Evidence for palliative enlargement of the right ventricular outflow tract in severe tetralogy of Fallot

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Abstract

Objective: If the pulmonary artery (PA) tree in patients with Fallot’s tetralogy (TOF) is extremely hypoplastic, a shunt procedure may be difficult and not desirable because of side-effects. Moreover, the failing catch-up growth of the pulmonary annulus is well known. In patients with a severe form of TOF, we performed palliative transannular patching of the right ventricular outflow tract. The early and long-term follow-up was evaluated.

Methods: Eleven patients (93 days (10–245 days); 3.5 ± 0.7 kg (2.5–4.3 kg)) had highly symptomatic TOF (Hb: 18 ± 2 g/dl, SO2: 68 ± 11%); angiographic diameters: RPA: 4.1 mm (2.5–6.4 mm), LPA: 3.4 mm (1.6–7.0 mm), PA trunc: 4.4 mm (2.5–7.0 mm). All 11 underwent transannular enlargement of the right ventricular outflow tract without closure of the ventricular septum defect. A PA index (cross-sectional area of the pulmonary arteries to BSA) was used to compare pre- and postoperative data. For follow-up, the patients were repetitively examined clinically and echocardiographically.

Results: Preoperative PA index was 87 ± 40 mm²/m² (normal: 330 ± 35 mm²/m²). Postpalliation angiograms (age: 10–14 months) demonstrated a significant catch-up growth in nine patients (PA index from 99 ± 40 to 310 ± 54 mm²/m²) and inadequate growth in two patients (PA index 63 and 115 mm²/m²). Perioperative mortality was zero. Ten patients (43 months; 6-105 months) underwent elective repair. Six patients received pulmonary homograft valves (6-15 years after repair) because of severe pulmonary valve insufficiency and severe RV dilation. Complications: One patient died 10 months postpalliation due to pneumonia, one patient received a pacemaker after repair and died (2 months post-repair) due to pacemaker failure, a 5-year-old patient died 1 month after repair due to sepsis. All eight long-term survivors (12-17 years) are in excellent clinical condition. Echocardiography revealed good RV function and near normal diameters at peak systolic pressures between 25 and 50 mmHg. Only one patient developed brady-arrhythmia; a pacemaker was implanted 8 years after repair and 2 years after homograft implantation. Conclusions: In a very severe form of TOF, palliative right ventricular outflow tract construction may provide the potential for complete repair. In the presented high-risk patient group, mortality was not related to the hypoplastic pulmonary arteries. Obviously, all patients need pulmonary valve implantation in the long run.

Keywords: Tetralogy of Fallot; Palliation; Hypoplastic pulmonary artery; Enlargement of right ventricular outflow tract; Vascular catch-up growth; Pulmonary valve replacement

1. Introduction

Since, the first palliation on a 16-months old, cyanotic patient in 1944 by Alfred Blalock [1] and since, the first total correction in 1954 by Lillehei and Dewall [2], the discussion continues about choosing a palliative procedure or a corrective procedure in Fallot patients with severe hypoplasia of the pulmonary artery tree.

The treatment of neonates and infants who are suffering from Fallot’s tetralogy with severe hypoplasia of the pulmonary vessels, still presents a challenge for the surgeon. We present early and long-term data of patients with a severe form of Fallot’s tetralogy, in which palliative transannular patching of the right ventricular outflow tract was performed and special attention was paid on the growth of the pulmonary arteries.

2. Patients and methods

Between 1965 and 1990, 688 Fallot patients were operated at the University Hospital of Dueseldorf. In 333 (48%) patients, a palliative shunt was performed.

To contribute to the discussion about the best preliminary palliative procedure, we performed a study on 11 patients
with extremely hypoplastic pulmonary arteries, in whom a transannular enlargement of the right ventricular outflow tract (RVOT) without closure of the ventricular septum defect was performed as initial palliative operation.

