

Types of Special Formulas for Metabolic Inborn Disorders



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Objectives

- **Metabolic Inborn Disorders**
- **Since the 1950s:**
 - PKU could be treated by diet restricting**
- **In many of these diseases diet is the primary and often the only treatment**

Amino Acid Disorders

- **Phenylketonuria (PKU)**
- **Maple syrup urine disease (MSUD)**
- **Tyrosinemia**
- **Homocystinuria/ Hypermethioninemia**

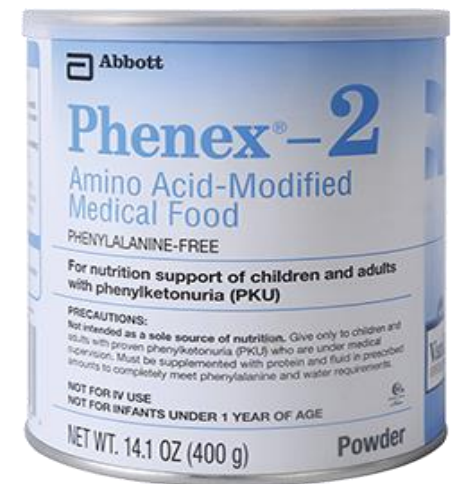
Phenylketonuria (PKU)

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Up to 3 years (As a supplementary feed)	Calories Pro Cho Fat Tyrosine	478 kcal 12 g 53 g 25 g 132 mg	EAA & non-EAA Fat (Vegetable & Coconut) Vitamins Minerals PUFA/ DHA/ ARA Lactose
- <u>Children over 1 year of age</u>	<u>Calories</u> Pro Cho Fat Tyrosine	402 kcal 27 g 42 g 14 g 239 mg	
1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water Not suitable as a sole source of nutrition Oral and Enteral use only			



Phenylketonuria (PKU)

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Infant Formula With Iron	Calories <u>Pro</u> Cho Fat <u>Iron</u>	480 kcal 15 g 53 g 22 g 9 g	EAA & non-EAA High Oleic Safflower, Coconut and Soy Oils Vitamins Minerals
- <u>Children, adolescents, and adults</u>	Calories <u>Pro</u> Cho Fat <u>Iron</u>	410 kcal 30 g 35 g 14 g 13 g	30% of TE (PUFA/DHA/ARA) <u>Gluten-free</u> <u>Lactose-free</u>
Not suitable as a sole source of nutrition Oral and Enteral use only			



Phenylketonuria (PKU)

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u>	<u>Calories</u>	507 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ARA Lactose
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
	Tyrosine	125 mg	
<u>- Children, adolescents, and adults</u>	<u>Calories</u>	424 kcal	
	Pro	31 g	
	Cho	41 g	
	Fat	15 g	
	Tyrosine	250 mg	
Not suitable as a sole source of nutrition Oral and Enteral use only COMIDA C: Up to 15 years			



Maple syrup urine disease (MSUD)

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u>	Calories	466 kcal	EAA & non-EAA Fat (Vegetable & Coconut) Vitamins Minerals PUFA/ DHA/ ARA Lactose
- Up to 3 years (As a supplementary feed)	Pro	13 g	
	Cho	50 g	
	Fat	27 g	
- <u>Children 1-8 year of age</u>	Calories	297 kcal	
	Pro	39 g	
	Cho	34 g	
	Fat	0.5 g	
1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water Not suitable as a sole source of nutrition Oral and Enteral use only Leucine, Isoleucine and Valine FREE			



Maple syrup urine disease (MSUD)

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u>	<u>Calories</u>	480 kcal	EAA & non-EAA High Oleic Safflower, Coconut and Soy Oils Vitamins Minerals 30% of TE (PUFA/DHA/ARA) <u>Gluten-free</u> <u>Lactose-free</u>
	Pro	15 g	
	Cho	53 g	
	Fat	22 g	
- <u>Children over 1 year of age and adults</u>	<u>Calories</u>	410 kcal	
	Pro	30 g	
	Cho	35 g	
	Fat	14 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Should not be fed to premature infants Leucine, Isoleucine and Valine FREE			



Maple syrup urine disease (MSUD)

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u>	<u>Calories</u> Pro Cho Fat	505 kcal 12 g 53 g 27 g	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ARA Lactose
<u>- Children, adolescents, and adults</u>	<u>Calories</u> Pro Cho Fat	418 kcal 31 g 39 g 15 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Leucine, Isoleucine and Valine FREE Sucrose Free COMIDA C: Up to 15 years			



