

A woman with long dark hair, wearing a light blue shirt, is sitting at a table and looking towards a young girl. The girl has short dark hair with bangs and is wearing a pink and white ruffled dress. She is holding a blue and yellow toy globe and appears to be speaking or laughing. On the table, there are various toys including a pink toy house and some colorful blocks. The background is softly blurred, showing a window with light-colored curtains.

Nutricia of Metabolic Milks

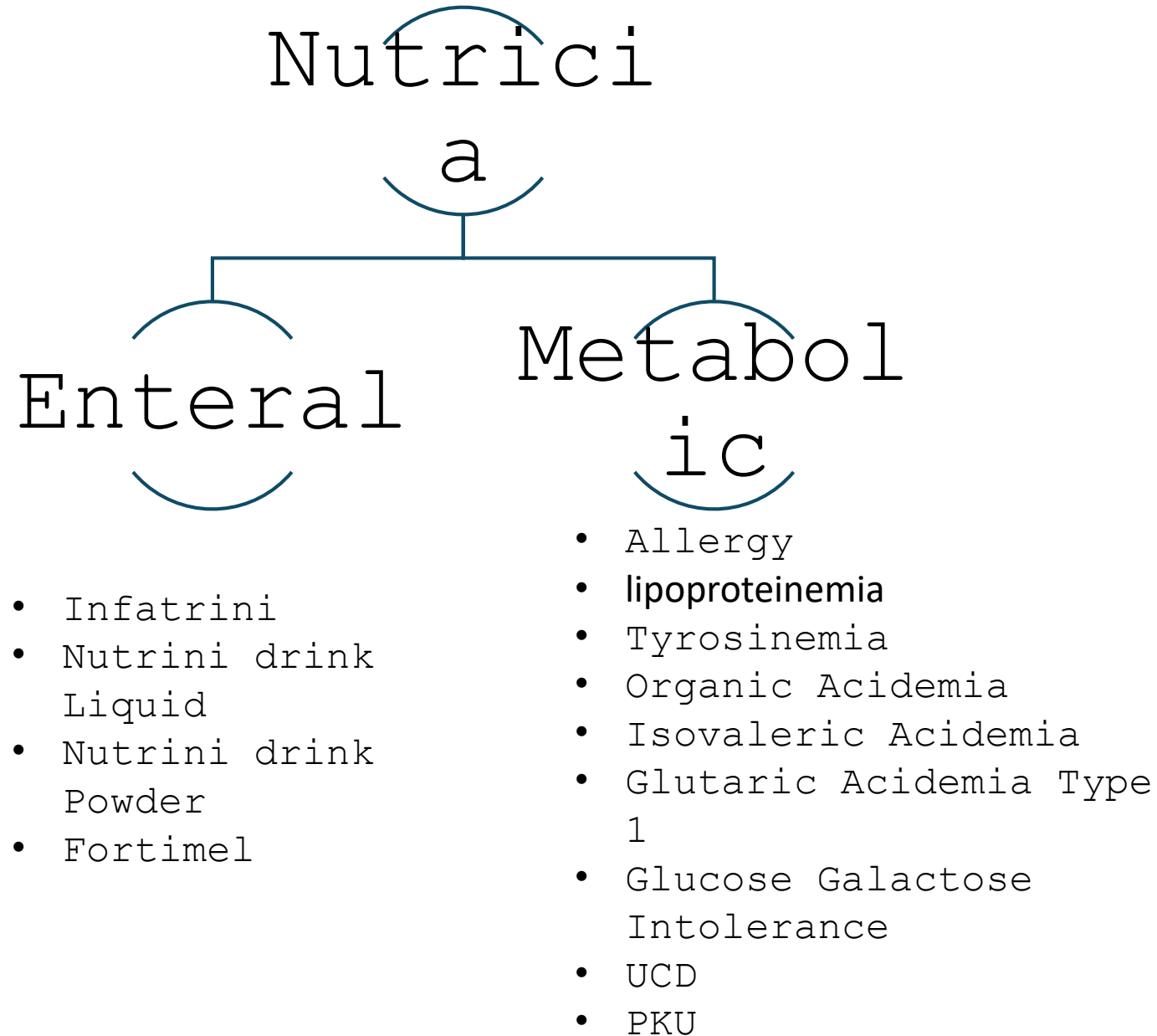
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First Line Manager

Nutricia Company

- Global Leader in specialized medical nutrition
- Evidence-based nutrition
- Pediatric



Nutricia current portfolio Market in Iran



Metabolic Products



Allergy

Allergy is an adverse **overreaction by the immune system** to a harmless substance (allergens) .

A **food allergy** occurs when specific components of a food (**typically proteins**) are recognized by immune system. Your immune system produces antibodies that detect the allergen, causing inflammatory reactions and the release of a chemical called **histamine**, both of which cause **allergic symptoms**.



Cow's milk protein allergy (CMPA)

- the abnormal response to proteins found in cow's milk or products containing milk proteins.
- the trigger molecules are two milk proteins called **whey** and **casein**. You or your child may be allergic to either one of these proteins, or both.
- The symptoms typically develop from **two hours** after consumption but can take **up to 72 hours**.