All patients suffered from a highly symptomatic tetralogy of Fallot (TOF). The haemoglobin was $18 \pm 2$ g/dl (mean $\pm$ SD) and the oxygen saturation was $68 \pm 11\%$. Six patients received preoperatively prostaglandins over 10-90 days. The RVOT enlargement was performed at the average age of 93 days (10-245 days). The patients weighed on average $3.45 \pm 0.72$ kg (range: 2.5-4.3 kg).

2.1. Angiography

Angiographies were analyzed pre- and postoperatively. The diameter of the right and left pulmonary arteries were measured just proximal to the 'upper lobe artery'. The calculated vessel area was divided by the body surface area (BSA; Nakata index [3]). The values were compared with a group of 59 patients without relevant cardiovascular abnormalities. A PA index of 120-150 mm$^2$/m$^2$ was indicative for palliative treatment, because a value of less than 150 mm$^2$/m$^2$ was associated with a high risk of death [4]. PA annulus was measured just at the base of the valve (Fig. 1).

2.2. Operative technique

Routinely, those patients who had a low PA index and a PA annulus index smaller than 120-140 mm$^2$/m$^2$, i.e. about 4 SD below the normal mean of $330 \pm 35$ mm$^2$/m$^2$, received a transannular enlargement during total correction according to the Rowlatt tables [5]. After cardiopulmonary bypass was established, cold Bretschneider’s cardioplegic solution was employed in all cases of the described group. To enlarge the RVOT, an autologous pericardial patch was used in all but one case after removing the pulmonary valve and widely resecting the infundibulum. In one case, a Goretex patch was used.

3. Results

Perioperative mortality was zero. Postpalliation angiography was performed after 5-14 months in all patients (Fig. 2). Pulmonary artery (PA) diameters were taken from the angiograms that showed an increase in the cardiovascular structures. The preoperative PA index

![Angiography of a patient with a severe hypoplasia of the pulmonary artery tree (diameter of the central right pulmonary artery (RPA): 4.0 mm; diameter of the central left pulmonary artery (LPA): 3.5 mm).](image1)

![Representative angiography after palliative reconstruction of the right ventricular outflow tract. The tract is nicely enlarged and the pulmonary vessels exhibit a good orthograde flow.](image2)
3.1. Complications

One patient died 10 months postpalliation due to pneumonia, one patient received a pacemaker after repair and died 2 months post-repair due to failure of the pacemaker; a 5-year-old patient died 1 month after repair due to sepsis.

3.2. Follow up

All eight long-term survivors (12-17 years) are in excellent clinical condition. Echocardiography revealed good right ventricular function and near normal diameters at RV peak pressures between 25 and 50 mmHg. Only one patient developed brady-arrhythmia. A pacemaker was implanted 8 years after repair and 2 years after homograft implantation.

4. Discussion

4.1. How should hypoplastic vessels be quantified?

The larger group of our patients included nine patients with secondary hypoplasia, i.e. the pulmonary arteries were not adequately developed owing to an infundibular and/or valvular pulmonary stenosis. In contrast, the two patients in the smaller group without catch-up growth after right ventricular outflow tract (RVOT) enlargement presented obviously a primary hypoplasia, i.e. the pulmonary arteries had presumably no potential for growth. Because antegrade flow was optimal and pressures in the central pulmonary arteries were elevated, a mechanism seemingly exists preventing adequate growth. Perhaps, further embryological and histological studies can shed some light on that mechanism.

Some pulmonary arteries are thick and somewhat hypoplastic. These vessels have exactly the same size in the operating room and on the angiogram. Other pulmonary arteries are soft and pliable. They are found to be larger in the operating room than on the angiogram (see discussion: [6]). With this diagnostic shortcoming, the preoperative decision is difficult.

By the majority, angiography underestimates the true size of the pulmonary artery because of the preoperatively decreased pulmonary blood flow [4]. This observation suggests that after palliation, those pulmonary arteries grew better, the smaller these vessels initially were. This finding was confirmed by several authors [7–10]. Our results are in concert with these results only in part, as in 2 of our 11 patients with the smallest pulmonary vessels, postoperative growth was only limited. Angiography can be replaced by MRI in older children but is difficult in small babies, especially in the ICU.

