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Case report

Prenatal dilatation of the pulmonary artery in the third trimester – important information for paediatric cardiologist or for neonatologist? Case report

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Abstract

We present a case of dilated main pulmonary artery diameter with accompanying cardiac anomalies and postnatal follow-up. The abnormalities in the left side of the heart concerned the left superior vena cava (LSVC), decreased left ventricular contractility SF LV (shortening fraction), and narrowing/accelerating flows through the aortic valve and aortic arch isthmus. In our case, coarctation of the aorta was suspected both pre- and postnatally due to the dominant picture of the accelerated flow velocities, but the patient did not require neonatal surgery. Based on a retrospective review of the case, we tried to explain our prenatal and early postnatal findings. After the exclusion of the structural congenital heart defects as a base of the dilatation of the main pulmonary artery, we should consider other pathophysiological mechanisms presented in the following report.

Key words: dilated pulmonary artery, coarctation of the aorta, false-positive CoA, case study, third trimester, prenatal cardiology, fetal echocardiography, fetal echo, prenatal diagnosis, differential diagnosis.

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Introduction

The broad pulmonary trunk in the third trimester of gestational age should be given special attention in the context of predicting the postnatal compromise and the assessment of cardiopulmonary efficiency of newborns [1–9]. An intrauterine environment, an in-utero infection, and fetal pulmonary hypertension can contribute to abnormal prenatal echocardiographic findings and may predict abnormal newborns' adaptation to ex-utero life [1–9].

We present a case of dilated main pulmonary artery diameter (up to 13 mm) with accompanying cardiac anomalies and postnatal follow-up.

Case report

A pregnant woman, G3P2, with positive medical reconnaissance for a right bundle branch block was directed to the Fetal Cardiology Centre in the 3rd Reference Hospital due to suspected heart defect of a fetus. The abnormalities concerning disproportions at the level of four cardiac chambers and

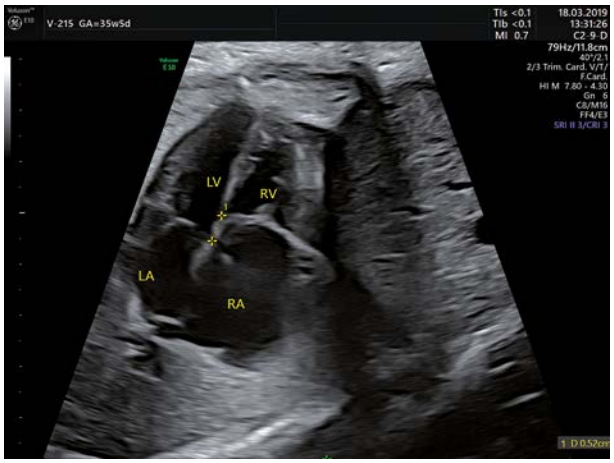

Figure 1. 4CV disproportion, dysplastic TV, 36 weeks of pregnancy, MDT-mitral valve-tricuspid valve distance 5.2 mm dysplastic TV

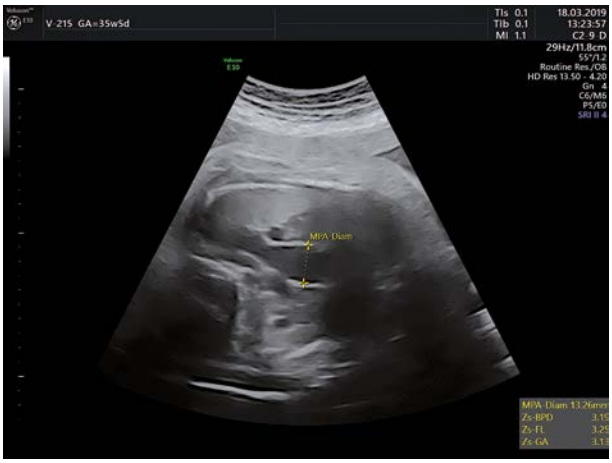

Figure 2. Dilatation of MPA at 36 GA as a predictor of heart defect or persistent pulmonary hypertension? In mediastinum there are 4 vessels (bilateral SVC, narrow aorta, and good size of the thymus)

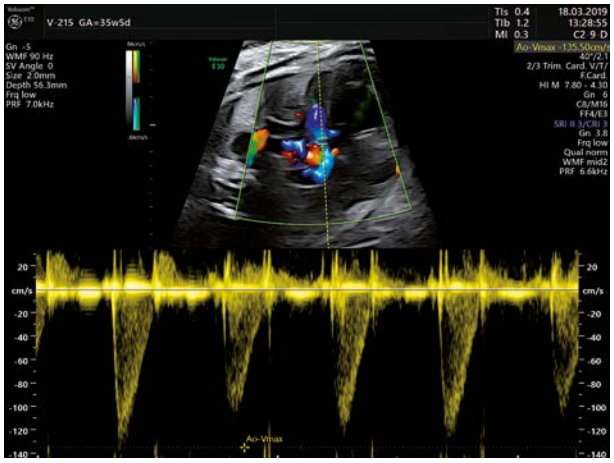

Figure 3. Increased AoV vel flow, 36 weeks, due to CoA?

