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● *Original Contribution*

THE THREE-VESSEL VIEW IN THE FETAL MEDIASTINUM IN THE DIAGNOSIS OF INTERRUPTED AORTIC ARCH

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Abstract—Interruption of the aortic arch (IAA) is difficult to detect and diagnose *in utero*. However, prenatal diagnosis may be beneficial because IAA is rapidly fatal (median age, 10 d) if left uncorrected. Our objective was to review the direct and indirect echocardiographic markers associated with IAA, focusing on the importance of the three-vessel view (3VV), which is obtained during routine ultrasound examination to rule out malformations. We analyzed the fetal echocardiograms of nine fetuses and compared them with 56 normal controls. In each fetus, there was a large discrepancy between the diameter of the larger, dilated pulmonary artery (PA) and smaller, narrow aortic arch (Ao). The calculated ratio of PA/Ao in fetuses with IAA was 2.6 ± 0.4 compared with 1.1 ± 0.09 in normal controls ($p < 0.0001$). The calculated ratio of PA/Ao in fetuses with IAA type A was 2.1 ± 0.09 and IAA type B 2.9 ± 0.2 ($p = 0.0007$). Discrepancy between PA/Ao diameters should raise the suspicion of aortic arch anomalies and a large discrepancy is a nearly pathognomonic sign of IAA type B. (E-mail: majkares@uni.lodz.pl) © **2011 World Federation for Ultrasound in Medicine & Biology.**

Key Words: **Interrupted aortic arch, Three-vessel view, Discrepancy, Congenital heart defect.**

INTRODUCTION

Interruption of the aortic arch (IAA) is a rare anomaly in the fetus and neonate that occurs in 3/100,000 live births (Morris and McNamara 1990). IAA was classified in 1959 by Celoria and Patton (1959) into three morphologic types: A, B and C, depending on the site of the interruption. In type A, interruption is distal to the left subclavian artery; in type B, interruption is between the left carotid and left subclavian arteries; and in type C, interruption is between the innominate artery and the left carotid artery. Type B is the most common type of IAA, accounting for 65% of cases, whereas types A and C occur less frequently—in 30% and 5%, respectively (Reardon et al. 1984). Other cardiac anomalies are often seen in association with IAA. Ventricular septal defects are commonly seen in association with IAA (Freedom et al. 1977; Marino et al. 1991; Reardon et al. 1984) but IAA can also be associated with complex congenital heart disease (Carotti et al. 2008; Lewin et al. 1997; Reardon et al. 1984), making the diagnosis more difficult.

Prenatal detection of IAA by echocardiography has been reported only a few times (Allan et al. 1994; Vogel et al. 2010; Volpe et al. 2003, 2010) and usually as case reports (Marasini et al. 1985; Paladini et al. 1998; Volpe et al. 2002). This condition is difficult to detect and diagnose *in utero* in even the most experienced tertiary care centers. However, prenatal diagnosis may be beneficial because IAA is rapidly fatal (median age, 10 d) if left uncorrected (Reardon et al.1984). Our objective was to review the direct and indirect echocardiographic markers associated with IAA, focusing on the importance of the three-vessel view (3VV).

MATERIALS AND METHODS

We analyzed the fetal echocardiograms of nine fetuses with antenatally suspected IAA, in whom the diagnosis was confirmed postnatally by angiography in the first days of life. There were three with type A, five with type B and one with type C IAA. The fetuses with

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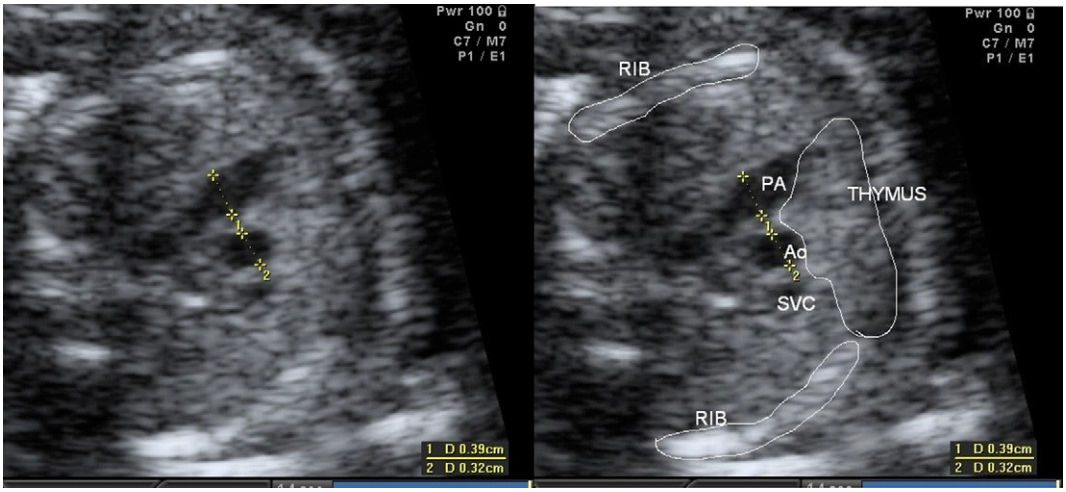

