● **Original paper - Pictorial Essay**

GREAT VESSELS ANOMALIES – PRENATAL ECHOCARDIOGRAPHY AND NEONATAL ANGIO-CT – A PICTORIAL ESSAY

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Abstract

Introduction: Fetal echocardiography is a method of choice for diagnosing cardiovascular anomalies prenatally. However, in the majority of cases, the complexity of a defect creates a diagnostic challenge. Moreover, postnatal validation of sonographic findings rarely can be obtained. Nevertheless, the feedback is vital for improving diagnostic capabilities. Thus, the aim of this research was to compare results of prenatal echocardiography with postnatal angio-CT in patients with anomalies of great vessels.

Material and methods: We retrospectively compared results of prenatal echocardiography and postnatal angio-CT in 10 patients with selected anomalies of the aortic arch. This was a qualitative analysis, thus discrepancies in recognized anomalies were compared between these two modalities.

Results: In 8/10 patient diagnoses were fully consistent. Nevertheless, the tiny caliber of vessels created a diagnostic challenge (e.x. to differentiate the hypoplastic aortic arch from the aortic arch interruption). In the remaining case, the discrepancy was due to a problem with complete visualization of all branches of the aortic arch in prenatal ultrasound.

Conclusions: Fetal echocardiography in tertiary center was a reliable method for assessment of great vessels anomalies. However, critically narrow vessels remain a diagnostic challenge and neonatal angio-CT seems to be the method of choice in cases of diagnostic doubts.

Key words: *prenatal cardiology, congenital heart defects, angio-CT, aortic arch, echocardiography.*

INTRODUCTION

In fetal echocardiography, the 4 chamber view and outflow tracts are usually well seen. It allows recognize most common cardiac pathologies such as hypoplastic left ventricle or complete atrioventricular canal, just to mention the most common prenatal heart defects[1]. However, there are anomalies that become diagnostic challenges for fetal echocardiography. Thus, answering for quarries whether there is an aortic hypoplasia with coarctation of the aorta or an aortic arch interruption might be problematic.

Therefore the aim of our work was to determine the accuracy of a fetal echocardiography in a tertiary center for fetal cardiology in diagnosing selected / anomalies of the aortic arch. This issue will be addressed by comparing diagnoses made with prenatal echocardiography and postnatal angio-CT as a gold standard method. Based on this comparison we will indicate causes of discrepancies, what may improve diagnostic efficiency of prenatal echocardiography.

MATERIAL AND METHODS:

It was a retrospective analysis of selected cases who had prenatal echocardiography at no more than 14 days before delivery followed by a neonatal angio-CT prior to any cardiovascular intervention. All examinations were performed in years 2014-2016.

Fetal echocardiography was performed in our referral center for fetal cardiology due to abnormalities picked up in obstetrical screening ultrasound. For scanning two, high-quality sonographic systems were applied: IU 22 Philips or GE 730 Expert, equipped with convex and cardiac probes. All examinations

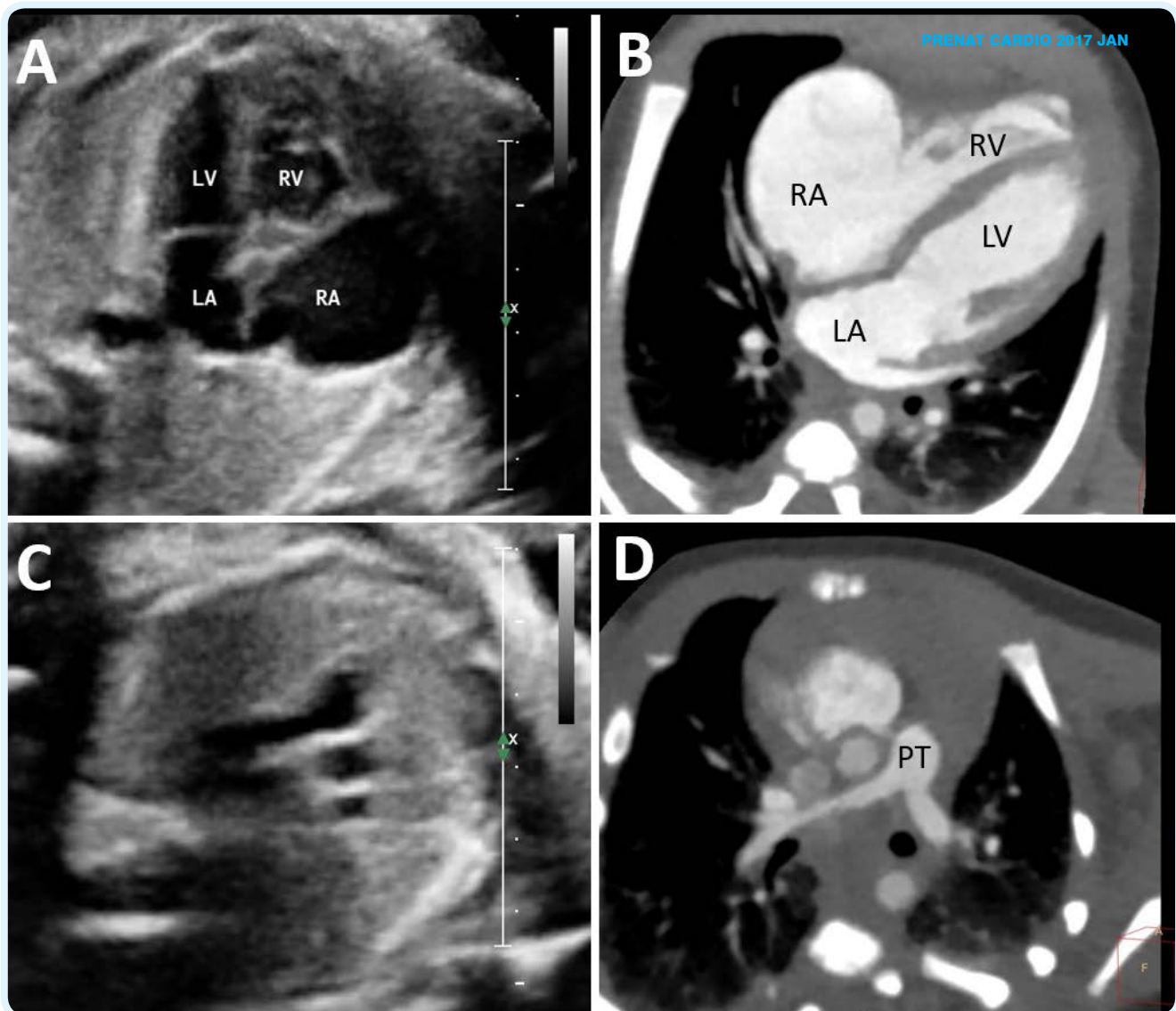*Corresponding author: majkares@uni.lodz.pl*

Fig. 1. RA dilatation due to atrial diverticulum recognized in prenatal ultrasound (A – four chamber view) and confirmed in neonatal angio-CT (B – transverse axis). Normal mediastinum in ultrasound (C – 3 vessel view) and angio-CT (D – transverse axis). LV – left ventricle, RV – right ventricle, LA – left atrium, RA – right atrium, PT – pulmonary trunk.

were performed by experienced echocardiographers (MRL, MS) who stored all acquired data.

