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Tetralogy of Fallot With Diminutive Pulmonary Arteries: Preoperative Pulmonary Valve Dilation and Transcatheter Rehabilitation of Pulmonary Arteries

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Objectives. **This study sought to determine the results of a novel transcatheter management approach in tetralogy of Fallot with diminutive pulmonary arteries.**

Background. **Tetralogy of Fallot with diminutive pulmonary arteries and severe pulmonary stenosis is rare and resembles tetralogy of Fallot with pulmonary atresia: There is a high incidence of aortopulmonary collateral channels, arborization abnormalities, stenoses and need for multiple operations. Because a combined catheter–surgery approach facilitates repair in these patients, such an approach may benefit those with diminutive pulmonary arteries and pulmonary stenosis.**

Methods. **Clinical, catheterization and surgical data were studied retrospectively for 10 such patients undergoing preoperative pulmonary valve balloon dilation, among other transcatheter interventions, from January 1989 to January 1995.**

Results. **Initially, the Nakata index ranged from 20 to 98 mm^2/m^2 (mean 67 ± 28 mm^2/m^2). The pulmonary valve was first balloon dilated (mean balloon/annulus 1.5 ± 0.3), and the mean initial valve annulus Z score (−4.0 ± 1) increased to −3.3 ± 1.1 ($p < 0.01$). Other interventions included branch pulmonary artery balloon dilation (7 patients, 23 vessels) and coil embolization of aortopulmonary collateral channels (8 patients, 31 collateral channels). At preoperative follow-up catheterization, the mean pulmonary annulus Z score was −3.1 ± 0.7, and the Nakata index increased to 143 ± 84 mm^2/m^2 ($p < 0.03$). All patients underwent complete surgical repair successfully. At a mean follow-up period of 2.6 ± 2 years, right ventricular pressure was <70% systemic in all patients and <50% systemic in seven.**

Conclusions. **In patients with tetralogy of Fallot, severe pulmonary stenosis and diminutive pulmonary arteries, initial pulmonary valve balloon dilation increases the annulus Z score and anterograde pulmonary blood flow and facilitates simultaneous coiling of aortopulmonary collateral channels and access for branch pulmonary artery dilation, all of which results in pulmonary artery growth, simplifying surgical management.**

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There is a wide spectrum of disease in tetralogy of Fallot, ranging from mild forms of infundibular obstruction and pulmonary valve stenosis to pulmonary atresia with hypoplastic or even absent pulmonary arteries (1–5). Most patients with tetralogy of Fallot can undergo complete primary repair during infancy (6–8) without need for a preliminary palliation. In contrast, patients with rare, severe forms with diminutive pulmonary arteries in whom the pulmonary blood flow is derived primarily from collateral channels usually need multiple interventions before complete repair (9,10) because the central pulmonary arteries may be inadequate to carry a full cardiac output after ventricular septal defect closure. Some have even advocated lifelong medical management for these patients (11).

Since 1984, we have managed most patients with tetralogy of Fallot, pulmonary atresia and diminutive pulmonary arteries with an initial right ventricle-to-pulmonary artery surgical graft, followed by balloon dilation of pulmonary artery stenoses and coil embolization of aortopulmonary collateral channels and, later, surgical closure of the ventricular septal defect in addition to repair of any remaining central obstructions (7,9). Such a management approach, combining interventional catheterization and operation, seems to enhance the chances of achieving a satisfactory repair (9).

On the basis of this favorable experience, we dilated the pulmonary valve in a group of 10 patients with severe tetralogy of Fallot, diminutive pulmonary arteries or multiple aortopulmonary collateral channels, thought to be unsuitable for primary complete surgical repair on presentation. In these patients, pulmonary valve dilation was an initial step for transcatheter pulmonary artery "rehabilitation," followed by coil embolization of aortopulmonary collateral channels, dila-

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tion of distal pulmonary artery stenosis and subsequent complete surgical repair.

Methods

Study group. All unoperated patients with tetralogy of Fallot who underwent any preoperative catheterization from January 1984 to January 1995 were identified from the Department of Cardiology data base. Patients with angiographic diagnosis of pulmonary artery hypoplasia or branch pulmonary artery stenosis were reviewed. Of those, patients with diminutive pulmonary arteries (Nakata index < 100 mm^2/m^2) who had undergone pulmonary valve balloon dilation as initial intervention were selected and constitute the study group. Because most patients with usual tetralogy of Fallot undergo primary repair after a diagnostic echocardiogram without the need for a previous catheterization, such patients were not included in this review. Patients with any previous surgical intervention (such as Blalock-Taussig shunts) or with diagnoses of pulmonary atresia, absent pulmonary valve syndrome, associated other major cardiac malformations (such as endocardial cushion defects, abnormalities in the ventricular loop, transposition of the great arteries, heterotaxia syndromes or hypoplastic ventricles) were excluded.

