Long-Term Outcomes of Patients With Absent Pulmonary Valve Syndrome: 38 Years of Experience

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**Background.** Absent pulmonary valve syndrome is associated with aneurysmal dilatation of the pulmonary arteries and compression of the tracheobronchial tree and may lead to significant respiratory compromise. We describe the outcomes of surgical correction of absent pulmonary valve syndrome and risk factors for mortality and reoperation.

**Methods.** A review of 52 patients with absent pulmonary valve syndrome who underwent surgical correction between 1975 and 2013 was conducted. The median age and weight at repair were 9 months (range, 4 days to 24.2 years) and 6.9 kg (range, 1.8 to 56 kg). Preoperative intubation was required in 15 patients (29%), and 21 patients (40%) underwent urgent repair. The pulmonary valve was replaced with a valved conduit in 16 patients (31%) or monocusp valve in 16 patients (31%). Valveless repair was performed in 20 patients (38%). Pulmonary artery reduction was performed in 39 patients (75%), and 2 patients (4%) underwent a Lecompte maneuver.

**Results.** The median follow-up time was 13 years (range, 1 month to 35 years). Early mortality was 18.8% (3 of 16) during 1975 through 1989, 19% (4 of 21) during 1990 through 2000, and 0% (0 of 15) during 2001 through 2013. Late mortality was 6.7% (3 of 45). Overall survival at 5, 10, and 20 years was 81.4% ± 5.6%. On multivariate analysis, preoperative ventilation (p = 0.009) was the only risk factor for overall mortality. Freedom from late reoperation at 5, 10, and 20 years was 79.7% ± 6.9%, 69.4% ± 8.2%, and 52.1% ± 9.8%, respectively. No difference in reoperation rates was found between valved conduit, monocusp, or valveless techniques. Risk factors for late reoperation on multivariate analysis were prematurity (p = 0.001) and neonatal primary repair (p = 0.007). Longer postoperative ventilation periods were predicted by preoperative ventilation (p < 0.001) and surgery during infancy (p = 0.01).

**Conclusions.** Long-term survival for absent pulmonary valve syndrome has improved during the last decade. Preoperative ventilation predicted longer postoperative ventilation and mortality.

Tetralogy of Fallot with absent pulmonary valve syndrome (APVS) is rare, making up 5% of the tetralogy of Fallot spectrum [1]. Patients with APVS have dysplastic or absent pulmonary valve, free pulmonary regurgitation, and annular stenosis [2, 3]. In addition, the characteristic feature of APVS is the aneurysmal pulmonary arteries (PAs) that often cause airway compression at the tracheal and bronchial level (Fig 1). Patients with APVS may present early in life with respiratory distress necessitating preoperative ventilatory support and urgent surgery [2, 3].

The surgical management of the enlarged PA and the optimal technique for right ventricle (RV)-to-PA connection remains debatable [4, 5]. We reviewed our experience with APVS repair during 38 years at the Royal Children’s Hospital.

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Patients and Methods

**Patients**

The Human Research Ethics Committee of the Royal Children’s Hospital approved this retrospective study. Between August 1975 and July 2013, 52 patients underwent complete repair of APVS at the Royal Children’s Hospital. The patients were subdivided into three groups, 1975 through 1989 (early), 1990 through 2000 (middle), and 2001 through 2013 (current), to determine the impact of the era on outcomes.

The median age at operation was 9 months (interquartile range [IQR], 4.6 to 18 months; total range, 4 days to 24.2 years). There were 3 neonates (5.8%), 24 infants (46.2%), 24 children (46.2%), and 1 adult (1.9%) at operation. There were 30 girls (57.7%). Nine patients (17.3%) were born prematurely (range, 29 to 36 weeks’ gestation; Table 1).

