One-Year Follow-up After Tetralogy of Fallot Total Repair Preserving Pulmonary Valve and Avoiding Right Ventriculotomy

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Background: We reviewed our revised surgical strategy for tetralogy of Fallot (TOF) total correction to minimize early exposure to significant pulmonary regurgitation (PR) and to avoid right ventriculotomy (RV-tomy).

Methods and Results: Since February 2016, we have tried to preserve, first, pulmonary valve (PV) function to minimize PR by extensive commissurotomy with annulus saving; and second, RV infundibular function by avoiding RV-tomy. With this strategy, we performed total correction for 50 consecutive patients with TOF until May 2018. We reviewed the early outcomes of 27 of 50 patients who received follow-up for ≥3 months. Mean patient age at operation was 10.2±5.0 months, and mean body weight was 8.8±1.2 kg. The preoperative pressure gradient at the RV outflow tract and the PV z-score were improved at most recent echocardiography from 82.0±7.1 to 26.8±6.4 mmHg, and from −2.35±0.49 to −0.55±0.54, respectively, during 11.1±1.6 months of follow-up after operation. One patient required re-intervention for residual pulmonary stenosis. Twenty-two patients had less than moderate PR (none, 1; trivial, 8; mild, 13), and 5 patients had moderate PR. There was no free or severe PR.

Conclusions: At 1-year follow-up, the patients who underwent total TOF correction with our revised surgical strategy had acceptable results in terms of PV function. The preserved PV had a tendency to grow on short-term follow-up.

Key Words: Congenital heart disease; Pediatrics; Tetralogy of Fallot; Valvular disease

Transannular right ventricular outflow tract (RVOT) reconstruction at the time of total repair of tetralogy of Fallot (TOF) induces significant pulmonary regurgitation (PR) and progressive right ventricular (RV) dilatation, which induces RV failure and fatal ventricular arrhythmia over time. Many surgeons have tried to reduce PR by minimizing the incision size on RVOT, preserving the pulmonary valvular function, or inserting cusp-like patch materials during TOF total repair. In addition to the pulmonary valve annulus and valvular function, we also focus on the function of the infundibulum for long-term RV function. We frequently found significant aneurysmal change in the infundibular area in most patients long after transannular RVOT-widening TOF total correction. This area showed a paradoxical contraction during RV systolic phase that interrupted the effective RV ejection and RV cardiac output.

Our hypothesis is that our revised surgical strategy to preserve the pulmonary valve and RV infundibular area might be helpful for preserving RV function in the long term. In the present study, we first examined the supposition that our revised strategy may produce tolerable pulmonary valve function in the short term.

