DO UMBILICAL CORD WRAPPED AROUND THE FETAL BODY CAN MIMIC SIGNS OF AORTAL COARCTATION?

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Abstract
Objectives: Coarctation of the aorta (CoA) is an irreversible congenital heart defect. Its prenatal diagnosis is not rare a subject to false-positive conclusion. We present a novel hypothesis explaining the basis of this error.

Methods: Ten cases of prenatal suspicion of the coarctation of the aorta (based on disproportion at the level of 4 chamber view and mediastinum) coincided with the umbilical cord wrapped around the fetal body were found in the Filemaker database of the Fetal Cardiology Department. Only single pregnancies were taken into account. In all cases another cardiac and extracardiac malformations were excluded.

Results: The mean maternal age was 29,6 years. The mean gestational age was 33 7/8 weeks. All fetuses were in a good cardiovascular condition. The usual position of the umbilical cord was neck, but they were also location such as nucha, abdomen or lower limb.

At birth, all newborns had normal anatomy of the heart. We conclude that the explanation of the false diagnosis was haemodynamic, resulting from the compression of the fetal neck by the umbilical cord that resulted in a disproportion of cardiac blood flow, “mimicking” CoA.

Conclusions: 1. Functional disturbances can mimic prenatal CoA. 2. Umbilical cord position (specially enlacing the fetus neck) should be taken into consideration in suspected cases of fetal CoA.

Key words: fetal echocardiography, coarctation of the aorta, prenatal diagnosis, perinatal outcome, umbilical cord, pediatric cardiology

INTRODUCTION
Coarctation of the aorta is a pathological narrowing of the aorta that changes flow through this great vessel. It usually affects the segment between the left subclavian artery and the ductus arteriosus. Spectrum of this congenital heart disease (CHD) ranges from only a slight narrowing to a hypoplastic or even an interrupted aortic arch¹. It is a common anomaly. The National Polish Register of Prenatal Cardiac Anomalies reports its 2.5% prevalence in fetuses afflicted with CHD in Poland².

There are two conceptional explanations of the origin of coarctation: ductus tissue theory, explains this anomaly by the migration of smooth muscle cells from the ductus to the aorta, and the hemodynamic theory, which assumes the reduced blood flow through the aorta in fetal life³.

The primary aim of our report was to determine whether physiological alterations of fetal and umbilical circulation have led to this discrepancy of pulmonary artery and aortic sizes resulting in false-positive diagnosis of coarctation.

METHODS
For the purpose of the study the Filemaker database of the Department was searched to find all cases with disproportion of the heart’s chambers coexisting with the umbilical cord enlacing the fetus. The fetus could not have other cardiac nor extracardiac abnormalities.

The images of three vessels view were analyzed. Each had to comprise the isoechochogenic area of the thymus and the mediastinum. Ten cases of prenatal suspicion of the coarctation of the aorta (based on disproportion at the level of 4 chamber view and mediastinum) coincided with the umbilical cord wrapped around the fetal body were found in the Filemaker database of the Fetal Cardiology Department. Only single pregnancies were taken into account. In all cases another cardiac and extracardiac malformations were excluded.

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the measurements of the great vessels in the mediastinum. The pulmonary artery and the aorta diameters were compared and the PA/Ao ratio was calculated for each of fetuses. The size of the aortic isthmus in sagittal plane were collected and the Z-score was found if possible (Fig. 1). The nomograms of Z-scores were obtained by http://fetal.parameterz.com/app website. The PA/Ao > 1.6 and Z-score lower than 1.65 of standard deviation were considered as indicating the coarctation.

**RESULTS**

Since January 2012 to March 2016 ten such cases have been examined. The mean age of the pregnant women was 29.6 years (see Table 1. below). The mean gestational age at the time of the first examination was 33 7/8 weeks. It is worth noting that only one case has been diagnosed in the second trimester, the other observations have been done in the third trimester. All ten fetuses had an efficient cardiovascular system (cardiovascular condition of the fetus number 7. improved in consequent examinations). The cardiovascular condition was described by Cardiovascular Profile Score. The patency of the foramen ovale and of the arterial duct was confirmed.

The photo of the umbilical cord is presented in Fig. 2. The most common location of the umbilical cord was neck of the fetus (8 out of 10). Surprisingly, in two cases in which the cord was wrapped around the abdomen, the observations were similar to these in ‘neck’ cases. Eventually, the fetuses follow-up was checked to see whether

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**Table 1.** Sonographic and echocardiographic features of prenatal MS and current case