The diagnosis of APVS was made by catheterization in the early era or by echocardiography in later eras. Fifteen patients (28.9%) underwent further PA and airway
assessment with computed tomography angiography. Antenatal diagnosis of APVS has improved with time, with the first antenatal diagnosis made in the early era in 1986 (1 of 16; 6.3%) compared with 4 of 21 (19%) and 10 of 15 (66.7%) patients in the middle and current eras, respectively.

All patients with APVS in our study had associated tetralogy of Fallot. Other cardiovascular anomalies included double-outlet RV (n = 2; 3.9%), right aortic arch (n = 20; 38.5%), anomalous coronary artery (n = 1; 1.9%), absence of the right superior vena cava (n = 1; 1.9%), anomalous origin of the left PA (n = 5; 9.6%), and anomalous pulmonary venous drainage (n = 1; 1.9%).

Eleven patients (n = 11; 21.2%) had underlying syndromal diagnoses including DiGeorge syndrome (n = 6; 11.5%), congenital rubella (n = 1; 1.9%), CHARGE syndrome (n = 1; 1.9%), Duane syndrome (n = 1; 1.9%), or undiagnosed syndromes (n = 2; 3.9%).

Seven patients (13.5%) had prior surgery including three noncardiac (right upper lobe lobectomy, n = 1; bowel resection and colostomy, n = 2) and four cardiac operations (PA plication, n = 2; Blalock-Taussig shunt, n = 1; left PA repair, n = 1). Of the patients who had prior cardiac surgery, the first patient was a 3-month-old girl weighing 5.5 kg in severe respiratory distress, who underwent PA plication in 1983 and complete repair at 3.5 years of age. The second patient was a 7-day-old premature boy weighing 2.3 kg in severe respiratory distress, who underwent PA plication in 1984 and complete repair at 4 months of age. The third patient was a 1-day-old boy weighing 3.2 kg, who underwent Blalock-Taussig shunt in 1999 and complete repair at 1 year of age. The fourth patient was a 6-day-old boy weighing 3.1 kg with a left PA coming from a separate origin from the patent ductus arteriosus, who underwent connection of the left PA to the main PA in 2003 and complete repair at 1 year of age.

Patients with airway compromise underwent urgent repair (n = 31; 59.6%). Preoperative intubation was necessary in 15 patients (28.9%) for severe airway compromise. Only patients younger than 1 year required ventilator support (all 3 neonates, 12 of 24 infants).

<p>| Table 1. Patient Characteristics and Perioperative Variables According to Era |</p>
<table>
<thead>
<tr>
<th>-------------------</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>n = 16</td>
<td>n = 21</td>
<td>n = 15</td>
</tr>
<tr>
<td>Demographics</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Median age repair (mo)</td>
<td>7.1</td>
<td>13.6</td>
<td>6.9</td>
</tr>
<tr>
<td>Neonates &lt;1 mo</td>
<td>2 (12.5%)</td>
<td>1 (4.8%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Infants 1 mo–1 y</td>
<td>8 (50%)</td>
<td>6 (28.6%)</td>
<td>10 (66.7%)</td>
</tr>
<tr>
<td>Children 1–18 y</td>
<td>6 (37.5%)</td>
<td>13 (61.9%)</td>
<td>5 (33.3%)</td>
</tr>
<tr>
<td>Adults &gt;18 y</td>
<td>0 (0%)</td>
<td>1 (4.8%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Prematurity</td>
<td>5 (31.3%)</td>
<td>2 (9.5%)</td>
<td>2 (13.3%)</td>
</tr>
<tr>
<td>Antenatal diagnosis</td>
<td>1 (6.3%)</td>
<td>4 (19%)</td>
<td>10 (66.7%)</td>
</tr>
<tr>
<td>Perioperative</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Preoperative ventilation</td>
<td>6 (37.5%)</td>
<td>5 (23.8%)</td>
<td>4 (26.7%)</td>
</tr>
<tr>
<td>Elective surgery</td>
<td>7 (43.8%)</td>
<td>14 (66.7%)</td>
<td>10 (66.7%)</td>
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<tr>
<td>Mean weight at operation (kg)</td>
<td>10.4</td>
<td>10.6</td>
<td>7.4</td>
</tr>
<tr>
<td>Technique</td>
<td></td>
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<tr>
<td>Valveless</td>
<td>8 (50%)</td>
<td>8 (38.1%)</td>
<td>4 (26.7%)</td>
</tr>
<tr>
<td>Monocusp</td>
<td>4 (25%)</td>
<td>8 (38.1%)</td>
<td>4 (26.7%)</td>
</tr>
<tr>
<td>Valved conduit</td>
<td>4 (25%)</td>
<td>5 (23.8%)</td>
<td>7 (46.7%)</td>
</tr>
<tr>
<td>Early mortality</td>
<td>3 (18.8%)</td>
<td>4 (19%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Late mortality</td>
<td>2/13 (15.4%)</td>
<td>0/17 (0%)</td>
<td>1/15 (6.7%)</td>
</tr>
<tr>
<td>Early reoperation</td>
<td>2 (12.5%)</td>
<td>4 (19%)</td>
<td>1 (6.7%)</td>
</tr>
<tr>
<td>Late reoperation within 1 year after repair</td>
<td>8/13 (61.5%)</td>
<td>5/17 (29%)</td>
<td>1 (6.7%)</td>
</tr>
<tr>
<td>Late reoperation within 2 years after repair</td>
<td>1/12 (8.3%)</td>
<td>1/17 (5.9%)</td>
<td>0/9 (0%)</td>
</tr>
<tr>
<td>Mean follow-up duration (y)</td>
<td>22</td>
<td>15</td>
<td>2.6</td>
</tr>
</tbody>
</table>

