

● Original Report

FETAL ECHOCARDIOGRAPHY IN UNCOMMON PRENATAL CARDIAC ANOMALIES: RIGHT ATRIUM DIVERTICULUM, INTERVENTRICULAR SEPTAL ANEURYSM, LEFT AND RIGHT VENTRICLE DIVERTICULUM – REPORT FROM REFERRAL CENTER FOR FETAL CARDIOLOGY IN POLAND

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Abstract

The prenatal detection of congenital anomalies of heart walls is very rare. We present a unique series of 8 cases with prenatal echocardiographic monitoring, treatment and postnatal follow-up, providing new insight into this "mysterious" heart problem..

Key words: diverticulum, interventricular septal aneurysm, left ventricle diverticulum, abnormal fetal heart wall

INTRODUCTION:

The prenatal detection of congenital anomalies of heart walls is very rare. We present a unique series of 8 cases with prenatal echocardiographic monitoring, treatment and postnatal follow-up, providing new insight into this "mysterious" heart problem.

MATERIAL & METHODS:

It was a retrospective analysis of our database from years 2014-2017 (plus one case from 2012). Total number of fetuses that underwent detailed fetal heart examinations in our unit during this time was 3135. The total number of fetal echo examinations in this period of time was 6419. During this time we had seven fetuses with fetal heart wall abnormalities. Additionally we also present one case that was supposed to be referred to our unit however there was fetal demise shortly after detection.

We present short medical review for each case with pictorial evaluation. The summary is presented in table nr 1 with prenatal data, perinatal management, delivery and postnatal data.

We also present the map of Poland with maternal home address.

No specific statistical data was performed for this series of cases

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**Case nr 0**

It was first pregnancy of healthy 25 year old woman, who had normal US scan at 12 weeks of gestation and was rescanned at 23 weeks (Fot.1 cine). This case was sent to our center for counseling via our internet data base from Rzeszów (400 km from our center). On the level of the 4 chamber view there was severe cardiomegaly, myocardial impairment and suspicion of an apical LV aneurysm. However one day later there was fetal demise. ("case 0", photo + schematic drawing + table nr 1)

Case nr 1

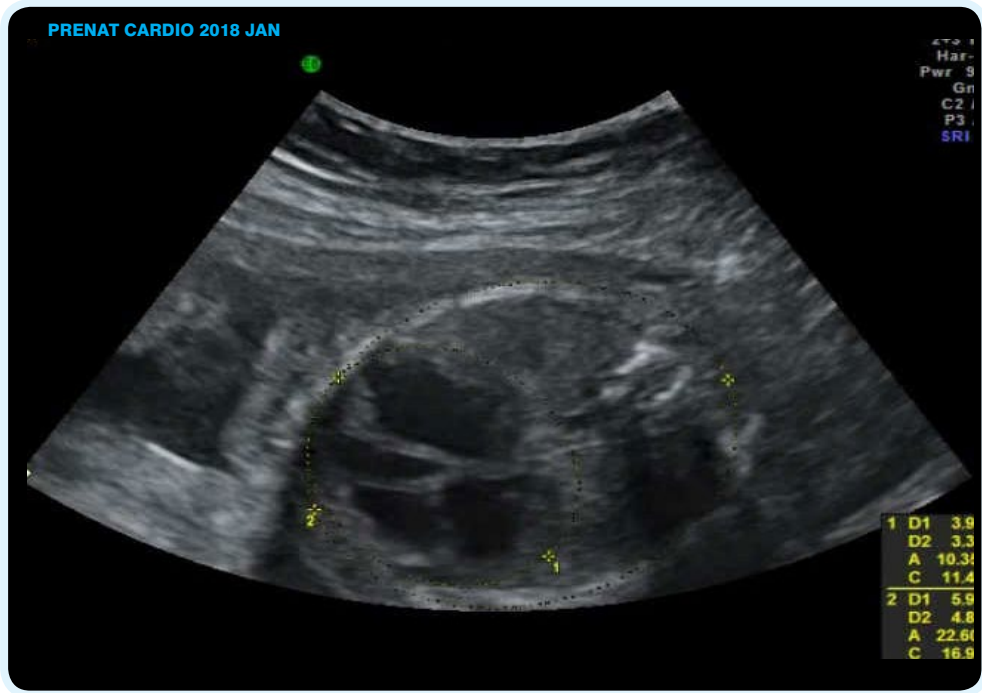
It was second pregnancy of 24 year old healthy woman (from first pregnancy healthy boy) who had normal first trimester of pregnancy, normal mid gestation scan and at 31 weeks of pregnancy "dilatation" of the right atrium was picked up during obstetrical ultrasound screening. The fetus was referred to our center. There was disproportion at the level of atria, ventricles and big vessels with an abnormal dilatation of the RA and partial filling of RA in CD (Fot. 2 Case 1 + schematic drawings+ table nr 1). As lack of blood flow in almost 50% of the "enlarged" right atrium resembled the presence of blood clot in this area, the pregnant woman underwent fetal echocardiography for the next 10 days on the hospital basis and due to accompanying elevated coagulogram values low



molecular weight heparin was introduced in the obstetrical department. No further progression was observed and follow-up was continued on an outpatient basis. After delivery the child underwent neonatal echo and CT angiography confirming prenatal findings but there was good blood filling of the entire heart (no clot after birth).

