

Late Outcomes After Surgical Repair For Tetralogy of Fallot with Absent Pulmonary Valve

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Background:

Tetralogy of Fallot with absent pulmonary valve (TOF-APV) accounts for only 3-6% of patients with tetralogy of Fallot, but is uniquely associated with aneurysmal dilation of the pulmonary arteries and potential for airway compromise and respiratory compromise. Improvements in surgical and medical management have resulted in improved early mortality after surgical repair of TOF-APV. Survivors however remain at increased risk for reoperation and chronic respiratory problems. The prevalence for these late complications in this population remains unknown. We sought to determine the long-term outcomes of patients after TOF-APV at our institution.

Methods:

Between 1977 and 2015, 35 patients underwent surgical repair for TOF-APV at the Children's Hospital of Wisconsin. Surgical repair consisted of closure of ventricular septal defect and repair and reconstruction of the right ventricular outflow tract (RVOT) with or without surgery to address airway compression. Early mortality was defined as death within 30 days after complete repair or prior to hospital discharge.

Results:

For the cohort (n=35), the median age at time of surgical repair was 4.5 months (4 days-27.8 years). Eleven (31.4%) required mechanical ventilation at time of complete repair. RVOT reconstruction included placement of an aortic or pulmonary homograft in 24 (68.6%) patients, bioprosthetic valve conduit in 3 (8.6%), monocusp valve in 2 (5.7%), and transannular patch in 2 (5.7%). Concomitant procedure to address airway compression was performed in 25 (71.4%) patients, including pulmonary artery plication in 16, Lecompte maneuver in 5, and both plication and Lecompte in 4. Patients that underwent Lecompte were more likely to be younger ($p<0.0001$) and require mechanical ventilation ($p<0.0001$) at time of repair. There were 2 early deaths: 1 at post-op day 12 due to acute pulmonary artery hemorrhage, and 1 at 5 months after repair (prior to hospital discharge) for respiratory failure. This resulted in an overall early mortality rate of 5.7%. After repair, 3 patients required tracheostomy, and for the remaining 32 patients the median length of post-operative mechanical ventilation was 4 days (range = 0 to 168 days). Among patients that required mechanical ventilation at time of repair, there was no association between Lecompte and duration of post-operative ventilation or need for tracheostomy.

For the 33 hospital survivors, the median follow-up after complete repair was 9.1 years (range = 1 month to 33.1 years). There were no late deaths. A total of 25 (75.8%) patients required a reoperation for cardiac or pulmonary indication, resulting in an overall freedom from reoperation of 49% at 5 years and 13% at 10 years. Multiple reoperations were common, with 6 patients requiring at least 2 reoperations, and 7 patients requiring 3 reoperations. The reoperations included pulmonary valve replacement in 21 (64%), pulmonary artery plication in 4 (12.1%), Lecompte maneuver in 2 (6%), pulmonary artery and/or airway pexy in 3 (9.1%), and closure of residual ventricular septal defect in 3 (9%). Interventional cardiac catheterizations were performed in 7 patients, including 6 for balloon pulmonary arterioplasty and 1 for percutaneous pulmonary valve (Melody) replacement.

Clinical follow-up data was available for 28 patients, of which 21 (75%) were classified as NYHA class I and 4 required chronic pulmonary medications. Spirometry was abnormal in all patients with obstructive (50%), restrictive (25%), or mixed physiology (25%). On cardiopulmonary exercise testing with treadmill Bruce protocol, the mean peak VO_2 % predicted was good at $80 \pm 16\%$.

Conclusions:

Early and late mortality after complete repair of TOF-APV is low. These patients however, remain at increased risk for reoperation and lung dysfunction as measured by spirometry. Nevertheless, survivors have relatively good functional status at late follow-up.