Case report

ISOLATED DOUBLE AORTIC ARCH (DAA) – PRENATAL DETECTION WITH POSTNATAL FOLLOW-UP, CASE REPORT AND LITERATURE REVIEW

Authors:
Julia Murlewska[1], Agnieszka Żalińska[1], Danuta Roik[2], Bożena Werner[3], Maria Respondek-Liberska[1]

1. Prenatal Cardiology Department Polish Mother's Memorial Hospital 2. Department of Pediatric Radiology, Medical University of Warsaw 3. Department of Pediatric Cardiology and General Pediatrics, Medical University of Warsaw

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Abstract

This case report presents a prenatal diagnosis with postnatal confirmation (by angio CT and computer reconstruction) of an isolated double aortic arch, with no blood disturbances and with no clinical symptoms after birth. Literature review was focusing on the possible symptoms in the future. Prenatal findings should be forwarded to neonatologist and pediatrician despite clinical silence.

Key words: *prenatal echocardiography, aortic arch, asymptomatic newborn*

INTRODUCTION

Double aortic arch (DAA) maybe detected prenatally by an experience sonographers and fetal cardiologists and maybe clinically silent during pregnancy and after birth, however neonatal medical history should not missed the prenatal finding.

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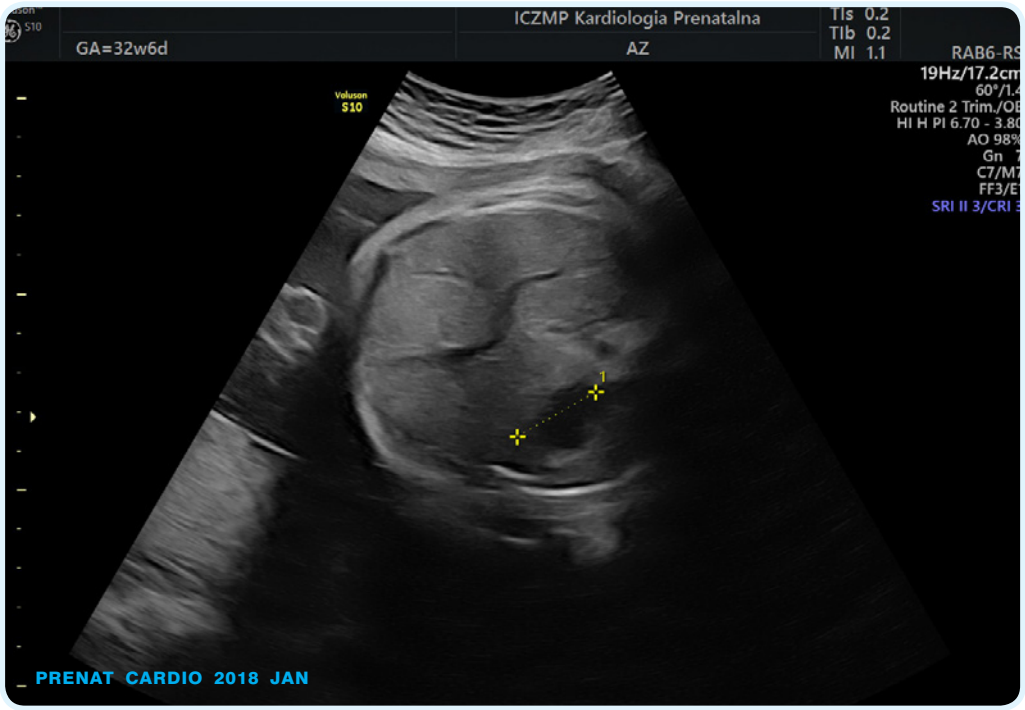
CASE REPORT

A 39-year old multigravida with a singleton pregnancy, had "normal" ultrasound scan in 1st trimester and at 21st week a routine obstetric ultrasound examination including basic fetal heart evaluation revealed right-sided aortic arch and a left arterial duct. The fetus was referred for targeted echocardiography at the fetal cardiology center in Lodz at 24 weeks. The family and obstetrical medical history were unimportant. There was normal extracardiac anatomy, normal four chamber view of the fetal heart size, with apex on the left, and the stomach on the left, and in the normal mediastinum (Fig. 1- 5) . However in upper mediastinum there was aortic bifurcation indicating double aortic arch with a dominant right arch and hipoplastic lfet arch, and antegrade flow in both aortic arches (Fig. 6-8). Cardiovascular



Fot.1. Fetus position cephalic and spine on the left: stomach on the left side - situs solitus

