# Surgical outcomes of absent pulmonary valve syndrome: An institutional experience

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## ABSTRACT

**Background :** **Absent pulmonary valve syndrome (APVS) is a variant of tetralogy of Fallot characterized by aneurysmal pulmonary arteries, which compresses the tracheobronchial tree, leading to respiratory symptoms. We report the mid-term outcomes of surgical correction of patients with APVS.**

**Subject and Methods :** **A total of 27 patients underwent surgery between 2001 and 2015, and they were followed up for a mean period of 6.4 ± 4.1 years. Out of the 27 patients, 14 (51.9%) were infants. The median age at repair was 9.8 months. Preoperative intubation was required in six patients (22.2%), and 11 patients (40.7%) had symptoms of respiratory distress. The pulmonary valve was replaced with a valved conduit in 15 patients (55.6%), monocusp valve in 6 patients (22.2%), and a transannular patch in 6 patients (22.2%). Reduction pulmonary arterioplasty was done in all patients.**

**Results :** **The overall 10-year survival was 82.1%. There was 81.1% overall freedom from re-intervention at 10 years. No statistically significant difference was found in 10-year survival ($P = 0.464$) and reoperation rates ($P = 0.129$) between valved conduit, monocusp, or transannular patch techniques. Older children had statistically significantly longer survival ($P = 0.039$) and freedom from re-intervention ($P = 0.016$) compared to infants. Patients without respiratory complications had 100% 10-year survival and 93.3% freedom from reoperation at 10 years compared to 55.6% and 60.1%, respectively, for patients with respiratory complications.**

**Conclusion :** **There has been improvement in surgical results for APVS over the years. However, it still remains a challenge to manage infants and patients with persistent respiratory problems.**

**Keywords :** Absent pulmonary valve, airway compression, aneurysmal pulmonary artery

## INTRODUCTION

Absent pulmonary valve syndrome (APVS) is a rare variant of tetralogy of Fallot (TOF), which accounts for approximately 3%–6% of patients with TOF. It is a congenital anomaly characterized by features similar to TOF, but with either rudimentary ridges or total absence of a pulmonary valve.[1,2] Another important feature of APVS is the marked aneurysmal dilatation of proximal pulmonary arteries, causing extrinsic compression of the tracheobronchial tree, leading to respiratory symptoms of variable severity. Patients presenting with profound respiratory distress earlier in life quite often require ventilatory support followed by urgent surgery.

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The surgical management of this congenital anomaly has evolved over the past few decades, however there is currently no general consensus on the method of repairing the dilated pulmonary arteries, and also the ideal technique for right ventricular outflow tract (RVOT) reconstruction remains debatable.[3-5] Although the mortality for surgical repair of APVS was high previously, especially in neonates and infants, the rates have come down significantly to 10%–20% in recent years.[6] However, these patients have been noted to face persistent respiratory symptoms and require further re-interventions in future. This study was carried out to report our center's experience of surgical management of APVS in a consecutive series of patients and to look at their outcomes.

## SUBJECTS AND METHODS

### Patients

Between January 2001 and December 2015, a total of 27 patients with a diagnosis of APVS who underwent total correction surgery at Institut Jantung Negara, Kuala Lumpur, Malaysia were included. A retrospective review of our electronic database and medical records were conducted after obtaining institutional ethics committee clearance. These patients were followed up for a period of up to 14.4 years (mean 6.4 years) from the time of surgery. Follow-up data were obtained via outpatient records and telephone consultations.

