Early and late results and the effects on pulmonary arteries of balloon dilatation of the right ventricular outflow tract in tetralogy of Fallot

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Background Balloon dilatation of the pulmonary valve remains controversial as a palliative procedure in tetralogy of Fallot.

Aims To determine the clinical outcome, growth of the pulmonary vascular tree and findings at surgery of balloon dilatation of the pulmonary valve performed as a palliation in infants with tetralogy of Fallot.

Methods and Results Thirty-three severely cyanosed infants (mean age of 3 months for the whole population, including seven neonates) underwent this procedure from June 1990 to January 1997. After balloon dilatation, systemic oxygen saturation increased from a mean value of 76 ± 9% to 88 ± 7% (P < 0.001). The procedure was accomplished without complications. Four patients had recurrent hypoxic spells after dilatation leading to surgical repair within 30 days of dilatation (three modified Blalock-Taussig shunts and one complete repair). A control study was performed at a mean of 6.1 ± 4.5 months after dilatation (three modified Blalock-Taussig shunts and one complete repair). A control study was performed at a mean of 6.1 ± 4.5 months after dilatation in 16 patients to establish growth of the pulmonary vascular tree (repeat catheterization in nine patients and surgical pulmonary annulus calibration in seven). The pulmonary annulus increased from a mean Z score of −4.1 ± 0.9 SD to −2.5 ± 1.1 SD (P < 0.001). Z score for the right pulmonary artery from −3.0 ± 0.6 SD to −1.9 ± 1.2 SD (P = 0.007) and the Z score for the left pulmonary artery from −2.7 ± 0.7 SD to −1.6 ± 1.3 SD (P = 0.021). At late follow-up (mean of 9 months after dilatation, range 3 to 54 months), 28 patients underwent complete repair. Trans-annular patching was required in 43% of patients. Four post-operative deaths were observed, none dilatation related.

Conclusion Balloon dilatation of the pulmonary valve is an effective and safe palliation in tetralogy of Fallot. It promotes growth of the pulmonary vascular tree, reducing the need for trans-annular patching and is recommended in symptomatic infants of very young age, with a small pulmonary annulus (Z value below −4 SD) and associated cardiac anomaly.

Key Words: Tetralogy of Fallot, balloon dilatation of the pulmonary valve.

Introduction

Balloon dilatation of the pulmonary valve remains controversial as a palliative procedure in tetralogy of Fallot. There are concerns regarding the efficacy and safety of this technique, in comparison with other palliation, such as creation of an aortopulmonary shunt. Another therapeutic option is primary complete repair which can be undertaken even in infancy, but with an increased risk for early post-operative death and frequent need for a trans-annular patch inducing pulmonary regurgitation. We report our experience with balloon dilatation of the pulmonary valve in symptomatic infants with tetralogy of Fallot and present our immediate and mid-term results. Changes in pulmonary vascular dimensions and the need for trans-annular patching are evaluated.

Material and method

Population

The retrospective study included 33 infants (15 girls and 18 boys) with tetralogy of Fallot who underwent balloon dilatation of the pulmonary valve.
dilatation of the pulmonary valve between June 1990 and January 1997. The mean age of catheterization was 3·3 ± 2·7 months (range 3 days to 11 months) and weight was 4·8 ± 1·3 kg (range 2·7 to 8 kg). All patients in whom it was intended to perform balloon dilatation of the pulmonary valve were included. Associated cardiac malformations were common (21 of 33 patients): right aortic arch (n=11), anomalous origin of the left anterior descending coronary artery from the right coronary artery (n=5), left superior vena cava (n=4), atrioventricular septal defect (n=3), multiple ventricular septal defects (n=2), atrial septal defect (n=1), large aortopulmonary collateral artery (n=1) and total anomalous pulmonary venous connection (n=3). Three of the patients had Down’s syndrome with an atrioventricular septal defect; two had DiGeorge syndrome; one had otomandibular syndrome, bilateral harelip and cleft palate with agenesis of corpus callosum and cerebral haemorrhage. Most of the patients systematically received beta-blockers (n=28) before cardiac catheterization. Balloon dilatation of the pulmonary valve was realized in all as a palliative procedure because of severe cyanosis (systemic oxygen saturation 76 ± 9% and haemoglobin level 15·4 ± 0·23 g dl$^{-1}$). Before this procedure, 14 of them had cyanotic spells.

