CASE REPORT

A Single Ventricle Defect: A Case Report from Fetal Echocardiography to Cardiac Surgery

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Abstract

Objective: The aim of the paper is to show a report of rare, life-threatening congenital heart defect (CHD), emphasizing the importance of fetal echocardiography for the diagnosis and treatment of CHD improving the outcome.

Case description: The diagnosis of the single ventricle, rudimentary left ventricle, common atrium, double outlet right ventricle (DORV), transposition of the great arteries (TGA), and mild pulmonary stenosis was made prenatally at routine fetal echocardiography scan and confirmed postnatally by pediatric cardiologist assessment, who indicated postnatal cardiosurgical treatment. According to planned treatment, the first step was establishment of the central shunt (polytetrafluoroethylene 3.5 mm) between the ascending aorta and the left pulmonary artery (LPA), followed by a bidirectional Glenn shunt. The patient was hemodynamically stable at discharge, treated with acetylsalicylic acid as antiaggregating agent. In the future, Fontan procedure as the third surgical procedure is planned.

Conclusion: Accurate prenatal diagnosis of CHD confirmed postnatally enabled to prepare medical team and parents for the treatment enabling survival of the infant and better parental coping with the diagnosis and treatment of severe life-threatening CHD.

Keywords: Congenital heart defects, Fetal echocardiography, Postnatal cardiac assessment, Treatment.

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Introduction

A single ventricle is a rare congenital heart defect (CHD) counting for 1–2% of all CHDs. A single ventricle is a result of the embryonic maldevelopment of ventricular separation and/or valve formation resulting in severe life-threatening CHD.[1,2] Blood from the right and left atria is mixing in the large common ventricle of the malformed heart.[1,2] Depending on the morphology of the single ventricle it can be assigned as morphologically left, denoted as double inlet left ventricle (DILV) or right, marked as double inlet right ventricle (DIRV).[1,2] Sometimes it is not possible to determine the morphology of the ventricle.[1,2] In most cases, there is an additional rudimentary outlet ventricle connected with the dominant ventricle through bulboventricular opening or ventricular septal defect (VSD).[3] Both great vessels arise functionally from large single ventricle, so aorta and pulmonary artery contain mixed desaturated blood with the same oxygen saturation in both vessels.[4,5] Therefore, transposition or normal position of the great vessels is not so important in terms of hemodynamics.[4,5] The single ventricle is supplying both the systemic and the pulmonary circulation, which may cause ventricular insufficiency with congestive heart failure.[4,5] Single ventricle comes very often with associated cardiac anomalies affecting adversely hemodynamics.[6–8] If pulmonary stenosis is not present, a large cardiac silhouette

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with a large left atrium and excessive pulmonary blood flow can be depicted by the X-ray. If pulmonary stenosis is present, the heart on the X-ray is of the normal size with decreased pulmonary vascularization. If there is the transposition of great vessels, a narrow mediastinum is present on X-ray.[6–8] The main finding on fetal and postnatal echocardiography is a large single ventricle with two atrioventricular (AV) valves. The single ventricle is best visualized in four-chamber and parasternal long axis view. Sometimes critical pulmonary stenosis and/or pulmonary atresia, and interrupted aortic arch could be diagnosed, as well as a severe coarctation of the aorta, and hypoplastic aortic arch.[6–8]

Aim

The aim of the paper is to present a rare, life-threatening CHD, pointing out the importance of prenatal diagnosis by fetal echocardiography, enabling timely postnatal confirmation of diagnosis and timely initiation of treatment with good outcome.

Case Description

The diagnosis of this CHD (single ventricle, rudimentary left ventricle, common atrium, double outlet right ventricle (DORV), d-transposition of the great arteries (d-TGA), and mild pulmonary stenosis) was established by the 2nd trimester fetal echocardiography (Figs 1 and 2). The course of pregnancy was uneventful. After the appearance of regular uterine contractions, the emergency cesarean section was performed at 37^{+6} weeks of gestation due to the breech presentation. Birthweight was 2995 gm, birth length was 47 cm, head circumference was 34 cm, and Apgar scores were 9 and 10 after one and 5 minutes. After the initial stabilization after birth, prostaglandin E1 was administered intravenously to keep the fetal shunts opened. There was no need for any other intensive therapy during the preparation for aortopulmonary 3.5 mm polytetrafluoroethylene shunt between ascendent aorta and left pulmonary artery (LPA). Patent ductus arteriosus (PDA) was not ligated and kept opened. After the surgery fetal echocardiography revealed dextrocardia, right atrial isomerism and functionally single ventricle with common AV valve, common atrium, pulmonary stenosis, mild common AV valve insufficiency, and central shunt. The second surgical procedure was done at the age of 1 month, when Glenn bidirectional shunt was created. The patient was discharged home clinically and hemodynamically stable, treated with 30 mg per day of acetylsalicylic acid. The Fontan procedure was scheduled at the age of 24 months.