Medical records, echocardiograms and angiographic and hemodynamic data were reviewed in all patients from the time of presentation to the most recent follow-up date. Approval for chart review was obtained from the Institutional Review Board.

Cardiac catheterization. At initial catheterization, after routine right- and left-sided hemodynamic analysis, patients had determination of 1) pulmonary valve annulus Z score before and after balloon dilation; 2) right and left pulmonary artery Z scores; 3) Nakata index; 4) McGoon ratio (12); 5) presence of aortopulmonary collaterals or patent ductus arteriosus; 6) oxygen saturation before and after pulmonary valve balloon dilation; and 7) other transcatheter interventions. The pulmonary artery size was determined by measuring the right and left pulmonary artery diameters immediately proximal to their first branch points at the initial angiogram. Magnification errors were corrected by relating vessel size to the known diameter of the angiographic catheter. The Nakata index was reported for each patient as the sum of the cross-sectional areas of both pulmonary arteries indexed to the body surface area (13). The normal Nakata index is 330 ± 30 mm^2/m^2. Pulmonary arteries with Nakata index less than 100 mm^2/m^2 were considered diminutive. The Z values obtained were based on the reported definition of Z = observed dimension minus mean normal dimension/standard deviation (14). Main pulmonary artery pressure and oxygen saturation were determined before and after dilation in some patients, with the exception of those with such severe stenosis that no pressure-recording catheter could be advanced across the valve before dilation. The presence of other sources of pulmonary blood flow by aortopulmonary collaterals or a patent ductus was determined by aortography or selective collateral injection.

At later preoperative catheterization, patients had repeat determination of 1) pulmonary valve annulus Z score; 2) right and left pulmonary artery Z scores; and 3) Nakata index. The initial measurements at the first catheterization before any intervention were compared with those immediately following transcatheter intervention and those at a preoperative catheterization.

Transcatheter interventions performed at preoperative catheterizations included pulmonary valve balloon dilation as previously described (15,16), branch pulmonary artery balloon dilation (17–20), coil embolization of aortopulmonary collaterals (21) in most patients and stent placement in the right ventricular outflow tract in one patient.

Parental informed consent was obtained before every procedure performed.

Operation. Operative notes and immediate and late postoperative results were reviewed. The surgical technique used for tetralogy of Fallot repair has been well described previously (22).

Follow-up. Follow-up information was obtained from hospital records and from referring physicians. At the latest follow-up, the results of electrocardiograms, echocardiograms, oximetry, available catheterizations and symptoms were reviewed. Right ventricular pressure by noninvasive (echocardiogram and Doppler) or invasive methods (cardiac catheterization) was determined in all patients at follow-up.

Statistical analysis. Results obtained are expressed as mean value ± SD. Pulmonary valve annulus diameter and right and left pulmonary arteries were expressed as Z scores (number of standard deviations from the mean normal for body surface area) (14,23). Initial results, immediate results after balloon dilation and late results at preoperative follow-up catheterization were compared and tested with a paired *t* test.

Results

Patient characteristics. Ten patients with diminutive pulmonary arteries (17 days to 12 years old, mean age 1.9 ± 3.7) underwent pulmonary valve balloon dilation as initial preoperative transcatheter intervention since June 1989. There were four boys and six girls. Age, weight and oxygen saturation at pulmonary valve balloon dilation, age at surgical correction and follow-up period after operation are summarized in Table 1.

Among associated anomalies were right aortic arch in four patients, aberrant right subclavian artery in one and restrictive ventricular septal defect with suprasystemic preoperative right ventricular pressures in three patients. Partial Di George syndrome was diagnosed in one patient. One patient had a persistent left superior vena cava to the coronary sinus.

Cardiac catheterization. *Pulmonary artery characteristics.* All patients had more than one catheterization, with an average total number of 2.6 per patient. All patients had diminutive pulmonary arteries (Fig. 1 and 2) with an average Nakata index of 67 ± 28 mm^2/m^2, ranging from 20 to

Table 1. Patient Characteristics

Pt No.	Age at BD (yr)	Gender	Weight (kg)	Oxygen Sat. (%)	Age at Operation (yr)	Postoperative Follow-Up (yr)
1	0.3	Male	5.6	82	0.8	4.5
2	0.2	Male	4.5	84	0.3	4
3	0.4	Female	5.9	64	1.2	4.2
4	2.5	Female	12.9	78	2.6	4.3
5	0.4	Male	5.9	82	0.5	3.9
6	0.8	Female	6.5	66	1.5	0.6
7	12	Female	30	89	12.3	0.3
8	0.1	Male	3.1	88	1.5	0.3
9	1.8	Female	9.2	78	3.4	3.2
10	0.2	Female	6	76	0.5	0.2
Average ± SD	1.9 ± 3.7		8.9 ± 7	78.7 ± 8	2.5 ± 3.6	2 ± 2.1

BD = pulmonary valve balloon dilation; Oxygen Sat. (%) = percent oxygen saturation at rest in room air, before any interventions; Pt = patient.

