The role of pulmonary artery anatomy in repair of tetralogy of Fallot

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Pulmonary artery anatomy is the key factor that determines the type of surgical treatment required in tetralogy of Fallot. Despite the fact that routine primary repair is now done on infants, inadequate pulmonary artery size can dictate the need for staged surgical repair in even the oldest age groups. From October 1986 to October 1998, 361 patients at our clinic underwent surgery to correct tetralogy of Fallot. A total of 292 cases were treated with primary repair, 69 surgeries were palliative, and 30 of these 69 underwent corrective surgery. The Nakata index was used as a pulmonary artery index (PAI), and PAI< 200 was the criterion for requirement of two-stage repair.

Of the 30 patients that underwent staged repair, the Blalock-Taussig shunt (BTS) procedure was used in 24; the remaining six patients had right ventricular outflow tract reconstruction (RVOTR). The mean age of all the palliative surgery patients was 3.4 years (range 6 months to 11 years), and of those who received corrective surgery was 5.5 years (range 2-12 years). These patients' PAI values were 181 ± 37.5 mm^2/m^2 and 359 ± 130.7 mm^2/m^2, respectively. The period between the two operations ranged from two months to four years.

Mortality rates were 2.8 percent for palliative surgery as a whole, 4.1 percent for primary repair, and 16.6 percent for staged repair. Our policy with regard to corrective surgery for tetralogy of Fallot is to do primary repair regardless of a patient's age and weight, except in cases where the pulmonary artery anatomy is appropriate for the patient's body size.

Key words: Fallot tetralogy, Blalock-Taussig shunt.

Surgical strategy in cases of tetralogy of Fallot has been discussed since the first repair was done by Lillehei in 1954[1]. A two-stage approach was adopted initially due to the excellent results that were achieved[2,3] but, in the early 1990s, several studies of primary repair revealed comparable results[4-8].

In two-stage repair, the most common palliative procedure is the Blalock-Taussig shunt (BTS), which is then followed by total repair at an older age. In an asymptomatic patient with a hypoplastic pulmonary artery, delay of corrective surgery does not lead to improvement of the pulmonary artery tree with age. Therefore, some researchers have advised right ventricular outflow tract reconstruction (RVOTR) and have disputed the importance of pulmonary artery size. In 1980, Naito et al.[9] developed a list of criteria to indicate the need for RVOTR in corrective repair, and four years later Nakata et al.[10] published a method for calculating a patient's pulmonary artery index (PAI). In this study, we discuss two-stage repair of tetralogy of Fallot with a focus on pulmonary artery anatomy.

Material and Methods

Between October 1986 and October 1998, 361 patients at our clinic underwent surgery for

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tetralogy of Fallot. Patients with pulmonary atresia or complex congenital cardiac anomalies were excluded from the study. Primary repair was carried out in 292 cases and 30 patients underwent two-stage repair. Palliative procedures alone were done on the remaining 39 patients, all except for two of whom were waiting for corrective surgery.

The BTS technique was used in 24 patients. In 21 cases this technique, the treatment of choice at our clinic in terms of palliative procedures, was done based on low PAI. Three patients with good PAIs had BTS operations under emergency conditions due to intractable cyanotic attacks. Goretex grafts were used in 23 patients, and one underwent a classical BTS procedure (Table I).

Table I. Palliative Procedures Performed

RVOTR	6 cases
BT Shunt	24 cases
Right modified	2
Left modified	21
Right classic	1

RVOTR: Right ventricular outflow tract reconstruction.
BT : Blalock-Taussig.

Right ventricular outflow tract reconstruction (RVOTR) was performed in six cases, the main indication for this approach being asymmetry of the left and right pulmonary arteries. In fact, two patients in this group had good PAIs but their left pulmonary arteries were very narrow, so RVOTR was performed in an attempt to encourage growth of the left pulmonary artery. Autogenous pericardium was used in five of the RVOTR cases, and the width of the pericardial graft was determined according to patient weight and height, as described by Naito[9]. In one patient, the width of the outflow patch was readjusted at the level of the pulmonary annulus after the termination of cardiopulmonary bypass; flow ratios were calculated in the operating room in order to prevent pulmonary overflow. The PAI and age information on the 30 patients prior to their palliative and corrective repair procedures are shown in Table II.

Primary repair was carried out in 292 patients. Ventricular septal defects were closed using dacron patches with interrupted sutures. Most cases had extensive myectomy in the septoparietal and ventriculo-infundibular fold regions, and the outlet septum was almost totally resected, as described by Kurosawa et al.[11]. Autogenous pericardium was used for outflow patches.

