Tetralogy of Fallot in the fetus: findings at targeted sonography

S.-J. Yoo*, Y.-H. Lee*, E. S. Kim^{\ddagger} , H. M. Ryu^{\ddagger} , M. Y. Kim^{\ddagger} , J. H. $Yang^{\ddagger}$, Y. K. Chun** and S. R. Hong**

Departments of *Ultrasound, [‡]Obstetrics and Gynecology, **Pathology, Samsung Cheil Hospital and Women's Healthcare Center, Seoul; Department of Radiology, Sejong Heart Institute, Kyunggi-do, Korea

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ABSTRACT

Objectives To evaluate the findings of tetralogy of Fallot in various fetal sonographic views.

Methods We reviewed the fetal sonograms and medical records of 20 fetuses with prenatal diagnosis of tetralogy of Fallot. We analyzed the indications for targeted sonography, the abnormalities seen in various sonographic views, the postnatal echocardiographic and angiographic findings and autopsy findings.

Results The most common indication for targeted sonography was an abnormal (n = 12) or inadequate (n = 3)finding on sonographic screening in which the abnormality was most frequently found on the three-vessel view (n = 9). The key pathological features of tetralogy of Fallot were uniformly demonstrated in the ventricular outflow tract, three-vessel and short-axis views. The ductus arteriosus was small in 70% of cases and not identifiable in the remaining fetuses. In three of six fetuses with no identifiable ductus, the ductus was shown to be absent at autopsy. The direction of ductal flow was variable.

Conclusion The key features of tetralogy of Fallot were always demonstrable in the ventricular outflow tract. three-vessel and short-axis views. The most common reason for referral was the abnormal three-vessel view.

INTRODUCTION

ORIGINAL PAPER

Tetralogy of Fallot is one of the most common types of cyanotic congenital heart disease, accounting for 8-10% of all congenital heart diseases among newborns¹. In most established cardiac centers, the operative mortality is less than 5% when the operation is performed after the first month of life². Prenatal diagnosis of tetralogy of Fallot aids parental counselling and enables delivery to be

planned at a pediatric cardiac center with early initiation of optimal treatment, which may further improve treatment outcome³⁻⁵.

Many papers and textbooks have introduced fetal sonographic clues to the diagnosis of tetralogy of Fallot. However, few have described its various fetal sonographic findings in detail. The purpose of the present study was to evaluate the findings of tetralogy of Fallot in various sonographic views.

MATERIALS AND METHODS

Between August 1995 and December 1997, 24 fetuses were diagnosed as having tetralogy of Fallot by targeted fetal sonography. Of these 24 fetuses, we studied 20 in which the prenatal diagnosis was confirmed by postnatal examination (n = 11) or autopsy (n = 9). The diagnosis of tetralogy of Fallot was based on the main anatomical feature - the anterosuperior and leftward deviation of the outlet septum relative to the rest of the interventricular septum, regardless of the degree of aortic overriding¹.

We examined the fetal heart in various imaging planes with continuous sweeping of the transducer, and routinely obtained the following six basic views: the transverse view of the upper abdomen, the four-chamber view, the threevessel view⁶, the left ventricular outflow tract view, the right ventricular outflow tract view, and the aortic arch view. We first obtained a transverse view of the upper abdomen. We then moved the transducer cranially along the long axis of the fetal body to obtain a four-chamber view and further upward to obtain a three-vessel view. We returned to the four-chamber plane and moved the transducer radially around the maternal abdomen to place the interventricular septum perpendicular to the sonographic beam axis (Figure 1). We then rotated the transducer clockwise or counterclockwise toward the cardiac apex and obtained a left ventricular outflow tract view. We slid the transducer a little toward the fetal head and obtained a right ventricular outflow tract view. We returned to the three-vessel plane and moved the transducer radially around the maternal abdomen to align the cross-sections of the ascending and descending aorta with the sonographic beam axis (Figure 2). We then rotated the transducer through 90° to obtain an aortic arch view. When this longitudinal aortic arch view was not obtainable, we obtained a transverse view of the aortic arch by sliding the transducer toward the fetal head from the threevessel plane. When an abnormality or abnormalities were found in the six basic views, we also obtained a short-axis view and performed color or power and pulsed Doppler examinations of the four cardiac valves and the ductus arteriosus.

We reviewed the screening and targeted sonograms and the medical records of the patients and their pregnancy outcome. We analyzed the indications for the targeted sonography, the abnormalities seen in various views of the screening and targeted sonography, the postnatal echocardiographic and angiographic findings and autopsy findings.