Figure 1. A, Right pulmonary artery angiogram demonstrates diminutive pulmonary artery. Note that contrast in right lower lobe is being washed by competing collateral flow. B, Selective angiogram of aortopulmonary collateral flow to left lower lobe demonstrates its dual supply with filling of diminutive central pulmonary arteries (arrow).

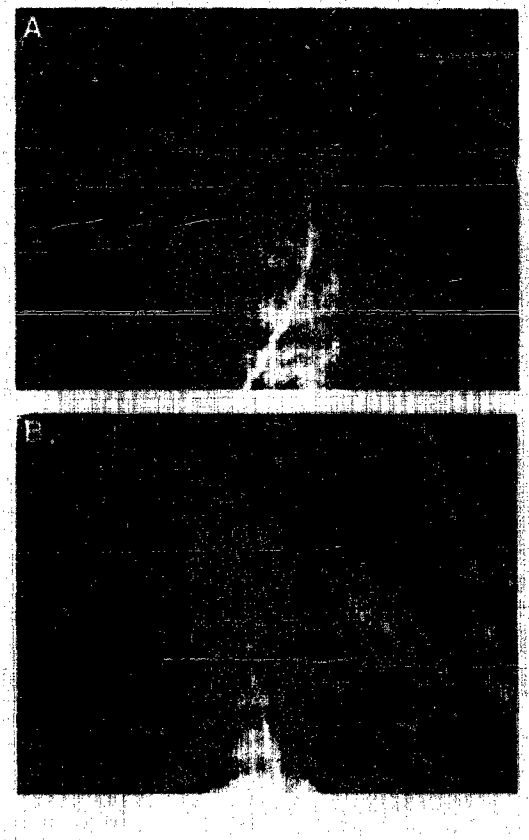


Figure 2. A, Before interventions. Pulmonary artery angiogram at initial catheterization demonstrates hypoplastic pulmonary arteries with severe stenosis of the proximal left pulmonary artery. Note diminished filling of left lung and right lower lobe as contrast is washed by aortopulmonary collateral flow. The pulmonary valve annulus is markedly hypoplastic with thickened leaflet (arrow). B, After interventions. Pulmonary artery angiogram of the same patient as A, after all transcatheter interventions and operation, demonstrates marked improvement in vessel dimensions and flow. Note that the aortopulmonary collateral channels have been coil embolized.

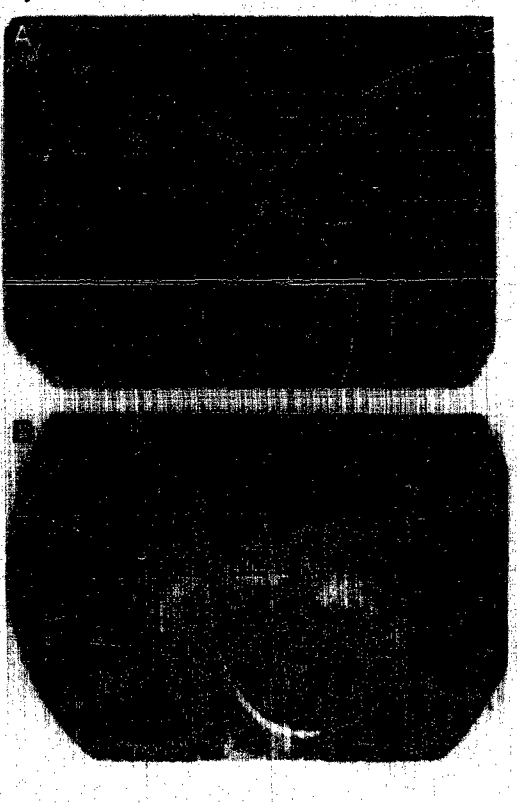


Table 2. Angiographic Characteristics and Summary of Transcatheter Interventions

Pt No.	Nakata Index (mm^2/m^2)	McGoon Ratio	MAPCs	Branch PA Stenosis BD	Coil MAPCs	Other Interventions
1	92	1.2	No	No	No	ASD device
2	81	0.9	Yes	No	Yes	No
3	98	0.8	Yes	Yes	Yes	No
4	69	1	Yes	Yes	Yes	No
5	91	0.6	Yes	No	Yes	No
6	28	0.6	Yes	Yes	Yes	No
7	80	0.7	Yes	Yes	Yes	No
8	42	0.9	Yes	Yes	Yes	Stent RVOT
9	20	0.6	No	Yes	No	ASD device
10	69	0.8	Yes	Yes	Yes	No
Average ± SD	67 ± 28	0.8 ± 0.2				

