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Complex TAPVC-Experience with Six Patients*

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ABSTRACT

Total anomalous pulmonary venous connection becomes a totally different subset when associated with complex congenital anomalies. The combination of two separate life-threatening congenital heart defects complicates the management of these patients. Six patients with total anomalous pulmonary venous connection associated with complex congenital heart disease were studied. There were 2 girls and 4 boys. Three of them were less than 5 kg in weight, and the other 3 were more than 5 kg in weight. Four patients had severe pulmonary arterial hypertension and 2 patients had pulmonary stenosis. Three patients had supracardiac type with a right vertical vein, one had drainage to the right atrium superior vena cava junction, one patient had supra cardiac type but split flow to both the superior vena cava and one patient had cardiac type. Three patients had double outlet right ventricle. Three patients had atrioventricular canal defect and 2 patients had preoperative pulmonary vein obstruction. All patients underwent rerouting of pulmonary veins. Concomitant procedures included intraventricular tunnel repair of ventricular septal defect and infundibular resection in double outlet right ventricle. Atrioventricular canal repair was done for Rastelli type A atrioventricular canal. Superior vena caval plasty, atrioventricular canal repair and pulmonary artery banding were done in unbalanced atrioventricular septal defect and large double outlet right ventricle. Intracardiac repair through transatrial approach was done for tetralogy of Fallot. Right ventricle-pulmonary artery conduit was done for truncus arteriosus. Single ventricle repair was done for corrected transposition of great arteries. There were 2 hospital deaths.

Keywords: Total Anomalous; AV Canal; Complex; Congenital; Pulmonary Vein

1. Introduction

Total anomalous pulmonary venous connection is characterized by the failed union of the pulmonary veins and incorporation of it by the developing left atrium in combination with a persistent embryologic connection between the pulmonary and systemic venous systems. The impact of the pathology depends on the degree to which the pulmonary venous drainage is obstructed and the magnitude of the left-to-right shunt. Accurate assessment of the anatomy and detailed surgical planning become essential when total anomalous pulmonary venous connection is associated with other complex congenital anomalies. This result in a combination of two separate life-threatening heart defects needs appropriate management. Most often the timing of surgery is dictated by the presence or development of pulmonary venous obstruction rather than the standardized protocols that apply to the majority of patients. It is our goal that this analysis will identify some important principles in the management of these complex patients.

2. Patients and Methods

Six patients with total anomalous pulmonary venous connection associated with complex congenital heart disease were diagnosed in our hospital between 2011 and 2013. Symptoms included delayed milestones, breathlessness, cyanosis, excessive sweating, poor weight gain and recurrent lower respiratory infections. There were 2 girls and 4 boys. Three of them were less than 5 kg in weight, and the other 3 were more than 5 kg in weight. Four patients had severe pulmonary arterial hypertension and 2 patients had pulmonary stenosis. Three patients had supracardiac total anomalous pulmonary venous connection with a right vertical vein, one had drainage to

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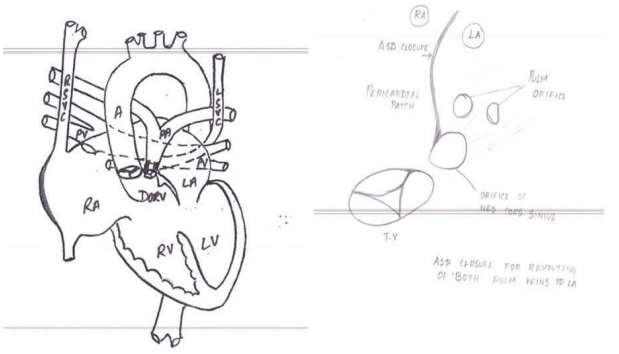
the right atrium superior vena cava junction, one patient had supra cardiac total anomalous pulmonary venous connection but split flow to both the superior vena cava and one patient had cardiac total anomalous pulmonary venous connection. Three patients had double outlet right ventricle. Three patients had atrioventricular canal defect and 2 patients had preoperative pulmonary vein obstruction. Patients' summary is given in **Table 1**.