Computed tomography examinations were performed with a The Brilliance iCT SP CT scanner (Philips, The Netherlands; slice thickness 0.9 mm, kVp 80, mAs 50–100, iDose 6) in the first week after birth. Patients were positioned prone with their hands above the head to minimize the radiation dose. Before the CT newborns had mild sedation by means of intravenous midazolam (0.1mg/kg) injection. The contrast agent (Omnipaque, 350mg iodine/ml; GE) was automatically injected (Medrad Stellant CT Injection System, Bayer HealthCare). The acquired datasets were evaluated by an experienced pediatric radiologist trained in heart CT evaluation (10 years of experience) who applied the IntelliSpace Portal 6.0 (Philips, The Netherlands) software. Median radiation dose was below 0.4mSv and consumption of contrast dye was 2ml/kg in all cases.

Reports from prenatal echocardiography and neonatal angio-CT, together with selected images, were compared by an independent specialist (PG) to evaluate similarities and discrepancies between modalities. Only qualitative elements of a diagnosis were assessed, not any direct measurements.

RESULTS

Patients

Ten cases were included into the analysis (6 males and 4 females). These 10 fetuses had 35 fetal echocardiography examinations. The first one was performed at mean of 25.6 (SD 4.6) hbd and the last one was performed at mean of 36.8 days (SD 1.1) hbd. A mean time between the last examination and the delivery was 10.8 days (SD 2.7).
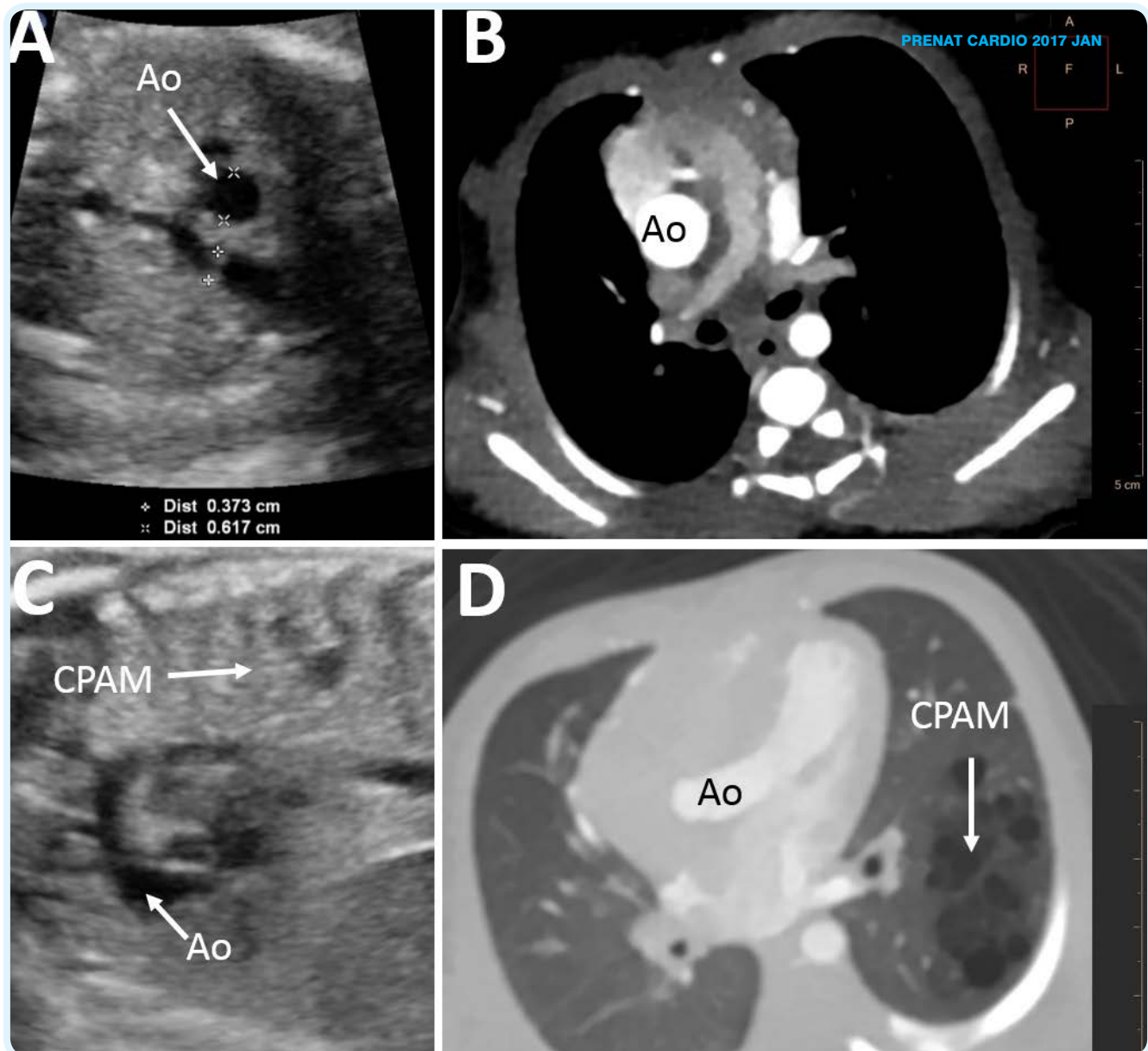
Fig. 2. Dilatation of the Ao (A – prenatal ultrasound, transverse view; B – neonatal angio-CT, transverse axis); C – prenatal ultrasound, longitudinal view; D – neonatal angio-CT, transverse axis) and coexistence of a congenital pulmonary airway malformations (CPAM) in the left lung. Ao – aorta.

There were 7 cesarean sections and 3 vaginal deliveries. The mean birth weight was 2992 g (SD 739 g), Apgar score was 8.2 (SD 1.2). There were no neonatal demises. Five newborns had cardiac surgery repair with good outcome. In remaining 5 patients treatment was postponed due to a good clinical conditions. Eventually, all patients were discharged home.

Cases

In eight patients aorta-associated anomalies were recognized, and in three patients mainly the pulmonary trunk was affected. One additional patient (Fig. 1) had no great vessels abnormality and was selected to present normal fetal mediastinum and normal great vessels, however, his major problem was an abnormal right atrium.

