Importance of Prenatal Diagnosis in Transposition of the Great Arteries: Case Report

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Abstract

Background: The purpose of the study is to highlight the prenatal diagnosis and the postnatal management of transposition of the great arteries, which is characterized by anomalous origin of aorta and pulmonary artery from the heart, and is one of the most frequently encountered form of conotruncal anomalies.

Case: Fetal echocardiography on the 32nd gestational week had revealed situs solitus, atrioventricular concordance, ventriculoarterial discordance and ventricular septal defect. During consultation with the pediatric cardiac surgery, the family is informed about the favourable results of the anatomic correction (Jatene) procedure performed by an experienced team in earlyterm following birth. The mother had decided to come to Istanbul for labor and gave birth to a 3300 gr. female baby on the 38th gestational week by cesarian section in a nearby center. The baby was then transfered to our center and was taken to arterial switch operation on the 7th day.

Conclusion: Prenatal diagnosis of transposition of the great arteries with fetal echocardiography is possible Prenatal diagnosis helps to achieve favourable early and longterm results in complex congenital cardiac anomalies by early surgical intervention.

Keywords: Transposition of the great arteries, Jatene procedure, arterial switch operation, fetal echocardiography, pregnancy.

Büyük arterlerin transpozisyonunda prenatal tanının önemi

Amaç: Konotrunkal anomalilerin en sık rastlanılan formlarından olan ve kalpten çıkan aort ile pulmoner arterin ters yerleşimiyle meydana gelen bir büyük arterlerin transpozisyonu olgusunun, prenatal teşhisi ve postnatal yönetiminin değerlendirilmesi amaçlanmıştır.

Olgu: Yirmisekiz yaşında, G1 P0, gebeliğinin 32 haftasında olan anne adayının fetüsüne obstetrisyen tarafından yapılan fetal ekokardiyografide; situs solitus, atriyoventriküler konkordant bağlantı, ventrikülоarteriyel diskordant bağlantı ve ventriküler septal defekt olduğu gözlendi. Pediyatrik kalp cerrahisi ekibi ile yapılan konsültasyonda aile, bebeğin doğum sonrası transpozisyon cerrahisinde deneyimli bir merkezde, erken dönemde uygulanacak anatomik tam düzeltme (Jatene) ameliyatı ile yüz güldürücü sonuçların alındığı konusunda bilgilendirildi. Doğumu takiben yenidoğan için gerekecek cerrahi müdahale için İstanbul'a gelen anne, 38 gebelik haftasında sezaryen ile 3300 gr. kız çocuğu dünyaya getirdi Postoperatif dönemde merkezimize nakledilen yenidoğan, doğumu takiben 7 gün 'arteriyel switch' ameliyatına alındı.

Sonuç: Büyük arterlerin transpozisyonunun fetal ekokardiyografi ile doğum öncesi tanısı mümkündür. Prenatal dönemde tanısı konulan kompleks konjenital kalp hastalıklarında, multidisipliner yaklaşım ve erken müdahale sayesinde, erken ve uzun dönemde yüz güldürücü sonuçlar alınabilmektedir

Anahtar Sözcükler: Büyük arterlerin transpozisyonu, Jatene ameliyatı, arteriyel switch ameliyatı, fetal ekokardiyografi, gebelik.

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Introduction

The congenital heart anomalies are seen between 0.4 to 1.1% among the new born and they form the most common natal abnormality group.[1,2] The congenital heart defect (CHD) is six times more frequent than the chormosomal abnormalities and four times more frequent than the neural tube defects,[3] and is also the reason of more than 20% of the perinatal mortalites related to the congenital malformation as well as more than half of the deadly malformations encountered during childhood.[4] Despite the fact that it is possible to diagnose major and minor heart abnormalities during the prenatal period, by systematical and repeated evaluation of the fetal heart,[5] only 10-20% of the babies born with heart abnormalities carry the known risk factors for these abnormalities. Most of the CHDs are outcomes of non-risk group pregnancies.[6] The transposition of the great arteries (TGA) is one of the most common conotruncal abnormality with a frequency up to 9-10% among all congenital cardiac anomalies.[7] TGA can be complete or congenitally corrected. The complete, or simple,transposition is defined as the abnormality of a heart having atrio-ventricular concordance and ventriculo-arterial discordance (origination of the pulmonar artery from left and the aorta from right ventricle).In congenitally corrected transposition,there is atrio-ventricular and ventriculo-arterial discordance (while the right atrium opens to the morphological left ventricle and the left atrium to the morphological rigt ventricle, the pulmonar artery originates from left ventricle and the aorta from the right ventricle).[8] The 70-75% of the patients having simple TGA do not have any other cardiac abnormality than a patent foramen ovale (PFO) or an atrial septal defect (ASD).A patent ductus arteriosus (PDA),present in the half of the patients,is closed functionally in the first month. The most common abnormality accompanying TGA, with a frequency of about 20%,is a ventricular septal defect (VSD).[9]