Corresponding author: juliamurlewska.jm@gmail.com

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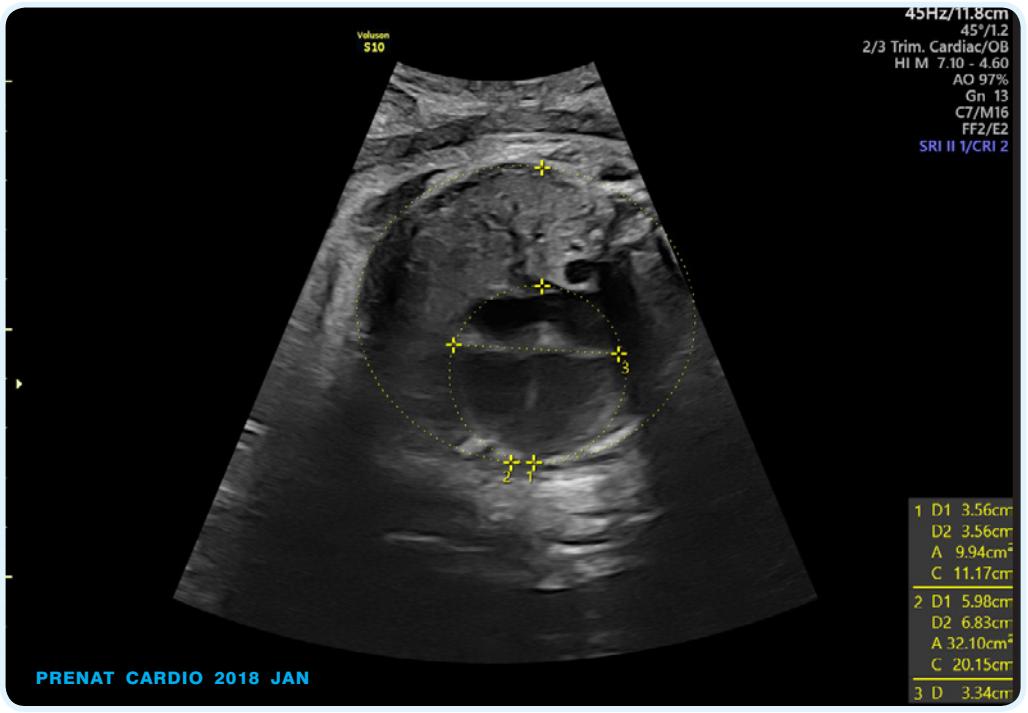
profile score was 10. The pregnancy follow-up was without any complications.

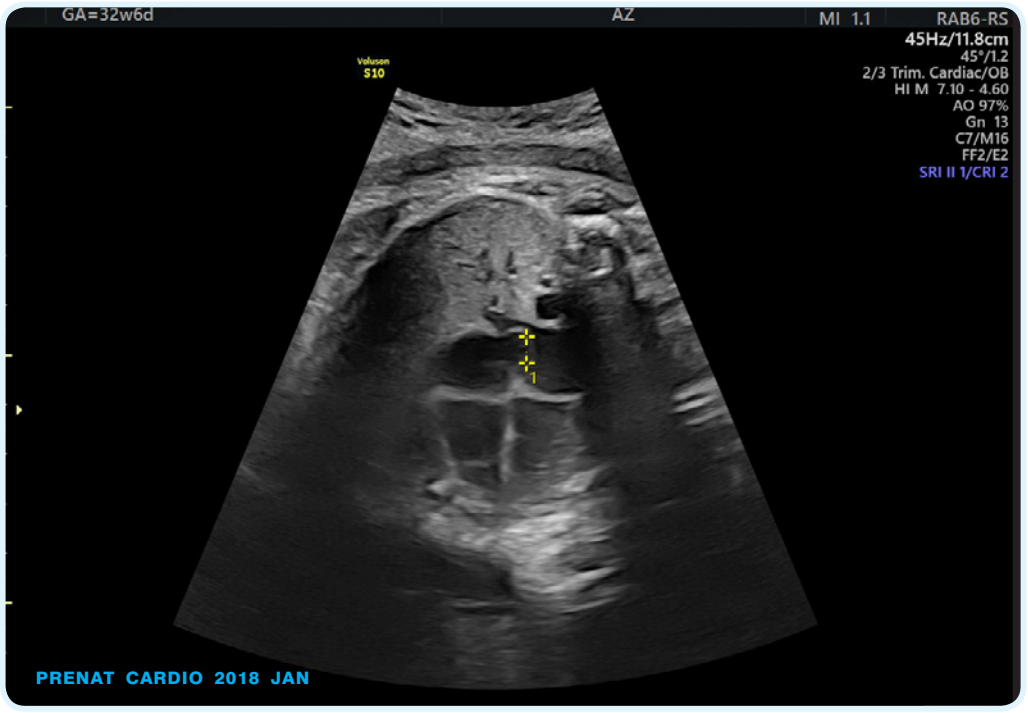
The baby-girl was delivered by elected cesarean section in our referral center due to 2 previous cs. Her birth weight was 3140g, Apgar scores of 10 at both 1st and 5th minutes. She was completely asymptomatic. Her screening neonatal echocardiography was assessed as "normal" and she was discharged home at 4th day of the hospital stay. Hospital neonatal medical history missed prenatally detected anomaly.

