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Case report

Prenatal diagnosis of criss-cross heart with congenitally corrected transposition of the great arteries – detection and fetal echocardiography monitoring with one-year postnatal follow-up

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Abstract

This paper presents an extremely rare and difficult case report of criss-cross heart (CCH) with a combination of congenitally corrected transposition of the great arteries (cc-TGA) of the fetal heart at 37 weeks of gestation. The impossibility of obtaining a proper 4-chamber view with opening of both the tricuspid and mitral valves in the same image as well as a parallel view of the great vessels confirmed later by neonatal volume-rendered computed tomography (CT) angiography were clues showing atrio-ventricular and ventriculo-arterial discordance. In addition to the heart defect, fetal echocardiography confirmed fetal well-being during subsequent exams and made it possible to plan vaginal delivery in a tertiary centre. Early cardiac surgery due to coexisting coarctation of the aorta (CoA) was performed on the 8th day and involved reconstruction of aortic arch and pulmonary artery banding. There were no clinical problems during the first year of postnatal life, despite abnormal intracardiac anatomy.

Key words: criss-cross heart, congenitally corrected transposition of the great arteries, coarctation of the aorta.

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Case report

This was a first pregnancy of a healthy woman. The fetus had routine ultrasound examinations performed at the 6th, 12th, and 20th weeks of gestation, which were determined as 'normal'. However, at the 27th week of gestation a ventricular septal defect was detected, which resulted in a referral to our centre for targeted echocardiography.

At the 37th week of gestation, the heart area/cardio-thoracic area (HA/CA) ratio was normal. Moreover, levocardia and situs solitus were observed both on the abdominal and atrial level. Both atria were found on their correct sides; however, it was impossible to visualize a tricuspid and mitral valve opening in the same plane. Two ventricles were present, but the morphologically left ventricle was found on the right side, and the hypoplastic morphological right ventricle was located on the left side.

Sequential analysis off-line in slow motion confirmed that blood flows were not parallel within the fetal heart – the blood from the right atrium was directed to the left ventricle, whereas

