

Idiopathic Constriction of the Fetal Ductus Arteriosus with Right Ventricular Failure; Rapid Resolution after Birth

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Premature constriction of the ductus arteriosus is rare, but it can occur during fetal life idiopathically or secondary to medications or structural lesions. Premature constriction of the ductus arteriosus can lead to progressive right heart dysfunction, heart failure, subsequent hydrops fetalis, and even fetal death. Herein, we describe a case of fetal ductus arteriosus constriction of unknown etiology with a severely enlarged and hypertrophied right ventricle, which resolved dramatically soon after birth.

Key Words : Ductus arteriosus, Prenatal diagnosis, Constriction, Pathologic

Premature constriction of the ductus arteriosus (DA) is a rare phenomenon. The true incidence might be larger than estimated because some cases have a subclinical or mild course. In only a few cases will ductal dysfunction come to the attention of the clinician.^{1,2} In other cases, a constriction of the DA occurs and can lead to cardiovascular dysfunction such as tricuspid valve regurgitation, pulmonary valve regurgitation, right heart failure, pulmonary hypertension, hydrops fetalis, and even fetal death. In the past, diagnosis of premature constriction of the DA was made eventually at autopsy.³ However, premature constriction of the DA can be diagnosed before delivery owing to the advancement of fetal echocardiography nowadays. Herein, we describe a case of fetal DA constriction of unknown etiology with a severely enlarged and hypertrophied right ventricle,

which resolved dramatically soon after birth.

Case Report

A 30-year-old woman, gravida 1, para 1, was admitted to Seoul National University hospital at 37 weeks and 1 day gestation for fetal surveillance. She had received prenatal care at the local hospital and was referred to our hospital owing to right heart dilatation on fetal ultrasonography. No personal or familial history of congenital heart disease was reported. She denied use of indomethacin or other prostaglandin inhibitors. Fetal echocardiogram at a gestational age of 37 weeks and 2 days showed that the right ventricle was severely enlarged and hypertrophied. A Doppler study revealed a prematurely constricted DA with continuous flow and a narrowest diameter of 1.3 mm and length of 6 mm (Fig. 1). The fetus also showed a moderate degree of tricuspid regurgitation with peak velocity of 4.4 m/sec and minimal pericardial effusion. No additional abnormalities were observed on fetal echocardiography.

After discussion, we decided to deliver this fetus; a

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3,100 g male neonate was born by vaginal delivery at 37 weeks and 3 days gestation. His Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. He was transferred to neonatal intensive care unit (NICU) for evaluation and close observation. His initial vital signs were stable with pulse oxygen saturation (SpO_2) consistently above 90% without any oxygen supply. He urinated well throughout the first day of life. His initial capillary blood gas analysis in NICU showed no significant acidosis or hypercapnea. His initial creatinine phosphokinase was 368 IU/L and

lactate dehydrogenase was 362 IU/L and there was no evidence of birth asphyxia. A postnatal chest radiograph showed severe cardiomegaly (Fig. 2A). On the postnatal transthoracic echocardiogram 3 hour after birth, the 2-dimensional images at birth revealed a severely enlarged, hypertrophied right ventricle with visually decreased contractility (tricuspid annular plane systolic excursion of only 4.9 mm) and a relatively small left ventricle on 4 chamber view with ejection fraction of 46.8% (Fig. 3A). The DA was nearly closed at this time. He started oral feeding

