Simple Aortopulmonary window: Outcomes of surgical repair at Vietnam National Children's Hospital

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ABSTRACT

Purpose: Evaluate the midterm outcomes of surgical repair for patients diagnosed with simple aortopulmonary window at Vietnam National Children's Hospital.

Methods: We retrospective review data of patients who underwent surgical repair for simple aortopulmonary window from May 2014 to December 2023 at Vietnam National Children's Hospital. Patients who had ventricular septal defect, interrupted aortic arch, aortic arch hypoplasia, double outlet of the right ventricle, or Berry syndrome were excluded from this study.

Results: There were 37 patients who underwent surgical repair in this study. The mean age at the time of surgery was 259.37 ± 702.59 days, and the mean weight was 5.04 ± 3.7 kg. According to the Society of Thoracic Surgeons Congenital Heart Database Committee classification, sixteen patients were type1, twelve were type2, and nine were type3. Two patients underwent ligated window, 3 had window division and sutures without bypass, and 32 had patch repair. There were 4 in-hospital deaths (10.8%), no late deaths, and no patients required cardiac reoperation during a mean follow-up of 62.09 ± 31.07 months.

Conclusions: Midterm survival beyond discharge of simple aortopulmonary window is excellent.

Keywords: *AP window, aortopulmonary window (APW).*

INTRODUCTIONS

Congenital anomaly of the aortopulmonary window (APW) is a rare disease, accounting for 0.1% to 0.2%. This anomaly was defined as a communication between the ascending aorta and the pulmonary artery in the presence of two individual semilunar valves (1). The defect can occur between the aorta and the main pulmonary artery or incorporate distal into the right pulmonary artery. Literature records that a significant number of patients diagnosed with APW were associated with other congenital heart defects such as ventricular septal defect, interrupted aortic arch, double outlet of the right ventricle, etc (2)(3)(4)(5). Given the rarity of APW, there were almost no reports in Vietnam about the surgical results and outcomes after APW repair. Therefore, we investigated the surgical management and midterm outcomes of simple APW repair in Vietnam National Children's Hospital.

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METHODS

From May 2014 to December 2023, patients were diagnosed with simple APW and underwent surgical repair at Vietnam National Children's Hospital. Data was retrospectively reviewed via patients' medical records and follow-up information was completed via telephone contact with the patient's family. According to the Society of Thoracic Surgeons (STS) Congenital Heart Database Committee classification described by Jacobs and colleagues (1), the type of APW was as follows: type I, a proximal defect just above the semilunar valve; type II, a distal defect adjacent to the right pulmonary artery; type III, a total defect from just above the semilunar valve to the right pulmonary artery, and type IV, an intermediate defect (**Figure 1**). Simple APW was defined as isolated APW, those have no associated cardiac lesions other than patent ductus arteriosus or foramen ovale.

Surgical techniques

The surgical repair was performed via midline sternotomy approach. There was a total of 5 patients who underwent APW repair without the use of cardiopulmonary bypass, depending on surgeons' perspective: 2 patients had APW ligation, and 3 had division and primary closure of both the aorta and pulmonary artery. The remaining 32 patients underwent APW repair with cardiopulmonary bypass. The aortic cannula usually requires placement near the base of the innominate artery. The branch pulmonary arteries were temporarily snared until cardioplegia arrest finished. The pulmonary artery trunk was opened and a patch of bovine pericardium was used to separate the aorta and the pulmonary artery by running polypropylene suture. The incision in the pulmonary artery was closed directly or patch if needed.

Follow-up

All patients were requested to complete their follow-up at the outpatient clinic at 1 month, 3 months, 6 months, and then annually after operation. Medical records were collected from the hospital database and contact with family's patients at the end of the study via telephone.

Statistical analysis

Continuous variables are expressed as mean with standard deviation. Categorical variables are presented as frequencies and percentages. Estimated for survival were made by the Kaplan-Meier method. Statistical analyses were performed using SPSS 20.0 software.