Case

A 28 year old patient,G1 P0,on her 32nd pregnancy week, was suspected of congenital heart defect in the fetus following a fetal echocardiography performed at an other centre is referred to Department of Obstetrics and Gyneacology, Faculty of Medicine, Kahramanmaras Sutcu Imam University. In the fetal echokardiography performed by sequential segmental analysis;situs solitus, concordance in atrio-ventricular relation and discordance in ventriculo-arterial relation is observed. No extra-cardiac abnormality is found in the evaluation made by the ultrasonography. The family is informed about the chromosome abnormality that can be seen in fetus having cardiac anomalies. Because of the short period left to the labor, the family did not require any invasive intervention aiming to define the caryotype. Following a consultation to Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Center, the family was informed in details about the heart disease of the fetus, the results of the complete surgical correction and the post operational situation of the newborn.It was decided that, should the labor took place in a hospital in Istanbul near the Center where the surgical operation will be performed, the loss of time will be minimized and the transfer of the newborn in order to prevent the problems that can be seen in the early stage would be easier. As the contractions of the mother began when she came to Istanbul on

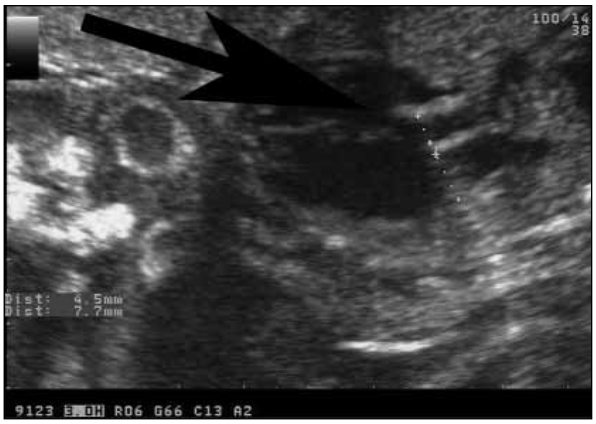

Figure 1. Great vessel output.

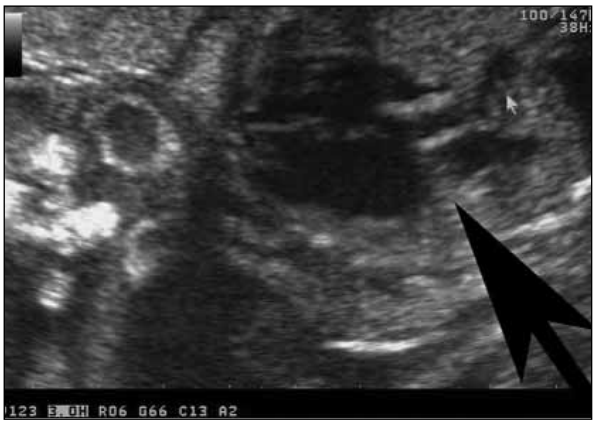

Figure 2. Great vessel output.

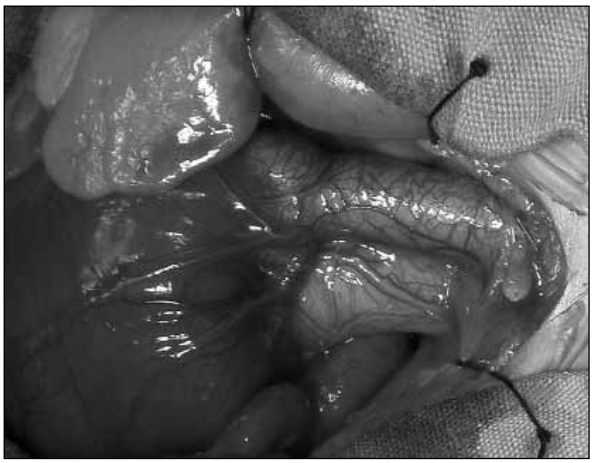

Figure 3. Great vessel output - anatomic view.