Due to maternal will to have the second opinion she was admitted to the Pediatric Cardiology Clinic in Warsaw.

Infant's echocardiogram and reconstruction of computerized tomography arteriography (angio-CT) confirmed the prenatal diagnosis; showed both right and left aortic arches around the trachea with the dominant and bigger right side; right arch measured 7mm, from which arised right subclavian and common carotid arteries (Figures 9-11). Left subclavian and posteriorly leading artery (Kommerel's diverticulum) arised from the left arch, which was 3,2mm in the narrowest segment between them and suggested the segmental atresia/ interruption of LAA- left aortic arch). The narrowest segment of the trachea was measuring 3mm at the level of the double aortic arch and post-stenotic segment was 5,5mm, angio-CT revealed also the asymmetrical pulmonary arteries; right measured 4,6mm and left 3,9mm. The infant did not present any clinical symptoms, with no stridor and dyspnea and was discharged on the 14th day with the planning follow up as an out-patient.



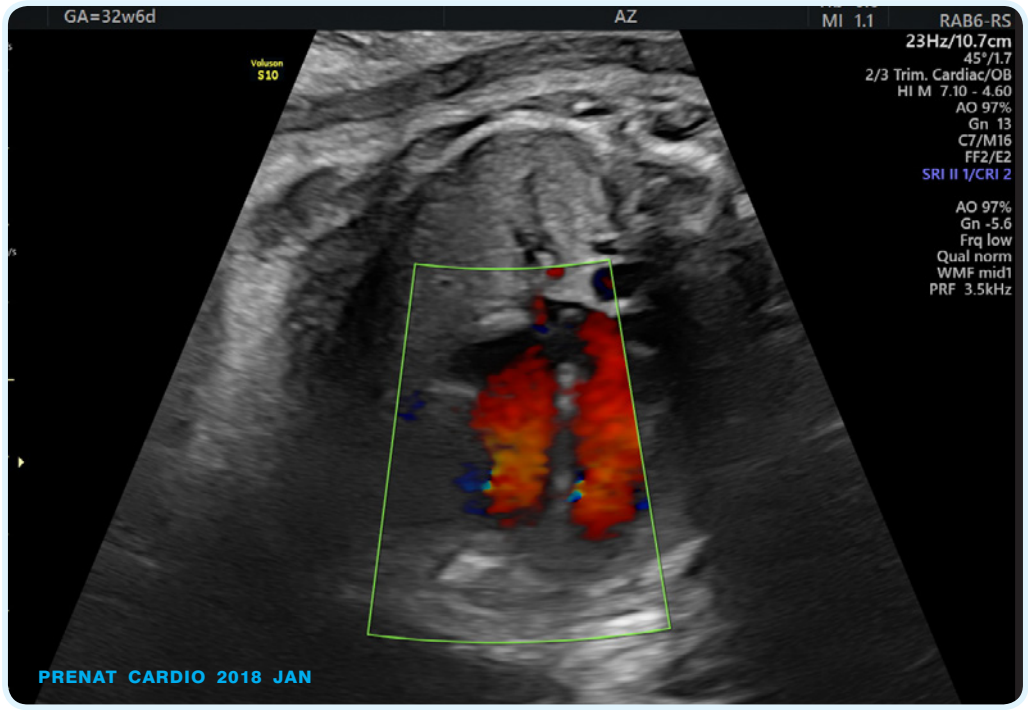
Fot.2. Normal heart area / chest area ratio = 0,3