<table>
<thead>
<tr>
<th>Patient's age [years]</th>
<th>Gestational age (last menstrual period) [weeks]</th>
<th>Estimated fetal weight (Hadlock et al.[25]) [g]</th>
<th>PA/Ao ratio</th>
<th>Isthmus of the aorta [mm], Z-score</th>
<th>CVPS</th>
<th>The umbilical cord enlacing</th>
<th>Other</th>
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<td>35 5/7</td>
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<td></td>
<td></td>
<td>The neck and thorax</td>
<td>Oligohydramnion</td>
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<td>38</td>
<td>2623</td>
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<td></td>
<td></td>
<td>The neck</td>
<td>Reverse flow in the aortic arch</td>
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<td>The neck</td>
<td>Hyperechogenic bowels</td>
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</table>

**Average** 29.6 33 7/8 2125.9 1.67 9

*Not measured because of the fetal position during the examination
newborns were suffering from any of the cardiac dysfunction, particularly from the coarctation of the aorta. The collected data are found in Table 2. All babies were delivered on time with a mean birth weight 3054.4 g. Female newborns consisted 6 out of 10 cases.

**DISCUSSION**

The problem of high incidence of false-positive diagnoses of the CoA caused difficulties since the beginning of the fetal ultrasonograms. The investigators have been trying to select the best method for the assessment of fetal aorta in each trimester.

Coarctation is suspected in the basic screening of the heart, i.e. by obtaining the four-chamber view, because examination shows that the right side of the heart is larger than the left and the disproportion is also seen in the great vessels in the mediastinum 4.5.6.7. The inference is strengthened if there is disproportion between the pulmonary artery and the aorta with the PA/Ao ratio equal to or larger than 1.6 should alert about CoA8,9. However, these indicators are not absolutely foolproof 10,11. The ultrasonographic diagnosis includes also Z-scores of the ascending aorta and aortic isthmus dimensions. Isthmal to ductal ratio may be helpful in exclusion of the CoA too12,13.

The enlarged right heart structures may be a result of late pregnancy14, restrictive foramen ovale15 and premature fetal closure of the arterial duct16.

Coarctation of aorta is considered to be an irreversible defect. However, there have been several cases of spontaneous regression of the CoA in infants17. How can we explain the absence of the suspected CoA just after a birth? False positive diagnoses of prenatal CoA have been reported several times 18,19, but we noted and are reporting for the first time that the prenatal condition – the umbilical cord enlacing the neck – may mimic CoA. How can the umbilical cord wrapping the fetal neck affect the fetal circulation?

Umbilical cord wrapped around the fetal neck causes disturbances in the blood supply to the cerebrum. In our hypothesis a decreased flow through cerebral
vessels leads to the alternations in ductus venosus functioning and its dilation (Fig. 3.).

The authors believe that the consequence of this condition is an enlargement of the right atrium and ventricle due to the redistribution of the blood, which may be a direct basis of a reduced flow through the aorta (Fig. 4.).

This hypothesis is drawn from the report published by Tchirikov et al. Scientists have found that the ductus venosus/umbilical vein blood flow ratio increases in the state of fetal hypoxia in the lambs, which means that larger part of the umbilical venous blood flow enters the DV during hypoxia, even if the velocity through the DV is decreased.20

The problems with umbilical cord encompass its knots and many morphological abnormalities.21 A true knot of the umbilical cord may lead to e.g. fetal hypoxia, intrauterine growth restriction and even to fetal death. Compression of the umbilical cord may threaten fetal life too, reducing the blood flow through the vein and arteries.22 Fetuses with the true knot of the umbilical cord are at higher risk of fetal distress and meconium stained amniotic fluid and fourfold higher risk of antepartum fetal death.23

Kobayashi et al. focused on the issue of the entanglement of the cord and its consequences. They mention higher frequency of Apgar scores in the 1 minute and 5 minute lower than 7 and umbilical artery pH lower than 7.1 in the group of fetuses whose neck or trunk was enlaced by the cord.24

Although earlier reports associated umbilical cord tightening up with other disturbances, we propose that umbilical cord enlacing the fetus may influence the image derived from an ultrasonographic examination and we suggest a haemodynamic explanation of the disproportion in the prenatal cardiac blood flow. This report is limited only to the investigation of the impact of entanglement of the cord on the fetal hypoxia, although there are several causes of this state, e.g. disturbances in the placental circulation.

Functional disturbances of blood flow may mimic prenatal narrowing of the aorta due to the appearing
disproportion between the heart’s chambers and great vessels. Not each of disproportions will result in diagnosis of the coarctation. Umbilical cord position and specially enlacing neck of the fetus should be ruled out as an alternative explanation, in cases suspected of fetal CoA.

CONCLUSIONS

1. Functional disturbances can mimic prenatal CoA.
2. Umbilical cord position (specially enlacing the fetus neck) should be taken into consideration in suspected cases of fetal CoA.

References

2. Website of The National Polish Register of Prenatal Cardiac Anomalies www.orpkp.pl

Division of work:
K. Więckowska: data search and first draft.
K. Zych-Krekora, M. Stodki: discussion, correction of the manuscript
M. Respondek-Liberska: concept of the research, correction of the paper, final version.

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