Case report

FETAL "AORTIC COARCTATION" AND DIFFERENT NEONATAL FOLLOW-UP IN 3 CASES

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Abstract

Heart defects which includes narrowing of aortic isthmus - aortic coarctation (CoA) are one of the most prevalent birth defects. Making a correct prenatal diagnosis of CoA is very difficult and problematic. We are still observing many false (+) and false (-) diagnoses. In presenting 3 cases with prenatal suspicion of CoA only one patient confirmed this defect in the postnatal life. In the fetal echocardiography inappropriate dimensions of great vessels and PA/Ao ratio are very relevant in the CoA diagnostics. Based on such suspicion before delivery we can select a group in which birth in the tertiary center, prostin infusion, control ECHO examinations and planned cardiac surgery will be needed. But wide differential diagnosis including pulmonary dilatation (due to pulmonary hypertension or fetal blood redistribution due to possible infection) is required.

Key words: *prenatal cardiology, complex heart defect, prenatal diagnosis, pulmonary hypertension, aortic coarctation, heart defect correction*

INTRODUCTION

Coarctation of the aorta (CoA) is relatively common disease , seen both prenatally and postnatally affecting 5-10% of infants with heart disability and it's three times more common in males [1,2]. In addition this lesion occurs in 48% of patients with Turner syndrome[3]. According to the Polish Registry of Fetal Cardiac Malformations in years 2004-2015 CoA was present in 2,4% of cases and was among the top 10 most common congenital fetal heart defects[4-7] . The true prevalence of prenatal CoA is difficult to assess due to both false (+) and false (-) diagnoses [8,9]. Narrowed lumen of aorta (more than 2/3) causes blood flow disorders. Rudolph et al. hemodynamic theory according to which the CoA occurs by supplying the lower part of the body through the ductus arteriosus (DA) and the associated lower blood flow through the isthmus is leading to aortic isthmus hypoplasia and its narrowing [10]. The first signal of prenatal aortic coarctation can be the disproportion between ventricles. Hypertrophic right ventricle pumps blood into MPA and then to ductus arteriosus which provide appropriate pressure of blood in the lower part of the body, causing smaller volume of the left ventricle and its narrow aortic arch. The similar situation might be seen in fetuses in the 3rd trimester, who after birth turned out to have normal heart anatomy and normal aortic arch.

We're presenting 3 cases of prenatal diagnoses of CoA (all with normal 1st trimester) and very different postnatal outcome.

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CASE NR 1

Prenatal findings

Thirty-two-year-old gravida II, para I (miscarriage at 5hbd, 2 years ago) was referred to our tertiary center due to suspicion of heart defect at 31 weeks of gestation.

First obstetric US at 12 weeks of gestation was described as normal, nuchal translucency was 1,7 mm. Next scan at 23rd week of gestation revealed a ventricular septal defect (VSD) and cytogenetic evaluation with normal karyotype- 46,XY.

The first ECHO in our referral center at 31hbd revealed male fetus with ductus arteriosus dependent congenital heart disease: there were two symmetrical atria with widely opened foramen ovale, and two morphologically different ventricles. The attachments of tricuspid valve (TV) tendinous chords was overlaying at the upper part of intraventricular septum suggesting

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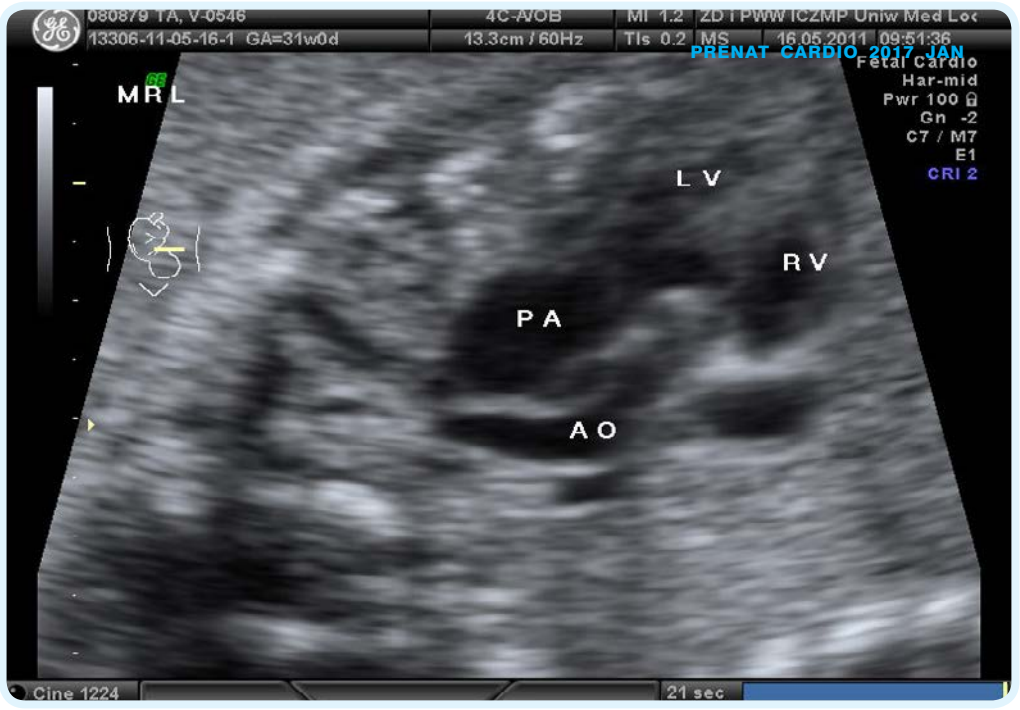

