Antegrade palliation for diminutive pulmonary arteries in Tetralogy of Fallot

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Abstract

Objectives: The purpose of this study was to evaluate the outcome following palliative reconstruction of right ventricular outflow tract in Tetralogy of Fallot (TOF) with diminutive pulmonary arteries with central and peripheral stenosis. Methods: Between 1986 and 1999 in 15 children with the diagnosis of TOF palliative reconstruction of the right ventricular outflow tract without closure of the ventricular septal defect (VSD) was performed. All patients were not suitable for an AP-Shunt because of a diminutive pulmonary vascular bed. Six patients were younger than 1 year at operation. Results: There was one hospital death (6.7%) in a child with additional aortic valve insufficiency in multi-organ failure. Although the postoperative course was prolonged (median duration on ICU: 8 days) and complicated by congestive heart failure, clinically the 14 patients discharged improved significantly. The arterial oxygen saturation increased from 67 to 93% (P = 0.001), the hemoglobin decreased from 16.1 to 13.3 g/l (P = 0.02) and hematocrit from 0.52 to 0.40 (P = 0.06). In control angiography, the McGoon Index increased in the average from 1.01 to 1.95 (P < 0.001). VSD closure was performed in 12 patients (median: 2.5 years after initial operation) with one perioperative death. A homograft had to be implanted in seven patients and a mechanical prosthesis in the right ventricular outflow tract in one. One late death occurred due to ventricular arrhythmia 12 years after antegrade palliation (11 years after corrective operation). Conclusions: The antegrade palliation seems to be an adequate strategy for the treatment of selected children with diminutive pulmonary arteries in TOF, who were not candidates for primary correction or an AP-Shunt. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Tetralogy of Fallot; Congenital heart defect; Palliative operation; Right ventricular outflow tract enlargement; Diminutive pulmonary arteries

1. Introduction

The treatment of patients with Tetralogy of Fallot (TOF) and diminutive pulmonary arteries remains a challenge. There are several therapeutic approaches to manage such patients: (1) non-surgical therapy (medical and/or balloon angioplasty), (2) primary repair, (3) systemic artery to pulmonary artery shunt (classic or modified BT-Shunt), and (4) right ventricular outflow tract enlargement without ventricular septal defect (VSD) closure (‘antegrade palliation’) [1–5]. The best therapeutic strategy depends on the clinical condition of the patient and the pulmonary artery anatomy. It is difficult to clearly define diminutive pulmonary arteries as the McGoon ratio [6] and the Nakata Index [7] are related to the more central portion of the pulmonary arteries only. But the evaluation of the peripheral pulmonary anatomy is mandatory. Therefore, the decision to follow the best therapeutic approach is based on the surgeon’s experience.

In this report, we describe the outcome of 15 consecutive patients following palliative enlargement of the right ventricular outflow tract without VSD closure in TOF with diminutive pulmonary arteries.

2. Material and methods

2.1. Patients

Between 1986 and 1999, 199 patients with TOF were referred for surgical treatment. Fifteen children (7.5%) were assessed as having diminutive pulmonary arteries (severely hypoplastic pulmonary arteries with central and peripheral stenosis). These children are the subject of this
report. All 15 children were symptomatic at the time of presentation and underwent palliative reconstruction of the right ventricular outflow tract without VSD closure (ante- 
grade palliation). There were six female and nine male children. The age ranged from 3 weeks to 14 years (mean age: 3.5 years). All patients showed cyanosis and tachypnea at presentation. The mean preoperative arterial oxygen saturation ratio was 67%, the mean hemoglobin was 16.1 g/l and the mean hematocrit value was 0.52. Previous procedures were performed in three children: interventional balloon angioplasty of pulmonary artery stenosis in two children and a Waterston shunt in one. All patients underwent preoperative cardiac catheterization. To describe the central portion of the pulmonary arteries the McGoon ratio (ratio of the summed diameter of the right and left pulmonary arteries to the diameter of the descending thoracic aorta) was measured [6]. The average of the McGoon ratio was 1.01 (range: 0.65–1.4). Multiple aortopulmonary collateral arteries were present in two children.

Data are presented as the mean ± standard deviation. Continuous variables were compared with Student’s t-test. P-values <0.05 were considered significant.

