Single-Stage Complete Repair versus Multistage Repair of Tetralogy of Fallot with Borderline Pulmonary Arteries

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ABSTRACT

Background: Tetralogy of Fallot is the most common cyanotic congenital heart defect. Borderline pulmonary anatomy has been associated with a higher risk of mortality and morbidity. Strategies to manage this condition—namely, single-or multistage repair—have long been debated.

Objective: The overall outcomes of patients with tetralogy of Fallot with borderline pulmonary arteries (McGoon ratio 1.3 to 1.7) with regard to the need for a single-stage or multistage repair and the outcome of each surgical management were evaluated.

Patients and methods: A retrospective, nonrandomized comparative study designed to evaluate patient outcomes comprised 60 patients with tetralogy of Fallot with borderline pulmonary arteries who underwent surgery at the Cardiothoracic Surgery Academy, Ain Shams University, Cairo, Egypt, between January 2016 and December 2017. After gaining approval from the affiliated ethical and research committee, and informed consent of the guardians, the patients were assigned into one of two groups. Shunt group included 30 patients managed surgically by a modified Blalock-Taussig (MBT) shunt as a part of a multistage repair, and repair group included 30 patients managed surgically by single-stage complete repair. The medical records of the patients were reviewed, and data relating to age, sex, weight, and preoperative oxygen saturation were collected. All patients underwent preoperative echocardiography and multislice computed tomography (CT) with angiography. The follow-up was performed in patients who received a shunt once the echocardiography showed acceptable pulmonary arteries.

Results: The patients’ age ranged from 5 to 50 months with a mean age of 18.63 ± 9.15 (19.84 ± 12.34 for the shunt group and 17.43 ± 8.54 for the repair group). The weight ranged from 5 kg to 18 kg with a mean of 9.6 ± 2.53 (8.82 ± 2.79 for the shunt group and 10.41 ± 2.63 for the repair group). The mean preoperative O2 saturation was 68.95% ± 7.8% for the shunt group and 87.93% ± 6.18% for the repair group. The median McGoon ratio was 1.4 for the shunt group and 1.6 for the repair group, the difference of which was highly significant (P < .0001). The mortality rate in our study was 10% (10% for the shunt group and 10% for the repair group). The morbidity incidence rate was 26.6% for the shunt and repair groups. The ICU stay ranged from 2 to 31 days, with a median of three days for the shunt group (mean 3.61 ± 1.91) and four days for the repair group (mean 6.07 ± 6.63 days). The calculated P value showed a significant difference between the two groups concerning ICU stay. The postoperative SO2 significantly increased to a mean of 85.58 ± 7.05 in the shunt group and 98.14 ± 3.36 in the repair group (P < .0001).

Conclusion: There was no statistically significant difference between multistage repair and single-stage complete repair regarding morbidity and mortality. Regarding ICU stay, patients in the single-stage had a better outcome. A McGoon ratio of 1.5 can be used as a guideline in the decision-making process.

INTRODUCTION

Tetralogy of Fallot is a congenital cardiac malformation consisting of a ventricular septal defect (VSD), aortic overriding, infundibular pulmonary obstruction, and right ventricular hypertrophy [Anderson 1981]. It is the most common cyanotic heart disease, with a prevalence from 0.21 to 0.36 per 1000 live births and comprising approximately 7 to 10 percent of all forms of congenital heart diseases [Ferencz 1985; Mitchell 1971].

Tetralogy of Fallot had been uniformly lethal, but this changed after the introduction of the systemic to pulmonary shunt by Alfred Blalock. On November 29, 1944, he performed the first operation on a cyanotic 1-year-old child with tetralogy of Fallot. The Blalock-Taussig shunt had a high rate of success, and, by 1950, 1000 operations involving this shunt had been performed by Blalock and his team [Taussig 1979].

The first intracardiac repair of tetralogy of Fallot was performed by C. Walton Lillehei and his team at the University of Minnesota using controlled cross circulation. In 1955, Kirklin and colleagues used the first pump oxygenator to perform a total repair [Neill 1994].

The mortality rate in cases managed by single-stage complete repair has decreased substantially. Among 10 surgeries performed by Dr. Lillehei, only 6 of the patients survived [Lillehei 1955]. However, mortality was 0.5% among...
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The medical records of the patients were reviewed, and age, sex, weight, and preoperative oxygen saturation data were collected. All patients underwent preoperative echocardiography and multislice computed tomography with angiography. Postoperative data, including oxygen saturation, ICU stay, hospital stay, morbidities, and mortality were collected.

