Case **report**

ROUTINE THIRD TRIMESTER FETAL CARDIAC EVALUATION: TIME FOR CONSIDERATION

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Abstract

Most heart defects form between 4 and 6 weeks after fertilization. The detection rate is still growing. Despite significant progress in prenatal diagnosis some cases still go undetected. We present two cases of similar defects: prenatally detected and undetected, both presenting with a normal four chamber view in mid-pregnancy. We compared the follow-up of both neonates along with sustained health and economic consequences. The dynamics of the development of heart defects during prenatal life suggests the legitimacy to perform additional, late echocardiography exams (35-38 weeks of gestation)

Key words: *prenatal diagnosis, heart failure, critical aortic stenosis, HLHS*

BACKGROUND:

The development of prenatal cardiology, which took place in the 80s and its incredible acceleration in the twenty-first century has brought, apart from progress, new dilemmas. On the one hand we have the instruments allowing us to precisely perform a prenatal examination and give a diagnosis, on the other hand, it turns out that many defects still go undetected.

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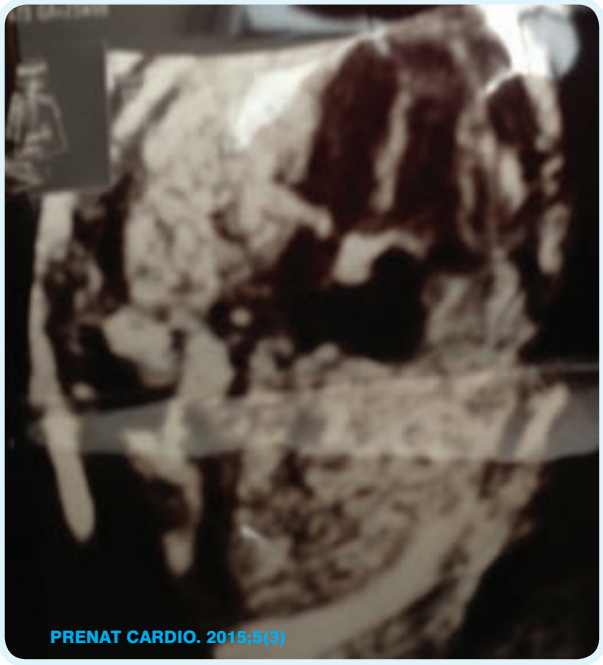
In the available literature we most commonly find descriptions regarding diagnostic or therapeutic success. This time we present a case of a prenatally undetected heart defect and the consequences regarding both neonatal follow-up and budget costs for the treating unit, compared to the analogous heart defect, which was diagnosed prenatally and treatment of the newborn was undertaken immediately after birth.

Case nr 1:

Patient aged 28, prima gravida, low risk pregnancy, family medical history irrelevant.



Fot.1: Image from correctly performed mid-pregnancy screening ultrasound at 21 weeks of gestation confirming normal fetal 4 chamber view (courtesy of the patient). Critical neonates condition after birth

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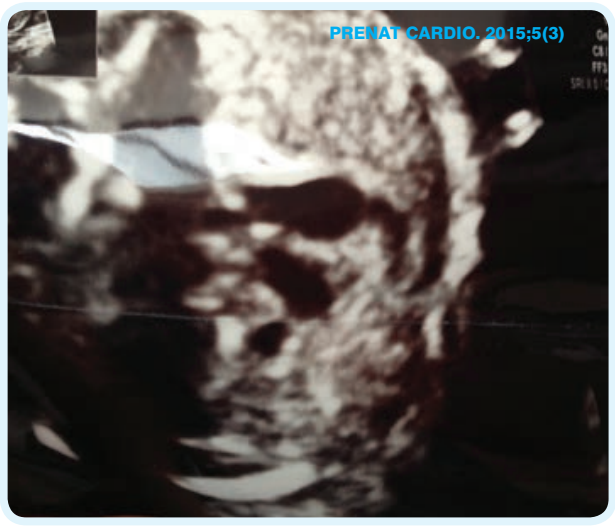
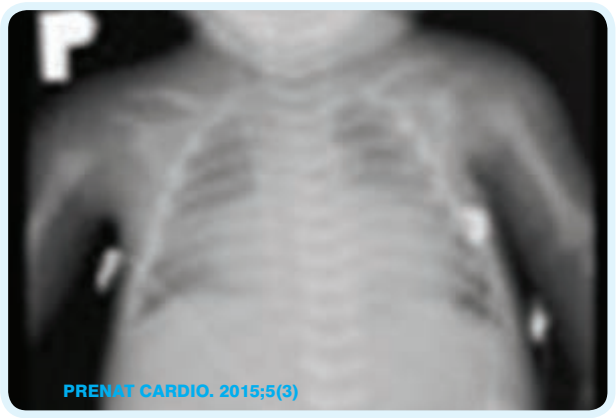


