PRENATAL DIAGNOSIS OF TAPVC ON MONDAY, DELIVERY OF TUESDAY AND CARDIAC SURGERY AT WEDNESDAY - A MODEL OF PERINATAL CARE IN 3RD TRIMESTER IN CASE OF FETAL/NEONATAL CRITICAL HEART DEFECT IN TERTIARY CENTER.

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is a congenital heart defect (CHD), in which all pulmonary veins connect to the systemic veins or to the right atrium/coronary sinus instead of the left atrium. We present a case report of fetus with prenatally diagnosed isolated infracardiac type of TAPVC in 38th week of gestation. In fetal echocardiographic examination performed in the Department of Prenatal Cardiology, the fetus presented lack of visible pulmonary veins connection to left atrium, abnormal venous confluence behind left atrium, additional vein leading from abdominal cavity to mediastinum and abnormal smooth Doppler blood flow in pulmonary confluence. The accurate prenatal diagnosis allowed to deliver the neonate at term, in tertiary center one day after diagnosis, and to perform surgical reposition of pulmonary veins the following day. The neonate was referred home in a good condition after 28 days of hospitalization. This case is a good example of the value of the 3rd trimester echocardiography.

Key words: *total anomalous pulmonary venous return, prenatal diagnosis, fetal echocardiography*

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a congenital heart defect (CHD), in which all pulmonary veins connect to the systemic veins or to the right atrium/coronary sinus instead of the left atrium. TAPVC is a relatively rare pathology. According to National Database for Fetal Cardiac Anomalies in Poland for years 2004-2015, TAPVC was diagnosed in 7 of 7106 registered fetuses. Among those cases, there were 5 cases of TAPVC associated with other cardiac defects, 1 false positive case and only 1 case of isolated TAPVC. We would like to present this case of isolated TAPVC, diagnosed in the Department of Prenatal Cardiology of Polish Mother's Memorial Hospital in Lodz as an example of model perinatal care in case of critical heart defect detected prenatally.

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CASE REPORT

A 20 years old gravida 2 para 1 (first pregnancy ended with miscarriage in 7th week of gestation), with no relevant medical conditions, was referred to Department of Prenatal Cardiology in 38th week of gestation. Until 31st week of gestation, the gravida had 4 obstetric ultrasound examinations, which revealed no cardiac or extracardiac abnormalities. NT at 13th week of gestation was 1,5 mm. The 5th and 6th ultrasound examinations performed in 35th week of gestation detected disproportion between pulmonary artery (PA) and ascending aorta (Ao asc) in favour of PA and disproportion between heart ventricles in favour of right ventricle (RV). The fetal echocardiographic examination performed in our unit in 38th week of gestation revealed a single male fetus in cephalic longitudinal position. The biometry was

