Cite as: Witkowski S, Węgrzynowski J, Krekora M, Maroszyńska I, Mazurek-Kula A, Grzelak P, Januszewska K, Iwona Strzelecka I, Respondek-Liberska M: Total anomalous pulmonary venous connection – prenatal echocardiography and neonatal follow-up. J Ultrason 2025; 25: 12. doi: 10.15557/JoU.2025.0012.

Submitted: 04.08.2024
Accepted: 20.02.2025
Published: 16.04.2025

Total anomalous pulmonary venous connection – prenatal echocardiography and neonatal follow-up

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DOI: 10.15557/JoU.2025.0012

Keywords
TAPVC; total anomalous pulmonary venous connection; prenatal diagnosis; prenatal echocardiography; fetal echocardiography; fetal echo

Abstract

Total anomalous pulmonary venous connection is a rare congenital heart defect that can be diagnosed prenatally and might be very important for perinatal management. In addition to other cardiac abnormalities (levocardia, situs inversus, small left ventricle, double outlet right ventricle, parallel great vessels, and hypoplastic aortic arch), total anomalous pulmonary venous connection of a subdiaphragmatic type was diagnosed during a prenatal echocardiography examination in the second half of pregnancy. Fetal echocardiography monitoring showed no signs of congestive heart failure. The neonate was born at 38 weeks of gestation at our tertiary center. Postnatal echocardiography revealed significant progression in neonatal hemodynamics, and early cardiac surgery, involving repair of the pulmonary veins, pulmonary artery banding, and aortic arch reconstruction, was performed with a good outcome. This case is an excellent example of the value of prenatal echocardiography.

Introduction

Total anomalous pulmonary venous connection (TAPVC) is a congenital heart defect (CHD) that accounts for 1–3% of all CHDs diagnosed prenatally and postnatally. TAPVC may be an isolated defect or occur alongside other cardiac abnormalities, such as isomerism.

TAPVC involves the flow of blood from the pulmonary veins to the right atrium through an abnormal vessel. In some cases, surgery may be required shortly after birth to save neonatal life. Echocardiographic examination is the method of choice for prenatal diagnosis, as it allows visualization of the incorrect vessel connection and assessment of blood flow using color Doppler[(1)]. According to the Polish Registry of Fetal Cardiac Pathology, between 2004 and 2023, TAPVC was diagnosed in only 14 fetuses (0.15%) out of 9,542 pregnancies with CHDs in the Registry. Of all the cases of TAPVC found in the Registry, as many as 12 cases (86%) were diagnosed at the Department of Prenatal Cardiology at the Institute of the Polish Mother's Memorial Hospital in Lodz (www.orpkp.pl).

Materials and methods

This is an analysis of a selected case of a pregnant woman who underwent prenatal echocardiographic examination three times at the Department of Prenatal Cardiology at the Institute of the Polish Mother's Memorial Hospital in Lodz. All examinations were conducted in 2023. The echocardiographic examinations were performed using the VOLUSON E10 ultrasound machine with both transabdominal convex and cardiac probes. All examinations were carried out by experienced echocardiographers.

Case report

A 26-year-old pregnant woman, in her first pregnancy, had gestational diabetes (GDM G1) and hypothyroidism. She underwent a routine obstetrical ultrasound examination in the first trimester. The results of the examination were described as a pregnancy without complications. The NT value at 13 weeks of gestation was 1.58 mm, and the nasal bone was present. There was no tricuspid valve regurgitation. The risk of trisomy based on ultrasound markers was not increased. Biochemical tests, including ß-hCG and PAPP-A, showed values within the normal range. No cardiac problems were observed at this stage of pregnancy.

The congenital heart defect was first suspected in the 20th week of gestation during the second routine obstetric ultrasound. In the 21st week, a physician from northern Poland, certified by the Polish Prenatal Cardiology Society, issued a medical report with the following information: situs inversus, a congenital heart defect in the form of HLHS, and possibly TAPVC. As a result, the pregnant woman was referred to our tertiary center in Lodz. During the course of the pregnancy, three fetal echocardiographic examinations were performed, and perinatal management was planned by our Fetal Team. The basic ultrasound and echocardiographic measurements are presented in Table 1.