Fot.2: Image from correctly performed mid-pregnancy screening ultrasound at 21 weeks of gestation - normal 3 vessels in mediastinal view (courtousy of the patient). Critical neonates condition after birth



Fot.3: Chest X-ray after performed cardiac surgery (14 days after delivery)

Prenatal diagnosis in local community obstetrical office (12 ultrasound exams) during pregnancy were described as normal.

First trimester ultrasound exams were at 10 and 12 weeks of gestation, than at 21 weeks of gestation - location, size, cardiac axis, and 4 chamber view normal. Proper heart function, right and left outflow tracts normal. Flow FO R-L." (Fot.1,2).

Following US exams were conducted at 24,27,30,33,36,38,39,40 weeks of gestation expecting the birth of a healthy newborn.

Vaginal delivery was at 40th week: green amniotic fluid, placenta with deposits, short umbilical cord were registered in medical history. The newborn's birth weight was 3100 g, 8 points in the Apgar scale. A few hours after delivery respiratory and circulatory failure occurred and the neonate was transported from local community hospital to larger city (61 km), and 2 days after birth was transported to our referral center (237 km) with suspected congenital heart defect. Echocardiography on the 3rd day of life showed: normal heart position, and venous return, foramen ovale 3.5 mm with left-to-right shunting 1m / s, tricuspid valve with normal flow, mild dual-stream regurgitation, hypoplastic mitral valve with a small opening, flow into the chamber appeared as reversal flow- no emptying of the chamber into the aorta, hypoplastic, atretic aortic valve, enlarged left ventricle, akinetic right ventricle borderline size with good contractility and normal pulmonary artery, however with significant pulmonary valve regurgitation, PDA 7mm with right-to-left shunt, ascending aorta 6 mm, 6.5 mm transversal aortic arch, descending aorta 4,4mm, without evidence of coarctation.

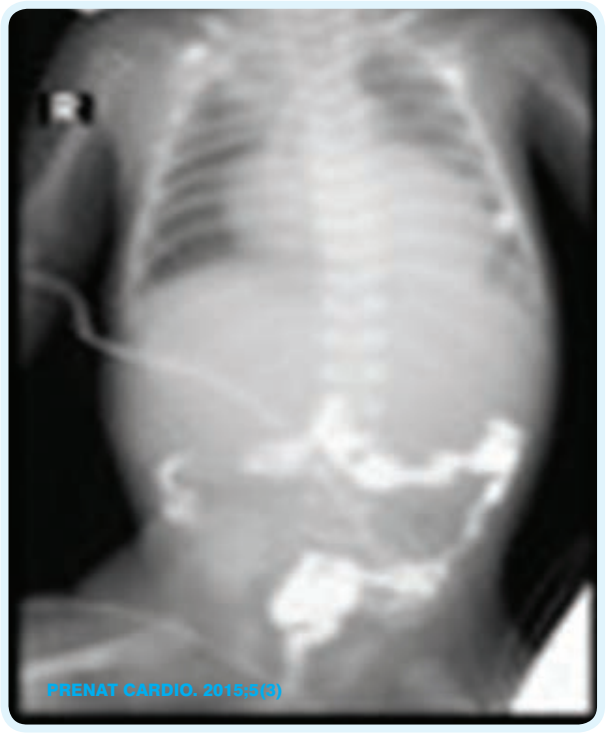
Conclusions: Enlarged akinetic LV with MV hypoplasia and aortic valve atresia with patent ductus arteriosus and foramen ovale.