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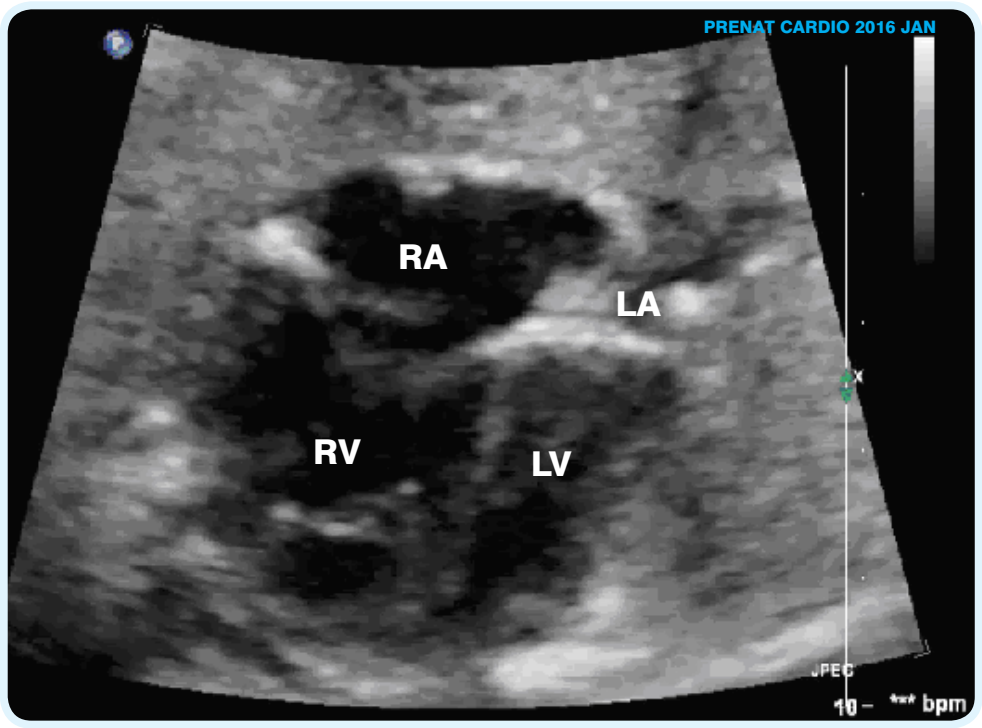
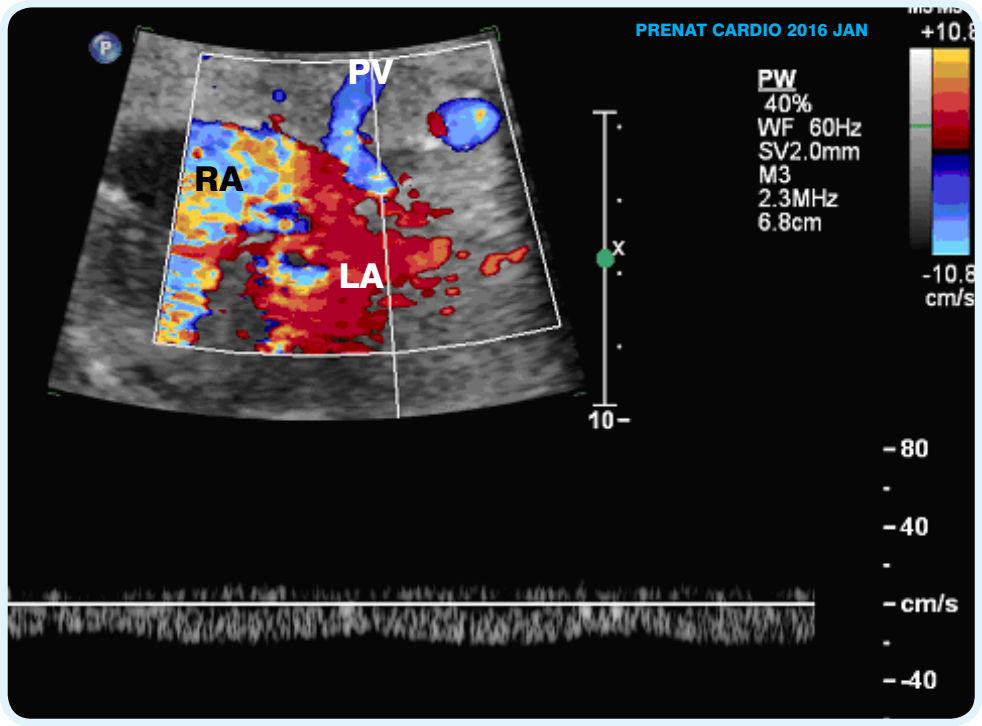


Fot 1: 4 chamber view of the fetal heart with slight disproportion at the level of atria and ventricles



Fot 2: Pulsed Doppler at the level of pulmonary vein "enetring" to the left atrium with very unusual pattern - no pulsation... no connection to the left atrium...no reflection of the changes of the left atrium blood pressure....

appropriate for gestational age. Fetus presented situs solitus, levocardia and no extracardiac abnormalities. The heart axis was 60°, HA/CA 0.33, AP diameter was 44 mm. A discrete disproportion between ventricles in favour of RV was confirmed. Heart atria were symmetrical, foramen ovale was wide, with right to left flow (Fot. 1). Connection of pulmonary veins to the left atrium was not visible. Instead, venous confluence behind left atrium was revealed by Colour Doppler, with abnormal monophasic flow in Spectral Doppler (Fot.2). Left and right ventricle outflow tracts were described as normal. In three vessels view, the examination showed a disproportion between pulmonary artery (7,5 mm) and aorta (5 mm). The additional vertical venous vessel was observed in mediastinum. It was parallel to descending aorta and was draining blood from fetal abdomen. The presence of pathological confluence of pulmonary veins and abnormal vertical vein leading from abdominal cavity to mediastinum indicated diagnosis of infracardiac type of TAPVC (Fot. 3). In long axis view, the examination revealed hypoplastic aortic arch with reversal flow form ductus arteriosus, meaning that the observed defect was ductus dependent.