All patients underwent rerouting of total anomalous pulmonary venous connection. All operations were performed under general anesthesia through a median sternotomy. Cardiopulmonary bypass was established with aortic and bicaval cannulation. Myocardial protection included cold blood cardioplegia. Deep hypothermic circulatory arrest was required in one patient. All patients were cooled to 28^0 celsius. For all types of total anomalous pulmonary venous connection, the anastomosis between the common pulmonary venous chamber and the atrium was performed by continuous suture with 7-0 polypropylene. In addition, the area of the anastomotic orifice was made as large as possible. Vertical vein was ligated and patent foramen ovale was left open in all cases.

Concomitant procedures included intraventricular tunnel repair of ventricular septal defect and infundibular resection in double outlet right ventricle with total anomalous pulmonary venous connection. Atrioventricular canal repair was done for Rastelli type A atrioventricular canal defect with total anomalous pulmonary venous connection. Superior vena caval plasty, atrioventricular canal repair and pulmonary artery banding was done in unbalanced atrioventricular septal defect and large double outlet right ventricle in which pulmonary vein was obstructed at the superior vena caval junction. Intracardiac repair through transatrial approach was done for tetralogy of Fallot with total anomalous pulmonary venous connection. Right ventricle-pulmonary artery conduit was constructed for truncus arteriosus with total anomalous pulmonary venous connection. Single ventricle repair was done for corrected transposition of great

Table 1. Patient summary.

Patient	Age	Sex	Weight in Kgs	Echocardiogram	Operation
1	2 Years	M	8	Situs solitus, double outlet right ventricle with large subaortic perimembranous ventricular septal defect, moderate pulmonary valve stenosis, unobstructed supracardiac TAPVC to right atrium superior vena cava junction, bilateral superior vena cava with left superior vena cava draining to unroofed coronary sinus. (**Figures 1** & **2**)	Intraventricular tunnel repair of the ventricular septal defect, Infundibular resection, Rerouting of TAPVC, Rerouting of left superior vena cava to right atrium, Pericardial patch closure of atrial septal defect.
2	9 Months	F	6	Situs ambiguous, right atrial isomerism, dextrocardia, common atrium, complete atrioventricular septal defect Rastelli type A, bilateral superior vena cava, supra cardiac TAPVC, 2 veins entering each of the superior vena cava, small patent ductus arteriosus, severe pulmonary hypertension. (**Figure 3**)	Rerouting of TAPVC, Atrioventricular canal repair. (Patient died on fourth postoperative day)
3	9 Months	M	4.5	Situs ambiguous right isomerism, partially obstructed right sided supra cardiac TAPVC to right superior vena cava, unbalanced atrioventricular septal defect with regurgitation with small left ventricle, double outlet right ventricle with severe pulmonary artery hypertension. (**Figures 4** & **5**)	Rerouting of TAPVC, Right superior vena caval plasty, Atrioventricular valve repair, Pulmonary artery banding.
4	6 Months	F	4.5	Situs solitus, tetralogy of Fallot with TAPVC draining to the coronary sinus. left superior vena cava draining to coronary sinus. (**Figure 6**)	Rerouting of pulmonary veins into left atrium, Intracardiac repair through transatrial approach, Rerouting of left superior vena cava to right atrium.
5	14 Years	M	30	Situs ambiguous, levocardia, right atrial isomerism, left superior vena cava to the roof of the left sided atrium, Unbalanced atrioventricular septal defect with rudimentary left ventricle, double outlet right ventricle with malposed great arteries, severe pulmonary valvular stenosis, unobstructed right sided supra cardiac TAPVC to right brachiocephalic vein. (**Figures 7** & **8**)	Rerouting of TAPVC with single stage Fontan surgery.
6	Newborn	M	3.2	Situs solitus, Type III-truncusarteriosus, right and left pulmonary artery arising separately from the common trunk, subtruncal ventricular septal defect, ostiumsecundum atrial septaldefect, partially obstructed right sided supracardiac TAPVC draining to right brachio cephalic vein, severe pulmonary arterial hypertension. (**Figure 9**)	Rerouting of TAPVC, Truncus repair with right ventricle-pulmonary artery conduit, Ventricular septal defect rerouted to aorta, Atrial septal defect closure. (Patient died on third postoperative day)



