BRIEF COMMUNICATION

High-definition Power Doppler Ultrasound Facilitates Prenatal Diagnosis of Interrupted Aortic Arch

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KEY WORDS
dilated pulmonary artery,
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ventricular septal defect

Diagnosing interrupted aortic arch in the fetus may be challenging. In most cases, all four chambers are present. Crossing of the outflow tracts is seen and the fetal scan can be mistaken for normal. High-definition power Doppler ultrasound apparently can facilitate visualization and detailed examination of the anatomical features of the interrupted aortic arch types in our present case.

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Brief Communication

We present a 30-year-old, Gravida 2 Para 1, woman who was referred to our prenatal diagnosis clinic at 26 weeks of gestation with a high risk of neural tube defect (1 in 91) and low risk for a second trimester biochemical screening test for Down syndrome. She reported an unremarkable past health and family history. Under three-vessel and left ventricle outflow tract view of fetal echocardiography, we detected a dilated pulmonary artery, a disproportionately dilated right atrium, a large subaortic ventricular septal defect (VSD), and a narrowing ascending aorta. High-definition power Doppler ultrasound identified only one defined arterial branch (possibly the innominate artery) arising from the ascending aorta (Fig. 1A) and another artery (possibly the left subclavian artery) arising from the descending aorta. A reverse flow was identified from the lower portion of the descending aorta to the aortic arch (Fig. 1B). After extensive counseling, the patient decided to keep pregnancy. The subsequent prenatal surveys were
uneventful. At 33 weeks of gestation, the patient received repetitive cesarean section because of preterm premature rupture of membranes and previous cesarean section. A male baby weighted 1,640 g was delivered, and the Apgar score was 5 and 7 at 1 minute and 5 minutes, respectively. The postnatal karyotype was normal and there was no DiGeorge syndrome (22q11 microdeletion). Prostaglandin E1 was given to maintain the patency of patent ductus arteriosus. An angiography was performed, which revealed type B interrupted aortic arch (IAA) (Figs. 2 and 3). Then, an operation was arranged. Type B IAA aortic arch reconstruction with direct end-to-side anastomosis between the descending aorta and the ascending aorta, along with patch repair of VSD with a Gore-tex patch and patent ductus arteriosus division were performed. Unfortunately, the symptoms/signs of heart failure were gradually present.

Fig. 1. Fetal echocardiographic images in this case showing (A) a normal four-chamber view; (B) and (C) another view of separation of the AAo, with only one defined arterial branch (the IA) arising from the ascending aorta; (D) the LSA (green arrow), which was separated from the ductal arch with a reverse flow from the DaO (detected by high-definition power Doppler ultrasound). AAo = ascending aorta; Dao = descending aorta; IA = innominate artery; LA = left atrium; LSA = left subclavian artery; LV = left ventricle; RA = right atrium; RV = right ventricle.

Fig. 2. The angiography showing (A) only the LSA coming from the aorta (long arrow) and balloon occlusion in the DaO; (B) the distal portion of interruption of the aorta (short arrow). DaO = descending aorta; LSA = left subclavian artery.
Although surgical repair was performed and extracorporeal membrane oxygenation was used, the patient expired on his 58th day.

IAA is defined as a lack of luminal continuity between the ascending and descending thoracic aorta. This discontinuity may be complete or may be spanned by an atretic fibrous band [1]. The condition is extremely rare, representing less than 1.5% of congenital heart disease cases [2]. This vascular anomaly was initially described in 1778 by Steidele [3] and was first surgically repaired in 1954 [4]. IAA is associated with a mortality rate of more than 90% at 1 year of age if untreated [4]. Rarely, patients with IAA can present in adulthood because of the presence of unusual collateral vessels [1,5]. Therefore, prenatal diagnosis is very important.

Nevertheless, diagnosing IAA in the fetus can be challenging. In most cases, all four chambers are present. Crossing of the outflow tracts is seen, and the fetal scan can be mistaken for normal. A number of anatomic features can facilitate the diagnosis. The most prominent anatomic hallmarks of IAA in the present series were a small aortic valve with a normal-size left ventricle, mildly enlarged right-sided structures, small aortic valve/pulmonary valve and ascending aorta/pulmonary artery diameter ratios, and the universal presence of a VSD, which always has been identified correctly as a posterior malalignment defect in cases of IAA Type B [6]. Moreover, a newer Doppler sonographic technique, known as high-definition flow imaging, was introduced [7]. This technique has the potential benefits of better axial resolution, fewer blooming artifacts, and improved sensitivity to small vessels, when compared with power Doppler [8]. In our case, we successfully identified IAA from other congenital heart abnormalities and clearly visualized the origin and course of the arterial branch by high-definition power Doppler ultrasound. In conclusion, the feasibility of prenatal characterization of IAA and its different types based on two-dimensional echocardiographic examination can be challenging and can present some limitations in thorough assessment. High-definition power Doppler ultrasound apparently can facilitate visualization and detailed examination of the anatomical features of the IAA types, including visualization of the neck vessels, thus supplying additional information with respect to two-dimensional sonography.

References