Shunt group was performed via median sternotomy. The choice of shunt size was based on the patient’s weight: a 4-mm shunt was used for individuals weighing 4–8 kg, and a 5-mm shunt was used for those >8 kg. Low-dose heparin (1 mg/kg) was given before opening the shunt. Air (21% O₂) was used for ventilation after declamping.

In single-stage complete repair, surgery was performed utilizing standard techniques of cardiopulmonary bypass, moderate hypothermia, and antegrade cold crystalloid cardioplegia. The transatrial transpulmonary approach was applied to all patients. First, the right atrium (RA) was opened, and the anatomy was examined. The VSD was closed, and infundibular resection was then performed via transatrial approach and, if required, was completed through a pulmonary arteriotomy. If the annulus was found to be inadequate, a transannular patch was utilized.

The follow-up was performed by echocardiography at discharge and at one month and six months after surgery. Multislice computed tomography with angiography was performed in patients who received a shunt once the echocardiography showed acceptable pulmonary arteries. The study was conducted between January 2016 and December 2017.

### Patients and Methods

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### Table 1. Comparison between Shunted and Repaired Patients with Regard to Preoperative Data

<table>
<thead>
<tr>
<th>Variable</th>
<th>Shunt Group (n = 30)</th>
<th>Repair Group (n = 30)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male, n (%)</td>
<td>17 (56.7)</td>
<td>21 (70)</td>
<td></td>
</tr>
<tr>
<td>Female, n (%)</td>
<td>13 (43.3)</td>
<td>9 (30)</td>
<td>.4216</td>
</tr>
<tr>
<td>Age, month</td>
<td>19.8 (12–24)</td>
<td>14.5 (12–21)</td>
<td>.338</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>8.8 (9–11)</td>
<td>10 (8–10)</td>
<td>.008</td>
</tr>
<tr>
<td>Preoperative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>O₂ saturation, mmHg</td>
<td>69 (68–72)</td>
<td>90 (85–92)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>MPA, mm</td>
<td>6.4 (6.2–7.1)</td>
<td>7 (5.7–8.5)</td>
<td>.183</td>
</tr>
<tr>
<td>RPA, mm</td>
<td>5.9 (5.3–6.3)</td>
<td>6.3 (5.5–7.8)</td>
<td>.092</td>
</tr>
<tr>
<td>LPA, mm</td>
<td>6 (5.7–7)</td>
<td>6 (5–7)</td>
<td>.994</td>
</tr>
<tr>
<td>McGoon ratio</td>
<td>1.4 (1.4–1.5)</td>
<td>1.6 (1.6–1.7)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>VSD, mm</td>
<td>11.8 (11.8–12.5)</td>
<td>9.15 (8–10)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>RVOT gradient, mmHg</td>
<td>79 (79–84)</td>
<td>80.5 (75–94)</td>
<td>.164</td>
</tr>
<tr>
<td>Aortic overriding degree</td>
<td>50 (50–50)</td>
<td>50 (40–50)</td>
<td>.513</td>
</tr>
</tbody>
</table>

Values are shown as median (range) where indicated. Bold indicates statistically significant P value.

MPA indicates main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; RVOT, right ventricular outflow tract.

### Table 2. Intraoperative Data*

<table>
<thead>
<tr>
<th>Variable</th>
<th>Shunt Group</th>
<th>Repair Group</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Variable, Shunt Group</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>n (%)</td>
<td>4</td>
<td>8 (26.7)</td>
<td></td>
</tr>
<tr>
<td>n (%)</td>
<td>5</td>
<td>22 (73.3)</td>
<td></td>
</tr>
<tr>
<td>Bypass time, min</td>
<td>99.16 ± 49.22 (25–293)</td>
<td>56.63 ± 24.12 (22–127)</td>
<td>.513</td>
</tr>
<tr>
<td>Cross-clamp time, min</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Values are shown as median (range) where indicated.
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Table 3. Comparison of Postoperative Data between Shunt and Repair Patients

<table>
<thead>
<tr>
<th></th>
<th>Shunt Group (n = 30)</th>
<th>Repair Group (n = 30)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>O₂ saturation, postop</td>
<td>85 (85–88)</td>
<td>99 (98–100)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>ICU stay, d</td>
<td>3 (3–4)</td>
<td>4 (3.75–5.25)</td>
<td>.0058</td>
</tr>
<tr>
<td>Hospital stay, d</td>
<td>8 (7.5–9)</td>
<td>9 (7–10)</td>
<td>.254</td>
</tr>
<tr>
<td>O₂ saturation, before discharge</td>
<td>86 (85.25–89.5)</td>
<td>99 (98.25–100)</td>
<td>&lt;.0001</td>
</tr>
</tbody>
</table>

Values are shown as median (range) where indicated. Bold indicates statistically significant P value.