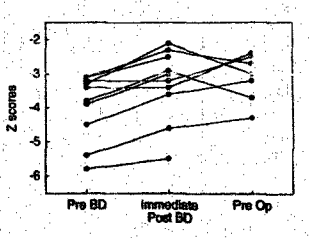
ASD = atrial septal defect; BD = balloon dilation; MAPCs = multiple aortopulmonary collateral channels; McGoon Ratio = Right pulmonary artery (PA) diameter + Left pulmonary artery diameter/Descending aorta diameter; Pt = patient; RVOT = right ventricular outflow tract.

98 mm^2/m^2 (Table 2). The right and left pulmonary artery Z scores ranged from −4.8 to −2.0 (mean −2.8 ± 0.9) and −4.5 to −0.5 (mean −2.2 ± 1), respectively. The McGoon ratio was below 1.5 for all patients, ranging from 0.6 to 1.2 (average 0.8 ± 0.2). Eight patients had multiple aortopulmonary collaterals. All lung segments supplied by aortopulmonary collaterals had dual supply by the native central pulmonary arteries as well (Fig. 1).

Pulmonary valve balloon dilation. Before balloon dilation, the pulmonary valve annulus Z score ranged from −5.8 to −3.1 (average −4.0 ± 1). Pulmonary valve dilation using a balloon/annulus ratio of 1.5 ± 0.3 resulted in a small immediate increase in the valve annulus size (Fig. 3). In seven patients there was an increase in initial diameter of at least 25%. Immediately following dilation, the average pulmonary valve annulus Z score increased from −4.0 to −3.3 ($p < 0.001$). The average number of valve dilations per patient was 3, ranging from 1 to 10.

There was also documentation of a small increase in the oxygen saturation ≥4% immediately following the procedure in five patients. However, because of simultaneous coil embolization of aortopulmonary collaterals, the change in systemic oxygen saturation was not consistent. In eight patients with adequate measurement of the main pulmonary artery pressure both before and after balloon dilation, an increase from a mean of 19 ± 7 to 30 ± 12 mm Hg ($p < 0.02$) was observed.

Figure 3. Comparison of pulmonary valve annulus Z score determined for each patient at initial catheterization before balloon dilation (BD), immediately after dilation and at preoperative follow-up catheterization. Note that only seven patients had repeat determinations before operation. Even though there is a small immediate increase in the pulmonary valve annulus Z scores with balloon dilation in all but two patients, this change does not consistently persist at preoperative catheterization.



Coil embolization of aortopulmonary collateral channels. Eight patients underwent coiling of aortopulmonary collaterals, most of which were occluded during the same initial catheterization (six patients), although two patients had them coiled at subsequent catheterizations. The total number of aortopulmonary collaterals coil embolized was 31. Thirty were successfully occluded, with no flow or trivial residual flow by selective angiography following the procedure. One patient had a large collateral ligated via video-assisted thoracoscopic surgery as a result of unsuccessful coil occlusion.

Balloon dilation of branch pulmonary artery stenoses. Seven patients underwent branch pulmonary artery balloon dilation, with a total of 22 vessels dilated, including segmental, lobar and main pulmonary artery branches. The mean average vessel diameter increased significantly with balloon dilation ($p < 0.001$). The diameter increased more than 30% of the initial size in 15 of the 22 vessels dilated. All but two vessels had improvement in the diameter and angiographic flow with balloon dilation. The average diameter increase was 76 ± 88%. The average balloon-to-vessel diameter ratio was 2.3 ± 0.8.

Other interventions. One patient with multiple very large aortopulmonary collaterals and diminutive pulmonary arteries underwent placement of a stent in the right ventricular outflow tract at a second preoperative catheterization in an attempt to improve anterograde pulmonary blood flow and allow coil occlusion of the large collaterals.

Preoperative catheterization. At preoperative catheterization, the Nakata index improved from 67 ± 28 mm^2/m^2 at presentation to 143 ± 84 mm^2/m^2 preoperatively ($p < 0.03$)

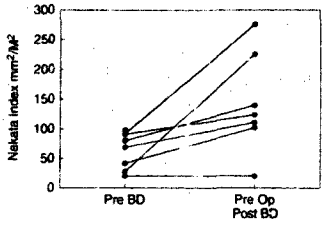


Figure 4. Comparison of Nakata index before balloon dilation (BD) and any interventions with values obtained at preoperative catheterization in seven patients. Note that there is a significant improvement in Nakata index in all but one patient before operation.

