidence is small, collectively they are a significant contributor to the infant mortality burden. In the absence of the requisite nutrition or delays caused by imports of special medical foods, the affected children are at risk for mental retardation, epilepsy, and severe deficiencies, even leading to death.

Many of the IEMs and hypoallergenic conditions require specialty foods, which are not readily available in India. Until recently, if a child was diagnosed with IEM, parents had to either get these shipped through acquaintances in the West or wait till their doctor got them directly from nutrition companies. Both solutions were not viable for all, and infants continued to suffer.

The Indian Academy of Pediatrics was in discussion with various industry partners and government bodies for the last year, and the efforts culminated in bringing IEM nutrition products to India. Last year in November, FSSAI in a landmark recommendation, allowed the import of foods for "Inborn errors of metabolism" and hypoallergenic conditions. Companies such as Abbott, Dannon, Mead Johnson and Nestle came forward through their global supply chain systems to expedite availability of nutrition products that could potentially save many children every year. An indicative list is provided in ***Table* I.**

IAP and our member pediatricians are relentlessly working towards enhancing the knowledge and information and availability of these products needed by healthcare professionals who provide these services to patients with IEM and allergy issues. These are severe nutritional emergencies and prompt initiation of appropriate therapy is still the best determinant of outcome for these patients.

We are glad to be part of the collaboration that is bringing IEM products into India to address the critical needs of babies with IEM. Rare metabolic disorders need immediate medical and dietary interventions. As neonatal screening is picking up in hospitals, we are confident that we will be able to monitor and track IEM incidence and help reduce acute illness and fatalities. It gives me great satisfaction as a pediatrician that together as a country, we have bridged the gap between access and demand for IEM portfolio and made a difference to a segment that's small but important to improving health outcomes for infants.

TABLE I INDICATIVE LIST OF DIETARY PRODUCTS AVAILABLE FOR MANAGING INBORN ERRORS OF METABOLISM

Medical condition	*Description*	*Products**
Maple syrup urine disease	Isoleucine, Leucine and Valine free diet powder	Ketonex-1, Ketonex-2MSUD 1-3
Glutaric Acidemia Type I	Lysine and Tryptophan free diet powder	Glutarex-1, Glutarex-2GA
Homocystinuria	Methionine free diet powder	Hominex-1, Hominex-2HCY 1-2, Hom 1-3
Isovaleric Acidemia, 3-MethylcrotonyI-CoA, Carboxylase Deficiency, 3-Methylglutacony I-CoA, Hydratase Deficiency	Leucine free diet powder	I-Valex-1, I-Valex-2LMD, IVA cooler
Methylmalonic Acidemias, Propionic Acidemia	Isoleucine, Methionine, Threonine and Valine free diet powder	Propimex-1, Propimex-2OA 1-2
Amino acid metabolic disorders	Protein and amino acid free diet powder (with and without fat)	Pro-Phree, PFD
Phenylketonuria (PKU)	Phenylalanine free diet powder	Phenex-1, Phenyl free 1-2 Phenex-2, PKU 1-3
Urea Cycle Disorders, Argininemia, Argininosuccinic Aciduria, Carbamoyl phosphate Synthetase Deficiency1, Citrullinemia	Non-essential amino acid free diet powder	Cyclinex-1, Cyclinex-2UCD 1-2
Tyrosinemia	Phenylalanine and Tyrosine free diet powder	Tyrex-1, Tyrex-2 Tyros 1-2
Galactosemia,	Galactose free formula	Ele Care, Ele Care Junior
3-Hydroxy Long Chain Acyl-CoA Dehydrogenase Deficiency	LCHAD Deficiency	Pro ViMin
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	
Disaccharidase deficiencies; Disorders of carbohydrate metabolism, Sucrase/isomaltase deficiency, Fructose intolerance	Protein hydrolysate formula base powder with iron for use with added carbohydrate.	RCF
Non-ketotic Hyperglycinemia, Lysinuric protein intolerance	Protein free formula	Pro-Phree
Glucose transport defect (Glut1 def), Pyruvate dehydrogenase complex deficiency	Low carbohydrate, sucrose, fructose, sugar free formula	RCF

**Product names are the registered trade names of the respective manufacturers.*