RSVC-Right superior vena cava LSVC-Left superior vena cava A-Aorta
PV-Pulmonary vein PA-Pulmonary artery RA-Right atrium RV-Right ventricle
LA-Left atrium LV-Left ventricle ASD-Atrial septal defect

Figure 1. Patient no one showing double outlet right ventricle with large subaortic perimembranous ventricular septal defect, moderate pulmonary valve stenosis with TAPVC. Procedure done is also shown.

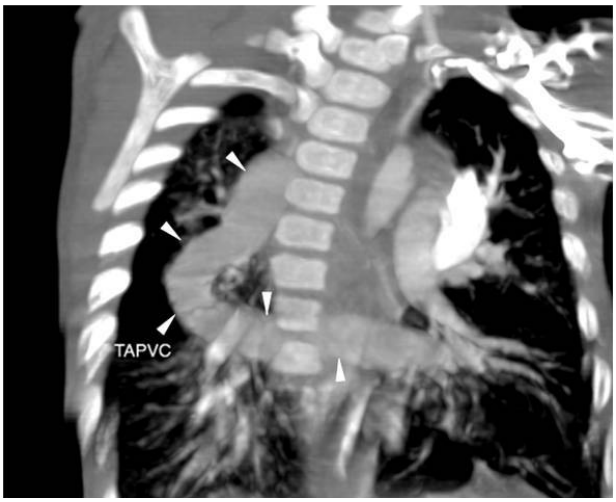


Figure 2. MR Angiogram of patient no one showing TAPVC.

R-Right superior vena cava
L-Left superior vena cava
P-Patent ductus arteriosus
A-Aorta
PV-Pulmonary vein
PA-Pulmonary artery
CA-Common atrium
RV-Right ventricle
LV-Left ventricle

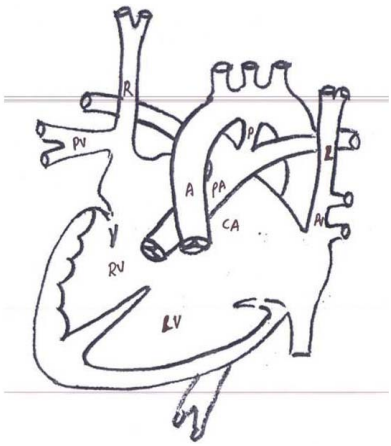
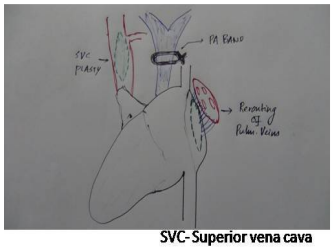
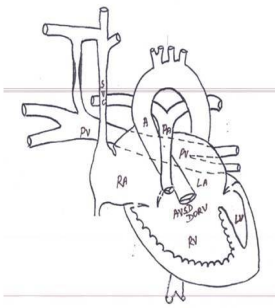


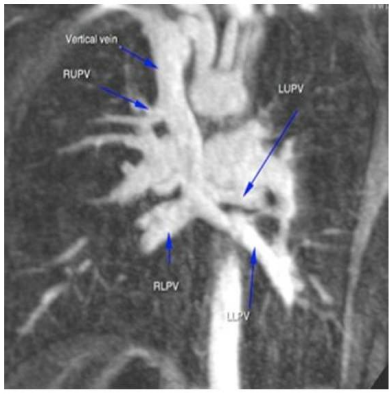
Figure 3. Patient no 2 showing dextrocardia, atrioventricular septal defect, patent ductus arteriosus, common atrium and TAPVC.





