Prenatal echocardiographic diagnosis of situs inversus totalis and transposition of the great arteries
A case report

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A case of situs inversus totalis and transposition of the great arteries (TGA) was diagnosed prenatally at 25 weeks' gestation. Postnatal echocardiographic examination confirmed the antenatal findings. This case underscores the importance of recognizing situs abnormalities during obstetric and fetal echocardiographic examination, as they are often associated with cardiac anomalies. Accurate prenatal diagnosis of structural heart defects is extremely important in family counselling and in planning obstetric and postnatal treatment.

Key words: transposition of great arteries, prenatal, fetal echocardiography, situs inversus.

The use of echocardiography in the human fetus has paralleled the development of echocardiographic technology and has permitted prenatal diagnosis of structural and functional abnormalities of fetal circulation. Thus the most complex cardiac defects can be investigated noninvasively, allowing appropriate prenatal counselling and postnatal treatment, if appropriate.

We report a case of prenatal diagnosis of situs inversus totalis and transposition of the great arteries (TGA) and stress the importance of accurate sequential diagnosis.

Case Report

A 24-year-old female was referred to our center for specialized fetal echocardiography because of gestational diabetes mellitus. She was not insulin dependent. Previous obstetric ultrasound examination revealed a normal four-chamber view of the heart and no extracardiac abnormalities. The echocardiographic examination at 25 weeks of gestation showed the heart on the right side of the thorax with a rikght-sided apex (Fig. 1). In the abdomen, the arrangement of the viscerae and abdominal vessels was mirror-image of normal, with the stomach to the right and liver to the left side of the fetus. There was no abnormality on the four-chamber view. The atrioventricular connectilon was concordant. The foramen ovale was seen in the atrial septum, with the flap valve within the left atrium. Imaging of the arterial trunks showed the great arteries in parallel fashion rather than in a spiral relationship (Fig. 2). The aorta originated from the right ventricle, and the pulmonary artery originated from the left ventricle and was typically bifurcated (Fig. 3). Mitral-pulmonary continuity was present. Pulsed and continuous wave Doppler examinations were normal. The echocardiographic findings were compatible with situs inversus totalis and TGA.

Based on the above diagnosis, the family was counselled and the obstetrician informed. The family decided to continue with the pregnancy. Subsequent follow-up was carried out jointly by the pediatric cardiologist and the obstetrician. A term baby was delivered vaginally, uneventfully. Birth weight was 4.5 kg, length 51 cm and Apgar score 9 at 5 minutes. Mild-to-moderate cyanosis was present at birth. Cardiovascular examination revealed a short systolic murmur at the upper left sternal border. Second heart sound was single. Pulses were equal in both upper and lower extremities. Postnatal echocardiographic examination was performed during the first hours of life and confirmed the intrauterine

diagnosis. The ductus arteriosus was open and the atrial septal defect was of adequate size. At two hours of age the baby was referred to a cardiac surgical center and operated in optimal conditions on day seven.

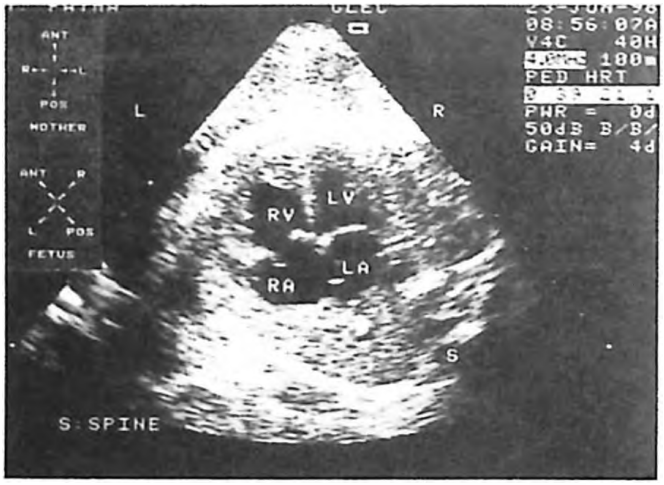

Fig. 1. Transverse section-through the fetal chest. Four-chamber view of the fetal heart; spine is on the left side of the mother. Note the fetal heart and apex on thi right side (fetal head is inferiorly located). S: spine, R: right side of the fetus; L: left side of the fetus; RA: rikght atrium; LA: left atrium; RV: right ventricle; LV: left ventricle.

