Phenylketonuria, also called PKU, is a rare inherited disorder that causes an amino acid called phenylalanine to build up in your body.

Without the enzyme necessary to process phenylalanine, a dangerous buildup can develop when a person with PKU eats foods that are high in protein. This can eventually lead to serious health problems.

For the rest of their lives, people with PKU need to follow a diet that limits phenylalanine, which is found mostly in foods that contain protein.

PKU symptoms can be mild or severe and may include:

- Intellectual disability (formerly called mental retardation)
- Delayed development
- Behavioral, emotional and social problems
- Psychiatric disorders
- Neurological problems that may include seizures
- Hyperactivity
- Poor bone strength
- Skin rashes (eczema)
- A musty odor in the child’s breath, skin or urine, caused by too much phenylalanine in the body
- Fair skin and blue eyes, because phenylalanine cannot transform into melanin — the pigment responsible for hair and skin tone
- Abnormally small head (microcephaly)

Foods that are not allowed:

- All meats such as: beef, lamb, pork, ham, bacon, chicken, fish and fish products, organ meats (liver, heart, kidney)
- Eggs
- All dairy products including: cottage cheese, cheese, milk, yogurt, ice cream, pudding, etc.
- Nuts and seeds
- Legumes
- Ordinary breads, flour cakes, and biscuits (made with yeast and/or gluten)
- Soya-Foods such as TVP (meat substitutes)
- Any food containing aspartame such as: diet sodas, diet jams, diet lemonades, etc.
Galactosemia

Galactosemia is a condition in which the body is unable to use the simple sugar galactose. People with galactosemia are unable to fully break down the simple sugar galactose. Galactose makes up half of lactose, the sugar found in milk.

If an infant with galactosemia is given milk, substances made from galactose build up in the infant’s system. These substances damage the liver, brain, kidneys, and eyes.

Persons with galactosemia cannot tolerate any form of milk (human or animal). They must be careful about eating other foods containing galactose.

Infants with galactosemia can develop symptoms in the first few days of life if they eat formula or breast milk that contains lactose.

Symptoms:
- Convulsions
- Irritability
- Lethargy
- Poor feeding
- Poor weight gain
- Yellow skin and whites of the eyes (jaundice)
- Vomiting

Infants can be fed with:
- Soy formula
- Meat-based formula or Nutramigen
- Another lactose-free formula

Calcium supplements are recommended.

Some Foods and Ingredients That Contain Galactose:
- Butter
- Buttermilk
- Buttermilk solids
- Casein Cheese - including cottage cheese, cream cheese and other cheese based products
- Cream
- Curds
- Dry milk
- Dry milk protein
- Garbanzo beans - can also be called chickpeas
- Ghee
- Hydrolyzed protein
- Ice cream
- Lactalbumin - can also be called milk albuminate
- Lactoglobulin
- Lactose
- Some margarines
- Milk
- Milk chocolate
- Milk solids
- Nonfat dry milk
- Nonfat dry milk solids
- Nonfat milk
- Organ meats - these include liver, heart, kidney, brains, sweetbreads, and pancreas. These are often listed as “meat byproducts” on labels.
- Sherbet - contains nonfat dry milk. This is different from sorbet, which is more like a fruit ice and is often acceptable.
- Sour cream
- Whey and whey solids
- Yogurt

For more information:


http://www.npkua.org/