SVC- Superior vena cava
PV-Pulmonary vein
PA-Pulmonary artery
A-Aorta
RA-Right atrium
RV-Right ventrice
LV-Left ventricle
A-Left atrium
AVSD-Atrioventricular septal defect ,
DORV-Double outlet right ventricle

Figure 4. Patient no three showing unbalanced atrioventricular septal defect, double outlet right ventricle and obstructed TAPVC. Procedure done is also shown.



RUPV-Right upper pulmonary vein
LUPV-Left upper pulmonary vein
RLPV-Right lower pulmonary vein
LLPV-Left lower pulmonary vein

Figure 5. CT angiogram of patient no 3 showing all four pulmonary veins and vertical vein.

RSVC-Right superior vena cava
LSVC-Left superior vena cava
A-Aorta
PV-Pulmonary vein
PA-Pulmonary artery
RA-Right atrium
LA-Left atrium
RV –Right ventricle
LV-Left ventricle

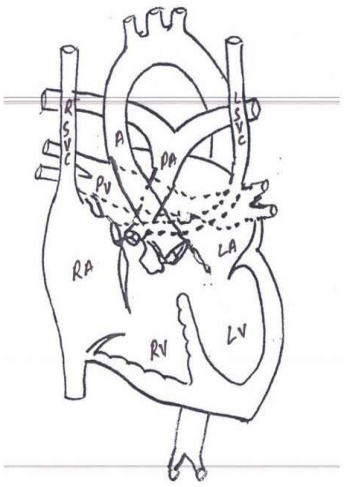


Figure 6. Patient no four showing tetralogy of Fallot with TAPVC.

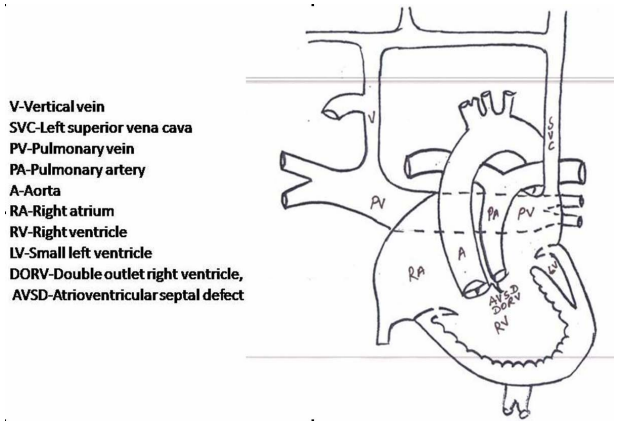


Figure 7. Patient no five showing double outlet right ventricle, atrioventricular septal defect, malposed great arteries and TAPVC.

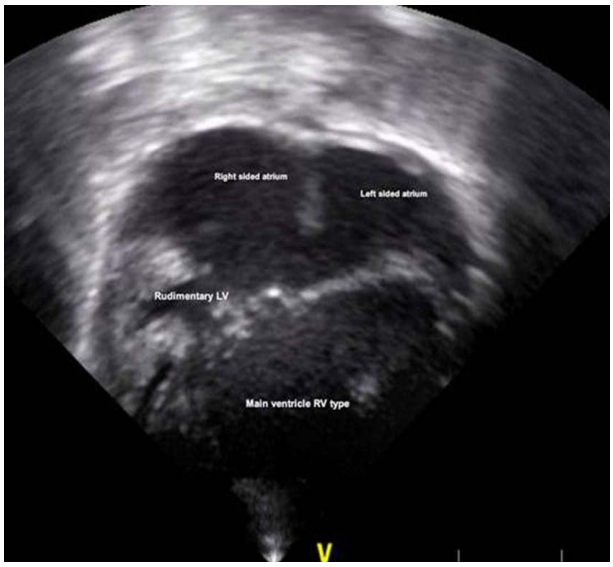


Figure 8. Echocardiogram of patient no five showing unbalanced atrioventricular septal defect with rudimentary left ventricle.