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Discussion

The aim of the third surgical procedure is to separate right from the left heart by the Fontan procedure. Before an upper cavopulmonary anastomosis (upper bidirectional Glenn anastomosis) can be made as the first step of Fontan palliation, palliative therapy is usually needed if pulmonary and systemic perfusions are not balanced.[9] The Fontan procedure is usually performed in patients with a single ventricle either anatomic or functional, which may have the rudimentary ventricle with less than 30% of its expected volume.[10] The functionally single ventricle is supplying systemic high resistance circulation, as well as pulmonary low resistance circulation to support long-term survival.[10] Such balanced circulation occurs only rarely without surgical intervention which in the most cases is mandatory (Fontan procedure).

It involves diverting the venous blood from the inferior vena cava (IVC) and superior vena cava (SVC) to the pulmonary arteries without passing through the morphologic right ventricle; that is, the systemic and pulmonary circulations are placed in series with the functional single ventricle.

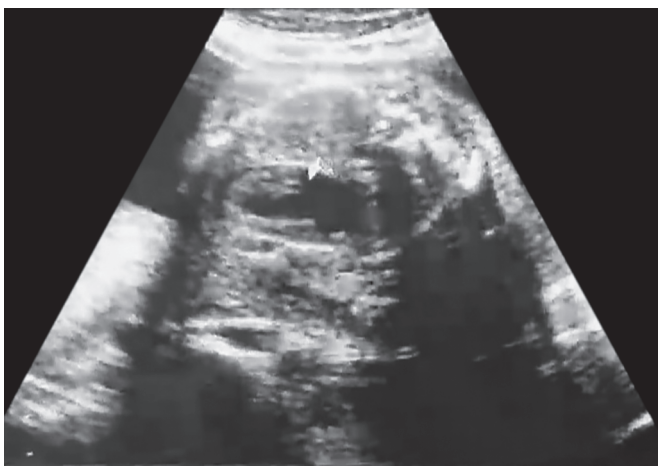

Fig. 1: Single ventricle of fetal heart at 22 gestational weeks

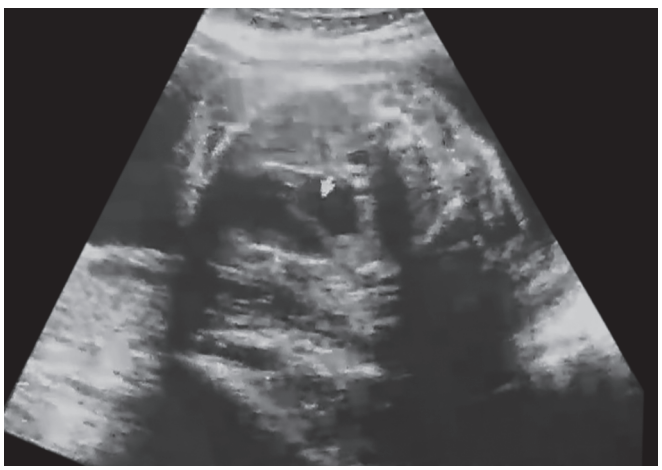

Fig. 2: Common valve of the enlarged univentricular heart (fetal echocardiography finding at 22 gestational weeks)

The arterial oxygen saturation is then around 75–85 %.[11,12] This procedure is made at the age of 2–6 months after a decrease in pulmonary resistance.[11–13] This procedure is generally scheduled after 2 years of age.[11–14]

There have been isolated reports of septation of the main ventricle in the literature.[11–14] In these cases, an artificial interventricular septum is made from a synthetic patch that divides the main ventricle into two compartments. However, the akinetic synthetic patch can never fulfill the function of a real muscular septum. In some cases, only a cardiac transplant remains as the last treatment option. The long-term prognosis depends primarily on the pulmonary blood flow. If there is pulmonary stenosis with balanced hemodynamics between the pulmonary and systemic circulation, there may be relatively few symptoms until late adulthood in some cases.[15] However, most patients die in infancy from the consequences of congestive heart failure or hypoxia. The later complications after the completion of the Fontan procedure include arrhythmias, dysfunction of the systemic ventricle, protein-losing enteropathy, and the risk of thrombosis. The 10-year survival after a Fontan procedure is from 60–80 %, respectively.[16]