The patients’ demographics and preoperative data are shown in Table 1. The patient age ranges from 5 to 50 months with a mean age of 18.63 ± 9.15 (19.84 ± 12.34 for the shunt group and 17.43 ± 8.54 for the repair group). There were 21 infants (35%) and 39 children (65%). The weight ranged from 5 kg to 18 kg, with a mean of 9.6 ± 2.53 (8.82 ± 2.79 for the shunt group and 10.41 ± 2.63 for the repair group), with a highly significant difference between the two groups (P = .008). Additionally, the O₂ saturation was 68.95 ± 7.8% for the shunt group and 87.93 ± 6.18% for the repair group; again, there was a highly significant difference between the two groups (P < .0001). The median McGoon ratio was 1.4 for the shunt group and 1.6 for the repair group (Table 1).

Most shunt group patients received a 5-mm shunt (73%), and most patients (86.6%) had the shunt placed on the right side (Table 2).

The mean bypass time in the repair patients was 99.16 ± 49.22 minutes, and the cross-clamp time was 56.63 ± 24.12 minutes (Table 2).

The postoperative oxygen saturation improved significantly in both groups (median, 83% in the shunt group; 99% in the repair group). The ICU stay ranged from 2 to 31 days (median, 3 days for the shunt group and 8.61 ± 1.91; and 4 days for the repair group (mean 6.07 ± 6.63 days). The overall hospital stay ranged from 5 to 50 days (median, 8 days for the repair group (mean 8.47 ± 3.54); and 4 days for the repair group (mean 11.37 ± 9.62). The comparative study between the two groups revealed a highly significant increase in the ICU stay in the repair group compared to the shunt group (P = .0058) but revealed a nonsignificant difference with regard to overall hospital stay (P > .05) (Table 3).

There was no significant difference in postoperative morbidity between the shunt and repair groups (Table 3).

In the shunt group, 8 patients (26.6%) suffered from postoperative complications. Shunt obstruction occurred in 1 patient after discharge, who needed another operation. Renal impairment requiring peritoneal dialysis occurred in 1 patient and eventually resolved; this resulted in a more extended ICU stay (9 days) for this patient. Chest infection occurred in 2 patients, wound infection occurred in 2 patients, and bleeding occurred in 2 patients (Table 4).

In the repair group, 8 patients (26.6%) suffered from postoperative complications. Two of these patients suffered from brain insult and chest infection, both of whom required more extended ICU stays (31 days and 27 days). Another patient suffered from chest infection. Localized pericardial effusion occurred in 1 patient, but reintervention was not needed. Two

### RESULTS

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patients had a wound infection, one patient suffered from postoperative bleeding and required intervention, and one patient suffered from supraventricular tachycardia (Table 4).

The incidence of mortality in our study was 10% (10% for the shunt group and 10% for the repair group) (Table 4). We used oxygen saturation and the NYHA score to assess patients after surgery. A comparative study between pre- and postoperative measurements for the shunt and repair groups revealed that the average SO2 significantly increased during serial O2 saturation measurements, with a highly significant difference \((P < .0001)\) (Figure).

Approximately 73.3% of the shunt group and 83.3% of the repair group had an NYHA score of Grade 1 (Table 5).

**DISCUSSION**

Even with the recent significant advances in congenital heart surgery, the management of tetralogy of Fallot remains challenging, especially regarding the choice between performing single-stage complete repair or performing systemic to pulmonary shunt surgery and delaying complete repair. Identifying when each surgical strategy should be applied and which patients would benefit most from both strategies are still not fully elucidated. The choice of repair or shunt will affect the practical decision, notably whether to perform a transannular patch, and will also be affected by resources, equipment, and experience of the surgeons at the institution. Our study showed that the morbidities, whether for the shunt group or the repair group, was comparable to those from developing countries but higher than those from developed countries. Additionally, according to mortality rates, our results are higher than those reported worldwide, especially those from developed countries [Gladman 1997; Jahangiri 1999; Maghur 2002; Singh 2014; Hirsch 2000; Dyamenahalli 2000; Anagnostopoulos 2007; Allam 2014; Sasikumar 2014; Saygi 2015].