Tyrosinemia

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u> <u>- Up to 3 years (As a supplementary feed)</u>	Calories	466 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA Lactose
	Pro	13 g	
	Cho	50 g	
	Fat	23 g	
<u>- Children 1-8 year of age</u>	Calories	309 kcal	
	Pro	25 g	
	Cho	51 g	
	Fat	0.5 g	
1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water Oral and Enteral use only Not suitable as a sole source of nutrition Tyrosine and Phenylalanine Free			



Tyrosinemia

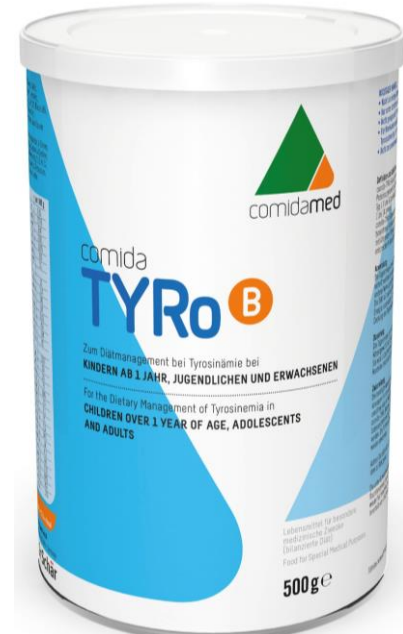
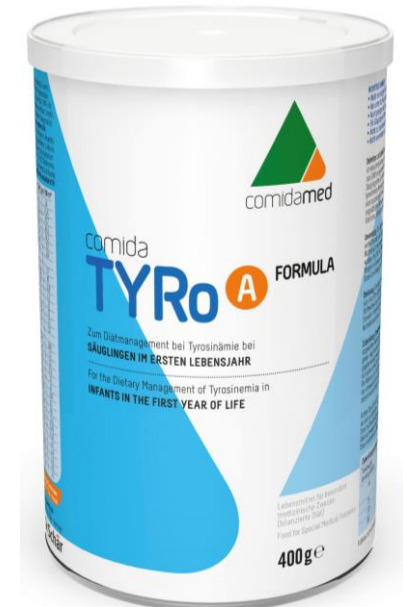
Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Infant Formula With Iron	Calories	480 kcal	EAA & non-EAA High Oleic Safflower, Coconut and Soy Oils Vitamins Minerals 30% of TE (PUFA/DHA/ARA) <u>Gluten-free</u> <u>Lactose-free</u>
	<u>Pro</u>	15 g	
	Cho	53 g	
	Fat	22 g	
	<u>Iron</u>	7.6 mg	
- <u>Children, adolescents, and adults</u>	Calories	410 kcal	
	<u>Pro</u>	30 g	
	Cho	35 g	
	Fat	14 g	
	<u>Iron</u>	13 mg	

Oral and Enteral use only
 Not suitable as a sole source of nutrition
 Should not be fed to premature infants
 Tyrosine and Phenylalanine Free



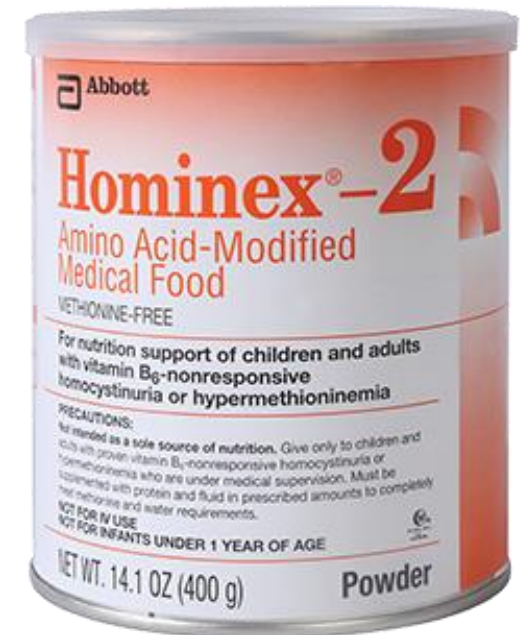
Tyrosinemia

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u>	<u>Calories</u>	506 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ ARA Lactose
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
<u>- Children, adolescents, and adults</u>	<u>Calories</u>	420 kcal	
	Pro	31 g	
	Cho	40 g	
	Fat	15 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Phenylalanine and Tyrosine FREE			