RESULTS

From May 2014 to December 2023, simple APW was diagnosed and surgical repair was performed in 37 patients at Vietnam National Children's Hospital. There were 19 boys (51.4%). The mean age and mean weight at operation were 259.37 ± 702.59 days, and 5.04 ± 3.7 kg, respectively. Seven patients (18.9%) were born prematurely at or before 36 weeks' gestation.

Respiratory distress was detected in 14 patients (37.8%), 10 patients required preoperative mechanical ventilation before operation, and one patient had cardiogenic shock when admitted to the hospital. Four patients (10.8%) required emergency operations due to severe congestive heart failure and respiratory distress. Five patients had extracardiac anomalies, included 2 patients with anorectal anomalies, 1 had congenital

diaphragmatic hernia, 1 had congenital inguinal hernia, and 1 had microcephaly syndrome. Patients' demographics and preoperative conditions are described in **Table 1**.

There were sixteen patients (43.2%) were type1, twelve (33.4%) were type2, and nine (24.4) were type3 according to the STS Database classification. One patient had pulmonary artery banding before definitive repair due to difficulty in infection control and severe congestive heart failure. The mean diameter of APW was 10 ± 3.92 mm. All patients underwent surgical repair via the midline sternotomy approach. Except for 5 patients (13.5%) who had APW ligation or division without cardiopulmonary bypass, the remaining 32 (86.5%) underwent APW repair using mild-moderate hypothermia cardiopulmonary bypass. The mean aortic cross-clamp time, bypass time and operation time were 49.7 ± 25.7 minutes, 77.6 ± 50.4 minutes, and 173.9 ± 60.7 minutes, respectively. The patent foramen ovale was left open in 18 patients (48.6%). Two patients need open chest and those two patients also require extracorporeal membrane oxygenation support. There were 12 patients (32.4%) had postoperative low-cardiac output syndrome, 2 (5.4%) had bleeding that required reoperation, 14 (37.8%) had liver function failure, and 13 (35.1%) had acute kidney failure that required peritoneal dialysis. The details information on perioperative variables is described in **Table 2**.

The mean ventilation time, postoperative ICU stay time were 90.5 ± 123.7 hours, and 49.7 ± 80.7 days, respectively. Four patients (10.8%) died in 30 days after operation, giving the survival of 89.2% at 10 years (**Figure 2**). Two patients (5.4%) required postoperative reoperation with 1 patient had partial anomalous return of the left superior pulmonary vein and 1 patient had conduit exchange with severe conduit stenosis after the operation. Two patients need postoperative extracorporeal membrane oxygenation support. There was no patient loss follow-up in 33 survivals with the mean time of follow-up being 62.1 ± 31.1 months. All survival patients had normal echocardiography with Ross class I at the latest follow-up.

COMMENTS

APW is a rare congenital heart anomaly, as many single-institution series of small numbers of patients underwent surgical repair for an extended period (2)(3)(4)(6). The mean age of 259 days (8.6 months) in our study is comparable to some recent studies from developing countries (6)(7)(8). This reflects our poor capacity to diagnose the disease in our conditions, in contrast to developed nations' ability to detect it early in neonates or small infancy. The diameter of the aortopulmonary window in our study is relatively big, comparable to the size of prior studies from prosperous countries (2)(3). This resulted in 29 patients (78.4%) were hospitalized with varied degrees of malnutrition, 14 patients (37.8%) with respiratory distress, 8 patients (21.6%) with pneumonia, and 4 patients (14.8%) required emergency surgery. As a result, the mean duration of mechanical ventilation following surgery in this research is longer than in other studies, with the primary cause being to treat pulmonary hypertension after surgery. Therefore, the necessity to diagnose APW lesions earlier in the community and increase the ability to screen

newborn children would be the key priority for developing nations to limit late surgery, which has numerous risks of early mortality and complications.

