Outcomes of prenatally diagnosed tetralogy of Fallot: Implications for valve-sparing repair versus transannular patch

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OBJECTIVES: To assess outcomes of prenatally diagnosed tetralogy of Fallot and determine factors associated with the choice to undergo a valve-sparing repair versus transannular patch, and the use of prostaglandins at birth.

METHODS: All cases at The Hospital for Sick Children (Toronto, Ontario) with a fetal diagnosis of tetralogy of Fallot from 1998 to 2006, were reviewed for demographic and fetal echocardiographic data to determine factors associated with the valve-sparing repair and need for perinatal support.

RESULTS: Sixty-four fetuses met inclusion criteria (median gestational age 22 weeks) with 47 live births. Twenty-six underwent valve-sparing repair (median age 5.7 months) and 14 underwent transannular patch repair (median age 4.5 months). There were seven deaths before surgery and one post-transannular patch repair. One patient required a transannular patch repair after the initial valve-sparing repair. Twelve of 29 (41%) patients received prostaglandins at birth. Type of surgical repair, use of prostaglandins and postnatal death were among the outcomes investigated. The mean pulmonary valve (PV) z-score was -3.0 ± 2.0 and the mean PV/aortic valve (AoV) ratio was 0.65 ± 0.10 . Lower PV z-score ($P=0.04$), smaller PV/AoV ratio ($P=0.04$) and the presence of nonantegrade arterial duct flow ($P=0.02$) were associated with prostaglandin use. A higher PV/AoV ratio was associated with valve-sparing repair ($P=0.04$). Fetal z-scores of the PV, AoV and right pulmonary artery at 29 to 32 weeks gestational age correlated with respective postnatal z-scores ($P=0.01$).

CONCLUSION: Fetal echocardiographic variables were associated with the use of prostaglandins and valve-sparing repair in fetuses with tetralogy of Fallot, and at 29 weeks, correlated with postnatal valve diameters.

Key Words: Echocardiography; Fetal; Prenatal; Tetralogy of Fallot; Valve-sparing

Tetralogy of Fallot is a common form of congenital heart disease amenable to full surgical repair. Although transannular patch repair has a good short-term outcome, there is mounting concern that long-standing severe pulmonary insufficiency may lead to progressive right-sided heart failure, reduced functional status and reduced life expectancy (1-3). Many patients require a pulmonary valve (PV) implantation later in life to relieve the symptoms of exercise intolerance and right heart failure (4-8). A newer surgical approach, valve-sparing repair, shows promise as a potentially superior option to

Les issues de la tétralogie de Fallot diagnostiquée pendant la période prénatale : Les conséquences d'une réfection épargnant les valves par rapport à une pièce transannulaire

OBJECTIFS : Évaluer les issues de la tétralogie de Fallot diagnostiquée pendant la période prénatale et déterminer les facteurs associés au choix d'entreprendre une réfection épargnant les valves plutôt que de poser une pièce transannulaire, ainsi que d'utiliser des prostaglandines à la naissance.

MÉTHODOLOGIE : Les auteurs ont revu tous les cas de diagnostic fœtal de tétralogie de Fallot au *Hospital for Sick Children* (Toronto, Ontario) entre 1998 et 2006 afin d'obtenir les données démographiques et échocardiographiques fœtales pour déterminer les facteurs associés à la réfection épargnant les valves et le besoin d'un soutien périnatal.

RÉSULTATS : Soixante-quatre fœtus respectaient les critères d'inclusion (âge gestationnel médian de 22 semaines), qui incluait 47 naissances vivantes. Vingt-six ont subi une réfection épargnant les valves (âge médian de 5,7 mois) et 14, une réfection par pièce transannulaire (âge médian de 4,5 mois). Sept décès ont eu lieu avant l'opération et un après la réfection par pièce transannulaire. Un patient a dû subir une réfection par pièce transannulaire après la réfection épargnant les valves. Douze des 29 patients (41 %) ont reçu des prostaglandines à la naissance. Le type de réfection chirurgicale, le recours aux prostaglandines et les décès postnatals faisaient partie des issues explorées. L'écart réduit moyen de la valeur pulmonaire (VP) était de $-3,0 \pm 2,0$ et le ratio moyen entre la VP et la valve aortique (VAo), de $0,65 \pm 0,10$. L'écart réduit inférieur de la VP ($P=0,04$), le plus petit ratio entre la VP et la VAo ($P=0,04$) et la présence d'un débit du canal artériel non antérograde ($P=0,02$) s'associaient à l'utilisation de prostaglandines. Un ratio plus élevé entre la VP et la VAo s'associait à une réfection épargnant les valves ($P=0,04$). Des écarts fœtaux réduits de la VP, de la VAo et de l'artère pulmonaire droite entre 29 et 32 semaines d'âge gestationnel étaient corrélés avec des écarts postnatals réduits respectifs ($P=0,01$).