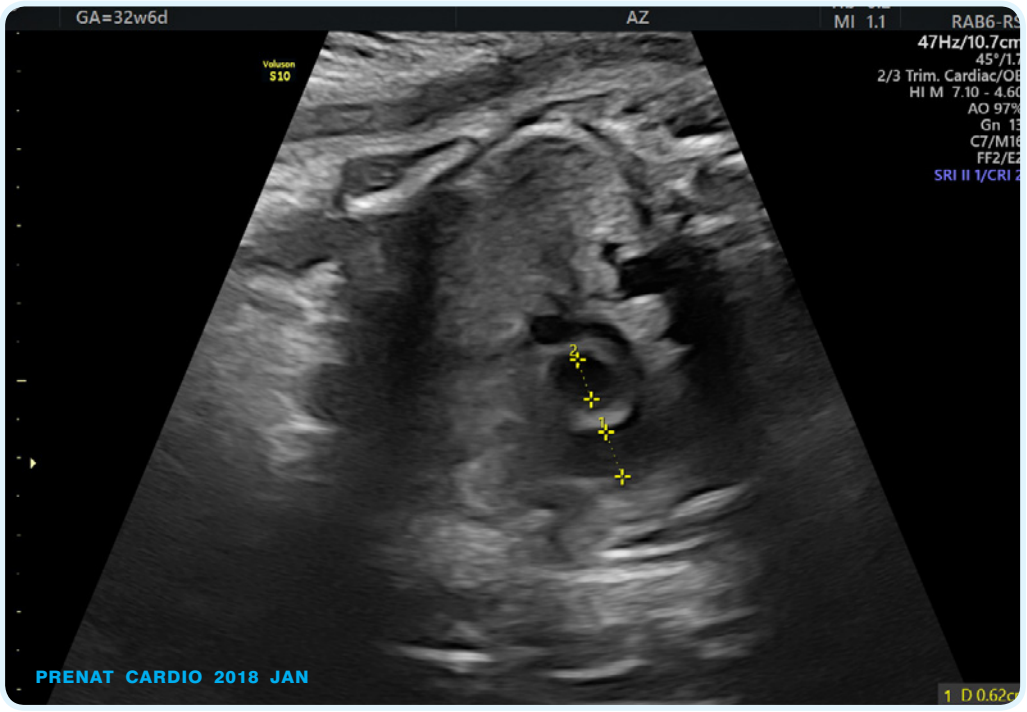
Fot.3. Normal 4 chamber view with wide opened foramen ovale

DISCUSSION

The prenatal detection of double aortic arch was reported several times. Hunter L., et all. [1] presented the earliest prenatal detection at 21 weeks, but generally the average time for fetal echocardiographic diagnosis was 24,5 weeks [1,2].



Fot.4. Normal intracardiac blood flows in color Doppler



Fot.5. Mediastinum scan at the level of three vessels (aorta and pulmonary artery of similar size

Fetal mediastinum may be visualized in 13-14th week of gestation , and right aortic arch maybe detected by ultrasound that early [3]. However the majority of cases with DAA have a dominant right arch and the minor left one is quite difficult to identify in the first trimester of pregnancy. So presumably, DAA could not be yet detectable on that time, but shortly afterwards, in the later gestational weeks, when the anterograde flow is commonly demonstrated in both aortic arches by color Doppler imaging. It is much more difficult to detect prenatally than other aortic arch anomaly like interrupted aortic arch[4].

The pregnancy follow-up usually is with no complications, apart from one case, presented by Gou Z. et all.,[2], in which the fetus died at 27 weeks because of severe intrauterine infection.

Symptomatic prenatal DAA may mimick CHAOS- congenital high-airway obstruction syndrome. Fetal obstruction that blocks trachea could be recognized as lung enlargement/ hiperechogenicity, flattened/ everted diaphragms, dilated distal airways, mediastinal compression, and polihydramnios, ascites and hydrops [5,6,7]. In utero or intrapartum treatment-EXIT procedure were not ascertained[4]. DAA is a planned congenital heart disease, and it never, if isolated required urgent intervention[8,9].

The postnatal confirmation of the prenatal diagnosis is made by echocardiography, angio-CT and MRI (magnetic resonance). Bronchoscopy is recommended to reveal compression of the trachea.

Double aortic arch may form complete or partial, vascular ring which could cause tracheal and esophageal compression.

Most of the patients are asymptomatic through the whole life, but in selected cases surgery treatment is proposed for correction of vascular ring, with favourable long-term prognosis[1,10,11]. Surgery repair successfully eliminate symptoms in 70% cases[2]. Usually, approaches via a left thoracotomy or thoracoscopic surgery is approached, however sternotomy is preferred in patients with associated tracheal stenosis, as surgery requires implementation

