Postoperative Outcomes of Children With Tetralogy of Fallot, Pulmonary Atresia, and Major Aortopulmonary Collaterals Undergoing Reconstruction of Occluded Pulmonary Artery Branches

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Background. Patients with tetralogy of Fallot, pulmonary atresia, and major aortopulmonary collaterals (TOF/PA/MAPCAs) undergoing unifocalization surgery are at risk for prolonged postoperative respiratory failure. We sought to understand whether patients undergoing reconstruction and incorporation of occluded pulmonary arterial branches were at risk for worse postoperative outcomes.

Methods. We performed a retrospective chart review to identify patients who underwent unifocalization or unifocalization revision with incorporation of occluded pulmonary artery branches. Patients with and without occluded branches were compared, with a focus on clinical outcomes.

Results. We studied 92 patients who underwent unifocalization procedures between 2010 and 2014, 17 (18%) of whom underwent reconstruction of occluded pulmonary artery branches. Patients with and without occluded branches were compared, with a focus on clinical outcomes.

Conclusions. Occluded pulmonary arterial branches can be safely recruited into the pulmonary vasculature in patients with TOF/PA/MAPCAs without a significant difference in postoperative outcomes compared with patients who did not have an occluded branch. Incorporation of occluded branches may also facilitate ultimate complete intracardiac repair in this complex population of patients.

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reconstruction of occluded PA branches would therefore be more likely to have prolonged respiratory failure, longer hospital stays, and higher rates of surgical reexploration than patients whose unifocalization procedure did not include such vessels.

**Patients and Methods**

All patients with TOF/PA/MAPCAs undergoing unifocalization or PA reconstruction surgery, or both, from January 2010 through June 2014 were included for analysis. We excluded patients with non-TOF anatomy or preoperative respiratory failure. We excluded patients with preoperative respiratory failure because the need for mechanical ventilation preoperatively may lengthen the duration of postoperative mechanical ventilation owing to the prolonged need for sedatives, central lines, and possible deconditioning. We included only the first operation at our institution for patients who underwent more than one surgery during the study period to avoid multiple event bias.

All patients undergoing unifocalization surgery underwent preoperative catheterization at which time PA and MAPCA anatomy were detailed and occluded PA branches were identified if present. The distribution of each occluded PA branch was carefully detailed and only considered for reconstruction if it supplied at least one major lung segment. We identified patients who had occluded PA branch interventions by reviewing the operative note. We compared postoperative outcomes of patients with reconstructed occluded vessels to patients with only nonoccluded vessels undergoing the same types of surgery. To reduce confounding related to the type of surgery, we excluded patients without previous interventions who were able to undergo complete unifocalization and intracardiac repair as their first and only operation.

![Fig 1. Reperfusion pulmonary edema chest radiograph (CXR) scoring form.](image)

<table>
<thead>
<tr>
<th>Patient number:</th>
<th>CXR Date:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years):</td>
<td></td>
</tr>
<tr>
<td>Opacity Score</td>
<td>0 (none)</td>
</tr>
<tr>
<td></td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>3 (dense)</td>
</tr>
<tr>
<td>L upper lobe</td>
<td></td>
</tr>
<tr>
<td>L lingula</td>
<td></td>
</tr>
<tr>
<td>L lower lobe</td>
<td></td>
</tr>
<tr>
<td>R upper lobe</td>
<td></td>
</tr>
<tr>
<td>R middle lobe</td>
<td></td>
</tr>
<tr>
<td>R lower lobe</td>
<td></td>
</tr>
</tbody>
</table>

Pleural effusion(s): Y _ N __

Location(s): _________________________

Size: Small _____ Moderate _______ Large _______

Scoring Legend:

- 0: no opacity
- 1: opacity not obscuring vasculature
- 2: same as vasculature
- 3: obscuring vasculature
operation. We included all patients who were able to undergo complete intracardiac repair with revision unifocalization. We reviewed postoperative angiograms, routinely performed 6 months after surgery, for the patients who had undergone reconstruction of occluded branches to confirm patency of these vessels.

A pediatric radiologist blinded to the clinical course evaluated five chest radiographs for each patient in the occluded PA branch group: the preoperative and postoperative days 0, 1, 2, 3, and 4. The evaluations were performed in accordance with previous studies from our institution using a standardized grading system (Fig 1). Each lung segment was evaluated for the development of a radiographic opacity and graded from 0 to 3. We considered a positive finding to be the development of a pulmonary opacity not present on the preoperative chest radiograph of grade 2 or greater in the lung segment supplied by the reconstructed occluded PA branch [4]. Chest radiographs were excluded at the radiologist’s discretion when the lung segments could not be evaluated in the presence of pleural effusion or pneumothorax.