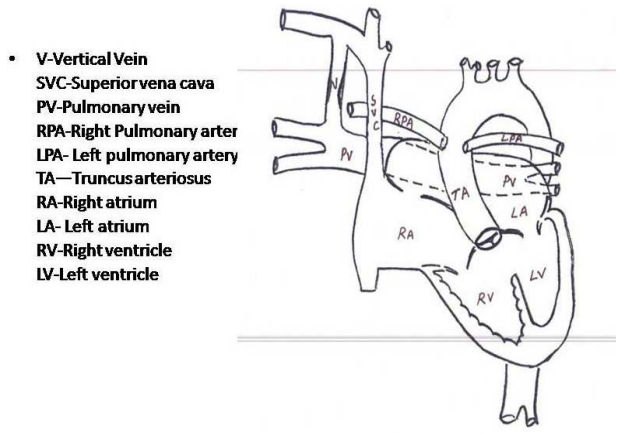


Figure 9. Patient no six showing truncus arteriosus with TAPVC.

arteries with total anomalous pulmonary venous connection. There were 2 hospital deaths. Cause of death in both the patients was pulmonary hypertensive crises.

3. Discussion

Complex heart defects involving venous, intracardiac and arterial pathways can be successfully repaired in a single stage. Complete repair of the very rare association of complex congenital anomalies like truncus arteriosus, tetralogy of Fallot, complete atrioventricular canal and corrected transposition of great arteries with total anomalous pulmonary venous connection is possible and customized technical solutions are often necessary to achieve good results [1]. Untreated, the prognosis for this patients is rather bleak, with half of the patients succumbing within 1st year of life [2,3]. The relative efficacy of surgical treatment was a point of debate for several decades. The subsequent standardization of protocols at many centers throughout the world has improved the outlook for these patients with multiple congenital heart defects substantially [4].

When total anomalous pulmonary venous connection is associated with other complex heart defects the risk for early postoperative death phenomenally increases. Aggressive postoperative management after a very meticulously planned corrective/palliative surgery is needed to improve the outcome. The 10-year survival rate in heterotaxy syndrome has been reported as 39% with total anomalous pulmonary venous connection and 64% without total anomalous pulmonary venous connection [5]. A report of 377 patients undergoing total anomalous pulmonary venous connection repair found that surgical outcomes have improved over time due to improvements in surgical techniques and perioperative management [6]. The 5-year survival rate after total anomalous pulmonary venous connection repair in the biventricular heart has improved to 97% since 2000. However, surgical results for complex congenital anomalies associated with total anomalous pulmonary venous connection have not yet been satisfactory, with a reported 3-year survival rate of only 47% [7].

Total anomalous pulmonary venous connection is usually associated with right isomerism in 30%, other heart anomalies in 20% and is isolated in more than 50%. Congenital anomalies associated with total anomalous pulmonary venous connection include viscerocardiac situs abnormality, aortocaval juxtaposition, atrioventricular septal defect, common atrium, double-outlet right ventricle, dextro-malposed great arteries, pulmonary stenosis or atresia, coarctation of aorta, hypoplastic left heart, parachute mitral valve and straddling mitral valve. The diagnosis of anomalous pulmonary venous connections in the setting of heterotaxy should be actively considered, especially in neonates with right isomerism [8].

In this series of six patients total anomalous pulmonary venous connection was associated with tetralogy of Fallot, truncus arteriosus, unbalanced AV canal, corrected transposition of great arteries, double outlet right ventricle and complete AV canal with dextocardia.