The fetal echocardiographic examinations were performed by Dr. Jerzy Węgrzynowski and Prof. Maria Respondek-Liberska.

The first echocardiographic examination at our tertiary center took place at 24.2 weeks of gestation. The male fetus presented with situs inversus and levocardia (Fig. 1). Both the SVC and IVC had the

Tab. 1. Basic ultrasound parameters in three examinations in the analyzed fetus

Selected parameters	Ultrasound and echocardiographicexamination at 24.2 weeks of gestation (LMP)	Ultrasound and echocardiographic examination at 30.5 weeks of gestation (LMP)	Ultrasound and echocardiographic examination at 37.3 weeks of gestation (LMP)
EFW [g]	589 (6th centile)	1511 (17th centile)	3044 (54st centile)
CVPS	10/10	10/10	10/10
Fetal position	Cephalic	Breech	Breech
Placental thickness [mm]	23 (normal range up to 40 mm)	40 (normal range up to 40 mm)	40 (normal range up to 40 mm)
AFI [cm]	20 (normal range: 5–25)	16 (normal range: 5–25)	16 (normal range: 5–25)
AP [mm]	23 (around 50th percentile)	29 (around 50th percentile)	29 (around 50th percentile)
PA [mm]	4.5 (Z-score = 0.10)	7.2 (Z-score = 1.57)	7.2 (Z-score = –0.91)
AO [mm]	2.6 (Z-score = –3.18)	3.3 (Z-score = –3.60)	3.3 (Z-score = –5.99)

EFW – estimated fetal weight; CVPS – cardiovascular profile score; AFI – amniotic fluid index; AP – transverse measurement of fetal heart; PA – pulmonary artery; AO – aorta

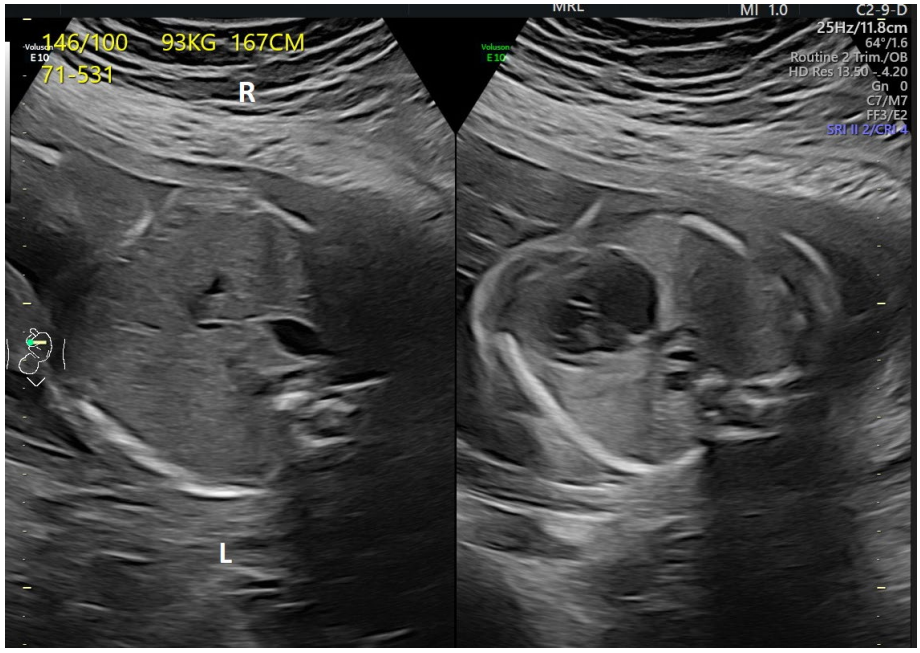

Fig. 1. *Situs inversus with levocardia during prenatal screening at 24.2 weeks of gestation*

same diameter of 2.7 mm at this time of examination. The flow of blood through the foramen ovale was correct.