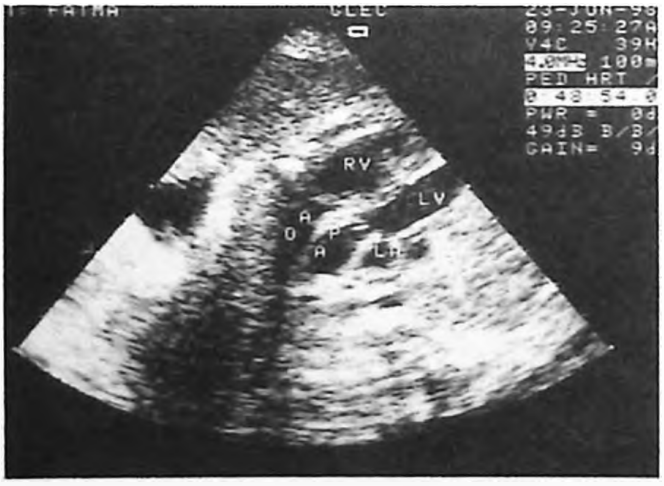

Fig. 2. View of the outflow tracts, color-Doppler flow. Note the parallel orientation of the aorta (Ao) and pulmonary artery (PA). S: stomach.

Discussion

Since the early 1980s, fetal echocardiography has proved to be a reliable tool for prenatal diagnosis of congenital heart diseases with a high degree of accuracy[1-4]. Over this time, complex heart malformations have been clearly described in the fetus. The prenatal diagnosis of TGA is possible and has been reported[1-4].

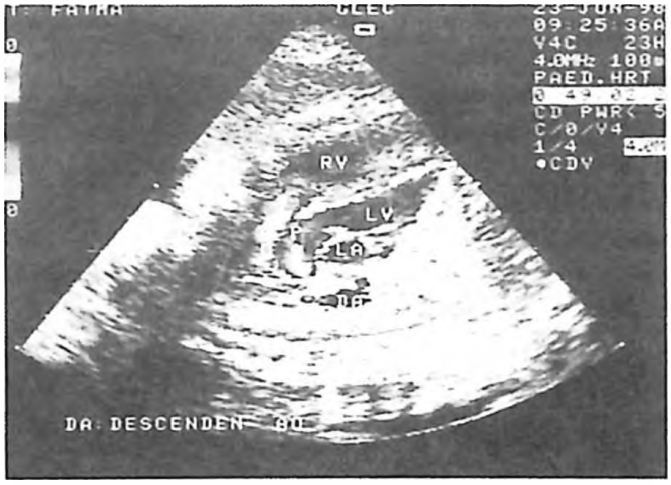

Fig. 3. Long axis view of the ventricular outflow tracts. The pulmonary artery (PA) arising from the left ventricle (LV) and the aorta (AO) arising from the right ventricle (RV). LA: left atrium.

This case, as far as we know, is the first case in Turkey that has been intrauterine diagnosed as situs inversus totalis and TGA.

In order to make the diagnosis, the transducer must be angled superiorly (towards the baby's head) to allow visualization of the ventricular outflow tracts[3], and thus determine the ventriculo-arterial connection. However, fetuses with simple transposition are frequently not referred for specialized fetal echocardiography, as most obstetric ultrasound screening programs rely on visualization of the four-chamber view alone and this view shows no obvious abnormalities. The diagnosis is therefore often missed in routine obstetric screenings. Furthermore, as the condition is well tolerated in the fetus, there is no evidence of heart failure or hemodynamic compromise that might lead to prenatal cardiac referral. During detailed examination of the heart, it is important to determine visceral and atrial situs as part of the sequential chamber localization[5]. Determination of fetal laterality and position of the viscerae during obstetric ultrasound examination, however, is not performed routinely[6]. This is exemplified in our cases where previous obstetric ultrasound had failed to show the abnormal position of the viscerae. Overall frequency of situs inversus is estimated at 1/10,000 births and is often associated with congenital heart defects[5-7]. In utero diagnosis of situs anomalies during obstetric ultrasound examination should be an indication to explore cardiac anomalies. In our patient the heart was located on the same side as the stomach. This can frequently be mistaken as the left side

if fetal laterality is not considered. When the right and left sides of the fetus were determined according to fetal orientation within the uterus, we noticed the mirror-image arrangement of all viscerae.

Accurate prenatal diagnosis of structural heart defects is extremely important for planning obstetric and postnatal treatment.

This case emphasizes the importance of fetal echocardiographic examination in complex cardiac pathologies where early surgical intervention is required.

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