Congenital Ductus Arteriosus Aneurysm Diagnosed at 38 Weeks of Gestation

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Abstract

A 25-year-old pregnant woman (G2P1) was diagnosed with fetal ductus arteriosus aneurysm (DAA) at 38 weeks of gestation. Patients have been referred to our hospital with the diagnosis of oligohydramnios. She did not have any medical or surgical history. Her diabetes and hypertension were well controlled with oral hypoglycemic agents. A 25-year-old pregnant woman (G2P1) was referred to our hospital with the diagnosis of neonatal period. In the same year, Mielke et al. reported a case of an abnormally S-shaped kinking of the ductus arteriosus with increased systolic and diastolic Doppler flow velocities. Since then, several case reports and a retrospective study have been reported.

DAA likely develops especially in the third trimester perhaps due to abnormal intimal cushion formation or elastin expression. It can be potentially fatal because of its association with severe complications. It is characterized by a fusiform and saccular dilatation of the ductus arteriosus (DA). Although the true incidence of congenital DAA is still unknown, recent reports suggest it may be much more common than previously thought.

Although imaging of the ductus arteriosus (DA) by ultrasonography has been possible for many years, it is only since 1995 that DAA has been reported using fetal echocardiography. In 1995, Puder et al. described the first case of DAA detected at 39 weeks of gestation and it spontaneously closed during the neonatal period. In the same year, Mielke et al. reported a case of an abnormally S-shaped kinking of the ductus arteriosus with increased systolic and diastolic Doppler flow velocities. Since then, several case reports and a retrospective study have been reported.

Introduction

Ductus arteriosus aneurysm (DAA) is characterized by a fusiform and saccular dilatation of the ductus arteriosus (DA). Although the true incidence of congenital DAA is still unknown, recent reports suggest it may be much more common than previously thought.

Although imaging of the ductus arteriosus (DA) by ultrasonography has been possible for many years, it is only since 1995 that DAA has been reported using fetal echocardiography. In 1995, Puder et al. described the first case of DAA detected at 39 weeks of gestation and it spontaneously closed during the neonatal period. In the same year, Mielke et al. reported a case of an abnormally S-shaped kinking of the ductus arteriosus with increased systolic and diastolic Doppler flow velocities. Since then, several case reports and a retrospective study have been reported.

Keywords:
Ductus arteriosus aneurysm; Obstetrics; Fetal echocardiography

Case Report

A 25-year-old pregnant woman (gravida 2, para 1) have been referred to our hospital with the diagnosis of oligohydramnios. She did not have any medical or surgical condition and there were no obstetrical complications. Both nuchal translucency and quad tests were all within the normal limits. A fetal US examination was performed to check fetal growth and measure the amniotic fluid index. At that time, a fusiform dilatation of the DA was incidentally detected. Fetal echocardiography was performed with a Mindray DC-7 scanner equipped with transabdominal 4-8 MHz curvilinear probe and the DAA was confirmed on the three-vessel view of the fetal heart (Figure 1). Its diameter was 12 mm.
Figure 1: Ductus arteriosus aneurysm detected in three vessel view.

Since the original description of the three-vessel view, there have been other reports confirming its usefulness and expanding its diagnostic role in fetal echocardiography. With slight angulation of the transducer more anatomical details are demonstrated, including the transverse aorta, ductus, superior vena cava and trachea in the three vessels and trachea view [9,10].

Figure 2: Ductus arteriosus aneurysm detected in sagittal cross-sectional image.

The sagittal view of the ductal arch demonstrated a dilated DA at the distal end (Figure 2).

On the spectral Doppler analysis, both the systolic and the diastolic flow velocities of the DAA were within the normal limits.

Figure 3: Turbulent flow in the DAA.

Turbulent flow in the DAA was detected when using color Doppler (Figure 3). There were not any other intracardiac abnormality.

Cesarean section has been decided because of fetal distress findings in non-stress test. The asymptomatic male newborn weighted 3300 g with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. Clinical examination was normal. No other cardiac abnormalities were detected. The DAA was confirmed by postnatal echocardiography within 24 hours after birth showed a PDA with a particularly tortuous course, and a restrictive pulmonary artery end was noted with partial involution of the aneurysmal DA. Flow was left to right. Three days after birth the mother and baby were discharged in a healthy way.

Discussion

Congenital ductus arteriosus aneurysm has been considered a rare cardiovascular lesion. Although the incidence of neonatal DAA was previously reported to be 0.8%, the true incidence is unclear, depending on the criteria of diagnosis [11]. In a previous study, there were three cases of congenital DAA in a review of 200 consecutive third-trimester US examinations (1.5%) [12]. In another longitudinal series that included 509 fetuses and more that 3000 US examinations, the prevalence of congenital DAA was 2.2% [13].

DAA has been uniformly identified in the third trimester and has not been detected earlier in gestation. This suggests a late onset of prenatal development may be the result of abnormal intimal cushion formation or defective elastin in ductus arteriosus [5]. DAA may be observed in the patients with connective tissue diseases such as Marfan, Ehler-Danlos and Larsen syndromes [14]. Previous studies showed the association of trisomy 21, 13 or Smith-Lemli-Opitz syndrome with DAA [15]. High risk factors associated with DAA include newborns with large for gestational age, maternal DM and mothers with blood group A [6].

Three-vessel view is helpful on diagnosis of DAA [16]. In our case, the DAA was confirmed by the three-vessel view on the fetal echocardiography, although it was incidentally detected during routine US evaluation of the fetal growth and the amniotic fluid index.

Although the majority of affected neonates are clinically asymptomatic, several reports demonstrated potential serious complications including spontaneous rupture, thromboembolism, erosion into airways, infection, and compression of surrounding vessels, airways and nerves [17]. Lund et al. cite a 31% complication rate in their earlier review of 65 cases of DAA previously diagnosed in children under 2 months of age, although it is rarely fatal [18]. It may also be associated with chromosomal anomalies and, more importantly, connective tissue disorders which may be progressive and place the infant at higher risk of spontaneous aneurysm rupture [19].

To avoid such lethal complications, surgical intervention may be considered if

1. The ductus arteriosus with DAA remains patent beyond the neonatal period,
2. The DAA is associated with connective tissue disease,
3. There is evidence of thrombus extension into other vessels or thromboembolism or
4. There is significant compression of adjacent structures [20].

Although it does not appear to warrant intervention in the majority of cases, it would be wise to document complete closure of the aneurysmal duct prior to neonatal discharge. Like most cases, the case presented here was asymptomatic, closed spontaneously and did not require treatment.

In addition to surgical considerations, close follow-up of the neonate is important because the majority of cases may spontaneously resolve after a period of time. The use of indomethacin has been reported to close DAA successfully in one case report; however the definitive effect of it on DAA is uncertain [21].

In summary, increased use of fetal echocardiography has detected more cases of congenital DAA. The majority of affected neonates are clinically asymptomatic and tend to progressively diminish in size of DAA and spontaneous closure, especially in cases of smaller DAA [22]. However, serious complications can occur during waiting periods indicating surgical intervention, especially in cases of persisting DAA beyond the neonatal period or associated connective tissue diseases. Finally, indomethacin treatment may be used to successfully close DAA in premature infants even though its definitive effect on DAA is still uncertain [20].

References