● **Case** report

PRENATAL DETECTION AND POSTNATAL VERIFICATION OF UNUSUAL INTRACARDIAC TOTAL ANOMOLOUS PULMONARY VENOUS CONNECTION (TAPVC) IN COMPLEX HEART DEFECT WITH DEXTROCARDIA - CASE REPORT

Authors:
Aneta Krasoń[1], Joanna Płużańska[2], Maciej Łukaszewski[3], Jadwiga Moll[4], Maria Respondek-Liberska[2,5]

1.Prenatal Cardiology Department, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland. Individual training in 2018. 2. Department of Prenatal Cardiology, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland 3. Department of Medical Imaging, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland 4. Department of Cardiology, Polish Mother's Memorial Hospital Research Institute, Lodz, Poland 5. Medical University of Lodz, Department of Diagnoses and Prevention Fetal Malformations, Lodz, Poland

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Abstract

Prenatal diagnosis of total anomolous pulmonary venous connection (intracardiac) was diagnosed in fetus with dextrocardia and complex heart defect, which allowed fetal echocardiography monitoring, planning the time and place for delivery as well as early cardiac surgery. The differences between prenatal and postnatal evaluations were underlined. Despite life treathening condition neonate was asymptomatic without any heart murmur for the first 3 days after delivery.

Key words: *congenital heart disease, total anomalous pulmonary venosus connection (TAPVC), restriction, fetal echocardiography*

INTRODUCTION

Total Anomalous Pulmonary Venous Connection-TAPVC is a congenital heart defect, characterized by abnormalities in the pulmonary venous return, with connections to systemic veins or right atrium.

The point of connection between pulmonary veins and systemic venous circulation is the basis for anatomical classification. Additional defects are diagnosed in 30% of newborns with TAPVC, defects such as VSD, ASD, common vetricle are the most frequently diagnosed. TAPVC accounts for 1-3% of live births with congenital heart disease. Neonates with TAPVC are usually in good clinical condition just after delivery, depending on the type of TAPVC and level of restriction on the collector but serious deterioration, resulting mainly from pulmonary oedema and hipoksemia begins within hours or days.

The prenatal detection and diagnosis of this defect is extremely important, it allows for in utero transportation of gravida to tertiary center, optimatal time of delivery, perinatal management and preparation of "Heart Team" how to make use of time before neonatal deterioration.

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Prenatal diagnosis of TAPVC is possible however rare even in tertiary fetal cardiology centers [1,2,3,4].

CASE REPORT

21- year old prima gravida, first trimester obstetrical scan at 12 wks of gestation was reported as normal. At 25 weeks of gestation, abnormal fetal four chamber view was detected and HLHS syndrome was suspected. Because of this the pregnant woman and her fetus were referred to our center. Based on targeted fetal echocardiography examination critical complex congenital heart disease was diagnosed[5,6]. The apex of fetal heart was directed to the right side meaning dextrocardia. HA/CA 0,3- was normal. Defect was of univentricular morphology with arising aorta and pulmonary artery. There was mitral atresia, hypoplastic left ventricle, large VSD, and intracardiac type TAPVC was suspected, due intracardiac "bump" at the level of the left atrium, abnormal course of pulmonary veins behind the wall of the left atrium with abnormal turbulent venous blood flow at the level of the left atrium - velocity 120 cm/sec and prominent pulmonary veins. It was suspected that the pulmonary veins drained together into the heart at the