In 8/10 patient diagnoses made with prenatal ultrasound were consistent with postnatal angio-CT findings. Detailed information about evaluated cases and differences in stated diagnoses are presented in table 1 and in figures 2-11. In general, main difficulties in prenatal echocardiography concerned differentiation between significant narrowing (e.x. a hipoplastic aortic arch or a pulmonary trunk) and complete discontinuity of the vessel (e.x. an interrupted aortic arch or pulmonary atresia with no pulmonary trunk). Furthermore, difficulties in fetus positioning resulted in problems with recognition of vessels topography or the aortic arch branching pattern. Nevertheless, in all cases, prenatal echocardiography supported clinicians with information vital to plan the postnatal management. Discrepancies between diagnoses did not change radically the treatment strategy (e.x. prostaglandins will be administered in children born with hypoplastic aortic arch as well as with interrupted aortic arch to sustain the patency of ductus arteriosus).
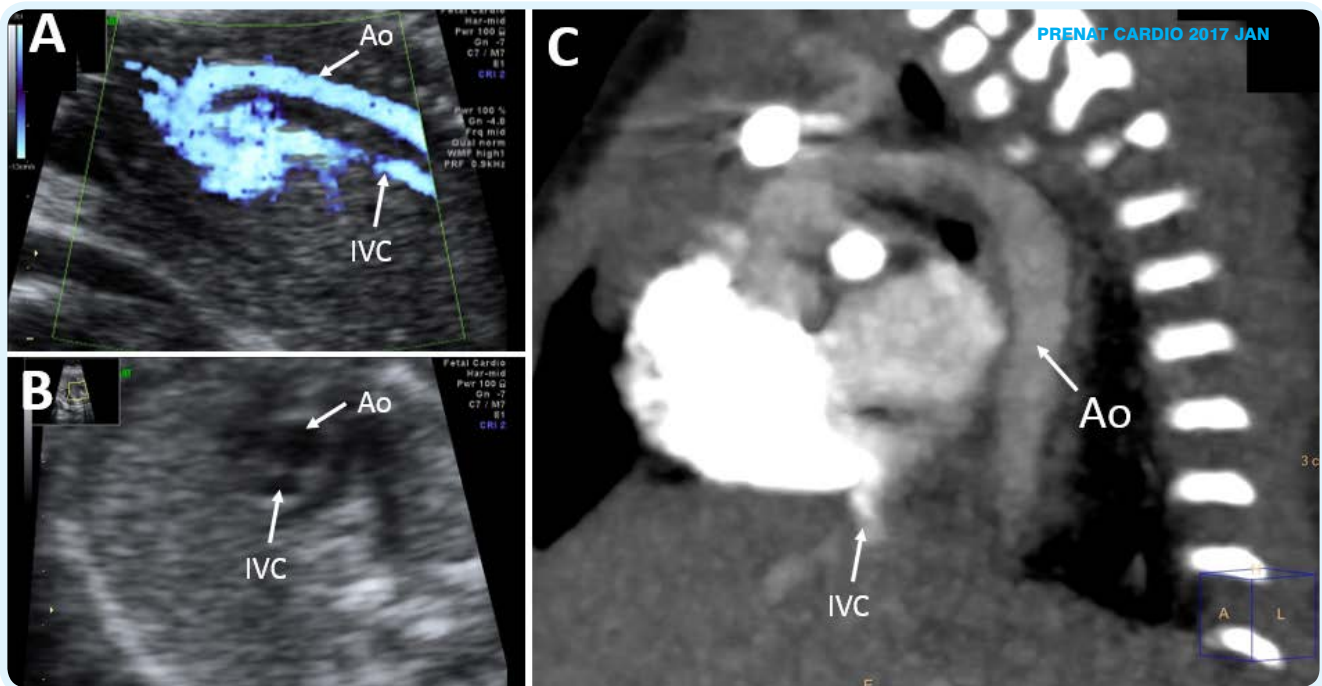
Fig. 3. A Double aortic arch suspected in prenatal ultrasound based on parallel alignment of the Ao and IVC (A – longitudinal view; B – mediastinum view). The correct diagnosis was of a right aortic arch was made based on neonatal angio-CT (C – sagittal reconstruction).

DISCUSSION

In fetal echocardiography, we usually start with assessing the situs of the stomach and the fetal heart apex. Then we evaluate a 4 chamber view, fetal mediastinum and finally systemic and pulmonary veins connections. Both, structural and functional evaluation is equally important to make conclusions about the diagnosis and prognosis [2]. If the pathology of the cardiovascular system is recognized it has to be categorized according to a fetal heart defects classification. It helps parents and a medical team to plan a postnatal care. It is vital because some defects require treatment in first days of life and in other cases treatment can be postponed for 2nd or 3rd week of postnatal life or patient can be treated conservatively [3,4].

To make a reliable diagnosis a meticulous scanning technique is required. For detecting anomalies concerning the major vessels in the mediastinum Yoo et al. [5] introduced the three vessels and trachea view. Its value was confirmed by several authors [6,7,8] Nevertheless, others suggested that 3 and 4 dimensional off-line analysis or fetal heart MRI should be applied [8-11]. Despite all the above, we showed that even by a "simple echocardiography" without 3 dimensional or STICK evaluation, it is possible to recognize an aortic arch coarctation, an interruption of the aortic arch [12] or to show the MAPCAs in cases of pulmonary atresia [13]. We accounted some errors, however, they did not bring any harm to newborns because they did not change the initial management. On the other hand, they remained us how important is a validation of echocardiographic findings.

In the past, to verify our prenatal diagnoses we used an autopsy results[14] or reports from the surgical theater[15].

However with modern imaging modalities nowadays, we may compare the prenatal echocardiographic imaging with excellent quality and more objective postnatal images such as obtained by neonatal angio-CT [16]. Its side effect is, however, an exposition to an appreciable dose of radiation. Moreover, an administered contrast agent is potentially harmful to kidneys. Finally, angio-CT provides only information on morphology and not a function of cardiovascular system. Thus, it does not allow for example to estimate a gradient in case of aortic arch coarctation. Hence, this modality should be used as a supplementary technique for final clarification in extremely difficult cases or for purpose of cardiosurgery planning [17,18].

So far in the literature, our research is one of the very first. By the comparison of the prenatal echocardiography with the postnatal angio-CT, we were able to obtain the "missed" link to finally verify the prenatal evaluation. Angio-CT reconstruction and its visualization side by side with prenatal images allowed fetal echocardiographer to better interpret their findings. It also allowed appreciating most common causes of omissions in diagnosis. Firstly, if a tiny canal is present hipoplasty may be confused with total interruption of the continuity. Secondly, due to difficulties in fetus alignment recognition of great vessels topography might lead to misinterpretations.

To avoid this fields of potential error we advise not only to scan with great caution but to perform a multiple scanning during the course of the pregnancy. The fetal heart may evolve during the entire pregnancy [19] and the last fetal echocardiography before the delivery provides the most reliable diagnosis [3,4]. In our series of 11 fetuses, we performed 37 fetal echocardiography examinations. It
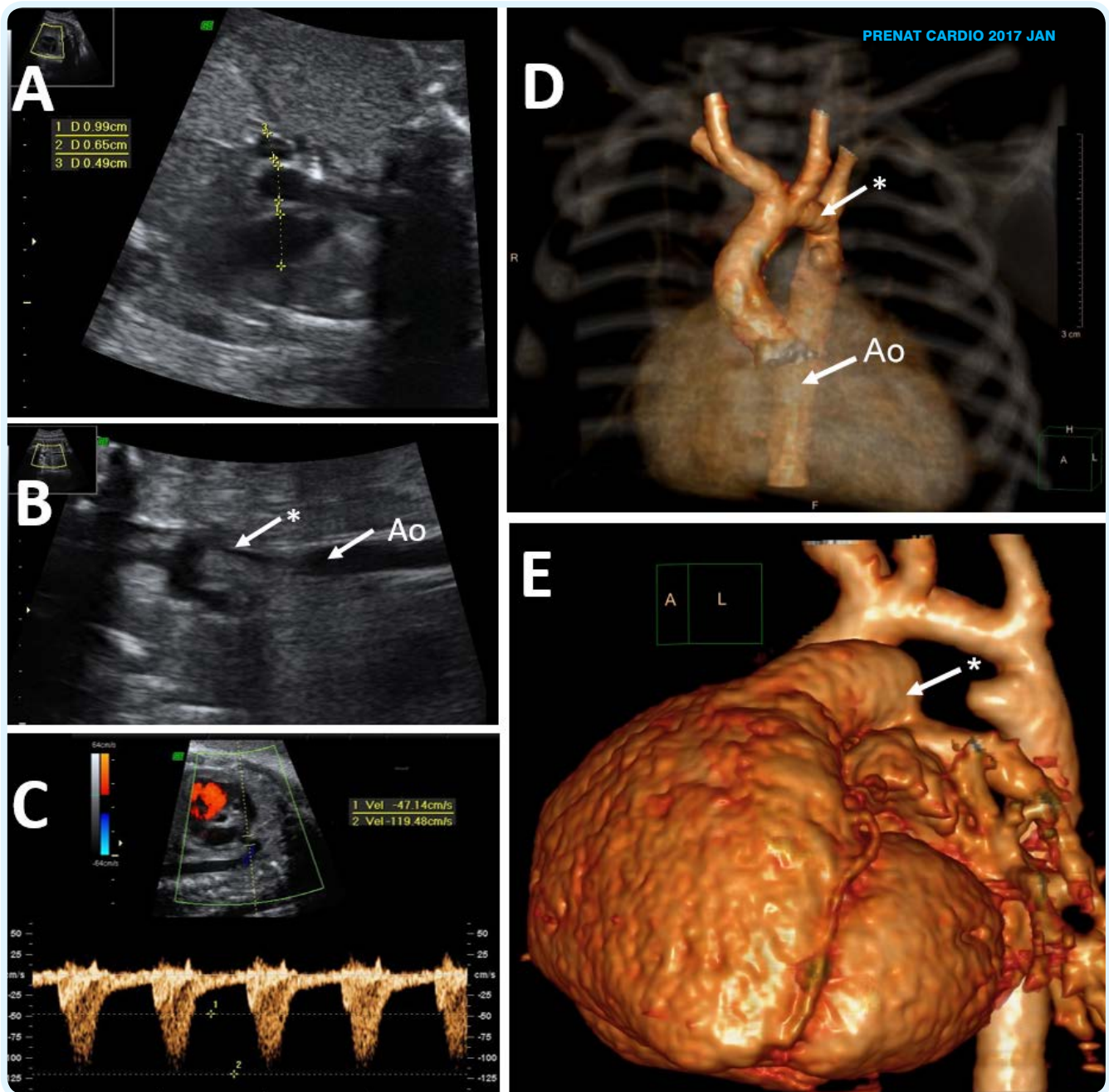
Fig. 4. An abnormal narrowing of the Ao arch with short transversal part and bovine type of cephalic branches (two instead of classical three) suspected in prenatal ultrasound (A – transverse view; B – longitudinal view). Colour and Pulse Wave Doppler analysis (C) confirmed a narrowing of the Ao isthmus. Slight CoA was also confirmed in neonatal angio-CT, however „3rd cephalic vessel" was present. (D and E – 3D reconstructions, anterior view). Ao – aorta; CoA – coarctation of the aorta; * - place of coarctation.