Fig. 1. Disproportion between big arteries at 31weeks – case nr 1.

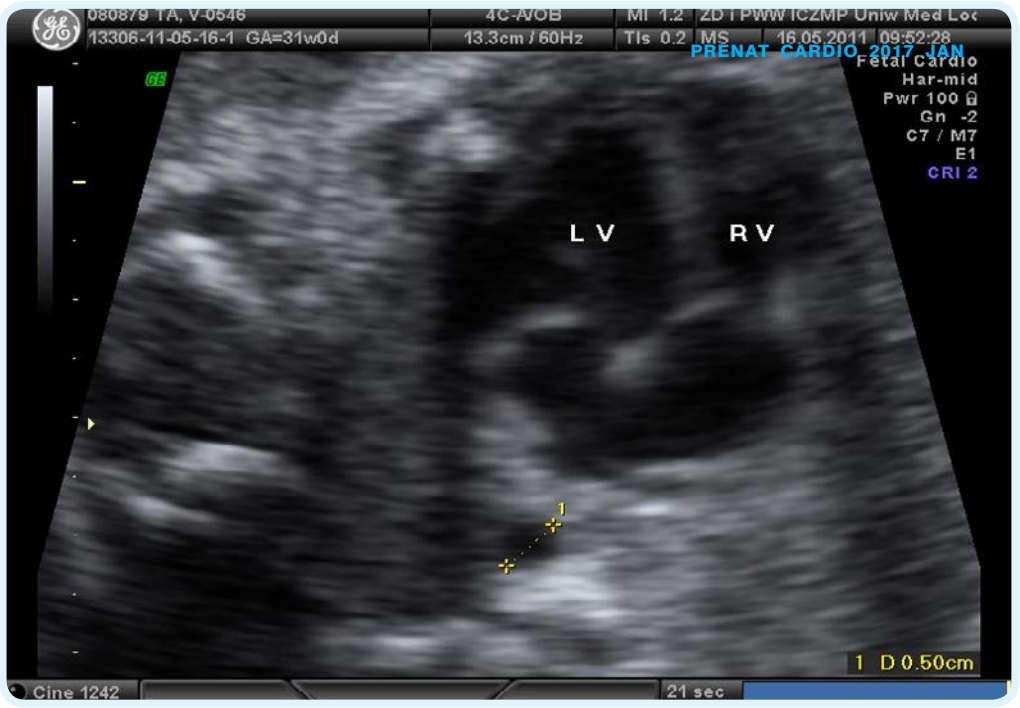

Fig. 2. Four chamber view – case nr 1 : almost symmetrical ventricles: LV slighty larger than RV (in CoA usually RV is bigger than LV)

straddling tricuspid valve. The inlet portion of ventricular septum was intact however there was big outlet VSD with aortic valve malalignment. At the level of fetal mediastinum we measured vessels dimensions: pulmonary artery (PA) was 8mm, aorta (Ao)-4mm, superior vena cava (SVC)- 4mm and there was large thymus. The ductus arteriosus arch was wide in the longitudinal axis, crossing hypoplastic aorta, partly visualized. At 34hbd hypoplastic "gothic" aortic arch was seen suggesting coarctation and hypoplasia in the transverse part of aorta. Summarizing this CHD was assessed as severe one, according to new classification[11,12], meaning the ductal dependent circulation required prostin (prostaglandin E1) infusion for neonate and planned neonatal cardiac surgery with repair of aortic arch, however without emergency just after delivery. (Fig.1,2,3).

Postnatal outcome

The baby was born in our center by forceps delivery at 38hbd with birth weight 3100g and Apgar score 8. Until 12th day of postnatal life he was on prostin infusion. Postnatal diagnosis confirmed prenatal findings: VSD=12 mm, CoA=2,5-2,7mm (PG=15 mmHg), hypoplastic aortic arch=4-5mm and narrowing of the left ventricle output tract up to 3,3 mm. Foramen ovale (FO) diameter was 4,7 mm, pulmonary flows was increased. Patient was qualified for pulmonary banding procedure and palliative Crawfoord operation in our Pediatric Cardiac Surgery Department, which was performed at 27th day with no complications.

