Summary. Aortic ostium stenosis is a group of the congenital heart malformations which is characterized by the disorder of the blood outflow from the left ventricle (infundibular-subvalvular stenosis), on the valve level (valve stenosis) and the ascending portion of the aorta (supravalvular stenosis). According to International Statistical Classification of Diseases -10, the following is defined: Q23.0 Congenital aortic valve stenosis; Q24.4 Congenital subaortic stenosis; Q23.4 Left-sided heart hypoplasia syndrome (Atresia or a marked hypoplasia of the ostium or of the aortic valve with the ascending aortic portion hypoplasia and with the left ventricle malformation (with stenosis or mitral valve atresia)). Frequency of the aortic ostium stenosis among all the congenital heart malformations ranges from 2% to 11%. Interaction with other defects. It is often combined with the patent ductus arteriosus and coarctation of the aorta, ventricular septal defect, atrial-septal defect. Valvular aortic stenosis can be observed under different chromosome aberrations and congenital syndromes. Clinical picture. Highly expressed hemodynamic disorders develop under stenosis of the aortic ostium with the diameter less than 1/2- 1/4 of its proper size. Block to the blood-flow from the left ventricle leads to the pressure rise in it, hypertrophy and to the subsequent degenerative changes in the myocardium.

The defect can go quite for a long time asymptotically but the physical exertions may provoke hemodynamic disorders which is the reason of a sudden death, connected with the dramatic coronary blood-flow deficiency and arrhythmogenesis.
and asystolia. Most children with the moderate stenosis are born with normal body weight and height and don’t lag behind in physical and psychic development at a later stage. Weak peripheral pulse, strengthened apex beat, systolic murmur in the second intercostal space to the right of sternum can be identified. The ultrasonic research shows the following signs: deformation and fusion of the aortic valve cusps; restriction of its flexibility; contraction of the aortic valve ostium in systole with the transversal section of the root; cupular cusps curve; appearance of the systolic turbulent flow in the ascending portion of the aorta; registration of the transaortic pressure gradient, etc. **Clinical manifestation** of the malformation reveals itself only by the schoolhood. Electrocardiogram data: electric axis deviation to the left or normogram, features of the left ventricle hypertrophy, S-T segment and T wave depression with the successive traits of the left atrium hypertrophy at a later stages. Echocardiography shows deformation of the valve cusps, with its protrusion in the aorta lumen. Dopplerechocardiography registers a high-speed turbulent blood-flow on the level of the valve. X-Ray data: the lung pattern is usually without changes; intensified lung pattern of the venous vessels bed is possible in case of a severe stenosis and left ventricle heart failure; well-shaped waist of the heart is quite a trait for this heart defect, with the apex stilted over the diaphragm. **Differential diagnostics** is led between different variants of the stenosis, including idiopathic hypertrophic subaortic stenosis as well as with the ventricular septal defect, coarctation of the aorta, pulmonary artery stenosis.
**Treatment.** Children don’t need any drug therapy if showing no symptoms, though, if there are any, the main method of treatment is a surgical correction. Heart glycosides should be used extremely carefully in case of the acute left ventricle cardiovascular insufficiency evolvement! **Prognosis.** Natural course of the defect promotes progression of the obstruction size with the average lifespan between 30-32 years. The prognosis of the light and moderate stenosis is quite favourable. The postsurgical survival with the severe stenosis in the run of fifteen years ranges from 85% to 95%. Sudden cardiac death is registered in 2-10% of the unoperated cases of the subaortic stenosis.