shows how difficult it was to complete prenatal diagnosis and on the same token shows the necessity to monitor hemodynamic changes during fetal life.

This study has a few limitations. The study population was limited because the majority of newborns diagnosed with fetal echo did not undergo chest CT. Secondly, it should be bared in mind that after birth blood hemodynamics changes dramatically. Thus some differences in interpretation might be due to physiological changes.

Summarizing, the comparison of finding from the fetal echocardiography with the neonatal angio-CT is a good method to confirm the reliability of the prenatal diagnosis.

Although there might be some discrepancies between this techniques cooperation of cardiologists and radiologists may help to establish the correct diagnosis. This cooperation is vital in such a difficult area as the fetal/ neonatal heart and in our opinion, it should become mandatory in the learning path for echocardiographers as well as for the radiologists working in the field of cardioimaging.
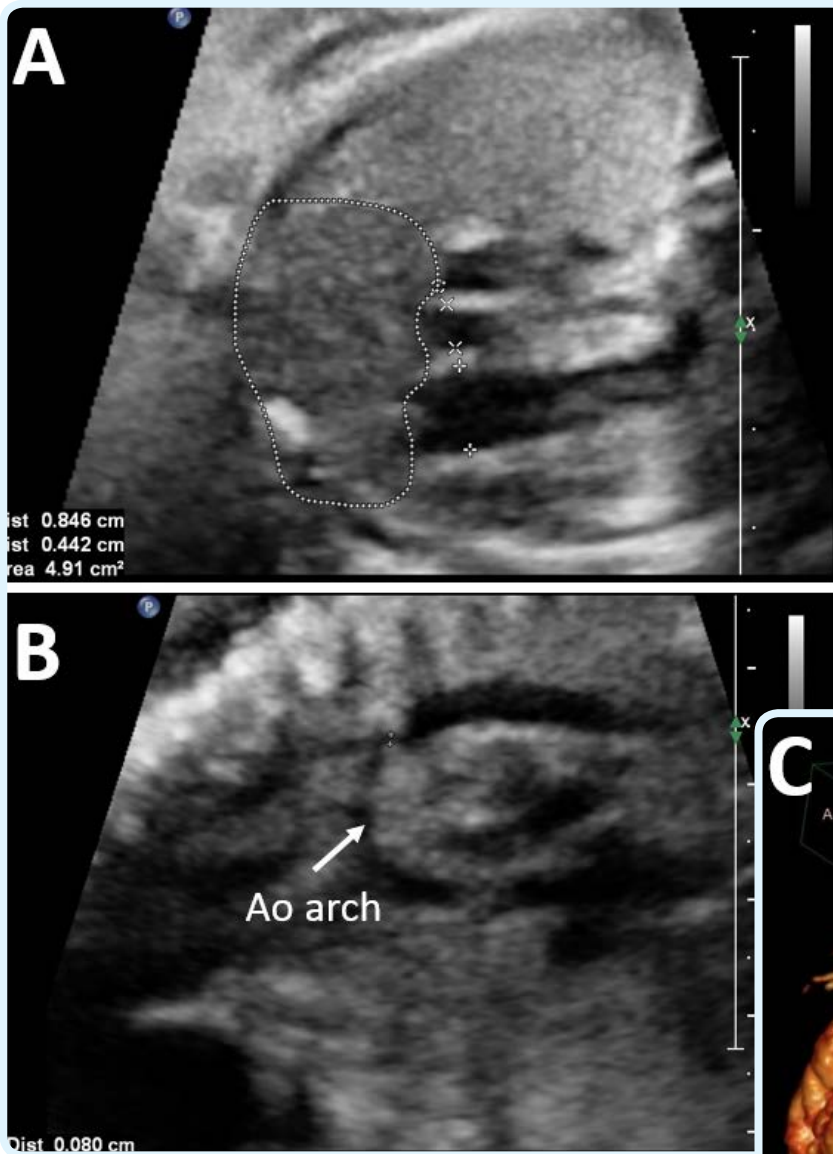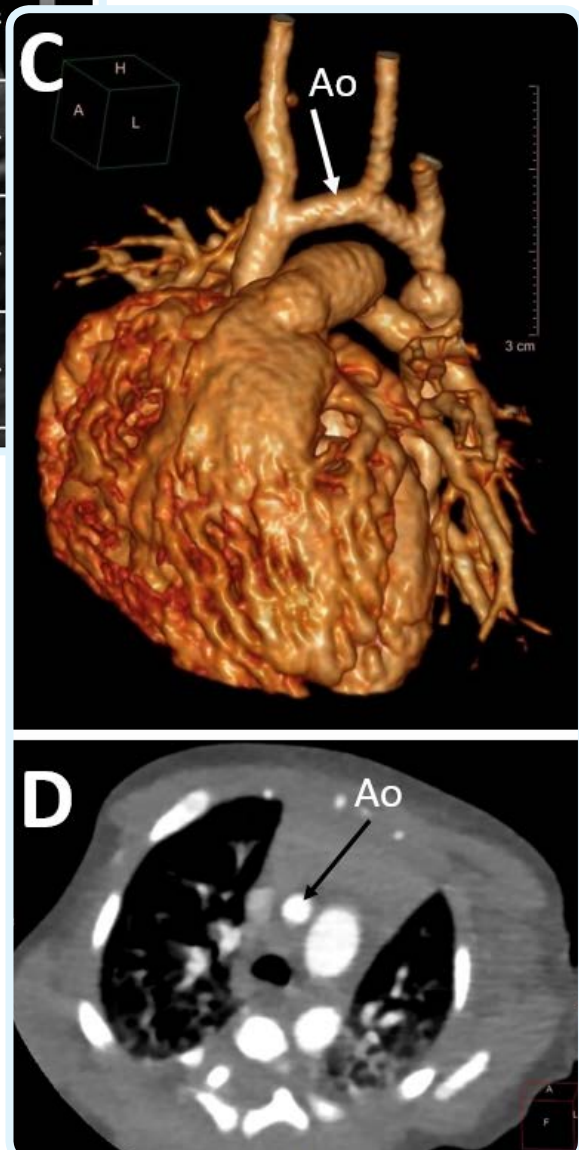
Fig. 5. Hipoplastic Ao arch recognized in prenatal ultrasound (A - 3 vessels view; B – longitudinal view) and confirmed in neonatal angio-CT (C - 3D reconstruction, anterolateral view; D – transverse axis). Ao – aorta.

