ANTENATAL ECHOCARDIOGRAPHIC DIAGNOSIS OF AN AORTOPULMONARY WINDOW COMBINED WITH PULMONARY STENOSIS IN NEONATE. A CASE REPORT

Author:
Katarzyna Więckowska[1], Jerzy Węgrzynowski[2], Katarzyna Zych - Krekora[3], Joanna Kwiatkowska[4], Maciej Słodki[3,5], Maria Respondek-Liberska[6,3]

1. Medical University of Lodz, Medical Faculty 2.Szczecin Zdroje Hospital 3. Department of Prenatal Cardiology, Polish Mother's Memorial Hospital Research Institute 4. Department of Pediatric Cardiology and Congenital Heart Disease, Medical University, Gdańsk 5. The State School of Higher Professional Education in Plock 6. Department of Diagnoses and Prevention Fetal Malformations Medical University of Lodz

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Abstract

In this report a case of aortopulmonary window (APW) diagnosed at 26 hbd is presented. APW supported the pulmonary circulation in neonate afflicted with pulmonary stenosis. To our knowledge, this is the first report in the literature referring to observing their coincidence in fetal life.

Key words: *aortopulmonary window, pulmonary stenosis, prenatal diagnosis*

INTRODUCTION

Aortopulmonary window (APW) is a rare anomaly in the development of the great arteries. This lesion of the septum between the aorta and pulmonary artery placed above the level of ventriculoarterial connection is uncommon: the prevalence of the defect accounts for about 0,1% of all cardiac abnormalities[1].

Pulmonary stenosis, accounting for around 9% of postnatal congenital heart diseases, causes an obstruction of blood flow into the pulmonary circulation. It occurs mostly at the level of pulmonary valve[1].

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The aim of this report is to present the case of a newborn afflicted with pulmonary stenosis, which was diagnosed with APW at 26th week of gestation.

CASE REPORT

Prenatal findings

Thirty-two-year-old gravida III, para III was referred to our tertiary center due to ventricular asymmetry detected at 20 weeks by screening ultrasound. Until then, the pregnancy was uneventful.

The first fetal echo performed at 26w1d revealed cardiomegaly (HA/CA=0,4, AP of 28 mm), pericardial effusion and heart axis of 50 degrees. The foramen ovale was of 7 mm and its flap was flaccid and aneurysmal. There was domination of the left ventricle. The aortic and pulmonary valves were of 5,8 mm and 5 mm respectively. In the mediastinum in addition to the three vessels there was also the innominate vein. Thymus of good size was documented. Aortic arch was captured in the longitudinal view.

Flows through the atrioventricular valves were normal, although a trace of tricuspid regurgitation was present. In the narrower vessel a suspicious turbulent (V max to 100 m/s) flow was visible in both longitudinal and short axis view. Moreover, there was a coronary artery in plane view of intraventricular septum.

On the basis of this examination the diagnosis of aortopulmonary fenestration was made. Treatment with digoxin and steroids at 29 weeks of gestational age were recommended.

At 33w3d of GA, cardiomegaly persisted. There was a Chiari's network present in right atrium. The left ventricle still dominated over the right and it had similar trabeculation (Fig. 1). Foramen ovale was patent of 8 mm, the right-to-left flow of 50 cm/s was documented (Fig. 2).

M-Mode ultrasound showed poor contractility of right ventricle. In the mediastinum the turbulent flow between

Corresponding author: katarzyna.wieckowska@stud.umed.lodz.pl

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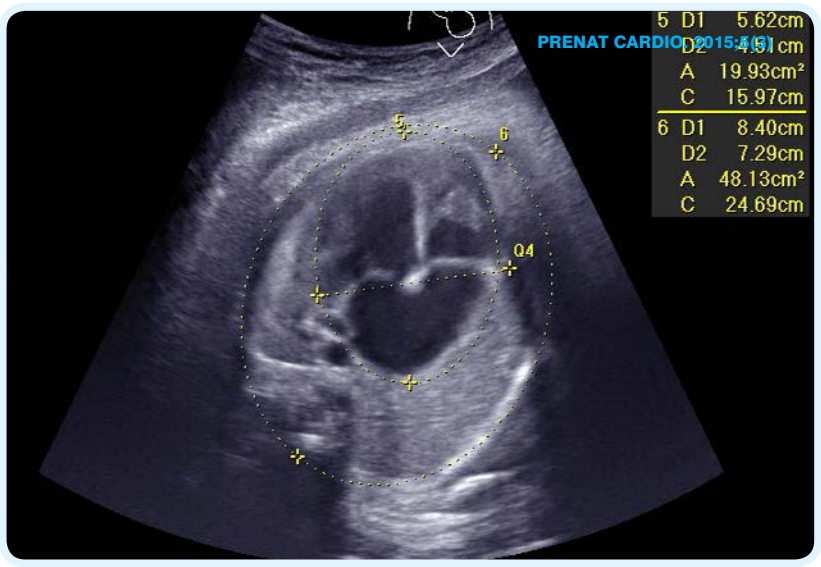