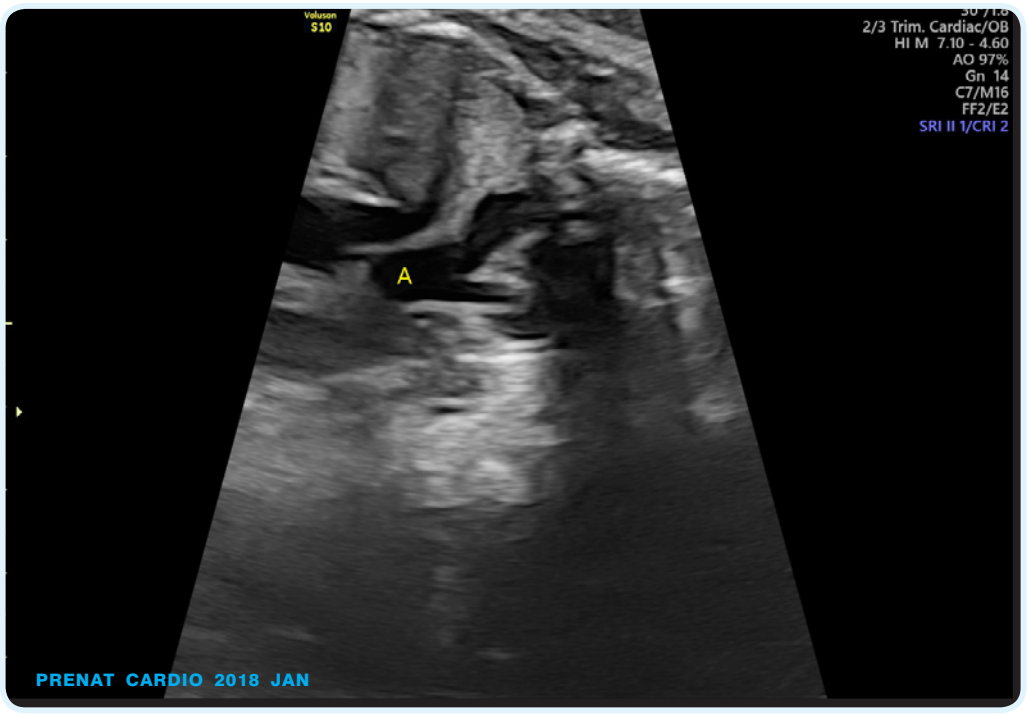
of cardiopulmonary bypass[10,12]. In repair of DAA, the division is made in the hypoplastic arch segment, which is usually the left one[10].

Majority reports are from adults and airway management and weaning from the ventilator could be challenges for anesthesiologists in the perioperative period, as the condition of these patients was often serious[11,13]. But Hunter L. et all.[1] reported term newborn, who developed stridor at 2 weeks of life and had successfull surgery so early[1].

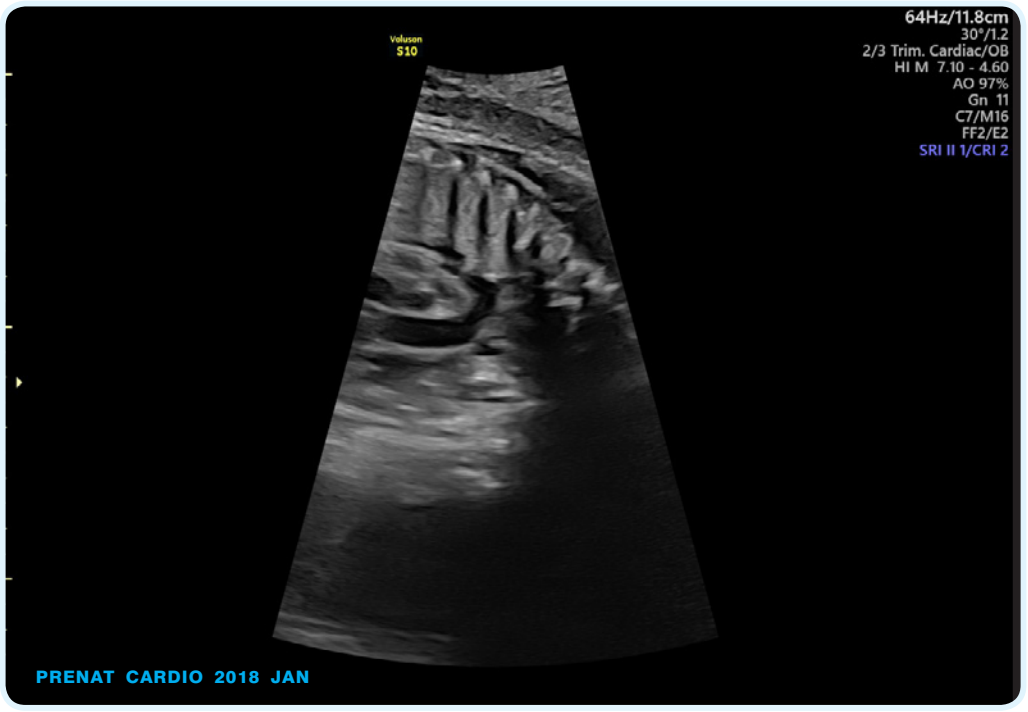
As prenatal and early postnatal diagnosis of DAA could be missed, in cases of symptoms like feeding intolerance, dyspnea or stridor since birth, "asthma", bronchiolitis, repeated pneumonia, these patients could be unsuccessfully treated with inhaled bronchodilators, oral corticosteroids or chest physiotherapy for several months [10-12,14-16].