Lifelong follow-up is necessary for patients with a single ventricle. In the rare cases of balanced hemodynamics, surgical treatment can be postponed.[16,17]

After completion of the Fontan procedure, most patients can handle normal everyday activities, however, their ergometry test might show reduced tolerance of physical activity. Competitive sports should be avoided as well as contact sports in patients treated with oral anticoagulants.

Dextrocardia is a rare cardiac anomaly with an estimated incidence of 1 in 10.000–12.000 of liveborn, with the prevalence of univentricular anatomy between 16 and 25 %.[18] Fetal and postnatal echocardiography are important to make a diagnosis, while fetal magnetic resonance imaging fetal electrocardiography, and fetal magnetocardiography (fMCG) may contribute to a more accurate description of heart structure and rhythm disturbances when indicated.[18,19]

According to the recommendations of The Association for European Pediatric and Congenital Cardiology (AEPC), fetal echocardiography should be done between 11 and 15 gestational weeks, and after the 20th week, every pregnant woman can be evaluated. Transvaginally fetal heart can be assessed at the gestational age of 8–9 weeks.[19] The best gestational age for fetal heart assessment is between 24 and 28 weeks.[19]

Prenatal detection of fetal congenital defects is important because they are a leading cause of infant mortality, and CHD among them is most frequently detected with significant influence on morbidity and mortality.[18] Generally, most of CHDs diagnosed in a postnatal period can be detected in the fetus, apart from some minor lesions, like Secundum atrial septal defects, and patent ductus arteriosus.[18]

CHDs are associated with an increased risk for adverse neurodevelopmental outcomes, which has been attributed to factors such as associated chromosomal defects, syndromes, postnatal cardiac insufficiency, some perioperative factors in infants who require surgical treatment, and possibly in utero hemodynamic abnormalities.

Parental counseling gives the parents an opportunity to get a reasonably realistic prognosis, learn about treatment options before and after birth, make decisions concerning the management, and plan for specific needs at and after birth.[18–20]

In a meta-analysis of eight observational studies concerning the influence of prenatal diagnosis of critical CHD on neonatal mortality, it has been shown that prenatal diagnosis reduced mortality prior to planned cardiac surgery compared with infants who were diagnosed postnatally.[21] Prenatal diagnosis of CHD has also been associated with a reduction in neonatal morbidity, including severe acidosis.[21] Infants with patent ductus arteriosus (PDA) dependent CHDs can benefit from early postnatal intervention (prostaglandin E1, balloon atrial septostomy, balloon valvuloplasty, and pacing) to prevent the closure of the DA or any other complication, which may cause mortality.[18]

Our patient has a complex CHD with dextrocardia, a common AV valve with insufficiency, which further complicates the surgical treatment in this case. The heart in our patient has been assessed morphologically, functionally, and hemodynamically. The most important treatment is palliation saving life with Norwood procedure performed in the first two weeks of life, with the mortality between 5 and 15% in the most developed medical institutions. A prerequisite for such a good outcome after the operation is a good child condition before the surgery. Dextrocardia, although rare in the population, is often noticed in patients with a single ventricle.[19] Although, dextrocardia represents anatomic variation when isolated, it is often associated with multiple cardiac and extracardiac malformations.[19] According to the data from the literature, the outcome of palliative operations in patients with a single ventricle is worse if dextrocardia is present compared to the patients with normally positioned heart[16,17,20]

Conclusion

We presented an unusual neonate with univentricular heart with dextrocardia who was successfully treated in the neonatal period. Early and accurate prenatal diagnosis enabled timely postnatal intervention and proper preparation of the patient for cardiac surgery. The patient had two palliative procedures without complications and recently preparing for the third procedure. By presenting this case we wanted to emphasize how important is the prenatal diagnosis and good cooperation between the obstetrician, pediatric cardiologist, neonatologist, anesthesiologist, and pediatric cardiological surgeon to improve the outcome with timely interventions. The parents were well prepared before the

baby was born, which made it easier for them to cope with severe stress due to the life-threatening CHD of their child.

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