Figure 1. Disproportion between ventricles and LV enlargement

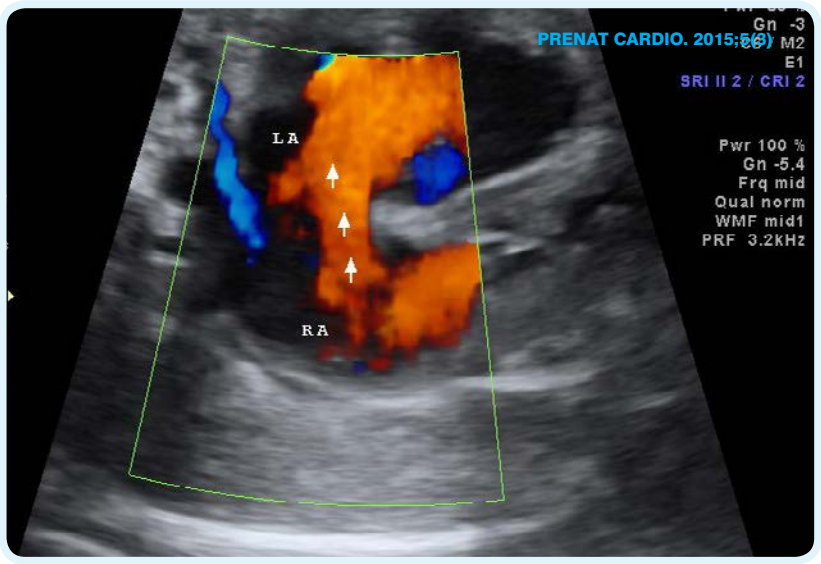


Figure 2. The right-to-left flow through the foramen ovale

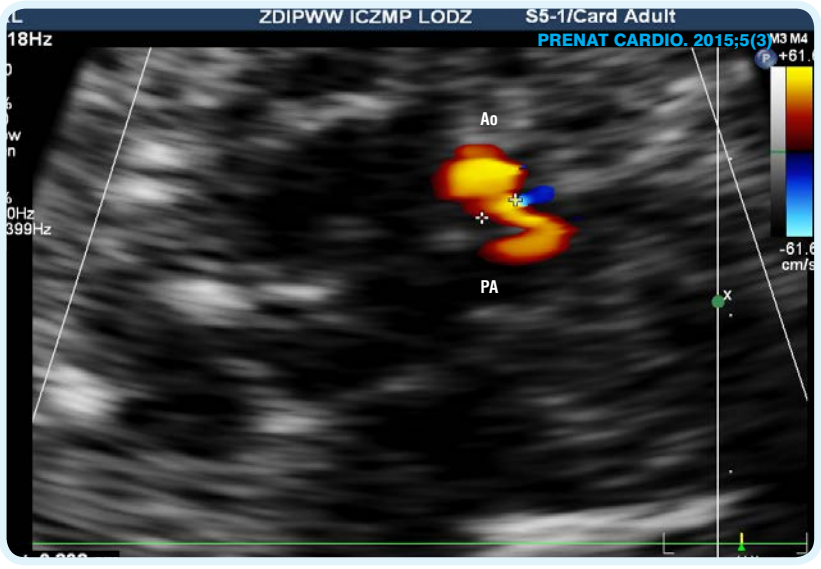


Figure 3. Aortopulmonary window detected in fetal life

pulmonary artery and aorta of maximum velocity of 100 cm/s and about 2 mm of diameter (Fig. 3). Fetal physiology was good, the fetus presented frequent and forceful respiratory movements of diaphragm. Digoxin treatment was continued.

Three weeks later still the dominance of left ventricle was present. The appendages of the atria were difficult to assess (the LAA seemed to be on the right side). The turbulence of flow between the great arteries had a velocity of 115 cm/s. The maximum velocity of flow through pulmonary valve was 130 cm/s. The contractility of the right ventricle was dramatically poor (SF=3%).

