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Original Article

Surgical management of anomalous pulmonary venous connection to the superior vena cava - early results

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ABSTRACT

Background: The anatomical variability in patients with anomalous pulmonary venous connection to superior vena cava presents a surgical challenge. The problem is further compounded by the common occurrence of postoperative complications like arrhythmias and obstruction of the superior vena cava or pulmonary veins. We present our experience of managing this subset using the two patch and Warden's techniques.

Patients and methods: Between June 2011 and September 2012, 7 patients with APVC to the SVC were operated in our institute. After delineating the anatomy, five of them had a two patch repair and two were managed with Warden's technique.

Results: There was no in-hospital mortality or early mortality over a mean follow-up of 9.66 ± 3.88 months (range 6–15 months). All the patients on follow-up had unobstructed pulmonary venous and SVC drainage on echocardiography and all of them were in normal sinus rhythm.

Conclusions: Anomalous pulmonary venous connection to superior vena cava is a challenging subset of patients in whom the surgical management needs to be individualized. The detailed anatomy must be delineated using echocardiography with or without CT angiography before deciding the surgical plan. This entity can be repaired with excellent immediate and early results. However, these patients must be closely followed up for complications like systemic and pulmonary venous obstruction and sinus node dysfunction.

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1. Introduction

Anomalous pulmonary venous connection (APVC) to superior vena cava (SVC) with its anatomical heterogeneity mandates an individualized approach. The commoner variant of APVC is partial anomalous pulmonary venous connection (PAPVC) to superior vena cava (SVC), which can be repaired with standard operative procedures. However', repair of total anomalous pulmonary venous connection (TAPVC) to SVC is challenging. The problem is further compounded by the common occurrence of postoperative complications like arrhythmias and obstruction of the SVC or pulmonary veins.[1–3]

The variants that are difficult to repair include drainage of all of the pulmonary veins directly to the SVC without a pulmonary venous confluence, drainage of one or more pulmonary veins to the high SVC (near the innominate vein), and drainage of one or more pulmonary veins to a small right SVC (usually in association with a persistent left SVC) without a bridging vein.[1]

We herein present early results of surgical management of APVC to SVC at our institute.

2. Patients and methods

Between June 2011 and September 2012, 7 patients with APVC to the SVC were operated in our institute. There were two female and five male patients, with age ranging from 1 year to 21 years (mean 9.71 ± 8.78 years) (Table 1). TAPVC to SVC without posterior pulmonary venous confluence was present in 3 patients (Fig. 1) and PAPVC to SVC was present in the rest. The associated defects included tricuspid regurgitation, atrial septal defect (ASD) [ostium secundum (OS) and sinus venosus (SV) type] and left SVC. All patients underwent trans thoracic echocardiography for assessment of the anatomy. Cardiac catheterization was undertaken in three cases with pulmonary arterial hypertension (PAH). Cardiac Computer Tomography angiography was done for one patient to further delineate the anatomy (patient no. 1).

All surgeries were performed via median sternotomy approach. After vertical pericardiotomy the anatomy was assessed. SVC and innominate vein were fully mobilized in patients planned for Warden's technique. The SVC cannula was placed at the junction of the innominate vein and SVC, and the inferior vena cava cannula was placed at the right atrial-inferior vena cava junction.

All procedures were performed using mild hypothermic cardiopulmonary bypass. Cardioplegic arrest was achieved with cold blood cardioplegia. All patients having PAPVC with SV ASD were repaired using two patch technique. The adult patient (patient no. 1) with TAPVC to SVC with an OS ASD was repaired using the modified two patch technique. In this patient the patch was fashioned from a Dacron tube graft to ensure unobstructed pulmonary venous drainage. The remaining two patients with TAPVC to SVC were repaired with Warden's technique.