Symptoms of cow's milk allergy

- ✓ raised red bumps of skin - **hives**
- ✓ itchy, red, rash of the skin - **dermatitis** or **eczema**
- ✓ swelling of the face
- ✓ abdominal pain-**colic**
- ✓ blood in the stools
- ✓ vomiting
- ✓ diarrhoea



Monogen

- A complete nutrition formula
- low in long chain triglycerides (LCT=16%) and high in medium chain triglycerides (MCT=84%)
- linoleic acid (LA) and alphalinolenic acid (ALA), and supplemented with docosaheaxaenoic acid (DHA) and arachidonic acid (AA)
- Whey protein
- Lactose free
- as a sole source of nutrition or as a supplementary feed for children and adults



Indication

- ✓ Hyperlipoproteinaemia type 1
- ✓ Long chain fatty acid oxidation disorders (LCFAODs)
- ✓ Chylous ascites
- ✓ Chylothorax
- ✓ Fat malabsorption
- ✓ Cystic Fibrosis

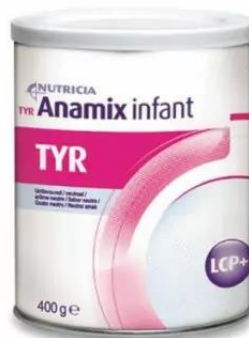


Tyrosinemia

- Tyrosinemia is an error of metabolism, usually inborn, in which the body cannot effectively break down the amino acid **tyrosine**.
- Tyrosinemia is inherited in an autosomal recessive both parents carried defective gene
- 3 type of tyrosinemias
 - Type I tyrosinemia
 - Type II tyrosinemia
 - Type III tyrosinemia
- All 3 forms result from dysfunction of various genes in the phenylalanine and tyrosine catabolic pathway



- Diagnosed as a result of newborn screening
- Symptoms
 - ☐ failure to gain weight and grow at the expected rate (failure to thrive),
 - ☐ fever
 - ☐ diarrhea
 - ☐ vomiting
 - ☐ abnormally enlarged liver (hepatomegaly), and yellowing of the skin and the whites of the eyes
- Elevated plasma Tyrosine Phenylalanine and Methionine levels



Organic Acidaemia / MMA / PA

- Methylmalonic Acidaemia (MMA) and Propionic Acidaemia (PA) are disorders of propionate metabolism.
- The degree of severity varies and clinical symptoms ranges from mild neurological impairment through to coma and death.
- The aim of dietary management is to limit production of propionate by restricting methionine, threonine, valine and isoleucine by means of a low protein diet.
- The protein requirements are met by supplementing the diet with methionine, threonine, valine and isoleucine free amino acid mixture if necessary



Isovaleric Acidemia

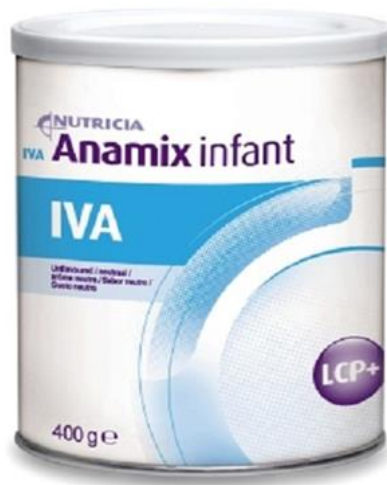
- ✓ autosomal recessive
- ✓ normal metabolism of the branched-chain amino acid
leucine is disrupted
- ✓ the deficiency of enzyme that is responsible for the
third step in processing leucine (essential amino acid)
symptoms
 - poor feeding
 - vomiting
 - Seizures
 - lack of energy that can progress to coma



Treatment

Dietary protein restriction, particularly leucine.

During acute episodes, **glycine** is sometimes given, which conjugates with isovalerate forming isovalerylglycine, or **carnitine** which has a similar effect



Glutaric Aciduria type I

- Glutaric Aciduria Type I (GAI) is a rare inherited disorder caused by a deficiency of glutaryl-CoA dehydrogenase
- resulting in progressive neurological deterioration
- The aim of dietary management is to limit lysine and tryptophan by means of a low protein diet.
- The protein requirements are met by supplementing the diet with lysine free and tryptophan low amino acid mixture if necessary.



Glucose Galactose Intolerance

- Glucose-galactose malabsorption is a rare and autosomal recessive condition in which the cells lining the intestine **cannot take** in the sugars **glucose and galactose**
- protein is found mainly in the **intestinal tract** and, to a lesser extent, in the **kidneys**, where it is involved in transporting glucose and the structurally similar galactose across cell membranes
- the **sodium/glucose cotransporter protein** is involved in the process of glucose uptake in the intestinal cells due to a sodium gradient across the membrane



- Mutations that prevent the sodium/glucose cotransporter protein result in a buildup of glucose and galactose in the intestinal tract.
- This failure of active transport prevents the glucose and galactose from being absorbed
- In addition, the water that normally would have been transported across the brush border with the sugar instead remains in the intestinal tract to be expelled with the stool, resulting in **dehydration of the body's tissues and severe diarrhea** resulting in life-threatening **dehydration** increased **acidity of the blood and tissues (acidosis)**, **weight loss** and **kidney stones**

Treatment

Low galactose, glucose diet



THANK YOU