Follow-up

At the age of 7-month mild recoarctation was suspected. At the age of 2,5-years- he had subaortic muscular obstruction, causing a gradient of 100 mmHg. At the age of 3 years final cardiac repair was carried out (University of Munster, prof. E. Malec): closure of outlet VSD, myomectomy of the LVOT mass and debanding. During cardiac surgery double orifice of TV was seen. Currently he is 5 years old, good looking with normal blood pressure on the upper and lower extremities, with normal chest without cardiomegaly. In echocardiography 4

Fig. 3 Schematic view of 4 chambers and great vessels of case nr 1.

chamber view is almost "normal", distored by anusual tricuspid valve (there is shortened septal leaflet), with moderate tricuspid valve regurgitation (up to 2 m/sec), good LV function and widely opened aortic arch with good flow in descending aorta up to 1,6 m/sec. He stays on pharmacologic treatment (enalapril and diuretics) and cardiological check-up is recommended every 6 months.

CASE NR 2

Prenatal findings

Thirty-seven-year-old gravida II, para II was referred in the third trimester to our tertiary center. At 35th week of gestation the heart defect was detected. Until then the pregnancy was uneventful.

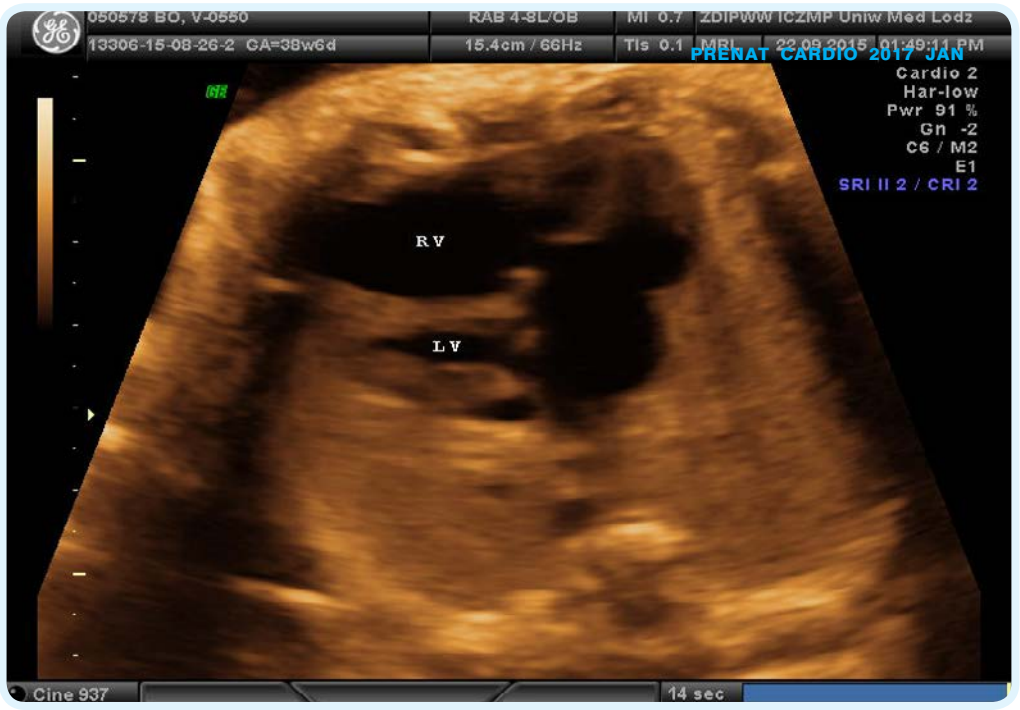

Fig. 4. Right ventricle enlargement and small left ventricle at 38w6d – case nr 2