The effect of undiagnosed early enough double aortic arch may take a long time for different workups: testing for various immunodeficiences, laryngoscopies, naso-laryngo-fiberoscopies, chest radiographies, gastric emptying studies, which nonetheless will demand echocardiography, CT- multidetector computed tomography thorax and MRI for the final, but delayed confirmation of the congenital anomaly[10,12,14].

Before the era of prenatal detection, the diagnosis was established as late as in teenager patient (14-year old), or 44 year-old patient, or even at 79 and 82 years old [16-20].

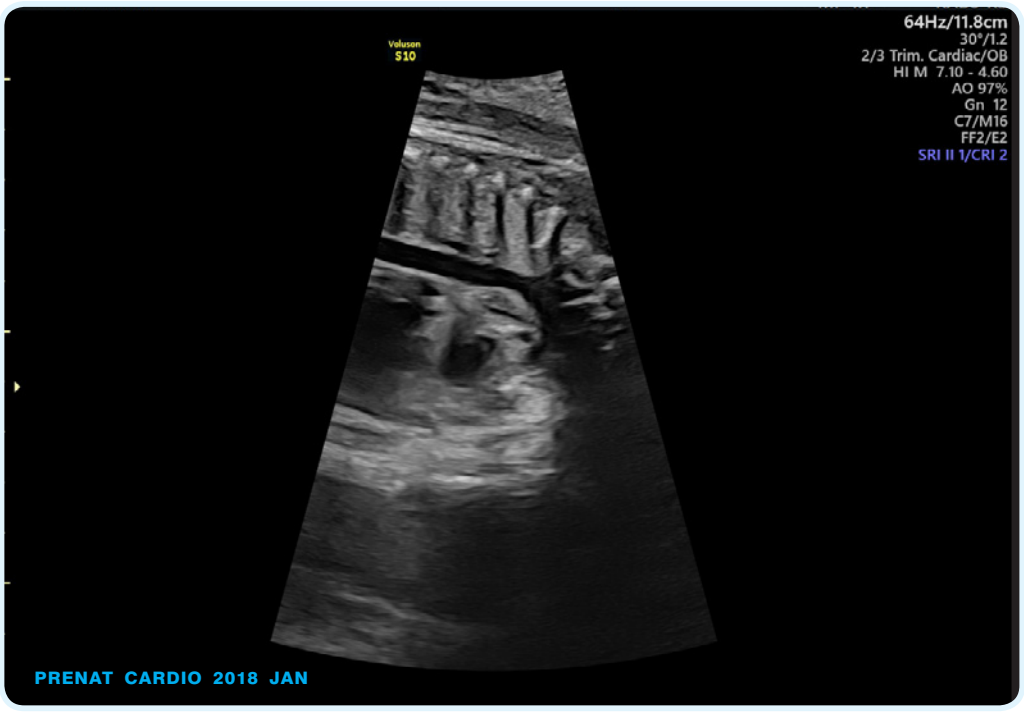


Fot.6. Long axis view mixed with short exis view – Aortic bifurcation

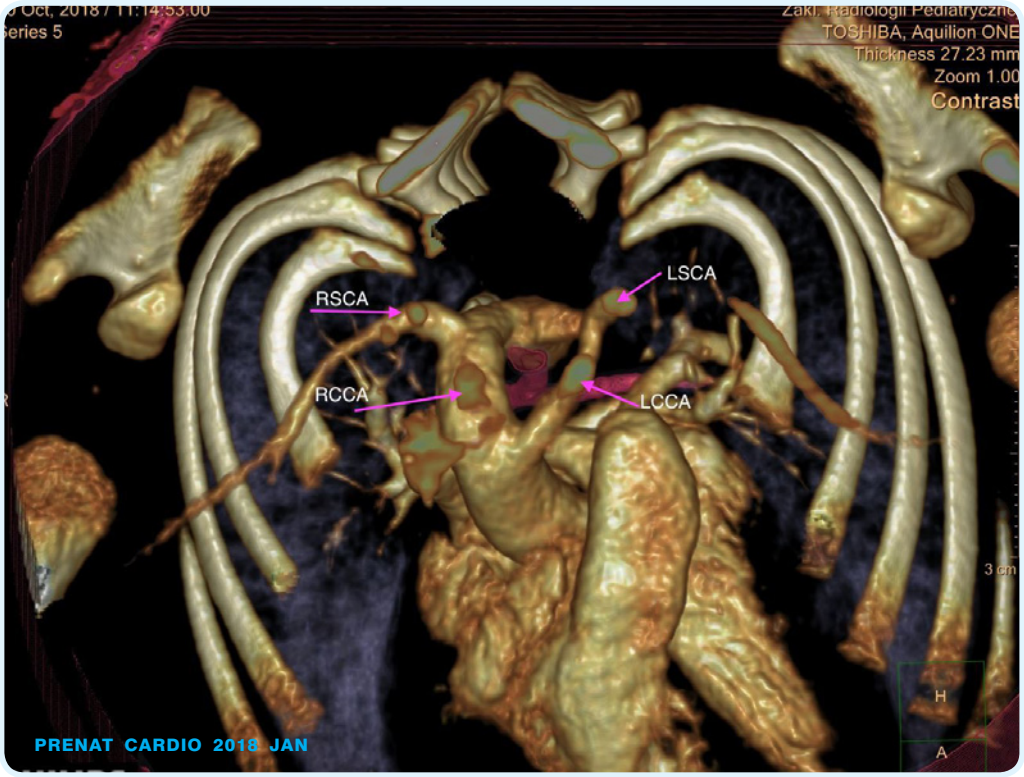


Fot.7. Long axis view - "Normal " Right Aortic Arch

Adult patients were dealing for many years with the heartburn, chest pain, "asthma", chronic pulmonary disease, pulmonary fibrosis, recurrent episodes of severe mucus obstruction, inspiratory stridor, wheezing, recurrent respiratory infections (due to trachea compression) or dysphagia, reflux, choking episodes, swallowing disorders, vomiting or food intolerance (due to esophagus compression)[17-20].



Fot.8. Lond axis scan at 32nd week of gestation - hypoplastic left aortic arch



Fot.9. Computerized tomography arteriography (angio-CT) of the chest showing right-sided dominant aortic arch and left aortic arch encircling the trachea and esophagus with their branches: RSCA- right subclavian artery, LSCA- left subclavian artery, RCCA- right common carrotid artery, LCCA- left common carotid artery