One week later (36w2d/35w1d) the function of the right ventricle improved to SF RV of 16%. Distended branches of the main pulmonary artery were discovered (right and left PA of 4 mm) in the mediastinum and a comb-pattern spectral Doppler through peripheral pulmonary vessels. The suspicious retrograde flow in RVOT was observed.

Fetal hemodynamics suggested higher pressure in pulmonary artery than in the aorta (there was R- L shunt at the level of APW). Postnatal care was discussed with the parents to be: planned delivery in the tertiary care center, the possibility of development of respiratory failure, observation in the NICU and the possibility of administration of nitric oxide.

A week later pulmonary regurgitation was revealed (Fig. 4).

Postnatal outcome

The baby girl was delivered by SC at 39w2d with birth weigh 3 130 g and Apgar score 10/10/10. Couple hours later she had 80% of oxygenation and was transferred to the Pediatric Cardiology Department. The dilated cardiomyopathy was suspected based on the first neonatal echocardiography.

The main postnatal findings were: monstrously dilated left atrial appendage, surrounding the left side of the main pulmonary artery and free wall of the left ventricle, bidirectional flow between the atria: alternately from the left atrium to the right, and from the inferior vena cava to the left atrium. Pulmonary valve was significantly narrower than aorta (but expanded in the next three days) and had three leaflets of thickened edges. Contractility of the ventricles was within normal limits. The left ventricle had