PA to the main PA in 2003 and complete repair at 1 year of age.

Patients with airway compromise underwent urgent repair (n = 31; 59.6%). Preoperative intubation was necessary in 15 patients (28.9%) for severe airway compromise. Only patients younger than 1 year required ventilator support (all 3 neonates, 12 of 24 infants).
Operative Technique
At operation, median weight was 6.9 kg (IQR, 4.9 to 9.6 kg) and body surface area was 0.38 m² (IQR, 0.29 to 0.45 m²). Surgery was performed through sternotomy with standard cardiopulmonary bypass, hypothermia, and cold blood cardioplegia. Median cardiopulmonary bypass time was 133 minutes (IQR, 95 to 161 minutes), median aortic cross-clamping time was 69 minutes (IQR, 51 to 92 minutes), and mean minimal temperature was 26° ± 5°C (16° to 33°C). Seven patients required circulatory arrest (range, 5 to 68 minutes) during the operation, all of which occurred before 1995.

The right atrium was opened. Infundibular muscle bands were resected to relieve right ventricular outflow tract (RVOT) obstruction. The ventricular septal defect was closed using Dacron (C.R. Bard, Haverhill, PA), Gore-Tex (W.L. Gore & Assoc, Flagstaff, AZ) or pericardial patch. Atrial septal defects were closed in all patients.

Surgical reduction of the aneurysmal PA was performed (Fig 2) in 39 patients (75%) by pulmonary plication or resection. The Lecompte maneuver was performed in 2 patients (3.9%). The pulmonary valve was managed by valveless repair (n = 20; 38.5%), monocusp (n = 16; 30.8%), and valved conduit (n = 16; 30.8%). In the valveless repair, a transannular patch was made of pericardium or resected PA. In a monocusp repair, a valve was fashioned from either pericardium (n = 11), Gore-Tex (n = 1), aortic homograft (n = 2), or pulmonary homograft (n = 2). The types of conduits were Hancock valved conduit (Medtronic Inc, Minneapolis, MN; n = 3), cryopreserved pulmonary homograft (n = 6), Contegra conduit (Medtronic Inc; n = 6), and Gore-Tex (n = 1). The size of the valved conduit ranged from 9 to 22 mm. Concomitant repair at the time of APVS surgery included tricuspid valve repair (n = 1), total anomalous pulmonary venous drainage repair (n = 1), and left PA reimplantation (n = 3).