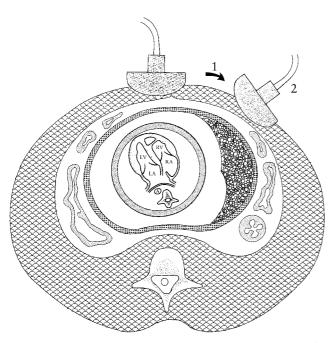


Figure 1 Diagram showing standard scanning maneuver for ventricular outflow tract views in a fetus in supine cephalic presentation. The examination starts from the four-chamber view obtained by the transducer at position 1. The transducer is moved radially (curved arrow) around the maternal abdomen to position 2, bringing the interventricular septum perpendicular to the sonographic beam. By counterclockwise rotation of the transducer at position 2 toward the fetal cardiac apex, the ventricular outflow tract views can be easily obtained. A, descending aorta; LV, left ventricle; RV, right ventricle; LA, left atrium; RA, right atrium. From reference 7 with permission

RESULTS

The indications for targeted sonography are listed in Table 1. The majority of the targeted examinations were performed because of abnormal (60%) or inadequate (15%) findings on sonographic screening. The abnormal finding during screening was found most commonly in the threevessel view (nine out of 12). The threevessel view abnormalities consisted of either one or a combination of abnormal vessel alignment, a small main pulmonary artery and a large ascending aorta, and/or a right-sided descending aorta. The ventricular outflow tract view abnormality was the origin of the aorta from the right ventricle. The four-chamber view abnormality was the leftward deviation of the cardiac axis with right ventricular hypertrophy. The

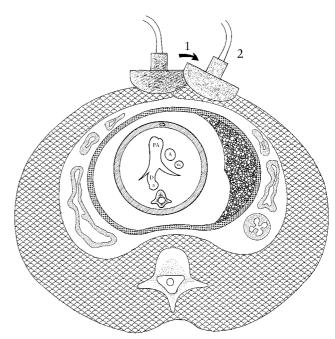


Figure 2 Diagram showing standard scanning maneuver for aortic arch view in a fetus in supine cephalic presentation. The examination starts from the three-vessel view obtained by the transducer at position 1. The transducer is moved radially (curved arrow) around the maternal abdomen to position 2, where the cross-sections of the ascending (A) and descending aorta (a) are aligned perpendicular to the sonographic beam. By 90° counterclockwise rotation of the transducer toward the fetal feet, the aortic arch view can be easily obtained. PA, main pulmonary artery; D, ductus arteriosus; VC, superior vena cava. From reference 7 with permission

Table 1 Indications for targeted sonography

Indication	Number of patients
Abnormal three-vessel view	9
Abnormal ventricular outflow tract view	2
Abnormal four-chamber view	1
Inadequate screening sonography	3
Advanced maternal age	1
Increased maternal serum α-fetoprotein	1
Twin pregnancy	1
Multiple anomalies	1
Known Down's syndrome	1

Table 2 Findings at targeted sonography

Sonographic views	Sonographic findings	Number of fetuses
Four-chamber view	leftward deviation of the cardiac apex	8
	thick right ventricular wall	10
	ventricular septal defect	4
	dextrocardia with situs inversus*	1
Ventricular outflow tract views	ventricular septal defect	20
	overriding aorta	20
Three-vessel view	abnormal vessel alignment	17
	small pulmonary artery and large aorta	20
	right-sided descending aorta	9
Short-axis view	subpulmonary stenosis due to deviated outlet septum	15
	atretic subpulmonary outflow tract	2
	small pulmonary valve annulus in the presence of doubly committed ventricular septal defect	3
	muscular outlet ventricular septal defect	1
Ductus arteriosus view	left aortic arch with left ductus arteriosus	7
	right aortic arch with left ductus arteriosus	6
	right aortic arch with right ductus arteriosus	1*
	not identifiable	6

^{*}Fetus with situs inversus

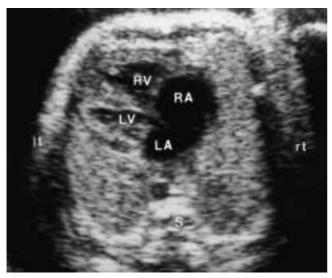


Figure 3 Right ventricular hypertrophy. Four-chamber view shows leftward deviation of the cardiac axis and thick right ventricular free wall. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; lt, left; rt, right; S, spine

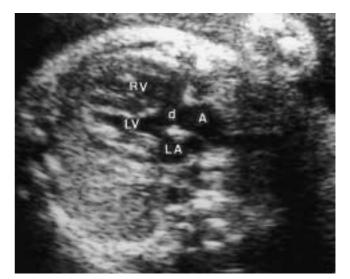


Figure 4 Perimembranous ventricular septal defect and overriding aorta. Left ventricular outflow tract view shows a ventricular septal defect (d) immediately below the aortic root (A). Approximately 25% of the aorta overrides the ventricular septum. LA, left atrium; LV, left ventricle; RV, right ventricle

diagnosis was made before 20 weeks of gestation in two fetuses, between 20 and 25 weeks in nine, between 25 and 30 weeks in four and after 30 weeks in five. The average gestational age at diagnosis was 26.4 weeks.