At 14th day of postnatal life cardiac surgery cardiac surgery was performed: pulmonary artery banding

Comparison of the cases	Case 1 SA/HLHS	Case 2 SA
Prenatal diagnosis	Yes	Yes
Type of pregnancy	Low risk pregnancy	High risk pregnancy
Number of USG	14	6
Number of Echocardiograms	0	5
Detection of heart defect	No	Yes
Week of detection		30 hbd
Delivery at referral centre	No	Yes
	Neonatal follow-up	
Type of delivery	Vaginal delivery	CS
Week of delivery	40 weeks of gestation	39 weeks of gestation
Gender	Boy	Boy
Birth weight	3100	3320
Transport of neonate after delivery	2x 61 km , 237 km	no
Number of interventions	14th day – cardiac surgery, 16th and 46th days cardiac catheterization and Rashkind procedure 2 x *	1st day: cardiac catheterization and aortic balloon valvuloplasty
Complications after procedure	2 x cardiac arrest	None
Patient status	Severe	Good
Discharged home	Currently still being treated in the Department of Cardiac Surgery	21days after delivery

Table 1. Comparison of neonatal follow-up based on the two cases, both with normal four chamber view and 3 vessels view in midgestation

* 14 days– banding of the pulmonary arteries, stent placement into PDA; 16 days – first Rashkind procedure; 46 days – second Rashkind procedure



Fot. 4: X-ray: gastrointestinal tract passage (44 days of life)

(Fot.3) with stent placement into the ductus arteriosus. At 16th day of postnatal life due to decrease in oxygen saturation Rashkind procedure was performed. During the procedure, cardiac arrest with successful resuscitation, occurred twice. On 44th day radiological gastrointestinal tract passage was performed due to poor weight gain (Fot. 4). At 46 days of postnatal life a second Rashkind procedure was performed (Table 1).

At 101 days of postnatal life our patient was is still in the hospital.

Case nr 2:

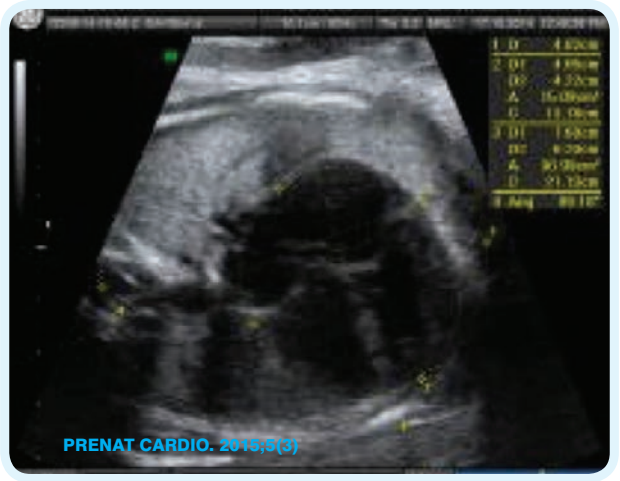
Pregnant woman aged 32, G 4 P 2: high risk pregnancy-poor obstetric history: G1 spontaneous abortion, G2 (2012) healthy son, G3 spontaneous abortion,

G4 US at 1st and 2nd trimester - no abnormalities, 4 chamber view and mediastinum were normal;

At 30th week of gestation because of past history the patient was referred for prenatal echocardiogram and left ventricle outflow tract obstruction was detected (exam performed by physician with the Section of Fetal Echocardiography and Prenatal Cardiology with the Certificate of the Polish Ultrasound Society).