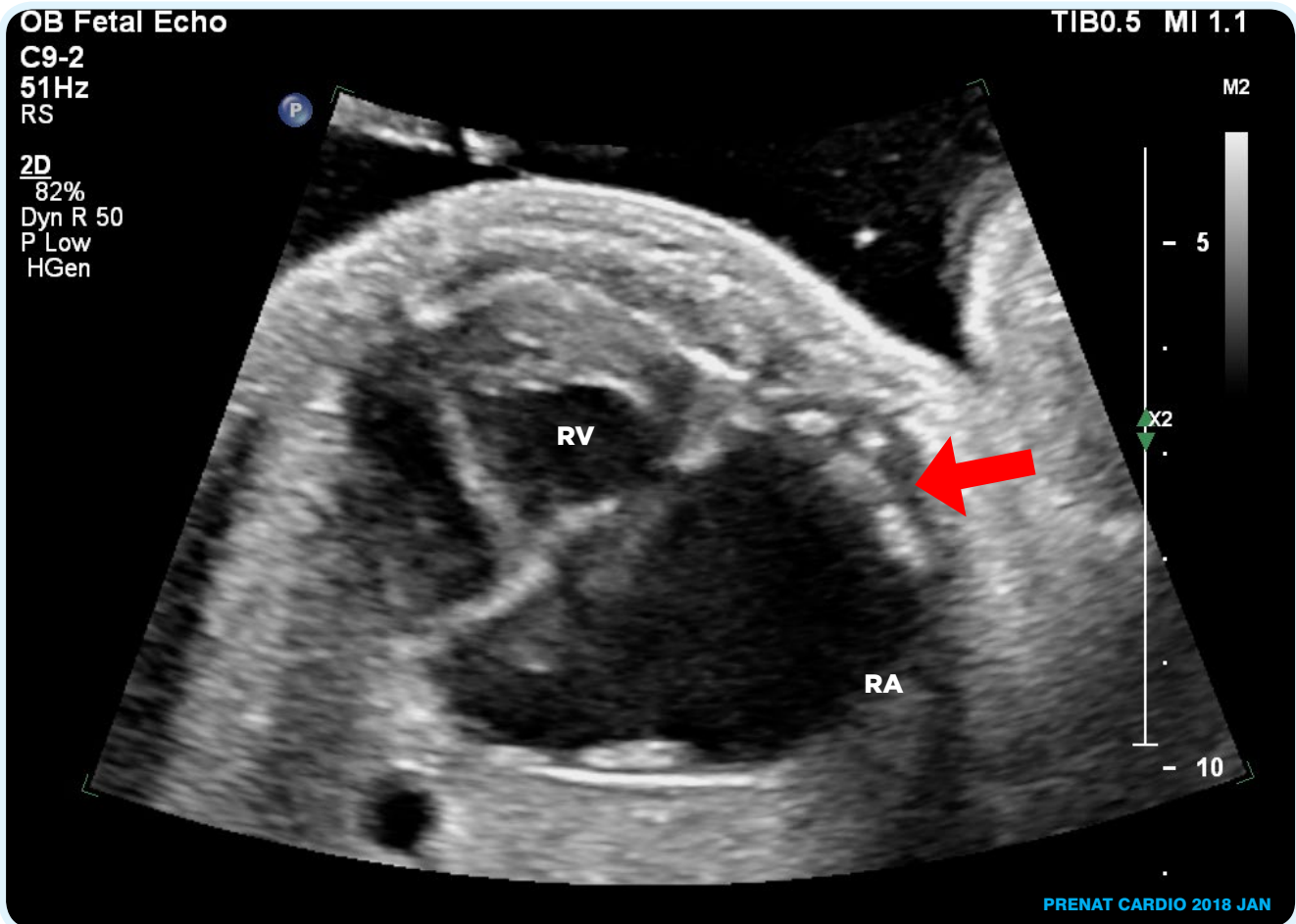
Case nr 2

It was first pregnancy of healthy woman age 31 with normal 13 and 20 week scans. At 29 weeks of gestation during obstetrical screening abnormal 4 chamber was detected and fetus was referred to our center for targeted echocardiography.



Fot. 1 (cine): Case 0 - Fetus with suspected myocarditis due to general hypokinesis, however there is lack of contractility around apex (arrow). Next day fetal demise

Cinematic content - place cursor over above graphics and click to play



Fot. 2. Case 1 - 4chamber view with dilatation of the right atrium (RA)



Fot. 3 (cine): Case 2 - Abnormal 4 chamber view with "dilatation" of the RV

valve regurgitation. The newborn baby was discharged home of the day 10th

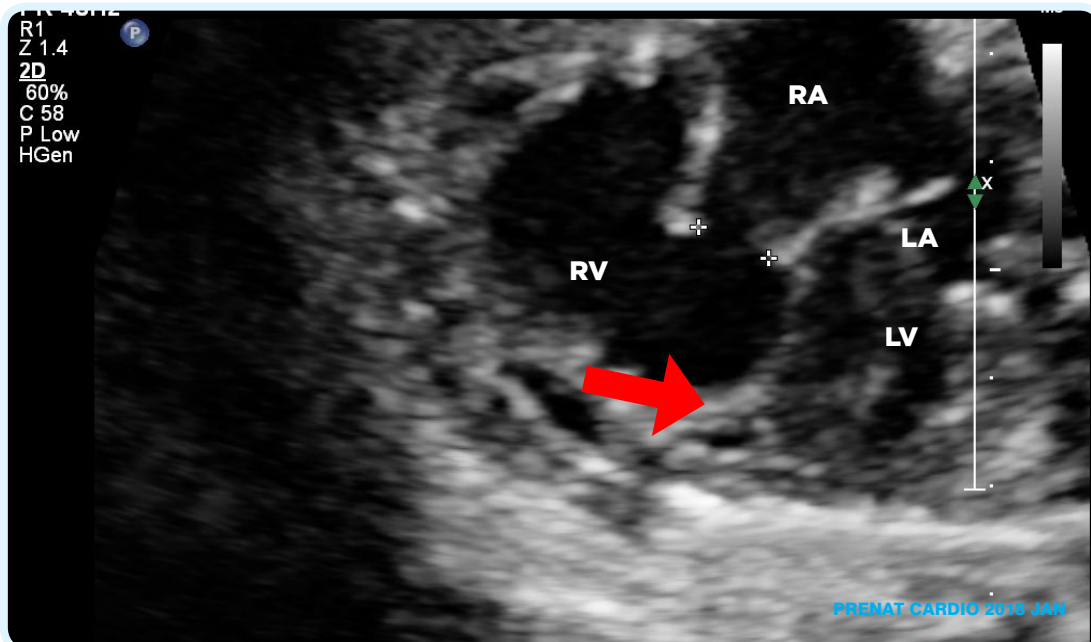
Two years later his mother is currently expecting second child, with no abnormal findings on fetal echocardiography.

Case nr 3 (2015)

20 year old woman, in her first pregnancy, who had frequent US exams due to her uropathy. At 35th week of gestation fetal cardiomegaly was detected and fetus was

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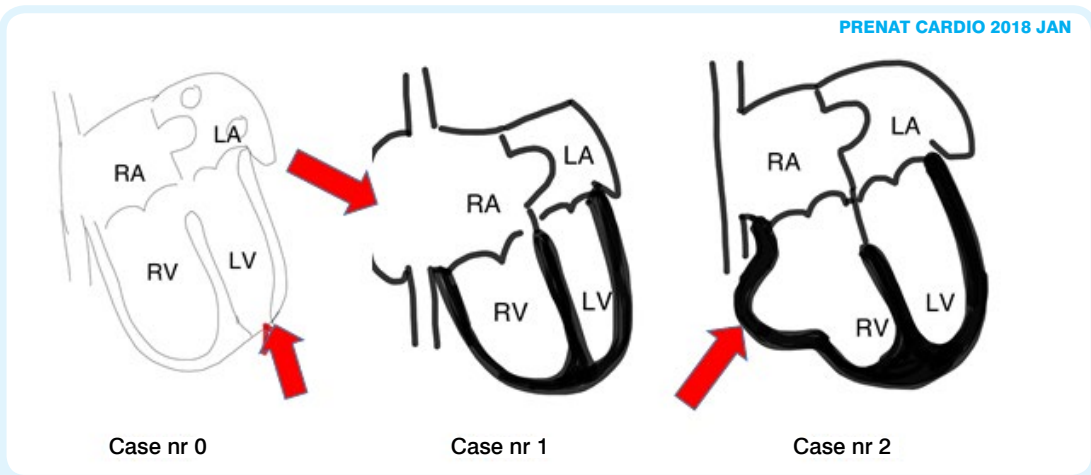
Fot. 4. Case 3 - Abnormal 4 ch view with huge dilatation of right ventricle (RV) and right atrium (RA) and compressed left ventricle (LV)