Out of the 27 patients, 14 (51.9%) were infants, while the other 13 (48.1%) were children older than 1 year of age. The median age at repair was 9.8 months (range, 1.5–72 months). There were 19 (70.4%) males, which made up majority of the patient cohort. Two patients (7.4%) were born prematurely, whereas 25 (92.6%) of them were born at term. The mean body surface area (BSA) at the time of surgery was 0.42 ± 0.17 $m^2$. In this APVS group, 11 patients (40.7%) presented with symptoms of respiratory distress, out of which six patients required preoperative intubation and ventilation, while one was placed on bi-level positive airway pressure ventilation support [Table 1]. All the patients who had respiratory distress (7 infants, 4 older children) were operated on urgently during the same hospital admission. Other associated cardiovascular anomalies in our patient cohort included an absent left pulmonary artery (PA) ($n = 1$), atrial septal defect ($n = 6$), right aortic arch ($n = 2$), left superior vena cava ($n = 1$), and major aorto-pulmonary collateral arteries ($n = 1$). Four patients also had underlying syndromic disorders such as DiGeorge syndrome ($n = 2$), Noonan syndrome ($n = 1$), and CHARGE syndrome ($n = 1$).

The diagnosis of APVS was established by preoperative two-dimensional transthoracic echocardiography in all patients. Computed tomography angiogram and flexible bronchoscopy were done in the seven patients who presented with respiratory distress and required ventilatory support. This was done mainly to rule out other causes of respiratory distress. It was noted that although the PA dilatation was only mild to moderate in four of them, it was significant enough to cause major airway compression due to the underlying tracheobronchomalacia.

**Table 1: Patient demographics and perioperative variables**

| Variable | *n* (%) |
|---|---|
| Age (months), median (range) | 9.8 (1.5-72) |
| BSA ($m^2$), mean±SD | 0.42±0.17 |
| Gender | |
| Male | 19 (70.4) |
| Female | 8 (29.6) |
| Premature baby | |
| Preterm | 2 (7.4) |
| Term | 25 (92.6) |
| Infant | 14 (51.9) |
| Older children | 13 (48.1) |
| Preoperative ventilation | |
| No | 20 (74.1) |
| Yes | 6 (22.2) |
| BiPAP | 1 (3.7) |
| Respiratory distress | |
| No | 16 (59.3) |
| Yes | 11 (40.7) |
| Types of pulmonary artery arterioplasty | |
| Anterior resection of RPA + Lecompte maneuver | 1 (3.7) |
| Anterior resection of RPA and LPA | 5 (18.5) |
| Anterior resection of MPA, LPA, RPA | 21 (77.8) |
| Types of RVOT reconstruction | |
| Transannular patch | 6 (22.2) |
| Monocusp valve | 6 (22.2) |
| Valved conduit | 15 (55.6) |

RPA: Right pulmonary artery; LPA: Left pulmonary artery, MPA: Main pulmonary artery, RVOT: Right ventricular outflow tract, BSA: Body surface area, BiPAP: Bi-level positive airway pressure, SD: Standard deviation

### Surgical technique

All the 27 patients did not undergo any palliative procedure, and a single-stage primary repair was carried out in them. The surgical correction was done via a median sternotomy with aortic and bicaval cannulation, standard cardiopulmonary bypass, mild-to-moderate systemic hypothermia (28°C–32°C), and intermittent antegrade cold blood cardioplegia. None of the patients required any circulatory arrest to perform the repair.

A right atriotomy was done together with a longitudinal incision along the main PA (MPA) and the RVOT to assess the intracardiac anatomy. Any atrial septal defect or patent foramen ovale, if present, was closed with an autologous pericardial patch or direct suture technique. Next, the hypertrophied infundibular muscle bands were resected to relieve the RVOT obstruction, followed by closure of the ventricular septal defect (VSD) using a Gore-Tex (W.L. Gore & Assoc., Flagstaff, AZ, USA) patch.

The aneurysmal PAs were repaired by performing a reduction pulmonary arterioplasty via anterior resection

in all patients. RVOT reconstruction was then done using either a right ventricle to PA (RV-PA) valved conduit or a transannular patch with or without a monocusp valve. The monocusp valve was constructed using either autologous pericardium (treated with 0.6% glutaraldehyde for 10 min) or pulmonary homograft. Transannular patch repair alone was done using autologous pericardium (treated with 0.6% glutaraldehyde for 10 min). The types of RV-PA conduits used were Contegra bovine jugular vein valved conduit (Medtronic Inc., Minneapolis, MN, USA), Labcor stentless valved pulmonary conduit (Labcor Laboratõrios, Belo Horizonte, Brazil), and cryopreserved valved pulmonary homograft.