Respiratory symptoms were the most common; in 91% of DAA, gastrointestinal symptoms in 40% of cases [2,13]. Respiratory arrest/ apparent life-threating event (ALTE) has been reported in 7% cases with DAA[10].

DAA with right dominance is seen in 70% cases, left dominance in 20-25% and the most rarely recognized are balanced types. DAA results from failure of regression of usually, the right of the dual aortic arches. The other vascular disorders include right arch/ left ligament, innominate artery compression, and pulmonary artery sling[15,17,21].

The first anatomical case of DAA was described by Hommel in 1737 and Gross was the first one, who performed surgical correction of DAA in 1945 [22].

DAA is usually an isolated cardiac anomaly, but its association with other cardiovascular anomalies is approximately in 20% of cases including: a ventricular septal defect-VSD, atrial septal defect-ASD, PDA, tetralogy of Fallot (ToF), transposition of the great arteries (TGA), pulmonary atresia-PA and common arterial trunk-CAT [1,23-25].

Current recommendations suggest that microarray testing should be performed in patients with arch anomalies. Association with 22q.11 deletion in aortic arch abnormalities has been reported[10,24,25].

We present this case due to main two reasons :

- an isolated double aortic arch (DAA) is rare anomaly difficult to detect and diagnose prenatally
- despite prenatal targeted fetal echocardiography, this diagnosis in asymptomatic newborn was not included in the hospital medical record, suggesting the need to improve the gap between fetal cardiac diagnostic center and neonatology.