Cardiac catheterization

Cardiac catheterization was performed under local anaesthesia after premedication (midazolam 0·3 mg kg$^{-1}$ body weight). Pressures were recorded in the right ventricle, pulmonary artery, left ventricle and aorta. Multiple angiograms were performed in the right ventricle in anteroposterior and lateral views, in the left ventricle in the left anterior oblique view and in both coronary arteries. The diameter of the pulmonary annulus (on lateral projection) and the origins of the pulmonary arteries before the first lobar branch (on an anteroposterior projection) were calculated at end systole. A flexible tip J guidewire (Radifocus, Terumo*, Leuven, Belgium) was placed in the distal right or left pulmonary artery (usually the right) by means of an end-hole catheter (a right 5-French coronary Judkins catheter). This catheter was replaced over a guidewire with a balloon catheter advanced and positioned across the pulmonary valve annulus. The maximal balloon size was chosen according to the size of the pulmonary annulus. The balloon diameter ranged from 5 to 10 mm (mean 7 ± 1 mm) with a ratio of balloon size to pulmonary annulus diameter ranging from 100 to 200% (mean 132 ± 28%). The balloon was inflated with diluted saline contrast material and deflated immediately (Fig. 1). Balloon dilatation of the pulmonary valve was performed between one to seven times, usually a mean of three times. After dilatation, a pressure pullback tracing across the pulmonary outflow tract was recorded. Similar measurements of the diameters of the pulmonary annulus and pulmonary arteries were calculated in 16 patients during a second catheterization (n=9) or surgical repair with annulus calibration (n=7). These measurements were performed at a mean of 6·1 ± 4·5 months after dilatation, but the patients who had required a Blalock-Taussig shunt after dilatation were not included in the study of pulmonary artery growth.

Figure 1 Straight anterior image of pulmonary valve dilatation showing two waists: one at the level of the stenotic pulmonary valve and the other at the level of the narrowed infundibulum.

Statistical analysis

All results are expressed as mean ± one standard deviation (SD) and P values <0·05 were considered significant. The size of the pulmonary annulus and the pulmonary artery just proximal to the first lobar branch were calculated and also transformed to a Z value, that is the number of SD from the mean normal value for the same body surface area size$^{1,2}$: $Z$ value=observed dimension minus mean normal dimension/SD around the mean normal dimension. The mean normal dimension and SD refer to values in normal persons of the same body surface area as the patient and were obtained from Sievers’ regression equations$^{3}$. The normal Z values in 95% of the reference population lay between −2 and +2. Differences between pre- and post-dilatation values were assessed using paired Wilcoxon tests. Linear regression was also performed between the ratio of balloon size to pulmonary annulus diameter and the increase in the pulmonary annulus with time.
Results

Immediate results

The pulmonary valve could be crossed in all patients. After balloon dilatation of the pulmonary valve, a significant increase in systemic oxygen saturation was noticed increasing from 76 ± 9% to 88 ± 7% (P < 0.001). The right ventricle-to-pulmonary artery pressure gradient decreased from 73 ± 12 to 52 ± 13 mmHg (P < 0.001). Ten of the 14 patients with hypoxic spells had persistent cyanosis after inflation controlled with intravenous propranolol. Four patients had transient arrhythmia during inflation (atrial tachycardia in two patients; bradycardia in one patient and complete heart block in one patient). Finally, the guidewire caused a small infundibular perforation in one patient during the procedure, but there were no clinical consequences. Patients receiving beta-blockers prior to dilatation continued their treatment after dilatation to prevent cyanotic spells. Despite propranolol therapy, adequate palliation did not last in four patients, all with predilatation spells. As a result, early surgical repair was necessary (within the 30 days of balloon dilatation): three infants had a modified Blalock-Taussig shunt and another, with adequate anatomy, underwent a complete corrective repair.