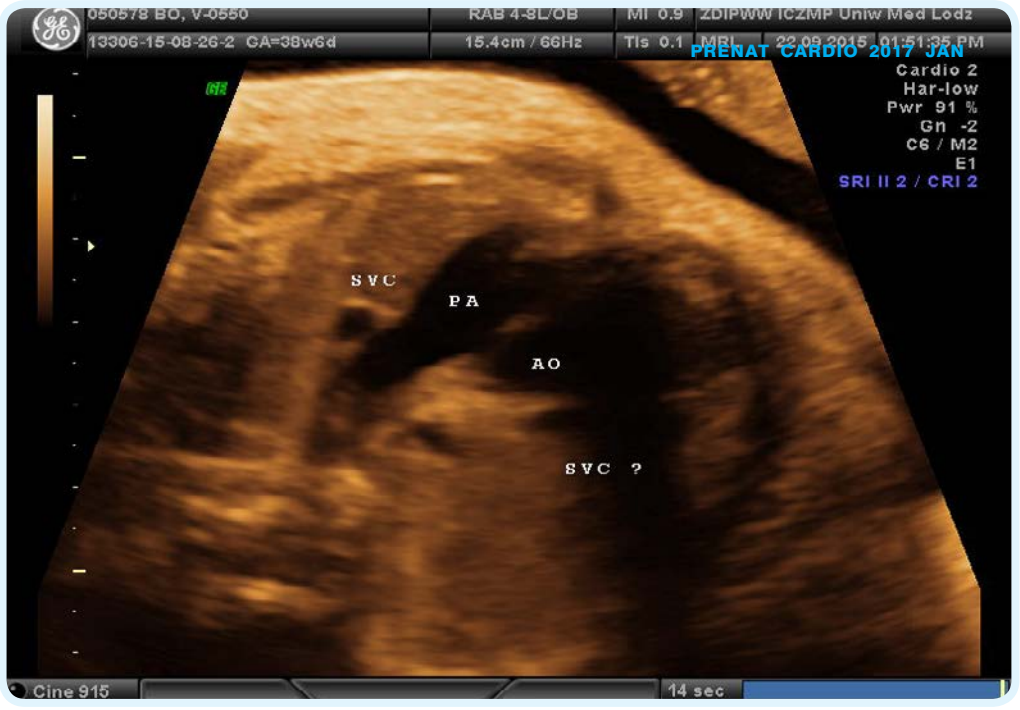

Fig. 5.Fetal mediastinum with additional left superior vena cava at 38w6d – case nr 2.

The first fetal echo at 35th week revealed abnormal biometry of the fetus (35/32-small for gestational age (SGA)), polyhydramnion (amniotic fluid index (AFI)=21) and congenital heart defect: There was chambers disproportion : right atrium was bigger than left atrium and right ventricle was bigger than left. The FO was widely opened. The aortic valve was 5,3 mm (Z-score= -1,22) and pulmonary valve was 8,3mm (normal range)- Z-score =1,31 . Aortic arch was described as hypoplastic. In the longitudinal view wide ductus arteriosus was shown. At the level of mediastinum there were four vessel- in addition to main pulmonary artery, aortic arch and superior vena cava there was also left superior vena cava. The thymus was estimated as in a good size. FHR was 140/min, sinus rhythm. 9 points in the CVPS was given due to pericardial effusion of 4 mm. Otherwise fetus was in a good condition presenting classical features of the face.

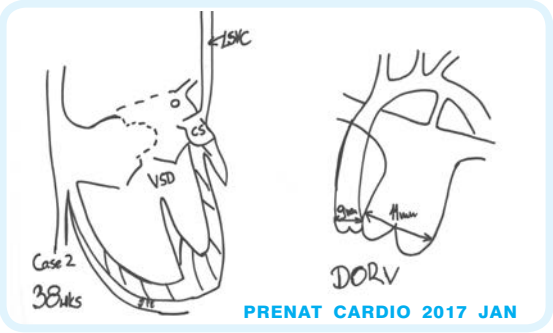

Fig. 6. Schematic view of 4 chambers and great vessels – case nr 2.

prostin dose was raised to 0,02 g/kg/min, due to lower oxygenation. Baby was transferred to cardiosurgery clinic at 21th day still with prostin infusion. Rashkind balloon atrial septostomy procedure was attempted , however was unsuccessful. At 22th day patient underwent Norwood operation. After induction of anesthesia milrinone infusion was required. Initially disconnection of extracorporeal circulation proceeded with dopamine and milrinone infusion. There occurred prolonged surgical hemostasis despite blood products transfusion. This resulted in postponed chest closing. After operation the circulation was unstable despite noradrenalin infusion. In few hours blood insufficiency showed with anuria and acidosis. What's more hypotension occurred with no

At 38th week according to the biometry there was 35th week, meaning normal gain weight from last examination. AFI was 16, HA/CA(heart area/chest area) was 0,4, aortic valve was 9 mm increased (Z score=2,21) and pulmonary valve was also increased 11 mm (Z score=3,22).

From prenatal cardiology point of view there was severe congenital heart defect[12], probably CoA, but not critical and due to heart size and dilatation of pulmonary valve[13] respiratory problems were suspected on the 2nd or 3rd day after delivery (Fig. 4, 5, 6).