Methods

Surgical Strategy and Approach

The strategy was to preserve the pulmonary valve function and annulus to minimize PR and to minimize right ventriculotomy (RV-tomy) even if RV-tomy seemed to be required according to the previous surgical strategies at the time of TOF total correction. Before 2016, when the size of the pulmonary valve was too small (z-score <−3) or the ratio of RV inlet/ascending aorta pressure (p-RVi/AAo) was >0.7 after the total correction of TOF in the operating room, we did not hesitate to perform transannular RVOT widening with a limited RV-tomy method to relieve the pressure gradient through the pulmonary valve. We found, however, that the RV pressure gradually dropped down to an acceptable level in many patients, even if p-RVi/AAo was >0.7 in the immediate postoperative period in the operating room, and also found that an aneurysmal change...
occurred over time in the infundibular area where we carried out RV-tomy and widened with a patch. These aneurysmal changes showed a paradoxical contraction. Therefore, we changed our strategy and applied it to all patients who underwent total repair for TOF from February 2016, as follows. First, regardless of the z-score of the preoperative pulmonary valve annulus, we avoided trans-annular RVOT widening. To achieve this, we made a more extensive commissurotomy first,\textsuperscript{12} and, if needed, we extended the additional small meticulous incisions parallel to the pulmonary arterial wall to relieve the tethered leaflet commissure area. This procedure could make the pulmonary valve orifice wider and pulmonary valve leaflet more mobile, even if the pulmonary valve was bi-leaflet. Second, we avoided making a separate RV-tomy, in order to make it easier to resect the infundibular muscle and to carry out patch widening at the infundibular area, which many surgeons have used to preserve the pulmonary annulus.\textsuperscript{6,9,10,12} as well as avoiding an RV-tomy that extends from the vertical main pulmonary artery incision for transannular RVOT widening. We usually use bicaval venous drainage; and ventricular septal defect (VSD) closure was performed first through right atriotomy using a glutaraldehyde-treated autologous pericardial patch with multiple interrupted sutures. After VSD closure, we resected the parietal band and some parts of the conal septum just outside of the VSD patch margin (Figure 1A,B). We then performed RVOT widening, including resection of the hypertrophied muscle in the RV cavity. The vertical incision was made from just distal of the pulmonary valve annulus to the pulmonary arteries bifurcation area. Before or after pulmonary valve commissurotomy was performed according to the situation (Figure 1C,D), the hypertrophied RV muscle, including the septal band and conal septum, was resected through this pulmonary arteriotomy site (Figure 1E,F). Then, the glutaraldehyde-treated autologous pericardial patch, which was prepared in a normal size (z-score around zero) using a Hega dilator, was applied to widen the main pulmonary artery. The body temperature was maintained at approximately 28–30°C. We did not let the atrial septum open in all patients during the study period.

Patients
From February 2016 to May 2018, 50 consecutive patients with TOF underwent total repair with our revised surgical strategy. Of these, we retrospectively reviewed 27 patients who had postoperative echocardiographic data for ≥3 months after the total correction of TOF. In the operating room, we measured the pressure of the AAo using the aortic root cannula, which was used for cardioplegia infusion after aorta cross-clamping. The pressure of the RV inlet was measured using a direct 23-G needle puncture. The patients’ body surface area (BSA) was calculated using the Mosteller method (BSA(m²)=(height(cm)×weight(kg)/3,600)\textsuperscript{0.5}). The z-score of the patient’s cardiac structures was calculated based on the echocardiographic measurements, according to Pettersen et al.\textsuperscript{14} We observed the changes in the echocardiographic parameters, such as pressure gradient, via the pulmonary valve, the size of the pulmonary valve annulus (z-score), the degree of PR, and the tricuspid annular plane systolic excursion (TAPSE) and the degree of tricuspid regurgitation to determine RV function after the total repair. In the present study, PR was classified into 4 different grades according to the regurgitant amount on echocardiography as reviewed by pediatric cardiologists: trivial; mild; moderate; and severe or free. Compared to other cardiac valves, the grading of PR tends to be subjective, and is dependent on the cardiologist. The grading system was as follows: when the width of regurgitant flow occupied >50% of the pulmonary annulus diameter, or regurgitant flow was shown from the branch pulmonary arteries, it was graded as “severe” or “free” PR. If the regurgitant flow was observed with an amount of physiological regurgitation,
Changes in pulmonary regurgitation (PR) during follow-up. Postop., postoperative.

Table. Change in Patient Characteristics

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Before operation</th>
<th>Discharge</th>
<th>Last follow-up (most recent echocardiography)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>10.2±5.0 (2.5–44.9)</td>
<td>20.6±4.7 (7.3–53.2)</td>
<td>12.1±1.3 (6.6–21.6)</td>
<td>0.520±0.04 (0.37–0.63)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Body weight (kg)</td>
<td>8.8±1.2 (5.5–17.1)</td>
<td>26.8±6.4</td>
<td>0.001</td>
<td></td>
</tr>
<tr>
<td>BSA (m²)</td>
<td>0.4±0.04 (0.31–0.70)</td>
<td>32.8±5.5</td>
<td>0.55±0.54 (−3.26–1.95)</td>
<td></td>
</tr>
<tr>
<td>PG at RVOT (mmHg)</td>
<td>82.0±7.1</td>
<td>26.8±6.4</td>
<td>0.001</td>
<td></td>
</tr>
<tr>
<td>PV annulus (z-score)</td>
<td>−2.35±0.49 (−4.21–0.11)</td>
<td>0.001</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Data given as mean±SD (range). BSA, body surface area; PG, pressure gradient; PV, pulmonary valve; RVOT, right ventricular outflow tract.