### Statistical analysis

All data collected were analyzed using the IBM SPSS statistical software version 22 (Armonk, New York, USA: IBM corp). Continuous variables were expressed as mean ± standard deviation, median, and range. Categorical variables were presented as frequencies and percentages. Kaplan–Meier survival curves were used to estimate patient survival over the years and to calculate their freedom from re-intervention. Differences between survival curves in subgroups were identified by using the log-rank test. The Cox proportional hazards regression model was used to perform multivariate analysis to identify risk factors for overall mortality and reoperation. The variables evaluated included sex, age during surgery, BSA, cardiopulmonary bypass time, aortic cross-clamp time, preoperative ventilation, presentation with respiratory distress, presence of genetic syndromes, type of PA arterioplasty, and type of RVOT reconstruction. $P < 0.05$ was considered statistically significant.

## RESULTS

### Surgery

The mean cardiopulmonary bypass time was 173.2 ± 55.5 min (range, 79–311 min), and the mean aortic cross-clamping time was 102.7 ± 34.8 min (range, 50 to 212 min). In repairing the aneurysmal PAs, 21 patients (77.8%) underwent anterior resection of the MPA, right PA (RPA), and left PA (LPA). Five patients (18.5%) had anterior resection to the RPA and LPA, while one patient (3.7%) had anterior resection of the RPA together with a Lecompte maneuver to bring the PA anterior to the aorta [Table 1]. The last approach was used in the patient with absent LPA who underwent a single-lung repair. The RVOT was reconstructed by using a RV-PA valved conduit in 15 patients (55.6%); out of which 11 were Contegra valved conduits, two were Labcor valved pulmonary conduits, and two were pulmonary homografts. Six patients (22.2%) underwent monocusp valve implantation (autologous pericardium: $n = 5$, pulmonary homograft: $n = 1$), and valveless repair with a transannular patch was performed in another six patients (22.2%) [Table 1]. In older children, four patients (30.8%) received a valved conduit, five patients (38.4%) had a monocusp valve, and four patients (30.8%) had a transannular patch repair. In the infant group, 11 out of the 14 patients (78.6%) received a valved conduit for their RVOT reconstruction and together with another patient who had a monocusp valve, 12 infants (85.7%) had a competent pulmonary valve post repair [Figure 1].

### Hospital course

The median length of hospital stay was 11 days (range, 5–92 days). Postoperatively, patients were mechanically ventilated for a median duration of 3 days (range, 1–61 days), and they were nursed in the intensive care unit for a median time of 5 days (range, 2–72 days). All patients had a flexible bronchoscopy postsurgery, and they were only extubated after ensuring that the airways were patent while on minimal ventilatory support. Out of the 27 patients, 10 (37%) suffered from respiratory complications marked by prolonged mechanical ventilation (more than 7 days), respiratory failure, and bronchopneumonia. Infants (80%) made up the majority of these patients who developed respiratory complications. They were intensively managed with various methods including regular bronchodilators, positive pressure ventilation, ventilation in a prone position with postural drainage, and effective antibiotics. Other postoperative complications noted were low cardiac output syndrome ($n = 1$), junctional ectopic tachycardia ($n = 2$), moderate mitral regurgitation