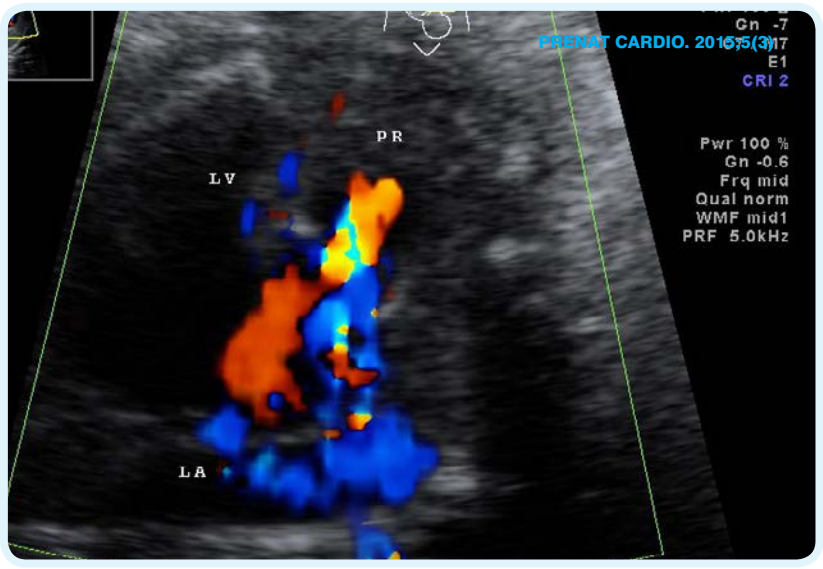


Figure 4. Pulmonary valve regurgitation at 37 hbd

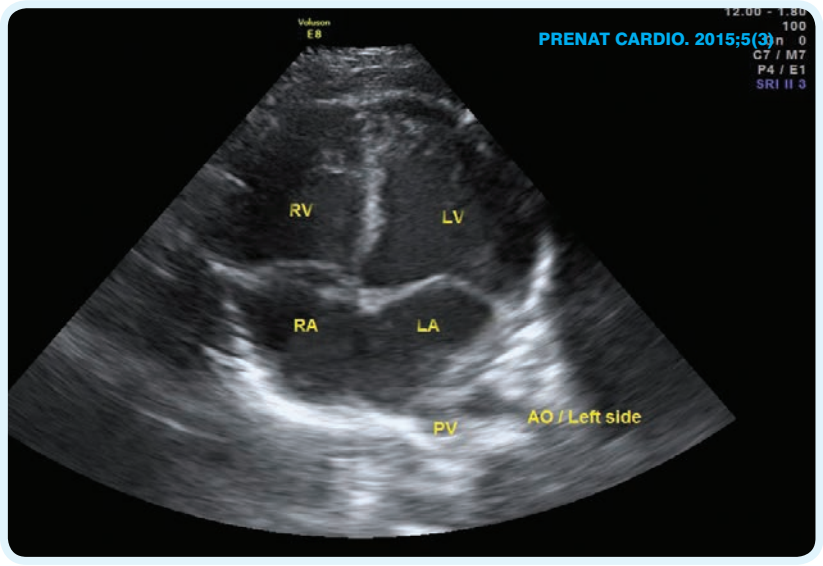


Figure 5. Four chambers view with annotations at the age of 6 weeks

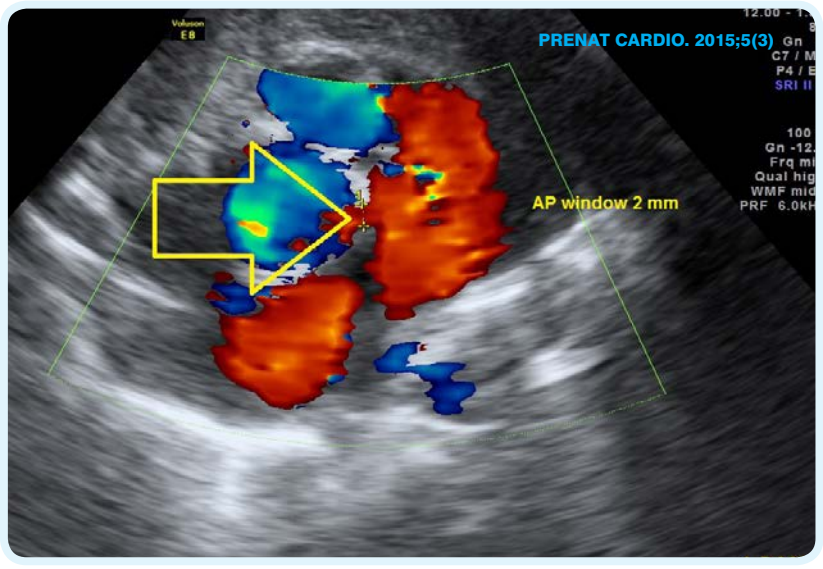


Figure 6. APW of 2 mm at 6 weeks of postnatal life

an abundant trabeculation and numerous lacunes. In conclusion, the CHD in this child was described as a defect with diminished pulmonary circulation, presenting certain features of HRHS.

The condition of the neonate improved after oxygen therapy, there were no overt signs of cardio-respiratory insufficiency. The baby was discharged home in good clinical condition with gaining weight.

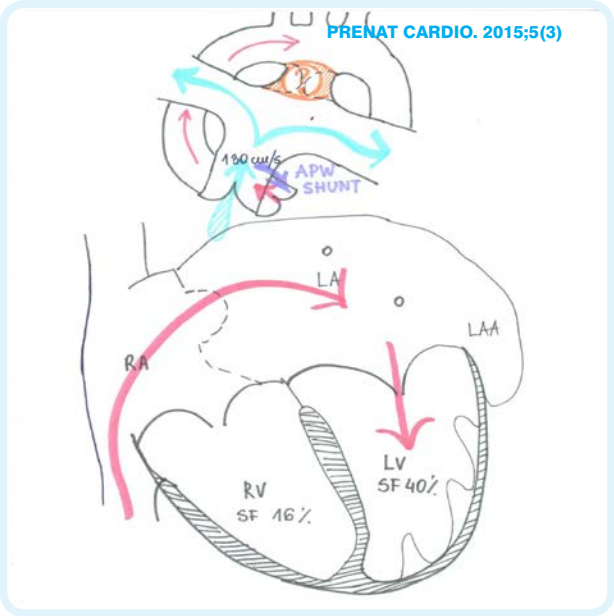
In the sixth week of her life, in the four chamber view the convertion of atrial appendages was noted (Fig. 5). The APW of 2,2 mm was still present above the level of arterial valves (Fig. 6). The pulmonary valve seemed to be dysplastic and there was an increased Doppler velocity across the valve up to 3,15 m/s (Fig. 7).