Fig. 1. Normal 3VV with exact points used to measure the PA and Ao diameters. SVC, superior vena cava.

suspected IAA were selected retrospectively from our electronic database of fetal echocardiograms (File Maker Pro 4, Microsoft Corp., Redmond, WA, USA). A control group consisting of 56 fetuses with structurally normal hearts by fetal and neonatal echocardiography was selected for comparison. The 3VV was routinely obtained as part of a comprehensive fetal ultrasound to rule out malformations in second trimester and also as a part of examination in the third trimester. The 3VV was obtained following the technique described by Yoo et al. (1997). After obtaining a four-chamber view, the transducer was slid cephalad along the fetal mediastinum. An adequate 3VV was defined by demonstration of a round (transverse) cross section of the ascending aorta and superior vena cava and an oblique section of the main pulmonary artery in a straight line (Fig. 1). Detailed retrospective analysis of fetal echocardiograms, with focus on the measures of the mediastinal great vessels, was performed. We measured vessel in the largest diameter seen in 3VV. The *t*-test was used for continuous variables, with $p < 0.05$ used to define statistical significance. We received informed consent from each mother before fetal echocardiography and our study was approved by the hospital's ethics committee.

RESULTS

The mean maternal age was 29 (±3.4) y. Cesarean sections were performed in five of nine pregnancies. Mean birth weight was 3220 ± 516 g. There were no preterm deliveries. All neonates were given prostin E1 (alprostadil) to prevent ductus arteriosus closure. Angiography was performed by the third postnatal day of life to confirm the diagnosis of IAA. All nine neonates underwent surgery to correct the heart defect. All of the neonates were discharged from the hospital. All of the fetuses diagnosed with IAA were referred for fetal echocardiography because of an abnormal four-chamber view. Four fetuses also presented with polyhydramnios (amniotic fluid index 25 cm, 26 cm, 26 cm and 33 cm). The median gestational age at the time of fetal echocardiography was 32.5 ± 3.6 weeks. All nine fetuses were

Table 1. Detailed fetal ultrasound and echo findings that lead to the diagnosis of IAA

Case	IAA type	GA (wk)	HA/CA	AFI (cm)	Diagnosis	3VV	PA/Ao ratio	"Y" sign of Ao	Narrow Ao	Bronchogram	Abnormal shape of DA	Additional vessels
1	A	37	0.3	33	VSD + R	Abnormal	2.1		Yes		Yes	
2	A	27	0.3	16	VSD + DORV	Abnormal	1.94	Yes	Yes	Yes	Yes	
3	B	31	0.3	16	VSD	Abnormal	3			Yes	Yes	
4	A	37	0.3	26	VSD	Abnormal	2.1		Yes		Yes	Collaterals
5	C	35.6	0.3	9	VSD	Abnormal	3	Yes	Yes		Yes	
6	B	30	0.3	25	VSD	Abnormal	2.8		Yes			Art. lusoria
7	B	32	0.3	15	VSD	Abnormal	3	Yes				Vena azygos
8	B	33	0.3	18	VSD	Abnormal	—					
9	B	30	0.3	26	VSD	Abnormal	2.56					

GA = gestational age; AFI = amniotic fluid index; R = right aortic arch; DA = ductus arteriosus.
In case 8, only the PA was seen, so we were not able to estimate the ratio.