CONCLUSION : Les variables échocardiographiques fœtales s'associaient à l'utilisation de prostaglandines et d'une réfection épargnant les valves chez des fœtus ayant une tétralogie de Fallot et, à 29 semaines, étaient corrélées avec le diamètre postnatal des valves.

transannular patch repair. Although fetal echocardiography has become an essential diagnostic tool when congenital heart disease is suspected, we are still limited in our ability to accurately predict postnatal outcome based on fetal data (9).

The purpose of the present study was to determine fetal echocardiographic factors associated with optimal surgical repair in this population, and determine fetal echocardiographic factors associated with the use of prostaglandins at birth in fetuses diagnosed with tetralogy of Fallot. Optimal surgical repair was defined as valve-sparing repair (10).

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Figure 1) Outcome of fetuses prenatally diagnosed with tetralogy of Fallot or tetralogy-type configuration. TAP Transannular patch

METHODS

Inclusion and exclusion criteria

All cases at The Hospital for Sick Children in Toronto, Ontario, with a fetal diagnosis of typical tetralogy of Fallot, or double-outlet right ventricle with pulmonary stenosis and ventricular septal defect (tetralogy-type), between January 1998 and May 2006, were identified from the fetal cardiac database. These were reviewed for clinical and fetal echocardiographic data to determine factors associated with valve-sparing repair. The present study was approved by the Research Ethics Boards at The Hospital for Sick Children and Mount Sinai Hospital in Toronto.

Fetuses with concomitant pulmonary atresia, major aortopulmonary collateral arteries with no pulmonary arterial flow, absent PV, atrioventricular septal defect, atrial isomerism, ventriculoarterial discordance, hypoplastic left ventricle or aortic stenosis at the time of first echocardiography were excluded from the study.

Demographic data

All medical charts were reviewed to determine the spectrum of anomalies and outcomes. Fetal echocardiogram reports, videotapes and/or digital studies were reviewed. Adverse outcomes were defined as post-natal death, in utero demise, use of prostaglandins at birth, postoperative complications such as arrhythmias, cardiac dysfunction and residual cardiac lesions.

Fetal echocardiographic measurements

Two-dimensional echocardiograms were reviewed offline by a single investigator (AB) blinded to patient outcome. Archived videotapes were reviewed using the Vingmed System Vivid-7 (GE Medical Systems, GE Vingmed Ultrasound AS, Norway). Digital echocardiograms were reviewed using KinetDx version 3.1 (Siemens Medical Solutions, USA). Two separate measurements were taken for the tricuspid valve, the PV and the aortic valve (AoV), branch pulmonary arteries and pulmonary trunk in each view, including the outflow tract and sagittal views. Given the variability of fetal echocardiography views, the maximal measurement of valve size was chosen. Doppler measurements were recorded when available for the arterial duct and right ventricular outflow tract. Significant right outflow obstruction was defined as turbulent flow at the subvalve and/or PV levels of greater than 80 cm/s. Ductus arteriosus flow was defined as right-to-left, left-to-right or bidirectional. Valvular regurgitation was defined as none-to-mild or moderate-severe. Ventricular function was qualitatively defined as good, moderately reduced or poor. Progression of disease was defined as the development of flow reversal or bidirectional flow in the ductus arteriosus, the development of pulmonary atresia, the development of turbulent flow in the right outflow tract, or

Doppler gradients of 80 cm/s or greater. The z-scores were calculated for the PV, tricuspid valve and AoV based on the methods detailed by Schneider et al (11). Postnatal z-scores were calculated using either body surface area, or both height and weight measured at the time of the first echocardiogram.

Statistical analysis

Data are presented as means $\pm$ SDs, medians with maximum and minimum values, or frequencies as appropriate. Associations among transannular patch repair, use of prostaglandins at birth and postoperative morbidity, with demographics, gestational, physiological and medical characteristics were sought using Student's *t* test, and Fisher's exact, χ^2 and linear regression models. Changes over time in echocardiography measurements were estimated through a mixed linear regression model adjusted for repeated measures. Survival analysis was performed using Kaplan-Meier nonparametric estimates. All statistical analyses were performed using SAS statistical software version 9.1 (SAS Institute Inc, USA). $P < 0.05$ was considered significant.