Outcome

A further two patients had modified Blalock-Taussig shunts, respectively, 2 and 5 months after dilatation. The first patient had miscellaneous malformations with bilateral harelip and cleft palate. The modified Blalock-Taussig shunt was constructed before planned plastic repair of the face, but unfortunately the patient died after shunting, from congenital neurological anomalies. The second patient had an atrioventricular septal defect and trisomy 21.

Growth of the pulmonary annulus

Before balloon dilatation of the pulmonary valve, the pulmonary annulus diameter was 5.5 ± 1.3 mm with a mean Z value of 4.3 ± 1.1 SD. In patients who underwent the control study, the pulmonary annulus diameter increased from 5.8 ± 1.3 mm to 9.0 ± 1.7 mm (P = 0.007) and the mean Z value from -4.1 ± 0.9 SD to -2.5 ± 1.1 SD (P < 0.001). A weak correlation was found between growth of the pulmonary annulus and balloon size to pulmonary annulus ratio (r=0.50).

Growth of the pulmonary arteries

Before balloon dilatation of the pulmonary valve, the right pulmonary artery diameter was 4.4 ± 1.2 mm and the left pulmonary artery diameter 4.4 ± 1.3 mm with, respectively, a mean Z value of -2.9 ± 1.1 SD and -2.6 ± 1.3 SD. In those patients who underwent the control study the diameter of the right pulmonary artery increased from 4.1 ± 0.6 mm to 6.4 ± 1.3 mm (P = 0.005) and the mean Z value from -3.0 ± 0.6 SD to -1.9 ± 1.2 SD (P = 0.007). Similarly, the diameter of the left pulmonary artery increased from 4.1 ± 1.0 mm to 6.5 ± 1.5 mm (P = 0.005) and the mean Z value from -2.7 ± 0.7 SD to -1.6 ± 1.3 SD (P = 0.021).

Operative findings and follow-up

Twenty-eight patients, including four patients with prior Blalock-Taussig shunts, underwent complete corrective surgery at 13.4 ± 9 months of age, a mean of 9 months after balloon dilatation of the pulmonary valve (range 3 to 54 months). In one patient, the surgeon noted a tear in one of the pulmonary cusps. This was not related to the large balloon (the ratio of balloon size to pulmonary annulus diameter was 1.28 for this patient). Twelve patients (43%), including two of the four in whom an anastomosis was implanted, required pulmonary trans-annular patching. Two of them had pulmonary valve insertion: one monocusp and one heterograft valve. When present, branch pulmonary artery stenoses were repaired with patch enlargement (n=3). The mean ratio of post-operative right ventricular to left ventricular systolic pressure was 0.60 ± 0.21 in patients requiring trans-annular patching, compared to 0.62 ± 0.12 in the remainder (P=ns). Three hospital deaths occurred after complete repair. In two patients, in whom low cardiac output was the cause of death, one, with associated atrioventricular septal defect, suffered a myocardial infarction due to coronary artery compression by the heterograft. The third patient with severe Fallot and a large left aortopulmonary collateral artery died from right ventricular failure and tamponade. The four remaining children, including the patient with atrioventricular septal defect, are clinically doing well and still awaiting complete repair. One of them has undergone correction of the associated total anomalous pulmonary venous connection without repair of the tetralogy 6 months after dilatation.