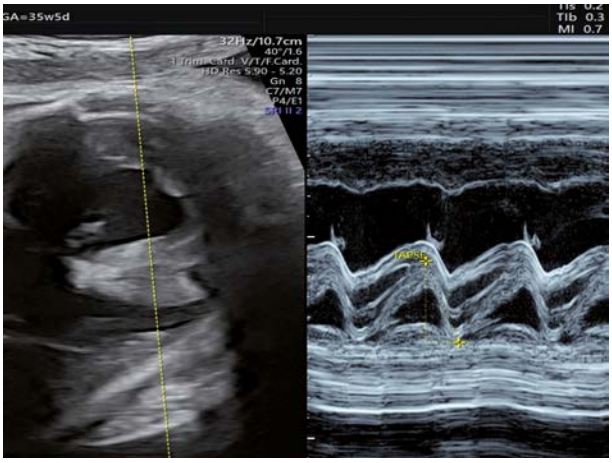

Figure 4. Increased TAPSE measurement at 36 weeks of pregnancy due to dysplastic TV?

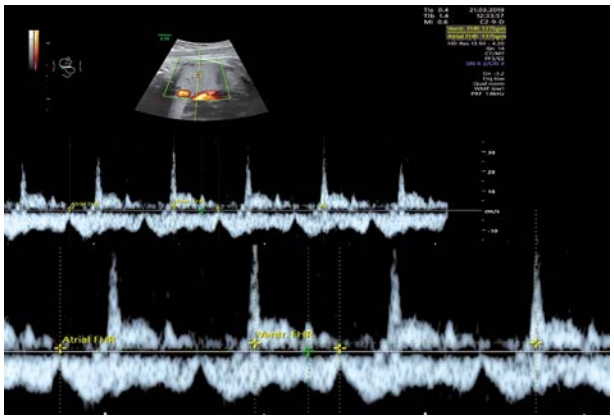

Figure 5. PVs flows, FHR ventricles = FHR atria = 137 bpm, 36 weeks

three vessels of the upper mediastinum were detected at the 21st week of the pregnancy. The first specialised echocardiographic examination in the tertiary centre in our hospital took place at the 36th week. Then, attention was paid to the dominance of the right side of the heart (Figure 1), with a broad pulmonary trunk main pulmonary artery diameter (MPA dia) of 13 mm, Z-score of +3.13 for gestational age (GA) (Figure 2), and narrow branches of the pulmonary trunk, as well as to the difference between dimensions of the atria and ventricles > 2 mm, without signs of cardiomegaly heart area/chest area (HA/CA) = 0.35.

The abnormalities in the left side of the heart concerned the left superior vena cava (LSVC), decreased left ventricular contractility SF LV = 23% (shortening fraction), and narrowing/accelerating flows through the aortic valve and aortic arch isthmus. The maximum flow velocities through the aortic valve amounted to 135.5 cm/s (Figure 3), via ductus arteriosus/aortic isthmus (DA/AoI) 156 cm/s prenatally. The distance between the mitral valve (MV) and tricuspid valve (TV) insertion to the interventricular septum (IVS) exceeded 5 mm. The tricuspid annular plane systolic excursion (TAPSE) rate was significantly elevated, at 14 mm. The tricuspid valve was assessed as dysplastic (Figure 4).

The venous pulmonary flows were accelerated, v_{max} 46 cm/s, while the ventricular and atrial heart function were compatible FHR = 137 bpm (Figure 5) and ventricular-atrial conduction time was extended (PR interval = 169 ms). The assessment of

the fetal circulatory compromise in the cardiovascular scale CVPS amounted to 9/10 points (–1 for SF LV) (Table 1).

Except for the cardiac abnormalities, hydrops testis and fetus large for gestational age (LGA) were diagnosed; LGA: 35w5d/38w4d – 3502 ±511 g. AFI was 16 mm. Placental thickness was 33 mm. Based on prenatal findings of reactive hydrocele, prenatal antibiotic vaginal therapy was proposed in the end stage of pregnancy. The spontaneous delivery took place seven days after the echocardiography at the 37th week of the pregnancy. The newborn had a birth weight of 3700 g, Apgar 7, 8. Assuming prenatal coarctation of the aortae as a ductal-dependent lesion, Prostin was given for 12 days (doses from 0.02 to 0.01 mg/kg/min).