RESULTS

The outcomes of all pregnancies are shown in Figure 1. During the study period, there were 461 cases of prenatally and postnatally diagnosed tetralogy of Fallot at The Hospital for Sick Children. Of those, 87 (19%) were diagnosed prenatally. Seventy-six had typical tetralogy of Fallot and 11 had tetralogy-type configuration. Twenty-three cases were excluded from the study, including 12 with pulmonary atresia, three with an atrioventricular septal defect, three with an absent PV, two with dextrotransposition of the great arteries, one with a hypoplastic left ventricle, one with right atrial isomerism and one with aortic stenosis diagnosed at the first echocardiogram. As a result, a total of 64 cases were available. Of these, 10 fetuses were terminated, two of which were of twin pregnancies, six had additional abnormalities and four cases were otherwise normal. The six fetuses with additional abnormalities had omphalocele ($n=1$), talipes ($n=1$), pentology of Cantrell ($n=2$) and trisomy 18 ($n=2$). In addition, four pregnancies resulted in in utero demise, and three were lost to follow-up before birth.

The median gestational age at the time of the initial fetal echocardiogram was 22 weeks (range 18 to 35 weeks). Forty-six pregnancies were singletons, and one was a twin pregnancy in which the other fetus had a normal heart and a normal amniocentesis. The most common reason for referral to the centre was suspected congenital heart disease on screening obstetrical ultrasound.

Live born characteristics are presented in Table 1. Additional cardiac anomalies were common, with a right aortic arch detected most frequently (22%). The median age at surgical repair was 5.3 months. Those who underwent a transannular patch repair did so earlier (median age 4.5 months, range 46 days to 23.2 months) than those who underwent a valve-sparing repair (median age 5.7 months, range 23 days to 9.9 months). One patient did not undergo surgical repair until 23 months of age because the parents initially refused repair. A valve-sparing repair was initially attempted; however, high right ventricular pressures once the patient was off cardiopulmonary bypass prompted the surgeon to immediately reintervene with a transannular patch.

Fetal and postnatal parameter correlation

Right pulmonary artery, AoV and PV values measured in echocardiograms taken between 29 and 32 weeks gestational age were found to have a strong correlation with their respective postnatal measurements taken on the first echocardiogram (median age two days, range zero to six months). Figure 2 illustrates the correlation between prenatal and postnatal values.

Postsurgical follow-up

The median duration of follow-up following surgery was 12 months (range three days to 5.1 years), with follow-up to December 2006. At

TABLE 1
Clinical characteristics of live-born infants (n=47)

Characteristic	
Male:female, n/n	21/26
GA at diagnosis, weeks, median (range)	22 (18–35)
GA at birth, weeks, median (range)	36 (29–41)
Birth weight, g, median (range)	2660 (780–4470)
Fetal diagnosis, n	
Tetralogy of Fallot	41
Tetralogy-type	6
Prostaglandins given at birth	12/29 (41)
Ventilated at birth	14/31 (45)
Abnormal chromosomes	9/47 (19)
Microdeletion 22, n	3
Trisomy 21, n	2
Trisomy 13, n	2
Trisomy 18, n	1
3/21 translocation (unbalanced), n	1
Additional cardiac anomalies	17/45 (38)
Right aortic arch, n	10
LSVC to coronary sinus, n	5
Branch PA stenosis, n	4
Extracardiac abnormalities	15/30 (50)
Omphalocele, n	5
Hydronephrosis, n	3
Tracheoesophageal fistula, n	3
Craniofacial, n	2
Orchidism, n	2

Data presented as n/n (%) unless otherwise indicated. Denominators indicate the number of patients with available data. GA Gestational age; LSVC Left superior vena cava; PA Pulmonary artery

the most recent clinic visit, 25 of 31 (71% of the valve-sparing repair group and 92% of the transannular patch repair group) had moderate to severe pulmonary insufficiency, 10 (29%) had a residual atrial or ventricular shunt and seven (20%) were on cardiac medications. Three patients were lost to follow-up following surgery because they returned to their countries of origin.

Postoperative complications within both groups included pleural effusions in 10, chylothorax in five and arrhythmia in eight, all of which were supraventricular in origin. Only two patients continue to require medical treatment for their arrhythmia. There was no significant difference in the frequency of complications between the two groups.