At the level of the heart, the fetus presented with a small left atrium, an ostium primum atrial septal defect (ASD), a common atrioventricular valve (AVC), and asymmetry in the three-vessel view, with a wide pulmonary artery (PA) compared to a narrow aorta (AO) in a parallel position.

The aortic arch had a 'gothic' shape. The aorta displayed a Y sign, which suggested interruption of the aortic arch (IAA) (Fig. 2, Fig. 3, Fig. 4). The pulmonary vein collector was located behind the wall of the left atrium, and the veins coursed under the diaphragm, indicating an infracardiac type of TAPVC.

In the second examination at 30.5 week of gestation, a normal fetal growth trend was observed, with no signs of congestive heart failure or turbulent flow within the fetal heart.

During the third examination at 37.3 week of gestation, there was no progression, and fetal well-being was confirmed, so there was no necessity for an early delivery. Other projections from the fetal ultrasound examination can be found in Fig. 5 and Fig. 6.

The fetal cardiac condition was discussed during our Fetal Team's meeting, as the pregnant woman decided to deliver at our hospital in Lodz due to the need for neonatal surgery.

After 20 days from the last echocardiographic examination at our Department of Prenatal Cardiology, a caesarian section was performed at 38 weeks of gestation due to breech presentation. The neonatal birth weight was 3,300 g, and the Apgar scores were 5/6/7. The newborn was admitted to the Clinic of Neonatology, Intensive Care, and Pathology of Newborns, where prostaglandin E1 infusion was started in the delivery ward. Postnatal echocardiographic and Angio CT examinations confirmed the prenatal findings. Cardiac surgery was performed on the 7th day of life and involved repair of

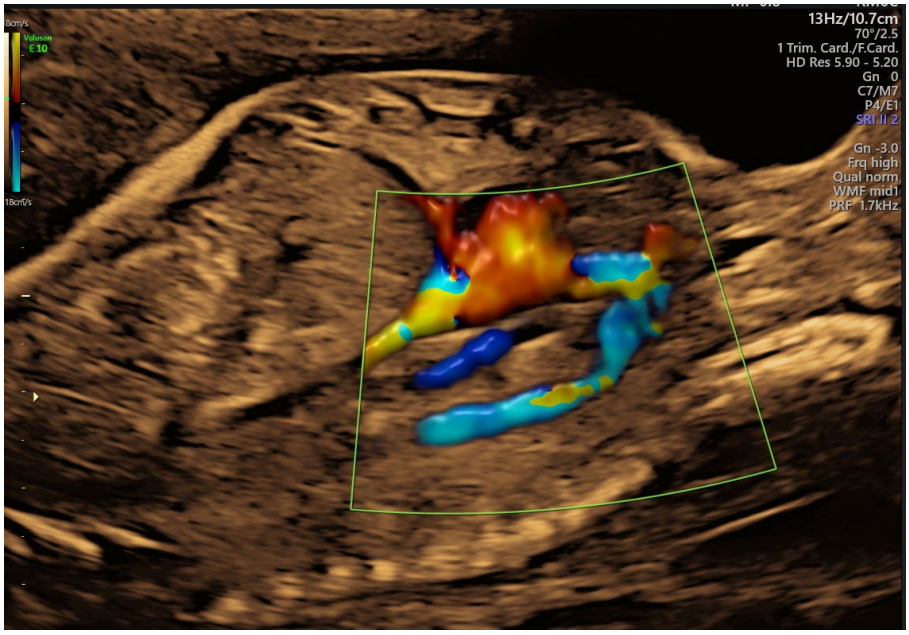

Fig. 2. *Abnormal vessel found during prenatal ultrasound screening at 24.2 weeks of gestation*