Corresponding author: asia.pluzanska@yahoo.com

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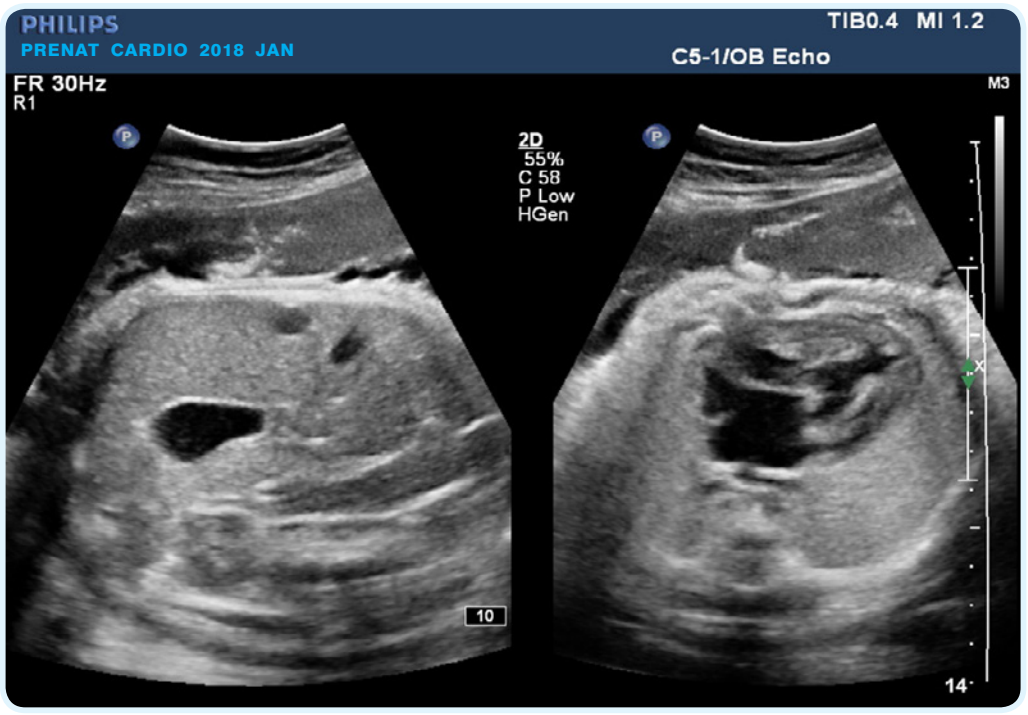
level of the atria. Additional interrupted aortic arch type B (IAA) was diagnosed, with disproportion between great vessels, (PA>Ao). FHR was regular 140 beats/min, there was no fetal arrythmia. Blood flow through the single A-V valve was normal without insufficiency. Peripheral dopplers were normal. No extracardiac malformations were detected. Fetal echocardiography was performed every 3 weeks in our Department confirming fetal well being despite echocardiographic signs of restriction at the level of the collector and grossly enlarged pulmonary veins.



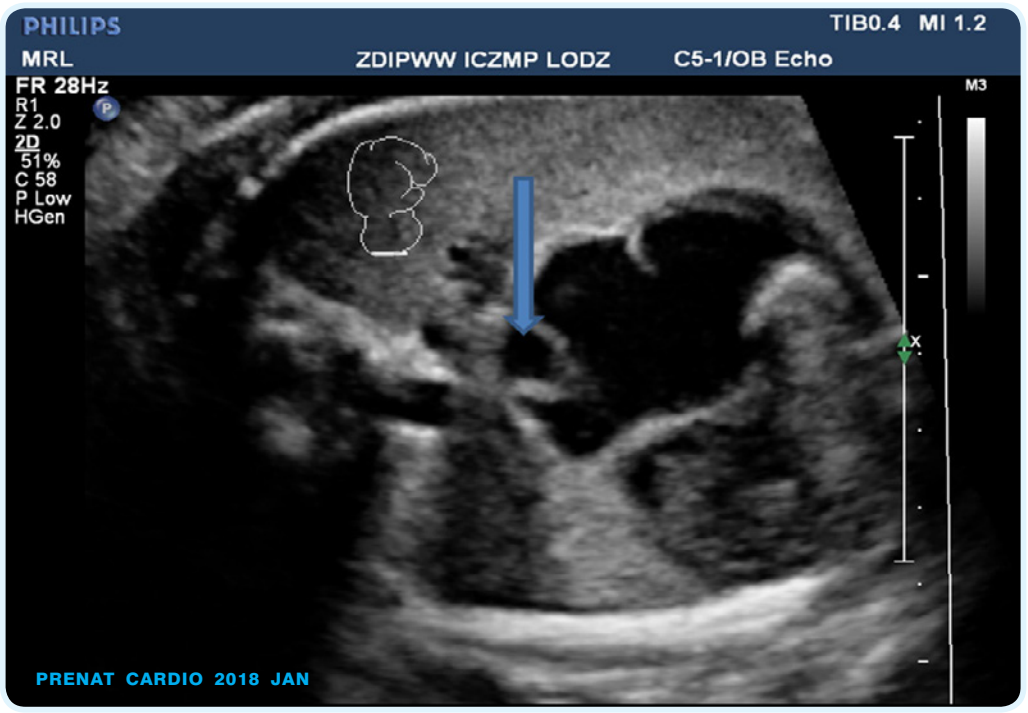
Fot. 1. Our echo exams- dextrocardia, abnormal 4 chamber of view, stomach on the left