Although we found no difference between the shunt and repair groups regarding morbidity and mortality, there was still a significantly shorter ICU stay for the shunt group. It should be noted when analyzing these results that the repair group in this study had a higher weight, higher preoperative SO2, and higher McGoon ratio. Additionally, there was a significant improvement in morbidity and mortality after shunt operation, which was comparable to that of single-stage repair.

We were limited to tetralogy of Fallot patients with borderline pulmonary arteries and those who had a McGoon ratio between 1.3 and 1.7. Even with the development of other methods to estimate pulmonary arterial size, the McGoon ratio is still used in Egypt, as it is available and practical and can be calculated by the surgeon. Additionally, it is still used in many reports, especially those from developing countries [Chen 2007].

We found that the median McGoon ratio was 1.4 for the shunt group and 1.6 for the repair group, which was significantly different \((P < .0001)\). This suggests a ratio of 1.5 as a differentiating point for determining the optimal surgical strategy.

Recent studies revealed that single-stage complete repair could be performed with a low risk of mortality and with survival rates equal to those of two-stage repair, even in younger patients [Stewart 2005; Kanter 2010; Karl 1992].

One of the critical challenges is the size of the pulmonary arteries. The application of MBT shunts has been found to promote the growth of pulmonary arteries, decrease the use of transannular patches, and facilitate subsequent repair [Kanter 2010; Jahangiri 1999; Ishikawa 2001; Sabri 1999; Ross 2015]. Complications of the MBT shunt still exist, thus adding another prognostic factor that should be kept in mind, especially as it relates to pulmonary artery distortion and stenosis [Gladman 1997].

Even with the growing interest in primary repair, the data regarding aggressive patch augmentation of the hypoplastic pulmonary arteries found that this strategy may lead to late-onset stenosis and higher rates of reintervention [Wilder 2016]. Additionally, the occurrence of pulmonary incompetence with the use of transannular patches has an unfavorable

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**Table 5. Comparison between Shunt and Repair Groups According to Postoperative NYHA Score**

<table>
<thead>
<tr>
<th>NYHA score, n (%)</th>
<th>Shunt Group ((n = 30))</th>
<th>Repair Group ((n = 30))</th>
<th>(P)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 1</td>
<td>22 (73.3)</td>
<td>25 (83.3)</td>
<td>.6024</td>
</tr>
<tr>
<td>Grade 2</td>
<td>4 (13.3)</td>
<td>2 (6.7)</td>
<td></td>
</tr>
<tr>
<td>Grade 3</td>
<td>1 (3.3)</td>
<td>0 (0)*</td>
<td></td>
</tr>
<tr>
<td>Grade 4</td>
<td>0 (0)*</td>
<td>0 (0)*</td>
<td></td>
</tr>
</tbody>
</table>

*Patients with this score died during the study.*
effect on the long-term recovery after repair [Singh 2014; Horneffer 1990; Frigiola 2004].

Shunted patients required fewer transannular patch repairs despite undergoing more new initial operations [Laas 1985].

If the two-stage strategy is to be used, patients should be thoroughly followed up after shunt operation to avoid and detect any complications until complete repair can be implemented.

**Study Limitations**

The study still requires further development in determining whether adding the postoperative repair results of a two-stage strategy will make a difference, if there is an actual decrease in the use of transannular and pulmonary patches with this strategy, and if these benefits outweigh the risk of undergoing two operations. Additional limitations of the study are as follows: the limited number of patients, the limited follow-up time, and the intraoperative decision-making at the discretion of the operating surgeon rather than strict adherence to the protocol.

**Conclusion**

There was a significant improvement after the MBT shunt with no significant difference between the shunt and single-stage complete repair regarding morbidity and mortality. However, regarding the ICU stay, the multistage repair was better with fewer postoperative severe complications. There is no significant difference between multistage repair (after the first stage) and single-stage complete repair. Proper decision-making can be performed using the McGoon ratio; however, intraoperative evaluation of the pulmonary arteries should still be performed. A McGoon ratio of 1.5 could be used as a cutoff in management decision-making.

The MBT shunt is a palliative first-stage surgery and is still a good option in managing tetralogy of Fallot (even with the increased interest of single-stage complete repair), especially in complex cases and in poorly equipped institutions, with good postoperative results. Patient follow-up after MBT shunt is significant.

Every effort should be made to improve the environment of heart surgery in Egypt, and more studies are needed to identify specific guidelines about tetralogy of Fallot management, primarily in developing countries.

**REFERENCES**