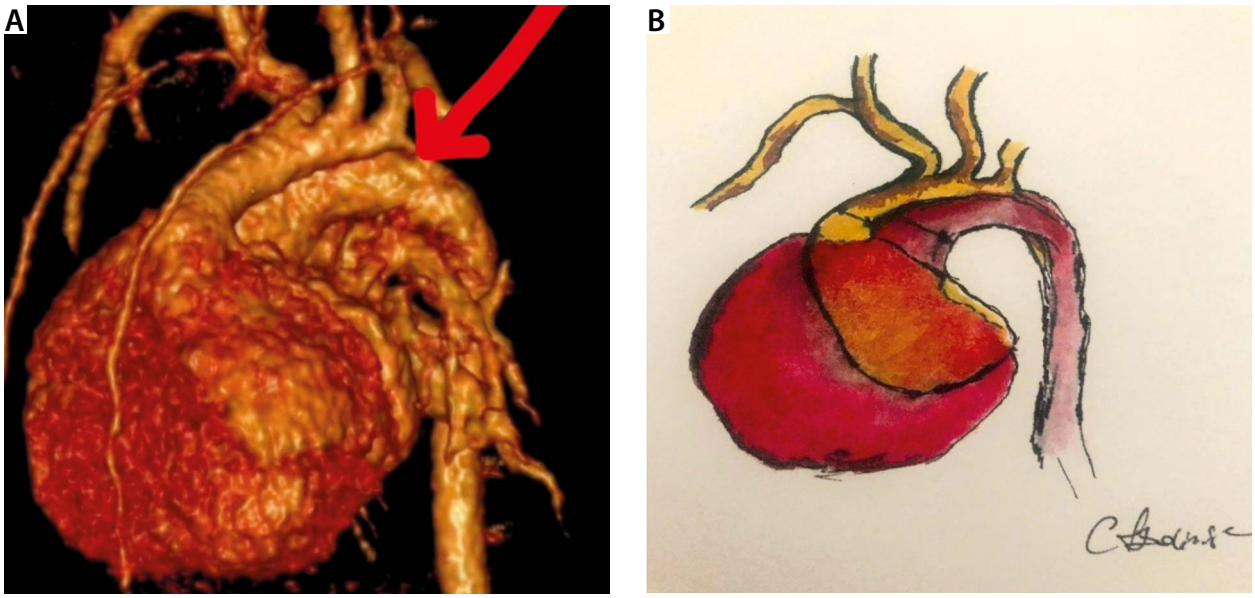

Figure 1. Postnatal confirmation. **A** – Newborn volume-rendered computed tomography angiography on the 5th day of postnatal life: the hypoplastic right ventricle/left ventricle are in a superior-inferior relation to each other due to an abnormal ventricular rotation in the criss-cross heart malformation. The aorta (indicated by an arrow) in front emerges from the upper chamber, which is a hypoplastic right ventricle: ventriculo-arterial discordance. The red arrow is pointing at coarctation of the aorta. **B** – A graphic assessment by AZ

Table 1. Prenatal echocardiographic parameters

Parameters	1st fECHO	2nd fECHO	3rd fECHO	4th fECHO	5th fECHO
GA by LMP		33.4	37.0	38.0	39.4
GA by US	31.2	34.0	35.5	36.0	36.3
UMB PI	1.06	1.1	0.96	0.73	0.9
Fetal presentation	cephalic	cephalic	cephalic	cephalic	cephalic
AFI [cm]		13	6 .5↓	7 .2↓	4↓
EFW [g]	1798 .1	2231 ±326	2754 ±402	2760 ±403	2919 ±426
FHR [bpm]	150	140	140	138	104
Axis of heart	–	–	75°	75°	–
AP	–	–	44	45	46
HA/CA	0.36	–	0.45↑	0.4↑	0.3
PA [mm]	8.1	10	13	13	13
AO [mm]	5	4.7	7	7	7
TV [mm]	7.5	7	–	–	–
MV [mm]	8.4	11	–	–	–
FO [mm]	–	4.3	R-L	R-L	R-L
CVPS	10	10	10	10	10
Speckle tracking analysis Global Sphericity Index (BAL/TL)				1.24	1.21
Speckle tracking analysis FS LV [% ±SD]				Abnormal LV contractility: Decreased LV 24 segment fractional shortening < 5th centile/< –1.65 Z score in 6-24 segments	Abnormal LV contractility: Decreased LV 24 segment fractional shortening < 5th centile/< –1.65 Z score in 1-24 segments
Speckle tracking analysis FS RV [% ±SD]				Abnormal RV contractility: RV Fractional Shortening < 5th centile/< –1.65 Z score in 9-24 segments	Abnormal RV contractility: RV Fractional Shortening < 5th centile/< –1.65 Z score in 4-24 segments
Global strain LV [%]				–1.56	–1.85
Global strain RV [%]				–1.86	–7.02

fECHO – fetal echocardiography, US – ultrasonographic scan, UMB PI – pulsatility index of umbilical artery, AFI – amniotic fluid index, EFW – estimated fetal weight, FHR – fetal heart rate, HA/CA – heart area/cardio-thoracic area ratio, PA – pulmonary artery, AO – aorta, TV – tricuspid valve, MV – mitral valve, FO – foramen ovale, CVPS – cardio-vascular profile score