The sonographic abnormalities are listed in Table 2 and shown in Figures 3-15. Six basic views were obtainable in all fetuses. The four-chamber view was abnormal in 18 cases. The ventricular outflow tract and three-vessel views were abnormal in all fetuses. The shortaxis view was abnormal in all 19 fetuses in which it was obtainable.

The visceroatrial situs was normal in 19 fetuses. There was one fetus with situs inversus that was missed on sonography. The ventricular septal defect was of the perimembranous type in 15 cases (including one with atrioventricular septal defect), doubly committed juxta-arterial type in three cases and muscular outlet type in two cases. The type of defect was confirmed to be correct at postnatal examination or autopsy in 18 fetuses. The muscular outlet defect that was present in two fetuses was misdiagnosed as a perimembranous defect. The largest dimension of the ventricular septal defect was seen on the left ventricular outflow tract view in 14 fetuses with a perimembranous defect and one with a muscular outlet defect (Figure 4), at the short-axis view in three fetuses with a doubly committed juxta-arterial defect (Figure 8) and one with a muscular outlet defect (Figure 9) and at the four-chamber view in one fetus with an atrioventricular septal defect.

All showed variable degrees of overriding of the aorta in the left ventricular outflow tract view; 25% overriding in one (Figure 4), 50% overriding in nine, 75% overriding in four and more than 75% overriding in six cases.

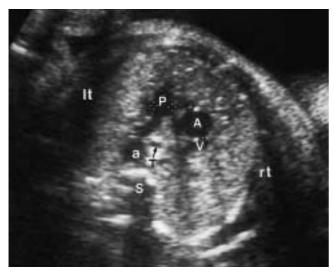


Figure 5 Abnormal alignment and size of the three vessels. Three-vessel view shows small main pulmonary artery (P) and large ascending aorta (A). The aorta is displaced anteriorly, and the pulmonary artery posteriorly. a, descending aorta; S, spine; T, trachea; V, superior vena cava; lt, left; rt, right

In 18 fetuses, right ventricular hypertrophy was suspected because of leftward deviation of the cardiac apex and/or thick right ventricular free wall (the right ventricular free wall) being thicker than the left ventricular free wall) in the four-chamber view (Figure 3).

Subpulmonary stenosis caused by anterosuperior displacement of the outlet septum was seen in the short-axis view in 15 fetuses with a perimembranous, atrioventricular or muscular outlet defect (Figures 7 and 9). In all three fetuses with a doubly committed juxta-arterial defect, the pulmonary valve annulus was smaller than the aortic valve annulus (Figure 8). It was difficult to evaluate the presence of pulmonary valve stenosis. Pulmonary atresia was diagnosed at the initial scan in two fetuses (Figure 10). Two fetuses developed pulmonary atresia identified on follow-up examination during gestation (Figure 14). One was found to have unsuspected pulmonary atresia at birth. One developed pulmonary atresia on the 4th day of postnatal life.

The main pulmonary artery was smaller than the ascending aorta in the three-vessel view in all cases (Figures 5 and 6). In 17 fetuses, the alignment of the vessels seen at the three-vessel view was abnormal because of anterior displacement of the aorta and posterior displacement of the pulmonary artery (Figure 5).

The descending aorta was on the right anterior aspect of the spine in nine fetuses, all of which showed a right aortic arch at the transverse aortic arch view. One of these was the case with situs inversus.