Freedom from reintervention for the valve-sparing repair group from the time of repair was 100% at six months and one year, and 80% at three years, while in the transannular patch group, freedom from reintervention remained at 88% from six months and beyond. A Kaplan-Meier survival curve for all live born infants is shown in Figure 3, indicating an overall survival of 82% from three months and beyond. The overall survival for the live born infants without documented chromosomal anomalies was 94% at one month, and 88% at three months and beyond.

Factors associated with a valve-sparing repair and neonatal support

Live born infants included in the analysis of predictors for valve-sparing repair versus transannular patch repair were those who underwent surgical intervention (n=40). Analysis of the use of prostaglandins at birth included those who underwent surgical intervention (n=40) and those who died postnatally (n=7). Fetal echocardiographic results comparing valve-sparing repair versus transannular patch repair and the use of prostaglandins at birth are shown in Tables 2 and 3, respectively. All measurements were taken from the initial fetal echocardiogram.

Figure 2) Correlation of echocardiographic z-scores for the right pulmonary artery, aortic and pulmonary valves ($R^2=0.20$, 0.33 and 0.21 , respectively) at 29 to 32 weeks gestational age and initial postnatal imaging

Figure 3) Kaplan-Meier curve displaying survival from birth to six years for all live borns (n=47)

TABLE 2
Univariate associations between echocardiographic variables and valve-sparing repair over transannular patch in live born infants with tetralogy of Fallot or tetralogy-type configuration (n=40)

	Valve-sparing repair (n=26)		Transannular patch repair (n=14)		P
	Mean $\pm$ SD	n	Mean $\pm$ SD	n	
PT/AoV ratio	0.66 $\pm$ 0.14	24	0.57 $\pm$ 0.12	14	0.05
PV/AoV ratio	0.65 $\pm$ 0.10	23	0.57 $\pm$ 0.12	12	0.04

AoV Aortic valve; PT Pulmonary trunk; PV Pulmonary valve

In our cohort, there was information available on prostaglandin use in 29 of 47 patients. The mean PV z-score by fetal echocardiography for the entire cohort was -3.0 ± 2.0 compared with a PV z-score of -4.5 ± 2.2 for the infants who received prostaglandins. The mean tricuspid valve z-score was -1.0 ± 1.5 and the mean AoV z-score was 2.4 ± 1.5 for the entire cohort. Figure 4 shows a box plot representation of mean PV/AoV ratios in both groups and Figure 5 shows a graph of the probability of undergoing a valve-sparing procedure based on the PV/AoV ratio.

TABLE 3

Univariate associations between echocardiographic variables and the use of prostaglandins at birth in live born infants with tetralogy of Fallot or tetralogy-type configuration (n=29)

	No PGE (n=17)		PGE (n=12)		P
	Value	n	Value	n	
Fetal arterial duct shunt*, n (%)	0 (0)	16	3 (33)	9	0.04
PV/AoV ratio	0.64±0.10	17	0.55±0.12	10	0.03
PT z-score	-2.2±1.3	16	-3.4±1.0	10	0.02
PV z-score	-2.3±2.8	16	-4.2±2.0	10	0.04

Data presented as mean ± SD unless otherwise indicated. *Retrograde/bidirectional versus antegrade arterial duct shunt. AoV Aortic valve; PGE Prostaglandins; PT Pulmonary trunk; PV Pulmonary valve

Figure 4) Pulmonary/aortic valve ratios for the valve-sparing and transannular patch groups. Box plots show the 25th, 50th and 75th percentiles. Minimum and maximum values are denoted, and mean values of each group are represented by a '+'

Factors associated with postnatal death

Table 4 outlines the factors associated with postnatal death. Seven deaths occurred before surgery, all due to noncardiac-related causes (two trisomy 13, one trisomy 18, one 3/21 translocation, one necrotizing enterocolitis, one lung hypoplasia and one liver failure). Of the 40 patients who underwent surgical intervention, 26 underwent an initial valve-sparing repair and 14 an initial transannular patch repair. One patient with a significantly stenotic left pulmonary artery underwent initial valve-sparing repair at three months of age. At the follow-up echocardiogram, there was asymptomatic fibromuscular right outflow obstruction and it was decided that the patient would undergo conversion to transannular patch repair at 18 months.