Discussion

For many years, controversy has surrounded the optimum time and appropriate type of surgery for symptomatic children with tetralogy of Fallot. Classically, construction of a modified Blalock-Taussig shunt was recommended in symptomatic cyanosed infants and correction was delayed until 1 or 2 years of age. More recently, there has been a tendency to propose earlier and primary complete surgical repair even in infancy. In 1973, Barrat-Boyes and Neutze reported such surgical repair in 18 infants, in 44% of whom a pulmonary trans-annular patch was needed. Subsequently, many...
investigators advocated a similar approach\textsuperscript{[5–12]} but such early surgical repair frequently involves opening the right ventricular outflow tract. The need for trans-annular patching has ranged from 36 to 100%, with an estimated mean figure of about 71% (Table 1). In one series\textsuperscript{[12]} a trans-annular patch was required in only a few, but probably related to surgical preference, in the opinion of the authors. In addition, very young age has been considered a risk factor for post-operative death, especially if a trans-annular patch is required\textsuperscript{[7,13]} Opening of the right ventricular outflow tract commonly induces pulmonary valve incompetence, which is usually well tolerated for a long period of time\textsuperscript{[14]}. However, this may also influence long-term morbidity, because pulmonary valve incompetence results in right ventricular enlargement and is also implicated in the pathogenesis of right ventricular dysfunction, ventricular arrhythmia and the risk of sudden death during long-term follow-up\textsuperscript{[15–17]}. Finally, trans-annular patching may increase the frequency of re-operation because of significant patch-induced pulmonary regurgitation\textsuperscript{[18,19]} Recently, Sousa Uva et al.\textsuperscript{[2]} did not recommend such primary repair in infants with very low weight (less than 3 kg), with a severely hypoplastic pulmonary arterial tree (Z value less than $-4$ SD) or a major associated cardiac anomaly. It is of interest that 31 of our patients fulfilled all these criteria, including 70% patients with a Z value for pulmonary annulus less than $-4$ SD at dilatation. In patients with associated cardiac anomaly, balloon dilatation is of interest. For example, a trans-annular patch may damage an anomalous origin of the left anterior descending coronary artery from the right coronary artery, leading to myocardial ischaemia. Moreover, atroventricular septal defect combined with tetralogy of Fallot complicates the repair and is a risk factor for death, especially in patients with Down’s syndrome. Both of these clearly indicate delaying complete repair, and balloon dilatation of the pulmonary valve appears a good therapeutic life saving option.

On the other hand, the other classical option in symptomatic infants is to perform a Blalock-Taussig shunt. Such an anastomosis is considered to reduce cyanosis and to promote the growth of the pulmonary vascular tree, but usually in a small number of patients\textsuperscript{[20–25]}. In contrast, some authors found that, after shunting, hypoplastic pulmonary arteries remained relatively small\textsuperscript{[26]} and the need for trans-annular patching was not always reduced at the time of complete repair\textsuperscript{[27]}. In addition, occlusion of the shunt has also been reported in the immediate post-operative period or during follow-up leading to shunt failure\textsuperscript{[21,23,25]}. Finally, there are concerns about the development of pulmonary artery stenosis at the level of the anastomosis in the long-term follow-up, as reported in 15% of shunts\textsuperscript{[28]}, which remains a potentially serious problem.

The other therapeutic option is to perform balloon dilatation of the pulmonary valve, which is a well established treatment for pulmonary valve stenosis in children\textsuperscript{[20]}. In tetralogy of Fallot, balloon dilatation is proposed as palliation similar to a Blalock-Taussig shunt. The reason is that balloon dilatation not only increases forward flow to the lungs with relief of cyanosis but should also promote growth of the pulmonary vascular tree significant enough to reduce the need for trans-annular patching at the time of complete repair. Inevitably, there are many obstructions in a series in tetralogy of Fallot. To overcome these obstacles, balloon dilatation of the pulmonary valve can be achieved without the risk of subsequent pulmonary vascular obstructive disease\textsuperscript{[30]}.

Balloon dilatation of the pulmonary valve as a palliative procedure in tetralogy of Fallot was first performed by Lababidi\textsuperscript{[31]} in 1983. In 1988, Qureshi et al.\textsuperscript{[32]} reported a series of 15 patients treated by balloon dilatation. The authors noticed an increase in systemic oxygen saturation and a decrease in the systolic gradient across the right ventricular outflow tract. In 70% of patients, corrective operation could be delayed, or no further palliation was required in the follow-up period. In 1991, Sreram and associates reported on a wider series of patients\textsuperscript{[67]} from the same centre\textsuperscript{[33]}. Primary success was obtained in 86% of the patients.