The two outcome factors to be considered are mortality and postoperative recurrent pulmonary vein obstruction. Recent reports about complex congenital heart disease and total anomalous pulmonary venous connection have indicated an improvement in the outcomes over time. They showed that survival at 5 years after total anomalous pulmonary venous connection repair was 79% and the risk factors for mortality were younger age, pulmonary atresia, preoperative obstructed total anomalous pulmonary venous connection, body weight less than 3.5 kg at operation, concomitant systemic pulmonary shunt, postoperative pulmonary vein obstruction and infracardiac or mixed total anomalous pulmonary venous connection [9].

Body weight and age at operation was identified as a risk factor for death. Of 226 patients with repaired total anomalous pulmonary venous connection in the Society of Thoracic Surgeons database, the surgical outcome in body weight ranging from 1 to 2.5 kg at operation was worse than that in body weight ranging from 2.5 to 4 kg [10]. Increased body weight at operation may also reduce perioperative complications such as intracranial hemorrhage, renal dysfunction, and coagulopathy [10]. In our series three patients weighed above 5 kgs and three patients weighed less than 5 kgs. Both the child whom died was less than 5 kgs. However, waiting for an increase in body weight before operation may not be advantageous due to an increase in preoperative morbidities [11]. Age at operation tends to affect early surgical outcomes [11]. However in our series all the children were slightly older except the truncus arteriosus patient who was referred to us in the newborn period.

Nonconfluent pulmonary arteries and pulmonary atresia were identified as a risk factor for death in patients operated for total anomalous pulmonary venous connection [12]. Because of the varied anatomic variations in the total anomalous pulmonary venous connection anatomy, especially when associated with complex congenital heart malformations, accurate preoperative diagnosis and determination of the configuration of the pulmonary vein morphology are essential for planning on surgical treatment of these challenging patients; else the postoperative course could be complicated [13]. Jenkins *et al.* reported that small pulmonary vein confluence was a strong negative predictor of survival in patients with total anomalous pulmonary venous connection including those with two-ventricle anatomy. Generally, mixed total anomalous pulmonary venous connection has smaller pulmonary vein confluence(s) and this may contribute to its worse prognosis [14]. Most of the patients in our series had good pulmonary vein confluence.

It is important to have a regulated pulmonary blood flow in total anomalous pulmonary venous connection. However patients requiring manipulation of pulmonary blood flow are difficult to manage. Concomitant systemic pulmonary shunt or pulmonary artery banding was identified as an added risk factor for hospital death. In the management of complex cardiac abnormalities, it is essential to have an optimal pressure-volume relationship in the heart (preload and after load) to gain better quality of life [15]. It may be difficult to sustain an adequate circulation in patients who have had total anomalous pulmonary venous connection repair with concomitant systemic pulmonary shunt, especially in patients diagnosed with preoperative pulmonary vein obstruction and functional univentricle physiology with right ventricular morphology. We consider that intensive management is important to adjust the pulmonary blood flow in these complex patients. Among our patients infundibular resection was done in two patients in two patients and PA banding was done in one patient. Vertical vein if present was ligated and patent foramen ovale was left open in all patients.

Atrioventricular valve regurgitation, which required surgical intervention, was a significant risk factor for postoperative recurrent pulmonary vein obstruction. Volume unloading after the atrioventricular valve repair reduces the size of the atrium and may stretch or distort the pulmonary vein(s) [9]. Atrioventricular regurgitation is associated with systemic ventricular dysfunction and subsequent pulmonary hypertension. Surgical outcomes are therefore influenced by the ability to repair atrioventricular valve regurgitation at total anomalous pulmonary venous connection repair [15]. We had 2 patients with atrioventricular valve regurgitation. In both the patients repair was done successfully, however one died. This patient continued to have persistent pulmonary hypertension despite a satisfactory anatomic repair. This patient eventually died from a combination of hypoxemia and respiratory failure.