Definitions
Early mortality was defined as death occurring within 30 days of surgery or before hospital discharge. All other deaths were considered late. Early reoperation was defined as further surgery required before discharge. Any reoperation performed after hospital discharge was considered late. Neonates were younger than 1 month at repair, infants between 1 month and 1 year, children were older than 1 year but younger than 18 years, and adults were 18 years of age and older. The degree of RVOT obstruction at discharge was determined by continuous wave Doppler and defined as trivial (peak velocity, <2 m/s), mild (peak velocity, 2 to 3 m/s), and moderate (peak velocity, 3 to 4 m/s).

Data Analysis
All data were analyzed with Stata version 10 (Stata Corp, College Station, TX). Descriptive statistics for continuous variables were expressed as mean ± standard deviations (range), whereas skewed continuous data are presented as medians (IQR). Categorical variables are summarized as frequencies and percentages. The Student’s t test was used to compare two group means, and Pearson χ² test was used for inferential comparison.

Logistic regression analysis was used to determine risk factors for binary outcome, linear regression analysis for continuous variables, and Cox proportional hazards analysis for time-related outcomes. Kaplan-Meier actuarial survival curves were used to analyze and plot time-related end points. Variables tested in the univariate model included sex, age at surgery, weight, bypass time, cross-clamp time, preoperative ventilation, monocusp reconstruction, valveless repair, valved conduit, PA surgery, infant group, neonatal group, prematurity, associated syndromes, and antenatal diagnosis. A cutoff probability value of 0.07 was used for variable inclusion and exclusion in the multivariable model.

Fig 2. Surgical management of the pulmonary arteries in absent pulmonary valve syndrome. Reduction of the proximally dilated pulmonary artery to the hilum only (A1) with persisting posthilum dilatation causing distal bronchial obstruction (A2). Reduction of pulmonary artery past border of the hilum (B1) relieves distal bronchial obstruction (B2). (LPA = left pulmonary artery, RPA = right pulmonary artery.)
stepwise method was used such that variables were removed until all probability values in the multivariable model were significant. All tests were two-tailed; probability values of less than 0.05 were considered significant.

Results

Survival

Early mortality was 13.5% (7 of 52). There were 2 sudden deaths (failed resuscitation after cardiac arrest), and in 5 patients care was withdrawn after prolonged ventilation for persistent airway obstruction. Early mortality was 18.8% (3 of 16) during 1975 through 1989, 19% (4 of 21) during 1990 through 2000, and 0% (0 of 15) during 2001 through 2013. There was a trend toward a reduction in early mortality with no deaths since 2001. Table 1 outlines characteristics of the patients and operative techniques. Antenatal diagnosis improved in the current era ($p < 0.001$). Early mortality occurred in 2 (66.7%, 2 of 3) neonates and 5 (20.8%, 5 of 24) infants. There were no deaths (0%, 0 of 25) in patients older than 1 year at operation. There were significantly fewer ($p = 0.016$) patients older than 1 year (60%, 15 of 25) who required PA surgery compared with infants and neonates (88.9%, 24 of 27). Univariate analysis is outlined in Table 2. Multivariable analysis was not performed owing to the small numbers of operative deaths.

There were 3 late deaths (6.7%, 3 of 45) at 3.4 months, 6.2 months, and 29 years after surgery. The causes of late deaths were pneumococcal sepsis in 2004, failure to wean off bypass after residual ventricular septal defect closure in 1980, and unknown cause in 2004, respectively. Kaplan-Meier analysis demonstrated survival of 81.4% ± 5.6% (95% confidence interval [CI], 67.4 to 89.9) at 5, 10, and 20 years (Fig 3). Cox proportional hazard regression multivariate analysis identified preoperative ventilation (hazard ratio, 8.83; 95% CI, 1.74 to 44.7; $p = 0.009$) as the only risk factor for overall death. Survival according to repair technique is displayed in Figure 4.

