Case report

RIGHT ATRIAL THROMBOSIS IN THE FETUS WITH DILATATION OF THE RIGHT ATRIUM WITH *IN UTERO* SPONTANEOUS RESOLUTION - A CASE REPORT WITH NEONATAL FOLLOW-UP

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PRENAT CARDIO. 2015 DEC;5(4):26-33
DOI 10.1515/pcard-2015-0004

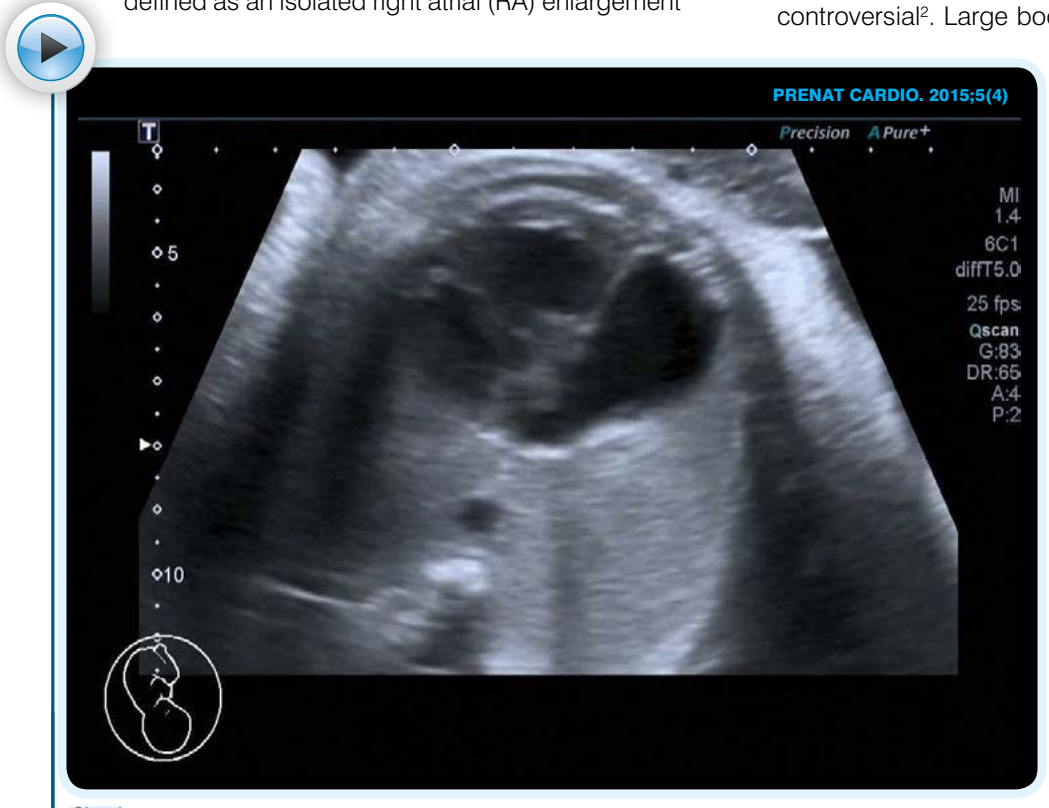
Abstract

We present an extraordinary case of congenital enlargement of the right atrium diagnosed at 31 weeks of gestation. This case emphasizes the fact that timing of the detection of this particular cardiac malformation is of capital importance to tract the optimal treatment strategy in order to monitor further progression (in this case accompanying thrombosis) and prevent complications.

Key words: *right atrial enlargement, prenatal diagnosis, fetal echocardiography, idiopathic, congenital anomaly*

INTRODUCTION

Dilatation of the right atrium (DRA) is a rare anomaly, defined as an isolated right atrial (RA) enlargement of unknown etiology in the absence of other cardiac defects, usually diagnosed by chance[1,2]. Whether this anomaly is congenital or acquired still remains controversial[2]. Large body of literature describes IDRA as an idiopathic dilatation of the RA, congenital enlargement of the RA or congenital RA-diverticulum[1-3]. So far, IDRA has been reported antenatally, in newborns, children and young adults.[1, 4-6] The clinical presentation of DRA ranges widely from the lack of any symptoms to significant atrial tachyarrythmias, cardiac failure or even sudden death[1, 2, 4]. Thus, the appropriate management of this rare anomaly is of capital importance to optimize fast diagnosis and treatment strategies.



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CASE REPORT

A 24 year old gravida 2 para 2 (first pregnancy uncomplicated, medical and family history irrelevant), was referred to our tertiary prenatal

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Submitted: 2015-11-30, accepted: 2015-12-20

cardiology center in Lodz at 33 weeks of gestation because of increasing disproportion at the level of the atria in favour of the right atrium and suspected tumor within. Up until 31 weeks of gestation all test results were described as normal.

How to cite this article:
Płużańska J, Jaguszewska K, Binikowska J, Oleś A, Łukaszewski M, Respondek-Liberska M. Right atrial thrombosis in the fetus with dilatation of the right atrium with in utero spontaneous resolution - a case report with neonatal follow-up. Prenat Cardio. 2015 Dec; 5(4):26-33