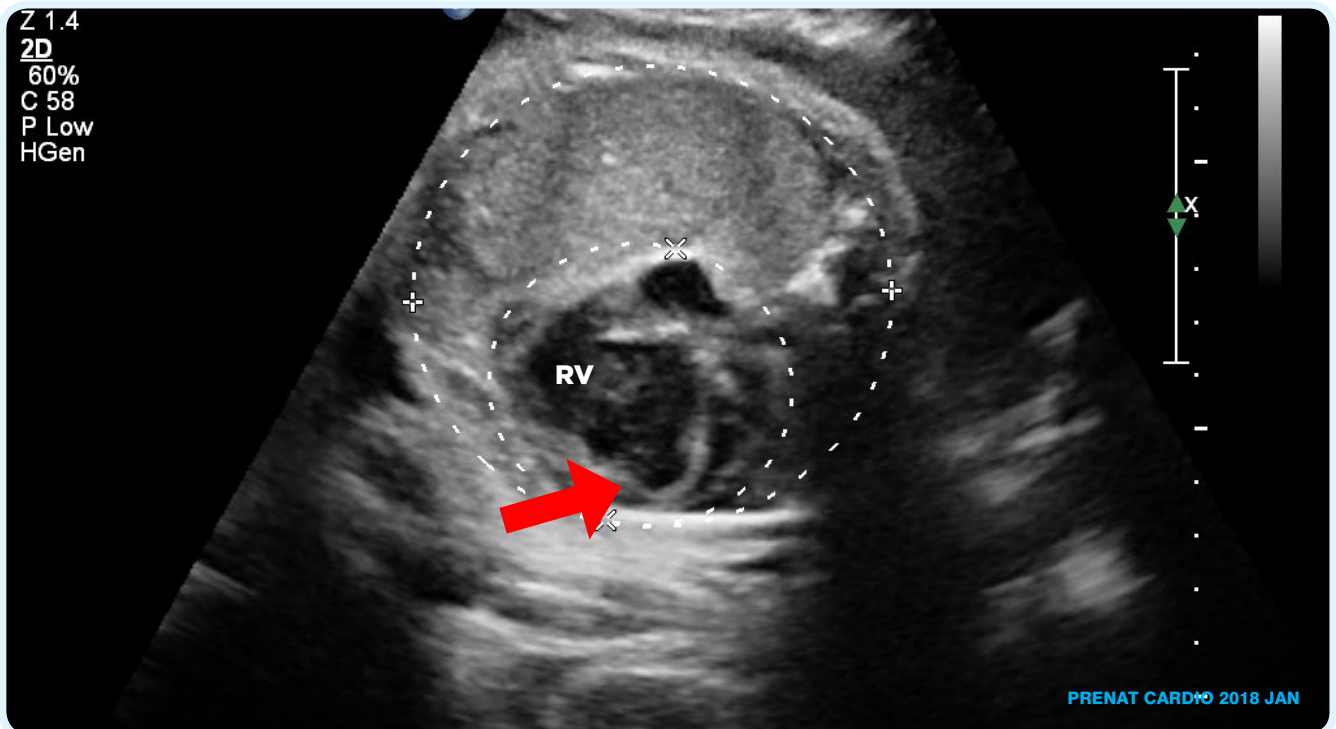


The fetal heart was enlarged and there was an abnormal shape of the right ventricle wall suggesting "additional pouch" or "aneurysm" with almost no blood flow in this area, fetus presented with abnormal heart rhythm. (Fot. 3 Case 2 + schematic drawings + table nr 1)

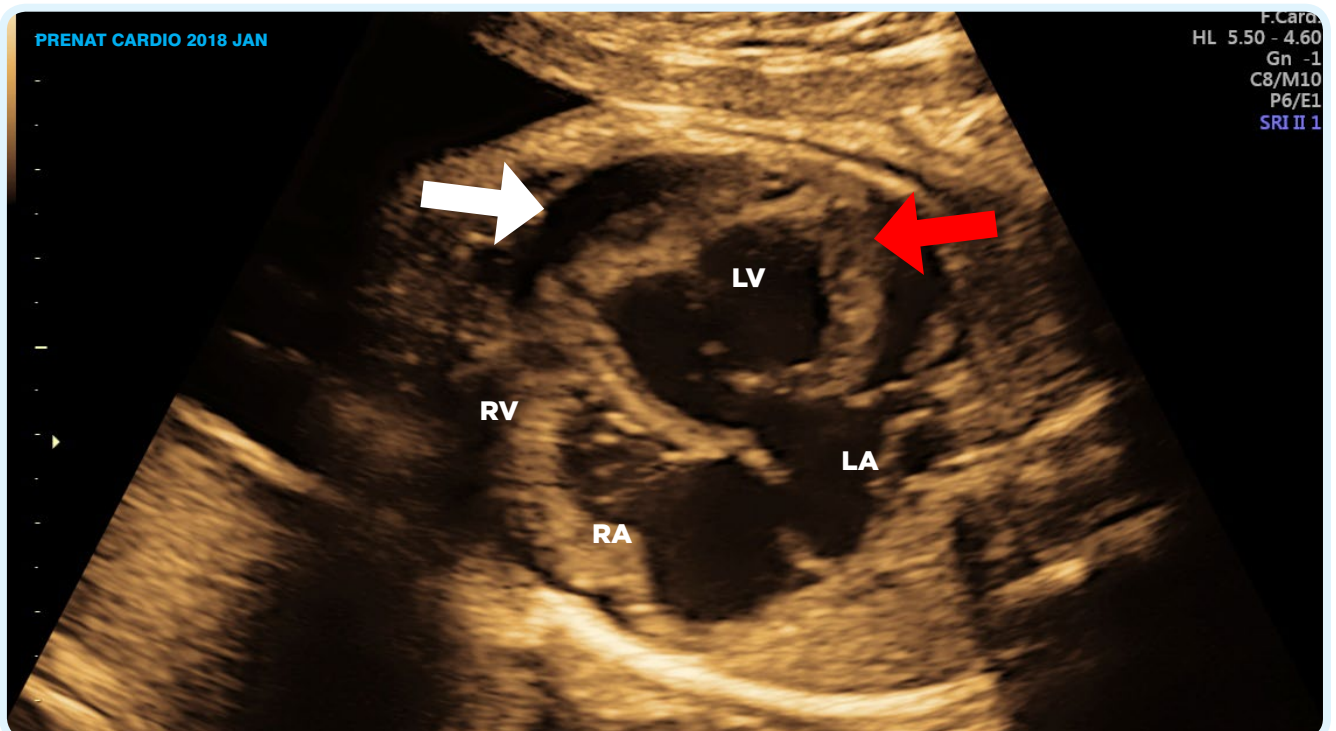
After delivery neonatal echo showed thickened right ventricle with trivial tricuspid

Fig.1. Schematic drawings of prenatal abnormalities in fetal echocardiography (cases 0 / 1 / 2)





Fot. 5. Case 3 - Fetal heart cardiomegaly HA/CA ratio 0,5 and abnormal 4 chamber view (1st and 2nd trimester US with no abnormalities; red arrow points aneurysm in lower part of the septum)