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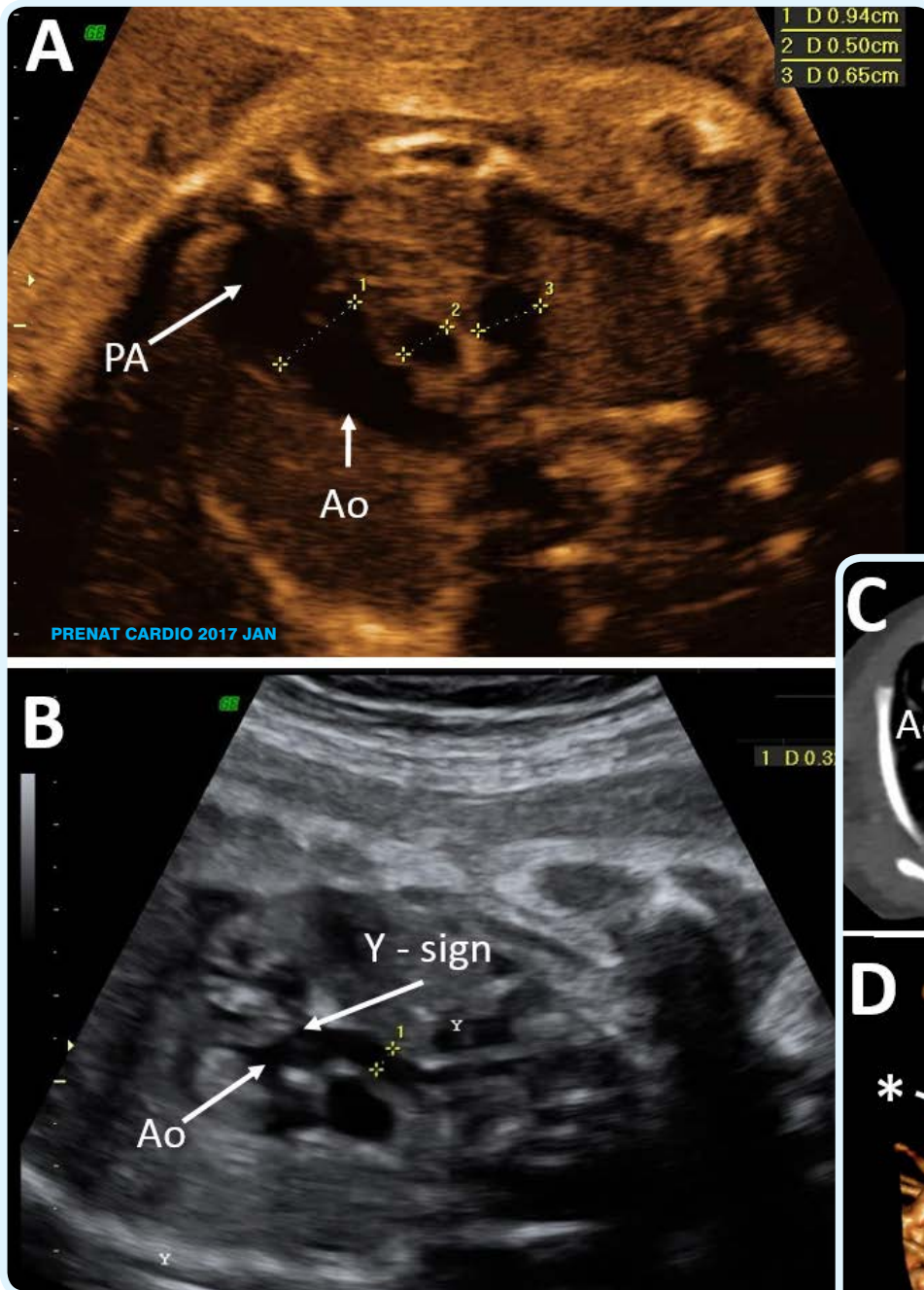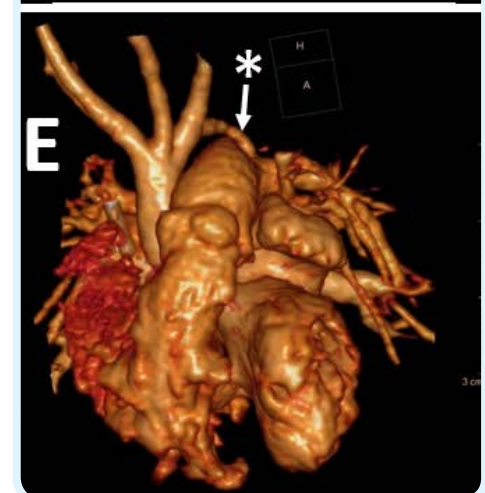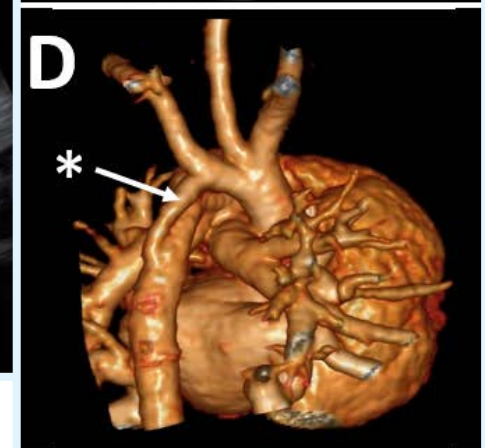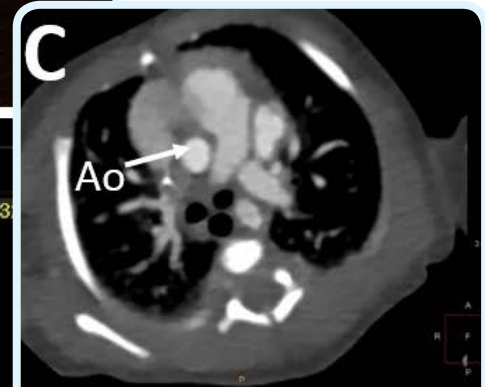
Fig. 6. Hipoplastic and interrupted Ao arch recognised in prenatal ultrasound (A – transverse view; B – longitudinal view with Y sign). Neonatal angio-CT confirmed a diagnosis (A – transverse view; B - 3D reconstruction, posterior view; C – 3D reconstruction, anterior view). Ao – aorta; PT – pulmonary trunk.