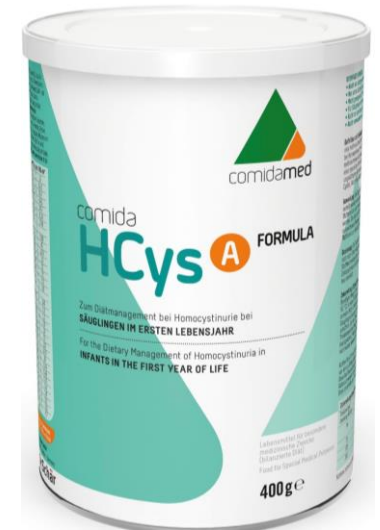
Homocystinuria/ Hypermethioninemia

Indications	100 gr		Nutrition Information
<u>Children, adolescents, and adults</u>	Calories	410 kcal	EAA & non-EAA High Oleic Safflower, Coconut and Soy Oils Vitamins Minerals 30% of TE (PUFA/DHA/ARA) Gluten-free Lactose-free
	<u>Pro</u>	30 g	
	<u>Cho</u>	35 g	
	<u>Fat</u>	14 g	
	<u>Iron</u>	13 mg	
	<u>Cystine</u>	900 mg	
Oral and Enteral use only Not suitable as a sole source of nutrition Methionine free			



Homocystinuria/ Hypermethioninemia

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u>	<u>Calories</u>	506 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ ARA Lactose
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
	<u>Cystine</u>	550 mg	
<u>- Children, adolescents, and adults</u>	<u>Calories</u>	420 kcal	
	Pro	31 g	
	Cho	40 g	
	Fat	15 g	
	<u>Cystine</u>	1500 mg	
Oral and Enteral use only Not suitable as a sole source of nutrition Methionine free			



Urea Cycle Disorder

Indications	100 gr		Nutrition Information
- <u>Up to 6 months, Adult</u>	Calories	360 kcal	<u>EAA</u> Vitamin C Electrolytes + Ca Lactose
	Pro	25 g	
	Cho	65 g	
	<u>Fat</u>	0 g	
Oral and Enteral use only Not suitable as a sole source of nutrition Chronic Renal Failure Nonessential amino acid-free			



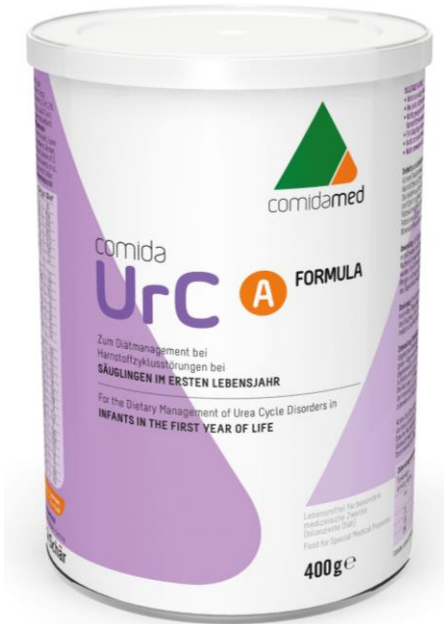
Urea Cycle Disorder

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Infant Formula With Iron	Calories	510 kcal	<u>EAA</u> 35%-43% of energy as Fat Vitamins Minerals PUFA/ MUFA/ ARA Gluten-free Lactose-free
	Pro	7.5 g	
	Cho	57 g	
	Fat	25 g	
	Iron	9 mg	
- <u>Children, adolescents, and adults</u>	Calories	440 kcal	
	Pro	15 g	
	Cho	45 g	
	Fat	17 g	
	Iron	17 mg	
Oral and Enteral use only Not suitable as a sole source of nutrition Should not be fed to premature infants Nonessential amino acid-free			



Urea Cycle Disorder

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u>	Calories	492 kcal	<u>EAA</u> Vitamins Minerals PUFA/ MUFA/ ARA Lactose
	Pro	7.5 g	
	Cho	58 g	
	Fat	25 g	
	<u>Cystine</u>	430 mg	
	<u>Tyrosine</u>	720 mg	
- <u>Children, adolescents, and adults</u>	Calories	421 kcal	
	Pro	16 g	
	Cho	47 g	
	Fat	17 g	
	<u>Cystine</u>	530 mg	
	<u>Tyrosine</u>	1800 mg	
Oral and Enteral use only Not suitable as a sole source of nutrition Nonessential amino acid-free Enriched with cystine and tyrosine			