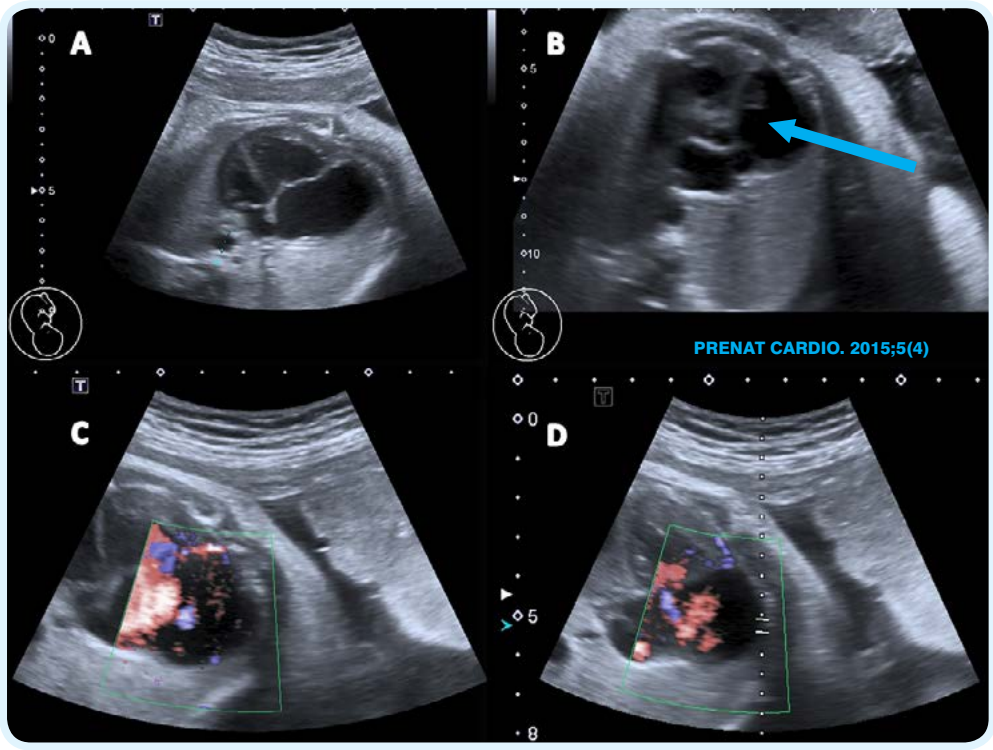


Fig.1.

The ultrasound examination in the Department of Prenatal Cardiology revealed a single male fetus in cephalic longitudinal position with appropriate for gestational age biometry. It showed a grossly enlarged right atrium and moderately enlarged right ventricle with right-to-left flow through the foramen ovale. An enlarged pulmonary artery and hypoplastic aortic arch with scarce flow (secondary to pulmonary dilatation?) and a hokey stick like ductus arteriosus with elevated Vmax up to 160cm/s were also noted. Systemic and pulmonary venous return were described as normal, HA/CA=40, AP=46mm. The patient was referred to the obstetric department at the Polish Mother's Memorial Hospital in order to conduct tests screening for coagulation disorders. Test results showed slightly elevated fibrinogen (5,82 g/l) and d-dimer levels (2109 μg/l), hemoglobin level of 10,9 g/dl and thrombocytes=235 10^9/l, urine sample showed erytrocyturia and leukocyturia. Echocardiographic examinaton after 8 days confirmed a dominating hypokinetic right atrium but without progression of cardiomegaly. The free wall of the atrium was visualised with an opening into a diverticulum of the right atrium, blood flow through the atrium and around a spherical mass was visualized- suspected blot clot. Control echo at 36 weeks showed polyhydramnion and hyperechogenic bowel. It revealed bidirectional flow through the foramen ovale along with turbulence within ductus arteriosus, the