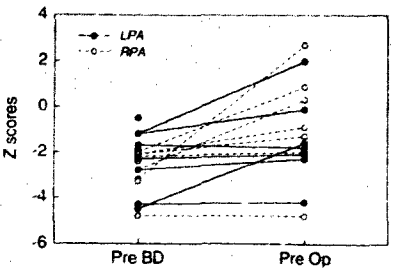


Figure 5. Left pulmonary artery (LPA) and right pulmonary artery (RPA) Z scores before any interventions and at preoperative follow-up catheterization for each patient. BD = balloon dilation.

(Fig. 4). Similarly, the right pulmonary artery Z scores changed from -2.8 ± 0.8 to -0.73 ± 2 ($p < 0.05$), and the left pulmonary artery Z scores increased from -2.2 ± 1.3 to -1.5 ± 1.9 ($p < 0.07$) (Fig. 5, Table 3). The average pulmonary valve annulus Z score, which had increased from -4 ± 0.1 before dilation to -3.3 ± 1.1 immediately after dilation, was -3.1 ± 0.7 preoperatively (Fig. 3).

Complications. There were no deaths. No tetralogy of Fallot cyanotic spells were observed during or after interventional catheterization. One patient, in whom a large aortopulmonary collateral could not be closed in the catheterization laboratory, had transient congestive heart failure and pulmonary edema requiring an additional 10 days in the intensive care unit with positive-pressure mechanical ventilation for 5 days. Because of collateral "steal" of descending aorta blood flow, this same patient had necrotizing enterocolitis, managed medically. During an attempt at coil embolization of the aforementioned large collateral, a coil was dislodged in the pulmonary artery distal collateral end. It was successfully retrieved, but damage to the vessel wall (contrast staining) without extravasation was observed by angiography. Subsequently, this large collateral was successfully ligated by video-assisted thoracoscopy. Another patient developed significant lobar pulmonary edema secondary to branch pulmonary artery balloon dilation and also required transient mechanical ventilation. Occlusion of the femoral arteries bilaterally was a complication in one patient with multiple aortopulmonary collaterals, which were coil embolized via arterial access. One patient had a transient pulse loss, and three had unilateral femoral vein occlusion diagnosed at a later catheterization.

Surgical management. Nine of 10 patients underwent complete single-stage surgical correction at an average of $8.5 \pm .6$ months (range 1 to 23 months) following the initial catheterization. One patient had a staged surgical repair with initial right ventricle-to-pulmonary artery homograft followed by ventricular septal defect closure 7 months later. The corrective surgical procedure involved ventricular septal defect closure in all and transannular right ventricular outflow tract patch in eight patients. One of these eight patients had augmentation of the outflow tract patch with use of a unicuspid aortic homograft. Two patients had placement of a right ventricle-to-pulmonary artery homograft.

Outcome and follow-up. All patients underwent complete operation at an average of 6 months after the first interventional catheterization. The mean follow-up period after operation was 2 years, ranging from 1 month to 4.9 years. At follow-up, the right ventricular pressure, estimated by echocardiogram (10 patients) or catheterization (6 patients), or both, was ≤50% systemic in 7 patients and between 50% and 70% in 3 patients, 2 of whom had right ventricle-to-pulmonary artery homografts placed, which were becoming relatively small with time. No patient had a right ventricular pressure higher than 70% systemic at follow-up. The average oxygen saturation at follow-up was 96%.

During follow-up, two patients had a residual atrial septal defect closed with a clamshell device in the cardiac catheterization laboratory without complications 4 and 10 months following complete tetralogy of Fallot repair, respectively.

Discussion

We report the results of a novel management approach for patients with severe tetralogy of Fallot and pulmonary stenosis who are thought to be unsuitable for complete surgical repair in one stage, involving use of transcatheter techniques.

Management of patients with tetralogy of Fallot. Tetralogy with diminutive pulmonary arteries and multiple aortopulmonary collaterals, in the absence of pulmonary atresia, is rare (2,3). Such patients share common characteristics with patients

Table 3. Average Values of Pulmonary Valve Annulus, Right and Left Pulmonary Artery Z Scores and Nakata Index Before Catheter Intervention and at Preoperative Catheterization

	Precatheter Intervention	Preoperative Catheterization*	p Value (*t* test)
PVA Z score	-4.0 ± 1	-3.1 ± 0.7	< 0.01
RPA Z score	-2.8 ± 0.8	-0.7 ± 2.3	< 0.05
LPA Z score	-2.2 ± 1.3	-1.5 ± 1.9	< 0.07
NI	67 ± 28	143 ± 84	< 0.03

*Values at preoperative follow-up catheterization after transcatheter interventions had been performed. LPA = left pulmonary artery; NI = Nakata index (mm^2/m^2); PVA = pulmonary valve annulus; RPA = right pulmonary artery.