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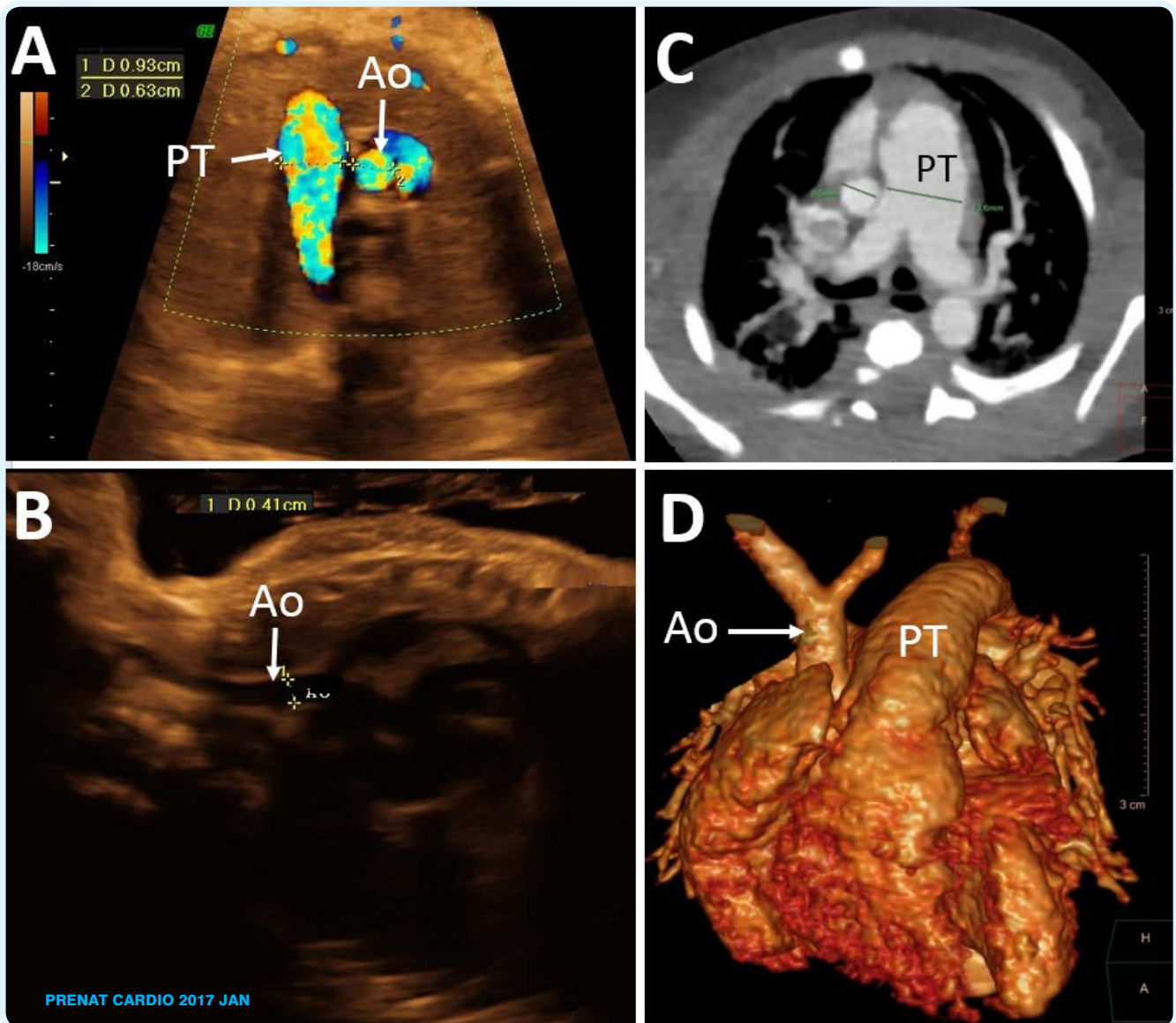
Fig. 7. Interrupted Ao arch recognised in prenatal ultrasound (A – transverse view through the mediastinum with narrow Ao and enlarged PT; B – longitudinal view with Ao traveling cranially with no Ao arch visible). Diagnosis confirmed in neonatal angio-CT (C – transverse axis with dilatation of the PT; D – 3D reconstruction, anterior view). Ao – aorta; PT – pulmonary trunk.