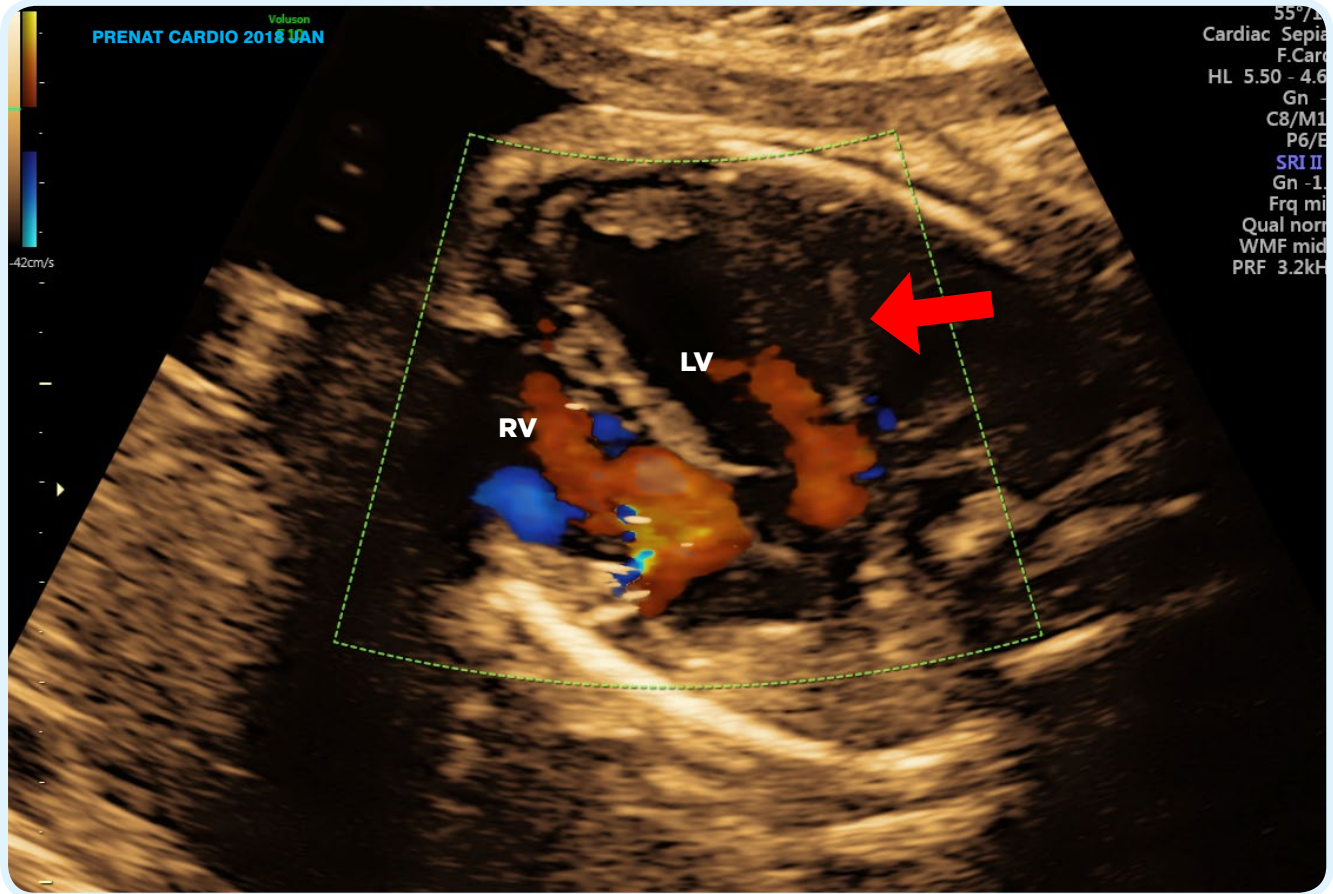


Fot. 6. Case 4: Abnormal 4 chamber view with aneurysm of LV (red arrow), pericardial effusion (white arrow) and cardiomegaly

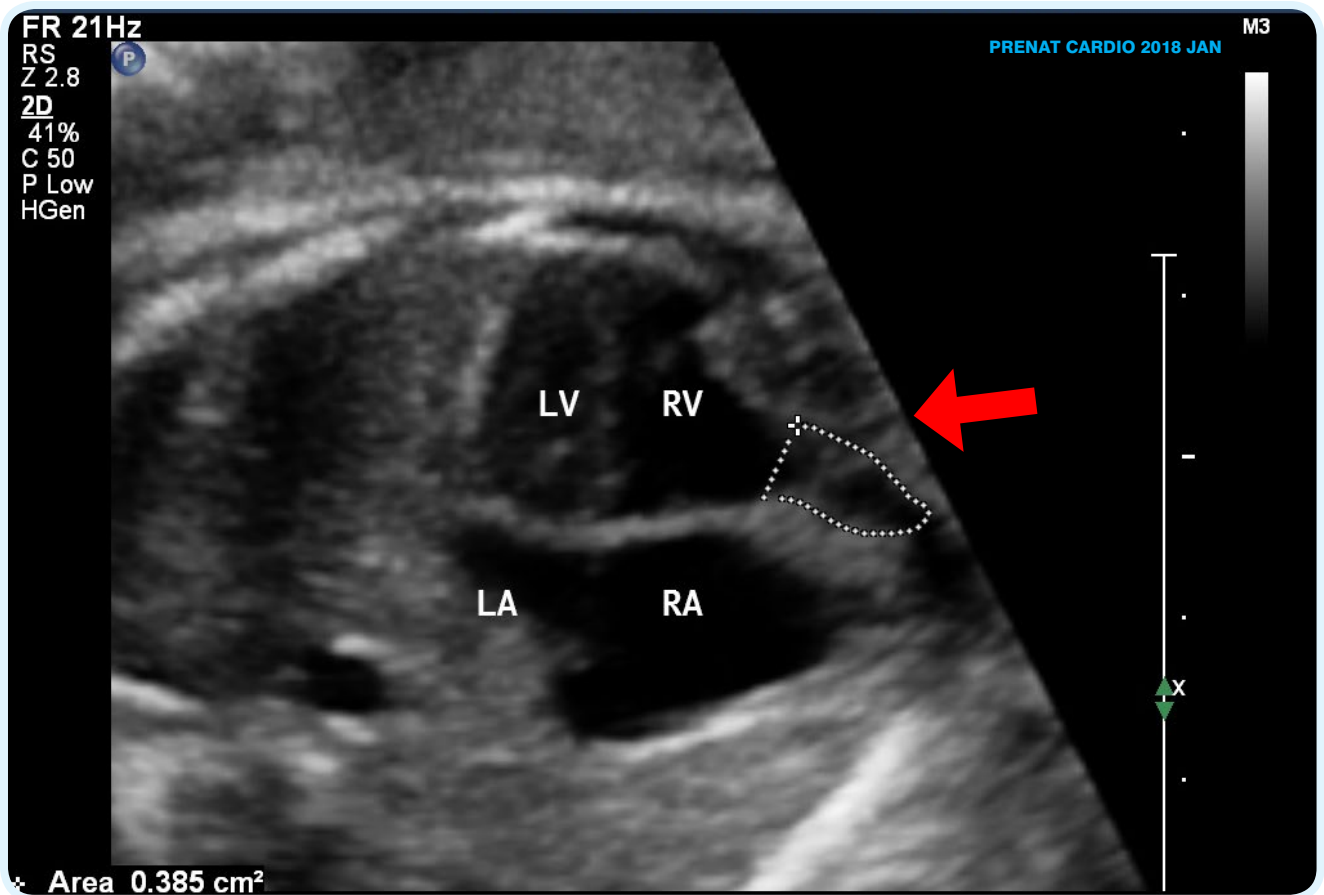
referred for targeted echocardiography to our center. This case was already published in 2016⁵ as a reason for emergency CS due to aneurysm of the muscular interventricular septum, however here we present new unpublished photos (Fot. 4/5 Case 3+schematic drawing+table nr 1)

Case nr 4:

It was second pregnancy of 26 year old woman (from first pregnancy 7 year old healthy boy), who had normal first trimester US and at 20 weeks of gestation aneurysm of the left ventricle was detected by obstetrical screening ultrasound and fetus was referred to our center.



Fot. 7. Case 4: 4 chamber view with color Doppler. Limited blood flow in color Doppler in left ventricle, comparing with the good filling of the right ventricle