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with tetralogy and pulmonary atresia and represent a group with a higher degree of severity, with features that overlap tetralogy with pulmonary atresia and tetralogy with pulmonary stenosis. Patients in whom the pulmonary arteries are thought to be too small to undergo complete correction generally undergo initial palliative surgical interventions. Even though surgical shunts have been shown to increase pulmonary artery growth (24,25) in patients with small pulmonary arteries, the effectiveness for diminutive pulmonary arteries (i.e., less than 100 mm^2/m^2) remains unproven. In addition, various studies, including postmortem studies, have demonstrated markedly abnormal pulmonary artery development in patients with shunts (26), including pulmonary artery distortion, inhomogeneous perfusion and multiple potential postoperative complications (27–30). In patients with hypoplasia or branch pulmonary artery stenosis, an aortopulmonary shunt offers very limited possibilities for subsequent transcatheter balloon dilation. Since 1984, we have opted to surgically establish right ventricle-to-pulmonary artery continuity and then, via transcatheter techniques, rehabilitate the pulmonary arteries so that a complete surgical correction can follow successfully (9). A similar approach was used in these reported 10 patients. Instead of a surgical right ventricle-to-pulmonary artery homograft or conduit, patients underwent pulmonary valve balloon dilation, which facilitated distal pulmonary artery transcatheter preoperative "rehabilitation." Patients who have pulmonary arteries of normal size or with mild or moderate hypoplasia generally undergo early complete surgical correction in one stage, with no need for initial palliation (6,7).

A recent report of intrauterine development of pulmonary atresia in fetuses with tetralogy of Fallot and pulmonary stenosis demonstrates that this is a progressive disease (4) with a potential increase in severity with time. Early intervention oriented to optimize the supply of pulmonary blood flow in the most physiologic fashion may halt this pathologic process, "normalize" pulmonary artery growth during infancy (31) and should therefore theoretically be beneficial. Thus, early repair of tetralogy of Fallot during infancy is thought to be the optimal management approach. When the pulmonary arteries are diminutive and early complete repair is precluded, transcatheter interventions may improve pulmonary artery growth and allow subsequent successful surgical repair.

Pulmonary valve balloon dilation in tetralogy of Fallot. Reported effects of pulmonary valve balloon dilation in patients with tetralogy of Fallot and "normal" pulmonary artery size have included increased pulmonary blood flow and oxygen saturation, increased pulmonary valve annulus and improved growth of the pulmonary artery branches (32–39). Even though the patient populations are markedly different, our immediate procedural results do not differ significantly from those reported by others in terms of immediate annular diameter change and pulmonary artery growth. However, the pulmonary valve annular growth remained very abnormal after dilation in our series of patients with severe tetralogy of Fallot. The pulmonary valve annulus Z score, which had increased immediately after balloon dilation, had not improved further at all at preoperative catheterization. Others have reported the potential advantage of pulmonary valve balloon dilation in avoiding a transannular patch at the time of surgical correction (39). This report of patients with severe forms of tetralogy of Fallot suggests that even though there is an immediate increase in pulmonary valve annulus with balloon dilation, its growth remains abnormal, and a transannular patch may still be required.

Study limitations. Because these patients had multiple transcatheter interventions, the effect of each of them independently cannot be determined. Thus, pulmonary artery growth may have occurred secondary to the effects of increased anterograde flow, balloon dilation or intrinsic vessel growth. In addition, changes in systemic oxygen saturation observed following balloon dilation of the pulmonary valve were influenced by simultaneous coil embolization of aortopulmonary collaterals and balloon dilation of peripheral stenosis.

It was not possible to identify a proper control group of patients with similarly small pulmonary arteries and aortopulmonary collaterals adequately assessed preoperatively who had been managed with palliative shunts before complete repair. Patients with such characteristics are rare, and the few patients identified differed substantially from this cohort and could not be used for comparison. The variability in patient age at the time of initial intervention in this series was dependent on the time of patient referral. Patients who were followed up at our institution from birth underwent pulmonary valve balloon dilation early in the newborn period, according to our policy of early intervention as a means of restoring normal pulmonary artery growth and development (6,7,22). Furthermore, the small number of patients limits our ability to perform analyses to study the factors predictive of success of this management approach.