her 38th pregnancy week, she is taken to a voluntary cesarean section operation in Clinic of Obstetrics and Gyneacology, Haydarpasa Numune Hospital, where the staff was already informed about the case,and gave birth to a baby girl of 3300 grams. During the following day,in the control echocardiography of the new born after her transfer to the Center where she will be operated, the findings of the prenatal period were confirmed. After the conclusion of the preoperative preparations,the new born underwent an anatomic total correction surgery (Jatene Operation) on the 7th day following birth and she was taken to the intensive care unit after the procedure was concluded without any problems.

Discussion

In fetal cardiac anomaly detection,it is shown that the percentage of demonstrating the disease by four chamber view is 26%,[10] but taking the great arteries in consideration raises this rate to 83%.[11] The prenatal diagnosis of complete transposition is difficult because of the impression of the normal four chamber view of the fetal heart. The parallel outflows of the great arteries and the view of the non-crossing of each other in ultrasonography have an important role in the diagnosis (Figures 1 and 2). Double outlet right ventricle is the most difficult and also is the most confusing cardiac anomaly in the differential diagnosis. Especially in the cases that are accompanied by a VSD, the diagnosis becomes even more difficult. The diagnosis should be reached by considering the displacement of the atrio-ventricular valves and the aorta.[12] The reconstructive operative procedures applied for TGA are anatomic (Jatene) and physiologic (Sennig and Mustard) correction operations. The insufficiency of the systemic ventricle and of the atrio-ventricular

valve,arrhythmias and baffle obstructions that can be seen after physiologic reconstruction operations are the main reason for the tendency towards the Jatene operation in the transposition surgery.[13] (Figure 3). The left ventricle functions, which are normal during the birth, decreases in the first month following birth parallel to the fall in the pulmonary vascular resistance. After this moment, the left ventricular muscle mass and its capacity of contraction, which are very important for the Jatene operation, further decreases and this may lead to changes in the surgical strategy. That is why timing is one of the most important factors in the transposition surgery; the ideal timing for Jatene operation is, therefore the first 14 days after birth but it can be prolonged until the 21st day.[14] In the presented case,even though no problem occurred until urgery, prostaglandin E1 (PGE1) perfusion might be required to keep the PDA patent in cases where the arterial oxygen saturation is low, and meanwhile in the presence of a restrictive PFO, the balloon atrial septostomy (BAS) is applied. In simple transposition cases, where prenatal diagnosis is made, the course may be very different.It was demonstrated in a study which covered 23 complete transposition cases that; 6 pregnancies is terminated (26%), 3 fetus died in utero (13%), 8 cases died in the neonatal period (35%) and 6 cases survived (26%).[15] In our case, following the pediatric surgery consultantion, the family decided not to terminate the pregnancy. The information that about 10-15% of the fetus having a congenital heart disease could have a chromosomal anomaly was given to the family.[6] The family stated that the karyotype structure of the fetus wouldn't change their mind after this stage of pregnancy,and they have refused any invasive intervention for prenatal chromosomal diagnosis. Even though, the frequency of accompanying extra-cardiac malformations in these cases is about 10%, no other abnormality in the prenatal ulrasonography and the postnatal evaluation was found in the presented case. In the prenatal diagnosis,presence of an accompanying anomaly in the ultrasonography (especially the face, kidneys and the gastro-intestinal system), the fetal karyotype, history of congenital cardiac anomaly and drug usage during pregnancy (lithium, etc.) should be considered. After all these necessary evaluations, a reason that could explain the cardiac anomaly in our case was not found. As conclusion, the prenatal diagnosis of the TGA which necessitates early surgical intervention in the first 2-3 weeks following birth is possible by fetal echocardiography. The fetal cardiac echocardiography performed in the prenatal period, is a golden standard in diagnosis for the prevention of complex congenital heart abnormalities which may have a mortal course and also, in the early definition of the surgical intervention strategies which may be required after birth. The prenatal and postnatal management of these cases are facilitated by the multidisciplinary approach and the cooperation of an experienced obstetrician in fetal echocardiography and an experienced surgical team.

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