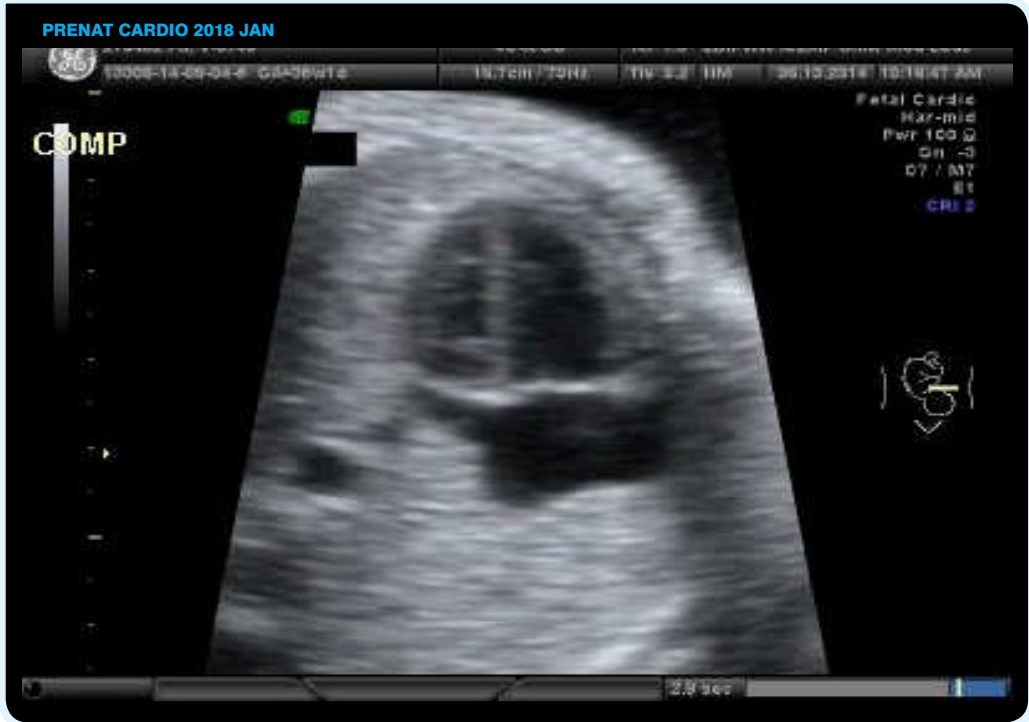


Fot. 8. Case 5 - Abnormal 4 chamber view with RV diverticulum



Fot. 9 (cine): Case 5 - cine of the 4 chamber view

In fetal echocardiography there was cardiomegaly, pericardial effusion, and in 4 chamber view there was abnormal wall of the left ventricle. (Fot 6/7 Case 4 + schematic drawing+table nr 1) After counselling transplacental treatment was initiated, fetal condition improved and the gravida decided to deliver in her tertiary center (200 km from our town).

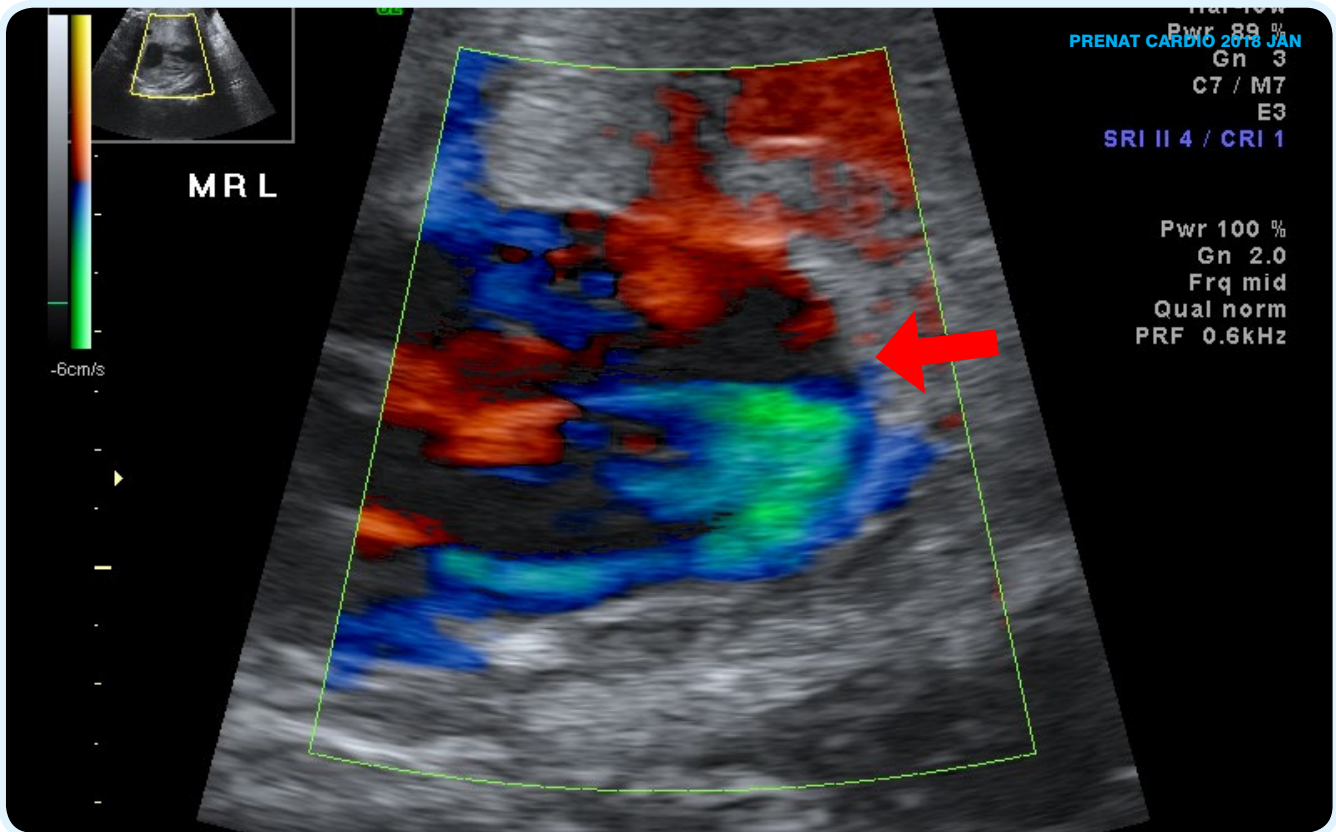


Case nr 5:

32 year old prima gravida with no underlying medical conditions was referred to our center for further examination because of bright spot in LV seen upon obstetrical scanning. Targeted fetal echocardiography revealed slight disproportion in favour of the right side. In the area of the right atrial appendage the right ventricle seemed to bulge inwards in

Cinematic content - place cursor over above graphics and click to play

the form of an aneurysm/diverticulum. (Fot 8/9 Case 5 + schematic drawing + table nr 1). The patient was monitored on an outpatient basis. Neonatal echo confirmed prenatal diagnosis of right ventricular diverticulum.



Fot. 10. Case 6 - 4 chamber view of the fetal heart with tissue Doppler: red and blue colors show good myocardial contractility; there is no contractility (grey area) around an apex