A ductus arteriosus was identified in 14 fetuses and was not identifiable in six fetuses. The identifiable ductus arteriosus was invariably small and present on the left, regardless of the position of the aortic arch when situs solitus was present. The ductus arteriosus arose from the undersurface of the aortic arch in five of the seven fetuses with situs solitus, a left aortic arch and an identifiable left ductus





Figure 6 Right aortic arch. (a) Three-vessel view shows that the main pulmonary artery (P) is smaller than the ascending aorta (A). There is minimal malalignment of the vessels. The descending aorta (a) is located at the right anterior aspect of the spine (S). (b) Transverse aortic arch view shows the aortic arch (Aa) on the right side of the trachea (T). V, superior vena cava; lt, left; rt, right

(Figure 11). The ductus arose from the junction of the aortic arch and descending aorta in the remaining two fetuses. The ductus arose from the undersurface of the right aortic arch in the fetus with situs inversus. In six fetuses with a right aortic arch and an identifiable left ductus, the ductus was seen between the left innominate artery and the left pulmonary artery (Figure 12). Among six fetuses with no identifiable ductus arteriosus, the ductus was shown to be absent by autopsy in three cases and small by autopsy or postnatal angiography in two cases. In one case with pulmonary atresia and absent ductus, major aortopulmonary collateral arteries were demonstrated on fetal sonography (Figure 15). There was no comment on the ductus in the autopsy record in the remaining case.

Doppler waveforms from the ductus arteriosus could be obtained in only five fetuses. In the remaining fetuses, Doppler waveforms could not be obtained, as the ductus arteriosus was aligned almost perpendicularly to the sonographic beam axis. The ductal flow was from the

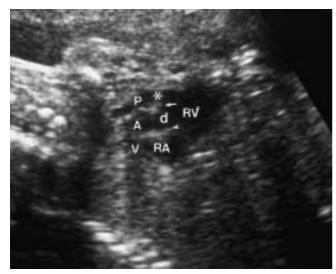


Figure 7 Subpulmonary stenosis. Modified short-axis view shows that the subpulmonary outflow tract (asterisk) is encroached upon by the deviated outlet septum (arrow). The aortic and tricuspid valves are in direct contact (arrowhead) at the posterior margin of the ventricular septal defect (d), which is characteristic of the perimembranous defect. A, ascending aorta; P, main pulmonary artery; RA, right atrium; RV, right ventricle; V, superior vena cava



Figure 9 Muscular outlet ventricular septal defect. Modified short-axis view shows that the defect (d) is bounded by muscular rims (arrow and arrowhead) on both sides. Neither the pulmonary nor the tricuspid valve is in direct contact with the aortic valve. It was misinterpreted as a perimembranous defect. P, main pulmonary artery; A, ascending aorta; V, superior vena cava; RA, right atrium; RV, right ventricle

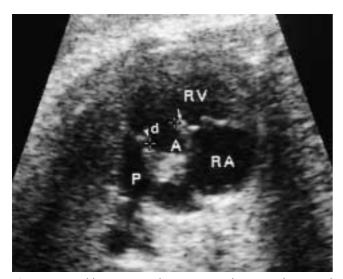


Figure 8 Doubly committed juxta-arterial ventricular septal defect. Short-axis view shows that the ventricular septal defect (d) involves the most cranial part of the ventricular septum. The aortic and pulmonary valves are in direct contact (arrowhead) through the defect. There is a posteroinferior rim (arrow) between the aortic and tricuspid valves. The pulmonary valve annulus is small. A, aortic root; P, main pulmonary artery; RA, right atrium; RV, right ventricle

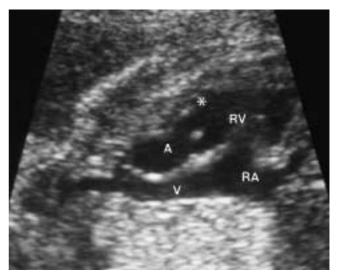


Figure 10 Pulmonary atresia. Modified short-axis view shows the atretic subpulmonary outflow tract (asterisk). A, ascending aorta; RA, right atrium; RV, right ventricle; V, superior vena cava

pulmonary artery into the aorta in two cases, from the aorta into the pulmonary artery in two cases and bidirectional in one (Figure 13). Doppler examination of the cardiac valves in general was of little help in understanding the underlying pathology or in predicting the outcome.

Karyotyping was performed in 15 fetuses. There was one case each of trisomy 18 and trisomy 21. Extracardiac anomalies were associated with six cases (30%) that included the two trisomic fetuses.