The fact that pulmonary regurgitation after TOF repair is common—especially in patients who required a transannular patch reconstruction at the time of repair—presents another aspect of our experience. Hence, in the long run, the patients of this study had severe pulmonary valvular insufficiency that required valve replacement.

As indications for pulmonary valve replacement (PVR) after TOF repair, we included symptoms as decreased exercise tolerance and poor functional class, development/progression of arrhythmias (especially ventricular arrhythmias), evidence of deteriorating RV function, and development/progression of tricuspid regurgitation. In a recent study from the Mayo clinic, RV size was reduced in most patients after PVR. However, this reduction did not reach statistical significance, in spite of the markedly improved RV function [11].

Our preference over the years has been to advise PVR relatively early trying to avoid the expected, irreversible anatomic and pathologic changes in the myocardium.

4.2. Is catch-up growth of the pulmonary arteries within reach?

Palliative reconstruction of the RVOT seems to be a good alternative to treat neonates and infants with severely hypoplastic pulmonary arteries. This reconstruction was already suggested in 1986 to achieve adequate pulmonary arterial enlargement in patients with pulmonary atresia, ventricular septal defect, and hypoplastic, confluent pulmonary arteries [12].

The disadvantages of causing a pulmonary insufficiency and the inevitable need for re-operation (correction) after the initial procedure with the heart-lung-machine did not play a major role in our hands. Obviously, close monitoring...
of the patients after palliative RVOT enlargement is mandatory [13-15].

4.3. Can catch-up growth of the pulmonary annulus be achieved to circumvent transannular RVOT reconstruction?

A rule of thumb to calculate the pulmonary artery tree was already suggested in 1975 [16] and was later termed 'McGoon ratio' [17]. According to this rule, the sum of the diameters of both pulmonary arteries is divided by the diameter of the thoraco-abdominal aorta: if the ratio is greater than one, total correction is feasible.

The pulmonary annulus index and the index of the two pulmonary arteries correlated only poorly ($r=0.3$), suggesting that the hypoplasia of the pulmonary annulus is only loosely coupled with the hypoplasia of the right and left pulmonary artery. The above study also showed that a reduction in the extent of transannular patch enlargement was not attainable by performing a Waterston-shunt or a Blalock-Taussig-shunt.

As to theoretical considerations, a catch-up growth of the pulmonary annulus cannot take place after systemic-pulmonary shunts. Yet, these considerations are not unanimously accepted: one observation is supportive [10] while others are not [7,8].

On the other hand, performance of a functioning Waterston-shunt or a classical Blalock-Taussig-shunt is extremely demanding in neonates with severe hypoplastic pulmonary arteries. One study reports on a high percentage of patients with significant alterations of the pulmonary artery anatomy as a result of previously performed systemic-pulmonary shunts [18].

4.4. RV dilatation and arrhythmia

Persisting RV dilatation and dysfunction is known to be substrates for ventricular arrhythmias and sudden death.

For patients with known paroxysmal ventricular tachycardia, complex ventricular premature beats, or a history of syncope or sustained palpitations, we perform a preoperative electrophysiology study. If ventricular arrhythmias were present postoperatively—in particular in the presence of RV dilatation—prophylactic implantation of an automatic cardioverter-defibrillator is considered seriously after additional electrophysiological examination.

For patients with documented preoperative supraventricular arrhythmias, we prefer to perform concomitant appropriate antiarrhythmic procedures as right-sided maze, cryoablation, division of accessory conduction pathways at the time of PVR.

4.5. Summary and conclusion

(1) The above method for estimating the size of the pulmonary vessels is adequate and permits assessment of the severity of TOF and of the success of surgery. (2) Enlargement of the RVOT provides the best possibility for symmetrical growth. (3) In very severe forms of TOF, palliative right ventricular outflow tract construction may provide the potential for complete repair. (4) In our high-risk patient group mortality was not related to the hypoplastic pulmonary arteries. (5) Obviously, all patients need pulmonary valve implantation in the long run.

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References