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Staged Procedures versus Primary Repair for Tetralogy of Fallot and Small Left Ventricle

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ABSTRACT

Introduction: This study was conducted to address whether staged surgical intervention or primary repair would be preferred for treating patients with tetralogy of Fallot and small left ventricle.

Methods: We retrospectively assessed 28 patients with tetralogy of Fallot and small left ventricle at the Chinese PLA 208th Hospital, Changchun, between January 2005 and December 2008. Of this cohort, 11 patients had undergone a systemic-to-pulmonary arterial shunt with a polytetrafluoroethylene interposition graft (off pump), followed by ancillary repair procedures as a surgically staged group; the remaining 17 patients underwent primary intracardiac repair via cardiopulmonary bypass. The oxygen level as measured by pulse oximetry (SpO2), the McGoon ratio, and the left ventricular end-diastolic volume index (LVEDVI) were measured before shunt procedures and at 6 to 18 months afterwards. All data were analyzed statistically with the paired Student t test. The 2 groups were compared with respect to LVEDVI values and McGoon indices at baseline by the independent t test.

Results: None of the patients died as a direct result of the surgery, but 1 fatality due to pneumonia occurred 4 months after shunting. Mean postshunt values for SpO2, the McGoon ratio, and the LVEDVI increased significantly from 76.8% ± 9.0% to 87.5% ± 2.4% ($P < .01$), from 1.42 ± 0.27 to 1.62 ± 0.32 ($P < .05$), and from 20.1 ± 7.0 mL/m2 to 34.3 ± 7.4 mL/m2 ($P < .01$), respectively. Nine patients were fully repaired within the 6- to 18-month time frame after shunting, while 1 patient awaited definitive surgery for pulmonary hypoplasia. In the primary-repair group, in which the increases the LVEDVI and McGoon ratio were comparatively higher, 1 patient died of hypoxemia.

Conclusion: A LVEDVI 20 mL/m2 may be an indication for primary repair in patients with tetralogy of Fallot. With values <20 mL/m2, staged procedures (ie, shunt with a polytetrafluoroethylene interposition graft) can promote left ventricle development and allow safe transition to the final repair.

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INTRODUCTION

Tetralogy of Fallot is the most common of the congenital heart diseases that lead to cyanosis [Fyler 1980]. Although there is debate as to whether the left ventricle is actually hypoplastic in this condition, early in 1970, Kirklin [1970] equated a postoperative low output with a reduced left ventricular size. Although excessively small left ventricles are seldom encountered with tetralogy of Fallot, small left ventricles do affect surgical outcomes. For our purposes, the diagnosis of small left ventricle was based on the left ventricular end-diastolic volume (LVEDV). We describe our experience with staged versus primary-repair operations in this setting.

MATERIALS AND METHODS

From January 2005 to December 2008, 182 patients with tetralogy of Fallot underwent operation at the Chinese PLA 208th Hospital, Changchun. Of these patients, 28 were associated with a LVEDV index (LVEDVI) <30 mL/m2 and were analyzed retrospectively. Eleven of the 28 patients (median age, 9.3 years; median weight, 23.6 kg) had undergone systemic-to-pulmonary arterial shunting, and the other 17 patients (median age, 4.8 years; median weight, 14.7 kg) were treated with primary surgical repair. Our investigation included reviews of hospital charts, echocardiographic exams, operative reports, and follow-up records. In each instance, diagnosis was based on Doppler echocardiography and cardiac catheterization results. All patients experienced transient episodes of breathlessness, fatigue, increasing cyanosis, progressive arrhythmias, and paradoxical emboli. Electrocardiograms showed right ventricular enlargement, and cardiothoracic ratios (range, 0.45-0.70) were calculated from chest radiographs. LVEDV estimates were obtained on 3 occasions by Simpson means through Doppler echocardiography (LVEDVI = Mean LVEDV Value Body Surface Area), and McGoon ratios were estimated at 6 to 18 months postsurgically.

The data were recorded as the mean SD, and paired Student t tests or independent t tests were then used with SPSS software (version 11.0; SPSS, Chicago, IL, USA) for statistical analyses. Statistical significance was set at P values <.05.

Surgical Procedures

In our study group, primary surgical repair was achieved with cardiopulmonary bypass, whereas shunting was performed off pump via median incisions. Central aortopulmonary shunts (3 patients) and shunts from the ascending aorta to the right pulmonary artery [Iyer 1991] (8 patients) were performed by using Gore-Tex grafting. Ventricular septal defects were closed with a Dacron patch, and right ventricular outflow tracts were relieved by placement of either pericardial patches (3 patients) or transannular patches (14 patients). Atrial septal defect, patent foramen ovale, and left superior vena cava were the only additional defects found in some patients.