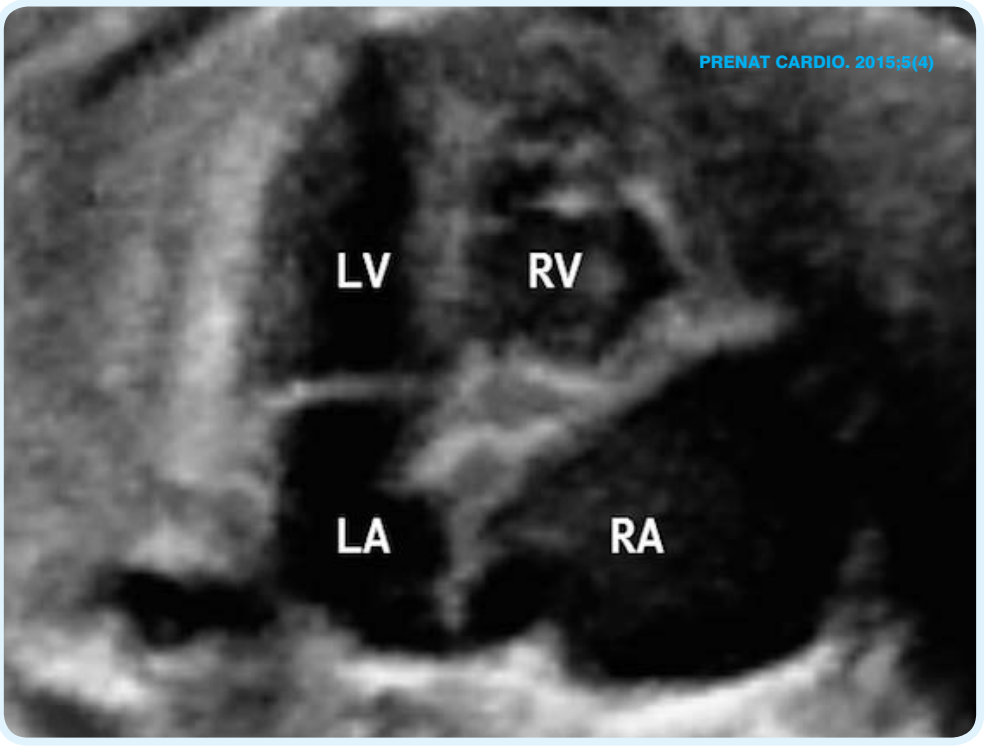


Fig. 2: Prenatal echocardiogram: 4 chamber view

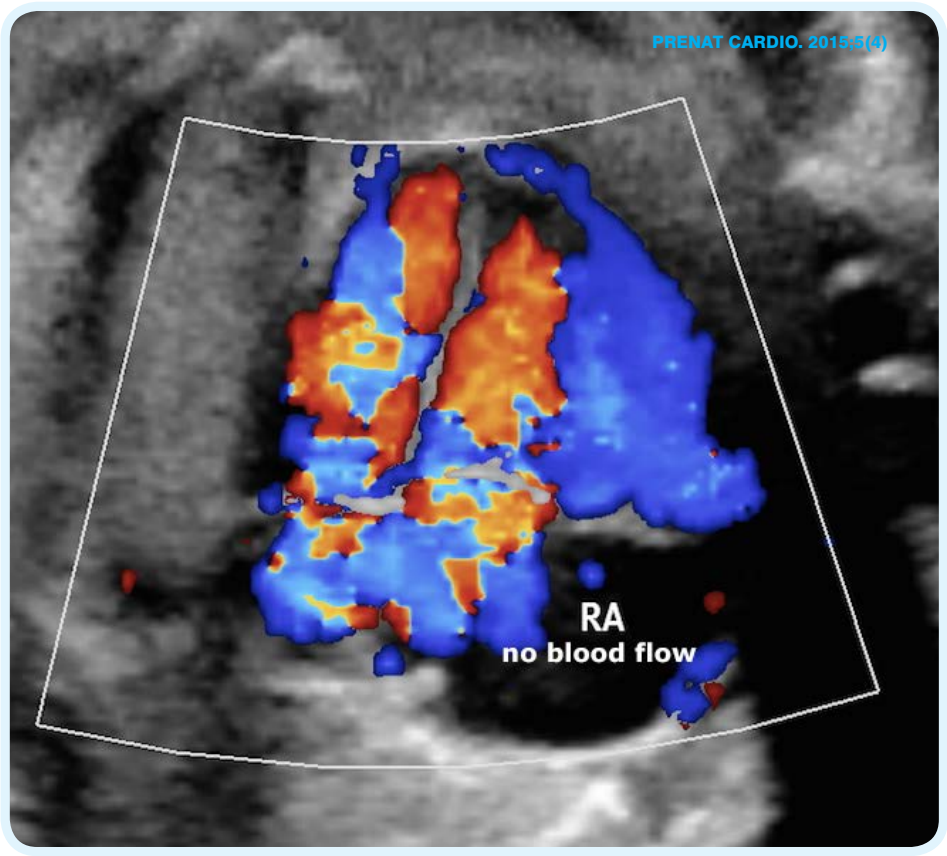


Fig. 3 Prenatal echocardiogram. Diminished flow through right atrium

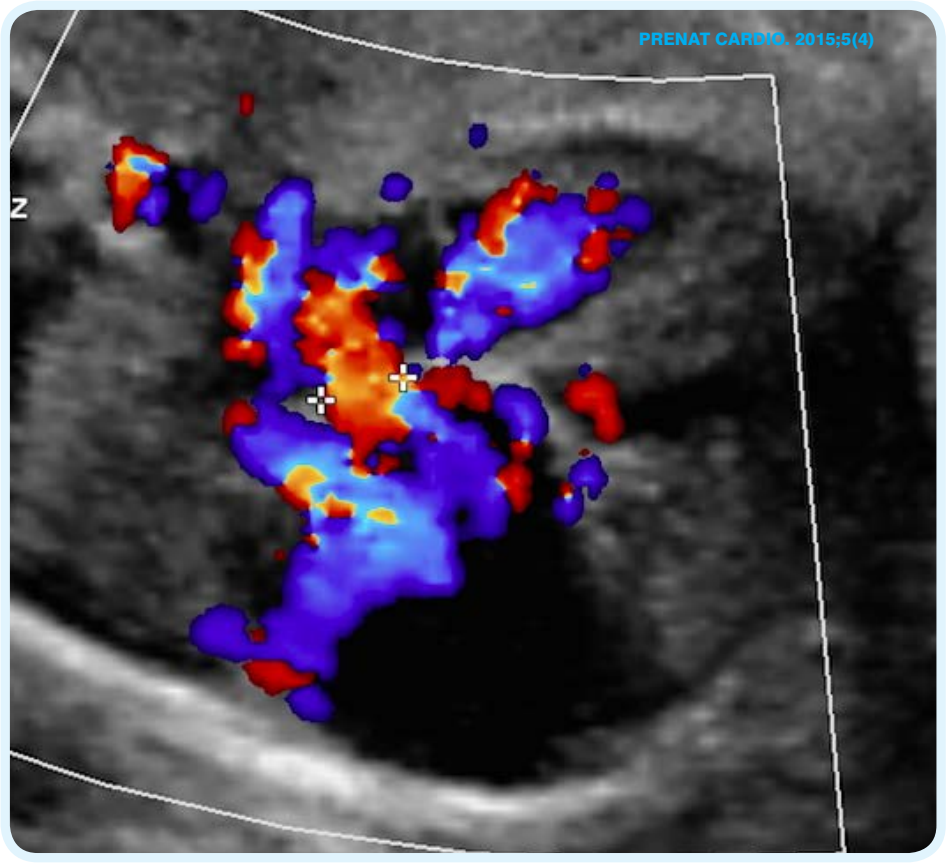


Fig. 4: Prenatal echocardiogram. Foramen ovale + diminished flow across right/left atrium

suspected clot was still visible. Umbilical knot was suspected. The patient was referred once more for monitoring to the obstetrical department.

At 39 weeks of gestation suspected clot was not visible, HA/CA=0,5, AP=55mm, aorta at the level of the 3 vessel view 7mm, pulmonary artery 11mm with periodic bidirectional flow through FO and turbulence within the ductus arteriosus. Seven days later at 40 weeks of gestation, male neonate weighing 4180g was delivered by cesarean section with an Apgar score of 10/10 points. Saturation levels 95-97%, heart rate 150bpm, screening for infection was conducted, and broad-spectrum antibiotics were administered until active inflammatory process was excluded. Cardiac enzymes, d-dimers and platelets were slightly elevated with a tendency to decrease.