At 33rd week: a normal 4 chamber view was still present, and Doppler blood flow through the aortic valve was 1,5 m / sec.

At 36th week again there was normal 4 chamber view and mediastinum, however at the level of aortic valve the blood flow was 3 m / sec by continuous wave Doppler.



Fot. 5. Normal 4 chamber view in the third trimester (case 2). Critical neonates condition after birth

At 38th week there was still a normal four chamber view, heart size was at the upper limit of normal, and blood flow through the aortic valve was similar to that at 36 weeks.

The 39th week echo: 4 chamber fetal heart normal with preserved left ventricular contractility (SF 52%), with hypertrophy of the left ventricle heart muscle and septum thickness in diastole was 7.8 mm, with V max for aortic valve 3.8 m / sec.

From the point of view of prenatal cardiology at 39 weeks this was a critical defect requiring elective aortic balloon valvuloplasty after delivery.

Two days later caesarean section was performed and newborn weighing 3320 g was delivered, Apgar score was 9 points, and he underwent aortic balloon valvuloplasty on the first day of postnatal life with a very good effect. Discharged home after 21 days of hospitalization.

The boy in the first 12 months of postnatal life did not require cardiac surgery and had mild aortic stenosis with good LV function

DISCUSSION

Most heart defects form between 4th and 6th week after fertilization[1]. They occur in one case per 100 pregnancies, genetic syndromes however such as Down syndrome considerably less - one case per thousand [2,3,4].

Recent years have seen significant progress in the detection of heart defects by use of ultrasound [1,5,6,7,8,9,10,11,12,13,14]. This progress is most evident in the case of HLHS (or hypoplasia of the left ventricle, hypoplastic left heart) [15.16]. This defect is the most often prenatally diagnosed heart defect not only in the world but also in Poland[17,18,19]. It amounts to 1,4-8,6% of all congenital heart defects. According to Polish Registry of Fetal Cardiac Anomalies (www.orpkp.pl) the detection rate of this defect increased from 22 cases in 2004 and 79 in 2012, up to 92 in the year 2014 [19,20].

The detection of HLHS may seem relatively simple. This defect can be visualized as early as 12-13 weeks of gestation and routinely at 18-22 weeks. Hypoplastic left heart syndrome in obstetric fetal heart screening studies presents itself as an incorrect image of the four heart chambers with a significant predominance of the right side. If the underdevelopment of the left heart is an isolated defect, the fetus develops normally, without hemodynamic disturbances [21,22]. The defect is clinically "silent" for the obstetrician, the gravida and even for the neonatologist in the first day of postnatal life.

Fetuses with left heart pathologies require monitoring because the echocardiographic image may be quite different at 20 and 40 weeks of gestation [23,24]. Neonate with isolated HLHS usually does not require any treatment in utero, is usually born at term with normal weight and no evidence of heart failure or abnormal auscultatory symptoms.

HLHS may not present in the classical way but can evolve during pregnancy [23,24,25], especially if the defect occurs with critical aortic stenosis and progressive reduction of the left ventricle. [26,27,28]

In some cases, prenatal aortic valve balloon valvuloplasty may diminish progress in the development of defects, and balloon septostomy can help improve outcomes for children with restrictive foramen ovale or intact atrial septum [29].

The defect, detected between 18-22 weeks and qualified as aortic stenosis, can change its character up to the time of delivery and evolve into the classic defect in the form of hypoplastic left heart [1,8,9,10]. The above reports provoke the conclusion that, in addition to routine cardiac echocardiography in the period of 18-22 weeks, repeat echo should be performed routinely at 28-35 weeks of gestation. By doing so, defects that were "silent" in both the 1st trimester and mid-pregnancy, can be detected in the 3rd trimester.

"Late" prenatal diagnosis in the 3rd trimester gives a chance for transport in utero, preparation of the mother and her fetus but above all, the team of clinicians needed for optimal perinatal management (case 2).