RESULTS

None of the patients who underwent staged procedures died from the surgery, but 1 fatality due to pneumonia occurred at another center 4 months after the shunting procedure. Another patient required a repeat thoracotomy 8 hours after shunting because of hemorrhage. This patient was discharged 3 weeks later, however, after recovery from an incisional infection. All of the patients' clinical symptoms otherwise improved with shunting, and digital oxygen levels as measured by pulse oximetry (SpO2) rose. After 6 to 18 months, echocardiography evaluations showed that shunted vessels were patent and that left and right pulmonary arterial diameters had increased considerably relative to their preoperative baseline values. LVEDVI values increased by 70% (Table 1). Nine patients were fully repaired within the 6- to 18-month time frame, while 1 patient awaited definitive surgery for pulmonary hypoplasia.

Table 1. The Oxygen Level as Measured by Pulse Oximetry (SpO_2), the McGoon Ratio, and the Left Ventricular End-Diastolic Volume Index (LVEDVI): Comparisons of Pre- and Post-Shunt Values

	Before Shunt	After Shunt	P*
SpO_2, %	75.5 ± 8.4	87.3 ± 2.4	.000 (t = 5.169)
McGoon ratio	1.38 ± 0.24	1.62 ± 0.32	.026 (t = 2.607)
LVEDVI, mL/m^2	19.0 ± 6.2	34.0 ± 7.6	.000 (t = 5.700)

*Independent Student t test.

One postsurgical death from hypoxemia was recorded in the primary-repair group, but the remaining 16 patients recovered successfully and were discharged. The mean McGoon ratio was higher in this group, compared with the group of patients who underwent staged shunting procedures (1.85 ± 0.33 versus 1.42 ± 0.27). The mean LVEDVI was also higher in the primary-repair group (26.2 ± 2.6 mL/m2 versus 20.0 ± 6.9 mL/m2; Table 2).

Table 2. The McGoon Ratio and Left Ventricular End-Diastolic Volume Index (LVEDVI): Shunt versus Primary-Repair Groups

	Shunt Group	Repair Group	P*
McGoon ratio	1.38 ± 0.24	1.86 ± 0.33	.000 (t = 4.142)
LVEDVI, mL/m^2	19.0 ± 6.2	26.2 ± 2.6	.003 (t = 3.630)

*Independent Student t test.

DISCUSSION

Most treatment centers recommend primary repair of tetralogy of Fallot as early as possible [Jonas 2004; Song 2009]. Generally, it is believed that an LVEDVI 30.0 mL/m2, a McGoon ratio 1.2, a pulmonary arterial index 150.0 mm2/m2, or existing coronary artery anomalies (making it difficult to suture/patch the right ventricular outflow tract) are indications for palliative surgery. In patients with tetralogy of Fallot and severe cyanosis, however, the LVEDV is typically normal or mildly diminished. Only in rare cases are the left ventricle and the mitral valve truly hypoplastic, which entails an LVEDVI <30.0 mL/m2 as an obstacle to reparative surgery. Furthermore, the physiological basis of a small left ventricle is rather complicated. Although pulmonary stenosis restricts flow to the left atrium, thereby reducing the left ventricular blood volume and promoting a small left ventricle, significant degrees of left ventricular hypoplasia are likely unrelated to such dysfunction [Kouchoukos 2003] and may simply represent inherent defects, as with other complex congenital heart diseases.

Some experts do feel that an LVEDVI 30.0 mL/m2 is a requirement for primary repair. Naito et al [1985] described a series of 91 patients with tetralogy of Fallot treated with intracardiac repair between 1978 and 1981. One patient with an LVEDVI of 21.0 mL/m2 died from left-sided heart failure. In addition, 3 patients with LVEDVI values of 28.0 mL/m2, 30.0 mL/m2, and 32.0 mL/m2 underwent successful primary repair, although they required atrial pacing and catecholamine. Similarly, Jonas and DiNardo [2004] maintained that because patients with tetralogy of Fallot and pulmonary stenosis had sufficient oxygenation via the true pulmonary arteries to survive preoperatively, the true pulmonary arteries and pulmonary bed were adequately developed for a single-stage complete repair. They also believed that with a left ventricle length at least 80% of the distance from the aortic valve to the apex of the heart and mitral valve dimensions greater than 2.0 to 2.5 z scores below normal, the left ventricle should be capable of full cardiac output. A study by Graham et al [1977], on the other hand, concluded that when the LVEDV is <55% of the predicted normal value, a shunt procedure may be indicated as the initial surgery, with subsequent repair taking place 1 or 2 years later so that the left ventricle might enlarge in the interim.