DISCUSSION

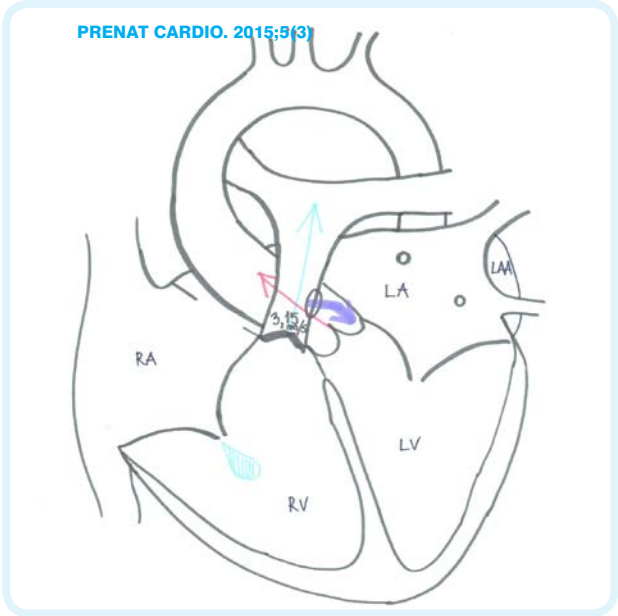
The prenatal diagnosis of aortopulmonary window is very difficult to make in fetal life[1]. An obstetrician cannot detect this lesion when performing the basic ultrasound. Although the AP window may be noticed directly during the scan in a tertiary care center[2], generally it is diagnosed through indirect signs, such as retrograde pulmonary perfusion[2] and unusual pulmonary valve regurge[3]. The AP window is frequently associated with other major cardiac defects: interrupted aortic arch, ventricular septal defect and Fallot syndrome[4].

Although the antenatal observation of APW is rare, reports exist of cases of fetal echocardiographic findings suggesting this condition[2,3,5,6,7,8]. Some scientists claim that observation of this kind of defect has little impact on further management in newborn as APW does not require emergency therapy and can be easily detected in postnatal echocardiography[6]. However, it may influence prognostic information[6] and surely contributes a better outcome in cases of APW coexisting with other lesions[5]. In each case surgical treatment is needed, preferably an operation in neonatal period, so as to prevent secondary damage such as irreversible pulmonary vascular changes[2].

In the presented case, aortopulmonary window coexisted with prenatal cardiomegaly and left ventricle dominance, and after delivery evident mild pulmonary stenosis with diminished RV were present.



