Correction of diagnosis from d-TGA with echocardiography to cc-TGA with levocardia in situs inversus with cMRI

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Fetal heterotaxy syndrome is associated with a wide spectrum of cardiac pathology such as transposition of the great arteries that can be assessed in utero[1][2]. As prenatal detection of TGA reduces mortality and morbidity after birth[3], detailed anatomical assessment of fetal cardiovascular structures is necessary. In fetal heterotaxy syndrome, however, especially hepatic venous connections and inferior vena cava to atrial connection remain difficult to define prenatally by echocardiography[4]. In these cases, fetal cMRI may provide an additional diagnostic benefit.

CASE PRESENTATION

Fetus at gestational age of 34+5 weeks with suspected congenital heart disease (CHD) without underlying maternal risk factors for development of CHD. During routine obstetric ultrasound screening at 2nd trimester, a situs ambiguous with right-sided stomach and left-sided gall bladder but left-sided heart has been diagnosed. Subsequently referral for fetal echocardiography and fetal cMRI was made.

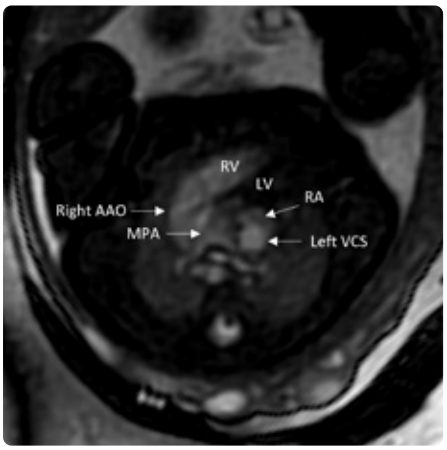

Figure 1: Axial bSSFP Cine: left-sided right atrium with junction of left vena cava superior, atrioventricular discordance with left-sided left ventricle and right-sided right ventricle, ventriculoarterial discordance with the right ventricle connected to the right ascending aorta and the left ventricle connected to the main pulmonary artery. AAO: ascending aorta; LV: left ventricle; MPA: main pulmonary artery RA: right atrium; RV: right ventricle; VCS: vena cava superior

INVESTIGATION

Fetal echocardiography in gestational week 34+5 could specify the underlying heart defect to a situs ambiguous with left-sided heart and left-sided juxtaposition of aorta and inferior vena cava, but normal venoatrial and atrioventricular connection. Further, a double outlet right ventricle (DORV) with malposition of the great arteries and obstructed pulmonary outflow tract has been observed.

On the same day, a fetal cMRI showed a total situs inversus *(Figure 1)* with levocardia and a cc-TGA: left-sided systemic veins were connected to the left-sided right atrium and pulmonary veins were connected to the right-sided left atrium, following by atrioventricular and ventriculoarterial discordance with the left-sided left ventricle connected to the main pulmonary artery (obstructed) and the right-sided right ventricle connected to the right aorta *(Figure 2)*.

DIFFERENTIAL DIAGNOSIS

Fetal cMRI yield additional clinically relevant information by evaluating venoatrial connections and diagnosing a CC-TGA in situs inversus. Diagnostic benefit was given due to the large FOV of fetal cMRI that allowed the evaluation of the great thoracic vessels in their courses and at the point where they join the heart.

TREATMENT

Immediately after birth, the newborn was provided with prostaglandins to maintain patency of the ductus arteriosus. 6 weeks after birth a Blalock-Taussig procedure was performed to increase the pulmonary arterial blood flow due to the pulmonary outflow tract obstruction.

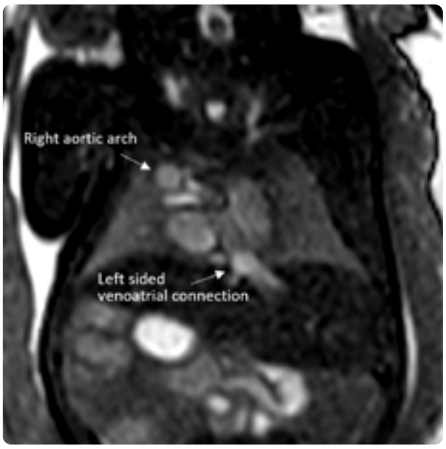

Figure 2: Coronaral bSSFP Cine: complete situs inversus with right aortic arch and left venoatrial connection

OUTCOME AND FOLLOW-UP

Due to prenatal diagnosis, the delivery took place at our university hospital, where the newborn could receive special treatment in the neonatal intensive care unit. After cardiac surgery, regular expert cardiological follow-up is scheduled.

TAKE HOME MESSAGES

As several CHD may affect the systemic or pulmonary vein system, it is crucial to ensure high imaging quality for the assessment of these structures. Since venoatrial connections may be difficult to assess by fetal echocardiography, fetal cMRI offers an additional diagnostic benefit to define the underlying heart defect in these cases.

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