With newer surgical techniques and better management strategies, the outcomes of patients with total anomalous pulmonary venous connection and associated congenital anomalies have improved dramatically. However, when complex congenital cardiac malformations exist in association with total anomalous pulmonary venous connection, it still remains a clinical challenge especially if it was obstructed [9]. There is a paucity of knowledge about this subset of patients. We had 2 cases of preoperative obstructed total anomalous pulmonary venous connection of which one child died. Both the cases were obstructed at the junction with the superior vena cava.

The repair of total anomalous pulmonary venous connection was performed by a single surgeon, and conformed to the technical principles of appropriate (not excessive) dissection of the pulmonary vein and minimal manipulation, taking small bites with nonabsorbable sutures and cutting off the pectinate muscle which is proper to the single right atrium of RAI, if necessary, to minimize the thickness of the anastomosis and precise anastomosis under short and intermittent circulatory arrest, if necessary. Surgical approaches such as primary sutureless repair reported by Yun and colleagues may be indicated for patients with mixed total anomalous pulmonary venous connection, especially when obstructed. However, it is not clear in the literature if primary sutureless repair will have any significant impact on the prognosis of mixed total anomalous pulmonary venous connection. The practice of cutting off the pectinate muscle is based on the theory that the thickness of the anastomosis contributes to the occurrence of anastomotic stenosis and recurrent pulmonary vein obstruction [16].

Univentricle or heterotaxy syndrome has been identified as a risk factor for reoperation in repaired pulmonary vein stenosis. The study found that 79% of reoperations for pulmonary vein stenosis occurred within 6 months of the initial repair [17]. Another report of 20 patients identified that postoperative pulmonary vein obstruction occurring within 6 months of the operation was a risk factor for death [18]. Residual pulmonary vein obstruction might cause subsequent pulmonary hypertension, with even worse outcomes [19]. Therefore, postoperative pulmonary vein obstruction should be repaired as soon as possible, before the pulmonary hypertension becomes persistent. Another report found that high resistance of the pulmonary vasculature, caused by postoperative pulmonary vein obstruction, was the main factor influencing death in 14 autopsies of patients diagnosed with univentricle and total anomalous pulmonary venous connection [20].

We think that pulmonary vein wall cannot be normalized immediately and that the various complications might occur due to the poor general condition of the patient, even if the pulmonary vein obstruction is relieved. Recurrent pulmonary vein obstruction occurs due to several intrinsic factors and compression from the adjacent structures like the aorta, atrium, bronchus, or vertebrae. Conventional total anomalous pulmonary venous connection repair may be associated with later intimal hyperplasia localized to the anastomotic site. Improvement or modification of surgical repair technique may improve the results. For example, a sutureless technique that avoids trauma to the pulmonary vein wall and minimizes the risk of distortion at the anastomosis has been reported [20]. This technique may be able to prevent postoperative pulmonary vein obstruction, although long-term surgical outcomes are unclear. More than the type of total anomalous pulmonary venous connection it is the postoperative pulmonary vein obstruction that worsens the prognosis. With our technical principles, none of the patients developed anastomotic stenosis.

4. Conclusion

Improvement or modification of the primary repair technique is required to avoid postoperative pulmonary vein obstruction, such as using sutureless technique, especially in patients of whom total anomalous pulmonary venous connection is associated with complex congenital cardiac anomalies. Prevention of postoperative pulmonary vein obstruction is important in this complex group of patients. Postoperative pulmonary vein obstruction should be repaired as early as possible, before the pulmonary hypertension becomes persistent. Careful postoperative management is required to control the pulmonary blood flow, especially with a concomitant systemic pulmonary shunt or pulmonary artery banding. Significant atrioventricular regurgitation should be addressed to improve postoperative morbidity and mortality. Total anomalous pulmonary venous connection associated with complex heart disease can be successfully repaired in a single stage. We therefore consider that complete evaluation of the pulmonary venous anatomy, corrective surgery at the appropriate time, aggressive perioperative management and close follow-up after hospital discharge are essential to improve the surgical outcome when total anomalous pulmonary venous connection is associated with complex congenital cardiac anomaly.

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