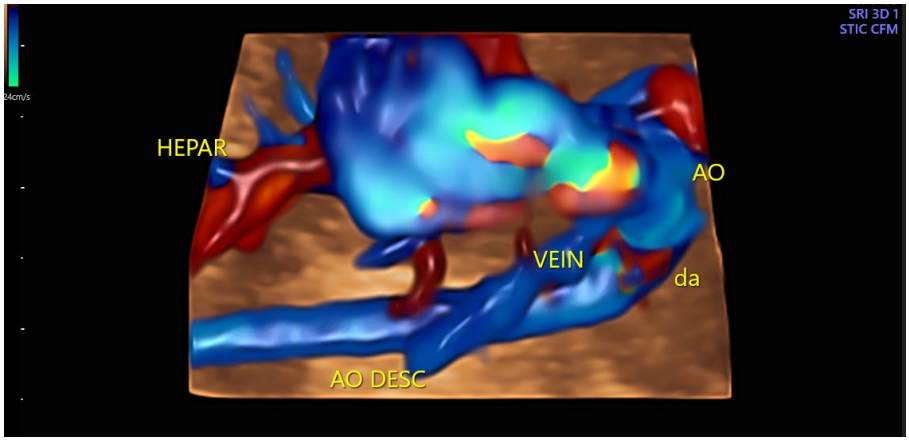

Fig. 3. *Abnormal vessel in 3D presentation at 24.2 weeks of gestation*

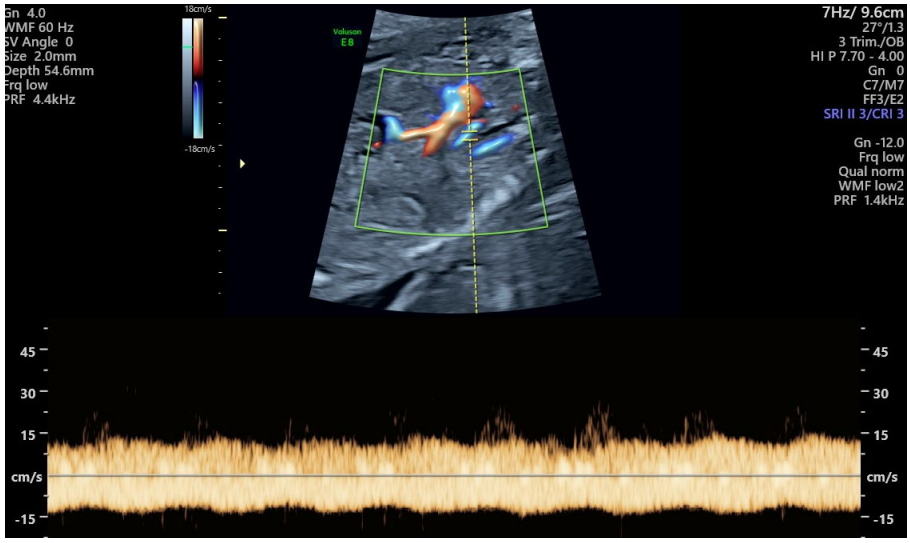

Fig. 4. *Fetal ultrasound image showing an abnormal vessel with typical venous flow of blood – no increased velocity until the 37th week of pregnancy. Ultrasound image from examination at 21.3 weeks of gestation*

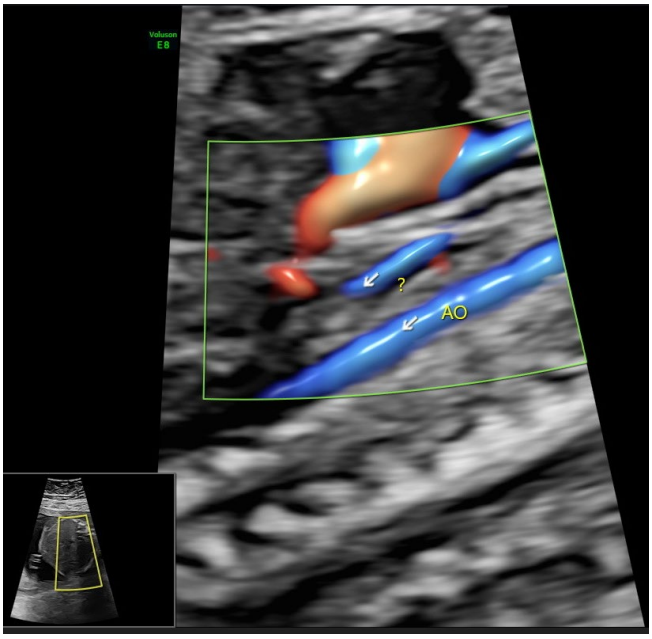

Fig. 5. *Fetal ultrasound image showing an abnormal vessel. Ultrasound image from examination at 21.3 weeks of gestation*