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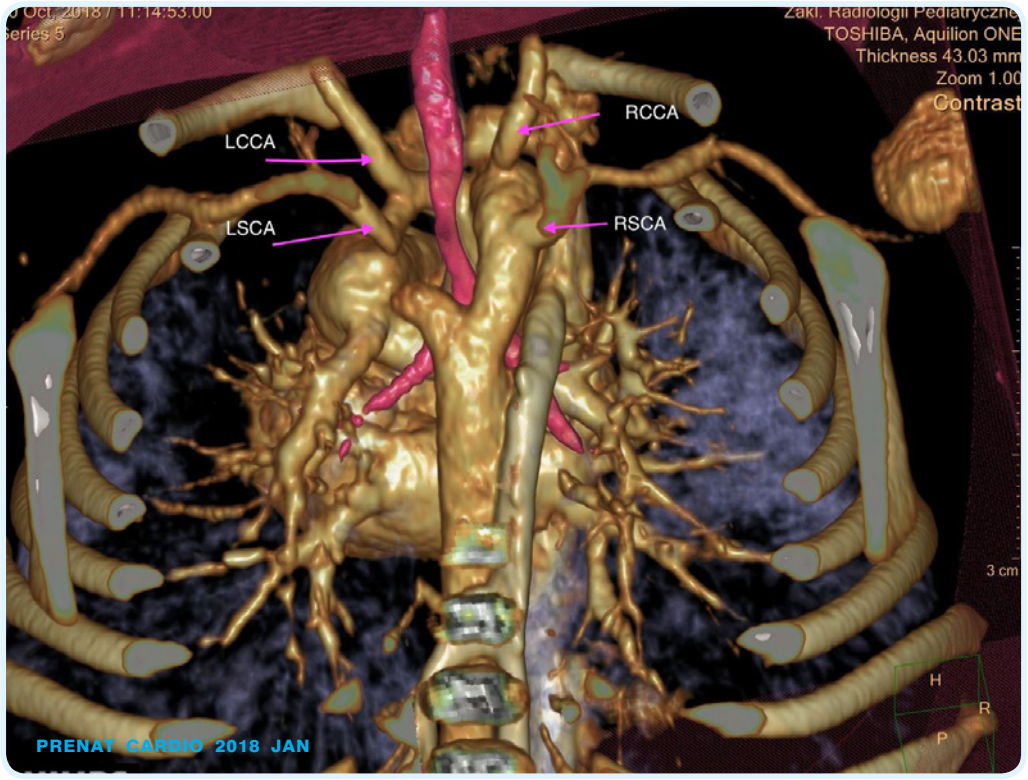
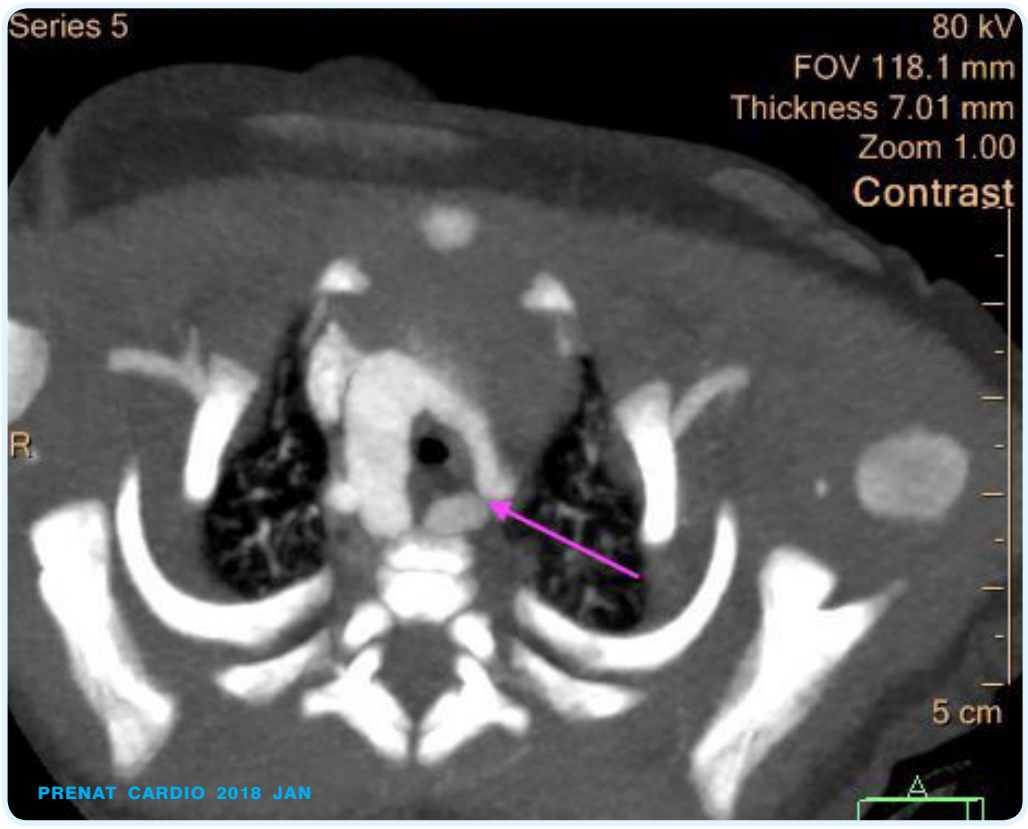


Fot.10. Computerized tomography arteriography (angio-CT) of the chest showing right-sided dominant aortic arch and left aortic arch with the left subclavian and the wide posteriorly leading artery (Kommerel's diverticulum).



Fot.11. The thoracic CT showed the presence of a double aortic arch that completely surrounded the trachea with the indicated segmental atresia between the left subclavian artery and the posteriorly leading artery (Kommerel's diverticulum).

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Division of work:

Julia Murlewska - first draft, literature search

Agnieszka Żalińska - work with the manuscript

Danuta Roik - postantal angio-CT reconstruction, work with the manucript

Bożena Werner - work with manucsript, final version

Maria Respondek-Liberska - work with the manuscript, final version

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