Postnatal echocardiographic examination and 3D angio-CT examinations (Figures 6, 7) revealed narrowing of the mitral valve, and mild MV stenosis with incorrect histogram up to 1.4 m/s (mean PG – 3 mm Hg). The presence of LSVC, tricuspid valve with elongated leaflets, characterised with the dysplasia and with signs of regurgitation and valvular cusp prolapse with difficult visualisation of the posterior leaflet were described. There was a high located intramembranous ventricular septal defect of about 2 mm and narrowing of the aortic isthmus of about 3 mm with colour Doppler flow velocity up to 2.45 m/s (PG – 24 mm Hg). During the hospitalisation period, the reduction percentage of the N-terminal brain natriuretic peptide (NTproBNT), which predicts long-term mortality and readmission in heart failure patients, was seen.

In the next control postnatal echocardiograms (in the first two weeks of the newborn's life) no escalating gradient in the aortic isthmus was observed. Laboratory findings confirmed neonatal infection, and antibiotic therapy with gentamicin and ampicillin was given for two weeks. The total hospitalisation time amounted to 30 days, including 17 days at the Neonatal Intensive Care Clinic. The newborn was not qualified for cardiosurgery and was discharged after long-term pharmacological treatment (as mentioned before) and recommendations to attend a further Cardiology Outpatient Clinic.

Table 1. Prenatal echocardiographic parameters

Parameters	Results
GA LMP	35.5
GA US	38.4
HA/CA	0.35
AP [mm]	42
LV SF [%]	**23**
TAPSE	**14**
MAPSE	5
Fo [mm]	7
Fo-direction of flow	R → L
MPA [mm]	**13**
Zs-BPD	+3.19
Zs-FL	+3.25
Zs-GA	**+3.13**
Ao [mm]	7.73
Zs-BPD	+1.81
Zs-FL	+2.64
Zs-GA	+1.92
AoV v_{max} [cm/s]	135.5
AoI v_{max} [cm/s]	**156.89**
CVPS	9/10

Discussion

The broad pulmonary trunk should draw special attention in the context of predicting the postnatal condition and the assessment of cardiopulmonary efficiency of newborns [1–2]. A dilated MPA in the three-vessel view could be found in isolated pulmonary valve stenosis with post-stenotic dilatation of the main pulmonary artery or in tetralogy of Fallot with absent pulmonary valve syndrome [3–8].

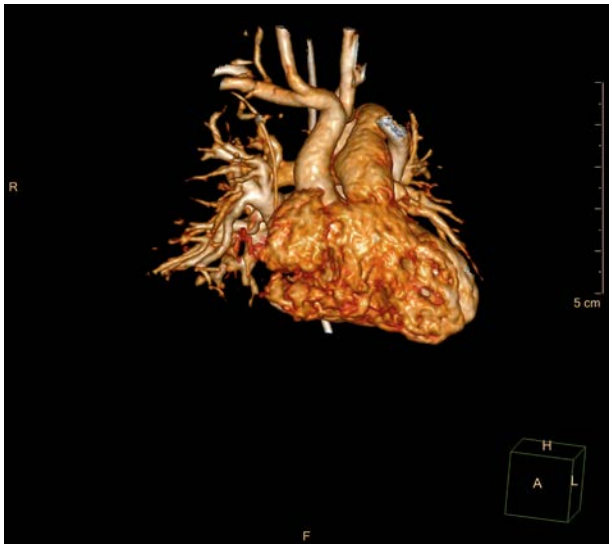

Figure 6. AngioCT, newborn: a picture of the coarctative nature of the isthmic segment (AoI 3 mm, v_{max} 245 cm/s)

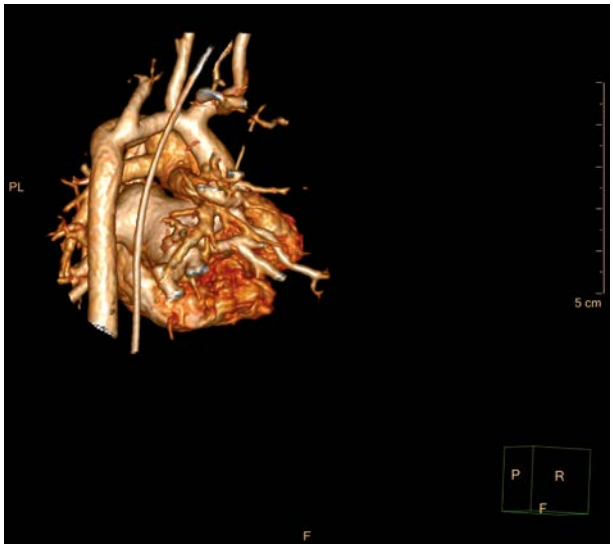

Figure 7. AngioCT, newborn: a picture of the coarctative nature of the isthmic segment (AoI 3 mm, v_{max} 245 cm/s)

