Introduction
Hypoplastic Right Heart Syndrome (HRHS) is a rare cyanotic congenital heart disease with undeveloped right parts of heart (hypoplastic ventricle, tricuspid and pulmonary valves, under-development of pulmonary vein and artery) often together with atrial septal defect. The survival of the newborn depends on the size of the defect in atrial septal and level of development of the right heart structures. HRHS is a condition, which demands early diagnostic and surgical investigation in neonatal period.

Key words: fetal right heart hypoplasia

Case report
A 36-year-old pregnant woman had unfavorable history: missed abortion, ectopic pregnancy and genetic pathology - MTHFR gene mutation and protein S deficiency. All her three pregnancies occurred after in vitro fertilization. The fetal ultrasound at 12 weeks of gestation revealed increased nuchal translucency 9.5 mm. Diagnostic amniocentesis was performed at 17 weeks of gestation. There were found no aneuploidies. Fetal cardiac sonography at 18 weeks revealed asymmetric heart chambers, hypoplastic right ventricle, high ventricle septal defect, narrow pulmonary trunk, pericardial fluid. A follow-up ultrasonography was performed every two weeks. The pregnancy continued to 39 weeks of gestation. A 3340 g female neonate was delivered vaginally, Apgar scores 9/9. A newborn condition required intensive care due to congenital heart disease. Postnatal echocardiography confirmed the antenatal ultrasound findings: enlarged left ventricle, hypoplastic right ventricle, perimembranous ventricular septal defect, tricuspid valve atresia, small patent ductus arteriosus. There were no indications for surgical heart investigation at the time. The newborn was discharged in good clinical condition. A newborn was again hospitalised at 8 months age and Glenn procedure was performed.

RHHS is a rare and clinically challenging fetal heart condition which requires early detection and intensive postnatal care.