

● Case report

PRENATAL DIAGNOSIS OF CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES ON THE BASIS OF FOUR CHAMBERS' VIEW - CASE REPORT.



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Abstract

Congenitally corrected transposition of great arteries is a rare congenital heart defect. The clue of the abnormality is the inversion of the ventricles which caused abnormal atrioventricular and ventriculoarterial connections. This defect is seldom identified prenatally, much more seldom than the simple transposition of the great arteries, even though, we can observe it, on the image of 4 chambers of the heart. Prenatal diagnosis of this defect, at the 24th week of pregnancy and during the routine ultrasound scan, is being described below. The echocardiographical features of the congenitally corrected transposition of great arteries are being presented, with reference to the differences in the image of the 4 chamber view.

Key words: prenatal diagnosis, four chamber view, corrected transposition of great arteries

SUMMARY

Corrected transposition of great arteries is a rare congenital heart defect. The heart of the abnormality is the inversion of the ventricles and caused by that abnormal atrioventricular and ventriculoarterial connections. This defect is seldom identified prenatally, much more seldom than the simple transposition of the great arteries, even though, we can observe it, on the image of 4 chambers of the heart. Prenatal diagnosis of this defect, in the 24th week of pregnancy and during the routine ultrasound scan, is being described below. The echocardiographical features of the congenitally corrected transposition of great arteries are being presented, with reference to the differences in the image of the 4 heart chambers.

CASE REPORT

Pregnant woman, aged 28, in the second pregnancy. The first pregnancy finished normally on time by caesarean birth, because of ectopic asphyxia of the foetus. During the first pregnancy, there was epilepsy diagnosed. The patient is taking anti-epileptic drugs on a permanent basis. Currently during the second pregnancy, because of the low concentration of the vitamin D (16 ng/ml) supplementation has been used. Prenatal examination (PAPPA test) which was carried out in the 13th week, did not show any abnormalities, the risk of genetic syndromes was low.

Ultrasound screening examination, which was carried out in 21st week, was postponed for 24th week, because of bad position of the fetus. In the 24th week, a satisfactory

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* image for the assessment of the anatomy of the heart of the foetus, was obtained. In the heart image, even though, 4 heart chambers were visible, abnormalities were found, which led to the identification: on the right side of the morphologically left ventricle with the pulmonary artery, and on the left side morphologically right ventricle with the aorta (Photo 1). Abnormal image of the upper

mediastinum was diagnosed – two arteries present (Photo 4), as well as parallel abnormal placement of the great arteries – (Photo 2,3). To verify the diagnosis and to establish further actions, the pregnant woman in the 28th week, was sent to echocardiographical examination to Department of Prenatal Cardiology, Polish Mother's Memorial Hospital Research Institute in Lodz, where previous diagnosis was confirmed. During 32nd week of pregnancy, in ECHO examination, there was a lack of continuity of the interventricular septum measuring the distance of 2mm, placed high in the muscle part. During the next echocardiographical examinations, no other abnormalities were diagnosed. The cardiovascular system function on the 10 points of the CVPS scale, had been maintained till the birth date. The birth took place in the 40th week of the pregnancy, using caesarean birth method – a 3800g baby was born. The newborn baby was given to Cardiological Supervisory Intensive Unit of Polish Mother's Memorial Hospital Research Institute, with the aim of further observation. They confirmed prenatal diagnosis. There were no disorders in the heart rhythm and no hemodynamic disorders, which were not recorded prenatally. The newborn baby was discharged from hospital in the 6th day of its life, in good general state, with the recommendation of further care in the cardiological clinic.

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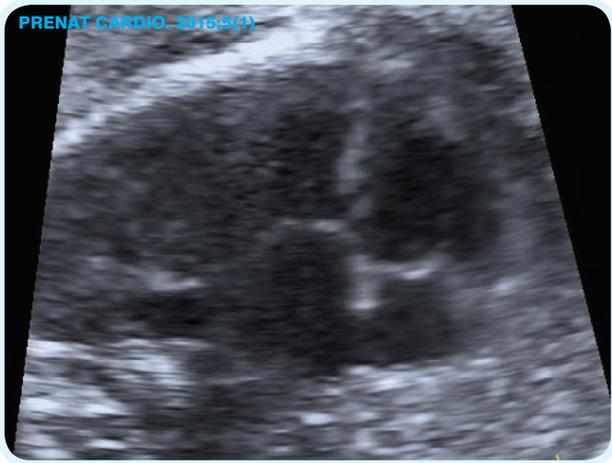


Photo 1. Four chamber view of the heart with corrected transposition of the great arteries

DISCUSSION

Corrected transposition of great arteries is an congenital heart defect. It occurs with frequency of less than 1% of all the heart defects¹. The problem of the abnormality is the inversion of the ventricles and caused by that abnormal atrioventricular and ventriculoarterial connections².