Organic Aciduria

- **Glutaric Aciduria**
- **Propionic Aciduria**
- **Isovaleric Aciduria**
- **Methylmalonic Aciduria**

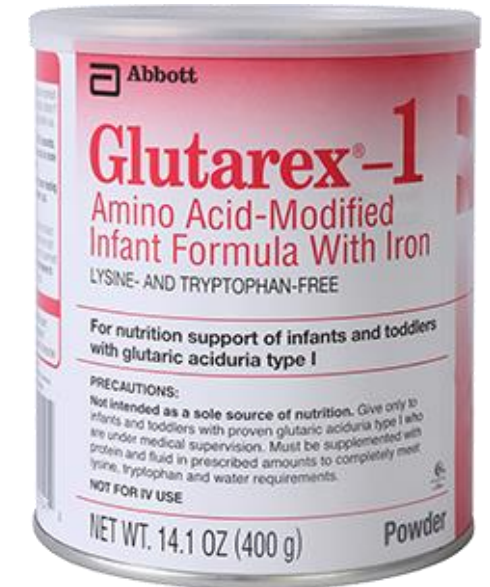
Glutaric Aciduria

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Up to 3 years (As a supplementary feed)	Calories	466 kcal	EAA & non-EAA Fat (Vegetable Oil) Vitamins Minerals PUFA/ DHA/ ARA Lactose
	Pro	13 g	
	Cho	50 g	
	Fat	23 g	
	Tryptophan	0.9 mg	
- <u>Children 1-8 year of age</u>	Calories	309 kcal	
	Pro	25 g	
	Cho	51 g	
	Fat	0.5 g	
1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water Not suitable as a sole source of nutrition Oral and Enteral use only Lysine free, Low Tryptophan			



Glutaric Aciduria

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u> Infant Formula With Iron	Calories	480 kcal	EAA & non-EAA 30%-40% of energy as Fat Vitamins Minerals DHA Gluten-free Lactose-free
	Pro	15 g	
	Cho	53 g	
	Fat	22 g	
	Iron	8 mg	
<u>- Children, adolescents, and adults</u>	Calories	410 kcal	
	Pro	30 g	
	Cho	35 g	
	Fat	13 g	
	Iron	13 mg	
Not suitable as a sole source of nutrition Oral and Enteral use only Lysine and Tryptophan Free			



Glutaric Aciduria

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u>	Calories	506 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ ARA Lactose Sucrose free
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
- <u>Children, adolescents, and adults</u>	Calories	421 kcal	
	Pro	16 g	
	Cho	40 g	
	Fat	17 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Lysine and Tryptophan FREE			



Propionic / Methylmalonic Aciduria

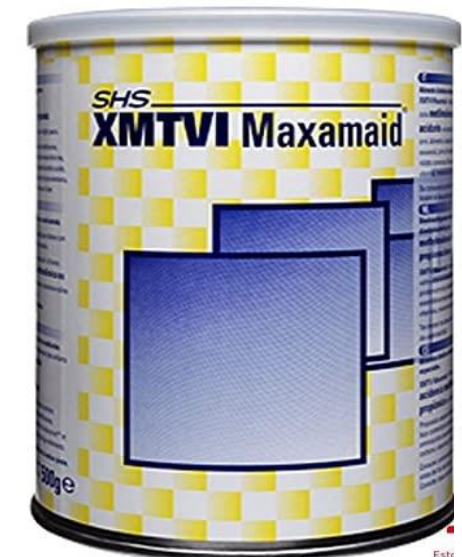
Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Up to 3 years (As a supplementary feed)	Calories	466 kcal	EAA & non-EAA Fat (Vegetable Oil) Vitamins Minerals PUFA/ DHA/ ARA Lactose
	Pro	13 g	
	Cho	50 g	
	Fat	23 g	
	Isoleucine	≥ 0.45 mg	
- <u>Children 1-8 year of age</u>	Calories	309 kcal	
	Pro	25 g	
	Cho	51 g	
	Fat	0.5 g	