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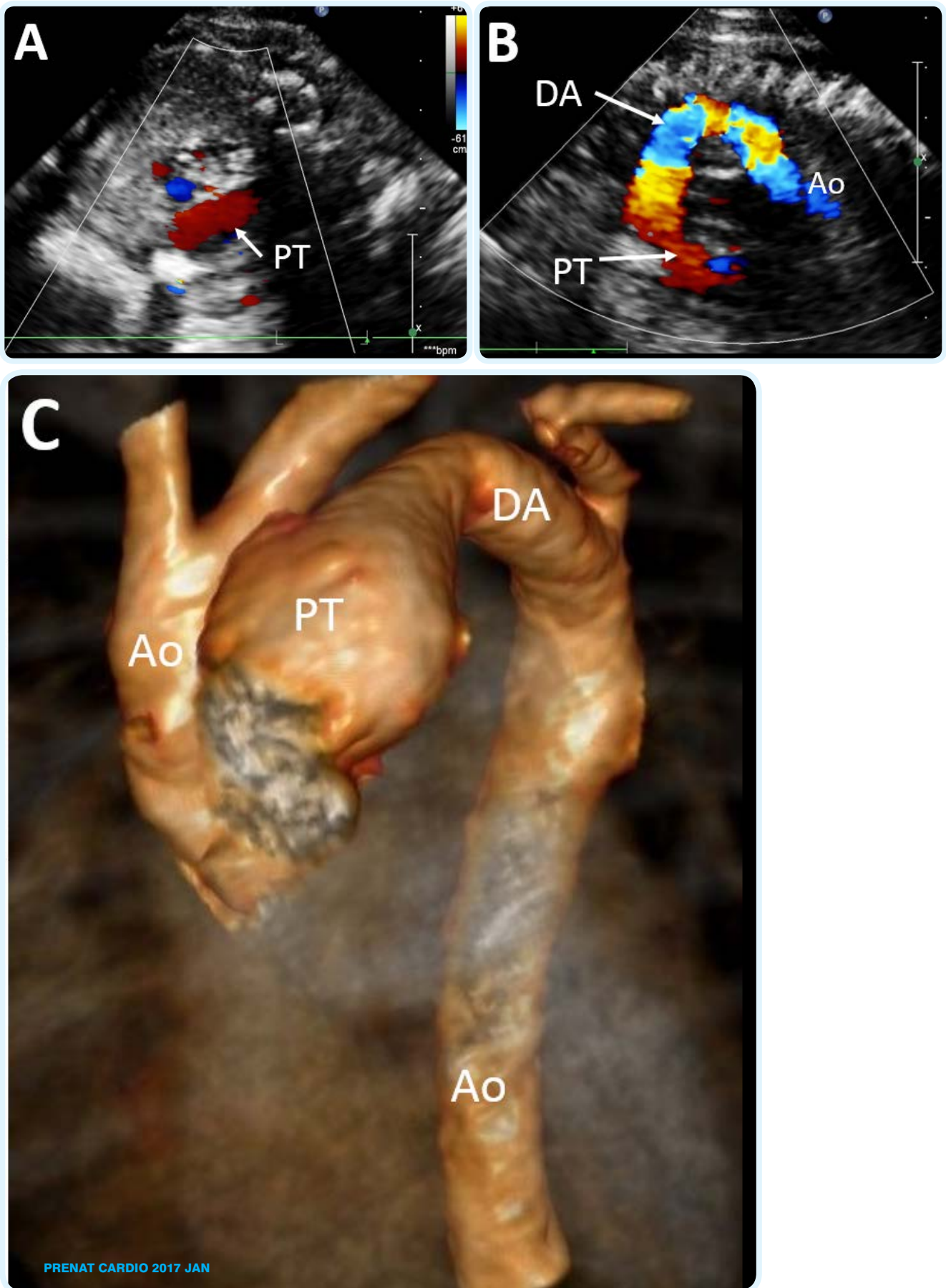
Fig. 8. Interrupted Ao arch recognised in prenatal ultrasound (A – transverse view in colour Doppler showing dilatation of the PT; B – longitudinal view with abnormal shape of ductus arteriosus – „broken hockey stick"). Neonatal angio CT confirmed prenatal findings (C – 3D reconstruction, anterior view). Ao – aorta; PT – pulmonary trunk; DA – ductus arteriosus.
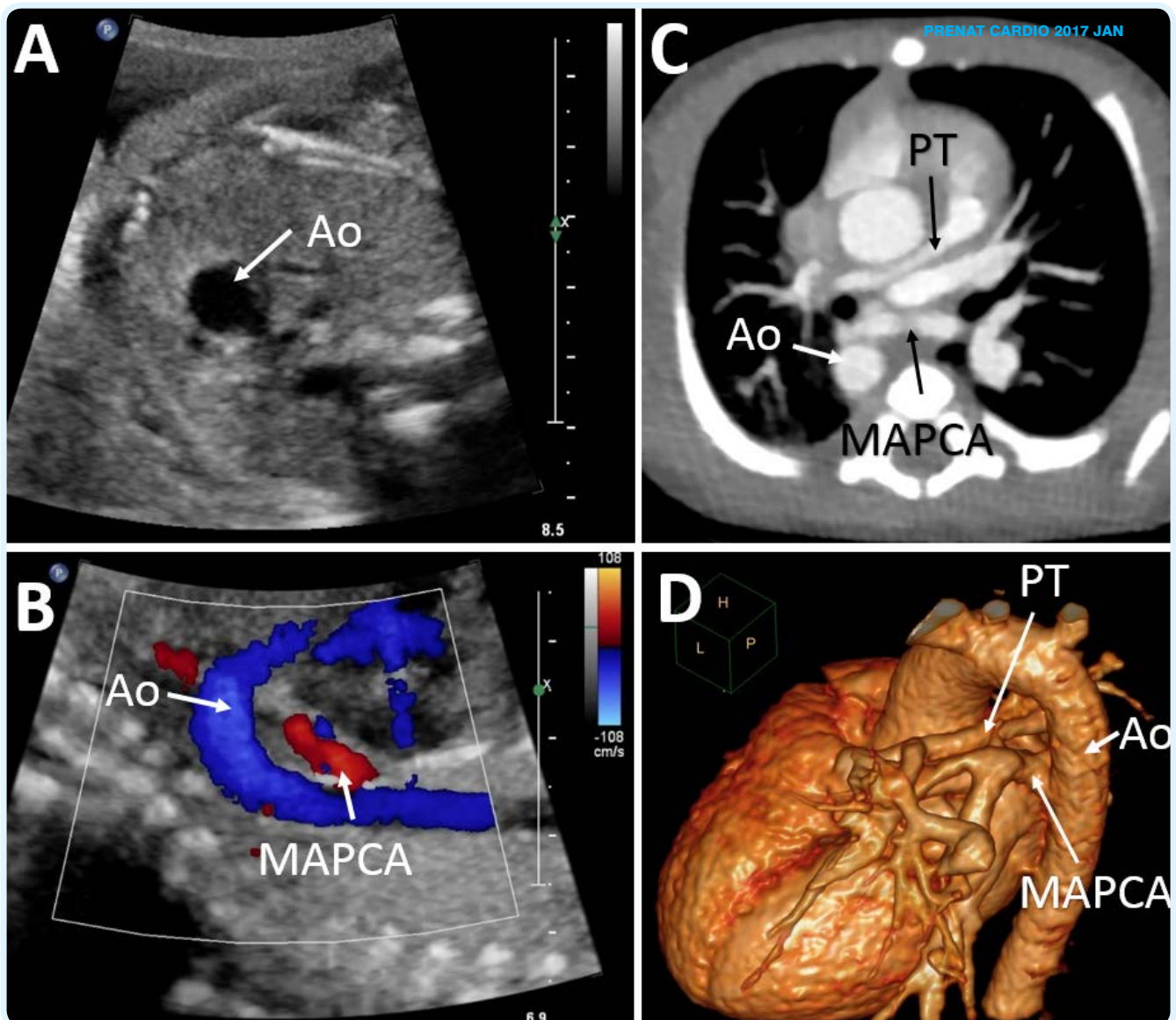
Fig. 9. Pulmonary atresia with MAPCA and lack of PT suspected in prenatal ultrasound based on visualisation of only one large vessel in the mediastinum (A – transverse view) and occurrence of abnormal branch starting from the ventral side of the Ao (B – colour Doppler, longitudinal view). Diagnosis confirmed in neonatal angio-CT (C – transverse axis atresia of the PT and MAPCA branching from the Ao; D – 3D reconstruction, supero-lateral view from the left side). Ao – aorta; MAPCA – major aortopulmonary collateral artery; PT – pulmonary trunk.
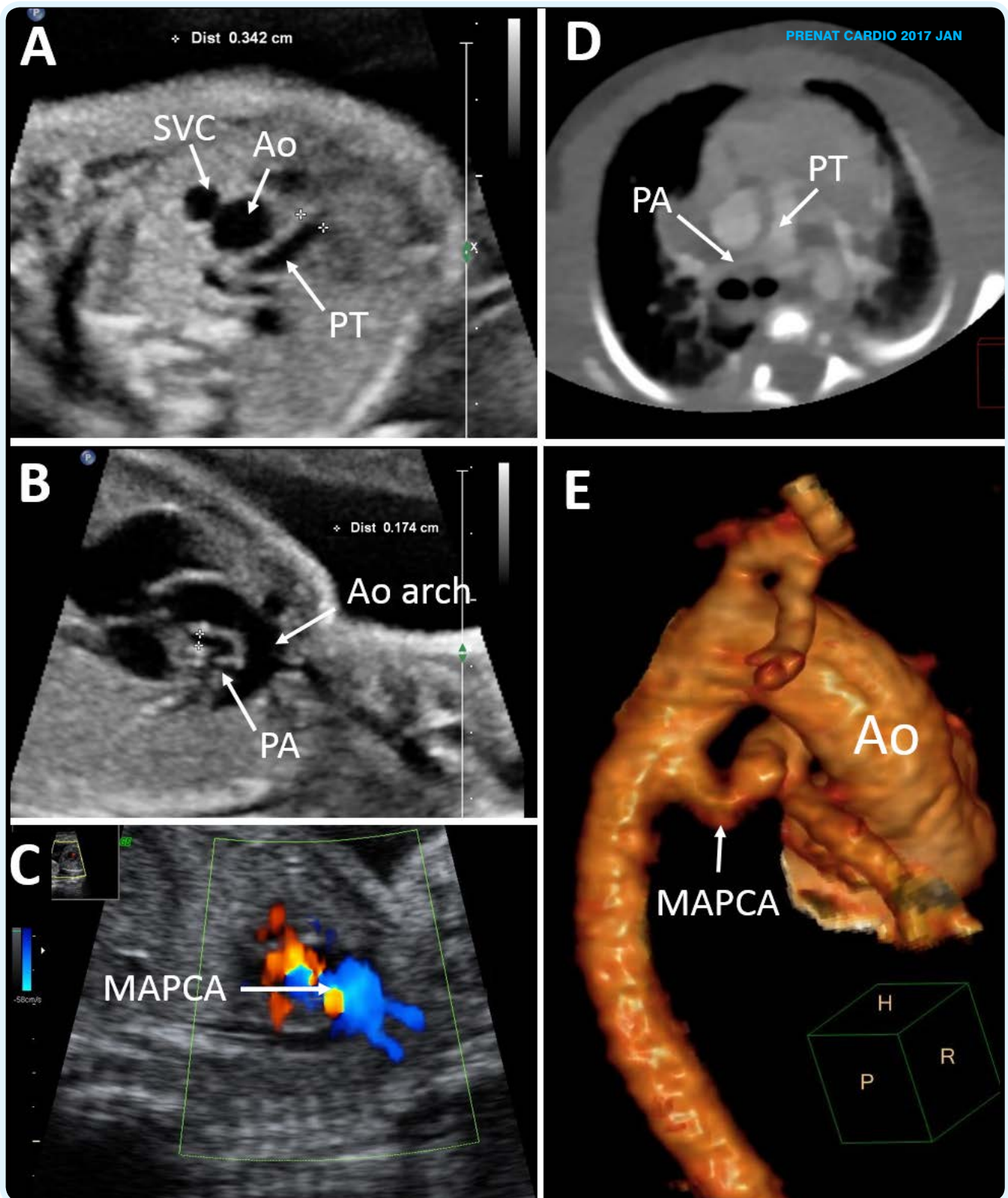
Fig. 10. Pulmonary atresia with MAPCA and vestigial PT recognised in prenatal ultrasound based on presence of enlarged Ao and small PT in mediastinum (A – transverse view), occurrence of small pulmonary arteries under the Ao arch (B – longitudinal view), and turbulent flow suggesting the presence of MAPCA (C – colour Doppler, longitudinal view). Diagnosis confirmed in angio-CT (D – transverse axis at the level of mediastinum; E – 3D reconstruction, lateral view). Ao – aorta; MAPCA – major aortopulmonary collateral artery; SVC – superior vena cava; PT – pulmonary trunk; PA – pulmonary artery.
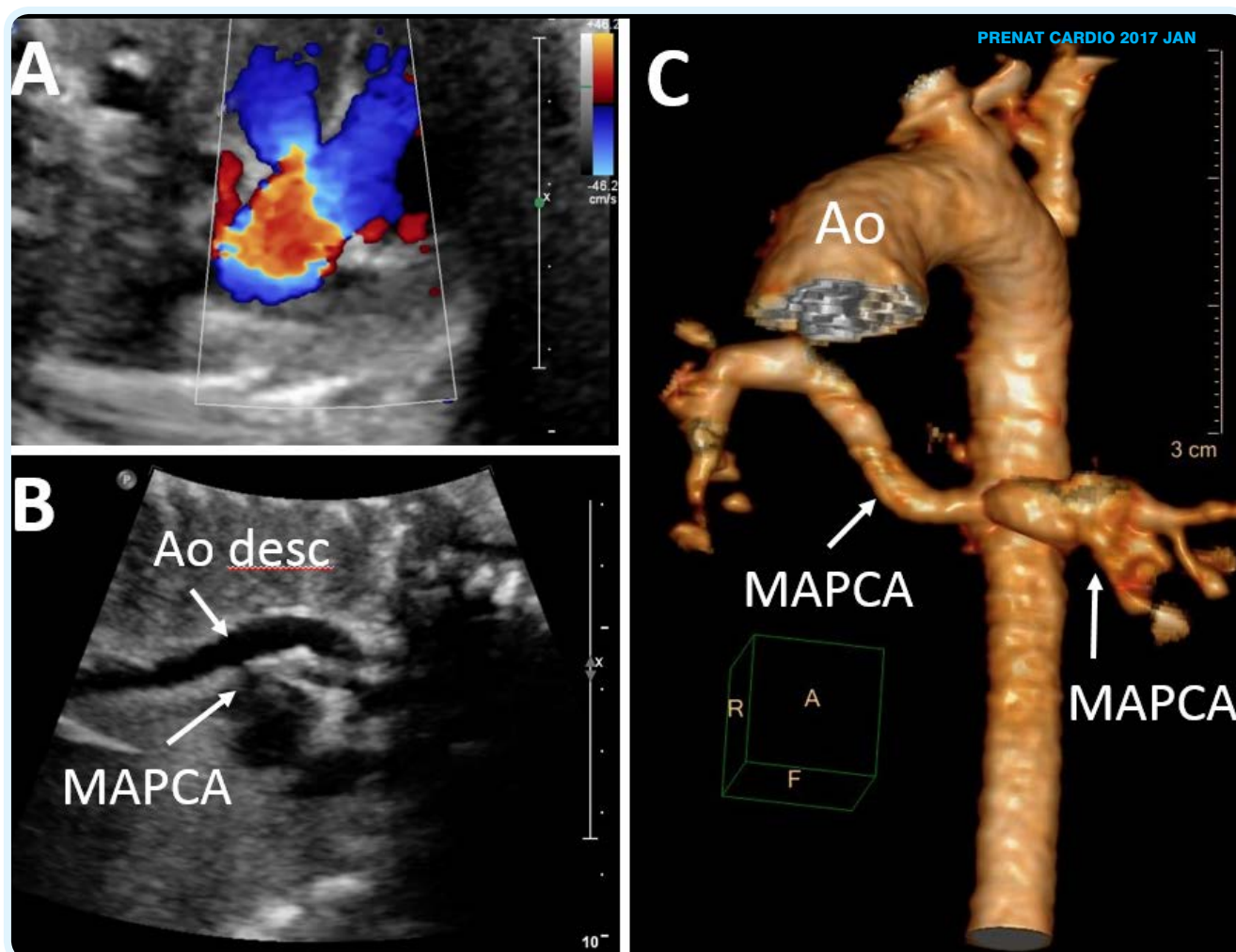
Fig. 11. Truncus arteriosus with MAPCAs recognised in prenatal ultrasound based on presence of an overriding „aorta" starting above the VSD (blood from both ventricles flows into one big vessel). A – colour Doppler, longitudinal view, abnormal vessel starting from the ventral side of the Ao; B – longitudinal view showing MAPCA starting from the desc Ao. Diagnosis confirmed in angio-CT (C – 3D reconstruction, anterior view). Ao – aorta; MAPCA – major aortopulmonary collateral artery; desc Ao – descending aorta.