Categoric variables were compared between groups using \( \chi^2 \) analysis or Fisher’s exact test, and the Wilcoxon rank sum test was utilized for continuous data. The Institutional Review Board at Stanford University approved this study. All statistical testing was conducted using SAS Enterprise Guide 4.3 software (SAS Institute, Cary, NC). A \( p \) value less than 0.05 was considered statistically significant.

Results

In all, 183 unifocalization procedures were performed between January 2010 and June 2014. We included 92 patients (41 male, 51 female) who met study criteria for analysis, 17 (18%) of whom underwent reconstruction of an occluded PA branch. In most cases, the occluded branch supplied only one lung segment, although in 1 case the entire left pulmonary artery was occluded by a previously placed right ventricle to pulmonary artery conduit (Fig 2). Baseline patient characteristics were similar between the two groups, although patients with occluded vessels were larger and older at the time of surgery (Table 1).

Patients with occluded PA branches underwent three types of surgery (Table 2). All except 1 of the patients with an occluded PA branch had undergone prior surgical or catheter-based intervention at other institutions. Compared with patients not having occluded branches, patients with occluded vessels were less likely to undergo complete intracardiac repair at the time of unifocalization revision and rather require staged unifocalization procedures before intracardiac repair.

During the operation in which patients underwent reconstruction of the occluded pulmonary artery branch, 2 patients had previous intracardiac repairs, and 5 patients underwent intracardiac repair at the time of reconstruction, indicating the pulmonary vascular resistance was low enough to safely close the ventriculoventricular defect (Fig 3). The right ventricular to left ventricular (RV/LV) pressure ratio in this group was 0.42 ± 0.06. The remaining 10 patients had staged unilateral reconstruction procedures and did not undergo intraoperative flow studies to assess suitability for ventriculocardiac defect closure at the time of the occluded branch intervention [5]. Of this group, 7 ultimately underwent successful intracardiac repair with mean RV/LV of 0.37 ± 0.05, indicating improved pulmonary vascular resistance over time. The 3 patients who did not undergo intracardiac repair. Patients with occluded PA branches underwent three types of surgery (Table 2). All except 1 of the patients with an occluded PA branch had undergone prior surgical or catheter-based intervention at other institutions. Compared with patients not having occluded branches, patients with occluded vessels were less likely to undergo complete intracardiac repair at the time of unifocalization revision and rather require staged unifocalization procedures before intracardiac repair.

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repair had multiple other abnormalities. One patient sustained severe neurologic injury after several cardiac arrests. Another patient is a young adult with restrictive lung disease and severe pulmonary hypertension ineligible for further surgery, and the last patient died of severe diastolic heart failure.

There was no significant difference in the clinical outcomes between patients with and patients without occluded vessels. In particular, duration of mechanical ventilation, need for surgical reexploration or extracorporeal membrane oxygenator support (ECMO) in the postoperative period, and duration of hospital stay were similar (Table 3).

Chest radiographs for all 17 patients with occluded vessels were reviewed. One patient was excluded because of a large pleural effusion. Ten of the remaining 16 patients (63%) had positive chest radiograph findings, that is, they had a radiographic opacity of grade 2 or higher within the first 96 hours postoperatively in the lung segment supplied by the previously occluded vessel. An example is shown in Figure 4.

Postoperative angiograms, routinely performed at 6 to 12 months after surgery, were available for review on 14 of 17 patients who underwent reconstruction of occluded PA branches. All accept 1 patient demonstrated patency of the reconstructed PA branches. Figure 5 illustrates examples.

Table 2. Type of Surgery

<table>
<thead>
<tr>
<th>Surgery</th>
<th>Without Occluded Vessel (n = 75)</th>
<th>With Occluded Vessel (n = 17)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral unifocalization revision with complete repair, type 1</td>
<td>65 (86%)</td>
<td>5 (29%)</td>
<td>0.001</td>
</tr>
<tr>
<td>Unilateral revision unifocalization, type 2</td>
<td>5 (7%)</td>
<td>10 (59%)</td>
<td></td>
</tr>
<tr>
<td>Bilateral revision unifocalization (history of complete repair), type 3</td>
<td>5 (7%)</td>
<td>2 (12%)</td>
<td></td>
</tr>
</tbody>
</table>

Comment

This study shows that occluded PA branches can be safely recruited into the pulmonary vasculature in patients with TOF/PA/MAPCAs, with no difference in early postoperative clinical outcomes compared with patients who underwent unifocalization without incorporation of an occluded vessel. Given the heterogeneous pulmonary blood flow in patients with MAPCAs, it is important to recruit as many lung segments as possible into the pulmonary circulation to optimize oxygenation in patients without complete repair, and to achieve the largest PA cross-sectional area possible [6]. Studies have shown PA cross-sectional area to be inversely related to the RV/LV pressure ratio, and lower RV/LV ratios are associated with improved survival after complete intracardiac repair [7, 8]. Although the majority of patients with occluded PA branches in this series underwent staged PA reconstruction procedures, 80% were ultimately able to undergo complete intracardiac repair. In MAPCAs patients, our strategy is to use an intraoperative flow study to determine candidacy for complete intracardiac repair, with a threshold mean PA pressure of less than 25 mm Hg used to qualify [5]. That most of our patients with occluded PA branches ultimately underwent complete repair indicates that having an occluded branch does not preclude achieving this benchmark.