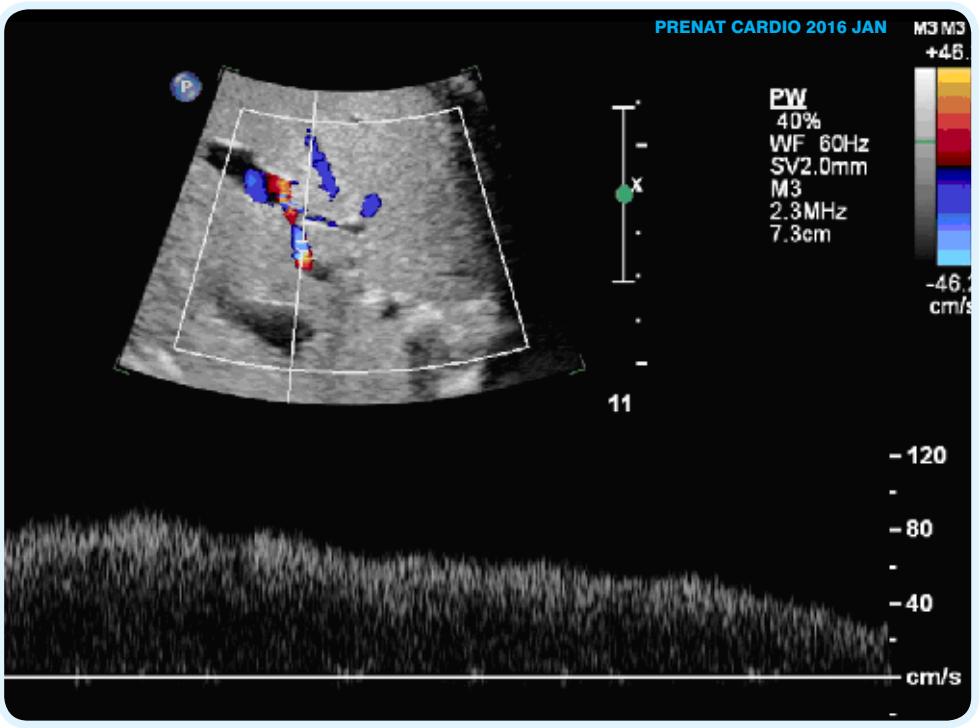
The same day gravida was referred to Obstetric Department of Polish Mother's Memorial Hospital for delivery. The next day, male neonate was delivered by caesarean section. The birth weight was 3120g and Apgar score was 8/8. Prostin iv was administered immediately after delivery. Diagnosis of infracardiac TAPVC was confirmed postnatally by neonatal echocardiography. Next day after delivery, the surgical reposition of pulmonary veins was performed in Department of Cardiac Surgery of our Institute. After the surgery, neonate was referred for further care to Intensive Care Unit. During the first week in ICU, neonate needed mechanical ventilation and administration of catecholamines. After 8 days of treatment, neonate was able to maintain respiratory and cardiovascular stability without invasive ventilation and catecholamines. The control postoperative echocardiographic examination revealed connection of pulmonary veins to left atrium, with PV Vmax 117 cm/s. Disproportion of ventricles in

favour of RV sustained, however contractility of both ventricles was described as normal. After 43 days of hospitalization, neonate was referred home in good condition.