The patient on day 6 of postnatal life was referred for further care and treatment to the Department of Cardiology in our hospital. Echocardiography showed a clear disproportion between atria, in the apical view 23x31mm, right ventricle without evidence of hypertrophy, no pathologic masses within the atria were visualized, which was later confirmed by angioCT (RA 33x30x29mm). Portal system doppler examination was also normal. Asprin and spironolactone were administered. The patient in good condition on day 16 of postnatal life with no overt signs of heart insufficiency was discharged home, with recommended control in cardiac clinic in four weeks and continued treatment with Spironolactone and Acesan.

DISCUSSION

Idiopathic dilatation of the right atrium is a rare congenital abnormality of uncertain etiology with strong adulthood

predominance. It was first reported by Bailey in 1955[1]. In the literature, there are only a few reports of IDRA diagnosed prenatally[1,2,7-19]. To our knowledge the first such case was the one presented by da Silva et al from 1992[12] of a fetus with dilated right atrium and massive tricuspid regurgitation, but asymptomatic after delivery and no accompanying TR. However, the earliest detection was made in the 21st week of gestation by Hoffmann et al[2]. The nomenclature is also various, some authors refer to this anomaly as idiopathic dilatation/enlargement of the right atrium others right atrial aneurysm or RA diverticulum. In 1965 Sumner R. et al described two essential criteria for IDRA diagnosis: isolated enlargement of the RA compared to other cardiac chambers and systematic exclusion of other cardiovascular and extracardial malformations that lead to RA dilatation[20]. Although the etiology of IDRA still remains unknown, researchers hypothesize two types of malformations: 1) secondary to degenerative process of unknown etiology affecting the atrial myocardium because of probable apoptotic process or 2) congenital absence of atrial myocardium and conduction tissue[21]. Blondheim D. noticed that the former etiology may have a more benign course, while the latter one may have associated conduction defects and poor long-term prognosis including sudden death. The clinical presentation of IDRA may vary from predominantly asymptomatic cases in the prenatal period with incidental diagnosis during echocardiography to severe symptomatic events of atrial tachycardia, arrhythmia ultimately requiring ablation or surgery, the most advanced cases can result in cardiac failure[1,13-18]. In asymptomatic cases, with only mild to moderate cardiomegaly, conservative approach from watchful waiting to long-term anticoagulation treatment, due to increased thromboembolic risk is required. To our knowledge this is the first case of suspected fetal thrombosis within the right atrium, as opposed to cases of clots in adults and during childhood[4,22]. One case diagnosed in the prenatal period with postnatally suspected thrombus required enoxaparin treatment[1].

