Antenatal diagnosis of postductal coarctation of the aorta A Case Report

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SUMMARY: Öztunç F, Eroğlu AG, Aksoy F, Saltık İL, Turan A. Antenatal diagnosis of postductal coarctation of the aorta: a case report. Turk J Pediatr 2001; 43: 67-69.

Fetal echocardiography can be used to detect congenital heart disease prenatally with a high degree of accuracy, and complex heart malformations have also been clearly described in the fetus. However, it is difficult to diagnose correctly or to exclude definitely aortic coarctation by fetal echocardiography. A 23-year-old woman was referred for fetal echocardiographic examination at 21 weeks' gestation after discovery of hydrops fetalis (nonimmune) on an obstetric ultrasound examination. Aortic isthmus appeared hypoplastic with a diameter ≤3rd percentile for gestational age. There was a narrowing within the descending aorta immediately distal to the origin of the ductus arteriosus. Color flow imaging demonstrated acceleration and turbulent flow and the peak pressure gradient was measured 83 mmHg by continuous wave Doppler in the same area. The pregnancy terminated in spontaneous abortion at 22 weeks' gestation. The fetus was stillborn. The autopsy findings confirmed the prenatal diagnosis. We conclude that together with the quantitative estimation of the aortic arch, color Doppler and continuous wave Doppler are helpful in diagnosis and estimation of the pressure gradient.

Key words: echocardiography, fetus, coarctation.

Fetal echocardiography can be used to detect congenital heart disease prenatally with a high degree of accuracy, and complex heart malformations have also been clearly described in the fetus[1]. However, a growing body of evidence has shown that it is difficult to diagnose correctly or to exclude definitely aortic coarctation by fetal echocardiography, even though there might be a high index of suspicion for the diagnosis in the presence of ventricular diameter imbalance or aortic arch hypoplasia [2-5].

We present the in utero echocardiographic features of a patient with severe postductal coarctation of the aorta and nonimmune hydrops.

Case Report

A 23-year-old woman, gravida 4, para 1, was referred for fetal echocardiographic examination at 21 weeks' gestation after discovery of hydrops fetalis (nonimmune) on an obstetric ultrasound examination. The first and second pregnancy had terminated spontaneously and both fetuses had been stillborn. Fetal echocardiography during those pregnancies and postmortem examinations had not been done. The fetal echocardiography in our case was performed with a 3.5 MHz transducer interfaced with an Acuson Sequoia C 256 ultrasound system. The fetus was found to have hydrops with marked ascites and pericardial effusion (Fig. 1). The right ventricle/left ventricle ratio was normal and the pulmonary artery/aorta ratio was above the normal range. The ascending and descending aortae were normal in size but the aortic isthmus appeared hypoplastic with a diameter ≤ 3rd percentile for gestational age (Fig. 2). A contraductal shelf was not visible. There was a narrowing within the descending aorta immediately distal to the origin of the ductus arteriosus. Color-flow imaging demonstrated acceleration and turbulent flow (Fig. 3), and the