At 38 weeks of gestation the boy was delivered by cesarean section, the birth weight was 3360g and Apgar score 10/9 points. Neonate had no heart murmer, no cyanosis, and no clinical symptoms of congenital heart defect, his oxygenation was 93% and HR 140/min, despite RTG signs of pulmonary oedema. His heart apex pulsation was on the right side of the chest. There was spontaneous breathing without apnoe for the next 3 days. Prostin infusion was provided since the first day of postnatal life, however on the day 4th baby needed intubation and mechanical ventilation.

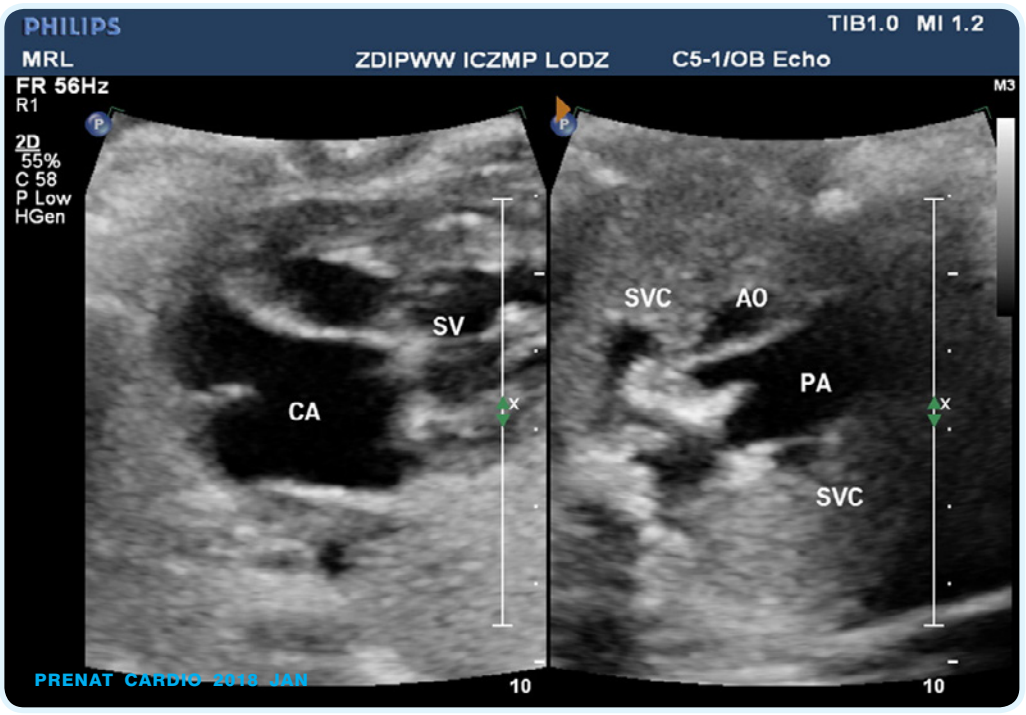
Neonatal echocardiography and angio-CT confirmed prenatal diagnosis (dextrocardia + mitral atresia + hypoplastic left ventricle + IAA type B and restriction in pulmonary venous drainage with enlarged pulmonary veins) and the baby was qualified for emergency 1st stage cardiac surgery: removal of obstruction at the atrial level, banding of the pulmonary arteries to prevent pulmonary overflow and stent insertion into the ductus arteriosus with next stage arch reconstruction after clinical stabilization.