Hospital Stay

The median length of hospital stay was 14 days (IQR, 9 to 28 days), median postoperative intubation time was 1.7 days (IQR, 0.6 to 4.9 days), and median length of intensive care unit stay was 2.9 days (IQR, 1.2 to 9 days). Median intubation was 11.9 days (IQR, 4.9 to 12 days) for neonates, 4.3 days (IQR, 1.7 to 10 days) for infants, and 0.83 days (IQR, 0.4 to 1.11 days) for patients older than 1 year. Multivariate analysis identified preoperative ventilation (coefficient, 1.62; 95% CI, 0.88 to 2.36; $p < 0.001$) and operation during infancy (coefficient, 0.9; 95% CI, 0.22 to 1.58; $p = 0.01$) as predictors of longer postoperative mechanical ventilation.

Major complications during hospital admission included pleural effusion requiring drainage ($n = 2$; 3.9%), pericardial effusion requiring drainage ($n = 4$; 7.7%), and complete heart block requiring permanent pacemaker ($n = 3$; 5.8%). Preoperative and postoperative echocardiographic dimensions of the PAs were compared in 38 patients. Postoperatively, main PA (10.8 ± 2.3 mm), left PA (9.1 ± 3.3 mm), and right PA diameter (9.3 ± 3 mm) were all significantly ($p < 0.001$) smaller than preoperative measurements (18.3 ± 5.4 mm, 16.1 ± 5.7 mm, and 18.5 ± 5 mm, respectively). The degree of pulmonary regurgitation at discharge was as follows: none in 4.1% (2 of 49), trivial to mild in 40.8% (20 of 49), and moderate to severe

Table 2. Univariate Analysis for Early Mortality

<table>
<thead>
<tr>
<th>Variable</th>
<th>p Value</th>
<th>Odds Ratio</th>
<th>95% Confidence Interval</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at operation</td>
<td>0.035</td>
<td>0.73</td>
<td>0.54–0.98</td>
</tr>
<tr>
<td>Weight at operation</td>
<td>0.019</td>
<td>0.37</td>
<td>0.16–0.84</td>
</tr>
<tr>
<td>Neonate</td>
<td>0.029</td>
<td>17.6</td>
<td>1.34–230.5</td>
</tr>
<tr>
<td>Prematurity</td>
<td>0.008</td>
<td>10.7</td>
<td>1.83–62.1</td>
</tr>
</tbody>
</table>

Fig 3. Overall survival after absent pulmonary valve syndrome (APVS) repair.

Fig 4. Survival after absent pulmonary valve syndrome (APVS) repair according to operative technique.
The degree of RVOT obstruction at discharge ranged from none to trivial in 53.1% (26 of 49), mild in 44.9% (22 of 49), and moderate in 2% (1 of 49) of patients. No information was available in 3 patients.

Reoperation
Early reoperations that occurred in 7 patients (13.5%) were pacemaker implantation (n = 3) in 1992, 1994, and 2010, relief of RVOT obstruction (n = 1) in 1997, and relief of airway obstruction (n = 3) in 1983, 1984, and 1990. In 2 of the latter 3 patients, the obstruction persisted distal to the main bronchi, and the patients died of respiratory failure. In the third patient, both branch PAs were further plicated distal to the site of previous plication with relief in residual bronchial compression (Fig 2). The patient survived and is currently asymptomatic.