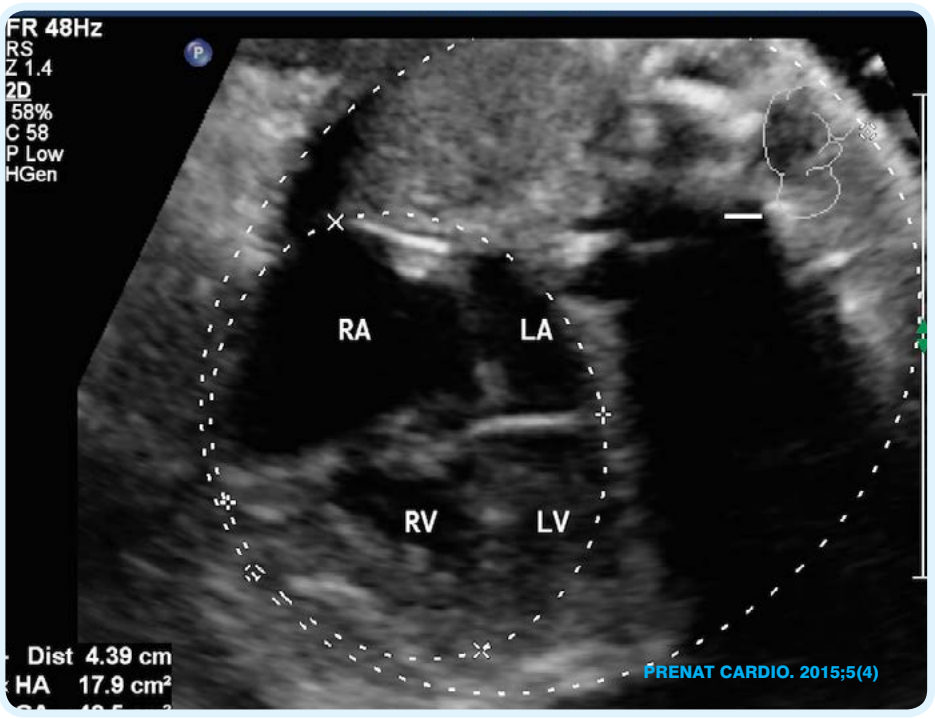


Fig. 5: Prenatal echocardiogram.HA/CA

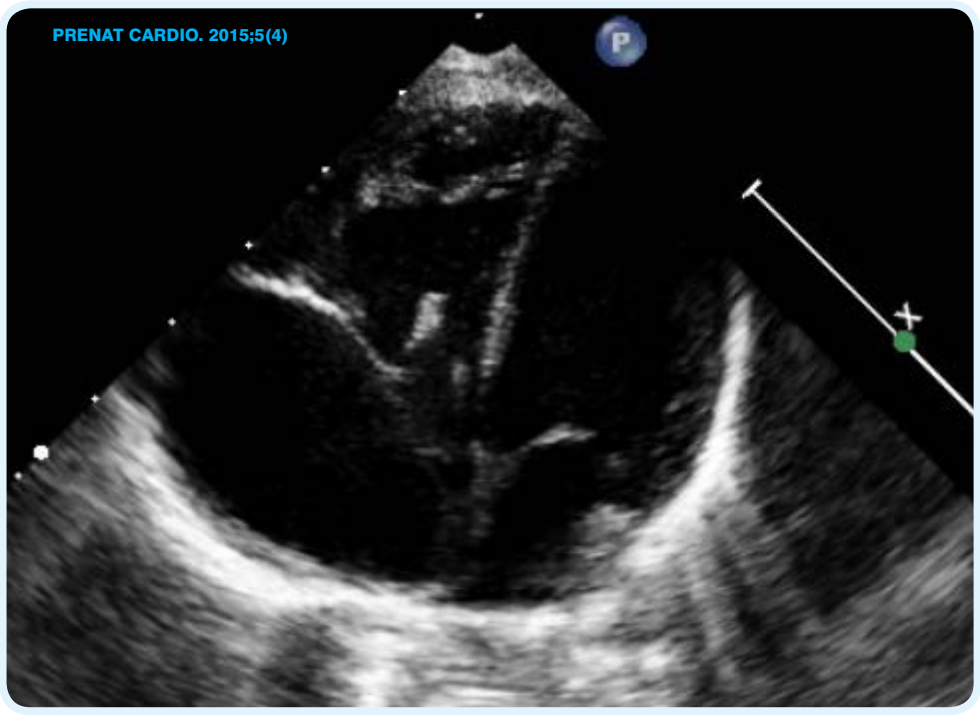


Fig..6: Postnatal echocardiogram in the Department of Pediatric Cardiology

Additional findings during the prenatal period reported in the literature include tricuspid regurgitation, fetal hydrops, intrauterine growth retardation, pericardial effusion, atrial tachycardia and other arrythmias, none of which were reported in our case[1,12,18]. Often the first interpretation of such an image is that of Ebstein's anomaly or pericardial effusion, abnormal pulmonary vein insertion must be excluded. Usually echocardiography is sufficient to make this differentiation but the most challenging cases may require

Name of first author	Year published	Prenatal diagnosis	Additional findings	Vaginal delivery or CS	Follow-up
SILVA[12]	1992	(+)	Massive TR	N/A	Asymptomatic without TR
SHAH[10]	1992	35 HBD RAD		38 hbd vd uneventful delivery	SVT (2hrs of life) Digoxin + flecainide- >cardioversion. WPW diagnosed
RUTLEDGE[11]	1997	N/A IDRA		N/A	Atrial flutter- sotalol treatment
REINHARDT-OWLYA[18]	1998	35 hbd, IDRA	Massive TR	N/A	Surgery at 10months. 4y.o asymptomatic
FERREIRA[17]	2005	22 hbd + . Right atrial aneurysm	Fetal hydrops, IUGR	CS 30hbd 2,250g, Apgar 4/7	Demise 10 hour of life
FORBES[1]	2007	case 1)18 hbd IDRA	Atrial tachykardia 28 hbd (digoxin mother + fetus). Signs of hydrops:32 hbd	32 hbd, cc, 2100g atrial tachykardia, 4weeks ventilatory support, discharge 7 weeks	9 weeks heart failure (pharmacotherapy) 6months: sugery RA reduction + MAZE 7m-3years pharmacotherapy 3years: radiofrequency ablation
	2007	case 2) 35 hbd IDRA	Moderate TR	VD, 3500g, tachypnoe + cyanosis	Suspected thrombus-> enoxaparin 5 weeks respiratory difficulty (airways compressed)->RA reduction surgery atrial tachycardia-> pharmacotherapy
	2007	case 3) 32hbd IDRA	Arrythmia	37 hbd, 3505g	2 weeks: PAC + AT ; 9 years- without progression or arrythmia
PAPAGIANNIS[13]	2008	RA aneurysm	None	40hbd , VD	Surgical treatment
LEE[14]	2010	RA aneurysm	None	40 hbd ; VD	Surgical treatment
WAGNER[8]	2011	22 HBD IDRA	None	Born at term	Asymptomatic at 12months
HOFMANN[2]	2012	20,5 HBD ; IDRA,	Abnormal flow L-> R FO	39 hbd, VD, 3020g Apgar 8/9/9	4 years old, no clinical symptoms
HERNANDEZ[19]	2012	N/A IDRA	None	Normal, at term, uneventful	18 months asymptomatic
BHUPALI[7]	2013	29hbd RA aneurysm	Mild TR	N/A	N/A

WALTER[15]	2013	39 hbd, IDRA	None	N/A	Asymptomatic, sTable atrial enlargement
		28 hbd, IDRA	None	N/A	Asymptomatic, rapid progression, surgical correction at 7 months
		32 hbd, IDRA	None	N/A	Unknown
		20 hbd, IDRA	None	N/A	Unknown
ENZENSBERGER[9]	2013	31 hbd IDRA	SVT 33 hbd. Digoxin+ Flecainide therapy	CC 38 hbd fetal distress apgar 8/9/9	SVT continued Digoxin Propafenon. Surgery 8 days + surgery 24months
NAGATA[16]	2015	23 hbd,	37 hbd pericardial effusion	Atrial flutter + respiratory distress (compression by RA)	18 days surgical treatment
	2015	30 hbd IDRA	None	Uneventful delivery	Day 11 atrial tachycardia ; 6 months no clinical symptoms
PLUZANSKA (this case)	2016	33 hbd.	Suspected thrombus within RA	Uneventful delivery. 40 hbd cc, Apgar 10/10	Prophylactic aspirin