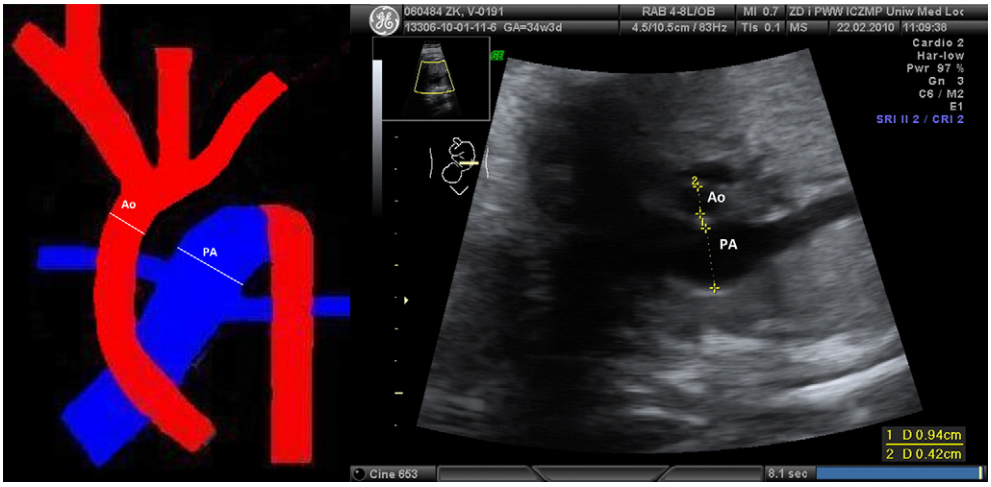

Fig. 2. Diagram and 3VV in a fetus at 34 weeks' gestational age with interrupted Ao type A confirmed after delivery. PA/Ao ratio 2:2.

diagnosed with additional cardiac malformations including ventricular septal defect ($n = 9$) and double-outlet right ventricle ($n = 1$). Ultrasound and echocardiogram findings are shown in Table 1. The heart area/chest area (HA/CA) ratio was normal in all nine cases (measured from the four-chamber view as the HA divided by CA). In addition, in all nine cases, the heart was in the normal position (levocardia). One fetus had a right aortic arch. In all nine cases, the 3VV view was abnormal. In each fetus there was a large discrepancy between the diameter of the larger, dilated pulmonary artery (PA) and the smaller, narrow aortic arch (Ao). The calculated ratio of PA/Ao in fetuses with IAA was 2.6 ± 0.4 compared with 1.1 ± 0.09 in normal controls ($p < 0.0001$). The calculated ratio of PA/Ao in fetuses with IAA type A was 2.1 ± 0.09 (Fig. 2), and in fetuses with IAA type B was 2.9 ± 0.2 (Fig. 3) ($p = 0.0007$). A "Y" sign of aorta describe also by Vogel et al. (2010) (aorta, innominate artery and left carotid artery) (Fig. 4) was well seen in three fetuses. We measured the Ao in long-axis view of the aorta and defined a narrow Ao in the third trimester of pregnancy as <4 mm (Hornberger et al. 1992). A narrow ascending aorta was seen in five fetuses.

In five fetuses the shape of ductus arterious was abnormal. We have termed the abnormal appearance of the ductus arteriosus the "broken hockey stick" sign (Fig. 5). In three fetuses there were additional vessels visualized in the 3VV or long-axis view between the descending aorta and the fetal heart. The additional vessels included "collaterals," arteria lusoria (aberrant right subclavian artery, leaving the aorta below the left subclavian artery and usually retroesophageal) and azygos vein.

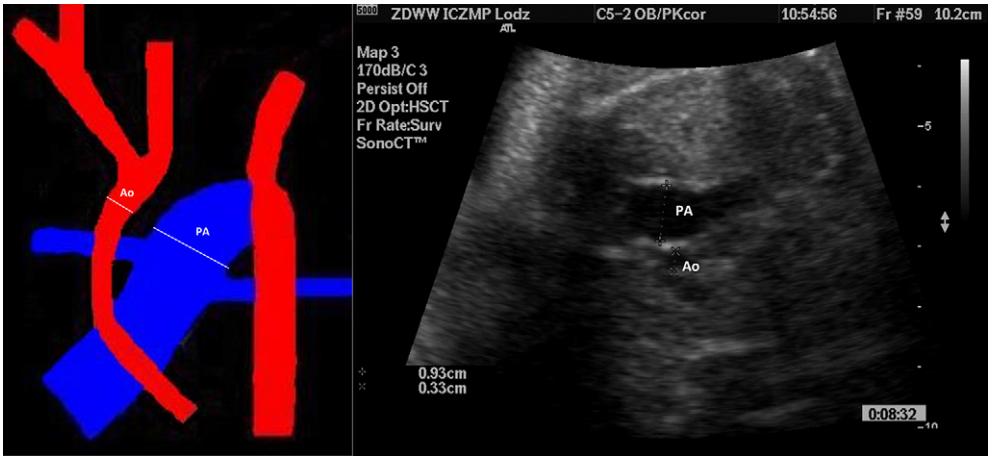

Fig. 3. Diagram and 3VV in a fetus at 32 weeks' gestational age with interrupted Ao type B confirmed after delivery. PA/Ao ratio 2:8.

