Hereditary fructose intolerance (fructose malabsorption, fruktosemiya) is a digestive pathology due to inborn errors of metabolism of fructose - deficiency of the enzyme aldolase B [8, 3]. It is an autosomal recessive disorder caused by a mutation ALDOB- gene located at locus q22.3 of 9th chromosome [9]. First discussed pathology was described in the 1956 year by Chambers and Pratt, and in 1969 year Odievre collected in world literature 57 cases of this disease [10].

According to the U. B. Kaiser [1], in patients with hereditary fructose intolerance identified eight types of structural genes deficit aldolase B. There are about 30 different mutations, the most common of which is A150R missense mutations, A175D and N335K, which together account about 80 % of mutant alleles [8]. The risk of having an affected child with parentsthouse carriers of pathogenic genes, with each new pregnancy is 25%.

Fructose (fruit, or fruit sugar) is a ketohexose of a monosaccharide group [3]. This is the sweetest of the sugars (1.5 times sweeter than sucrose and 3 times - glucose) substance soluble in water, widespread in nature (found in fruits and vegetables in a small amount, honey, chocolate) [4]. Fructose was first isolated from a "honey water" in 1972 by the Russian chemist T.E. Lovitz [5]. The discussed monosaccharide is a major source of carbohydrates for human body, used for the synthesis of fatty acids, stabilizes blood glucose levels, has a positive effect on the immune system, speeds up the metabolism of alcohol [4, 6].

Supplied in food fructose has three forms: a monosaccharide (fruit and honey); disaccharide, monosaccharide fructose which is in communication with a monosaccharide sucrose (white and brown table sugar) and fructans - polymers of fructose, oligo-and polysaccharides (wheat and some vegetables) [7].

Metabolism of fructose include multiple paths of its use for the synthesis of other substances (saccharides, lactose, etc.) and in supplying energy to the body part, wherein it is converted to glucose in the liver or its metabolic intermediates (glycogen, triacylglycerols).

Aldolase defect is accompanied by accumulation of fructose-1-phosphate, which inhibits the activity of phosphoglucomutase, converts glucose-1-phosphate into glucose-6-phosphate. In connection with the inhibition of glycogen breakdown occurs at the formation of glucose 1-phosphate, resulting in hypoglycemia develop. As a consequence, the accelerating lipid mobilization and oxidation of fatty acids.

Discussed the disease does not manifest itself until the child is fed breast milk, i.e. food does not contain fructose. Symptoms occur when added to the diet of fruit juices and sugar. While abdominal pain, regurgitation, vomiting, diarrhea may occur as early as 30 minutes after ingestion of food containing fructose.
Typical symptoms of fructose malabsorption are at air belching, flatulence, abdominal pain, nausea, vomiting, unstable stools or diarrhea, headache, fatigue and over time and weight loss [11].

Clinical aspects of fructosemia are very diverse from the difficult to diagnose forms to severe, including not only the defeat of the gastrointestinal tract, but also other organs and systems [9].

Currently, there are several methods for diagnosis of fructose intolerance. First of all this is history analysis: figuring out what foods cause the child disgust or symptoms of the disease, whether there are features of heredity. In the objective status of the possibility of fructose intolerance may indicate reduction in body weight and length, slow mental development, icteric staining of the skin, hepatomegaly. Conducting laboratory tests, should pay attention to whether the child has hypoglycemia, changes in the level of electrolytes, glycosuria, proteinuria, aminoaciduria. Sample load reveals fructose malabsorption in this kind of reduction in blood sugar levels after 20 and 60 minutes after ingestion of fructose (clinically with potliv child feels hunger). Fructose intolerance is diagnosed by a positive result and the hydrogen breath test [13]. Genetic testing identifies a mutation in the locus q 22.3 9th chromosome.

Identification of fructose intolerance and its elimination from the diet significantly improves intestinal symptoms. It should be remembered that the degree of manifestation of symptoms depends primarily on the amount consumed fructose.

Babies and children first year of life, being on the mixed and artificial feeding, use formula milk, no sugar, but with the lactose and maltose - dextrin. Instead of fruit juices and purees offer child glucose. Such children earlier than healthy, shows the introduction of solid foods in the meat, fish, eggs, cheese. Diet food shown to children 5-6 years, then under the supervision of a physician and blood biochemical parameters for possible inclusion in the power of fruit, starting with minimal amounts of controlled observation diary.

S. J. Shepherd et al. [22] developed dietary guidelines for patients with congenital fructose intolerance, according to which the eligible products are stone fruits (apricot, nectarine, peach, plum), berries (blueberries, blackberries, raspberries, strawberries), citrus (kumquat, grapefruit, lemon, lime, tangerine, orange) and a ripe banana, kiwi, passion fruit, pineapple, rhubarb, tamarillo, chestnuts. Allowed vegetables green beans, watercress, lettuce, leeks, cabbage, spinach. All prohibited medication containing sugar and sorbitol (pellets, dragees, powders, tablets).

Thus, hereditary fructose intolerance is more common than diagnosed in our practice. Having many clinical manifestations, this disease can be one of the causes of regurgitation syndrome, rumination, cyclic vomiting, intestinal colic and other dyspeptic manifestations in
infants and adults – functional digestive disorders, irritable bowel syndrome, liver cirrhosis, renal failure and etc. In modern conditions diagnostics fructose intolerance is real and timely administration of diets containing no fructose, can improve the health and condition of the patients.