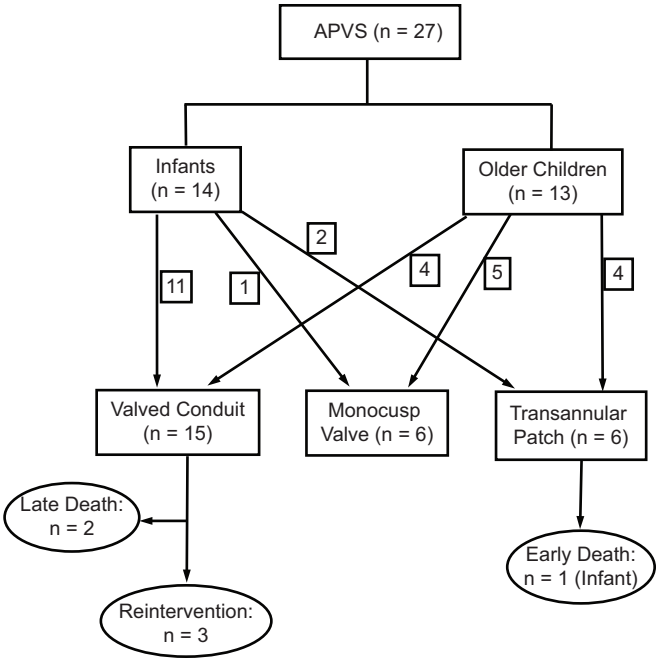


**Figure 1: Distribution of the types of right ventricular outflow tract reconstruction done in infants and older children and their outcomes (APVS: Absent pulmonary valve syndrome)**

which was managed medically ($n = 1$), and chylothorax requiring drainage ($n = 1$).

### Survival

The outcomes based on patient age group and the types of surgical repair done are summarized in Figure 1. There was one early mortality (3.7%) and two late mortalities (7.4%), all belonging to the infant group. Early mortality was defined as death occurring within the same hospital admission or within 30 days from surgery. All other mortalities were considered late. The early death occurred in an infant 5 days after surgery due to Type 2 respiratory failure and persistent lung collapse. Late deaths were reported in two older children at 10 months and 36 months postsurgery, who were operated on as infants. One died due to acute respiratory failure and the other due to severe bronchopneumonia with sepsis. Out of the three mortalities, two patients (66.7%) initially presented with respiratory distress symptoms and were mechanically ventilated preoperatively. All the three had underlying tracheobronchomalacia and respiratory complications after surgery.

Overall survival shown by the Kaplan–Meier curve at 5 and 10 years was 82.1% ±8.1% [Figure 2]. No statistically significant difference was found in 10-year survival using log-rank testing between valved conduit, monocusp valve, or transannular patch techniques [$P = 0.464$, Figure 3]; although all patients who had a monocusp valve had 100% freedom from mortality. Infants only had a 65.6% 10-year survival, which was significantly poorer compared to 100% 10-year survival in older children [$P = 0.039$, Figure 4]. Patients without postoperative respiratory complications had 100% freedom from mortality at 10 years compared to only 55.6% for patients with respiratory complications [$P = 0.007$, Figure 5]. However, on multivariate analysis, no independent risk factor was found to affect the overall survival.

### Re-intervention

Three patients (11.1%) out of the 27 required late reoperations following the initial surgery; all of them had a Contegra valved conduit implanted during infancy [Figure 1]. One patient underwent a conduit change with an aortic homograft due to infected conduit 3 years postsurgery, while another patient developed a large MPA-LPA aneurysm with thrombus 4 years later and had to go for a repair of the aneurysm and change of conduit with an aortic homograft. The third patient had conduit stenosis with high RVOT gradient 2 years after surgery and went for a conduit change with pulmonary homograft.

There was an overall 81.1% ± 8.1% freedom from re-intervention at 5 and 10 years [Figure 6]. No statistically significant difference was found in reoperation rates using log-rank testing between valved conduit, monocusp valve, or transannular patch techniques [$P = 0.129$, Figure 7]. The infants again had a statistically significantly lesser 10-year freedom from reintervention at 47.4%, compared to 100% for older children [$P = 0.016$, Figure 8]. Patients with respiratory complications had 60% freedom from reoperation at 10 years as opposed to 93.3% in those without respiratory complications [$P = 0.206$, Figure 9].