DISCUSSION

At 10 weeks of gestation the fetal heart can be examined usually by transvaginal ultrasound, but pulmonary vein assessment is not part of routine obstetric scanning for

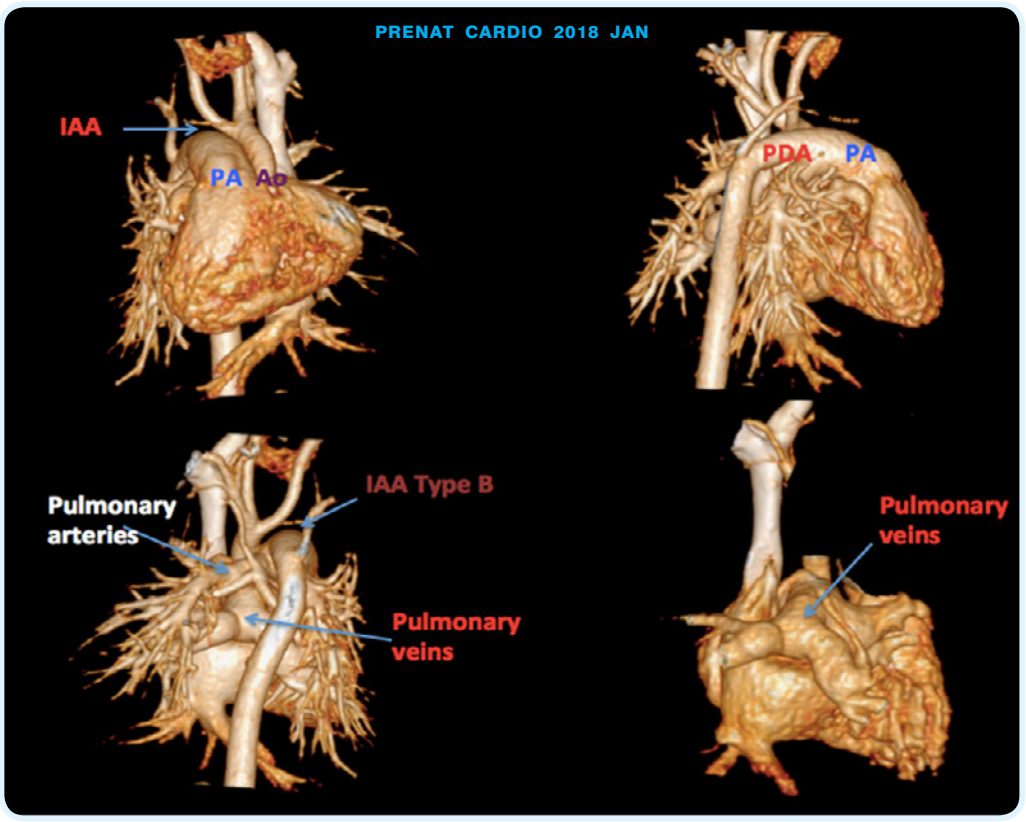


Fot. 2. TAPVC to the upper part of the atrium (arrow), wild connection between left and right atrium, common atrioventricular valve, small LV and large RV



Fot.3. Large septal defect, haemodynamic SV and common atrium (on the left), persistent bilateral SVC in mediastinum

fetal cardiac defects. It is also difficult in fetal cardiac tertiary centers[7-9]. The best imaging of the fetal heart is usually achieved between 18-22 weeks of gestation. Screening fetal heart examination protocols include evaluation for 4 chamber view and three vessels in the upper mediastinum, and are recommendend by American, European and Polish Societies[10,11].



Fot. 4 .Agnio CT

Fetal echocardiography allows for the diagnosis of most common cardiac pathologies such as hypoplasic left or right ventricule, atrioventricular canal, truncus arteriosus, tetarlogy of Fallot, transposition of the great arteries, etc[12]. However, there are anomalies such as TAPVR, which occur rarely, and that become diagnostic challenges. Evaluation of the fetal pulmonary veins is one of the most challenging parts of fetal echocardiography examination [1,2,13-17].