The outcome of the 20 fetuses is summarized in Table 3. Nine sets of parents chose termination of pregnancy after counselling. The termination was performed because of an abnormal fetal karyotype in two cases, because of multiple extracardiac anomalies in two cases and by parental decision in five cases. Eleven cases were born alive: nine at a tertiary cardiac center and two at our hospital. All of the five liveborns with pulmonary atresia received an urgent Blalock-Taussig shunt operation. Two of these died



Figure 11 Left ductus arteriosus arising from left aortic arch. Aortic arch view shows that a small ductus (DA) arises from the undersurface of the aortic arch (Aa) with an obtuse proximal angle. A, ascending aorta; a, descending aorta

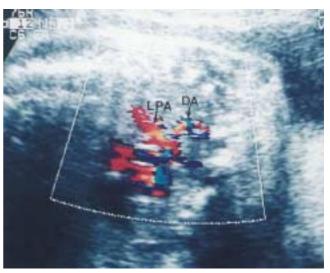
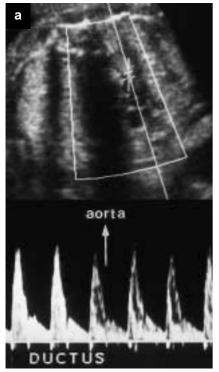
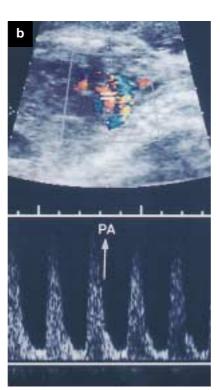


Figure 12 Left ductus arteriosus arising from the left innominate artery of the right aortic arch. Oblique coronal sonogram of the left thorax shows a small ductus (DA) connected to the left pulmonary artery (LPA). The connection of the ductus arteriosus to the left innominate artery was demonstrated in the adjacent plane





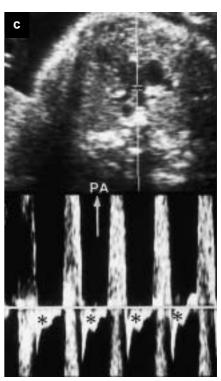


Figure 13 Various Doppler waveforms from the ductus arteriosus. (a) Unidirectional continuous forward flow from the pulmonary artery into the aorta. (b) Unidirectional continuous reversed flow from the aorta into the pulmonary artery (PA). (c) Bidirectional flow with reversed flow during the systolic phase and forward flow (asterisks) during the diastolic phase

immediately after the shunt operation. None of the cases died *in utero*.

DISCUSSION

The characteristic findings of tetralogy of Fallot are anterosuperior and leftward deviation of the outlet septum relative to the rest of the ventricular septum, resulting in an anterior malalignment type of ventricular septal defect, subpulmonary obstruction, overriding or dextroposition of the aorta and right ventricular hypertrophy^{1,2}. The fetal echocardiographic findings of the various features of tetralogy of Fallot have not been described in detail, but those of fetuses are likely to differ from those of the postnatal patients because of the different sonographic windows, the ever-changing fetal position, the small fetal heart size, the patency of the ductus arteriosus and perhaps the evolving nature of the lesions.

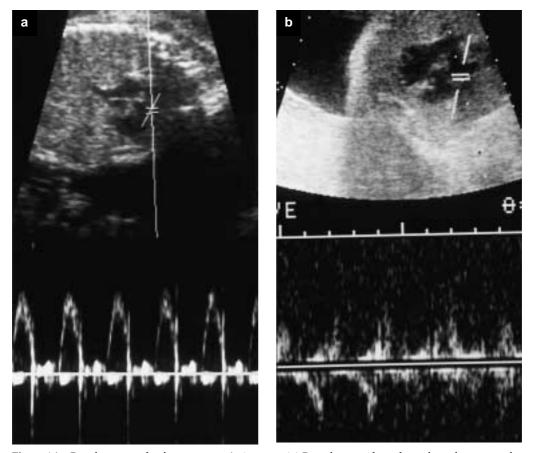


Figure 14 Development of pulmonary atresia in utero. (a) Doppler waveform from the pulmonary valve at 30 weeks shows forward flow. The blood flow through the ductus arteriosus was bidirectional as shown in Figure 13c, obtained from the same fetus. (b) Doppler waveform from the pulmonary valve at 33 weeks shows no forward flow. Signals above and below the baseline denote turbulent flow around the atretic pulmonary valve. Reversed flow through the ductus was demonstrated at a later gestation



Figure 15 Pulmonary atresia with a major aortopulmonary collateral artery. Color Doppler sonogram in the transverse plane shows a large collateral artery (c) arising from the descending aorta (a). The ductus arteriosus was absent. S, spine