Postnatal outcome

The female newborn was delivered by CS at 40th week with birth weight 2800g and Apgar score 8/8. Neonate was in a stable condition on prostin infusion since day 1st, nourished parentally, treated because of secondary infection (*Escherichia coli* and *Staphylococcus spp.* detected in the swab from endotracheal tube). Antibiotic therapy by sultamicillin and gentamicin was implemented since 5th day of postnatal life. At 9th day

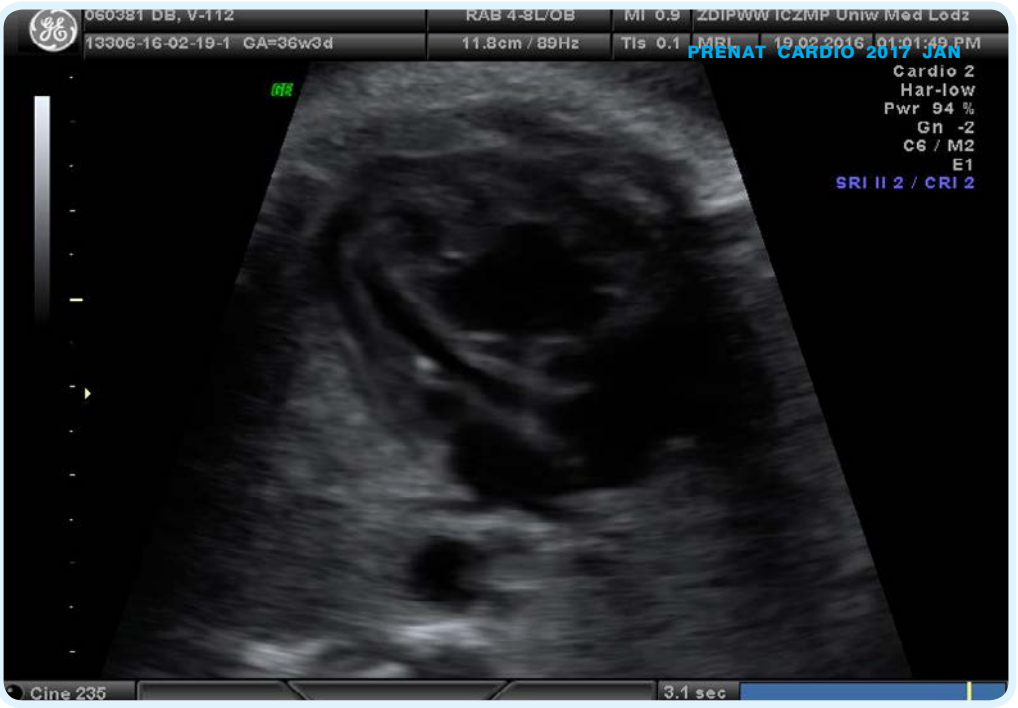

Fig. 7. Fetal 4 chamber view at 36w3d , showing RV >> LV – case nr 3.

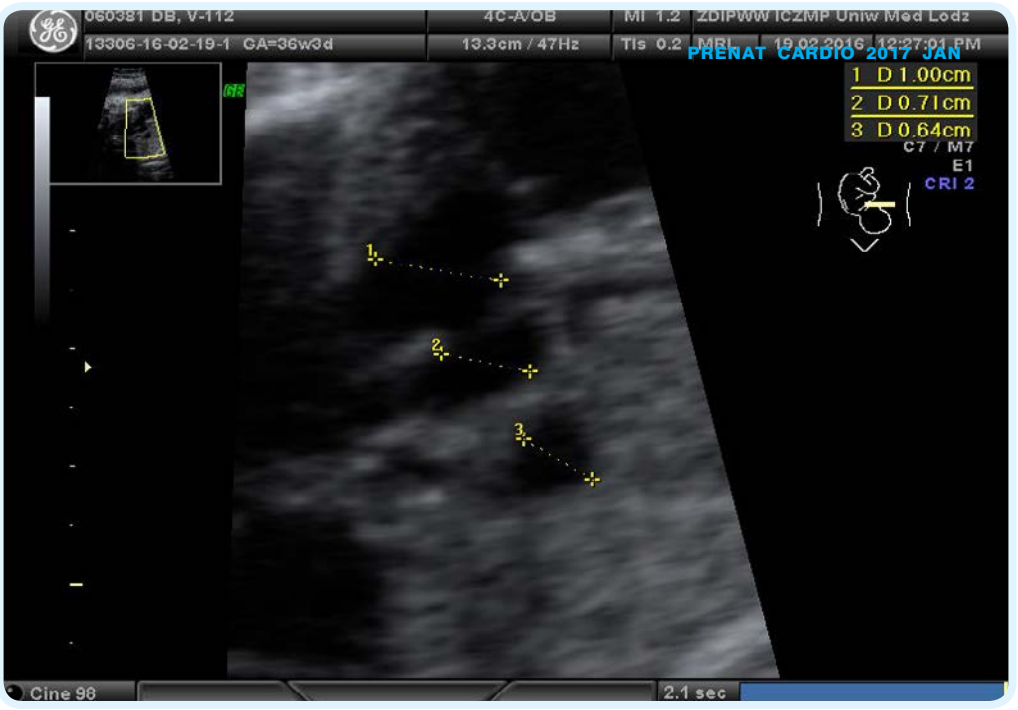

Fig. 8. Fetal mediastinum at 36w3d – case nr 3.