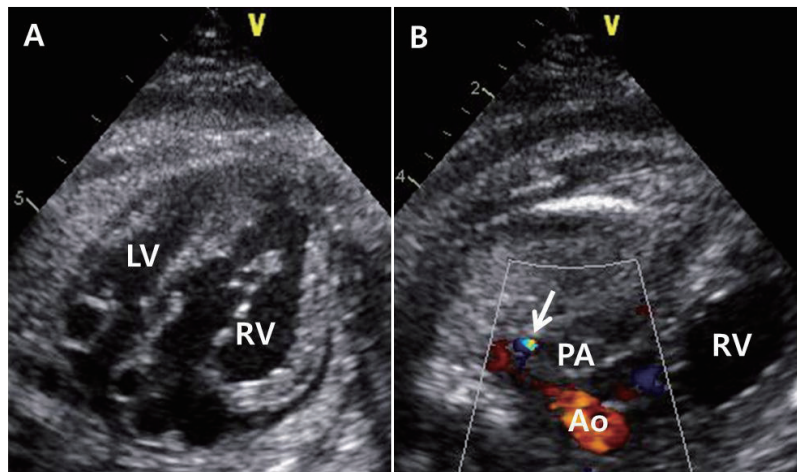


Fig. 1. Fetal echocardiogram at gestational age of 37 weeks and 2 days. A: The 2 dimensional image shows a severely enlarged and hypertrophied right ventricle on 4 chamber view. B: Color Doppler reveals prematurely constricted ductus arteriosus with a narrowest diameter of 1.3 mm and length of 6 mm (arrow). RV: right ventricle, LV: left ventricle, PA: pulmonary artery, Ao: aorta.

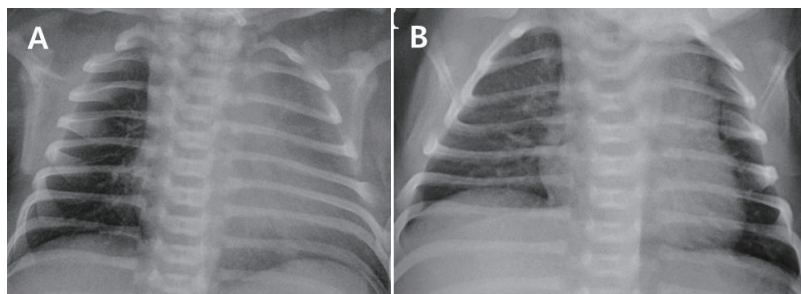


Fig. 2. Chest posterior-anterior (PA) radiograph. A: Chest PA radiograph at birth shows severe cardiomegaly with a cardiothoracic (CT) ratio of 66.2%. B: Chest PA radiograph at 2 weeks after birth shows that the heart shadow has regressed to normal with a CT ratio of 46.7%.

from the first day of life. A follow-up echocardiogram 2 days after birth showed that the contractility of the right ventricle had rapidly improved. However, the left ventricle showed decreased contractility (ejection fraction of 38.3%; Fig. 3B). As the neonate did not have any symptoms or signs of heart failure, we did not treat him with any supportive heart medication or prostaglandin. A follow-up echocardiogram 6 days after birth showed improved left ventricular contractility (ejection fraction of 53.1%) that had spontaneously become normal. The neonate was discharged 8 days after birth in good condition without any medication. At 2 weeks after birth, he was doing well and a chest radiograph showed a normal cardiac shadow (Fig. 2B). At 1 month of age, the right ventricle was normal with good function of both ventricles on 4 chamber view (Fig. 3C). There was no significant tricuspid regurgitation and the left ventricular ejection fraction was 63% at this time.

Discussion

In this neonate with right ventricular remodeling after premature constriction of the fetal DA, we observed dramatic improvement soon after birth. This improvement suggests that right ventricular remodeling

was reversible. In the fetal circulation, the DA allows most of the oxygenated right ventricular output to bypass the high-resistance pulmonary vascular system and flow to the systemic circulation, acting as a shunt connecting the pulmonary artery to the aortic arch.² Patency of the DA is thought to be an active process that is regulated by a balance between the opposing actions of oxygen and prostaglandin E.⁴ The stimulus for closure of the mature DA is exposure to the higher arterial oxygen content predominating in postnatal life. Closure is mainly caused by contraction of the muscular media, which consists mostly of muscle fibers that are oriented circumferentially. Whereas circumferential muscle fibers serve to narrow the lumen, the relatively few longitudinal fibers complete the closure by shortening the DA.²