Results

Pre- and Intraoperative Characteristics

Patient mean age was 10.2±5.0 months (range, 2.5–44.9 months), the mean body weight was 8.8±1.2 kg (range, 5.5–17.1 kg), and the mean BSA was 0.414±0.042 m² (range, 0.309–0.697 m²) at the time of total correction of TOF. The mean preoperative pressure gradient at RVOT was 82.0±7.1 mmHg, and the z-score of the pulmonary valve annulus was −2.35±0.49 (range, −4.21 to 0.11) on echocardiography. Small pulmonary valve annulus, that is, z-score <−3, was observed in 11 patients (40.7%).

Three patients had a 3-leaflet pulmonary valve, 21 patients had a bicuspid pulmonary valve and 2 patients had a monocuspid valve. All monocuspid and bicuspid valves were thickened and dysmorphic to a varying degree, and showed commissural fusion. Mean p-RVi/AAo was 0.58±0.07. p-RVi/AAo >0.65 was observed in 10 patients (37.0%), >0.7 was observed in 6 patients (22.2%), and >0.8 in 3 patients (11.1%). Of 9 patients with p-RVi/AAo >0.7 in the operating room, 4 patients had a small pulmonary valve annulus with z-score <−3 on preoperative echocardiography. According to our surgical strategy, we did not perform transannular RVOT widening to relieve the residual pulmonary stenosis (PS) for these high p-RVi/AAo patients. Meanwhile, 7 patients who had preoperative pulmonary valve annulus z-score <−3 had p-RVi/AAo <70% in the operating room after weaning off cardiopulmonary bypass (CPB).

Postoperative Changes During Follow-up

The mean follow-up duration (from the time of total correction to the time of most recent outpatient clinic visit) was 12.3±2.0 months. The mean duration from the time of operation to the most recent echocardiography was 11.1±1.6 months. The mean body weight was 12.1±1.3 kg (range, 6.6–21.6 kg), and the mean age at the most recent echocardiography was 20.6±4.7 months (range, 7.3–53.2 months). The mean BSA was 0.52±0.04 m² (range, 0.37–0.80 m²). The mean pressure gradient at the last echocardiography was 26.8±6.4 mmHg, and the z-score of the pulmonary valve annulus size was −0.55±0.54 (range, −3.26 to 1.95), and these were significantly improved not only from the preoperative status, but also from the immediate postoperative status (Table). This showed that with increased forward flow through the RVOT, the z-score of the pulmonary annulus size increased from −2.15±0.56 in the immediate postoperative status to −0.55±0.54 at the most recent follow-up (P=0.001).

At the last echocardiography, most patients had an acceptable degree of PR: no PR in 1 patient; trivial PR in 8 patients; mild PR in 13 patients; moderate PR in 5 patients; and no free PR in any patients (Figure 2). Additional repeated interventions were required to relie residual PS in 1 patient, whose preoperative pulmonary valve annulus z-score was −3.75. p-RVi/AAo was 0.7, and blood flow velocity through the pulmonary valve was 3.49 m/s, immediately after the operation in the operating room. The blood flow velocity through the RVOT was 4.2 m/s on echocardiography at discharge. Seven months after the total correction, although the functional status was acceptable, this patient’s PS was not relieved (blood flow velocity through the RVOT, 4.4 m/s on echocardiography; 52 mmHg on catheterization study). Thus, balloon pulmonary valvuloplasty was done by cardiologists; the pressure gradient was decreased to 39 mmHg on catheterization study, and
the RV systolic pressure was also mildly improved from 73 to 61 mmHg. The pressure gradient, however, was still significant, even 9 months after balloon angioplasty (flow velocity through the RVOT, 3.9 m/s on echocardiography; 30 mmHg on catheterization study), so this patient underwent repeated balloon angioplasty. The pressure gradient was successfully improved from 30 to 15 mmHg with a mild degree of PR. RV pressure also improved from 62 to 46 mmHg after balloon angioplasty on catheterization study.