There were 4 patients died in-hospital in this study, giving a survival rate of 10 years of 89.2%. Two patients died related to nosocomial infection, with positive culture of the endotracheal tube with bacteria. One patient had a complication with the left main coronary injury when the surgeon tried to divide the APW. An attempt was made to repair the injured coronary artery but was not successful. The patient was supported with extracorporeal membrane oxygenation but could not recover and was withdrawn from treatment at 2 weeks after surgery. The last patient was diagnosed with APW type 3 with a single coronary artery. The patient underwent division of the APW and right ventricular outflow tract reconstruction with a polytetrafluoroethylene conduit of 6mm. After the operation, the patient had a complete atrioventricular block and required a temporary pacemaker. However, the conduit was small and the patient needed extracorporeal membrane oxygenation support and later required conduit replacement of 8mm. However, the patient had a nosocomial infection suffered septic shock, and died with severe hemodynamic compromise. According to the postoperative data of this study, there were a significant number of patients who suffered from infection with 9 patients (24.3%) had pneumonia, and 3 patients (8.1%) had septic shock. As long as the patients required prolonged postoperative mechanical ventilation (mean ventilation time of 3.8 days), the risk of infection increased and the outcomes might be negatively influenced. However, our midterm results are comparable with previous studies (3)(4)(7)(8), but there is still tremendous opportunity to enhance survival outcomes following surgery.

Including the patient who required early reoperation of conduit replacement has been previously described, there was one more patient who required reoperation with residual partial anomalous right pulmonary vein connection, which produced the reoperation rate of 5.4%. During a follow-up of 62 months, there was no mortality and all patients completed the follow-up with NYHA class I.

CONCLUSIONS

The outcomes of surgical repair for APW are good and can be improved with better infection control. The midterm survival beyond discharge of simple APW is excellent.

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Table 1: Patients demographic

Preoperative variables	**n (%) or mean (SD)**
Age (days)	259.37 ± 702.59
Weight (kg)	5.04 ± 3.7
BSA (m^2)	0.28 ± 0.13
Prematurity	7 (18.9%)
Respiratory distress	14 (37.8%)
Mechanical ventilation	10 (27%)
Pneumonia	8 (21.6%)
Malnutrition	29 (78.4%)
Liver dysfunction	4 (10.8%)
Renal dysfunction	5 (13.5%)
Cardiogenic shock	1 (2.7%)
APW type	
Type1	16 (43.2%)
Type2	12 (32.4%)
Type3	9 (24.3%)
APW diameter (mm)	10 ± 3.92

Table 2: Perioperative variables

Variables	n (%) or mean (SD)
Procedures for APW repair	
Patch closure	32 (86.5%)
APW division with direct suture	3 (8.1%)
APW ligation	2 (5.4%)
Bypass time (minutes)	77.6 ± 50.4
Aortic cross clamp time (minutes)	49.7 ± 25.7
PDA ligation	11 (29.7%)
Sternum open	2 (5.4%)
ECMO	2 (5.4%)
Low cardiac output syndrome	12 (32.4%)
Pneumonia	9 (24.3%)
Sepsis	3 (8.1%)
Ventilation time (hours)	90.5 ± 123.7
ICU stay time (days)	49.7 ± 80.7
High frequency oxygenation ventilation	2 (5.4%)
Pulmonary hypertensive cirsis	5 (13.5%)
Reoperation	2 (5.4%)
Early death	4 (10.8%)

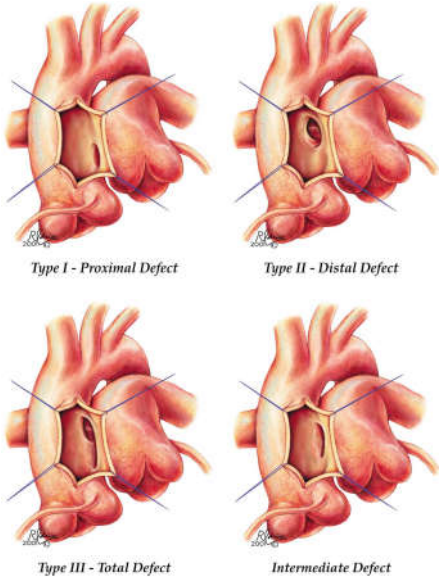

Figure 1: Type of APW [illustration by Idriss (4)] according to STS Congenital Heart Database Committee classification