3. Results

There was no in-hospital mortality or early mortality over a mean follow-up of 9.66 ± 3.88 months (range 6–15 months). The duration of the cardiopulmonary bypass and aortic cross clamping for Warden's Technique was 181 min and 104 min respectively and for two patch technique was 114.2 min and 64.6 min respectively (Table 2). Mean ICU stay was 3 ± 1.15 days and hospital stay was 10.14 ± 2.79 days. We routinely performed trans-thoracic echocardiography on day one and before discharge. There was no residual ASD. All patients received inodilators (Dobutamine and Milirinone) in the immediate postoperative period. The patients were later continued on ACE inhibitors and diuretics. On follow-up echocardiography unobstructed pulmonary venous and SVC drainage was present in all patients. None of the patients in our group had any postoperative rhythm problems and all of them were in sinus rhythm at discharge (documented by serial ECG). All patients remained in sinus rhythm on follow up evaluation with 12 lead ECG. Two patients (patient nos. 2 & 3) had RV dysfunction in the postoperative period which recovered with conservative management. RV dysfunction was probably due to moderate pulmonary artery hypertension present preoperatively. One patient (patient no. 2) required prolonged ventilation postoperatively due to lower respiratory tract infection (Table 2).

4. Discussion

The anatomical variability in patients with APVC to the SVC presents a surgical challenge. Furthermore the repair is complicated by the occurrence of obstruction of the SVC or

Table 1 – Summary of patient data.

Patient number	Sex	Age (years)	Pulmonary vein	Left SVC	ASD	PAH
1	Female	14	TAPVC to SVC	No	OS ASD	Mild
2	Male	2	TAPVC to SVC	Yes	SV ASD	Moderate
3	Male	1	TAPVC to SVC	No	OS ASD	Moderate
4	Male	5	PAPVC to SVC	Yes	OS ASD	No
5	Male	21	PAPVC to SVC	Yes	SV ASD	No
6	Male	21	PAPVC to SVC	No	SV ASD	No
7	Female	4	PAPVC to SVC	No	SV ASD	No

TAPVC = total anomalous pulmonary venous connection, PAPVC = partial anomalous pulmonary venous connection SVC = superior vena cava, OS ASD = ostium secundum atrial septal defect, SV ASD = sinus venosus atrial septal defect, PAH = pulmonary arterial hypertension.