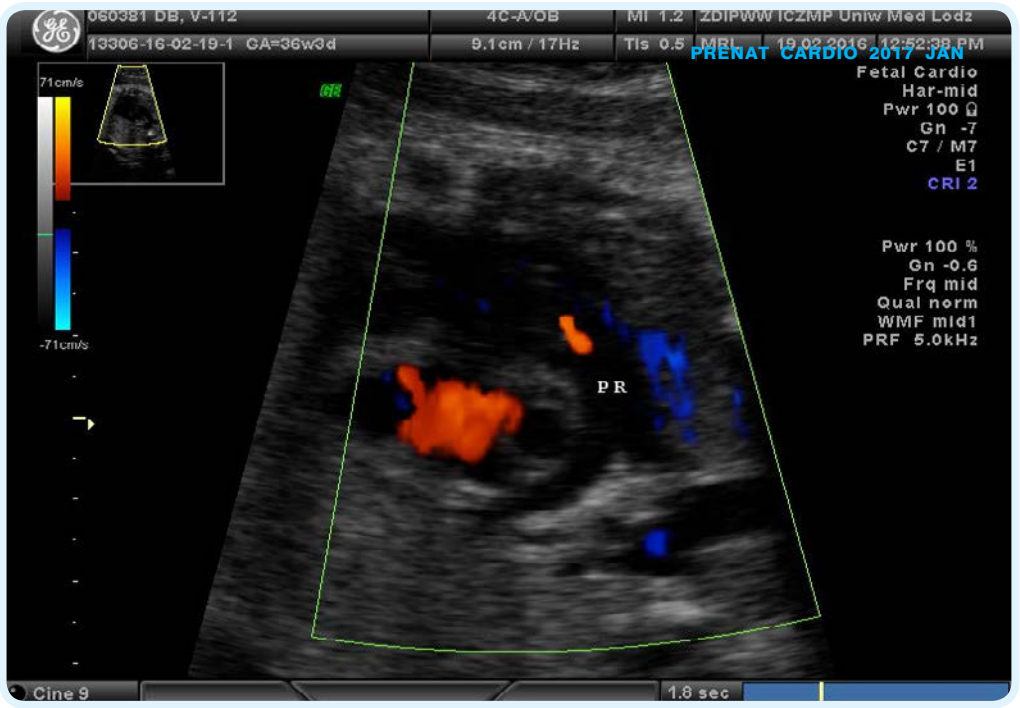

Fig. 9. Fetal pulmonary regurgitation at 36hbd – case nr 3.

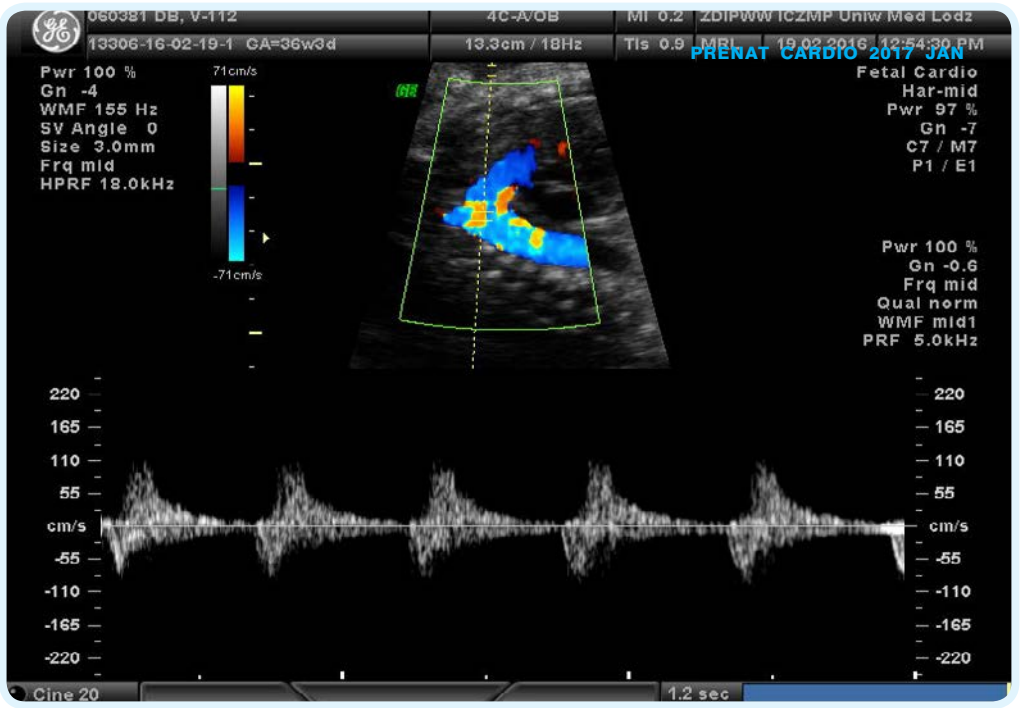

Fig. 10. Bidirectional flow at the level of aorta and the ductus arteriosus at 36hbd – case nr 3.