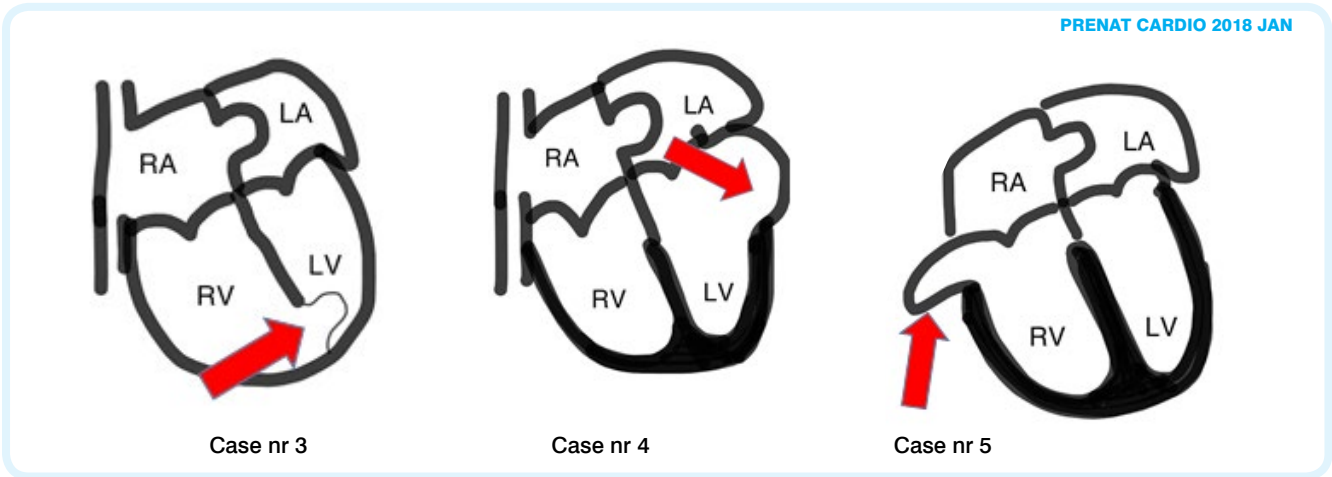


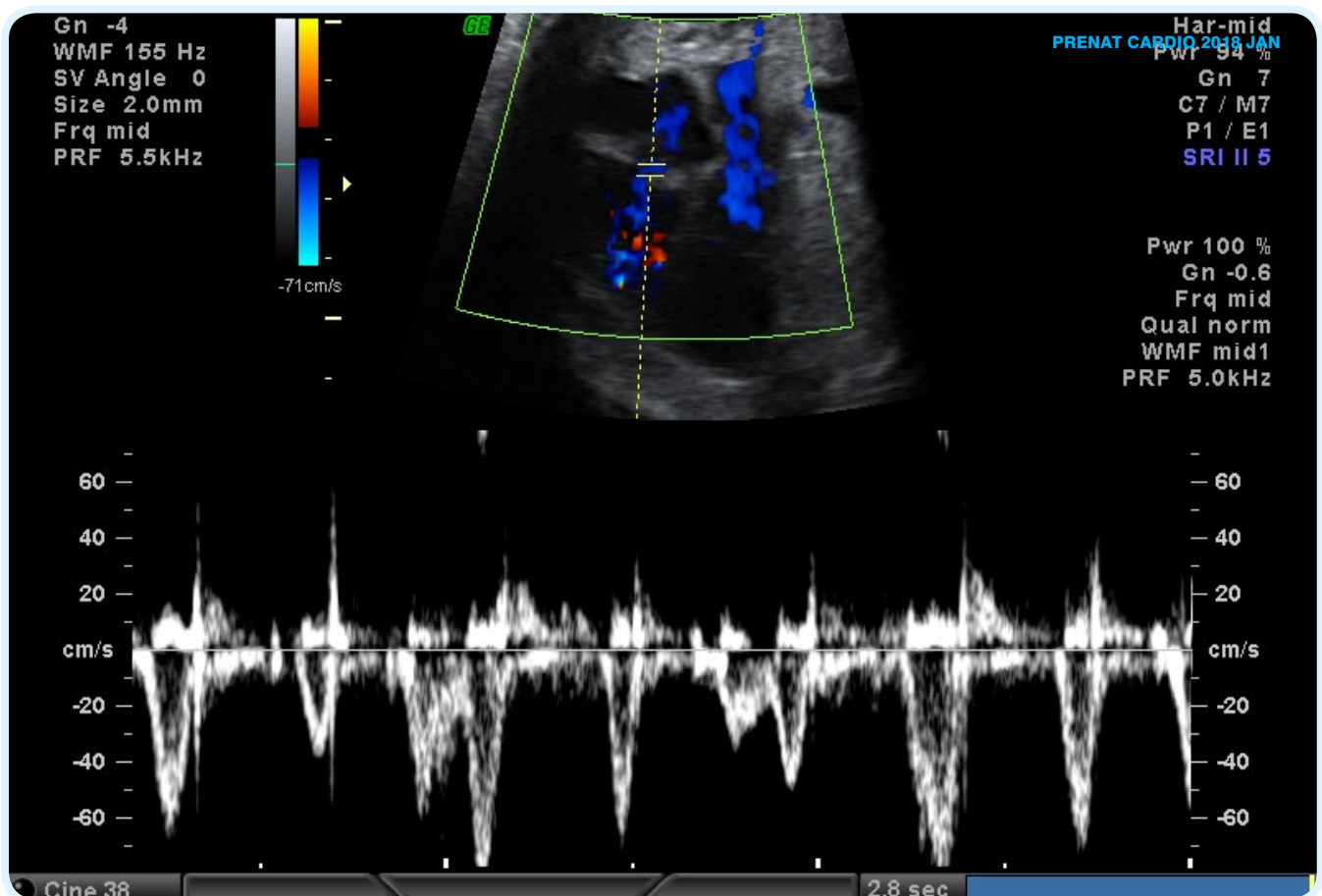
Fig.2. Schematic drawings of prenatal abnormalities in fetal echocardiography (cases 3 / 4 / 5)

Case nr 6:

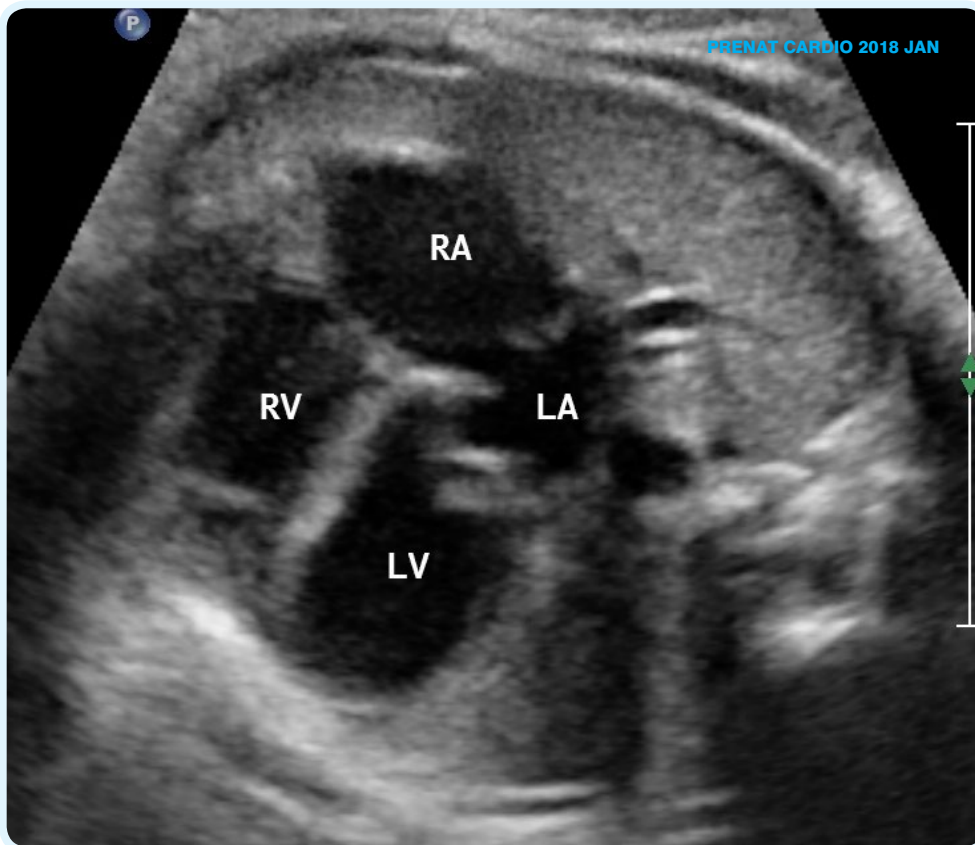
28 year old prima gravida with gestational diabetes due to fetal arrhythmia was admitted to her local hospital by her physician at 35 weeks of gestation where she was treated with antibiotics. She was later referred to our Center for fetal echocardiography after treatment completion. During scan fetus presented complex arrhythmia and LV apical aneurysm was diagnosed. (Fot 10/11 Case 6+schematic drawing+table nr 1)

Case nr 7:

