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Coexistence of Ductal Constriction and Closure of the Foramen Ovale In Utero

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Abstract. We report a fetus with an unusual combination of a narrow ductus arteriosus (DA) and foramen ovale. A pregnant mother was referred at 26 weeks of gestation for fetal pericardial effusion. Fetal echocardiography showed pericardial effusion, right atrial enlargement, right ventricular hypertrophy, and tricuspid regurgitation. The DA looked tortuous with S-shaped kinking. The atrial septum primum bulged into the left atrium. Color Doppler did not show any flow across the atrial septum. Cesarean section was performed at 31 weeks of gestation. Admission to intensive care was required after delivery, but the infant gradually improved and was discharged home without any sequela.

Keywords: Premature closure of ductus arteriosus — Premature closure of foramen ovale — Fetal pericardial effusion — Fetal congestive heart failure

In the normal fetal circulation, patency of the ductus arteriosus (DA) and the foramen ovale is vital for right-to-left shunting to bypass the pulmonary circuit. Constriction of the DA *in utero* may be an ominous event and has often caused congestive heart failure, fetal hydrops, and even fetal or neonatal death [7]. Ductal stenosis *in utero* unrelated to prostaglandin inhibitors is rare [5]. Prenatal restriction of the foramen ovale is a cardiac abnormality that rarely occurs with right heart failure, which also results in serious sequelae to the fetus [2, 12]. There are no reports of a fetus with both ductal constriction and premature closure of the foramen ovale *in utero*.

Case Report

A 35-year-old woman, gravida 4, para 2, was referred at 26 weeks of gestation for fetal pericardial effusion. Obstetric ultrasound examination showed normal fetal growth and amniotic fluid index, no other organ anomalies, a moderate pericardial effusion, and a small amount of ascites without hydrops fetalis (Fig. 1). On Doppler examination, the systolic/diastolic ratio of the fetal umbilical artery and middle cerebral artery was within normal limits. There was no history of medication during pregnancy, including antiinflammatory agents.

Fetal echocardiography showed a moderate pericardial effusion. The right ventricle was dilated and hypertrophied, with a decrease in shortening fraction. The right atrium was also dilated. In contrast, the left atrium and left ventricle were small. A moderate degree of tricuspid regurgitation (peak velocity, 4.1 m/sec) was noted. The DA appeared to be constricted and tortuous with S-shaped kinking. Ductal velocity was at least 3.0 m/sec at systolic phase and 0.44 m/sec at end-diastolic phase (Fig. 2). The pulsatility index was 2.32. The atrial septum primum bulged into the left atrium without any phasic motion and looked like an atrial septal aneurysm (Fig. 3). Color-flow mapping did not demonstrate normal right-to-left flow across the atrial septum.

It was decided to continue the pregnancy under close surveillance. Pharmacologic agents to dilate the DA were not given because of their compounding effect on the smooth muscle. At 31 weeks of gestation, a cesarean section was performed due to fetal distress. A male infant weighing 1.8 kg was delivered with a Apgar score of 1 at both 1 minute and 5 minutes. Intensive cardiopulmonary supportive care was given upon delivery. Postnatal echocardiogram showed that the DA was still tortuous but without significant stenosis. On Doppler examination, the bidirectional flow with low velocities across the DA was shown (Fig. 4). The septum primum bulged into the left atrium with little phasic motion, without any flow across the atrial septum on color-flow mapping.

Follow-up echocardiographic examination revealed a spontaneous closure of the DA, regression of right ventricular hypertrophy with functional improvement, and marked decreases in the amount of pericardial effusion and the severity of tricuspid regurgitation. The infant gradually improved and was weaned from the ventilator after 1 week but thereafter suffered from sepsis and jaundice. He was discharged home at 5 weeks of age. After discharge, he did well and gained weight. An echocardiogram performed at 4 months of age showed no abnormalities.

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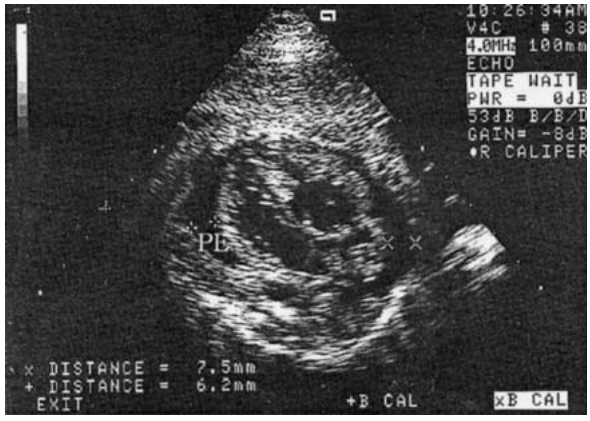

Fig. 1. Two-dimensional fetal echocardiogram showing a moderate amount of pericardial effusion (*PE*).

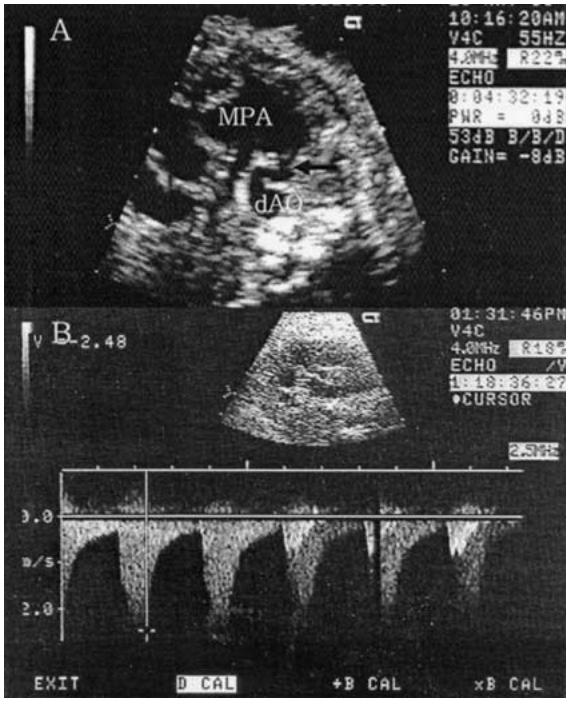

Fig. 2. (**A**) Two-dimensional fetal echocardiogram showing the S-shaped kinking of the ductus arteriosus with constriction. (**B**) Continuous-waveDoppler flow showing a peak velocity of 2.48 m/sec at the ductus arteriosus. *dAO*, descending aorta; *MPA*, main pulmonary artery.