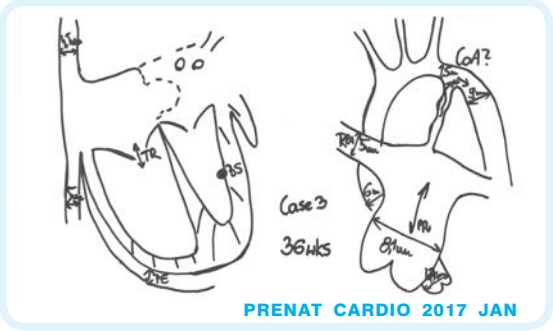

Fig. 11 Schematic view of 4 chamber and great vessels – case nr 3.

response on resuscitation. Patient died at 24th day of postnatal life, 2 days after surgical operation.

Autopsy

The autopsy revealed that the surgery was *lege artis*. Right ventricle was hypertrophic with fibrosis. Moreover high degree extension of both pulmonary arteries and the peripheral branches was noticed. Morphological changes provides for diagnosis of pulmonary venous hypertension (I stage)[2].

CASE NR 3

Prenatal findings

35-year-old gravida in the second pregnancy, second delivery (previous pregnancy was extrauterine, current pregnancy after IVF) reported to our Prenatal Cardiology Department at 36hbd. At 35th week of gestation suspicion of CoA was made.

The first ultrasonography in our tertiary center showed male eutrophic fetus with normal biometry. AFI was 22 suggesting polyhydramnion. In the four chamber view there was asymmetric ventricles with right side dominance. The FO was patent with 3,5 mm dimension. In the left ventricle there was noticed a bright spot. The mediastinum was described as untypical, the thymus was big at the upper limit of the normal range (39 x 19mm). Long tortuous ductus arteriosus was visible with shelf and turbulent flow. Aortic dimensions was: ascending-6mm, transverse-5mm, descending-9mm. The flow rate in the isthmus was 175 cm/s, reversal. There was large disproportion between aortic and pulmonary valve: AoV was 4,7mm and PV 8,1mm with pulmonary regurgitation (PR). What's more the right pulmonary artery (RPA) was also wide with 5mm diameter. Flows measured on mitral valve showed that E wave was bigger than A, suggesting impaired diastolic function. There was tricuspid valve regurgitation (TR) and pleural effusion in the pericardial sac (cardiovascular point scale: CVPS=8). The redistribution in

the fetal blood flows at the level of atria, ventricles and great arteries were evident, probably due to premature ductus arteriosus constriction, but neonatal echocardiography was recommended to rule out coarctation of the aorta.

Polyhydramnion, TR, PR, pericardial effusion, bright spot in LV, hydrops fetalis and intrahepatic umbilical vein up to 8 mm suggested also mild fetal infection. (Fig. 7,8,9,10,11).

Postnatal outcome

The baby was delivered by CS at 39th week of gestation (3 weeks after the last fetal ECHO) with birth weight 3360g and 9/9 in the Apgar score. Neonatal echo ruled out structural abnormality of the heart. Blood laboratory results were normal. At 4th day of postnatal life newborn was discharged from the hospital in a good clinical condition.

At the age of 5,5-months he was in a good condition with no abnormalities based on clinical examination. Echocardiography in our unit revealed : in the four chamber view symmetrical atria and ventricles, pulmonary blood flow by Doppler was 100cm/s with regurgitation up to 1,6 m/sec, wide pulmonary arterial branches. There was also prominent RV (in M-mode it was 15 mm). In his transfontanell ultrasound he had mild, symmetrical ventriculomegaly up to 10 mm.

The clinical data of 3 cases presented above are summarized in Table 1 and 2.

DISCUSSION

We presented three prenatally diagnosed cases in which, based on targeted echocardiography in tertiary center , CoA was suspected and only patient 1 confirmed a true CoA. Despite no asymmetry in the four chamber view, probably due to large VSD and no pressure gradient between RV and LV , he revealed coarctation and aortic hypoplasia. Targeted echocardiography in the 3rd trimester allowed to organizes delivery in a tertiary center, prostin infusion after birth and planned cardiac surgery in the same institution.