Total anomalous pulmonary venous connection (TAPVC) is a rare isolated congenital heart disease (CHD), accounting for 1-3% of all cardiac malformations. It is characterized by an anomalous connection of the pulmonary veins draining via one or more systemic veins to the right atrium, instead of the left atrium, or there may be abnormal connection to the coronary sinus[17].

There are several different possible types of pulmonary venous (PV) connections including supracardiac- type I- the four pulmonary veins merge into confluence posterior to the left atrium and connect via a vertical vein to the left brachiocephalic vein, which drains into the superior vena cava. Cardiac type 2- the pulmonary veins connect directly to the coronary sinus, which becomes dilated or connect into the posterior wall of the atrium. Infracardiac-type3- the four pulmonary veins form a confluence behind the atria, or mixture of these- type 4-is rare and involves a variety of pulmonary venous drainage where the left pulmonary veins drain

to the left innominate vein into the coronary sinus or directly to the right atrium.

Supracardiac anomalous pulmonary connection is the most common and accounts for about 45% of cases.

TAPVC may be present without pulmonary vein obstruction (which has much better prognosis) or may be present with an obstruction (meaning worse prognosis)[18]. Hemodynamic consequence of anomalous pulmonary veins connections has significant influence on the neonatal haemodynamic state as various degrees of mixing of pulmonary and systemic blood occurs, which leads to pulmonary oedema and cyanosis. TAPVC can be isolated or can occur in association with other cardiac anomalies. One of the most common associated cardiac abnormalities is atrioventricular septal defect, single ventricle, hypoplastic left heart syndrome and interrupted aortic arch.