Most patients had no cyanosis and a good functional class in the outpatient clinic. At the most recent echocardiography, the left ventricular (LV) ejection fraction was good, and the ventricular septal movement and configuration were acceptable. The degree of tricuspid regurgitation was none or trivial in all patients, and RV hypertrophy was gradually regressed. TAPSE on the most recent echocardiography, however, was mildly decreased (11.5±1.1 mm) compared with the normal range,14 and this was improved from the findings at discharge (6.3±1.2 mm), after operation. The z-score of the pulmonary valve annulus increased (P=0.001, Table).

**Discussion**

**Problems After TOF Total Correction and Surgical Strategy**

For the past several decades, the aims of surgical treatment for the patient with TOF have been to relieve the significant stenosis of RVOT and to eradicate the intra-cardiac shunt. To obtain adequate relief from RVOT stenosis, surgeons have not only performed pulmonary valvotomy, pulmonary artery angioplasty and removed hypertrophied infundibular muscles to the greatest extent possible, but they have also sacrificed the pulmonary valve annulus (transannular RVOT widening) in some cases. As the number of survivors after TOF total correction has increased, however, we find that significant PR after transannular RVOT widening in this population induces severe RV dilatation and RV failure as well as fatal ventricular arrhythmia over time.12,4,5,11

Because of the aforementioned problems, many surgeons have tried to preserve the pulmonary valvular function and minimize the RV incision by various surgical modifications.6,10,12,13,16,17 Some surgeons make a separate external incision in the infundibular area (RV-tomy) to remove more hypertrophied infundibular muscles through the RV-tomy site while preserving the pulmonary valve annulus.9,10 In the case of small pulmonary valve annulus and z-score <−3, however, preservation of the pulmonary valve annulus still seems to be difficult with sufficient relief of PS. Therefore, transannular RVOT widening has been seriously considered in these cases.

In addition to the problem of PR, we focused on the function of the RV infundibular area for the entire RV function. The RV-tomy area, which was widened by some of the patch materials used for RVOT widening during the total correction, shows significant aneurysmal changes over time. This can occur in both situations, that is, in the case of transannular RVOT widening or in the case of a separate patch widening of the RV-tomy area with pulmonary valve annulus preservation. From our own long-term observation on cardiac magnetic resonance imaging, we have the impression that this aneurysmal area seems to be less helpful for RV function because of paradoxical or akinetic motion during the cardiac phase.18,19 Because of the aforementioned 2 issues, that is, preventing the PR, and RV-tomy at infundibular area, we revised our strategy for the total correction of TOF. We need long-term follow-up data to evaluate the effects of preserving pulmonary valve function and RVOT infundibulum.