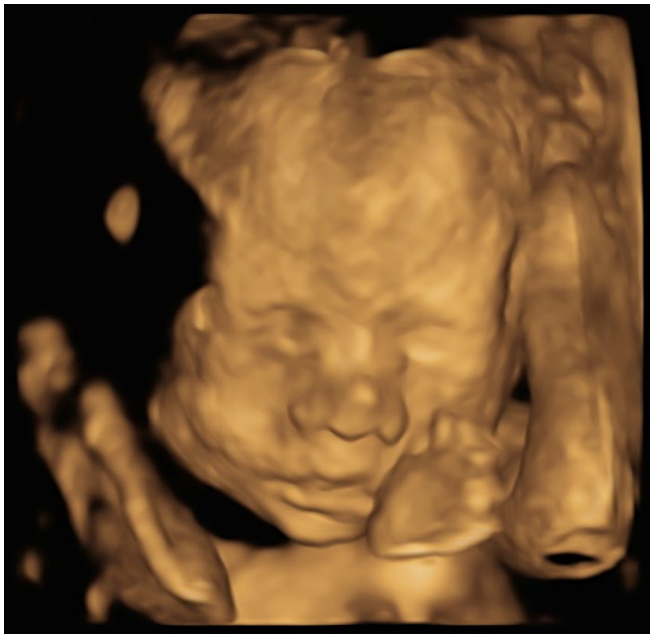

Fig. 6. *3D representation of the fetus during ultrasound examination at 24.2 weeks of gestation*

the pulmonary veins, pulmonary artery banding, and reconstruction of the aortic arch. All prenatal and postnatal data can be found in Table 2.

The neonatal echocardiographic examination showed an abnormal 8 mm-wide vessel within the liver (pulmonary collector) with typical venous flow and increased velocity, suggesting venous obstruction (Fig. 7, Fig. 8).

The measurements were as follows: aortic isthmus – 2.1 mm, VSD – 12.7 mm, ASD I – 2.5 mm, and ASD II – 10.5 mm. A common atrioventricular valve with regurgitation was present. The aorta and pulmonary trunk were positioned in parallel, as seen in L-TGA.

The chest X-ray examination showed an enlarged heart shifted to the left side. The lung fields presented some interstitial and parenchymal densities.

The neonatal angio-CT examination showed the following findings: Type III TAPVC, VSD, ASD, pulmonary veins (upper and lower on both the right side and left sides) forming a common wide collector under the diaphragm with stenosis, draining blood to the liver veins, SV, and a hypoplastic aortic arch. (Fig. 9)

After cardiac surgery, which involved aortic arch reconstruction and main pulmonary artery banding, a follow-up chest X-ray presented uniformly aerated lungs with some interstitial densities (Fig. 10).

Tab. 2. Comparison of prenatal and postnatal echocardiographic diagnosis

Prenatal diagnosis	Postnatal diagnosis
Situs inversus	Situs inversus
Levocardia	Levocardia
ASD	ASD
Infracardiac-type TAPVC	Infracardiac-type TAPVC
SV	SV
Hypoplastic aortic arch	Hypoplastic aortic arch / CoA
IAA	IAA
Hypoplastic left ventricle and small LA	HLHS
Common AV	Common AV
Missed l-TGA as part of prenatal diagnosis	l-TGA
No AV valve regurgitation	AV valve regurgitation