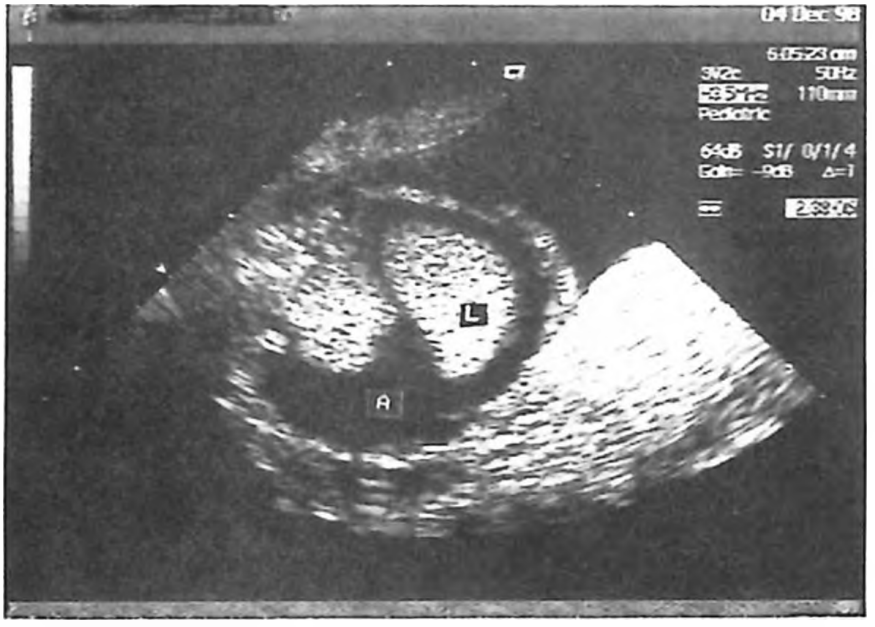

Fig. 1. Horizontal section across the fetal abdomen shows marked ascites. A: ascites; L: liver; S: spleen.

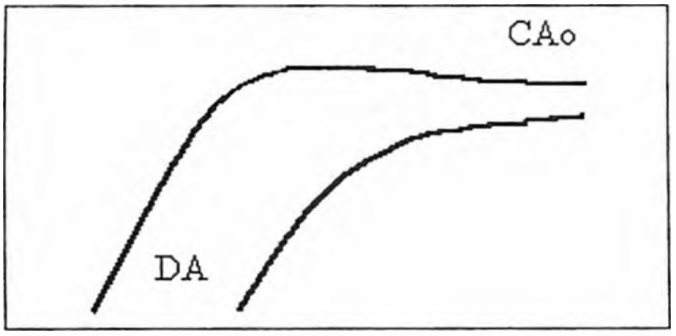

Fig. 2a. Color Doppler flow mapping shows turbulent flow in the coarctation area. CAo: coarctation of the aorta: DA: descending aorta.

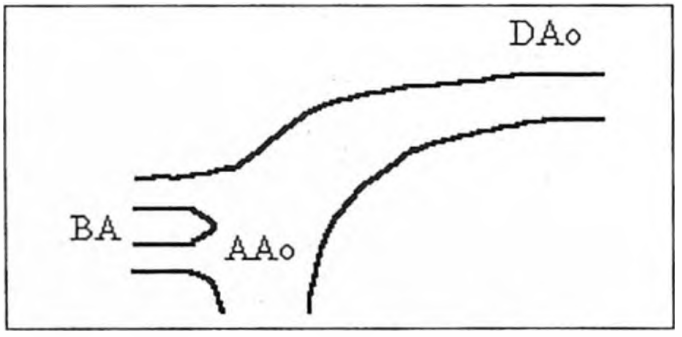

Fig. 2b. Image of dilated brachiocephalic arteries. AAo: ascending aorta: BA: brachiocephalic arteries; DAo: descending aorta.