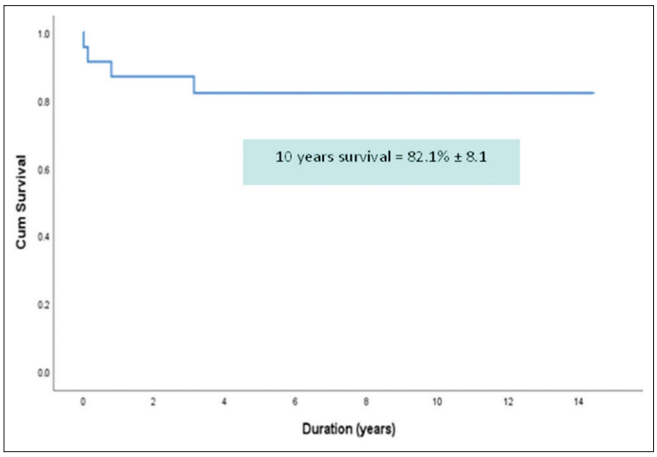


**Figure 2: Overall survival after absent pulmonary valve syndrome repair estimated by the Kaplan–Meier method**

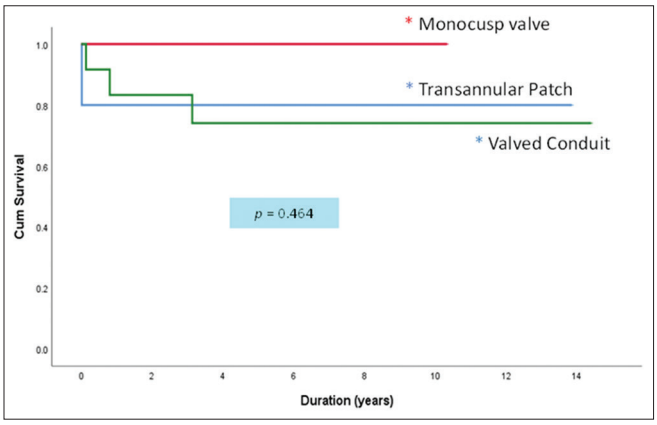


**Figure 3: Difference in survival based on the type of right ventricular outflow tract reconstruction**

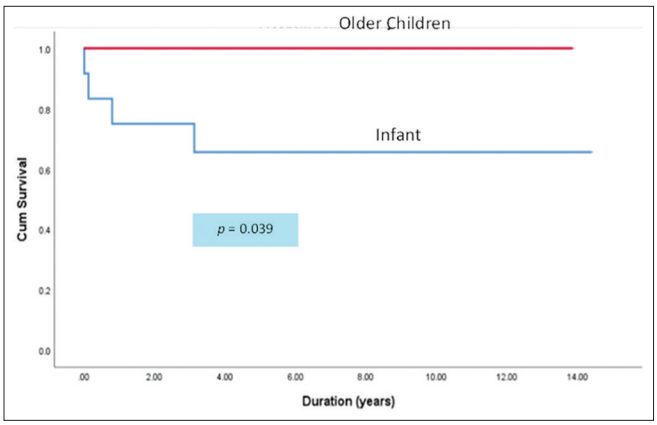


**Figure 4: Difference in survival based on the age of patients**

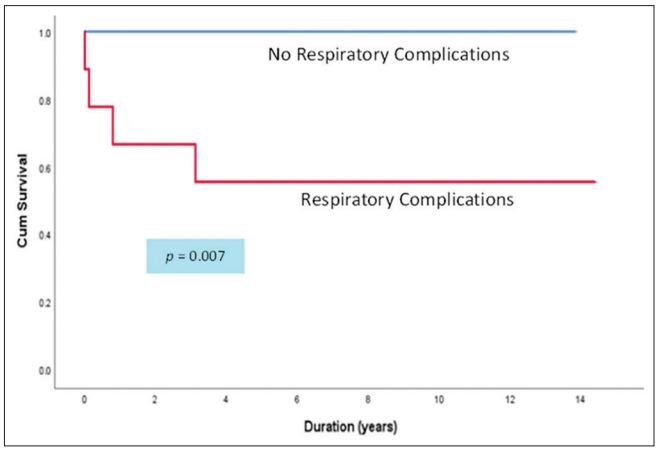


**Figure 5: Difference in survival based on respiratory complications**

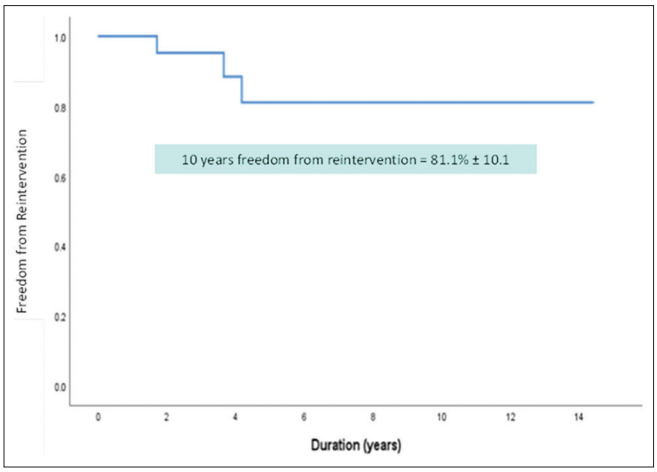