There was only one surgical death, which occurred in the transannular patch repair group. This patient was born at 41 weeks gestation with tetralogy of Fallot and concomitant aortic stenosis that had progressed during the pregnancy, but was not found at the initial 20-week fetal echocardiogram. The patient underwent balloon dilation of both the AoV and PV within a week of birth, and then required a second catheterization for stenting of the right ventricular outflow tract. At 3.5 months of age, the patient underwent a transannular patch repair

Figure 5) Probability of undergoing a valve-sparing procedure based on the pulmonary to aortic valve ratio derived from a logistic regression model of the procedure (valve-sparing or transannular patch) over the pulmonary to aortic valve ratio

TABLE 4

Kaplan-Meier estimates of univariate associations between echocardiographic variables and postnatal death in fetuses with tetralogy of Fallot or tetralogy-type configuration

Death (n=8)	EST±SE	P	HR
Younger gestational age at birth, weeks	0.491±0.198	0.02	1.61
Lower birth weight, kg	1.804±0.766	0.02	5.88
Extracardiac anomalies	2.396±1.119	0.04	10.97
Fetal arterial duct shunt	2.821±1.021	0.006	16.79
Smaller ascending aorta z-score	0.672±0.299	0.03	1.96
Chromosome anomalies	1.742±0.714	0.02	5.71

EST ± SE Estimate ± standard error (change in time-related survival with an increase of one unit in the independent variable); HR Hazard ratio (increase in time-related hazard of death by an increase of one unit in the independent variable)

due to persistent high pulmonary artery pressures; however, the patient died postoperatively.

Predictors of in utero demise

Within the sample, four fetuses died in utero at a median gestational age of 25 weeks (range 23 to 28 weeks). There was no obvious preceding arrhythmia in any of the cases. Of these fetuses, one had trisomy 14. A second case was a twin pregnancy in which the affected twin had concomitant esophageal atresia with normal chromosomes. A third fetus had Dandy-Walker syndrome, a massive cystic hygroma and bilateral pleural effusions. The fourth fetus had trisomy 21 and had no fetal heart activity at 26 weeks. Of these four fetuses, three had moderate to severe tricuspid regurgitation (TR), which was a significant predictor of in utero demise (P=0.03) compared with the live born infants.

Progression of disease in utero

Repeated fetal echocardiographic measurements were assessed for progression over time. Of the 62 patients for which an initial echocardiography was provided, 32 (52%) had one follow-up echocardiogram and 13 (21%) had more than two serial echocardiograms. Twelve fetuses demonstrated progression of the right outflow obstruction, while six fetuses improved and 13 did not demonstrate any significant change in severity of obstruction. One fetus initially diagnosed with pulmonary stenosis at the 20-week echocardiogram progressed to

pulmonary atresia by 25 weeks. In this fetus, flow through the ductus arteriosus was retrograde. One fetus developed progressive severe AoV stenosis. Of the 30 patients who had more than one echocardiogram, 26 had normal flow in the ductus arteriosus in both the first and last echocardiograms. There were no significant changes in echocardiographic parameters using mixed logistic regression analysis adjusted for repeated measures.

DISCUSSION

Tetralogy of Fallot is a common form of cyanotic congenital heart disease with a generally favourable outcome (12). However, suboptimal outcomes, and fetal or perinatal demise are not uncommon (13). As the field of fetal echocardiography grows, the ability to predict outcomes rather than simply diagnose a cardiac lesion becomes increasingly important.

Our results indicate that measurements derived from an echocardiogram taken between 29 and 32 weeks gestational age can predict initial postnatal AoV, PV and right pulmonary artery sizes. This demonstrates that a routine echocardiogram at 29 weeks gestational age in this population may be beneficial in predicting valve size and allowing for risk stratification for surgery type and time.

Factors associated with optimal surgical repair

Previous studies have suggested that sparing the PV represents the best surgical outcome for patients with tetralogy of Fallot (1,10,14). The focus of our study was to determine echocardiographic factors associated with postnatal outcome. In our cohort, 26 (65%) of 40 patients underwent valve-sparing repair, and a higher PV/AoV ratio at the initial fetal echocardiogram was associated with this outcome. Although there was an overlap between the valve-sparing repair and transannular patch groups in terms of the PV/AoV ratio, our findings suggested that a PV/AoV ratio of less than 0.65 was associated with the fetus being more likely to receive a transannular patch repair after birth. In a previous fetal study of 16 fetuses with tetralogy of Fallot by Hornberger et al (15) in 1995, progression of right outflow obstruction was found when the main pulmonary artery to ascending aorta ratio was less than 0.5 ($P=0.01$). Our series found that a ratio of less than 0.65 likely indicated more severe stenosis at birth and the need for transannular patch repair.