**Improvement of RV Pressure, Pulmonary Valve Annulus, and Residual PS**

Transannular RVOT widening has been seriously considered when the RV/LV pressure ratio is >70% or 80% in the operating room, immediately after total correction of TOF.7,9,13 Since February 2016, however, we have decided not to perform an additional procedure for residual PS relief unless the RV inlet pressure is higher than the systemic pressure (pressure of the AAo after CPB weaning in the operating room. We occasionally performed the transannular RVOT widening in the case of high p-RVi/AAo in the past (before 2016), but we found that the RV pressure decreased significantly during the recovery period in the intensive care unit in many patients. During weaning off CPB, we usually use inotropics or vasoconstrictors, such as epinephrine, or dopamine, which enhance the contraction of RV hypertrophied muscles. The remaining inflammatory reactions associated with the CPB itself also cause high resistance of the pulmonary vasculature, which reflects the high RV pressure. We hypothesize that these effects of inotropics or vasoconstrictors and the inflammatory reaction in the immediate postoperative period would regress over time and that the RV pressure would subsequently do the same. Therefore, if we can successfully take the patient off CPB at once and there are no significant structures to be resected or widened, except sacrificing the pulmonary valve annulus or RV infundibulotomy with patch widening, we wait for the RV pressure to regress, even when p-RVi/AAo is >70–80% in the operating room immediately after CPB weaning.

We expected that the patients who had a smaller preoperative pulmonary valve annulus z-score would have a tendency toward higher p-RVi/AAo after operation. We found, however, that even though the preoperative mean pulmonary valve annulus z-score was smaller than that in previous studies,6,9 only 1 patient required an additional intervention to relieve residual PS during the 13.7±6.4 months of follow-up, which involved our surgical strategy of aggressive commissurotomy and waiting for the spontaneous regression of RV pressure in the immediate postoperative period without additional surgical interventions. We are not aware of similar good results on long-term follow-up; if, however, we consider the tendency of the pulmonary valve annulus z-score to grow or increase (Table) without significant PS in most patients during the 1 year of follow up, we may still expect that the PS will not be significantly aggravated in the long term.

**Newly Developing PR**

Similar to other previous studies, we performed aggressive pulmonary valvotomy.6,7,9 In addition to widening the opening of pulmonary valve, we focused on the valve motion after the procedure. When the leaflets still stuck to the annulus and arterial wall, even after valvotomy, meaning that the leaflets would therefore show stiff movement, we could not expect an effective pulmonary valve orifice area for the forward flow from the RV. Therefore, if we considered a valvotomy to be insufficient to provide an adequate opening, we added a meticulous detaching procedure, that is, we detached approximately 1 mm of tethered dysmorphic...
pulmonary leaflet from the annulus and arterial wall. This detachment did not induce significant PR in the immediate postoperative period, and it was not significantly aggravated (no free PR) during follow-up in most patients. The proportion of patients with moderate PR on postoperative echocardiography (11.1±1.6 months after operation) was much smaller (18.5%; 5 patients with moderate PR) than in previous studies (approximately 36%). We also assume that the bicuspid or monocuspid valve would have worse valvular function. Despite, however, a larger proportion of bicuspid or monocuspid pulmonary valves (14.8% of 3-leaflet pulmonary valves; 4/27; 1 of these 4 patients had small pulmonary annulus, z-score=−3.25) compared with previous studies (approximately 36%), the proportion of patients with significant PR was smaller.

**Study Limitations**

This was a single-center retrospective study, involving only short-term follow-up results in a small number of patients. Long-term observation and evaluation are mandatory to prove the effectiveness of our revised surgical strategy. We were not able to evaluate the effectiveness of preserved RV infundibular area in this study, as we mentioned earlier, because it relates to future RV function. Although the changes in some parameters seemed significant (increased pulmonary annulus PS and low prevalence of significant PR compared with previous studies), the total number of enrolled patients was small. We plan, however, to keep using this strategy for future patients with TOF because the short-term follow-up results seem promising.

**Conclusions**

We were able to preserve the pulmonary valve function in most patients, with an acceptable residual PS and tolerable PR on 1-year follow-up in most patients when using our revised surgical approach for the total correction of TOF. It is mandatory to observe not only whether the preserved pulmonary valvular function is maintained during long-term follow-up, but also whether the preserved RV infundibular area may play a role in long-term RV function.

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**Disclosures**

The authors declare no conflicts of interest.

**References**