Patient nr 2 had RV dominance and CoA diagnosis was made because of hypoplastic aortic arch and narrowed isthmus[14]. However postnatal CoA was excluded. By retrospective analysis we focused our attention on the enlarged diameter of MPA what today gives us a suspicion of high

	Case1	**Case 2**	**Case3**
Prenatal findings:			
I trimester			
NT	1,7mm	1,8mm	1,5mm
Double test	Normal	Normal	Normal
II trimester	Normal	Normal	Normal
Gestational age at the time of detection anomaly	23 wks	26,1 wks	35,5 wks
Gestational age at the time of fetal ECHO diagnosis	26 wks	32,3 wks	36,4 wks
III trimester			
Last ECHO (biometry)	34,2	35,2	36,4
Gender	Male	Female	Male
Growth	AGA	AGA/SGA	AGA
RV>LV	No	Yes	Yes
PV (mm)	8	11	8,1
PV Z score	0,26	2,04	0,31
AV (mm)	4	9	4,7
AV Z score	-3,67	2,16	-2,2
PA/Ao	2	1,2	3,6
Ao arch	Hypoplastic	Hypoplastic	Normal
Y sign	Yes	Yes	No
CoA suspicion	**Yes**	**Yes + IAA**	**Yes + infection**
Mediastinum	3 vessels PA>>Ao	4 vessels	3 vessels
Thymus	Large	Large	Large
Extracardiac malformations (ECM)	SUA	-	Hyperechogenic bowels
PE	No	No	Yes
AFI	22	16	12
TR	No	No	Yes
CVPS	10	10	7
Nr of days between last ECHO and birth	25	14	20
Postnatal findings			
Week of delivery	38 th	40 th	39 th
Type of delivery	Forceps	CS	CS
Birth weight	3100g	2800g	3360g
Apgar score	8	8	9
Day of surgery	27th day-Crafoord and MPA banding	22th day-Norwood	-
Discharged home	41th day	Died at 24th day	At 4th day

Table 1. Clinical and echo data from 3 cases

	Case nr 1	Case nr 2	Case nr 3
Disproportion at the 4 chamber view	**No**	Yes	Yes
Disproportion at the mediastinum level	**Yes**	**Yes**	**Yes**
Cardiac surgery	**Yes (True CoA)**	Yes	No
Additional findings	2 orifice TV	LSVC; Postmortem pulmonary hypertension	Prenatal ductus arteriosus constriction and or mild infection? Postnatal mild ventriculomegaly on transfontanel US with no clinical symptoms

Table 2. Selected findings in prenatal CoA in 3 presented cases

probability of pulmonary hypertension[13]. Probable in the prenatal life early changes in the pulmonary tissue occurred. Progression of pulmonary hypertension was probably a cause of patient death.

Case 3 showed that narrowed aortic isthmus as a sign of blood redistribution probably due to intrauterine infection. Tricuspid and pulmonary regurgitation occurred due to volume overload and or myocardial impairment. Pleural effusion and E > A were a part of congestive cardiomyopathy. What's more, bidirectional flow through the aortic isthmus suggested that we have a right to suspect that there aortic isthmus obstruction can occur[15]. Next to regurgitation through the right heart, flows through DA was reversed and called as a shunt. It results that more blood supplies lungs which could provide also to pulmonary hypertension. Postnatal persistent DA and probability of mild pulmonary hypertension occurrence later in his life were the reasons to monitor this patient despite of his lack of clinical symptoms[16]. Neonatal infection wasn't confirmed after delivery by routine laboratory tests. Could prenatal findings be an endpoint of earlier fetal infection?

These three above cases shows that there isn't one correct easy scheme to diagnose prenatally CoA[17]. Truly diagnosed CoA should have PA/ Ao> 2,03 +/-0,48[8]. Only in case 1 this ratio was 2 which enabled to make a correct diagnose. In cases 2 and 3 value of PA/Ao ratio (there were 1,2 and 3,68, but false positive with narrowed aortic arch should be 1,6+/-0,23) didn't allow to exclude CoA assumption. The difference between Ao and PA dimensions in the three vessel view shows that PA/Ao is relevant parameter[18] in the CoA diagnosis. Thanks to that we could identify a group of patient who will need monitoring during prenatal life[19-21], prostin infusion and probably surgical intervention in the first days of postnatal life. We also believe that it is worth to make longitudinal observations in cases of false (+) prenatal diagnoses (based on case nr 3). Despite good statistical studies we are still observing false positive diagnosis in the clinical practice so every patient needs to be treated individually[22].

It's very important in the postnatal care to include ultrasound data from patient prenatal period[1].

False negative diagnoses of fetal CoA are also an important problem[23], but this was not the aim of our analysis in this report.

CONCLUSION

In case of suspicion of CoA based on screening prenatal ultrasound examination, detailed fetal echocardiography is recommended with postnatal follow-up, as a "gold standard".

Narrowing of aortic isthmus in fetal life in the 2nd half of pregnancy may be a structural anomaly but may be also temporary event due to functional abnormalities in fetal circulation.

Aortic coarctation is still a difficult diagnosis in prenatal life.

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Division of work:

Barbara Swięchowicz: first draft, data collection

Maria Respondek-Liberska: idea of the article

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