**Figure 6: Overall freedom from reoperation after absent pulmonary valve syndrome repair**

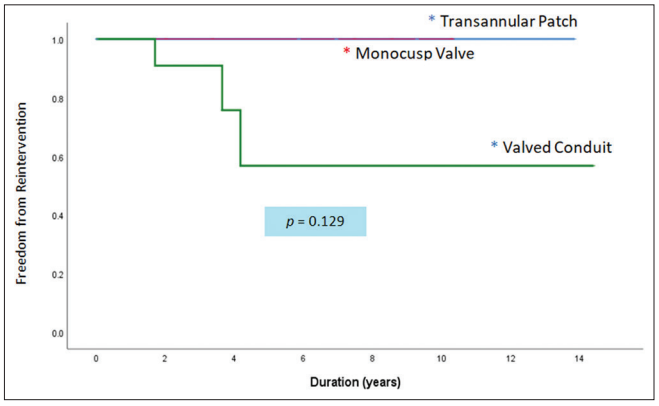


**Figure 7: Difference in freedom from reoperation based on the type of right ventricular outflow tract reconstruction**

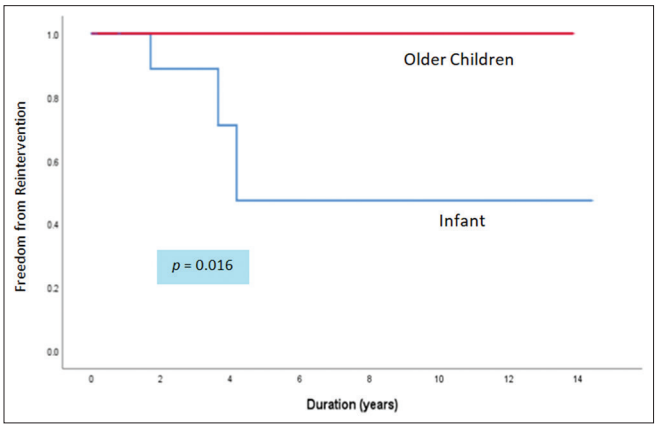


**Figure 8: Difference in freedom from reoperation based on the age of patients**

On multivariate analysis, we were again unable to identify any significant risk factor for re-intervention.