Factors associated with neonatal support

Forty-one per cent (12 of 29) of the live born infants were given prostaglandins. By using the PV/AoV ratio, PV z-score and the direction of ductal flow at initial presentation as predictors, we could potentially risk stratify patients requiring prostaglandins and plan perinatal care accordingly. The PV/AoV ratio at the first echocardiogram was significantly correlated with outcomes in our analysis, and appears to be independent of the direction of ductal flow.

It is clear that a smaller, more dysplastic PV eventually leads to more pulmonary outflow obstruction. The sequence of progression is not known. We anticipate that using the PV/AoV ratio in conjunction with flow across the ductus arteriosus, PV z-score and degree of right ventricular outflow obstruction would provide insight into the mechanisms of progression of disease in utero.

Factors associated with death

Only one of the eight deaths could be attributed to the underlying cardiac diagnosis, which included the rare finding of concomitant aortic stenosis. The other seven deaths were related to severe chromosomal or extracardiac abnormalities. Ductal shunting and smaller aortic scores thus appear to be fortuitous findings and cannot be used as factors predicting death, given the significant number of chromosomal and extracardiac abnormalities in the group. We recommend that chromosomal testing be offered to all mothers with a fetus prenatally diagnosed with tetralogy of Fallot due to the high incidence of chromosomal abnormalities and its association with death.

Factors associated with in utero demise

In our study, moderate to severe TR was a predictor of fetal demise. All four cases had major additional abnormalities and died in the late second to early third trimester. Indeed, severe TR may be attributed to the inherent abnormality of the tricuspid valve, and is well reported in association with specific congenital cardiac lesions (16,17). In otherwise normal cases with tetralogy of Fallot, we can counsel families that intrauterine demise is unlikely. In cases of prenatally diagnosed chromosomal and extracardiac abnormalities, parents should be counselled regarding the higher risk of demise later in gestation.

The findings of our study suggest that not only is it important to make the diagnosis, but it is now possible that fetal parameters such as PV/AoV ratio, PV size and flow dynamics are of value in predicting the use of prostaglandins at birth and the surgical approach. More diligent measurements of Doppler flow patterns might also clarify the etiology of progression of disease in tetralogy of Fallot. This can significantly aid in our understanding of fetal physiology as well as in the prenatal counselling of these families.

Study limitations

The study was limited by the relatively small number of cases in both groups, and by the retrospective nature of the study. Doppler patterns were not consistently measured and we did not have consistent follow-up fetal echocardiograms to determine factors that progressed in utero. Due to these limitations, a prospective multicentre trial is underway to validate our findings in a larger sample population with the consistent use of Doppler and follow-up echocardiograms. We chose valve-sparing repair as our primary end point because previous data have suggested its superiority over the transannular patch repair, although this is not conclusively proven. Interestingly, both interventional groups in follow-up revealed significant pulmonary insufficiency (71% of the valve-sparing repair group and 92% of the transannular patch repair group; $P=0.69$). While other studies have found significantly reduced rates of pulmonary insufficiency in the valve-sparing procedure (1,10,14), our study was not powered to illustrate this difference. Furthermore, a greater understanding of the natural history of pulmonary insufficiency and its deleterious sequelae is likely to be of value. We did not have information on prostaglandin use in all cases. The use of prostaglandins at birth may have been biased by the fetal cardiologist's recommendations to the obstetrician, or by the decision of the obstetrician or neonatologist at the time of delivery, and may not be an accurate indicator of disease severity.

CONCLUSIONS

Prenatally diagnosed tetralogy of Fallot has a good outcome with low short- and mid-term morbidity and mortality following valve-sparing repair or transannular patch repair in fetuses and live born infants who are otherwise normal. The mortality rate is high among cases with chromosomal and/or extracardiac anomalies. An echocardiogram performed at 29 weeks has a good correlation with initial postnatal AoV and PV diameters. A larger PV/AoV ratio was found to be associated with a valve-sparing repair, while a smaller ratio, abnormal flow in the ductus arteriosus and smaller PV z-score were found to be prenatal predictors of the use of prostaglandins at the time of delivery. These findings need to be validated prospectively but will ultimately have an impact on prenatal counselling and management.

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