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**Table 1 Need for pulmonary transannular patching in infancy**

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<th>Author(s)</th>
<th>Transannular patch</th>
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*Repair in infants less than 18 months of age (mean 7 months).
†Repair after balloon dilatation of the pulmonary valve.
Other series, including our patients, had similar results. Most of the patients showed marked clinical improvement with relief of cyanosis and the complete repair could be delayed.

Some have questioned the efficacy of balloon dilatation of the pulmonary valve on the growth of the pulmonary vascular tree. Many criteria have been proposed to describe such growth. We have chosen the Z value because it has the advantage of being independent of body growth during follow-up. Significant growth of the pulmonary annulus and pulmonary artery branches was observed in the current series, which is consistent with the data reported by Sluysmans et al. using a larger balloon size to pulmonary annulus ratio of 150% vs ours of 132%. In their series, the Z value of the pulmonary annulus increased from −4.8 SD to −2.7 SD at pre-operative catheterization, so that trans-annular patching was required in only 31% of the 16 infants repaired. In our experience, trans-annular patching was performed in 43% of 28 patients repaired, despite a larger Z value for the pulmonary annulus at control study. Although the reason for this is not entirely clear, this may reflect individual surgical preference. On the other hand, it is likely that more of our patients could have been repaired without opening the pulmonary annulus, using a similar criterion. In our patients who underwent the control study, it appears that trans-annular patching was placed when the Z value for the pulmonary annulus was −3 SD or less and was not placed when the Z value was greater than −2 SD. The differences observed between the different series may be related to the degree of hypoplasia of the pulmonary annulus and the choice of balloon size at dilatation. It is clear that growth of the pulmonary annulus and subsequent reduction of trans-annular patching is the major long-term advantage of this technique.

The major complication of the technique is perforation of the right ventricular outflow tract with cardiac tamponade. This is relatively uncommon but can be fatal. The use of a right coronary Judkins or Cobra catheter with an appropriate curve, in combination with a flexible guidewire, should reduce this risk and facilitate the crossing of the right ventricular outflow tract. Damage to the pulmonary valve leaflet following balloon dilatation has also been reported in 50 to 75% of patients. This may be the consequence of using a large balloon with a balloon size to pulmonary annulus ratio between 1.5 and 2. In our experience, this happened once, with the tear of one pulmonary cusp, but this was not related to a large balloon size to pulmonary annulus ratio (ratio 1.28 for this child). It is unlikely that this complication would compromise this technique. However, despite the reported correlation between balloon size to pulmonary annulus ratio and growth of the pulmonary annulus, it is likely that such complications can be avoided by using an appropriately sized balloon, and we recommend using a balloon size to pulmonary annulus ratio which should not exceed 1.5. Other complications have been reported, such as femoral vein thrombosis and septicaemia, which are not specific to this technique, but are similar to those observed with interventional procedures in infancy.

A transient decrease in systemic saturation during balloon inflation and in the hours following the procedure is frequently observed. It has been suggested that patients suffering hypoxic spells are not good candidates for dilatation. We found that 10 of the 14 (71%) patients with hypoxic spells before dilatation did not require urgent surgical operation after balloon dilatation of the pulmonary valve. It seems clear that most of them had improved after dilatation, as already reported.

**Conclusion**

Balloon dilatation of the pulmonary valve is a useful and effective palliative procedure in very cyanosed infants with tetralogy of Fallot. This technique increases systemic oxygen saturation, allows growth of the pulmonary annulus and reduces the need for trans-annular patching. We recommend this technique in symptomatic infants of very young age (less than 3 months), with a small pulmonary annulus (Z value less than −4 SD) and major associated cardiac anomalies, such as multiple ventricular septal defects, ativoventricular septal defect, anomalous origin of the left anterior descending coronary artery from the right coronary artery. For these patients, primary complete repair carries an increased risk factor for post-operative death. On the other hand, unsuccessful balloon dilatation may be an indication for creation of an aortopulmonary shunt.

**References**