Although examination of the four-chamber view is an excellent screening tool for detection of major congenital heart disease, it has limitations in detecting anomalies involving the ventricular outflow tracts and/or great arteries,

and therefore tetralogy of Fallot^{4,8-16}. This weakness can be compensated for if the examination is extended to include the ventricular outflow tract views^{4,10-16}. However, the left and right ventricular outflow tract views have often been considered difficult and time-consuming to obtain 14,16. To solve this problem, we developed a standard maneuver to obtain the ventricular outflow tract views more easily and more consistently. The essence of our maneuver for the ventricular outflow tract views was to use the standard four-chamber view, in which the interventricular septum is placed perpendicular to the sonographic beam axis. From this standard position, the left ventricular outflow tract view can be easily obtained by simple rotation of the transducer toward the fetal cardiac apex⁷. We also added the three-vessel view to our screening protocol. The threevessel view is an orthogonal transverse view of the upper mediastinum where the oblique section of the main pulmonary artery and the cross-sections of the ascending aorta and superior vena cava can be identified lying in a straight line. The main pulmonary artery section is consistently larger than that of the ascending aorta which, in turn, is larger than that of the superior vena cava. Our previous study emphasized the usefulness of the three-vessel view in the detection of the anomalies involving the ventricular outflow tracts and/or great arteries⁶. By developing standard maneuvers to obtain the ventricular outflow tract views

Table 3 Pregnancy outcome

Pregnancy outcome	Number of fetuses
$\overline{Born\ alive\ (n=11)}$	
Postnatal management	
one-stage total correction	3
total correction after Blalock-Taussig shunt	2
waiting for total correction without any intervention	2
waiting for total correction after Blalock–Taussig shunt	2
death after Blalock-Taussig shunt	2
Termination of pregnancy $(n = 9)$	
Reason for termination of pregnancy	
chromosomal abnormality with multiple anomalies	2
multiple major anomalies	2
parental decision	5

and including the three-vessel view in the routine views, we substantially improved our detection rate of congenital heart disease, without significant increase in the duration of the examination.

The key pathological features of tetralogy of Fallot can be demonstrated consistently in the ventricular outflow tract, three-vessel and short-axis views. The interventricular septal defect is almost always large and readily detectable. The defect is most commonly a perimembranous type and best shown in the left ventricular outflow tract view. When the defect is a doubly committed juxtaarterial or muscular outlet type, however, it may no longer be seen in the left ventricular outflow tract view, although it will be seen in the short-axis view as a discontinuity of the muscular outlet septum supporting the aortic and pulmonary valves. Overriding of the aorta can best be detected in the left ventricular outflow tract view. As the dilated ascending aorta is displaced rightward and anteriorly and the main pulmonary artery posteriorly, the vessel alignment seen at the three-vessel view is abnormal in most cases⁶. Encroachment of the right ventricular outflow tract by the deviated outlet septum is characteristically shown in the short-axis view as an anterior protrusion of the outlet septum¹². Of the four typical features of tetralogy of Fallot, right ventricular hypertrophy may not be apparent in fetal life⁴. It should be suspected, however, whenever the right ventricular free wall is thicker than the left ventricular free wall in the four-chamber view. It may also be suspected when the cardiac apex is deviated to the left¹⁷.

The main pulmonary artery shows variable degrees of hypoplasia because of diversion of the right ventricular output into the ascending aorta. This produces a significant disproportion in size of the great arteries with enlargement of the size of the ascending aorta, providing an important diagnostic marker of tetralogy of Fallot^{4,12,18,19}. The disproportion can be assessed by direct measurement of the great arterial diameters and calculation of the pulmonary artery to aortic diameter ratio^{4,18,19}. The disproportion can also be readily assessed by a quick comparison of the relative size of the two great arteries by simple inspection¹⁸. In this regard, the three-vessel view is superior to any other single view, as it demonstrates both great arteries in a single

plane⁶. In our series, the disproportionate size of the two vessels was apparent on the three-vessel view in all cases. In fetuses with pulmonary atresia and severe hypoplasia or aplasia of the main pulmonary artery, only two vessels can be seen in the three-vessel view as in truncus arteriosus⁶.

A right aortic arch is common in tetralogy of Fallot and can be suspected when the descending aorta is seen on the right anterior aspect of the spine at the three-vessel or four-chamber view⁶. The position of the aortic arch relative to the fluid-filled trachea can be confirmed by the transverse aortic arch view.