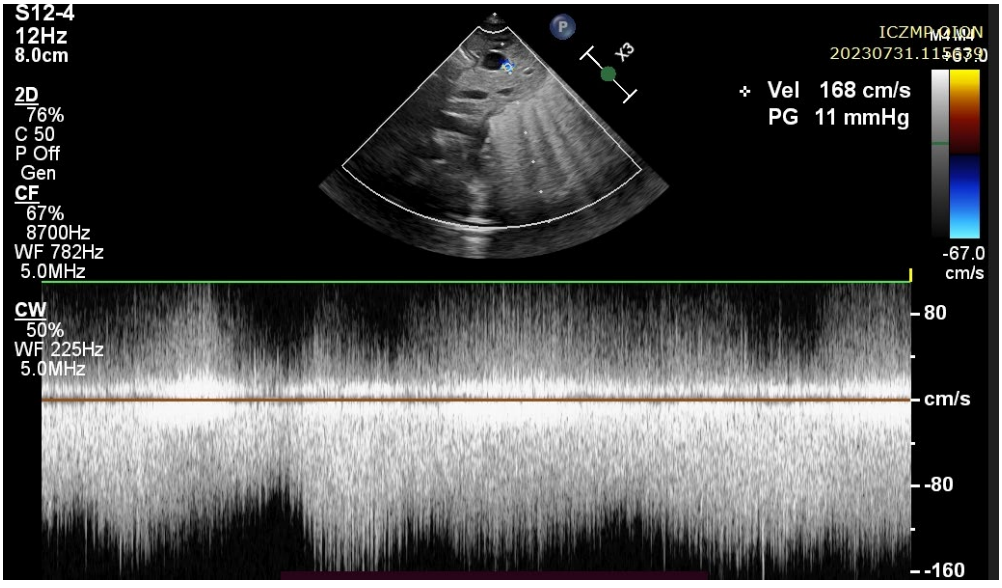

Fig. 7. *Postnatal ultrasound of a neonate showing a vessel with flow characteristics typical of blood flow restriction*

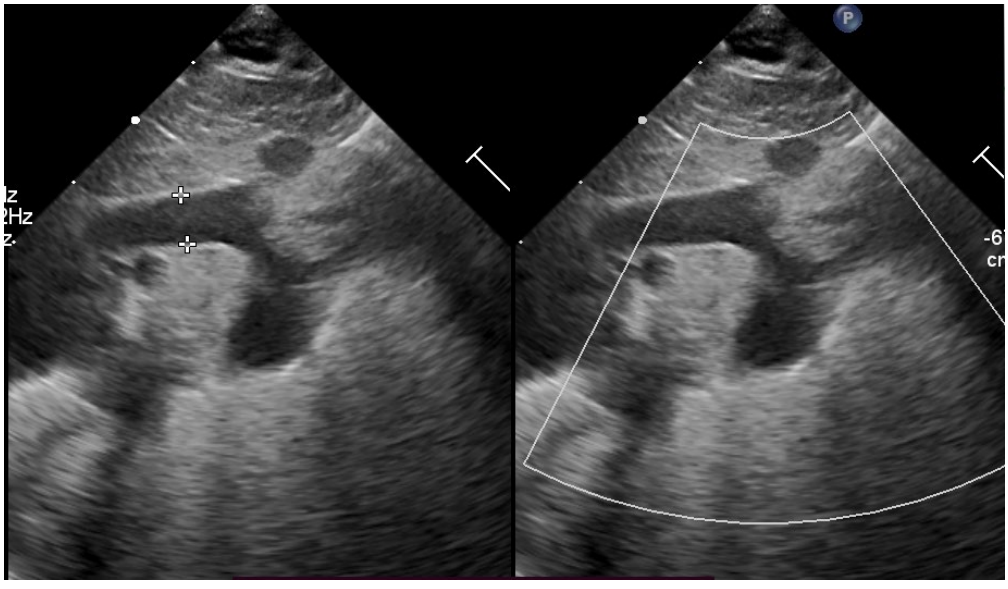

Fig. 8. *Postnatal ultrasound of a neonate with measurement of an atypical vessel*