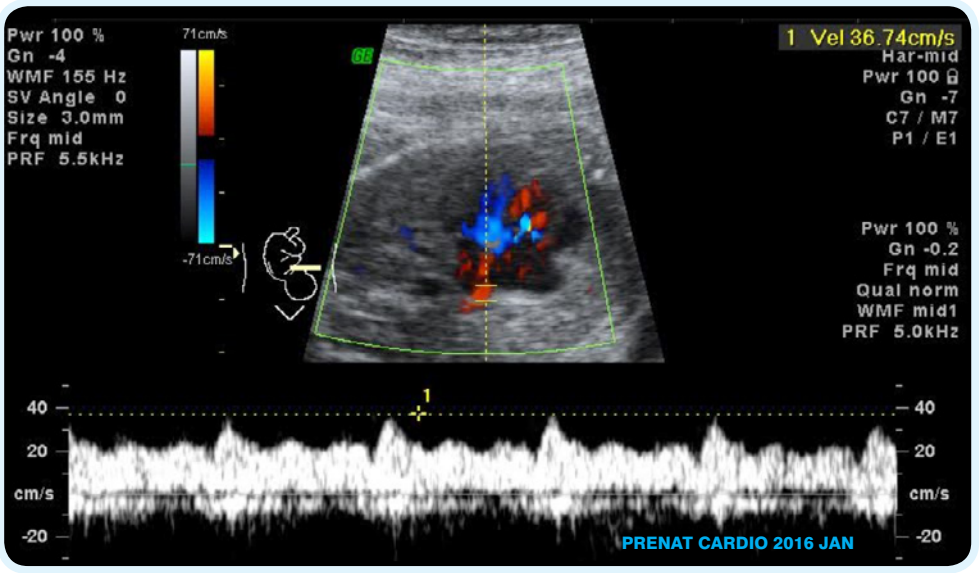
DISCUSSION

TAPVC is categorized into 4 types, depending on the location of abnormal connection of pulmonary veins (PV):

- supracardiac, in which PV connect to superior vena cava via a common vein (44%-48%)
- intracardiac, in which PV connect directly to right atrium or coronary sinus (16%-21%)
- infracardiac, in which PV connect to portal vein through common vertical vein leading to abdominal cavity (20%-26%)
- mixed (9%-12%) [1,2,3].



Fot 3: Pulsed Doppler at the venous vessel beflow diaphragma...again no pulsation...no blood pressure changes...the same pattern as in Fot 2...the same vessel?



Fot. 4 Pulsed Doppler of pulmonary veins showing normal 3 phasic blood flow related to different and changing blood pressure in left atrium in a healthy fetus in a similar gestational age

It may occur as the isolated defect or be a part of a complex CHD. It is commonly associated with right atrial heterotaxy[4,5]. TAPVC is a rare pathology, especially in isolated form, with only a few cases described in literature so far (Table 1)[6,7,8]. In case of TAPVC, the pulmonary venous obstruction may occur. It is present in 48%-55% of the cases and is more frequent in supracardiac and infracardiac types, due to the longer and more complicated way that blood needs to pass in order to reach the heart[9,10]. In order to prevent the increase of pulmonary blood pressure and consequent decrease of blood saturation leading to neonatal death, neonates with TAPVC and pulmonary venous obstruction should be delivered in a tertiary center and need to undergo a surgical intervention during the first hours after birth. This implicates, that TAPVC with pulmonary venous obstruction is a severe urgent CHD, according to the CHD classification by by Slodki et al.[11] The need of urgent intervention in this group of infants highlights the importance of prenatal diagnosis of TAPVC. Our experience so far had shown, that obstructed TAPVC associated with other heart abnormalities, despite good clinical condition of the newborns on first day of postnatal life, was a lethal condition on the following day[12]. There are several specific ultrasound findings, suggesting diagnosis of TAPVC. The most significant ones are: lack of visible connection of pulmonary veins to the left atrium, a visible venous confluence on four chambers view and the presence of vertical vein. The asymmetry of heart chambers and other abnormal vertical vessels in three vessels or abdominal view may also be present[13,14].