Table 1: Prenatally diagnosed cases of IDRA- literature review

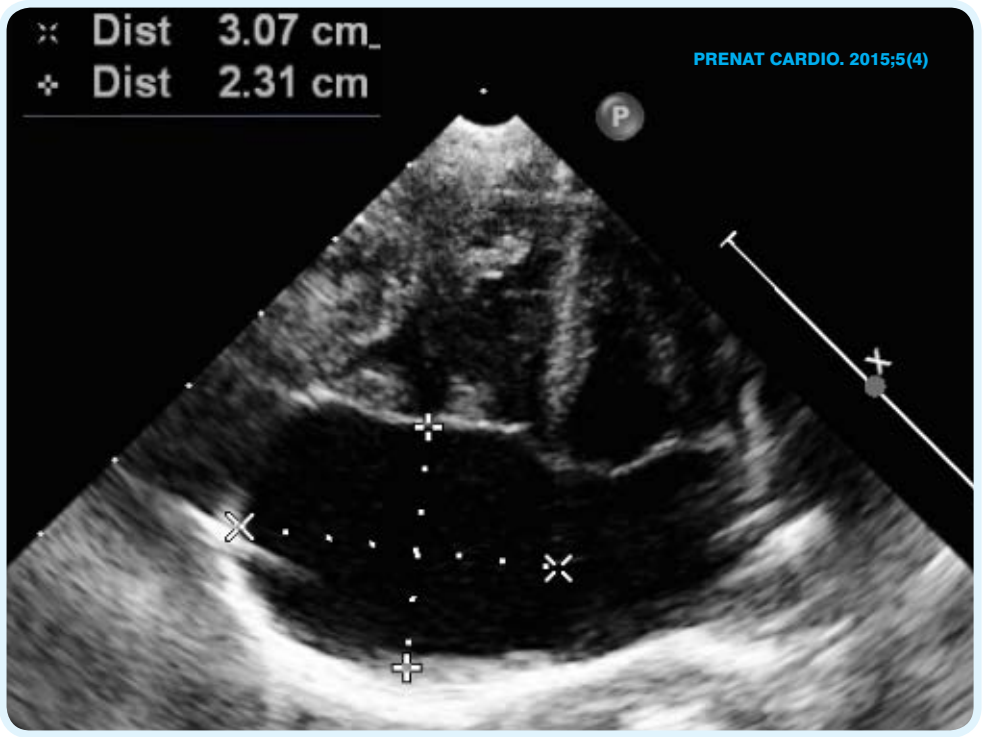


Fig. 7: Postnatal echocardiogram in the Department of Pediatric Cardiology

additional angioCT, MRI or even cardiac catheterization in the neonatal period. A summary of all prenatal cases reported in the literature to the best of our knowledge we present in Table 1

All IDRA cases need long-term follow-up with repeated echo exams and ECG and 24hour Holter-ECG monitoring to observe possible progression and occurrence of arrhythmias.

Our patient is under regular observation, for now conservative treatment and thrombolytic prophylaxis has been administered and fortunately no arrhythmias have been recorded. At four months of age the child is developing properly and no progression of dilatation has

been observed so far.

CONCLUSION:

1) Isolated dilatation of the right atrium is a rare prenatal finding and can be associated with clot formation in the fetus

2) All IDRA cases need long-term follow-up with repeated echo exams, ECG and 24hour Holter-ECG monitoring to observe possible progression and occurrence of arrhythmias.