### Follow-up

All patients who survived the initial surgery (26 out of 27) were followed up for a mean period of 6.4 ± 4.1 years (median 7.0 years, range 6 months to 14.4 years). One patient was uncontactable and was lost to follow-up. Taking into account the two late mortalities, the remaining 23 late survivors were found to be well in New York Heart Association functional Class I or II, with no significant limitations in daily activities. Eight patients (34.8%) have already started schooling and one (4.3%) is currently self-employed. However, four patients (17.4%) among the late survivors had been noted to have recurrent hospital admissions for respiratory symptoms such as bronchospasm and lower respiratory tract infections. The frequencies of their admissions have been reducing as they got older. They were followed up with yearly bronchoscopy and echocardiogram. Although their pulmonary arteries were not further dilated, they still had persistent tracheobronchomalacia and were given expectant management when needed with bronchodilators, prophylactic antibiotics, and noninvasive positive pressure ventilation. Two patients were on intermittent nasal continuous positive airway pressure device at home.

## DISCUSSION

Chevers in 1847 first described the syndrome of TOF with absent pulmonary valve,[7] which includes characteristics of anterior mal-alignment VSD, overriding aorta, right ventricular hypertrophy, pulmonary annular stenosis with the absence of a functional pulmonary valve causing pulmonary regurgitation, plus aneurysmal dilatation of the pulmonary trunk, and two main branch PA. Since then, there have been various surgical methods used to address this condition which reduce the compressive effect of the dilated PAs on the trachea and main bronchi. In the early years before the 1980s, infants who presented with severe respiratory distress commonly had palliative procedures such as transecting the RPA and bringing it anterior to the aorta and reattaching it to the MPA using an interposition graft,[8] or banding or ligation of the MPA with creation of a systemic-to-PA shunt.[9,10]

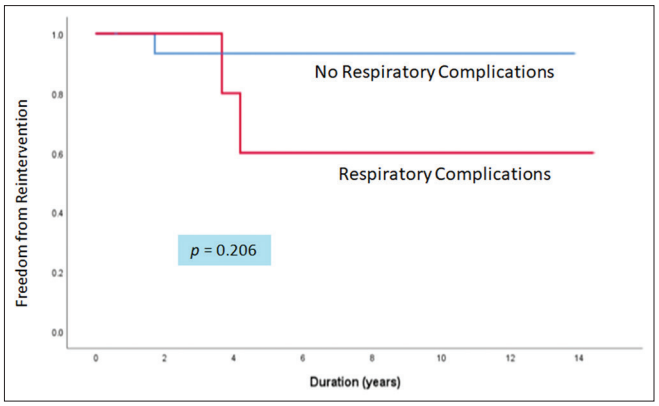


**Figure 9: Difference in freedom from reoperation based on respiratory complications**

In later years with the improvement in surgical techniques and the usage of homografts/conduits, the treatment of choice is to perform a complete surgical repair in all patients, including symptomatic neonates and infants. Due to the rarity of this condition and scarcity of large studies to propose management protocols, there is a huge variability in the surgical approaches to correcting APVS. It is generally agreed that almost all patients with dilated PAs will need some form of reduction pulmonary arterioplasty or plication to relieve the airway obstruction.[3,5,6,11,12] Another option advocated by Hraska *et al.*[13] is to perform a Lecompte maneuver by transecting the ascending aorta and translocating the PAs anterior to the aorta, away from the tracheobronchial tree.

The main controversy, however, lies in the best way of managing the pulmonary valve and RVOT; whether by using a RV-PA valved conduit, or monocusp valve implantation, or valveless repair with a transannular patch. In 1985, McCaughan *et al.*[14] recommended the establishment of pulmonary valvular competence in markedly symptomatic patients who were generally younger, while it was not so important in minimally symptomatic older children. Similarly, Hew *et al.*[15] and Jonas[11] have advocated a complete replacement of the main and branch pulmonary arteries with a valved pulmonary homograft for neonates presenting with severe respiratory distress, whereas for asymptomatic infants and older children, they found that such an aggressive approach is usually not necessary and even transannular patching may be sufficient for managing the RVOT. On the contrary, Godart *et al.*[16] recommended treatment with pulmonary arterioplasty without pulmonary valve insertion except in patients with pulmonary hypertension and achieved 92% overall survival. Chen *et al.*[17] also reported an overall survival of 89% and low postoperative morbidity in neonates and infants by using a valveless RV-PA connection with transannular patch.