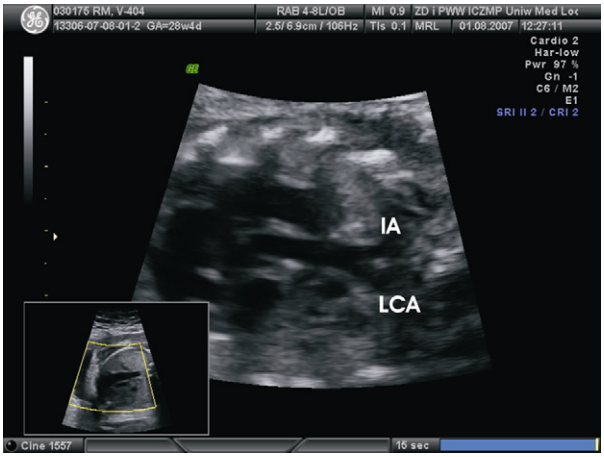

Fig. 4. "Y" sign. Note the aorta ascending without curvature and two vessels: innominate artery (IA) and left carotid artery (LCA), in a fetus at 28 weeks' gestational age, interrupted Ao type B confirmed after delivery.

DISCUSSION

The 3VV is easy to obtain and shows a transverse view of the fetal upper mediastinum, where normally the oblique section of the main PA and cross section of the ascending aorta and superior vena cava are arranged in a straight line (Yoo et al. 1997). In IAA, the Ao has a significantly smaller diameter than the main PA. Yoo et al. were the first to describe the usefulness of the 3VV to diagnose IAA because this view illustrates the abnormal vessel size. Yagel et al. (2002) demonstrated the clinical applicability of a slightly different view: the three-vessel and trachea (3VT) view, defined as a transverse view of the upper mediastinum slightly cranial to the usual 3VV. The advantage of this view, compared with the 3VV, is in making the diagnosis of a right Ao but there is no advantage over the 3VV when it comes to diagnosing IAA (Achiron et al. 2002). In our experience, PA and Ao size can be easily appreciated in either the 3VV or 3VT. Both planes allow for easy measurement of the proximal part of PA and ascending aorta.

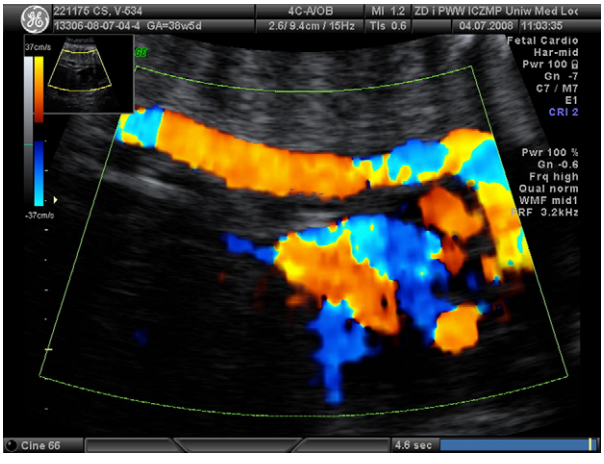

Fig. 5. "Broken hockey stick" sign in a fetus at 38 weeks' gestation.

Typically, in IAA the Ao has a significantly smaller diameter than the main PA. In a series reported by Volpe et al. (2002), the PA/Ao ratio was >1.4, whereas in our series the ratio was 2.6 (Fig. 6). The difference is probably because of the higher mean gestational age in our series—34 weeks compared with 25 weeks. In another series, the PA/Ao ratio was 1.92 at 25 weeks (Vogel et al. 2010). In normal pregnancy, we observed slightly increasing PA/Ao ratio from 1.1 in 20 weeks to 1.2 in 40 weeks (Zalel et al. 2004). This finding, increasing PA/Ao ratio at later gestational ages, may suggest that altered hemodynamics as a consequence of IAA causes more progressive dilatation of the main pulmonary artery in the second and third trimester of pregnancy.

Distinguishing between IAA and coarctation of the aorta (CoA) is possible but not easy (Vogel et al. 2010). In IAA, curvature of the ascending aorta is abnormal, coursing straight to its branches whereas in coarctation of the aorta the aortic arch is normally curved and is continuous with the descending aorta (Volpe et al. 2002). We agree with Volpe et al., but it is also possible to differentiate between the different types of IAA. In type B, the ascending aorta follows a straight course to the innominate artery and the left carotid arteries, whereas in type A there is a slight curvature after the origin of the innominate artery, related to the persistence of the aortic arch segment between the origin of the left carotid and subclavian arteries. For this reason, it is easiest to differentiate between CoA and IAA type B.