Fourteen (31.1%, 14 of 45) hospital survivors required late reoperation: closure of residual ventricular septal defect (n = 1), pulmonary conduit replacement (n = 11), and RVOT stenosis resection (n = 2). The median interval between initial procedure and reoperation was 4.8 years (IQR, 2.1 to 11.7 years). Second late reoperation was required in 6.7% (3 of 45) of survivors: 2 valved conduit replacements and 1 pulmonary valve replacement. Overall freedom from late reoperation was 79.7% ± 6.9% (95% CI, 61.8 to 89.9) at 5 years, 69.4% ± 8.2% (95% CI, 50.2 to 82.4) at 10 years, and 52.1% ± 9.8% (95% CI, 31.8 to 69.1) at 20 years (Fig 5). Multivariate analysis identified neonatal age at repair (hazard ratio, 37; 95% CI, 2.68 to 509.8; p = 0.007) and prematurity (hazard ratio, 26.5; 95% CI, 3.6 to 192.6; p = 0.001) as predictors of late reoperation. There was no difference in late reoperation rates between valved conduit, monocusp, or valveless techniques. Freedom from late reoperation according to era is shown in Figure 6. There was a trend toward fewer reoperations within 1 and 2 years after surgery (Table 1). However, this was not statistically significant.

Follow-Up
Median follow-up time for survivors (n = 42) after surgery was 13 years (IQR, 2.8 to 19.1 years). Two patients (4.8%) were lost to follow-up. All survivors were in New York Heart Association classification I or II.

Comment
First described in 1847 by Chevers [7], APVS associated with airway compression continues to remain a challenge. Few studies have been published, and the paucity of patients in each study makes it difficult to draw management guidelines. We reviewed our experience to emphasize the importance of extensive intralobar PA reduction and determine the strategy that eliminated operative mortality in the recent era. Some of the patients in this study have been previously reported [4].

Overall operative mortality was 13.5%, but was eliminated in the current era, with no operative mortality in 15 consecutive patients since 2001. The current literature cites an early mortality of 4% to 21% [5, 8–11]. Overall survival in our study was 81.4% ± 5.6% at 10 years. This is consistent with that reported by Alsoufi and associates [8] and Hew and colleagues [11] of 87% ± 5% and 78% at 10 years, respectively. In our study, multivariate analysis of overall survival identified preoperative ventilation to be a predictor of death. The various pulmonary valve management techniques were not risk factors for overall mortality. The need for preoperative ventilation was also significantly correlated with prolonged postoperative intubation. The requirement of preoperative ventilation...
for severe respiratory distress implied significant airway compression by the aneurysmal PA.

Other authors have also identified tracheobronchomalacia to be a significant cause of mortality [5] and have reported preoperative ventilation requirements or severe respiratory distress to be risk factors for poor outcome [8, 9, 11]. Airway compromise plays a key role in postoperative mortality and morbidity. Numerous techniques have been suggested [11–17] to eliminate airway compression, including anterior and posterior plication of aneurysmal PA, excision of PA wall segments, suspension of the PA to the retrosternal fascia, translocating the PA anterior to the aorta with Lecompte maneuver [6], or complete replacement of the mediastinal PA with a bifurcated pulmonary homograft. However, none of these techniques are effective unless the distal airway compression is eliminated. In our study, 5 of 7 operative deaths had persistent distal airway compression identified by postoperative bronchogram. This led us to introduce the technique of intrahilar reduction of the PAs. One patient in the middle era had an early reoperation using this technique and survived. No patients in the current era required early reoperation on the PAs. Thus, we emphasize the importance of intrahilar branch PA reduction (Fig 2). The Lecompte maneuver [6] was used in 2 patients in our series (both occurred within the current era) with no mortality. Endobronchial stents have been a proposed management option [18, 19] for persistent malacia. However, the stenting does not relieve distal bronchial compression and has not been used at the Royal Children’s Hospital. Our current surgical algorithm is to perform a preoperative echocardiogram and computed tomography scan, intrahilar PA reduction, postoperative bronchogram, and computed tomography scan in those who cannot be weaned off from mechanical ventilation.