Discussion

Chao et al. [1] reported the first case of intrauterine closure of the DA detected by Doppler echocardiography, which revealed markedly decreased blood flow through the tricuspid and pulmonary valves and no blood flow at the DA. Color-flow mapping reaffirmed these findings as well as the resultant increased

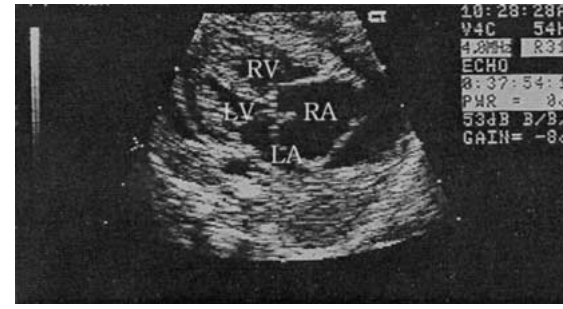

Fig. 3. Two-dimensional echocardiogram showing aneurysmal dilatation and bulging of the atrial septum into the left atrium without any interatrial communication. *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

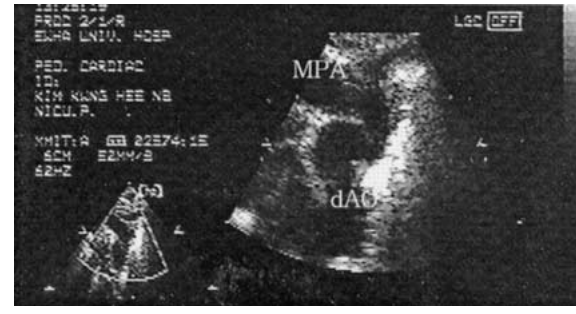

Fig. 4. Postnatal echocardiogram showing the S-shaped tortuous patent ductus arteriosus. *dAO*, descending aorta; *MPA*, main pulmonary artery.

right-to-left shunting across the foramen ovale. In five cases of ductal closure *in utero*, Leal et al. [6] found no flow in the DA, a dilated right ventricle with decreased function, and mild or moderate tricuspid and pulmonary insufficiency. Ductal closure results in blood flow redistribution characterized by reduced blood flow through the right heart and increased right-to-left shunting across the dilated foramen ovale [9]. Mielke et al. [8] reported an abnormally S-shaped kinking of the fetal DA with increased systolic and diastolic flow velocities combined with right heart dilatation and severe tricuspid valve insufficiency. These findings are very similar to those observed in our case. Mielke et al. did not describe the size of foramen ovale, but they thought that elevated peak systolic flow velocity in the ascending aorta may have been caused by increased left ventricular output due to right-to-left shunting across the foramen ovale. In our case, the foramen ovale appeared to be almost completely obstructed by an aneurysmal septum primum that bulged into the left atrium. This is unusual in the premature ductal closure because the right atrial pressure is higher than the left atrial pressure.

Isolated premature restriction of the foramen ovale *in utero* results in diminished foramen ovale

blood flow and venous congestion, which is associated with right-sided heart failure, causing pericardial effusion, arrhythmia, hydrops fetalis, and often death [2, 12]. Closure or restriction of the foramen ovale may occur primary or secondary to preexisting cardiac disease associated with elevated left atrial pressure, such as mitral obstruction, hypoplastic left heart syndrome, and coarctation of the aorta [3, 10]. In the primary restrictive foramen ovale, the atrial aneurymal structure consistently bulges into the left atrium [3], whereas in the presence of left ventricular or mitral flow obstruction, the aneurysm may bulge into the right atrium [10]. When the foramen ovale is closed *in utero*, almost all of the cardiac output must pass through the right ventricle, which results in increased blood flow into the right ventricle and the DA and reduced flow to the left side of the heart. Any resultant hemodynamically mediated morphologic abnormalities would theoretically depend on the time of onset and the severity of the narrowing of the foramen ovale [11].

In our case, it is not certain why the DA became narrow and tortuous, but the ductal narrowing seemed to be reactive. Although the DA had been narrow during fetal life, an immediate postnatal echocardiogram showed a tortuous but widened DA, which closed in a normal way after birth. Moreover, during fetal life, the peak velocity of tricuspid regurgitation was approximately 4.0 m/sec, indicating that the estimated right ventricular pressure was at least 64 mmHg. An unknown ductal constrictor or increased ductal muscular reactivity might be hypothesized as a cause.

If ductal constriction or premature closure of the foramen ovale are confirmed antenatally and a fetus is viable, prompt delivery appears to be the most appropriate course of action, but it should be based on several factors, including gestational age, fetal karyotype, the presence of other anatomic abnormalities or fetal hydrops, and the results of antepartum surveillance tests [4, 10]. In case of prematurity, the infant may suffer from hyaline membrane disease, but hemodynamics become stable after birth. Therefore, the time of delivery must be determined after careful consideration and collaboration between obstetricians and pediatric cardiologists.

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