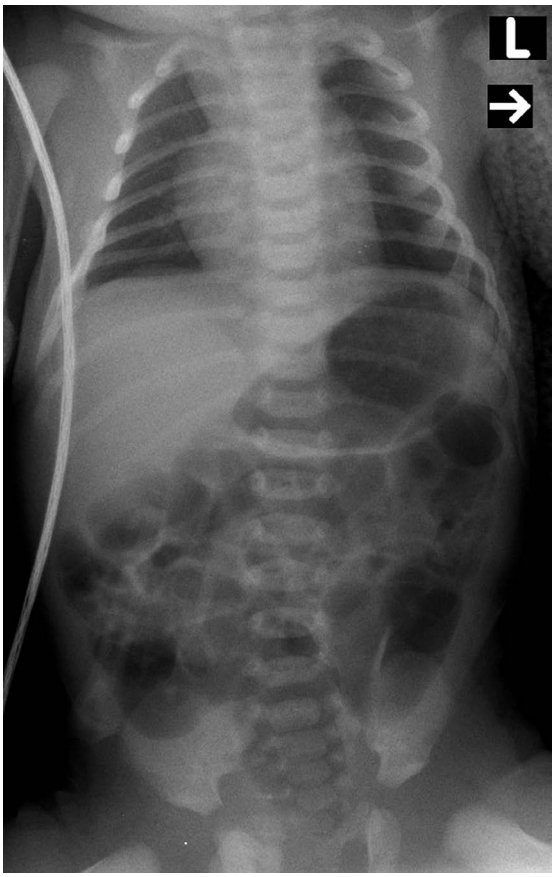

Figure 2. Chest X-ray of the heart of the newborn on the first day of postnatal life. The heart has an unusual presentation with no typical apex; however, cardiomegaly is not present

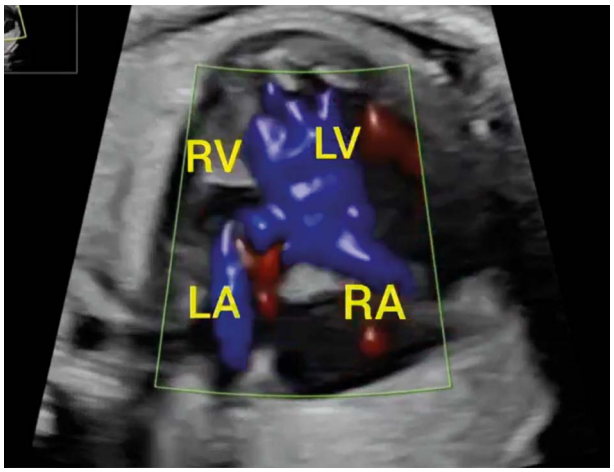

Figure 4. Criss-cross heart with congenitally corrected transposition of the great arteries: color Doppler blood flows from the left atrium through the tricuspid valve was directed to the right ventricle, from the right atrium through the mitral valve directly to the left ventricle, from the right atrium indirectly through the large VSD to the right ventricle and the blood was mixed between the ventricles

the blood from the left atria was directed to the right ventricle. The position of the hypoplastic right ventricle was superior – above to the left ventricle, which was inferior, and this was clearly confirmed by postnatal computed tomography (CT) angiography (Figure 1). So, there was a superior-inferior relationship between the ventricles. The aorta emerged from the mor-

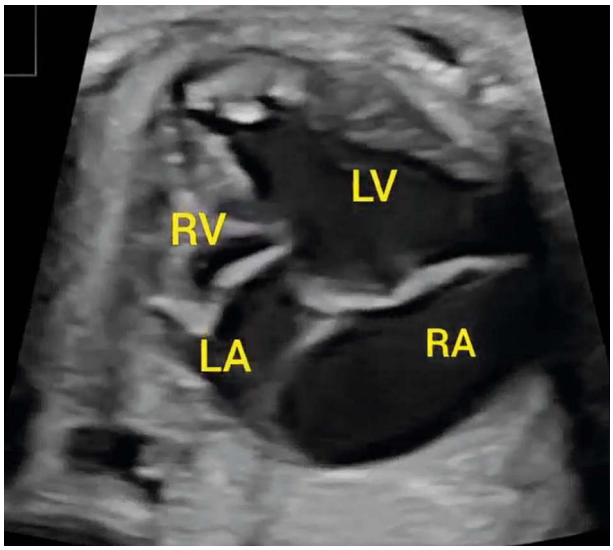

Figure 3. Criss-cross heart with congenitally corrected transposition of the great arteries: four chamber view: the left atrio-ventricular valve is inserted in perpendicular view to the right atrio-ventricular valve. Left atrium is connected by the twisted atrio-ventricular valve with the hypoplastic ventricle of the right ventricle's morphology. The right atrium is connected with the morphologic left ventricle. There are discordant atrio-ventricular connections: on the left side there is small right ventricular and on the right side there is large left ventricle. Right ventricle/ left ventricle are like up/down in relation to each other. Ventricular septal defect is presented in the middle "inflow" part of the septum From the morphological hypoplastic right ventricle is coming off the aorta and under the aorta is pulmonary trunk widening distally into the arterial duct