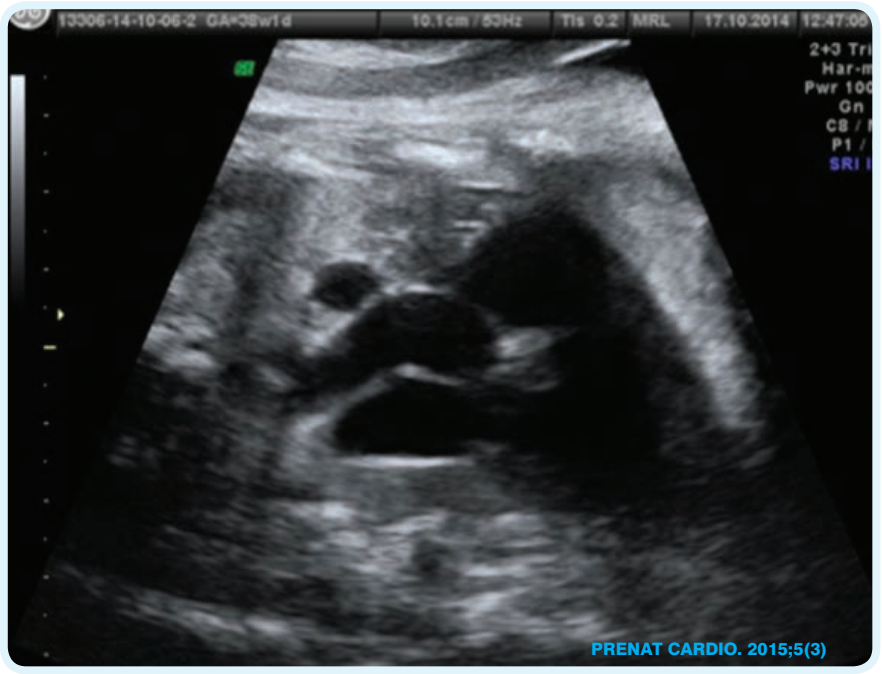
Prenatal cardiology in the 3rd trimester of pregnancy therefore seems reasonable, not only in the case of previously detected defects. Similar conclusions can be drawn on the basis of the article by Tworetzky et al. The authors describe that changes in the left ventricle were detected in the US between 28-35 weeks of gestation.

Another issue was a very low prenatal detection rate of defects in American multicenter studies published in 2015: 10 cases per 117 newborns (8.5%) with critical aortic stenosis [30].

Prenatal diagnosis of congenital heart disease improves results of subsequent cardiac surgery and significantly affects the follow-up of the neonate [31,32]. In Poland every year about 80-90 infants with HLHS require surgery for this reason in the first month of life. [33]

Detecting heart defects prenatally in the third trimester of pregnancy (33 and 36 weeks) during routine ultrasound, especially critical heart defects can dramatically affect the follow-up of patients [34].

The social aspect of such actions - is to shorten the length of stay of the newborn in the hospital. This implies



Fot.6. Normal mediastinal view in the 3 trimester (case 2). Critical neonates condition after birth

a reduction of direct costs- child's stay and indirect - absence of one parent from work (often both).Transport in utero reduces the cost of transporting newborns in critical condition (specialized transport teams, air transport) to the referral centers.

Would the prenatal detection of the heart defect in presented case 1 improve the prognosis for the newborn? Was the original pathology in this case aortic stenosis

or maybe myocarditis in the 2nd half of pregnancy? These questions are difficult to answer ex post. If the electronic stethoscope in the form of fetal echocardiography was used before delivery, transport by ambulance almost 300 km could have been avoided.