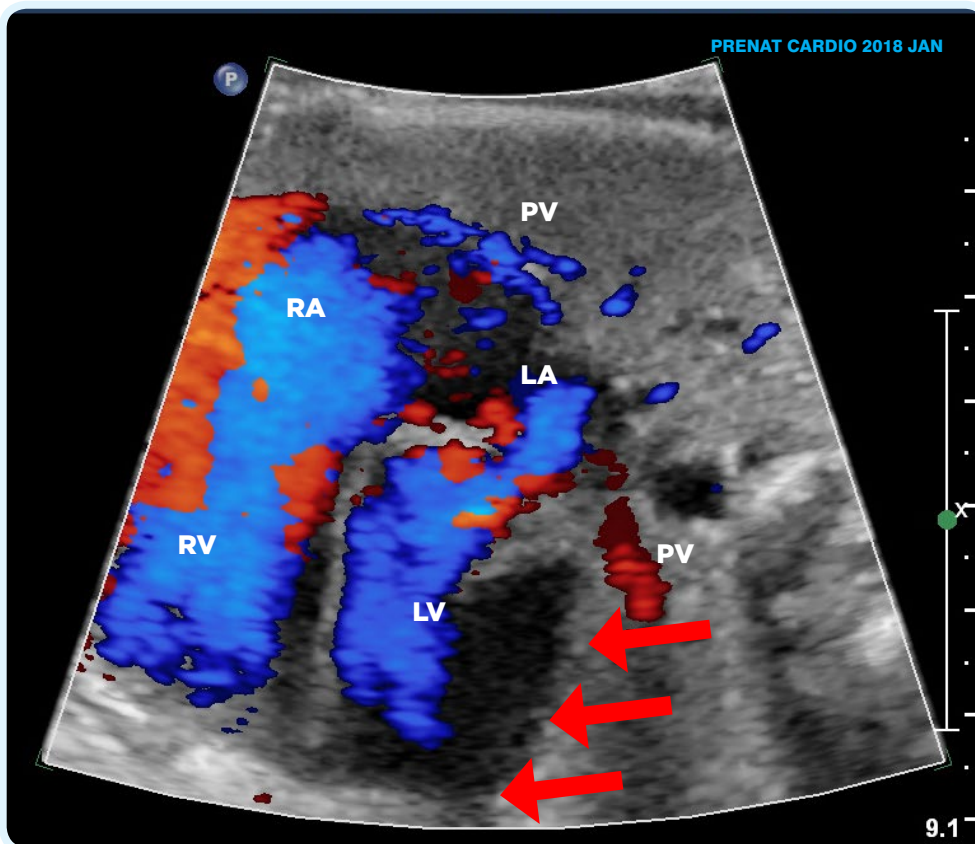
Midgestation scan of healthy, 28 year old gravida showed cardiomegaly, pleural effusion, first trimester scan was normal (NT=2,1mm, NB (+), 46,XX). The patient was referred to tertiary center for consultation and transplacental treatment was initiated. (Fot. 12/12 Case 7 +schematic drawings+table nr 1. After delivery the child was in good general condition, echocardiographic exam confirmed diagnosis with compromised left ventricular function (EF=31% Simpson method 4C ; 2C EF=18,5%).



Fot. 11. Case 6 - Doppler sample gate at the level of the tricuspid valve showing premature atrial contractions



Fot. 12. Case 7 - Fetal heart cardiomegaly, abnormal heart axis, enlarged left ventricle (LV)



Fot. 13. Case 7 - Fetal heart cardiomegaly, good blood flow in right atrium (RA), right ventricle (RV), left atrium (LA), but left ventricle (LV) is partially filled with blood; PV - normal pulmonary veins flow (PV); abnormal heart axis

Because of bidirectional flow across the patent ductus arteriosus, Prostin was administered in addition to Corotrope (0,4mcg/kg/min). TORCH and other intrauterine infections were excluded. Control echo showed good aortic and mitral flow with left-right-shunt across the patent ductus arteriosus and Prostin was discontinued. Over time Corotrope was replaced with oral medications (Carvedilol, Captopril, Spironol). The patient was discharged home in excellent clinical state.

DISCUSSION:

The prenatal detection of congenital anomalies of heart walls is very rare. The first prenatal diagnosis of a muscular IVS aneurysm was presented by Fujiwara in 2001 (16 years ago)¹. Two brothers diagnosed at 26 and 28 weeks of gestation and both after vaginal delivery were asymptomatic. Donofrio in 2002 published four patients spanning three generations affected with congenital myocardial disease². The youngest member of the family, diagnosed as a fetus with a large aneurysm of the muscular interventricular septum.

Both papers suggested screening family members of patients with muscular interventricular septal aneurysms to assess for silent familial myocardial disease. We plan to perform such investigations in families of our patients. In Poland in the Fetal Cardiac Anomaly Registry (www.orpkp.pl)³ in years 2004-2016 among 7249 fetuses the prenatal diagnosis of "LV aneurysm" was present in 3 cases and "RV aneurysm" also in 3 cases, so all together 6 cases/ 7249 is 0,08% (included data from our center). List of diagnoses

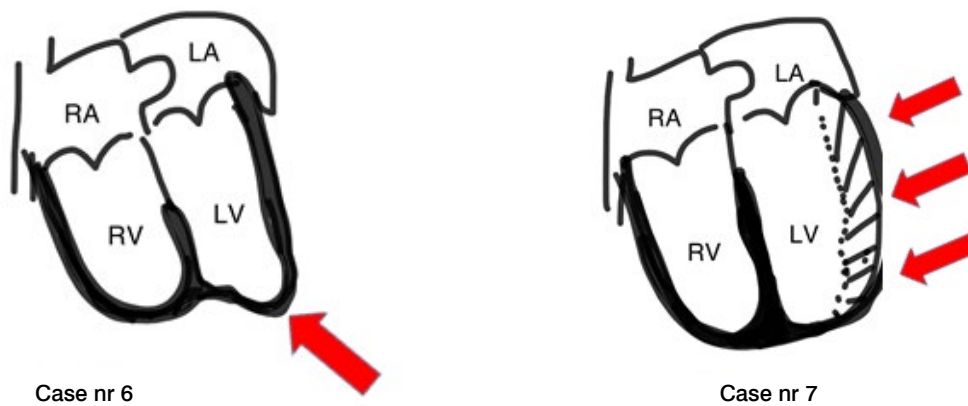


Fig.3. Schematic drawings of prenatal abnormalities in fetal echocardiography (cases 6 / 7)

adapted from European Association Pediatric Cardiology did not include "RA diverticulum".

There are several speculations in adult and pediatric literature about the possible cause of such abnormalities seen usually in clinically silent patients during their postnatal life⁴. Coronary artery abnormalities or embryological atypical myocardial development during the first trimester of pregnancy could "imprint" abnormal heart wall development. Are these abnormalities a base for postnatal "non-compaction" myocardial cardiomyopathy? Another interesting speculation could be created based on chart 1 (Map of Poland) – five fetuses were referred to our center from western part of Poland. An epidemiological inquiry would be interesting but unfortunately it is not possible in our Institute

In differential diagnosis one should take into consideration that "diverticula of the fetal heart" abnormalities are related usually to one wall or one chamber, whereas in typical myocardial hypertrophy it is usually an abnormality of the right chamber, left chamber and interventricular septum.

However this time we want to present this unique series of cases, which provides some new thoughts into this "mysterious" heart problem.

As there is not enough published data on this subject having in mind also intrauterine demise as possible outcome, we suggested to the families close echocardiography monitoring in the second half of pregnancies.