First author	Year of publication	Time of diagnosis	2D Ultrasound findings	Color Doppler findings	Extracardiac malformations	Delivery	Menagement	Follow up
D. Laux	2013	37 hbd	Vertical vein, PV confluence, small LA, asymmetry of ventricles	Normal		37-40 hbd	Surgical repair within the first 24 h	Alive and well
D. Laux	2013	26 hbd	Vertical vein, PV confluence	Normal			Surgical repair within the first 24 h	Alive and well
D. Laux	2013	25 hbd	Vertical vein, PV confluence, asymmetry of ventricles, dilated SVC	PV obstruction suspected: aliased flow in vertical vein, high-velocity continuous flow in Pulsed Doppler			Postponed surgical repair	Alive and well
D. Laux	2013	25 hbd	Vertical vein, PV confluence, small LA, dilated SVC	PV obstruction suspected: aliased flow in vertical vein, high-velocity continuous flow in Pulsed Doppler			Surgical repair within the first 24 h	Alive and well
D. Laux	2013	25 hbd	Vertical vein, PV confluence, asymmetry of ventricles, dilated SVC	Normal			Postponed surgical repair	Alive and well
D. Laux	2013	28 hbd	Vertical vein, PV confluence, small LA, dilated SVC	PV obstruction suspected: aliased flow in vertical vein, high-velocity continuous flow in Pulsed Doppler			Surgical repair within the first 24 h	Alive and well
D. Laux	2013	32 hbd	PV confluence, asymmetry of ventricles, dilated coronary sinus	Normal			Postponed surgical repair	Alive and well
D. Laux	2013	31 hbd	Vertical vein, PV confluence, small LA, asymmetry of ventricles	Normal			Surgical repair within the first 24 h	Alive and well
D. Laux	2013	24 hbd	Vertical vein, PV confluence, small LA, asymmetry of ventricles, SVC dilation	PV obstruction suspected: aliased flow in vertical vein, high-velocity continuous flow in Pulsed Doppler			Surgical repair within the first 24 h	Died in 6 weeks
D. Laux	2013	34 hbd	Vertical vein, PV confluence	Normal			Surgical repair within the first 24 h	Alive and well
K. M. Law	2007	19 hbd	RA and RV dilatation, absent PV connection to LA, additional vein in mediastinum	Turbulent flow in additional vein				Termination in 20 hbd
K. M. Law	2007	27 hbd	RA and RV dilatation, absent PV connection to LA, additional vein in mediastinum	Monophasic flow in PV	NT 4 mm in 1st trimester, normal karyotype	35 hbd	Emergency surgical repair	Neonatal death in postoperative period
L D Allan	2001	20 hbd	Ventricles disproportion in favor of RV, great arteries disproportion in favor of PA, PV connection to coronary sinus		Abnormal karyotype, multiple malformations	No data	Surgical repair in 3rd month of life	Death in 1st year of life, due to extracardiac malformations

Table 1: Reported in publications cases of prenatal TAPVC and their follow-up /part 1/

L D Allan	2001	20 hbd	Ventricles disproportion in favor of RV, great arteries disproportion in favor of PA, PV connection to coronary sinus		Abnormal karyotype, multiple malformations	No data	Surgical repair in 3rd month of life	Death in 1st year of life, due to extracardiac malformations
L D Allan	2001	20 hbd	Dilated coronary sinus, small disproportion in favor of right heart		Pericardial effusion	No data	Successful surgical repair	No data
L D Allan	2001	17 hbd	Ventricles disproportion in favor of RV, great arteries disproportion in favor of PA		Hydramnios, pleural and pericardial effusion	34 hbd		Death in first few hours after birth
L D Allan	2001	19 hbd	Heart chambers disproportion in favor of right heart, great arteries disproportion in favor of PA, dilated SVC			Delivery in cardiac center	Surgical repair	The neonate died at surgical repair

Table 1: Reported in publications cases of prenatal TAPVC and their follow-up /part 2/