It has been suggested that the etiology of IAA type B is distinct from both of the other forms of IAA (Van Mierop and Kutsche 1984; Volpe et al. 2010). The pathogenetics of type A IAA are believed to be more closely related to CoA and are thought to be caused by abnormal blood flow during embryogenesis (Fig. 7). On the other hand, IAA type B is caused by an abnormality

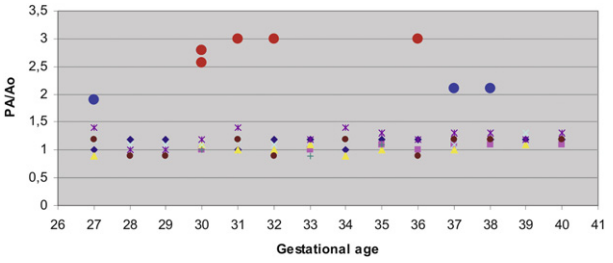

Fig. 6. Distribution of the PA/Ao ratio in 56 normal pregnancies and in eight fetuses with IAA type A (blue = 3 cases) and IAA types B and C (red = 5 cases). (In the ninth fetus, only a dilated pulmonary artery was seen, so the ratio was not calculated.)

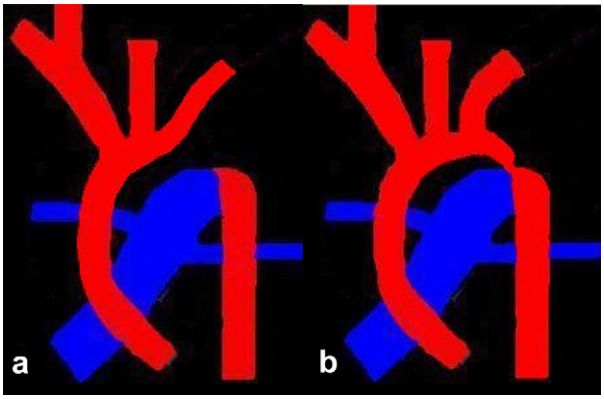

Fig. 7. Diagram of IAA type A (a) and CoA (b).

of neural crest cell migration. Vogel et al. (2010) demonstrated that it may be possible to distinguish between type A and B IAA based on the degree of PA/Ao discrepancy. They demonstrated that the size discrepancy between the PA and ascending aorta diameter was greater in type B IAA than type A IAA. This also held true comparing IAA type B with coarctation of the aorta. In our own recent study, we demonstrated the usefulness of measurement of the great vessels in diagnosing CoA in the third trimester of pregnancy (Slodki et al. 2009). Because IAA type A is more closely related to CoA than type B IAA, we speculate that the degree of PA/Ao diameter discrepancy may be useful in differentiating type B IAA from coarctation of the aorta but not between type A IAA and CoA. Our sample size would have been too small to test this hypothesis (Fig. 8).

The presence of other abnormalities including bronchogram, an additional vessel in mediastinum or "Y" sign of the aorta may contribute to the diagnosis of IAA and differentiate between types, but these findings are much more subtle compared with the PA/Ao diameter discrepancy. New techniques like 3-D multiplanar imaging or spatio-temporary image correlation (STIC) are promising in prenatal diagnosis of congenital heart disease (De Vore et al. 2004) and also in differentiating between IAA type A and B (Volpe et al. 2010). However,

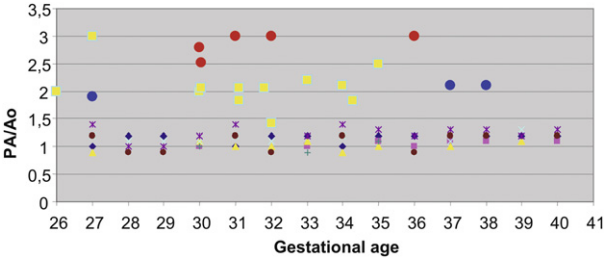

Fig. 8. Distribution of the PA/Ao ratio in normal pregnancy, IAA type A (*blue dot*), IAA types B and C (*red dot*) and CoA (*yellow square*).

neither of these techniques is perfect (Rizzo et al. 2008) and should not replace traditional measurements obtained from 2-D evaluation of the mediastinum.

CONCLUSION

The 3VV is the best view to diagnose IAA. Discrepancy between PA/Ao diameter should raise the suspicion of Ao anomalies, and a large discrepancy is a nearly pathognomonic sign of IAA type B.

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