Lp.	Diagnosis from prenatal echocardiography	Diagnosis from neonatal angio-CT	Discrepancies
1.	Diverticulum of RA	Dilatation of the RA	Non
2	Ao dilatation + Ao regurgitation + CPAM	Ao dilatation and CPAM	Non
3	Double Ao arch	Right aortic arch	There was no left Aortic Arch
4	CoA with hypoplastic transversal Ao arch and bovine type of cephalic branches	CoA	CoA but with normal Ao arch branching pattern
5	CoA	Hipoplastic Ao arch	Narrowing of the Ao was more extended
6	Hipoplastic Ao arch + IAA	Hipoplastic Ao arch	In angio-CT there was a contrast flow through the narrowed segment of the Ao
7	IAA	IAA	Non
8	IAA	IAA	Non
9	PA with no PT and MAPCA	PA with hipoplastic PT and MAPCA	In angio-CT there was a contrast flow through the hipoplastic PT
10	PA with hipoplastic PT and MAPCAs	PA with hipoplastic PT and MAPCA	Non
11	PA with truncus art and MAPCAs	PA with truncus art and MAPCAs	Non

Ao – aorta; CoA – coarctation of the aorta; CPAM – congenital pulmonary airway malformations; IAA – interrupted aortic arch; IVC – inferior vena cava; MAPCAs - major aortopulmonary collateral arteries; PA – pulmonary atresia; PT – pulmonary trunk; RA – right atrium; VSD – ventricular septal defect;

Table 1: Prenatal and postnadal data of 11 cases selected for comparative analysis