M. Respondek-Liberska et al. described a case of partial anomalous pulmonary veins connection to the superior vena cava, with the dilated superior vena cava and venous flow increased to 90 cm/s[15]. Kawazu Y. et al. described a novel and efficient ultrasound marker of TAPVC, the "post-LA space index". It has been proved, that if the ratio of the descending aorta-left atrium distance to the diameter of descending aorta is higher than 1.27, the diagnosis of TAPVC is confirmed with a sensitivity of 100% and specificity of 99%[16]. Doppler echocardiography may also be useful in diagnosis of TAPVC. In normal pulmonary veins flow there is 3 phasic blood flow related to different and changing blood pressure in left atrium (Fot. 4). The majority of fetuses with TAPVC have abnormal waveforms of blood flow in pulmonary veins in spectral Doppler. The turbulent flow with high velocity through the vertical vein is an important marker of obstructed TPVC[13,14]. However, in our case the venous blood flow was "stable" and did not reflect the blood pressure in left atrium due to lack of such connection. This observation, we do believe, was not reported before. More advanced ultrasound technology, such as four-dimensional echocardiography with B-flow imaging and spatiotemporal image correlation, may be a source of additional information about the anatomic details of TAPVC[17]. Despite of the advance in fetal echocardiography, detection and diagnosis of TAPVC in a fetus is still a challenging task. The study by Olsesn R. et al. showed, that all the cases of isolated TAPVC were overlooked in the fetal screening ultrasound examination[18]. In the group of 95 neonates with TAPVC described by Laux D. Et al., only 10 had a prenatal diagnosis confirmed by fetal echocardiography expert[6]. In the large study group of 424 cases of TAPVC described by Seale A. N.et al., only 8 (1,9%) were diagnosed prenatally. These studies prove, that screening fetal ultrasound is insufficient in detecting and diagnosing TAPVC, therefore fetuses with the increased risk of TAPVC should be examined by fetal echocardiography expert. The study by Seale A.N. et al. suggests, that the factor of the increased risk of TAPVC is a family history of CHD. Among the 422 lifeborn infants with TAPVC 17 (4%) had a relative witch CHD, and 6 of those 17 cases had a sibling with TAPVC[19]. Nowadays, due to the advancement of pediatric cardiac surgery and postoperative care, the children with isolated TAPVC without PV obstruction have a good prognosis. The mortality of surgically treated patients ranges from 3% to 19%[1,2,3,20,21]. Cases of isolated TAPVC presented in table 1, confirm that early cardiac surgery, preceded by prenatal diagnosis, usually results with good outcome[6,7,8]. The mortality is significantly higher in the group of patients with heterotaxy syndrome or other major heart defects, and ranges form 38% to 63%[2,3,5,20]. Other factors contributing to unfavourable prognosis are: earlier age and lower weight at surgery, pre and postoperative pulmonary venous obstruction, cardiac connection type and single ventricle physiology[1,2,3,9,10,20]. The experience of our institution indicates, that an isolated subdiaphragmatic anomalous pulmonary venous return could be detected and diagnosed prenatally in fetal cardiology center and as a critical cardiac condition can be electively delivered and without delay undergo rescue cardiac surgery. In busy obstetrical and cardiac surgery departments, the prenatal echocardiographic findings were appreciated and good cooperation of medical team allowed for favourable postnatal outcome. This case is also a good example of the value of the 3rd trimester echocardiography.

References

1. Seale AN, Uemura H, Webber SA, Partridge J, Roughton M, Ho SY, McCarthy KP, Jones S, Shaughnessy L, Sunnegardh J, Hanseus K, Berggren H, Johansson S, Rigby ML, Keeton BR, Daubeney PE; British Congenital Cardiac Association. Total anomalous pulmonary venous connection: morphology and outcome from an international population-based study. Circulation. 2010 Dec 21

2. Karamlou T, Gurofsky R, Al Sukhni E, Coles JG, Williams WG, Caldarone CA, Van Arsdell GS, McCrindle BW. Factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous connection. Circulation. 2007 Mar 27; 115(12)

3. Padalino MA, Cavalli G, De Franceschi M, Mancuso D, Maschietto N, Vida V, Milanesi O, Stellin G. Surgical outcomes of total anomalous pulmonary venous connection repair: a 22-year experience.J Card Surg. 2014 Sep; 29(5):678-85

4. Respondek-Liberska M. Atlas Wad Serca Płodu. 2011. Adi Art., Łódź

5. Khan MS, Bryant R, Kim SH, Hill KD, Jacobs JP, Jacobs ML, Pasquali SK, Morales DL. Contemporary Outcomes of Surgical Repair of Total Anomalous Pulmonary Venous Connection in Patients With Heterotaxy Syndrome. Ann Thorac Surg. 2015 Jun; 99(6)

6. Laux D, Fermont L, Bajolle F, Boudjemline Y, Stirnemann J, Bonnet D. Prenatal diagnosis of isolated total anomalous pulmonary venous connection: a series of 10 cases. Ultrasound Obstet Gynecol. 2013 Mar;41(3):291-7.

7. Law KM, Leung KY, Tang MH, Chau AK. Prenatal two- and three-dimensional sonographic diagnosis of total anomalous pulmonary venous connection. Ultrasound Obstet Gynecol. 2007 Oct;30(5)

8. Allan LD, Sharland GK. The echocardiographic diagnosis of totally anomalous pulmonary venous connection in the fetus. Heart. 2001 Apr;85(4): 433-7.