Conclusions. Initial pulmonary valve balloon dilation in patients with tetralogy of Fallot, severe pulmonary stenosis and diminutive pulmonary arteries increases the annulus Z score and anterograde pulmonary blood flow, allowing simultaneous coiling of aortopulmonary collaterals and easier access for branch pulmonary artery dilation, all of which improve pulmonary artery growth and both anatomic and physiologic preoperative conditions. Most patients with tetralogy of Fallot do not need palliative procedures or even preoperative cardiac catheterization. In contrast, patients with diminutive pulmonary arteries seem to benefit from an initial preoperative transcatheter palliation, although the small size of the series, the multiplicity of interventions performed and the lack of adequate control group make this conclusion tentative.

References

1. Van Praagh RV, Van Praagh S, Nebasar RA, Muster AJ, Sinha SN, Paul MH. Tetralogy of Fallot: underdevelopment of the pulmonary infundibulum and its sequelae. Am J Cardiol 1970;26:25–33.
2. Kirklin JW, Blackstone EH, Jonas RA, et al. Morphologic and surgical determinants of outcome events after repair of tetralogy of Fallot and pulmonary stenosis. J Thorac Cardiovasc Surg 1992;103:706–23.
3. Shimazaki Y, Blackstone EH, Kirklin JW, Jonas RA, Mandell V, Colvin EV

The dimensions of the right ventricular outflow tract and pulmonary arteries in tetralogy of Fallot and pulmonary stenosis. J Thorac Cardiovasc Surg 1992;103:692–705.