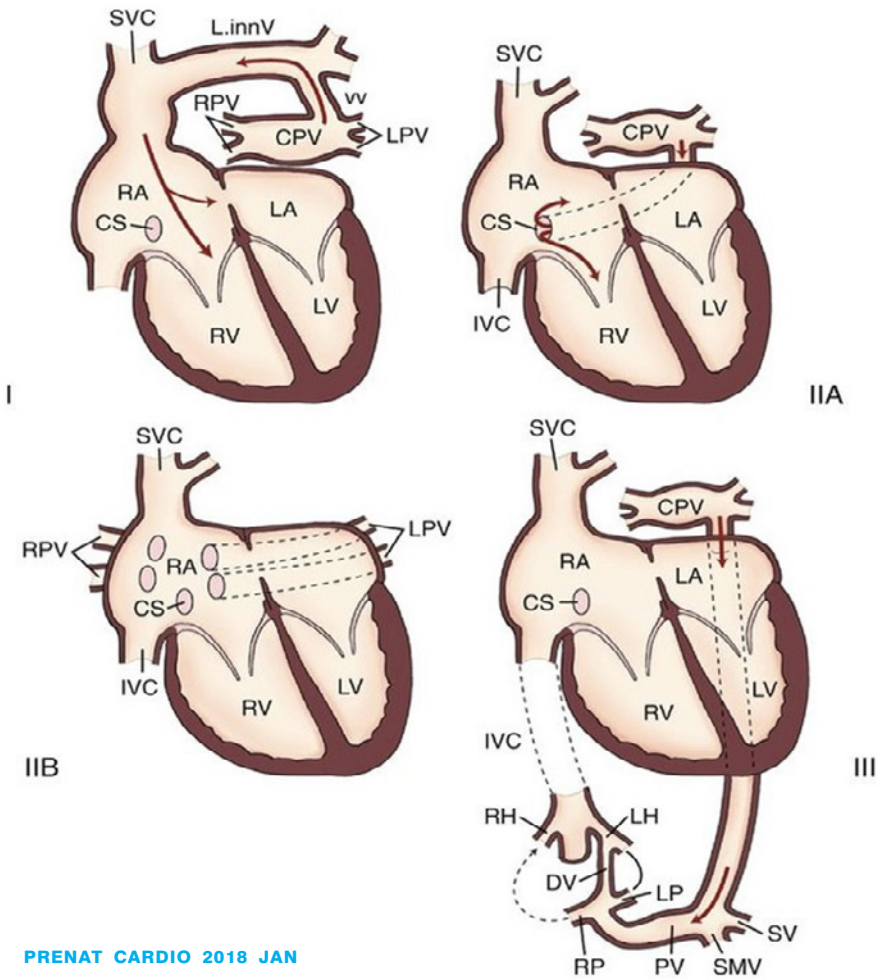


Fig.1. Schematic drawing of typical 3 types of abnormal pulmonary veins connections in hearts with levocardia.

In addition to morphology during fetal heart evaluation it was also very important to check the blood velocity in the pulmonary veins. Based on our experience and data from the literature a poor sign in terms of neonatal prognosis is an increased velocity suggesting pulmonary venous obstruction. So far we have had no survivors in cases of pulmonary venous blood flow >100 cm/sec[18]. In the presented case the maximum blood flow velocity was increased with small changes from one examination to another suggesting both functional and structural pulmonary vein stenosis (Table 1).

	1st fetal echo exam	2nd fetal echo exam	3rd fetal echo exam
EFW (g)	1599	2460	2879
HA/CA ratio	0,3	0,3	0,3
Pulmonary blood flow in veins in cm/sec	110	90	120

Table 1. Maximum blood flow velocity in 3 examinations

However postnatal evaluation in this case showed some remarkable differences such as: from prenatal point of view there was a wide communication between atria, whereas postnatal echo revealed an atrial septum.

Prenatally we suspected typical intracardiac total anomalous venous return with restriction at the outlet of the collector, however postnatal scans showed pulmonary veins drained into the left atrium instead of right atrium in order to fullfil the classical definition, which in cases of single ventricular heart morphology and no restriction at the level of collector would not require correction of TAPVC (Table 2). The patient had to undergo surgical atrial septostomy instead of the Rashkind procedure (which is the typical step in the hybrid procedure) because of the unusual structure of the left atrium. Cardiac surgery confirmed additional tissue dividing the left atrium, which gave the impression that the veins formed a collector and not a case of restriction at the level of the foramen ovale, and therefore interventional treatment was not an option.

	Prenatal diagnosis	Postnatal diagnosis
Heart position in the chest	Dextrocardia	Dextrocardia
Atrial septum	Small almost not visible "Common atrium" with two different appandages	Atrial septum present
Pulmonary venous drainage	Abnormal	Abnormal
Type of drainage	To the left atrium or to the right atrium?	To the left atrium
Pulmonary veins	Not very much dilated but well visible behind the left atrial wall	Very dilated behind the left atrial wall
Ventricles	Dominated RV, small LV connection via large VSD	The same
Ao	Present	Present
Ao arch	Interrupted type II	Interrupted type II
Signs of congestive heart failure	None	None (for 48 hours)

Table 2. Elements of prenatal and postnatal diagnosis in presented case

In this case complex fetal heart malformation, without extracardiac malformation, was detected by obstetrican and allowed fetal echocardiography monitoring in tertiary fetal cardiology center since 27 weeks of gestation to term delivery in referral hospital with obstetrical, neonatal, pediatric cardiology and cardiac surgery departments which allowed planned and early treatment.

Our experience shows that neonate with isolated TAPVC and collector restriction present with rapid clinical deteriotion and can be succesfully repaired on the second day after delivery after previous prenatal detection and diagnosis[16]. Whereas on the basis of this case, restriction of pulmonary venous drainage in case of univentricular morphology did not show any neonatal clinical symptoms in the first few days after delivery, despite life treathening condition. This is original observation from our case report and was not underlined before.

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Division of work:

A. Krasoń - first draft, literature search

J. Płużańska - work with the manuscript

M. Łukaszewski - postantal angio-CT reconstruction, work with the manucript

J. Moll - work with manucsript, final version

M. Respondek-Liberska - author of the fetal echocardiography, work with the manuscript, final version

All authors certify that there is no actual potential conflict of interest in relation to this article.