The morphology of the ductus arteriosus and the blood flow pattern through it in tetralogy of Fallot should be mentioned as, in most cases with tetralogy, the left ductus persists regardless of the position of the aortic arch¹. Therefore, when a right aortic arch is identified, one should search for a ductus between the left innominate artery and the left pulmonary artery. The ductus is characteristically small in fetuses with tetralogy of Fallot, reflecting the reduced blood flow through this vessel⁵. The direction of the blood flow through the ductus may be from the pulmonary artery to the aorta when the subpulmonary and pulmonary stenosis is mild. When the stenosis is severe, the ductal flow direction may be reversed^{19,20}. This may explain why the ductus arteriosus in tetralogy of Fallot commonly arises from the undersurface of the aortic arch as if it were a branch of the aorta²¹. In the present study, the ductus arteriosus was small in 14 cases and not identifiable in six cases. In cases with a left aortic arch and an identifiable ductus, the insertion of the ductus was situated more proximally on the undersurface of the aortic arch than in the normal fetus. We attempted Doppler examination of the ductus in every case to document the ductal flow patterns in tetralogy of Fallot. We failed, however, to obtain adequate Doppler waveforms in the majority of cases, as the ductus was small and its course was usually perpendicular to the incident sonographic beam. In the five fetuses in which the ductal waveforms could be obtained, the flow direction was forward, reversed or bidirectional. In cases of pulmonary atresia, the source of pulmonary arterial supply is either a ductus arteriosus or major aortopulmonary collateral arteries. When one or both lungs are supplied by the collateral arteries, the ductus arteriosus is almost always absent. The other entity that commonly occurs with an absent or atretic ductus arteriosus is tetralogy with absent pulmonary valve syndrome^{22–25}. It is interesting to note that the ductus is usually patent when absent pulmonary valve syndrome is not associated with tetralogy of Fallot^{23,24}.

Among the various sonographic features described here, the abnormalities of the three-vessel view are perhaps most easily detected. This is supported by the fact that the most common reason for referral from sonographic screening in our study population was the abnormal three-vessel view. In screening by obstetric sonography, the abnormalities of the three-vessel view together with the abnormalities of the left ventricular outflow tract view lead the sonographer to the correct diagnosis of tetralogy of Fallot in most cases. Conversely, it is important for the sonographer to be acquainted with other features that may provide a clue, or

clues, to the diagnosis when inadequate sonographic visualization of the fetal heart is achieved.

In utero progression of the severity of the lesions in tetralogy of Fallot has been well documented^{3,4,19,26}. The ascending aorta and less frequently the main pulmonary artery may be normal in size in early gestation^{4,19}. The pulmonary arterial growth usually parallels the normal range, but the decelerated growth, especially of the main pulmonary artery, strongly suggests progression of the severity of the disease. Hornberger and colleagues¹⁹ demonstrated that decelerated growth of the pulmonary arteries, accelerated growth of the ascending aorta, cessation of forward flow through the pulmonary valve and reversed flow through the ductus arteriosus were all predictive of ductus-dependent pulmonary circulation at birth and were thus associated with poorer outcomes. In the present study, pulmonary atresia developed in utero in two cases and postnatally in one. The two cases with surgical mortality were from the group with pulmonary atresia.

The prognosis of the baby with tetralogy of Fallot can be improved by prenatal diagnosis of the abnormality which enables planned delivery at a cardiac center where pediatric cardiologists can initiate appropriate management as early as possible. Despite this possibility, many of the parents in our series still opted for termination of pregnancy when the diagnosis was made early enough to make this an acceptable option.

REFERENCES

- 1. Mair DD, Edwards WD, Hagler DJ, Julsrud PR, Puga FJ. Tetralogy of Fallot and pulmonary atresia with ventricular septal defect. In Moller JH, Neal WA, eds. Fetal, Neonatal, and Infant Cardiac Disease. Norwalk: Appleton & Lange, 1990:639-69
- 2. Castaneda AR, Mayer J. Tetralogy of Fallot. In Stark J, de Leval M, eds. Surgery for Congenital Heart Defects. Philadelphia: WB Saunders, 1994:405–16
- 3. Allan L, Sharland GK. Prognosis in fetal tetralogy of Fallot. Pediatr Cardiol 1992;13:1-4
- 4. Lee W, Smith RS, Comstock CH, Kirk JS, Riggs T, Weinhouse E. Tetralogy of Fallot: prenatal diagnosis and postnatal survival. Obstet Gynecol 1995;86:583-8
- 5. Mielke G, Steil E, Kendziorra H, Goelz R. Ductus arteriosusdependent pulmonary circulation secondary to cardiac malformations in fetal life. Ultrasound Obstet Gynecol 1997;9:25-9
- 6. Yoo SJ, Lee Y-H, Kim ES, Ryu HM, Kim MY, Choi HK, Cho KS, Kim A. Three-vessel view of the fetal upper mediastinum: an easy means of detecting abnormalities of the ventricular outflow tracts and great arteries during obstetric screening. Ultrasound Obstet Gynecol 1997;9:173-82
- 7. Yoo SJ, Lee YH, Cho KS, Kim DY. Sequential segmental approach to fetal congenital heart disease. Cardiol Young 1999;9:440-54
- 8. Copel JA, Pilu G, Green J, Hobbins JC, Kleinman CS. Fetal echocardiographic screening for congenital heart disease: the importance of the four-chamber view. Am J Obstet Gynecol 1987;157:648-55