1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water

Not suitable as a sole source of nutrition

Oral and Enteral use only

Methionine, Threonine, Valine Free and low Isoleucine



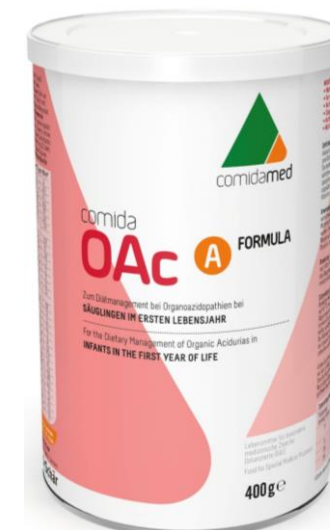
Propionic / Methylmalonic Aciduria

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> Infant Formula With Iron	Calories	480 kcal	EAA & non-EAA 30%-40% of energy as Fat Vitamins Minerals DHA <u>Gluten-free</u> <u>Lactose-free</u>
	Pro	15 g	
	Cho	53 g	
	Fat	22 g	
	Iron	8 mg	
- <u>Children, adolescents, and adults</u>	Calories	410 kcal	
	Pro	30 g	
	Cho	35 g	
	Fat	13 g	
	Iron	13 mg	
Not suitable as a sole source of nutrition Oral and Enteral use only Methionine and Valine Free Low in Isoleucine and Threonine			



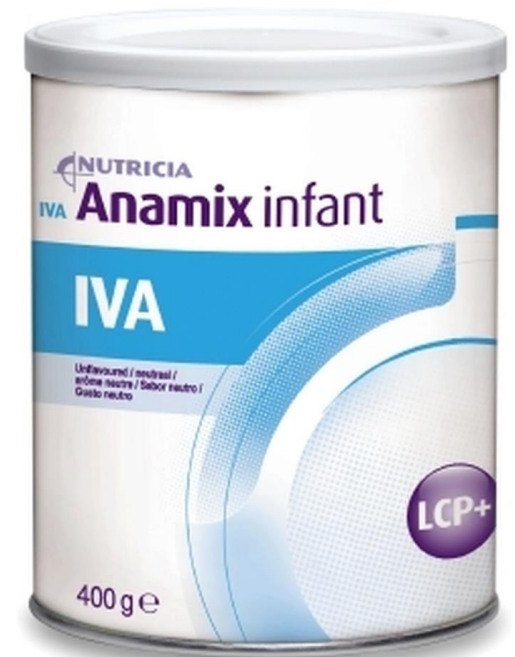
Propionic / Methylmalonic Aciduria

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u>	Calories	506 kcal	EAA & non-EAA Vitamins Minerals
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
<u>- Children, adolescents, and adults</u>	Calories	421 kcal	
	Pro	16 g	
	Cho	40 g	
	Fat	17 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Methionine, Isoleucine, Threonine and Valine Free			



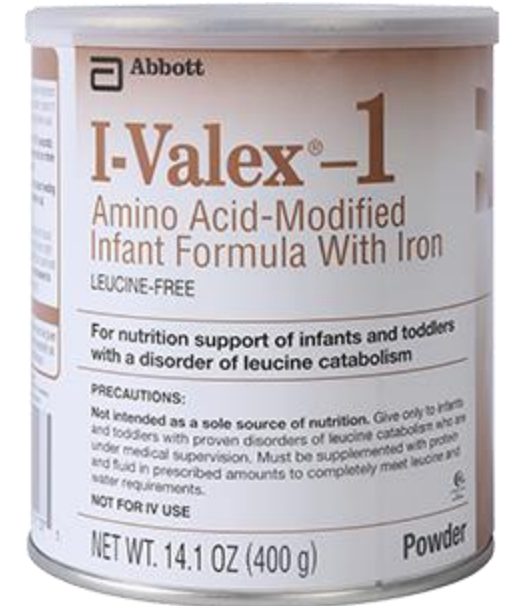
Isovaleric Aciduria

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - Up to 3 years (As a supplementary feed)	Calories	466 kcal	EAA & non-EAA Fat (Vegetable Oil) Vitamins Minerals PUFA/ DHA/ ARA Lactose
	Pro	13 g	
	Cho	50 g	
	Fat	23 g	
- <u>Children 1-8 year of age</u>	Calories	309 kcal	
	Pro	25 g	
	Cho	51 g	
	Fat	0.5 g	
1 unpacked scoop (5 g) of powder to each fluid ounce (30 ml) of water Not suitable as a sole source of nutrition Oral and Enteral use only Leucine-free			



Isovaleric Aciduria

Indications	100 gr		Nutrition Information
<u>- Birth to 12 months infants</u> Infant Formula With Iron	Calories	480 kcal	EAA & non-EAA 30%-40% of energy as Fat Vitamins Minerals DHA Gluten-free Lactose-free
	Pro	15 g	
	Cho	53 g	
	Fat	22 g	
	Iron	8 mg	
<u>- Children, adolescents, and adults</u>	Calories	410 kcal	
	Pro	30 g	
	Cho	35 g	
	Fat	13 g	
	Iron	13 mg	
Not suitable as a sole source of nutrition Oral and Enteral use only Leucine-free			