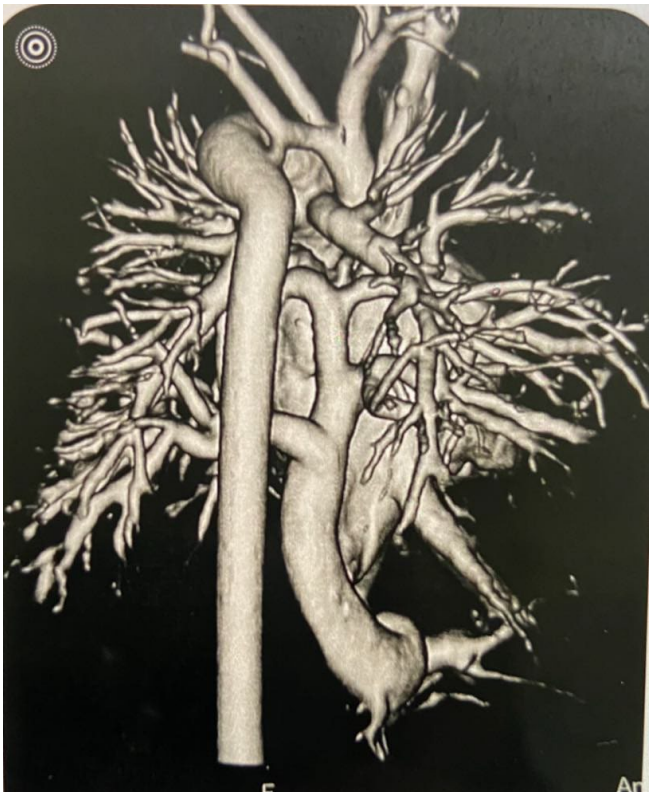

Fig. 9. *Angio-CT of the heart of a newborn with the collector described in prenatal examination reports*

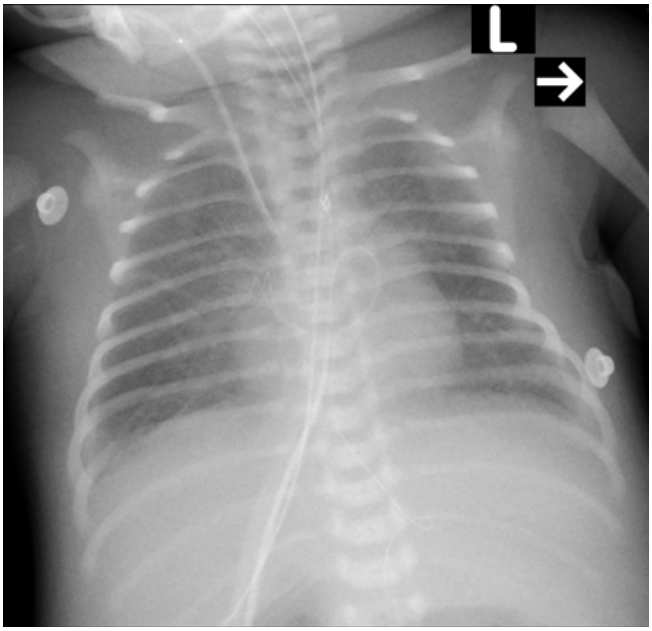

Fig. 10. *X-ray chest of a neonate on the 1st day of postnatal life showing uniformly aerated lungs with some interstitial densities*

Discussion

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart defect caused by the failure of the left atrium to incorporate pulmonary veins during heart embryogenesis, leading to improper connection of the pulmonary veins to the right atrium. There are four types of TAPVC.

- Type I. Supracardiac – the pulmonary veins join together to form a common vein that drains into the superior vena cava (SVC) above the heart. The SVC is the main vein that allows for blood flow from the upper part of the body to the heart. Blood flows to the right atrium through the superior vena cava. This is the most common type of TAPVC.
- Type II. Cardiac – the pulmonary veins drain blood to the right atrium directly or through the dilated coronary sinus.
- Type III. Infracardiac – the pulmonary veins join together to form an abnormal common pulmonary vein that passes through the diaphragm and connects to the inferior vena cava (IVC), portal vein, or ductus venosus, ultimately draining into the right atrium.
- Type IV. Mixed – the pulmonary veins join systemic veins and the right atrium through a combination of the above-mentioned pathways. This is the least common type of TAPVC.