2.2. Operative technique

All operations were performed via a median sternotomy with cardiopulmonary bypass. In 12 children deep hypothermic circulatory arrest and in three children moderate hypothermia (28°C) and cardiac arrest with cold hyperkalemic crystalloid cardioplegic solution was used. A short longitudinal right ventricular incision opening the infundibulum was made. In all but two children the ventriculotomy was extended across the main pulmonary artery trunk. After resection of infundibular muscle a patch (autologous pericardial in six and expanded polytetrafluoroethylene patch (Gore-Tex) in nine children) as small as suitable was inserted to enlarge the right ventricular outflow tract. Additionally in four children peripheral pulmonary artery stenosis was enlarged using an autologous pericardial patch. One child had simultaneous surgical closure of a main aortopulmonary collateral artery.

3. Results

There was one hospital death (6.7%) in a child with additional aortic valve insufficiency and multiple aortopulmonary collateral arteries due to congestive heart failure 8 days after antegrade palliation. The median duration on ICU was 8 days. The immediate postoperative course was mainly complicated by congestive heart failure. Eleven of 14 children were discharged with a medication of digitoxin and nine with diuretics. Clinically the 14 children discharged improved substantially. The arterial oxygen saturation increased from 67 ± 7.2 to 93 ± 5.9% (P < 0.001), the hemoglobin decreased from 16.1 ± 2.5 to 13.3 ± 1.2 g/l (P = 0.02) and the hematocrit decreased from 0.52 ± 0.1 to 0.40 ± 0.09 (P = 0.06).

Control cardiac catheterization was performed 4 weeks–24 months after the antegrade palliation depending on the functional status of the child (Figs. 1 and 2). The size of the pulmonary arteries (McGoon ratio) increased substantially from 1.01 ± 0.2 to 1.95 ± 0.5 (P < 0.001). Twelve of 14 children underwent VSD closure (total correction) after a median time of 2.5 years (range: 5 weeks–11 years). Additionally, implantation of a homograft was necessary in seven children and a mechanical heart valve prosthesis (St. Jude Medical) in one child. There was one perioperative death after correction due to right heart failure. Of the remaining two children, one is waiting for total correction and in the other child permission for VSD closure was denied by parents. The age at operation had no influence on these results.

The mean follow-up was 8.1 ± 3.5 years. One late death
occurred 12 years after antegrade palliation (11 years after VSD closure). The patient suffered from ventricular arrhythmias. The remaining ten children with VSD closure are doing well without signs of heart failure.

4. Discussion

Early corrective repair of TOF is nowadays a routinely used approach in patients with normally related pulmonary arteries [8–10]. Children presenting small pulmonary arteries and/or a hypoplastic pulmonary anatomy, can be treated successfully with staged correction: systemic-pulmonary artery shunt (Blalock–Taussig shunt) followed by complete repair [1,11]. However, treatment of children with TOF and diminutive pulmonary arteries with central and peripheral pulmonary stenosis is controversial. Most of these patients do not tolerate primary repair. Several approaches to develop such diminutive pulmonary arteries have been investigated. After the Blalock–Taussig shunt, growth of the pulmonary arteries was demonstrated by Gale et al. [12]. But others reported about pulmonary artery hypoplasia and pulmonary artery distortion and stenosis after the Blalock–Taussig shunt [13]. Thus, this seems to be not the optimal therapeutic strategy for all patients with TOF and diminutive pulmonary arteries.

Our approach in the treatment of TOF and diminutive pulmonary arteries with central and peripheral stenosis, as reported by others [2,4,5,14,15] has been to enlarge the right ventricular outflow tract using cardiopulmonary bypass as the first step. We called this the antegrade palliation. The antegrade palliation with increased early pulmonary artery blood flow offers substantially greater and uniform growth of the pulmonary valve annulus, of the central and peripheral portion of the pulmonary arterial tree, improves arterial oxygen saturation and increases angiogenesis of distal microvessels [4,5,16]. Our data support these hypotheses as 86% of the patients showed a sufficient pulmonary artery growth after antegrade palliation and underwent total correction. Moreover, implantation of a homograft was not necessary in four of 11 patients with successful VSD closure as a sign of adequate central pulmonary artery growth. These patients are doing well without signs of right heart failure.

Disadvantages of antegrade palliation resulting in congestive heart failure requiring prolonged ICU treatment in the immediate postoperative course are the requirements of cardiopulmonary bypass and the possible postoperative excessive pulmonary blood flow. An important limiting factor for postoperative excessive pulmonary blood flow is the pulmonary artery size itself. Therefore, the precise preoperative quantification of the pulmonary arteries is even more important, but remains uncertain.

The hospital mortality rate of 6.7% in this group of patients was favourable to others [4].