Premature constriction of the fetal DA will result in an instant increase in afterload of the right ventricle, leading to progressive right ventricular dysfunction with hypertrophy, dysfunction, dilation, tricuspid valve regurgitation, papillary muscle stress, and ischemia.^{1, 5, 6} An increase in pulmonary trunk pressure may change blood flow in the high-resistance, fluid-filled lungs.⁷ The intra-uterine increase in pressure leads to hypertrophy of the media, causing pulmonary hypertension and postnatal persistent pulmonary hyper-

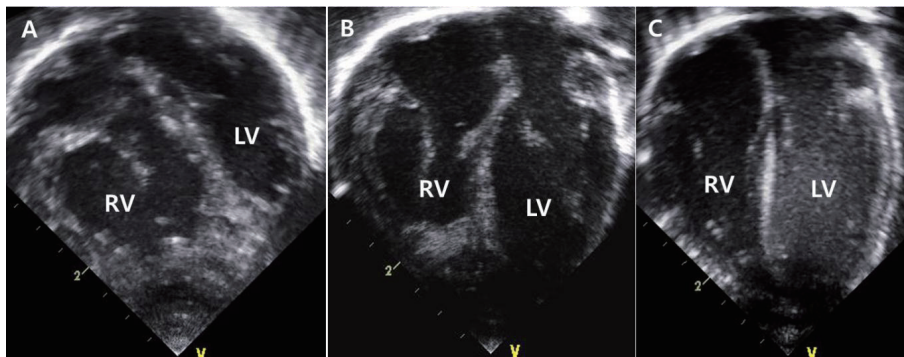


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tension of the neonate.⁸ After birth, these problems may cause inadequate pulmonary blood flow and atrial right to left shunting, and thus, can lead to significant neonatal hypoxia and cyanosis. But in this case the infant had no significant comorbidity after birth.

Non-steroid anti-inflammatory drugs are known to cause prenatal constriction of the DA, and many other medications, such as lithium, are suspected of causing constriction.^{9,10} In Korea, a case of prenatal constriction of the DA following maternal ingestion of non-steroid anti-inflammatory drug was reported.¹¹ And some foods have recently been associated with constriction of the DA.¹² However, most cases remain idiopathic because the causes of prenatal constriction of the duct are not clearly known. In our case, the mother denied the use of indomethacin and other medications known to cause constriction of the duct. As there was no other identifiable cause, this case remains idiopathic. There has been one case report of idiopathic ductal constriction in Korea with normalized echocardiographic finding 7 months after birth.¹³

According to previous case reports, fetuses with ductal constriction are usually managed by immediate delivery unless they are too premature, and their post-natal prognosis seems to be good. And a fetus without distress can be followed up rather than delivered.¹³ However, if a fetus in distress is too premature, a decision on delivery should balance fetal risk and post-natal risk.

Conclusion

In summary, we reported a case of premature fetal DA constriction of unknown etiology with a severely enlarged and hypertrophied right ventricle, which dramatically resolved within 1 month after birth. In the case of significant right ventricular hypertrophy

with dysfunction in the third trimester, clinicians should evaluate the shape and flow of the DA. If there is premature constriction of the DA with hemodynamic compromise in a fetus, immediate delivery of the fetus is a treatment for the recovery of heart function.

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= 국 문 초 록 =

출생 후 빠르게 호전된 특발성 태아동맥관 수축과 동반된 우심실 부전

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이두리 · 안경진 · 김기범 · 권보상 · 배은정 · 노정일

태아기에 발생하는 동맥관 조기수축은 그 빈도가 드물지만 약물이나 구조적인 병변에 의해 2차적으로 발생하거나 또는 특발성으로 발생할 수 있다. 동맥관 조기수축은 우심방기능저하, 우심방부전으로 이어져 태아수종이나 심지어 태아 사망까지 이어질 수 있다. 본 증례에서는 자궁내에서는 동맥관 조기수축이 우심방비대로 이어졌으나 출생 후 극적으로 호전된 증례를 보고한다.

중심 단어 : 동맥관, 산전 진단, 조기 수축