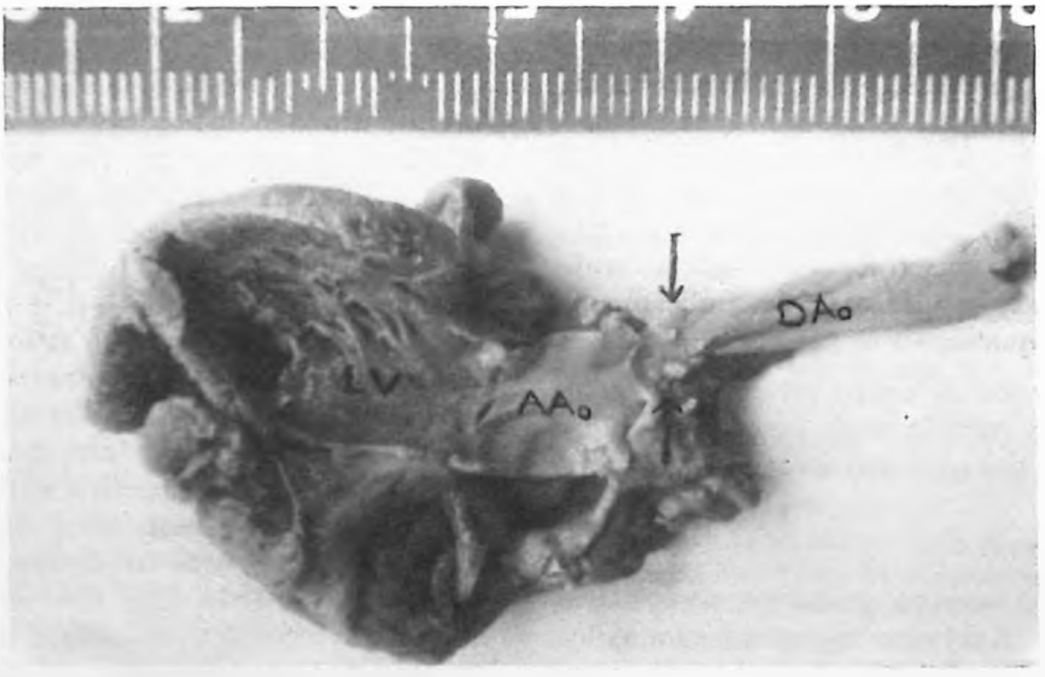

Fig. 3. Necropsy of the fetal heart shows postductal coarctation of the aorta.

peak pressure gradient was measured 83 mmHg by continuous wave Doppler in the same area. Color Doppler study revealed severe tricuspid regurgitation. The pulmonary valve velocity, the aortic valve velocity and the peak systolic flow velocity in the ductus arteriosus were increased. The diastolic velocity in the ductus arteriosus was normal.

The pregnancy terminated in spontaneous abortion at 22 weeks' gestation. The fetus was stillborn. The autopsy findings confirmed the prenatal diagnosis.

Discussion

Initial reports of antenatal detection of coarctation of the aorta consisted of isolated cases within several large series directed toward the in utero diagnosis of congenital heart disease[1]. In these earlier studies, the diagnosis of coarctation was based primarily on the two dimensional echocardiographic appearance of the arch at the site of the contraductal shelf and the accompanying area of the distal aortic arch hypoplasia[1]. Others[2-4] emphasized associated findings that might provide a clue to in utero diagnosis of aortic coarctation. A greater than usual discrepancy in ventricular size favoring the right ventricle and an increase in pulmonary artery size relative to the aorta were suggested by some investigators[2-4]. By spectral Doppler echocardiography, increased tricuspid valve flow velocities and decreased aortic flow were other in utero findings associated with coarctation. However, all of these are indirect observations and are not specific for this condition although they are consistent with present theories of the pathogenesis of aortic coarctation. Recent reports have emphasized quantitative hypoplasia of the isthmus, and transverse arch has been the most consistent observation and, therefore, the most definitive antenatal sign of postnatal coarctation[5].

In our case, there was a narrowing within the descending aorta immediately distal to the origin of the ductus arteriosus. The color-flow imaging demonstrated acceleration and turbulent flow and the peak pressure gradient was markedly increased by continuous wave Doppler in the coarctation area. Hypoplasia of the aortic isthmus and increased pulmonary artery size relative to the aorta were other in utero findings associated with coarctation.

When all the echocardiographic features of coarctation are present in early pregnancy, the diagnosis is highly likely to be correct and the outlook for postnatal survival is poor[4]. The mortality in these series is 64 percent in continuing pregnancies where the diagnosis was made before a gestational age of 24 weeks[4]. The prognosis of nonimmune hydrops in general is poor, with 82 percent mortality, and the atrioventricular valve regurgitation makes the prognosis even poorer[8]. This pregnancy terminated in spontaneous abortion at 22 weeks' gestation, and the fetus was stillborn.

We conclude that together with the quantitative estimation of the aortic arch, color Doppler and continuous wave Doppler are helpful in the diagnosis and estimation of the pressure gradient.

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