After discharge, close monitoring of the patients is mandatory to determine the optimal time of total correction. In younger patients the greatest increase in pulmonary arterial growth occurs soon after antegrade palliation [2,4]. Therefore, control cardiac catheterization was performed in the first year after antegrade palliation. In older patients cardiac catheterization was performed later on depending on the functional status and the echocardiographic findings. The majority of our patients showed a sufficient pulmonary artery growth and the VSD closure was performed after a median time of 2.5 years.

The antegrade palliation seems to be an adequate strategy for the treatment of selected children with diminutive pulmonary arteries in TOF.

References


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Appendix A. Conference discussion

Dr G. Sarris (Athens, Greece): Clearly, your approach is one option for patients with truly diminutive pulmonary arteries, especially in the peripheral period, I am wondering, however, if one encounters severe heart failure in the perioperative period, isn’t this an indication that at least some of these patients could have withstood a completely corrective operation?

Dr Vazquez-Jimenez: Well, the most difficult part of all this process is making the decision which patients are candidates for this. Once you open the right ventricular outflow tract and enlarge it, there is a big flow over the pulmonary arteries, but if you close the VSD, there was a study in 1993 made by Rome and Castaneda in which they divide the patients in four different groups. One of them was trying to make the complete correction. From nine patients mortality was seven patients. So it doesn’t function.

The problem is how big needs to be the communication between the right ventricle and pulmonary arteries. There was only one study in the literature made by the Japanese, it was very interesting, also in 1990, and they showed that if the communication tends to be very high, very big, the patients die in the intensive care unit because of right ventricle failure. So it needs to be a compromise. What is surprising is these patients are individually very different. Some of them we need to make the correction very soon, some of them, one patient didn’t grow up the pulmonary artery, so that is the one patient still waiting for VSD closure.

Dr S. Sano (Okayama, Japan): I think the main aim of your approach, the antegrade palliation, is to increase the size of the main pulmonary artery, on the other hand, usually Blalock–Taussig shunt increases the right and left pulmonary arteries, may increase the main pulmonary artery too. The aim to me, I have done a few patients the same, not tetralogy of Fallot, pulmonary atresia intraventricular septum, the aim I did was to increase the main pulmonary artery size and then in complete repair I could do like a transatrial–transpulmonary repair without pulmonary like a homograft or a valve. But even if the main pulmonary artery increased, why did you put like a homograft or the mechanical valve into these patients?

Dr Vazquez-Jimenez: We put the mechanical valve and homografts in these patients in the complete correction, but the first time we only opened the right ventricular outflow tract. And I think this is the aim of discussion in the literature, if it is better to put a shunt or make this procedure. The shunt needs to have at least some degree of growth in the pulmonary artery. So below 5 mm it will be very difficult to get a good shunt function. And if you open the right ventricular tract, the idea is the pulsatile flow is going not only to open the central portion of the pulmonary arteries, so as you see here, the peripheral portion also. So you can close the VSD afterwards.

Dr Sano: I think from my experience an RV-PA shunt doesn’t increase the pulmonary artery size compared to the classical BT shunt, because in the classical BT shunt you have pulsatile flow in diastole and also systole, but in the antegrade flows you have only systolic flow, not diastolic flow. So if your aim is to increase the pulmonary artery size, usually the BT shunt increases the pulmonary artery size more.

I told you, the aim of my antegrade approach in a few patients, not tetralogy of Fallot, was to increase the main pulmonary artery size and also the anulus of the pulmonary valve, and that means in complete repair you can create like a tricuspid or a bicuspid pulmonary valve and then to avoid the mechanical valve or homograft in the pulmonary circuit.

Dr Vazquez-Jimenez: Well, I think that this is a completely different problem here. This is not tetralogy of Fallot with pulmonary atresia but tetralogy of Fallot with an open valve. So the idea of this study was to look if these patients, if in the physiology of these patients the pulmonary growth is different than in pulmonary atresia, and we were very surprised to see the results.

Dr D. Metras (Marseille, France): It looks to me, if I remember well, that on the angiogram you showed preoperatively that there was a very thickened pulmonary valve. Now, would you consider in those cases just a balloon dilatation that would result in the same strategy of making the pulmonary artery grow rather than do a first operation?

Dr Vazquez-Jimenez: Well, for sure that is a strategy, but the experience we have with our cardiologists making a balloon dilatation in such a pulmonary valve was not the best one. So I will apply for our system at the moment. But, for sure, it is a good alternative. It is functional.