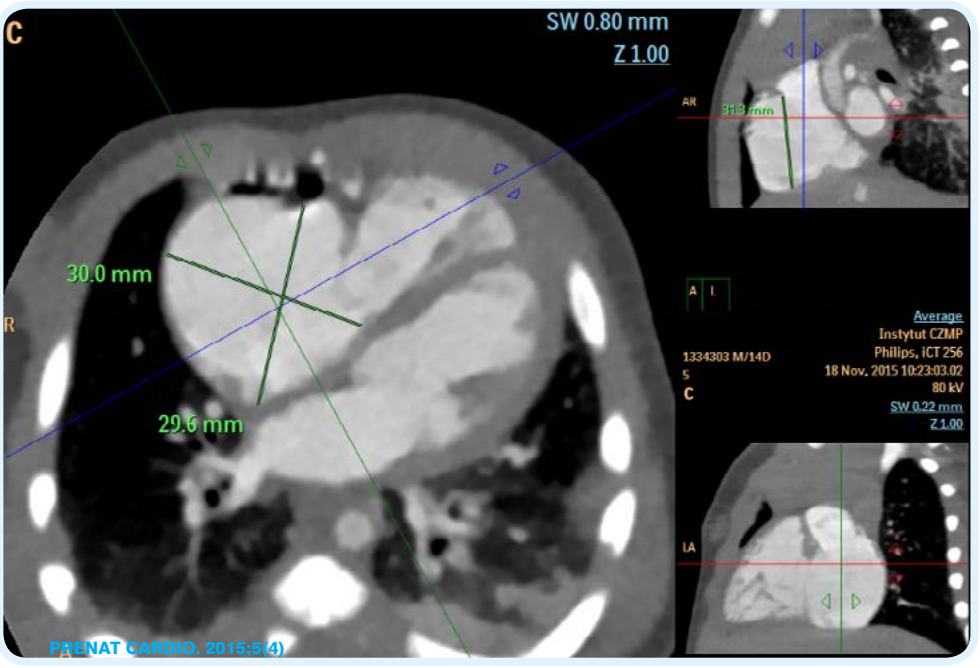


Fig..8: AngioCT measurments of right atrium in neonate

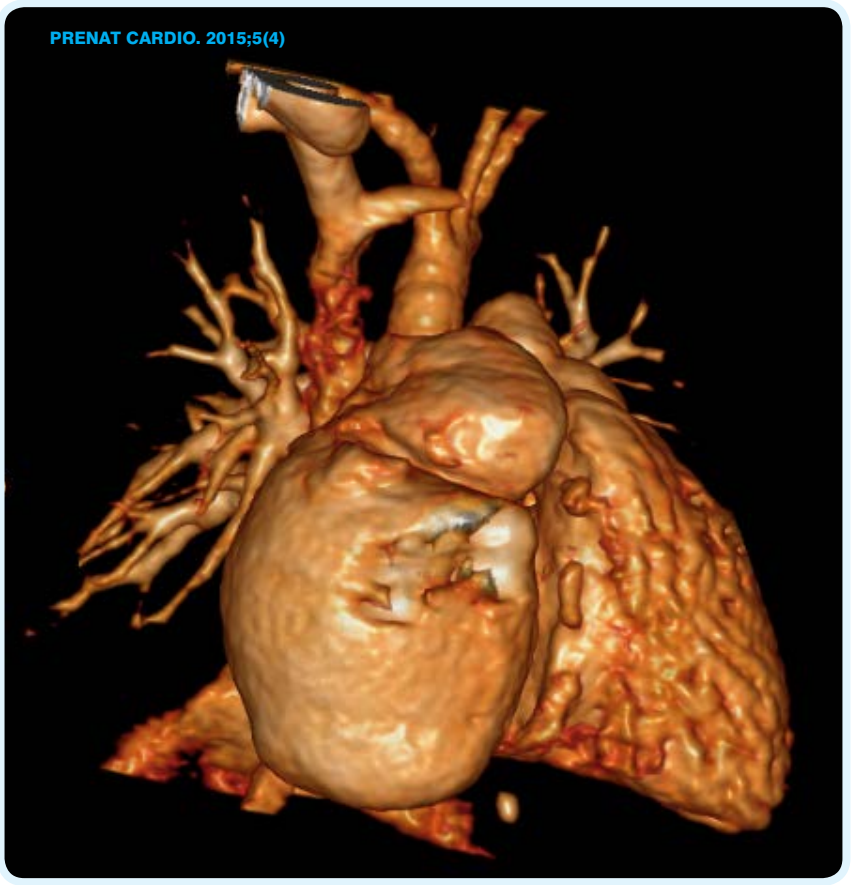


Fig..8: AngioCT 3D reconstruction, visible dillated right atrium

References:

1. *Forbes K, Kantoch MJ, Divekar A, Ross D and Rebeyka IM. Management of infants with idiopathic dilatation of the right atrium and atrial tachycardia. Pediatric Cardiology. 2007;28:289-96.*

2. *Hofmann SR, Heilmann A, Hausler HJ, Dahnert I, Kamin G and Lachmann R. Congenital idiopathic dilatation of the right atrium: antenatal appearance, postnatal management, long-term follow-up and possible pathomechanism. Fetal Diagnosis and Therapy. 2012;32:256-61.*

3. *Imren Y, Halit V, Kula S and Olgunturk R. Giant right atrial aneurysm: case report. International Journal of Cardiology. 2006;112:e66-8.*

4. *Binder TM, Rosenhek R, Frank H, Gwechenberger M, Maurer G and Baumgartner H. Congenital malformations of the right atrium and the coronary sinus: an analysis based on 103 cases reported in the literature and two additional cases. Chest. 2000;117:1740-8.*

5. *Pernot C, Hoeffel JC and Jacob F. Idiopathic dilatation of the right atrium revealed in childhood by dysrhythmias. Pediatric Radiology. 1977;6:52-4.*

6. *Marin-Garcia J and Allen RG. Idiopathic dilatation of the right atrium: postoperative follow-up in a child. Journal of Pediatric Surgery. 1983;18:196-8.*

7. *Bhupali AN, Patankar KB, Paranjpe FS and Tamhane AU. Giant right atrium in a foetus. Indian Heart J. 2013;65:493-5.*

8. *Wagner N, Kagan KO, Abele H, Hoopmann M, Kaulitz R and Hofbeck M. Fetal idiopathic dilatation of the right atrium. Ultraschall Med. 2011;32:81-2.*

9. *Enzensberger C, Kreymborg KG, Valeske K, Apitz C, Akinturk H, Schranz D and Axt-Fliedner R. Management of idiopathic giant dilatation of the right atrium with subsequent atrial tachycardia. Arch Gynecol Obstet. 2013;288:705-7.*

10. *Shah K and Walsh K. Giant right atrial diverticulum: an unusual cause of Wolff-Parkinson-White syndrome. Br Heart J. 1992;68:58-9.*

11. *Rutledge J, Robertson MA, Kantoch M and Dyck J. Idiopathic dilation of the right atrium: case report and survey of the literature. Can J Cardiol. 1997;13:855-7.*

12. *Silva AM, Witsemburg M, Elzenza N, Stewart P. Idiopathic dilatation of the right atrium diagnosed in utero. Rev Port Cardiol 1992; 11: 161-3.*

13. *Papagiannis J, Chatzis a, Sarris G. Giant right atrial aneurysm: a case report. Int J Cardiol. 2008;129:e5-e6. doi:10.1016/j.ijcard.2007.06.081.*

14. *Lee Y, Cho J, Kim G, Lee S, Hyun M, Kim Y. Surgical Repair of Giant Right Atrial Aneurysm in a Neonate. Korean Circ J. 2011;41:331. doi:10.4070/kcj.2011.41.6.331.*

15. *Walter C, Carretero JM, Bartrons J, et al. Idiopathic dilatation of the right atrium. Report of four fetal cases. Hospital Sant Joan de Déu. Barcelona. Spain. Abstract AEPC*

16. *Nagata H, Stambach D, Wilson GJ, Menon A, Jaeggi E. Fetal Diagnosis of Right Atrial Aneurysms. Can J Cardiol. 2015. doi:10.1016/j.cjca.2015.11.002.*

17. *Ferreira M, Barberato A, Barberato SH, Gomes CC, Costa SL, Krawiec A. Right Atrial Aneurysm Associated to Fetal Hydrops: Diagnosis through Fetal*

Echocardiography. Arquivos Brasileiros de Cardiologia 2005;85:2004-2005.