At our center, we used a valved conduit for a majority (78.6%) of the infants, while the methods of RVOT reconstruction were almost equally distributed for older children [Figure 1]. Although the choice of RVOT reconstruction method was mainly based on the individual preference of five different surgeons, we were more inclined to establish a competent pulmonary valve in infants (12 out of 14) to prevent volume overload of the RV and to aid a better postoperative recovery in this high-risk group. Nevertheless, numerous studies including ours have compared the various types of RVOT reconstruction techniques and found that they made no difference to the survival of these patients.[3-6,18] Although not significant, our series also found that using a valved conduit confers a relatively higher risk of re-intervention in 10 years compared to other techniques [Figure 7].

The overall outcomes from this study are comparable to other recent published series of APVS mainly with respect to 10-year survival, late death, and reoperation, with a relatively lower infant mortality rate [Table 2]. Multiple studies including ours have identified that neonates and infants with respiratory distress often require early mechanical ventilation, and these patients have poorer outcomes in terms of survival and reoperation compared to older children.[3-5,15,18,21] Respiratory complications and morbidity are mainly associated with postoperative recurrent respiratory infections and ventilator dependency. Despite successful PA plication or arterioplasty, persistent respiratory complications in some patients are linked to the underlying tracheobronchomalacia and intraparenchymal airway disease.[6,12] Identification of the cause of persistent airway obstruction in these patients can be done by performing a bronchoscopy, and the usage of intrabronchial expandable stents has been reported in the past to successfully treat this condition.[22-24]

### Limitations

This study is limited by it being a retrospective review and the results are influenced by selection bias and lack of randomization. Our study population is also small, due to the rarity of APVS and this being a single-center study. There are biases in terms of patient selection, surgeon preference for surgical methods of repair, and also advances in perioperative management during the study period. Data analyzed were also limited to what was available in the medical records.

## CONCLUSION

Despite improvement in surgical outcomes for the management of APVS over the years, the outcomes for infants seem poorer compared to those of older children. Furthermore, it still remains a challenge to manage infants and patients with persistent respiratory problems

**Table 2: Comparison with other series of surgical management of absent pulmonary valve syndrome**

| Study | Number of patients: Total/infants | Infant mortality (%) | 10-year survival (%) | Mean follow-up (years) | Late death | Reoperation |
|---|---|---|---|---|---|---|
| Snir *et al.* (1991)[19] | 22/8 | 25 | - | 3.6 | 1 | 0 |
| Watterson *et al.* (1992)[20] | 19/18 | 17 | - | - | 1 | 5 |
| Godart *et al.* (1996)[16] | 37/10 | 20 | - | 2.8 | 1 | 1 |
| Dodge-Khatami *et al.* (1999)[22] | 11/10 | 10 | - | 2.1 | 1 | 5 |
| McDonnell *et al.* (1999)[21] | 28/18 | 33 | 72 | 5.5 | 1 | 3 |
| Hew *et al.* (2002)[15] | 54/39 | 21 | 78 | 6.4 | 5 | 15 |
| Brown *et al.* (2006)[3] | 20/11 | 9 | 85 | 7.3 | 2 | 6 |
| Alsoufi *et al.* (2007)[18] | 62/29 | 10 | 87 | 8.0 | 5 | 18 |
| Hu *et al.* (2013)[6] | 42/22 | 14 | 92 | 5.2 | 3 | 2 |
| Yong *et al.* (2014)[5] | 52/27 | 26 | 81 | 13.2 | 3 | 18 |
| This study | 27/14 | 7 | 82 | 6.4 | 2 | 3 |

postsurgery, especially in a developing country. These patients may end up having multiple hospitalizations, and a continued long-term follow-up is crucial as many of these patients survive into adulthood and may need future re-interventions.

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**Conflicts of interest**

There are no conflicts of interest.

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