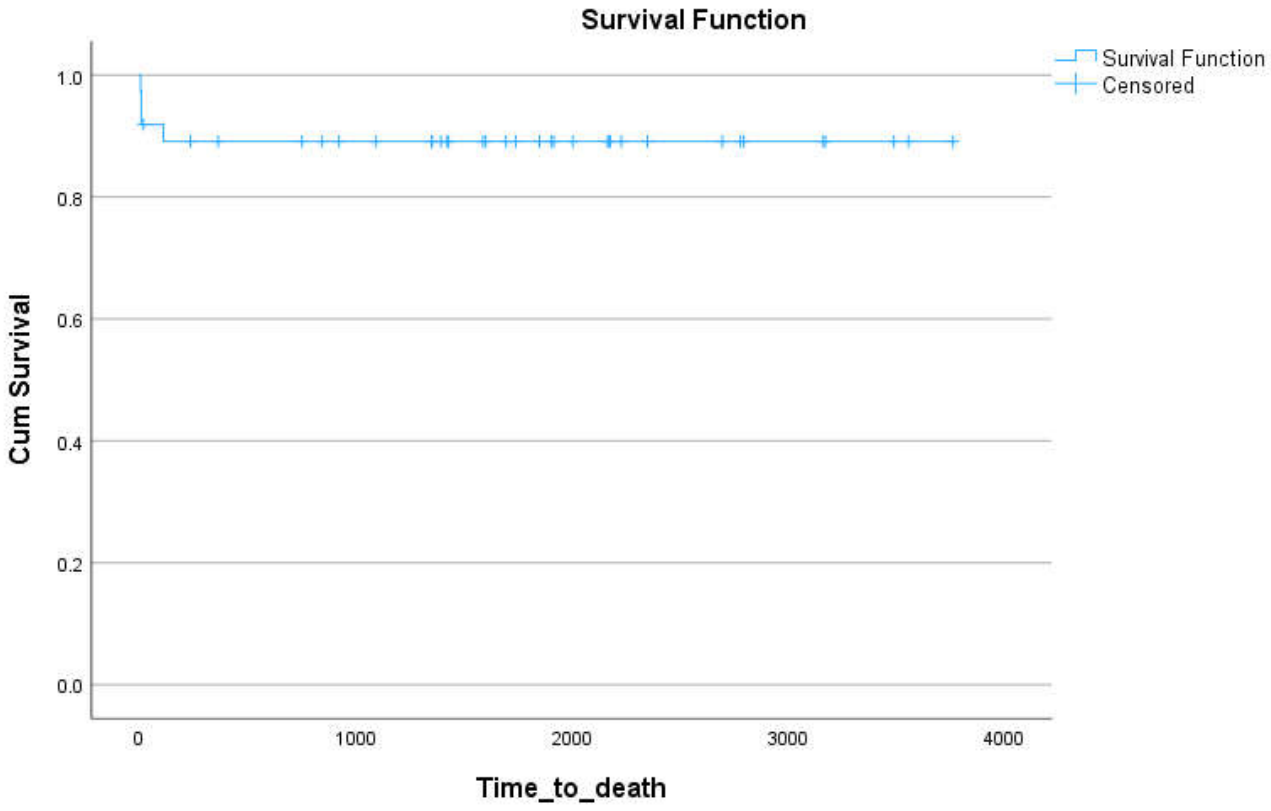

Figure 2: Kaplan-Meier survival of simple APW repair