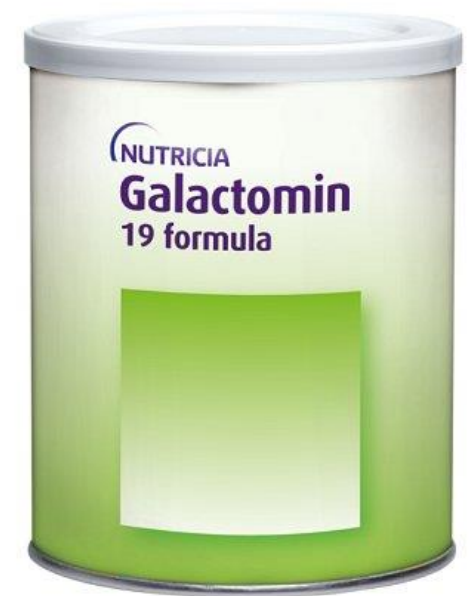
Isovaleric Aciduria

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u>	Calories	506 kcal	EAA & non-EAA Vitamins Minerals PUFA/ MUFA/ ARA Lactose Sucrose free
	Pro	12 g	
	Cho	53 g	
	Fat	27 g	
- <u>Children, adolescents, and adults</u>	Calories	421 kcal	
	Pro	16 g	
	Cho	40 g	
	Fat	17 g	
Not suitable as a sole source of nutrition Oral and Enteral use only Leucine-free			



Glucose-Galactose Malabsorption

Indications	100 gr		Nutrition Information
- <u>infants under 1 year of age</u> - <u>As a supplement for young children</u>	Calories	534 kcal	EAA & non-EAA
	Pro	15 g	Fat (Vegetable Oil)
	Cho	50 g	Vitamins
	Fat	22 g	Minerals
PUFA/ DHA/ ARA			
suitable as a sole source of nutrition			
Oral and Enteral use only			
Containing cow's milk protein, Carbohydrate (fructose)			
Minimal levels of lactose, galactose and glucose			



Glucose-Galactose Malabsorption

Indications	100 gr		Nutrition Information
- <u>Infants under 1 year of age</u> - <u>As a supplement for young children</u>	Calories	534 kcal	EAA & non-EAA
	Pro	15 g	Fat (Vegetable Oil)
	Cho	50 g	Vitamins
	Fat	22 g	Minerals
			PUFA/ DHA/ ARA
			Prebiotic FOS
			Lactose free
			Gluten free
Suitable as a sole source of nutrition Soy-based formula Sucrose and Galactose free Caw's milk protein allergy & Family history of allergy Post diarrhea feeding & recurrent diarrhea feeding Vegetarian diet			



Glucose-Galactose Malabsorption

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - <u>As a supplement for young children</u>	Calories	493 kcal	EAA & non-EAA
	Pro	14 g	Vitamins
	Cho	53 g	Minerals
	Fat	25 g	PUFA/ MUFA/ ARA
			Maltodextrin
suitable as a sole source of nutrition			
Oral and Enteral use only			
Lactose, sucrose, fructose and galactose Free			
Gluten, gliadin, milk proteins Free			



Long chain fatty acid oxidation disorders

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - <u>As a supplement for young children</u>	Calories	444 kcal	EAA & non-EAA
	Pro	13 g	Vitamins
	Cho	70 g	Minerals
	Fat	13 g	PUFA/ MUFA/ ARA LCT 16% MCT 84%
Suitable as a sole source of nutrition Oral and Enteral use only Hyperlipoproteinaemia type 1 Long chain fatty acid oxidation disorders Chylous ascites Chylothorax Fat malabsorption			



Long chain fatty acid oxidation disorders

Indications	100 gr		Nutrition Information
- <u>Birth to 12 months infants</u> - <u>As a supplement for young children</u>	Calories	466 kcal	EAA & non-EAA
	Pro	12.5 g	Vitamins
	Cho	62 g	Minerals
	Fat	17.5 g	PUFA/ MUFA/ SFA
			LCT <MCT
			Fiber
Suitable as a sole source of nutrition Oral and Enteral use only Hyperlipoproteinaemia type 1 Long chain fatty acid oxidation disorders Chylous ascites Chylothorax Fat malabsorption			



Thanks for Your Attention