Results

The mean PAI of patients that underwent palliative procedures was 181 ± 37.5 mm^2/m^2, and this rose to 359 ± 130.7 mm^2/m^2 prior to corrective surgery. The mean time interval between the two operations was 23 months (range 2 to 48 months).

Of the BTS patients, one underwent postoperative exploration for hemorrhage and two experienced late obstructions. Pulmonary artery distortion was diagnosed at the anastomosis sites in two other cases. In the RVOTR patient group, the growth of the left pulmonary artery was insufficient with regard to the right in one case, but in other instances the growth of both pulmonary arteries was adequate. In the primary repair group, transannular autogenous pericardial patches were used in 259 (88%) cases, with the right ventriculotomy closed primarily in only one patient. In the remaining 32 individuals, closure of the right ventriculotomy was achieved using autogenous pericardium. Of the patients who had undergone previous palliative procedures, a transannular patch was used in 26 (86%). The operative mortality rates were 4.1 percent (12 patients) in the primary repair group and 16.6 percent (5 patients) for the staged repair group. Overall mortality for all the palliative procedures done on tetralogy cases at our clinic was 2.8 percent (two of 69 patients) (Table II).

Discussion

The aim in surgery for tetralogy of Fallot is to close the ventricular septal defect completely and to relieve the right ventricular outflow tract. Any residual pathology can place the patient in

Table II. Patient Age and Pulmonary Artery Index (PAI) in Relation to Procedure and Associated Mortality

	Age	PAI	Mortality Rates (%)
Palliative procedure	3.4 yrs (6 mo-11 yrs)	181(± 37.5)	2.8*
Two-stage repair	5.4 yrs (2-12 yrs)	359 (± 130.7)	16.6
Primary repair	4.6 yrs (8 mo-14 yrs)	363 (± 54.3)	4.1

* overall mortality in 69 patients.

a critical condition, especially during the early postoperative period. To achieve total correction of this anomaly, the patient must either undergo a staged or primary repair strategy. Initially, two-stage repair gained popularity[2,3] based on earlier studies that reported low mortality rates, but today many experts cite the advantages of primary repair at any age[4-8].

The main purpose of staged repair of tetralogy of Fallot is to reduce the risk of mortality in the neonatal period. The palliative portion of this strategy, usually BTS, can be performed at relatively low risk during the neonatal stage. Corrective surgery two to three years after the first operation completes this accepted operative procedure[2]. Despite the excellent results achieved with staged repair, it has been discovered that palliative procedures are also associated with high mortality and morbidity rates. Complications with pulmonary artery distortion and partial or total occlusion after BTS operations are not uncommon. Authors have reported post-BTS pulmonary artery distortion rates of 18 to 36 percent[12-14].

In our series, pulmonary artery distortion was observed in two patients (8.3%), and total occlusion of the ipsilateral pulmonary artery was diagnosed in one individual (4.1%) through angiography. A total of 12.5 percent of the patients in our series had pulmonary artery abnormalities due to previous BTS operations.

In our study, the mortality rate for staged corrections was higher than that of primary repair, but the decision to perform staged repair was not related to patient age and body weight. The only indication for palliation in our series was low PAI. In his original article, Nakata[10] stated that a PAI above 100 indicated corrective surgery, but he still discussed the postoperative heart failure problems of patients whose PAIs were between 100 and 150. It is our experience that a PAI above 200 is safer in terms of the postoperative period. Wu[15] stated that tetralogy of Fallot could be corrected under any circumstances with a detailed evaluation of the patient, but we noted that, in cases with low PAI, underdevelopment of the pulmonary artery was not restricted to major branches. Under such circumstances, graft reconstruction to the level of the hilus does not relieve the pressure gradient between the pulmonary artery and the hilus. This results in elevated right ventricular pressure and high mortality.

Mortality is closely related to residual pulmonary stenosis, which is easily assessed by calculating RV/LV (right ventricular pressure, left ventricular pressure) ratio. An RV/LV greater than 0.8 coupled with a ratio of reconstructed pulmonary artery area to ideal pulmonary artery area for that body surface area of less than 0.7 predict increased risk of mortality[9,16]. These two parameters are directly related to PAI. Unless a patient has adequate pulmonary artery size, surgical procedures done proximal to the hilus do not result in good RV/LV results.

We conclude that the operative mortality in corrective surgery for tetralogy of Fallot depends on several factors. As we presented here, problems with pulmonary artery anatomy, especially distal stenosis, are the most important features to correct. We performed two-stage corrective surgery on 27 patients in order to obtain appropriate pulmonary artery sizes and decrease mortality, though this rate remains high compared to that for primary repair.

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