The second case with prenatally diagnosed congenital heart defect occurring with normal four chamber and mediastinal view (as case 1) by means of transport in

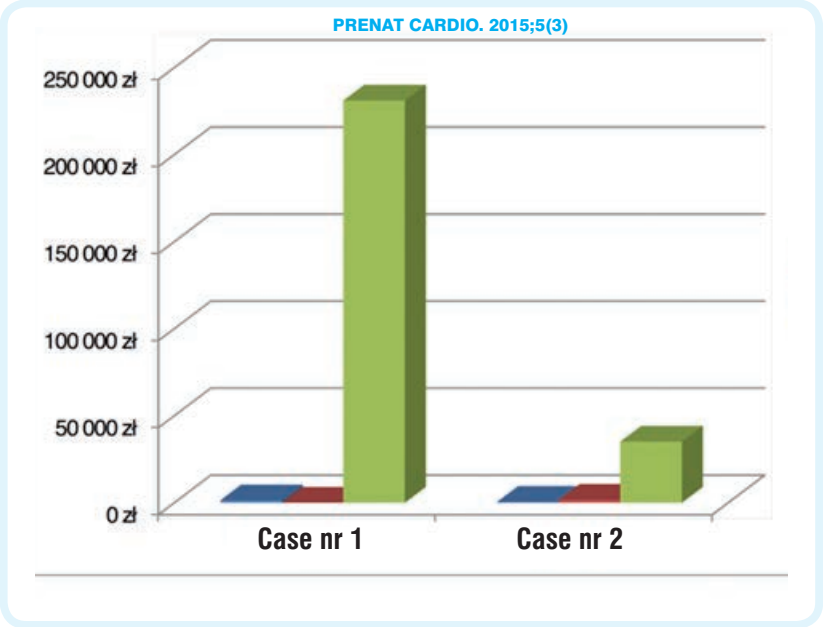


Figure 1: Costs of USG, Echo and hospitalization in presented cases

Costs	Case 1 SA/HLHS	Case 2 SA
US exams	1 400 zł	600 zł
Echocardiograms	0 zł	1 750 zł
Hospitalization	229 774,60 zł	32 849 zł
total	231 174,60 zł	35 199 zł

Table 2. Costs of US exams, Echo and hospitalization in presented cases with critical conditions after delivery

utero was qualified for aortic balloon valvuloplasty on the 1st day of life without the need for cardiac surgery and was discharged home 21 days after treatment. The prenatal cardiology team of specialists from different cities in Poland (Szczecin and Lodz), and specialist team at our referral center took part in the relay for his life and health. Properly monitored fetus in late pregnancy, scheduled time, location and type of delivery, avoiding premature birth and direct treatment after delivery, proves the effectiveness of prenatal diagnosis, especially in critical congenital heart disease.

Summary data regarding the cost of treatment of both newborns proves that the problem of prenatal diagnosis has also an important financial aspect. (Table 2), (Figure 1)

In both presented cases, the fact that the costs of their prenatal tests are comparable is quite obvious. However, the different quality in tests used in both cases has a diametric impact on the final result - the fate of both newborns (Table 1). Performing an ultrasound without fetal heart echocardiography may lead to diagnostic errors, defects may go undetected during the most appropriate time necessary in order to plan specialized treatment, consequently - generating unnecessary, enormous costs after birth.

CONCLUSIONS

Normal 4 chamber and mediastinal view at 1st trimester and mid-gestation ultrasound does not rule out the presence of heart defect in subsequent weeks of fetal or neonatal life.

1. The introduction of mandatory third trimester fetal echocardiography should be considered in order to avoid wrong decisions as to the place, time and type of delivery.

2. Late implementation of specialized cardiac intervention or cardiac surgery on the basis of case 1 and the high costs of prolonged hospitalization and rehabilitation, suggest the need to introduce mandatory echocardiography of the fetus before delivery in order to optimize transport in utero.

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Strzelecka I: data collection, first draft, literature search

Płużańska J: English version, submiting manuscript

Wegrzynowski J: detection of anomaly, data collection, work with manuscript

Moszura T: neonatal treatment, data collection, work with manuscript

Słodki M: work with manuscript

Respondek-Liberska M: concept of the research, work with manuscript, final version