18. Reinhardt-Owlya L, Sekarski N, Hurni M, et al. [Idiopathic dilatation of the right atrium simulating Ebstein's anomaly: apropos of a case diagnosed in utero]. Arch Mal Coeur Vaiss 1998;91:645–9.

19. Tejero-Hernández MÁ, Espejo-Pérez S, Suárez-de-Lezo-Cruz-Conde J. Congenital Aneurysm of the Right Atrial Appendage in a Newborn: a Rare Anomaly. Rev Española Cardiol (English Ed. 2012;65(1):99-100. doi:10.1016/j.rec.2011.03.028

20. Sumner RG, Phillips JH, Jacoby WJ, Jr. and Tucker DH. Idiopathic enlargement of the right atrium. Circulation. 1965;32:985-91.

21. Blondheim DS, Klein R, Plich M and Marmor AT. Familial idiopathic dilatation of the right atrium with complete atrio-ventricular block: a new syndrome? Cardiology. 2000;94:224-6.

22. Blaysat G, Villain E, Marcon F, Rey C, Lipka J, Lefevre M and Bourlon F. [Prognosis and outcome of idiopathic dilatation of the right atrium in children. A cooperative study of 15 cases]. Arch Mal Coeur Vaiss. 1997;90:645-8.

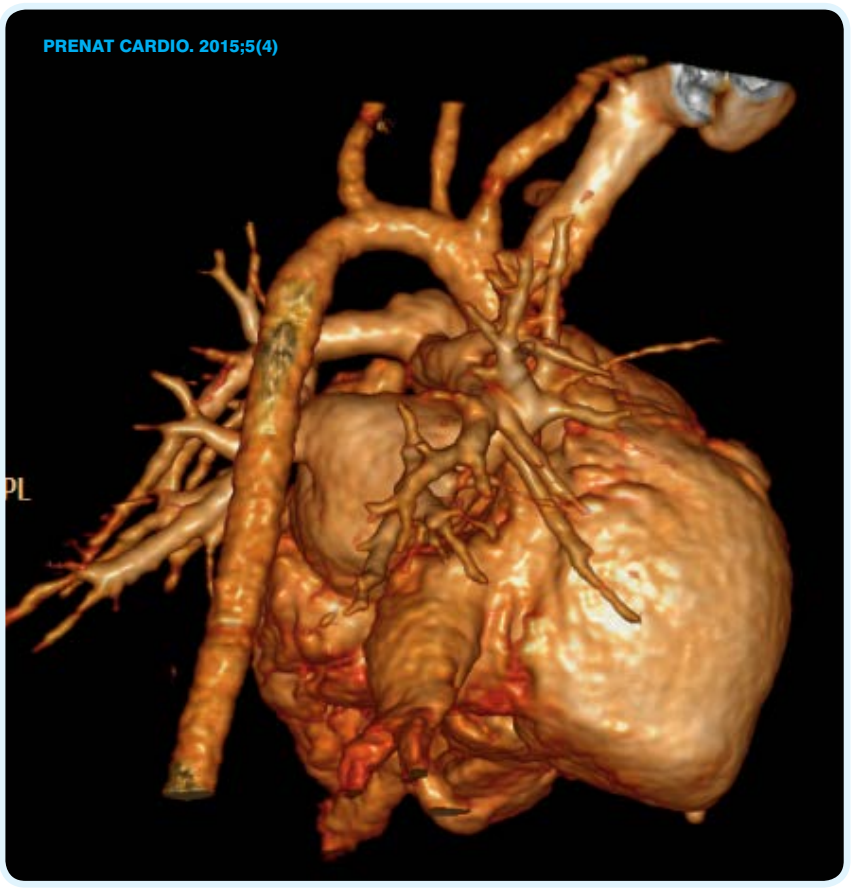
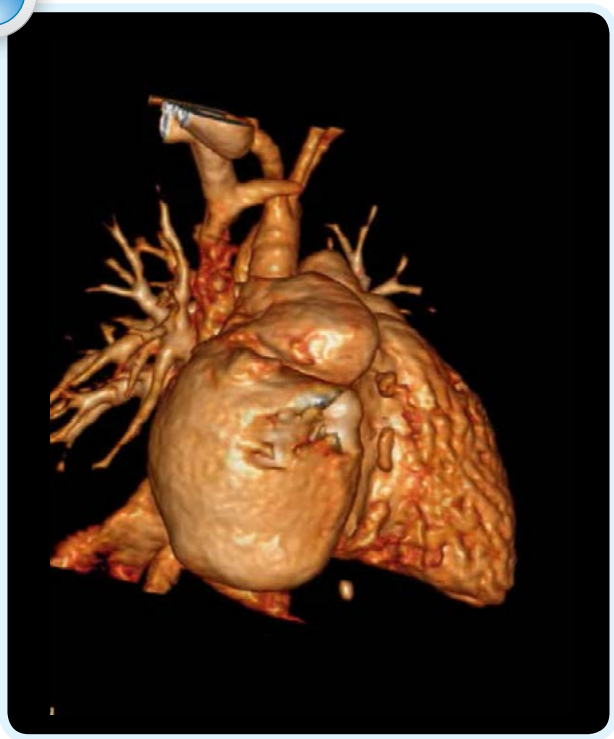


Fig..10: AngioCT 3D reconstruction



Cine 2. 3D heart reconstruction
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Płużańska J: first draft of case report, literature search and discussion, english version, submitting manuscript

Jaguszewska K: literature search & discussion

Binikowska J: postnatal echo, neonatal treatment, work with manuscript

Oleś Adam: angioCT, work with manuscript

Łukaszewski Maciej: angioCT, work with manuscript

Respondek-Liberska M: providing fetal echo exams and perinatal care, concept of the literature review presentation,