No consensus exists about the preferred method of RV-to-PA reconstruction. Some authors have suggested that pulmonary valve competence is important in reducing long-term risk of arrhythmias and late RV dysfunction along with improving early postoperative hemodynamics and reduction of persistent PA dilatation [5, 8, 20]. Brown and coworkers [5] (n = 20; 1984 to 2005) and Alsoufi and associates [8] (n = 61; 1982 to 2006) have suggested establishing a competent RV-to-PA connection as part of their management strategy of APVS. In contrast, Chen and colleagues [10] (n = 23; 1990 to 2005) have reported an overall survival of 89% and low postoperative morbidity while using a valveless technique. In addition, Hew and colleagues [11] (n = 54; 1960 to 1998) reported no mortality with a transannular patching technique in APVS patients beyond the neonatal period, despite residual pulmonary regurgitation. Although the valveless repair and implantation of monocusp or valved conduits were used throughout the study period, there was a tendency to use valved conduits more often during the recent decade. Our improved results during the last 12 years may be related to the combination of increased experience with this condition, the application of the extensive branch PA reduction technique described in Figure 2, and a trend toward increased utilization of valved conduits.

In this current study, freedom from reoperation was 69.4% ± 8.2% at 10 years. Similarly, Alsoufi and coworkers [8] and Brown and associates [5] reported freedom from reoperation at 10 years to be 59% ± 9% and 66%, respectively. When adjusting for follow-up time, there was a trend toward a reduction in late reoperation (Table 1) during the three eras. Risk factors for reoperation were neonatal age at repair and prematurity. Brown and colleagues [5] identified the use of pulmonary homograft and Gore-Tex monocusp patch as predictors for reoperation. However, we did not identify any difference between the various techniques, perhaps owing to relatively small number of patients in each group.

Survival beyond 1 year after repair offers good outcome. However, patients were likely to require late reoperation to replace the RV-to-PA tract or implant the pulmonary valve. Thus, a long-term follow-up is needed.

Limitations
This study is limited by its retrospective nature. Operative techniques, perioperative management, and imaging methods have varied during the long study period. Numbers of patients in each surgical technique group were small, thus limiting the power of the study to identify clinically significant risk factors.

Conclusions
Long-term survival for APVS has improved during the last decade. Death after 1 year of repair is rare. However, continued follow-up is required as a large proportion may require reoperations later in life. Preoperative ventilation predicted longer postoperative ventilation and mortality.

Yves d’Udekem is a Career Development Fellow of The National Heart Foundation of Australia (CR 10M 5339).

References
INVITED COMMENTARY

The paper by Yong and colleagues [1] from Royal Children’s Hospital, Melbourne, Australia in the current issue describes a retrospective review of 52 patients with absent pulmonary valve syndrome who underwent surgical repair at Royal Children’s Hospital between 1975 and 2013. The median follow-up was 13 years and extended as long as 35 years. Early mortality was eliminated in the last decade, and late survival has been stable at 5, 10, and 20 years at 81%. Preoperative ventilation was the only risk factor for overall mortality. Risk factors for late reoperation were prematurity and neonatal primary repair.

These encouraging results are good news for families who often face challenging pulmonary problems in the early years of life. Until now, there has been limited late information about this rare anomaly.

While the paper provides welcome news for patients and families, it is less helpful for caregivers who must decide between various treatment options. There are numerous controversies, such as: When is it appropriate to replace completely the mediastinal pulmonary arteries with a valved conduit? If a conduit is going to be used, should it be a bifurcated pulmonary homograft, or a Contegra bovine conduit or a monocusp Gore-Tex conduit adequate? What is the role of the Lecompte maneuver? What anesthetic maneuver or ventilator maneuvers are helpful to manage the severe air trapping that frequently results from the combination of bronchomalacia and distally dilated branch pulmonary arteries? How much of a problem is the management of chondromalacia and distally dilated branch pulmonary arteries? How much of a problem is the management of DiGeorge syndrome, both early and late? These questions are going to be difficult to answer because this anomaly is rare and exists in a wide spectrum of severity. Only a multiinstitutional study similar to those organized by the Congenital Heart Surgeons’ Society will be able to help to begin to answer these many questions.

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Reference