Main veins go to the right atrium that is connected with the mitral valve, with morphologically left ventricle, from which the pulmonary artery origins. Blood flows to the left atrium from the pulmonary veins and flows through the tricuspid valve to the morphologically right ventricle. The aorta origin from the right ventricle. Great arteries run parallel to each other^{2,3}. The oxygen-rich blood flow and the systemic circulation are correct.

The defect is rarely recognizable prenatally, especially when it is not accompanied by additional abnormalities. Isolated L-TGA occurs only in 9-16 %^{3,4,5}. In 50% cases, there is a defect in ventricular septum (VSD), which is very often the key to detect this abnormality⁶. In our case, the diagnosis of the L-TGA, was the key to additionally diagnose VSD. In similar percentage there is also present the valvular and subvalvular narrowing of the pulmonic valve^{6,7,8}. Ebstein's anomaly, coarctation of the aorta coexists less frequently. Due to the abnormalities in the conduction system, disorders and predisposition of the heart block occur very often^{3,9}.

The infant's condition is dependent on the types of the concomitant defects. In case of the lack of additional abnormalities, the development in the prenatal period is correct. With the presence of the atrioventricular block and the concomitant defects, the prognosis is unfavourable^{8,9,10}.

In mature age, it's difficult to predict the distant result of the defect. Heart rhythm disorders occur more often, in the form of bradycardia, which in 5% of cases, lead to the complete blockage of heart¹¹.

The result of the systemic circulation stress on the right ventricle, may also be the atrioventricular incompetence, especially of the tricuspid valve with the subsequent heart insufficiency^{2,12}.

Prenatal diagnosis of congenitally corrected transposition of great arteries still seems to be too rare. On the basis of the collected literature, the sensitivity of the prenatal diagnosis fluctuates around 14%^{5,13}. In 2005 and 2006, Sharland and Paladini published two independent examinations^{4,5}, 34 and 30 fetuses with L-TGA have been examined. Both authors underline the importance of the routine ultrasound scan,

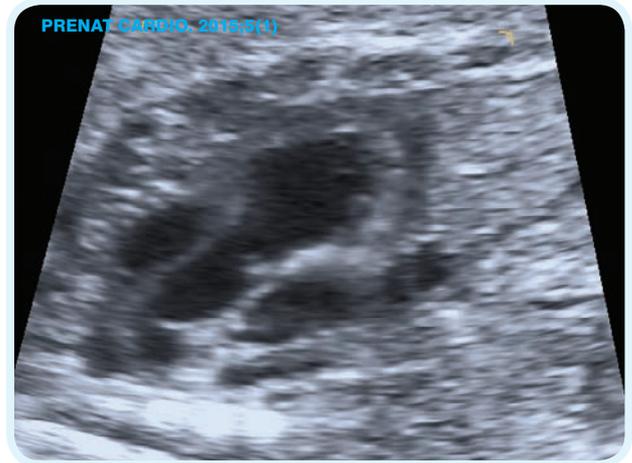


Photo 2. Left ventricular outflow track in heart with corrected transposition of the great arteries

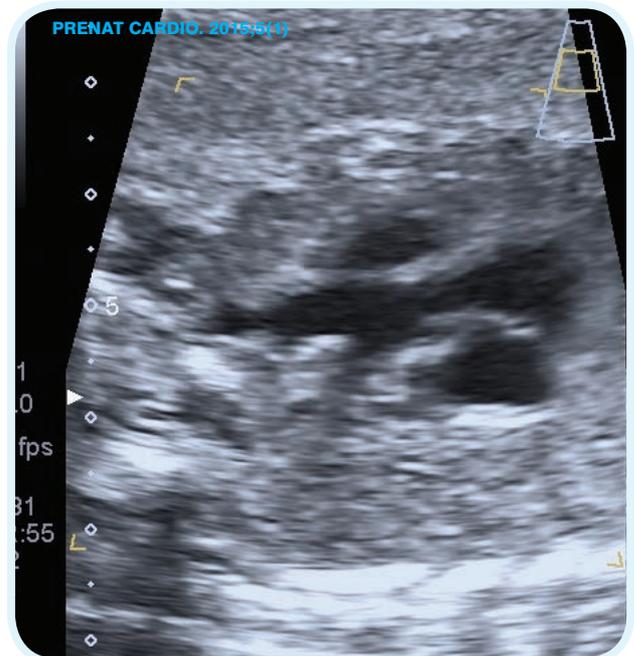


Photo 3. Right ventricular outflow track in heart with corrected transposition of the great arteries



Photo 4. Three vessel view in heart with corrected transposition of the great arteries

where the image of the four chambers of the heart and the morphological distinction of the right and left ventricle, may be the most important element in the differential diagnosis of this defect.