In our study, the mean LVEDVI was 26.2 ± 2.6 mL/m2 for the patients who underwent primary repair, with the lowest value being 20.0 mL/m2. Postoperative outcomes in this group were considered good, although 1 patient died of hypoxemia. We thus concluded that although an LVEDVI

value 23.0 mL/m2 was preferable, a value 20.0 mL/m2 was a minimum requirement for primary repair. Elsewhere, Yi et al [1993] have asserted that an LVEDVI <24.0 mL/m2 is prohibitive for primary repair and that values between 24.0 mL/m2 and 30.0 mL/m2 may influence recovery but do not preclude active treatment. Our rationale was that if the pulmonary vessels are well developed, an LVEDVI as low as 20.0 mL/m2 is permissible for primary repair. Nevertheless, McGoon and LVEDV indices at baseline were higher in our primary-repair group than in the staged group ($P < .01$ for both).

Patients who underwent staged procedures received systemic-to-pulmonary arterial shunts for a small left ventricle or for a small left ventricle and pulmonary artery combined. Classic and modified Blalock-Taussig shunts or a central shunt operation were used largely for palliation. Polytetrafluoroethylene (PTFE) graft was chosen for blood-compatibility and patency reasons. In 8 of these 11 patients, ventricular and pulmonary angiography examinations recorded right-to-left ventricle ratios of 1:1 (3 patients), 1:0.8 (2 patients), and 1:0.5 (3 patients); McGoon ratios were <1.2 (3 patients), 1.2 to 1.5 (3 patients), and 1.5 to 1.8 (2 patients).

A shunt procedure is more suitable when the LVEDVI of these patients is <20.0 mL/m2 and there is also angio- and echocardiographic evidence of a hypoplastic pulmonary artery. Jarmakani et al [1969] found that left ventricular size correlated with a left-to-right or bidirectional shunt, meaning that the LVEDVI reflected pulmonary growth and flow conditions. One patient (weight, 18 kg) with preoperative tricuspid incompetence died of episodic pneumonia after a shunt from the ascending aorta to the right pulmonary artery was placed with a 5.0-mm PTFE graft. Excessive postshunt pulmonary flow and a poor right ventricular function were blamed. In another 3 patients, in whom central aortopulmonary shunts with PTFE grafts were performed, excessive pulmonary flow and heart failure also developed but were managed successfully until the repairs could be done. The burden of care between the shunting and the surgical repair might have been reduced if the initial repair had been done.

An echocardiography evaluation was performed 1 week after each shunt procedure. The mean left ventricular end-diastolic dimension in the long axis had increased from 21.5 ± 4.9 mm to 25.5 ± 4.5 mm ($t = 3.43$; $P = .006$), whereas the McGoon ratio was more likely to improve when initial values were >1.5 or the patient was older than 10 years. The underlying reasons for this may be that the small left ventricles were already quite functional or that their postoperative dimensions had increased dramatically. Changes were noted, in fact, several days after the shunt procedures. With transposition of the great arteries, a shift in the isoform of heavy-chain myosin occurs within hours following banding procedures. Measurements of the rate of muscle protein synthesis peak approximately 48 hours after altering the pressure load, so the time anticipated for change is measured in days rather than months [Izumo 1988; Jonas 2004]. It is therefore feasible that the left ventricle performs similarly under volume load, but when the left ventricle is extremely small, surgical repair might prove harmful in terms of hemodynamics. Nomoto et al [1984] had concerns that the preoperative size of the left ventricle might contribute to morbidity within the first 2 weeks of repair of tetralogy of Fallot, but it did not influence left ventricle performance over the long term.

In summary, our findings indicate that shunting with a PTFE vessel can be done safely in patients with tetralogy of Fallot and small left ventricle if the LVEDVI is <20.0 mL/m2. This approach prepares the left ventricle for later definitive repair. After shunting, however, the patient's hemodynamics should be monitored to adjust shunt flow as needed or to opt for an earlier repair. When the LVEDVI is 20.0 mL/m2 and the pulmonary artery is well developed, reparative surgery can be undertaken directly. Older children or adults usually withstand primary repair, because their small left ventricles are relative to right ventricular and septal hypertrophy. Advances in imaging (magnetic resonance and 3-dimensional ultrasound) have improved estimation of left ventricular size, so that the treatment algorithms for patients with tetralogy of Fallot and small left ventricle are now easier to navigate.

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