9. Karamlou T, Gurofsky R, Al Sukhni E, Coles JG, Williams WG, Caldarone CA, Van Arsdell GS, McCrindle BW. Factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous connection. Circulation. 2007 Mar 27; 115(12):1591-8.

10. Hancock Friesen CL1, Zurakowski D, Thiagarajan RR, Forbess JM, del Nido PJ, Mayer JE, Jonas RA. Total anomalous pulmonary venous connection: an analysis of current management strategies in a single institution. Ann Thorac Surg. 2005 Feb; 79(2):596-60

11. Słodki M, Respondek-Liberska M, Pruetz JD, Donofrio MT. Fetal cardiology: changing the definition of critical heart disease in the newborn. J Perinatol. 2016 Mar 10

12. Góra E, Moczulska H, Janiak K, Słodki M, Respondek-Liberska M. Anomalous pulmonary venous return with stenosis in comlex left heart defect- lethal anomaly- 3 cases report. Polska Kardiologia Prenatalna ECHO PŁODU, 2013, 1 (8), 22-27

13. Ganesan S, Brook MM, Silverman NH, Moon-Grady AJ. Prenatal findings in total anomalous pulmonary venous return: a diagnostic road map starts with obstetric screening views. J Ultrasound Med. 2014 Jul

14. Valsangiacomo ER, Hornberger LK, Barrea C, Smallhorn JF, Yoo SJ. Partial and total anomalous pulmonary venous connection in the fetus: two-dimensional and Doppler echocardiographic findings. Ultrasound Obstet Gynecol. 2003 Sep;22(3):257-63.

15. Respondek-Liberska M1, Janiak K, Moll J, Ostrowska K, Czichos E. Prenatal diagnosis of partial anomalous pulmonary venous connection by detection of dilatation of superior vena cava in hypoplastic left heart. A case report. Fetal Diagn Ther. 2002 Sep-Oct;17(5):298-301.

16. Kawazu Y, Inamura N, Shiono N, Kanagawa N, Narita J, Hamamichi Y, Kayatani F.'Post-LA space index' as a potential novel marker for the prenatal diagnosis of isolated total anomalous pulmonary venous connection. Ultrasound Obstet Gynecol. 2014 Dec;44(6)

17. Volpe P, Campobasso G, De Robertis V, Di Paolo S, Caruso G, Stanziano A, Volpe N, Gentile M. Two- and four-dimensional echocardiography with B-flow imaging and spatiotemporal image correlation in prenatal diagnosis of isolated total anomalous pulmonary venous connection. Ultrasound Obstet Gynecol. 2007 Nov;30(6):830-7.

18. Olsen R, Doyle Z, Levy D, Anton T, Molkara D, Tarsa M, Sklansky M5, Pretorius DH. Anomalous Pulmonary Venous Return: Insights Into Prenatal Detection. J Ultrasound Med. 2016 Jun;35(6):1193-206.

19. Seale AN, Carvalho JS, Gardiner HM, Mellander M, Roughton M, Simpson J, Tometzki A, Uzun O, Webber SA, Daubeney PE; British Congenital Cardiac Association. Total anomalous pulmonary venous connection: impact of prenatal diagnosis. Ultrasound Obstet. Gynecol. 2012 Sep;40(3):310-8.

20. Jang SI, Song JY, Kim SJ, Choi EY, Shim WS, Lee C, Lim HG, Lee CH. The recent surgical result of total anomalous pulmonary venous return. Korean Circ J. 2010 Jan;40(1):31-5.

21. Fu CM, Wang JK, Lu CW, Chiu SN, Lin MT, Chen CA, Chang CI, Chen YS, Chiu IS, Wu MH. Total anomalous pulmonary venous connection: 15 years' experience of a tertiary care center in Taiwan. Pediatr Neonatol. 2012 Jun;53(3):164-70.

Division of work:

Maria Respondek-Liberska: idea of the article, diagnosis of the case, documentation of the case, correction of the manuscript

Łukasz Sokołowski: first draft of manuscript, review of the literature

Other authors: work with the manuscript

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