- 9. Sharland GK, Allan LD. Screening for congenital heart disease prenatally. Results of a 2 and 1/2-year study in the south east Thames region. Br J Obstet Gynaecol 1992;99:220-5
- 10. DeVore GR, Siassi B, Platt LD. Fetal echocardiography. VIII. Aortic root dilatation - a marker for tetralogy of Fallot. Am J Obstet Gynecol 1988;159:129-36
- 11. Benacerraf BR, Pober BR, Sanders SP. Accuracy of fetal echocardiography. Radiology 1987;165:847-9
- 12. Bromley B, Estroff JA, Sanders SP, Parad R, Roberts D, Frigoletto FD, Benacerraf BR. Fetal echocardiography: accuracy and limitations in a population at high and low risk for heart defects. Am J Obstet Gynecol 1992;166:1473-81
- 13. Achiron R, Glaser J, Gelernter I, Hegesh J, Yagel S. Extended fetal echocardiographic examination for detecting cardiac malformations in low risk pregnancies. Br Med J 1992;304: 671 - 4
- 14. Wigton TR, Sabbagha RE, Tamura RK, Cohen L, Minogue JP, Strasburger JF. Sonographic diagnosis of congenital heart disease: comparison between the four chamber view and multiple cardiac views. Obstet Gynecol 1993;82:219-24
- 15. Kirk JS, Riggs TW, Comstock CH, Lee W, Yang S, Weinhouse E. Prenatal screening for cardiac anomalies: the value of routine addition of the aortic root to the four-chamber view. Obstet Gynecol 1994;84:427-31
- 16. Rustico MA, Benettoni A, D'Ottavio G, Maieron A, Fischer-Tamaro I, Conoscenti G, Meir Y, Montesano M, Cattaneo A, Mandruzzato G. Fetal heart screening in low-risk pregnancies. Ultrasound Obstet Gynecol 1995;6:313-19
- 17. Shipp TD, Bromley B, Hornberger LK, Nadel A, Benacerraf BR. Levorotation of the fetal cardiac axis: a clue for the presence of congenital heart disease. Obstet Gynecol 1995;85:
- 18. Comstock CH, Riggs T, Lee W, Kirk J. Pulmonary artery-toaorta ratio in the normal and abnormal fetal heart. Am J Obstet Gynecol 1991;165:1038-44
- 19. Hornberger LK, Sanders SP, Sahn DJ, Rice MJ, Spevak PJ, Benacerraf BR, McDonald RW, Colan SD. In utero pulmonary artery and aortic growth and potential for progression of pulmonary outflow tract obstruction in tetralogy of Fallot. J Am Coll Cardiol 1997;25:739-45
- 20. Berning RA, Silverman NH, Villegas M, Sahn DJ, Martin GR, Rice MJ. Reversed shunting across the ductus arteriosus or atrial septum in utero heralds severe congenital heart disease. J Am Coll Cardiol 1996;27:481-6
- 21. Yoo S-J, Choi Y-H. Patent or persistent ductus arteriosus. In Angiocardiograms in Congenital Heart Disease. Oxford: Oxford University Press, 1991:69–75
- 22. Yagel S, Hochner-Celnikier D, Hurwitz A, Palti Z, Gotsman MS. The significance and importance of prenatal diagnosis of fetal cardiac malformations by Doppler echocardiography. Am J Obstet Gynecol 1988;158:272-7
- 23. Ettedgui JA, Sharland GK, Chita SK, Cook A, Fagg N, Allan LD. Absent pulmonary valve syndrome with ventricular septal defect: role of the arterial duct. Am J Cardiol 1990;66:233-4
- 24. Chiappa E, Gaglioti P, Todros T. Rudimentary pulmonary valve and ventricular septal defect associated patent ductus arteriosus. Pediatr Cardiol 1995;16:190-3
- 25. Rowland DG, Caserta T, Foy P. Wheller JJ, Allen HD. Congenital absence of the pulmonary valve with tetralogy of Fallot with associated aortic stenosis and patent ductus arteriosus: a prenatal diagnosis. Am Heart J 1996;132:1075-7
- 26. Rice MJ, McDonald RW, Reller MD. Progressive pulmonary stenosis in the fetus: two case reports. Am J Perinatol 1993; 10:424-7