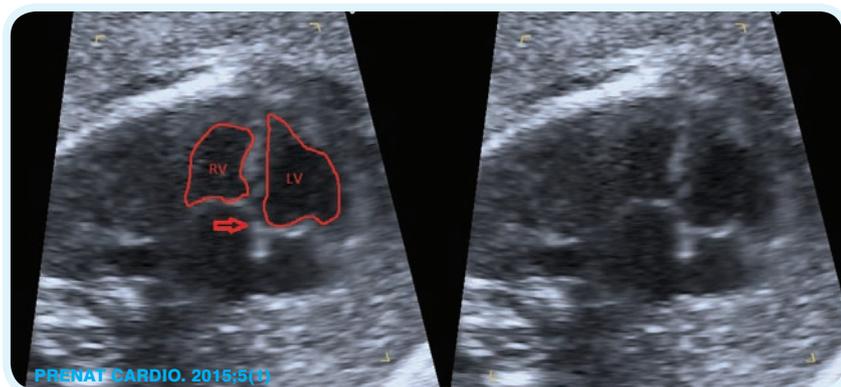


Photo 5. Abnormality in four chamber view in fetal heart with corrected transposition of the great arteries

Paladini showed three tips, which could make the distinction easier. The most important for him was the correct diversification of the trabecles. The observation of the different placement of the trabecular muscles in the right and left ventricle makes it a lot easier. In the morphologically right ventricle, muscles are heading from the tricuspid valve through the ventricle, sticking distally. In the morphologically left ventricle on the other hand, the stick to the side wall.

The next differentiation of this defect is the improper placement and the structure of the tricuspid valve. The third tip, suggested by Paladini, is dextrocardia which occurs in 25% cases.

In the case presented by us, it is the abnormal morphology of the heart's ventricles, that led us to the suspicion of the heart defect presence.

The ventricle which was round with characteristic crosswise trabeculation, with the shape similar to the pear, was placed on the left side, a little bit back (Photo 5) However, from the front of the chest, there was a ventricle which was longitudinal and similar to the banana. The next abnormality which was observed, was the unusual mediastinum image, where instead of the three vessels, there were only two big vessels observed. (Photo 4) The absence of the crossing of the big arteries was just the next step of the examination, which gave the impression of this defect (Photo 2, 3)

Without a doubt, important was the assessment of the comprehensive heart structure and the rest of the foetus's organs. In the first examination there were no, very commonly observed in this defect, ventricular septum defect and irregular heartbeat. In the next echocardiographic examination, which took place in the 8th week, there was ventricular septum defect observed. Till the end of the pregnancy, there were no irregular heartbeat nor hemodynamic failures. The next echocardiographic examinations, were not only verifying ones, but also crucial for the assessment of the foetus condition. Good condition of the foetus and no other disorders, allowed the pregnant woman to deliver the child full term.

Because of the irregular heartbeat and cardiac insufficiency occurring, in L-TGA patient's different periods of life, many scientists point out that there is a necessity of systematic observation of the foetus, since the diagnosis of the defect and through the next years^{2,8}.

In the conducted analysis Wan revealed, that around 70% of children who were L-TGA diagnosed, needed a cardiological intervention. In this group, there were big cardiosurgical interventions or pacemakers' implants. Presented analysis, makes it clear that, even rather simple 'mild' image of this

defect does not exempt from the obligation of the systematic observation of the foetus till the end of pregnancy time and during the infant period⁹.

Prenatal diagnosis of the L-TGA is possible on the basis of the careful and sharp analysis of the 4 chamber view. Contrary to D-TGA, in L-TGA abnormalities are always visible in the image of four chambers of the heart as well as in the image of three vessels.

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Małgorzata Soroka: search for references, submitting

Maciej Słodki: collecting data for the research, concept of the research, discussion, final version of the manuscript