Baby's heart - 6 weeks after the birth



Fetal heart at 37th week

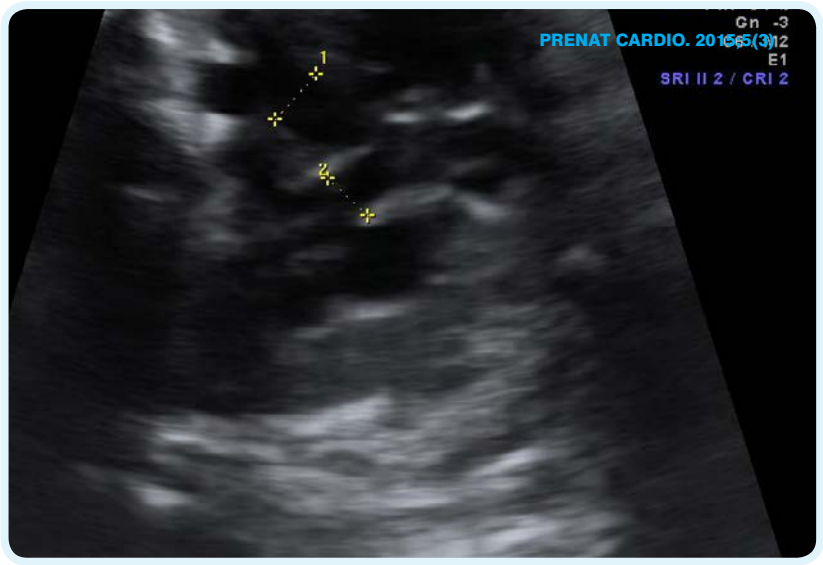


Figure 7. RPA and LPA in 37 hbd of GA

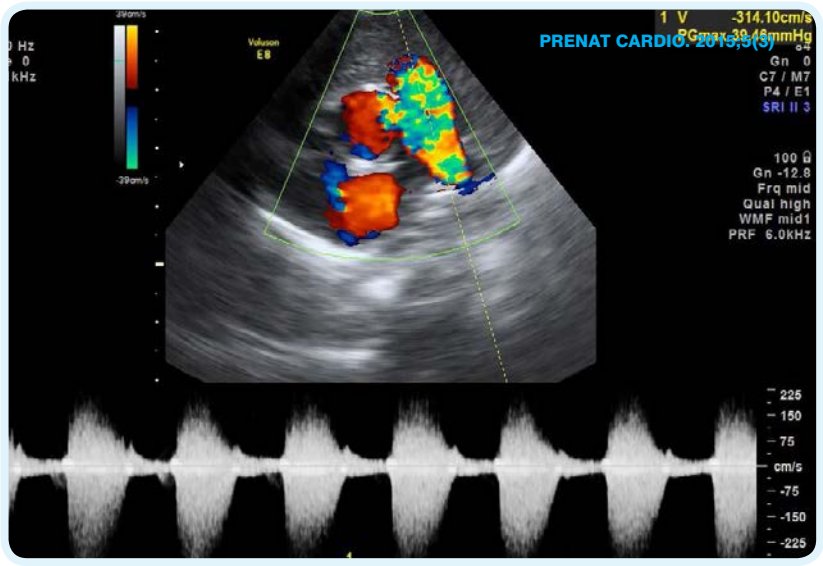


Figure 8. Doppler velocity at PV at the age of 6th weeks

The literature comprises just a few papers of coincidental APW and PS, one is dated back to eighties of the past century and it is related to adults[9]. The other two discuss the problem in neonates[10,11], but none of them raise the issue of the diagnosis in fetal life.

The haemodynamic explanation of prenatal findings with postnatal outcome in this case is very interesting and not easy. It is believed that the shunt across the AP window contributed to the left atrium and left ventricle overloading during prenatal life. It was the reason for the unusual abundant trabeculation of the LV. Compromising RV contractility was provoked by the great volume of the left ventricle (the right ventricle was compressed by the left heart's structures). On the other hand prenatally RV had an increased pressure due to ductal constriction or ductal agenesis (wide right and left pulmonary arteries were also a sign of this condition). Supposedly, this constriction could be also an effect of steroid therapy in 29 hbd.

During prenatal life flow across pulmonary valve was up to 1,3 m/s and there was pulmonary regurgitation described on the last examination. Usually in severe pulmonary stenosis or dysplasia there is also significant tricuspid valve regurgitation which was not seen in this case.

After delivery there was a redistribution of the blood in the circulatory system. Due to this redistribution the pulmonary flow was decreased, the hypoxemia and reduced levels of saturation occurred. The absence of patent DA could not be ruled out at this stage.

The retrospective analysis of the previous examinations revealed that the ductal arch in the longitudinal view was not documented clearly during pregnancy despite of good echogenicity and other conditions for testing.

The improvement of the right ventricle function on postnatal examinations makes the prognosis optimistic, PS may be less and less significant over months.

To conclude, to our knowledge this case of such an early diagnosis of combined APW and PS is the first to publish. The AP window could sustain pulmonary flow in condition of stenotic pulmonary valve in postnatal period. Was neonatal pulmonary stenosis just a coincidence with AP window or its prenatal consequence, it is hard to say, as there is no data in the literature. The ductal presence should be checked in a case of AP window.

Biometry [hbd]	26/25	33/32	35/35	36/35	37/36
EFW [g]	783	1876	2651	2858	2905
AP [mm]	28	40	45	47	48
HA/CA	0,4	0,4	0,38	0,4	0,4
Axis [deg.]	50		41		40
PE [mm]	2			2	2
A:V	01:03	01:01	01:01	01:03	01:03
TV [mm]	Traces of TR	13		13	TR
AV [mm]	5,8	8	9	7	7
AV [cm/s]			100		85
PV [mm]	5	9	8	8	10
PV [cm/s]			130	130	130
PR					PR +
Septum, movements	normal	normal	paradoxal	Not evaluated	Not evaluated
SF LV [%]		31	30	31	40
SF RV [%]		28	3	16	16
FO	7 mm	8 mm	13 mm		
Fo, Vmax+direction		50 cm/s,	50 cm/s,		
		R → L	R → L		
Shunt, R → L	2 mm, 100 cm/s	idem	2 mm, 115 cm/s		
Pulmonary veins	29 cm/s		49 cm/s	40 cm/s	20-36 cm/s
DV	Normal	Normal	Normal	Normal	Abnormal
MCA [cm/s]		30	32	24	76
MCA, PI			1,9	1,54	1,8
Tei RV	0,46	0,8	0,9	0,7	0,7
Tei LV	0,56	0,6	0,52	0,3	0,5
Digoxine	no	yes	yes	yes	yes
Steroids		Administration at 29 week			
Other medications		Amoxicillinum			

Table 1. Longitudinal fetal echocardiography monitoring

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Zych-Krekora K: data collection, work with manuscript

Kwiatkowska J: pediatric cardiologist, data collection, work with manuscript,

Słodki M: data collection, work with manuscript

Respondek-Liberska M: concept of the reasearch, provider of photos, work with the manuscript, final version