Table 2. Differential diagnosis in dilated main pulmonary artery (MPA)

Dilated MPA in differential diagnosis	References
Pulmonary valve stenosis with post-stenotic dilatation	• A dilated MPA in the three-vessel-view with the turbulent antegrade flow could be found in isolated pulmonary valve stenosis with post-stenotic dilatation of the main pulmonary artery [3–8, 31–32]
Tetralogy of Fallot with absent pulmonary valve syndrome (APVS)	• A dilated MPA in the three-vessel-view in tetralogy of Fallot with absent pulmonary valve syndrome with to and fro blood flow and patterns of severe stenosis and insufficiency of the pulmonary valve • Width of MPA does not correlate with the survival of the newborns with APVS, but affects the respiratory tract compression and respiratory dysfunction, probably secondary to the pulmonary valve regurgitation, seen prenatally [3–8, 33–36]
Ductal constriction	• Constriction of fetal ductus arteriosus is a risk factor for pulmonary hypertension in the newborn period with known severe consequences [37–42]
Pulmonary hypertension/congenital diaphragmatic hernia (CDH), liver herniation (LV)	• Larger MPA diameter best correlates with postnatal death and respiratory morbidity in CDH • Fetal MPA diameters are probably related to the severity of pulmonary hypoplasia in CDH • PA diameters appeared to be correlated with prognosis in infants with CDH + LH [25, 27, 43, 44]
Pulmonary hypertension/HLHS	• Enlarged MPA in fetal hypoplastic left heart syndrome • There were statistical differences between PA/AO ratio at 39 weeks of gestation in pulmonary hypertension and HLHS groups [28, 43]
Coarctation of the aorta-CoA	• Fetuses whose PA/Ao is greater than 1.6 are more likely to suffer from CoA [13, 16]

Fetal heart disproportion in the cross section of four heart chambers and three vessels of the upper mediastinum in favour of the right side in the third trimester of a pregnancy may be the variant of the norm, but then coarctation of the aorta should be excluded [9–18].

In our case, coarctation of the aorta was suspected both pre- and postnatally due to the dominant picture of the accelerated flow velocities, with the maximum velocity AoI = 156–246 cm/s, but the patient did not require neonatal surgery. Based on retrospective review of the case we tried to explain our prenatal and early postnatal findings. The prenatal diagnosis of fetal CoA is still very challenging, and it remains one of the most difficult cardiac defects to diagnose before birth (Table 2). The positive diagnosis of the fetal CoA was accompanied by additional abnormalities, including LSVC, which could have the potential of impairing the left ventricular filling and imitated CoA.

With the presentation of the dilated MPA, smaller right pulmonary artery (RPA), left pulmonary artery (LPA), shorter time-to-peak velocity, absent end-diastolic flow, and increased pulsatility index in the pulmonary artery, the other possible explanation could be fetal increased pulmonary resistance as a base of the later neonatal hypertension. Different type of persistent pulmonary hypertension should be taken into differential diagnosis to speed up NO (nitric oxide) inhalation with magnesium treatment during the transition time in the first days after birth to avoid respiratory morbidity.

Accelerating flows through the ductus arteriosus, tricuspid valve dysplasia/dysfunction, higher spectrum of the fetal pulmonary vein downflow, extended atrial conduction time, could be presented in the course of transient occlusion of the ductus arteriosus and in utero infection [19–30]. So, after the exclusion of the structural congenital heart defects as a base of the dilatation of the main pulmonary artery, we should consider other pathophysiological mechanisms, and neonatologists should be aware of this abnormal finding (Table 2).

Conclusions

The case presented by us draws attention to the value of echocardiographic evaluation in the third trimester of gestation, to better forecast and plan the care of a newborn baby. In the case of a very large dilatation of the pulmonary trunk and mild abnormalities of the fetus' left heart, we have to take into account significant difficulties in the process of physiological adaptation of the newborn to ectopic life. Not every case of disproportion at the level of three vessels of the upper mediastinum in favour of the pulmonary trunk after birth gives the classic picture of aortic coarctation requiring surgical treatment.

Conflict of interest

The authors declare no conflict of interest.

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Division of work

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