Fig 3. Type of surgery for patients with occluded pulmonary arterial (PA) branches.
Patients with occluded branches were more likely to undergo revision unifocalization as their first surgery at our institution. Patients with occluded vessels were also older and weighed more at the time of surgery. These findings are likely related to patients with occluded vessels having undergone unsuccessful attempts at unifocalization surgery or catheter-based intervention resulting in mechanical obstruction. There was no significant difference between the two groups with regard to the presence of central PAs, indicating that the baseline anatomy may have been equally unfavorable between the two groups. We did not attempt to evaluate the underlying cause or process leading to PA occlusion but speculate that a combination of factors may have been involved, including remodeling of the MAPCAs over time in association with progressively decreasing flow as patients became older and larger, as well as potential distortion after prior interventions. Major aortopulmonary collateral vessels in patients with TOF/PA are vulnerable to changes in blood pressure and flow resulting in histologic changes and vessel stenosis [9]. Regardless of the mechanism of occlusion, we have shown that patients who undergo reconstruction of occluded pulmonary artery branches can have an uncomplicated postoperative course similar to that of patients without occluded vessels and may be able to undergo full intracardiac repair.

The primary aim of this study was not to diagnose or differentiate the risk of RPE, but rather to examine potential differences in clinical outcomes between patients who we hypothesized had a higher risk for RPE and other forms of lung injury, including contusion and bleeding. Interestingly, in more than half of the patients with occluded vessels, a radiographic opacity developed in the lung segment supplied by the reconstructed occluded PA branch, suggesting that the lung segment was affected in some way. This opacity may represent lung contusion, atelectasis, or RPE. Nonetheless, the presence of radiographic opacity did not represent lung pathology in this group that was unique and sufficiently severe to alter the postoperative course. This finding is consistent with the results of previous studies from our institution, in which RPE occurred in 50% to 65% of cases and was not associated with differences in the postoperative clinical course [3, 4]. Our data suggest that although RPE may be relatively common after unifocalization surgery, its impact is generally mild and self-limited.

Study Limitations

Given the retrospective design of this study, we were limited to the use of the operative report to determine which patients underwent reconstruction of occluded vessels. We did not attempt to compare the incidence of RPE between patients with and patients without an occluded PA branch because the development of radiographic opacities after unifocalization surgery may be multifactorial and nonspecific. Given the risk of prolonged and complicated postoperative courses of patients undergoing unifocalization surgery, we chose to focus on clinical outcomes in an effort to understand whether our hypothesis was correct that these patients are at particularly high risk for complications. Also, comparison between the occluded PA group and the control group was potentially confounded by the differences in history of prior procedures. Our study was not designed to delineate the differences in outcomes between patients with and patients without iatrogenic causes for their PA branch occlusions. The majority of referrals to our institution are patients who have undergone previous

**Table 3. Clinical Outcomes**

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Without Occluded Vessel (n = 75)</th>
<th>With Occluded Vessel (n = 17)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duration of mechanical ventilation, days</td>
<td>5 (1–44)</td>
<td>4 (1–48)</td>
<td>0.8</td>
</tr>
<tr>
<td>Need for reoperation</td>
<td>15 (20%)</td>
<td>3 (18%)</td>
<td>0.9</td>
</tr>
<tr>
<td>Duration of hospital stay, days</td>
<td>15 (5–109)</td>
<td>20 (4–61)</td>
<td>0.7</td>
</tr>
</tbody>
</table>

Fig 4. Preoperative angiogram and postoperative radiograph example. (A) Preoperative angiogram demonstrates occlusion of the left lower lobe pulmonary artery on pulmonary venous wedge injection (arrow). (B) Chest radiograph on the first postoperative night demonstrating significant diffuse haziness of the left lung with increased opacity of the left lower lobe (arrow).
interventions. Given the natural history of MAPCAs, it would also be problematic to distinguish iatrogenic causes from progression of disease.

In conclusion, patients with TOF/PA/MAPCAs with occluded PA branches have postoperative outcomes similar to those of patients without occluded vessels. Occluded PA branches can be safely recruited into the pulmonary vasculature, although staged unifocalization procedures may be required. Future studies will be directed toward the identification, prevention, and treatment of respiratory complications associated with prolonged respiratory failure in this population and to the development of specific radiographic techniques to understand regional lung perfusion.

References