phological hypoplastic right ventricle and was parallel to the wide pulmonary trunk, which means that there was ventriculo-arterial discordance. The 2 arterial vessels in superior mediastinum had normal blood flow according to Doppler measurements. During subsequent four echo-sonographic exams (Table 1), there were normal umbilical Doppler blood flow parameters and normal heart rate up to the 38th week of gestation. Three weeks before delivery, the size of the pulmonary trunk was 13 mm (Z score +2.47) [1]. The development of the aorta stopped at the same time, and its maximum size was 7 mm (Z score –0.43). The aorta/main pulmonary artery (AO/MPA) ratio was 0.54, which suggested coarctation of the aorta (CoA). There was normal right-left blood flow through the foramen ovale, and the size of the ventricular septal defect (VSD) did not increase. In each fetal exam, the cardio-vascular profile score (CVPS) was 10, so a vaginal delivery was planned in our tertiary centre (prenatal cardiology, obstetrics, paediatric cardiology, and cardiac surgery departments). At the 40th week of gestation a spontaneous delivery occurred, and a male baby was born with a weight of 3000 g and an Apgar score of 10. The Prostin infusion was initiated just after birth to maintain the arterial duct patency. Postnatal echocardiography, chest X-rays, and volume-rendered CT angiography (Figures 1-4) confirmed the prenatal findings and also made it possible to confirm aortic coarctation type A. The aortic arch reconstruction surgery with pulmonary trunk banding was performed on the 8th day of postnatal life, and the child was asymptomatic during his first year of life (our postnatal follow-up).

Table 2. Reports of prenatal diagnoses of CCH and ccTGA in PubMed and our case

Authors	Year of publication	CCH + cc-TGA	Gest age at the time of detection	Additional anomalies	Postnatal follow-up
Ravi P, *et al.*	2017	1/5	23	DORV+ CoA TGA	PA band Stable at 8 months of age
Vorisek CN, *et al.*	2020	4/8	29 31 21 24	VSD DORV RAA ASD PS TGA TAPVC	Stable
Our case	2021	1	27	CoA	CoA repair and PA band 1 year of life: asymptomatic

CCH – criss-cross heart, cc-TGA – corrected transposition of the great arteries, CoA – coarctation of the aorta, VSD –ventricular septal defect, DORV – double outlet right ventricle, PS – pulmonary stenosis, RAA – right aortic arch, ASD – atrial septal defect, TGA – transposition of great arteries, TAPVC – total anomalous pulmonary venous connection, PA – pulmonary artery.

Discussion

Criss-cross heart (CCH) combined with corrected transposition of the great arteries (cc-TGA) is a cardiac malformation that is very difficult to diagnose both prenatally and postnatally because it is a rare and complex cardiac malformation. In the past, such a malformation was described only in autopsy examinations [2] or was detected by accident in adults [3-5]. In the years 2008-2020 progress in ultrasound technology and experience in fetal echocardiography enabled the diagnosis of CCH or cc-TGA prenatally [6-8]. However, the combination of criss-cross heart and corrected TGA was found only in individual cases (Table 2) [9, 10].

What is unique in our report is not only the diagnosis of abnormal heart anatomy, but also fetal heart monitoring by echocardiography. Besides the diagnosis, fetal echocardiography enabled safe monitoring and preparation of the pregnant woman and medical staff for vaginal delivery at term in the tertiary centre for obstetrics and cardiology. Further cardiological work-up is planned for our patient, but his medical history as well as other cases presented in the literature (Table 2) confirm that despite an abnormal fetal heart looping in early embryogenesis, it does not cause congestive heart failure both prenatally and postnatally and is not related to an abnormal karyotype. These data have crucial value for prenatal consultation purposes for parents-to-be as well as obstetricians, perinatologists, paediatric cardiologists, and adult cardiologists.

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Conflict of interest

The authors declare no conflict of interest.

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