4. Hornberger LK, Sanders SP, Sahn DJ, et al. In utero pulmonary artery and aortic growth and potential for progression of pulmonary outflow obstruction in tetralogy of Fallot. J Am Coll Cardiol 1995;25:739–45.
5. Groh M, Meliones JN, Bove EL, et al. Repair of tetralogy of Fallot in infancy. Effect of pulmonary artery size on outcome. Circulation 1991;84: Suppl III:III-206–12.
6. Walsh EP, Rockenmacher S, Keane JF, Hougen TJ, Lock JE, Castañeda AR. Late results in patients with tetralogy of Fallot repaired during infancy. Circulation 1988;77:1062–7.
7. Di Donato RM, Jonas RA, Lang P, Rome JJ, Mayer JE Jr, Castañeda AR. Neonatal repair of tetralogy of Fallot with and without pulmonary atresia. J Thorac Cardiovasc Surg 1991;101:126–37.
8. Hennein HA, Mosca RS, Urcelay G, Crowley DC, Bove EL. Intermediate results after complete repair of tetralogy of Fallot in neonates. J Thorac Cardiovasc Surg 1995;109:332–44.
9. Rome JJ, Mayer JE, Castañeda AR, Lock JE. Tetralogy of Fallot with pulmonary atresia: rehabilitation of the pulmonary arteries. Circulation 1993;88:1691–8.
10. Kirklin JW, Blackstone EH, Shimazaki Y, et al. Survival, functional status and reoperations after repair of tetralogy of Fallot with pulmonary atresia. J Thorac Cardiovasc Surg 1988;96:102–16.
11. Bull K, Somerville J, Ty E, Spiegelhalter D. Presentation and attrition in complex pulmonary atresia. J Am Coll Cardiol 1995;25:491–9.
12. Piehler JM, Danielson GK, McGoon DC, Wallace RB, Fulton RE, Mair DD. Management of pulmonary atresia with ventricular septal defect and hypoplastic pulmonary arteries by right ventricular outflow construction. J Thorac Cardiovasc Surg 1980;80:552.
13. Nakata S, Imai Y, Takanashi Y, et al. A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart disease with decreased pulmonary blood flow. J Thorac Cardiovasc Surg 1984;88:610–9.
14. Kirklin JW, Barratt-Boyes BG. Anatomy, dimensions, and terminology. In: Cardiac Surgery, 2nd edition. London: Churchill Livingstone, 1993:5–60.
15. Keane JF, Lock JE. Catheter intervention: balloon valvotomy. In: Lock JE, Keane JF, Fellows KE, eds. Diagnostic and Interventional Catheterization in Congenital Heart Disease. The Hague: Martinus Nijhoff, 1987:111–22.
16. Radke W, Keane JF, Fellows KE, Lang P, Lock JE. Percutaneous balloon valvotomy of congenital pulmonary stenosis using oversized balloons. J Am Coll Cardiol 1986;8:909–15.
17. Lock JE, Castaneda-Zuniga WR, Furhman BP, et al. Balloon dilation angioplasty of hypoplastic and stenotic pulmonary arteries. Circulation 1983;67:962–7.
18. Rothman A, Perry SB, Keane JF, Lock JE. Early results and follow-up of balloon angioplasty for branch pulmonary artery stenoses. J Am Coll Cardiol 1990;15:1109–17.
19. Zeevi B, Keane JF, Fellows KE, Lock JE. Balloon dilation of critical pulmonary artery stenosis in the first week of life. J Am Coll Cardiol 1988;11:821–4.
20. Lock JE. Catheter intervention: balloon angioplasty. In: Lock JE, Keane JF, Fellows KE, eds. Diagnostic and Interventional Catheterization in Congenital Heart Disease. The Hague: Martinus Nijhoff, 1987:91–110.
21. Fellows KE, Lock JE. Catheter intervention: septostomy, occlusion techniques and pericardial drainage. In: Lock JE, Keane JF, Fellows KE, eds. Diagnostic and Interventional Catheterization in Congenital Heart Disease. The Hague: Martinus Nijhoff, 1987:123–43.
22. Castañeda AR, Jonas RA, Mayer JE, Hanley FL. Tetralogy of Fallot. In: Castañeda AR, ed. Cardiac Surgery of the Neonate and Infant. Philadelphia: WB Saunders, 1994:215–34.
23. Sievers HH, Onnasch DG, Lange PE, Bernhard A, Heintzen PH. Dimensions of the great arteries, semilunar valve roots and right ventricular outflow tract during growth: normative angiographic data. Pediatr Cardiol 1983;4: 189–96.
24. Gale AW, Arcimegas E, Green EW, Blackstone EH, Kirklin JW. Growth of the pulmonary annulus and pulmonary arteries after the Blalock-Taussig shunt. J Thorac Cardiovasc Surg 1979;77:459–66.
25. Kirklin JW, Bargeron LM, Pacifico AD. The enlargement of small pulmonary arteries by preliminary palliative operations. Circulation 1977;56:612–7.
26. Rabinovitch M, Herrera-de Leon V, Castaneda AR, Reid L. Growth and development of the pulmonary vascular bed in patients with tetralogy of Fallot with or without pulmonary atresia. Circulation 1981;64:1234–48.
27. Mickell JJ, Oh KS, Siewers RD, Galvis AG, Fricker FJ, Mathews RA. Clinical implications of postoperative unilateral phrenic nerve paralysis. J Thorac Cardiovasc Surg 1978;76:297–304.
28. Trusler GA, Miyamura H, Culhan JA, Fowler RS, Freedom RM, Williams WG. Pulmonary artery stenosis following aorticopulmonary anastomosis. J Thorac Cardiovasc Surg 1981;82:398–404.
29. Smith CD, Sade RM, Crawford FA, Othersen HB. Diaphragmatic paralysis and eventration in infants. J Thorac Cardiovasc Surg 1986;91:490–7.
30. Ullom RL, Sade RM, Crawford FA. The Blalok-Taussig shunt in infants: standard versus modified. Ann Thorac Surg 1987;44:539–43.
31. Hislop A, Reid L. Pulmonary arterial development during childhood: branching pattern and structure. Thorax 1973;28:129–35.
32. Boucek MM, Webster HE, Ormond GS, Ruttenberg HD. Balloon pulmonary valvotomy: palliation for cyanotic heart disease. Am Heart J 1988;115: 318–22.
33. Qureschi SA, Kirk CR, Lamb RK, Arnold R, Wilkinson JL. Balloon dilation of the pulmonary valve in the first year of life in patients with tetralogy of Fallot: a preliminary study. Br Heart J 1988;60:232–5.
34. Rey C, Marache P, Francart C, Dupuis C. Percutaneous transluminal balloon valvuloplasty of congenital pulmonary valve stenoses with a special report on infants and neonates. J Am Coll Cardiol 1988;11:815–20.
35. De Greeter P, Weisburd P, Dillenseger P, Willard D. Valvulplastie pulmonaire percutenee palliative dans les formes neonateles de tetralogie de Fallot. Arch Fr Pediatr 1989;46:117–9.
36. Parsons JM, Ladusans EJ, Qureshi SA. Growth of the pulmonary artery after neonatal balloon dilation of the right ventricular outflow tract in an infant with tetralogy of Fallot and atrioventricular septal defect. Br Heart J 1989;62:65–8.
37. Sreeram N, Saleem M, Jackson M, Peart I, McKay R, Arnold R. Results of balloon pulmonary valvuloplasty as a palliative procedure in tetralogy of Fallot. J Am Coll Cardiol 1991;18:159–65.
38. Guntheroth WG. Balloon valvuloplasty as palliation in tetralogy. J Am Coll Cardiol 1992;19:234–7.
39. Sluysmans T, Neven B, Rubay J, et al. Early balloon dilation of the pulmonary valve in infants with tetralogy of Fallot, risk and benefits. Circulation 1995;91:1506–11.