Missing ductus venosus: a case report

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Summary

Background: The ductus venosus is a short vessel, present in the newborn infant on the dorsal surface of the liver, connecting the portal and umbilical circulation with the inferior vena cava. Agenesis of the duct is a rare anomaly. *Case:* A 28-year-old woman was referred to our department for the first trimester ultrasound evaluation. Detailed scanning revealed agenesis of the duct. Fetal echocardiography showed cardiac disproportion at the level of the ventricles. *Conclusion:* Agenesis of the duct can be related to either cardiac or congenital abnormalities.

Key words: Ductus venosus and aneuploidy or heart problems; Agenesis of ductus venosus; Ventricular disproportion; Congenital liver abnormalities.

Introduction

The embryological development of ductus venosus affects, to a great extent, the prenatal course and pregnancy outcome. Agenesis of the tiny duct is related to cardiac structural anomalies and various congenital anomalies. An abnormal course of the umbilical vein seems to be the first indication that the duct is probably not there. Some cases have been reported in the past [1].

Case Report

A 28-year-old woman was referred to our fetal medicine clinic in the first trimester (13+1 weeks) for a routine scan evaluation. She was a non-smoker, with a pre-pregnancy weight of 75 kg and a BMI of 26; no allergies or comorbidities were reported. Nuchal translucency (NT) was 1.8 mm, the nasal bone was present and blood flow across the tricuspid valve was normal. Maternal serum biochemistry (free beta hCG: 1.223MoM, PAPP-A: 1.2988MoM) gave her a low risk for Down's syndrome. Detailed fetal ultrasound (US) examination showed a single viable fetus with agenesis of the ductus venosus and an umbilical left iliac vein-inferior vena cava aberrant connection (Figure 1). Fetal echocardiography revealed ventricular disproportion. No hydrops had developed and the portal vein was absent. Fetal karyotyping detected no chromosomal abnormalities. After birth, the newborn's heart on US confirmed ventricular disproportion (the right side larger than the left). During the infant's neonatal period the intracardiac pressure reached the normal values and the heart size became normal.

Discussion

The most common congenital malformations are cardiac defects. The ductus venosus is a small fetal vessel that transfers oxygenated blood originating from the placenta to the fetal heart through the umbilical vein. Ductus venosus shunts a significant majority of the blood flow of the umbilical vein directly to the inferior vena cava. In this way, it allows oxygenated blood from the placenta to bypass the liver, thus playing a critical role in preferentially shunting oxygenated blood to the fetal heart and brain. The amount of shunting is determined by the diameter of the vessel, the pressure gradient and blood viscosity. The blood from the ductus venosus is then directed towards the left atrium with high velocity due to the small diameter of the vessel. This flow bypasses the right atrium, reaches the left atrium through the foramen ovale and enters the ascending aorta towards the coronary and brain vessels [1]. It is one of the three physiological shunts determining blood distribution during intrauterine life and is greatly involved in the regulation of fetal circulation, acting as a sphincter to protect the fetus from placental overcirculation. The volume of its flow is altered according to the pressure gradient between the umbilical vein and the heart. Absence of the ductus venosus leads to direct umbilical venous return into the heart. The prenatal diagnosis of an abnormal waveform on the ductus during first trimester scanning can be portrayed as a sign of an abnormal pregnancy course [2].

The absence of the ductus venosus is usually related to adverse pregnancy outcome [2]. Similarly, the abnormal flow pattern of the ductus is associated to structural, cardiac and chromosomal defects of the embryo in both singletons and multiple pregnancies [3]. The ductus venosus normal waveform is altered during the second trimester, mainly due to severe intrauterine growth restriction, twin-to-twin syndrome, heart defects and cardiac vessel abnormalities [4]. Today, the assessment of ductus venosus flow is proposed as an integral part of the 11-13-week scan. Ductus venosus flow assessment improves the performance of screening by NT thickness and serum PAPP-A and free -hCG [3, 4].

A negative a-wave on the ductus venosus waveform in combination with a backflow along the tricuspid valve can be a reliable indicator of serious cardiac abnormalities, i.e., ventricular disproportion and unfavorable fetal outcome [5]. Fetuses with no ductus venosus should be carefully scanned for any additional anatomical abnormalities [6]. Moreover, techniques such as two-phase helical computed tomography, magnetic resonance (MR) imaging and MR angiography can shed light on the vascular anomaly of ductus venosus [7].

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Figure 1. — Umbilical vein-left iliac vein-inferior vena cava aberrant connection.

In the early second trimester of pregnancy, fetal cardiac malformations can be detected by the ductus venosus Doppler waveform and the four-chamber view [8]. Going back in the past, sonographical detection of the nasal bone was part of first trimester prenatal testing, while in the second trimester the nasal bone length was put into consideration [9].

During the first trimester, the ductus venosus can be easily imaged with color Doppler and its flow waveform can be visualized by pulsed Doppler. It is identified as the part of the vessel bearing the highest blood velocity, following the umbilical vein. Ductus venosus has a typical waveform with three phases: a) the highest velocity peak that corresponds to the ventricular systole while the pressure gradient between the umbilical vein and the atrium is the highest; b) the peak of forward flow that corresponds to early diastole, throughout the opening of the atrioventricular valves and early passive filling of the ventricles; and c) the lowest velocity that corresponds to the atrial contraction, during late diastole. In this last phase the atrial pressure is high and there is a low pressure gradient [10, 11].

Fetuses with abnormal vascular connections in the liver parenchyma require detailed scanning and evaluation [12]. The number of cases of ductus venosus agenesis with a normal or abnormal course of the umbilical vein have been found postmortem, postnatally or prenatally [1].

Absence of the ductus venosus vessel is a very rare pathological finding, which may be compatible with normal fetal development. Non visualization of the vessel's anatomical position combined with increased amniotic fluid, an abnormal heart structure and a deviated umbilical vein during scanning suggests non formation of the ductus. Careful US assessment of the ductus venosus and the umbilical vein should be performed on every fetus with unexplained cardiomegaly, polyhydramnios, ascites or hydrops [13-15].

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