As there is a lot of data supporting first and 2nd trimester fetal echocardiography we again strongly advocate the value of the 3rd trimester echocardiography. In the observed cases fetal echocardiography provides a valuable tool for obstetricians in order to prolong pregnancy despite fetal cardiomegaly and structural

heart problems⁵. In utero transfer in these cases to a referral perinatal center was also important, especially for the fetus/neonate who was in critical state just before delivery. Postnatal severe condition was also present in case published by Carr⁶.

Such a short series of cases is probably not enough to recommend digoxin transplacental treatment, however none of the pregnant women showed any adverse effects or manifested complaints during the therapy (maternal

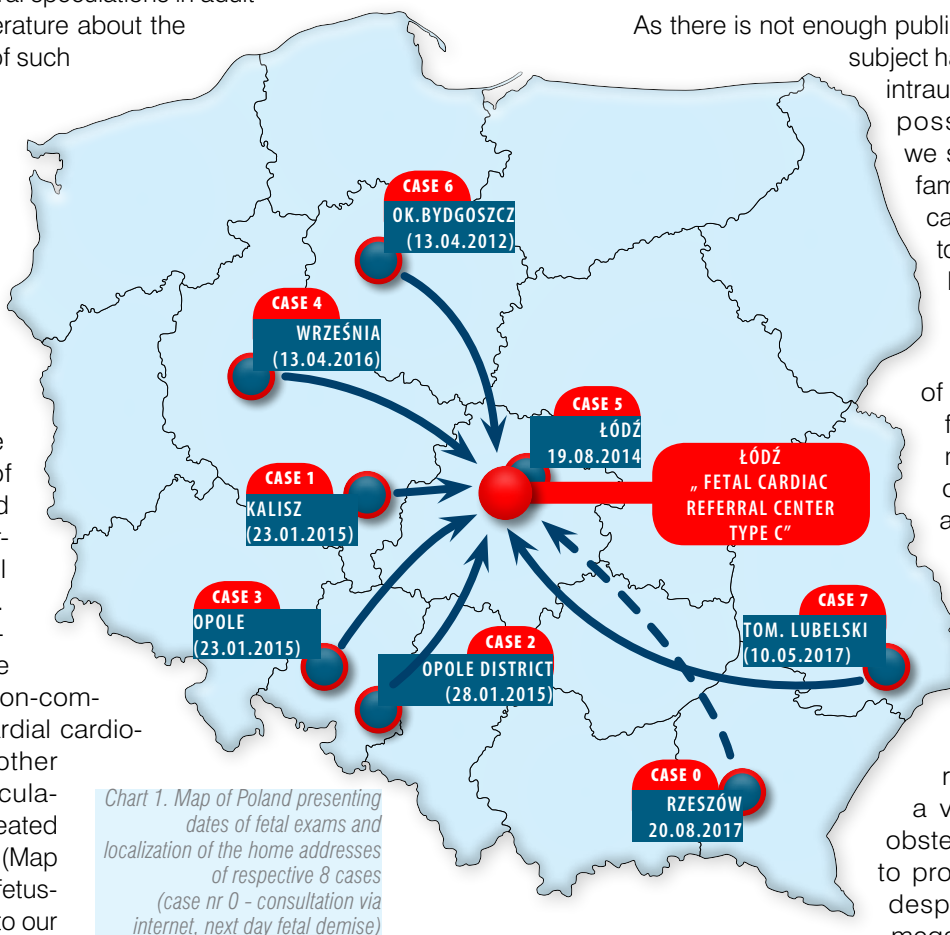


Chart 1. Map of Poland presenting dates of fetal exams and localization of the home addresses of respective 8 cases (case nr 0 - consultation via internet, next day fetal demise)

	Case nr 1	Case nr 2	Case nr 3	Case nr 4	Case nr 5	Case nr 6	Case nr 7
	Aneurysm of RA	Aneurysm of RA	Aneurysm of RV	Aneurysm of LV	RV diverticulum	LV aneurysm (apex)	LV dilatation
Gest age in weeks time of diagnosis	33	26	35	31	30	35	25
Heart rate	Sinus rhythm	Bigeminia	Sinus rhythm	Sinus rhythm	Sinus rhythm	Bigeminy, Trigeminy SVES	Sinus rhythm
Perinatal management	Digoxin, Steroids, Heparin	Observation	Emergency Delivery	Digoxin, Steroids	None	None	Digoxin, Heparin, Steroids
Delivery	CS	CS	CS	Vaginal	CS	CS	CS
Birth weight	4180	3180	3000	2900	1830	2940	4040g
Gender	Male	Male	Male	Male	Female	Female	Female
Apgar score	10/10	9/9	5/6/8	8/9	8/9	5/8	8/9
Neonatal condition	Good	Good	Cardio-respiratory failure	Good	Good	Good	Good
Hospitalisation (in days)	26	21	20	18	17	25	32
Follow-up	2 years	2 years	2 years	6 months	?	?	6 months
Clinical symptoms	None	None	None	None	None	None	None
Pharmacotherapy	Acesan	No cardiological drugs	Intensive Care Unit for 2 weeks. Dopamine, Corotrope Discharge: Acesan, Enarenal		No cardiological drugs	Cordarone	After delivery- Corotrope, Prostin. Discharged: Karwedilol, Spironol, Captopril

Table 1: The summary of seven presented cases with prenatal data, perinatal management, delivery and postnatal data; case "0" not included - referred via internet but no fetal echo in our center

digoxin level was in range 0,9 – 1,5 mikrogram/ml, their ECG did not show any significant abnormalities) and we do believe that it was a good option to prevent further progress of cardiomegaly and improved myocardial hypertrophy. Digoxin belongs to the drugs which currently are not used in adult cardiology but are still used in prenatal and pediatric cardiology⁷. By longitudinal observation in prenatal echocardiography M-mode as well as Tei indexes would be of great value. We performed 15 fetal echocardiography examinations in our group of fetuses with abnormal heart walls, but M-mode echocardiography was performed only in 6 examinations, Tei indexes were calculated only in 4 examinations, in one case due to fetal bigeminia evaluation of Tei index was not possible. The other limitation of our study was not using Tissue –Doppler. Only in one case we used STICK technique and 4D evaluation.

One may question the need for maternal low molecular weight heparin injection as there is no proof that heparin is transferred via placenta into the fetal circulation. However on the other hand when fetal cardiologist cannot see the blood flow in any chamber despite power angio Doppler using different transducers and in different scans, it is hard to forget that in such areas no blood flow may mean a blood clot. Also in many literature reviews different effects of heparin, for e.g anti-inflammatory, have been investigated. With accompanying maternal problems like for instance thrombophlebitis low grade heparin is recommended, we do believe that in presented cases the transplacental treatment was reasonable.^{8,9,10}

We believe that proper prenatal care, longitudinal echocardiography monitoring, prenatal treatment and planning the best time for delivery gives proper perinatal care in preparation for the postnatal live of our patients¹¹.

CONCLUSIONS:

Fetal heart aneurysm or diverticulum may be detected in 3rd trimester of pregnancy after normal first half of pregnancy and should be referred for targeted fetal echocardiography in a referral center.

Close fetal echocardiography monitoring in 3rd trimester in these cases may be of valuable help for obstetricians in the safe prolongation of the pregnancy, despite cardiac structural abnormality.

Should prenatal transplacental treatment be recommended in these cases, multicenter expertise would be necessary for the best future care of our special patients.

ACKNOWLEDGEMENTS

This way we would like to thank to the obstetricians performing basic fetal heart examination who did refer their case to our fetal cardiology center in Lodz, Poland.

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Division of work:

Respondek-Liberska M: concept of the manuscript, literature search and discussion, english version providing fetal echo exams and perinatal care, literature review ,

Plużańska J: first draft of manuscript, literature search and discussion, english version, submitting manuscript, neonatal care