In a newborn, blood from the pulmonary veins flows into the right atrium. Due to excess volume of blood in the right atrium, it becomes enlarged, while the left atrium and left ventricle are small[2–4]. Prognostic factors depend on the blood flow through the pulmonary veins. If the blood flow through the pulmonary veins is obstructed by the presence of narrowed pulmonary veins or a constricted common pulmonary vein, the neonatal outcome is usually poor. A newborn with severe narrowing of the pulmonary veins may suffer from pulmonary hypertension, metabolic acidosis, cyanosis, and heart failure, which may negatively affect their mental and physical development. Additionally, the incidence of upper respiratory tract infections is increased.

The presence of a widely opened foramen ovale and unobstructed ductus arteriosus (DA) can temporarily support circulation, allowing the newborn to maintain relatively stable condition for a short period.

TAPVC is a type of heart defect that requires cardiosurgical intervention. Without surgery, infants typically do not survive beyond their first year without surgery. The results of surgery are satisfactory.

TAPVC is independent of the patency of ductus arteriosus (DA) but if the flow through DA is made possible, a smaller amount of blood volume flows through the pulmonary circulation system, which decreases the risk of developing pulmonary hypertension[4,5].

Early diagnosis is crucial and can be achieved through prenatal echocardiographic examination. The most commonly observed findings in echocardiographic examinations for TAPVC in the 3rd trimester of pregnancy include a small left atrium and left ventricle, as well as an enlarged right atrium and right ventricle. Therefore, it is necessary to focus on the connections of the pulmonary veins to the left atrium. If it is difficult to visualize pulmonary veins, and the wall of the left atrium appears 'smooth', it is necessary to perform a detailed examination to search for an additional pulmonary collector, which may be located behind the left atrium or below the diaphragm. Another clue for TAPVC is a large distance between the left atrium and the aorta. It is also important to look for accelerated blood flow in the collector or in the superior vena cava or inferior vena cava.

In a three-vessels view, a vertical vein visible as a fourth vessel close to the pulmonary artery may suggest the supracardiac type of TAPVC. Another finding in this view is dilatation of the superior vena cava. In the abdominal view, an additional vessel (collector) can be found between the inferior vena cava and the aorta, which suggests the presence of the infracardiac type of TAPVC. The intracardiac type of TAPVC may be manifested as an enlarged coronary sinus (>3 mm in diameter).

When it comes to color Doppler, it should be used for imaging the direction of blood flow and detection of turbulent flow, which suggests obstruction.

In terms of spectral Doppler, TAPVC is characterized by the presence of a continuous monophasic pattern or abnormal pulsatility[4,6–11]. The assessment of maximum, mean, and minimum velocities in the vertical vein seems to be a significant factor in predicting pulmonary venous obstruction[12]. Echocardiographic markers for TAPVC should be excluded in cases of heterotaxy (both right and left atrial isomerism); however, the detection or diagnosis of an isolated anomaly is much more challenging. Despite the fact that fetal echocardiography is a complex examination that requires experience, it remains the gold standard for detecting TAPVC and can help to save neonatal life[13,14,15].

What is worth noting in the presented case is the good fetal condition without turbulent flow present during three echocardiographic examinations, and an increasing venous obstruction after birth, most likely due to diaphragm movements.

Another thing that must be emphasized is the fact that fetal echocardiographic monitoring just before birth plays an important role in assessing fetal condition. In our case, the last examination was performed three weeks before birth.

It is essential that every fetal cardiac center should perform fetal echocardiographic examinations a week before delivery – not only for conditions such as critical aortic or pulmonary stenosis, d-TGA, HLHS, or in conditions involving the restriction of the foramen ovale, but also in TAPVC due to the possibility of venous obstruction just before birth.

Conclusions

Fetal echocardiography is a challenging method for prenatal diagnosis of infracardiac TAPVC, and fetal echocardiographic monitoring should be continued until the very end of prenatal life.

Informed consent statement

Informed consent was obtained from all subjects involved in this study.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

Author contributions

Writing of manuscript: SW. Analysis and interpretation of data: SW. Final acceptation of manuscript: MRL. Collection, recording and/or compilation of data: SW. Critical review of manuscript: JW, MK, IM, AMK, PG, KJ, IS, MRL.

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