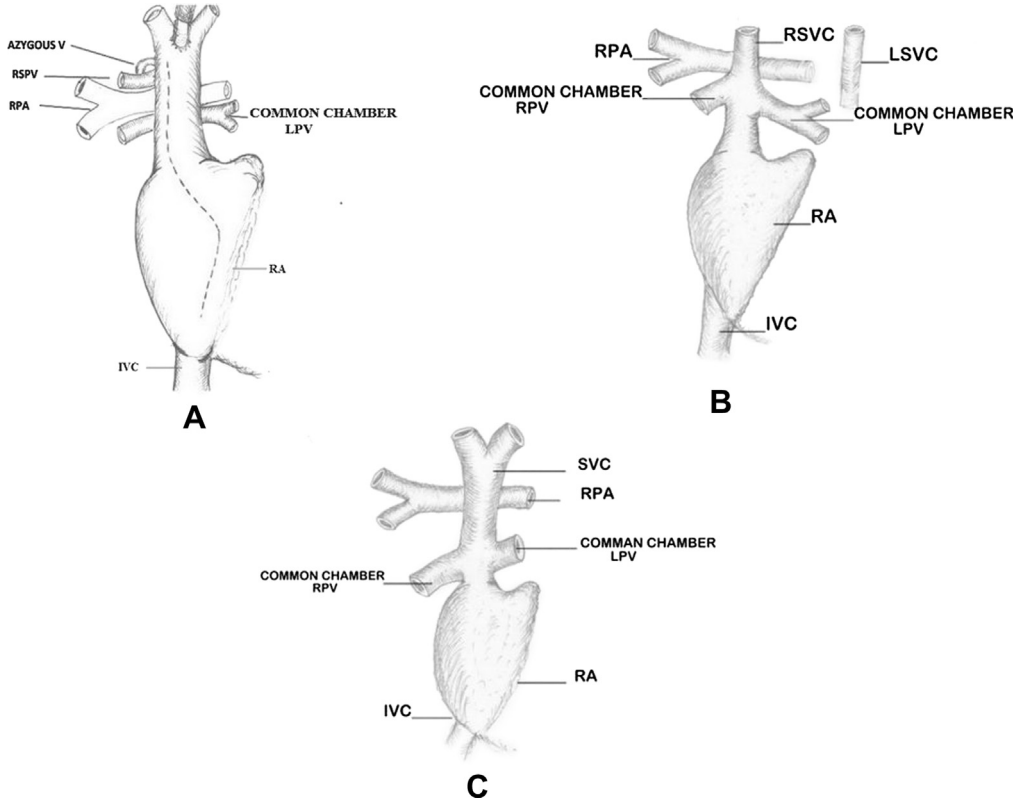

Fig. 1 – Anatomy of TAPVC to SVC in our group. A) Patient no. 1 showing separate opening of RPV and LPV higher up in SVC near RPA, on posterolateral aspect. B) Patient no. 2 showing separate opening of RPV and LPV in small Right SVC higher up in SVC near RPA and left SVC draining in coronary sinus. C) Patient no. 3 showing RPV and LPV in SVC near SVC RA junction. RSPV–right superior pulmonary vein, RIPV–right inferior pulmonary vein, RPV–right pulmonary vein, LPV–left pulmonary vein, RPA–right pulmonary artery, RSVC–right superior vena cava, LSVC–left superior vena cava, IVC–inferior vena cava, RA–right atrium.

pulmonary veins and postoperative arrhythmias.[1–5] Surgical repair for PAPVC to SVC includes closure of the atrial septal defect and redirection of the anomalous pulmonary veins into the left atrium. Care must be taken to avoid pulmonary venous or SVC obstruction and injury to the sinus node and its blood supply.[6]

However, application of these techniques to complex variants of APVC to SVC can result in an increased incidence of postoperative complications, and some forms are difficult to correct using standard methods.[7–10]

The variants which are challenging to repair include those that have all the pulmonary veins draining directly into the SVC without a pulmonary venous confluence and those where one or more pulmonary veins drain into the high SVC near the innominate vein.[1] Patients with TAPVC to SVC, especially with one of the pulmonary veins draining

Table 2 – Summary of management and outcome.

Patient number	Surgery performed	Post-operative rhythm	ACC time (min)	CPB time (min)	Complication
1	Two patch technique	NSR	104	148	No
2	Warden technique	NSR	125	204	LRTI prolonged ventilation & RV dysfunction
3	Warden technique	NSR	83	158	RV dysfunction
4	Two patch technique	NSR	37	103	No
5	Two patch technique	NSR	114	158	No
6	Two patch technique	NSR	34	81	No
7	Two patch technique	NSR	34	81	No

NSR = normal sinus rhythm, ACC = aortic cross clamp, CPB = cardiopulmonary bypass, LRTI = lower respiratory tract infection, RV = right ventricle.

separately in the high SVC, usually have a large SVC and a common channel which is placed high and cephalad to the left atrium. This anatomy precludes a common channel to the left atrial anastomosis. Furthermore, the presence of OS ASD which is remote to the opening of pulmonary veins makes rerouting difficult.

Single patch technique employs one patch to baffle the pulmonary venous blood to the left atrium from the SVC. It can be associated with SVC narrowing and subsequent SVC obstruction. Rhythm disturbances are known complications of this surgery and are a result of intraoperative damage to the sinoatrial node or its blood supply. There are reports wherein an incision in the SVC or across the cavoatrial junction, without any apparent injury to the node or the SA nodal artery, caused sinus node dysfunction in follow-up period. This may be due to fibrosis in that area.[11]

Double patch technique was introduced by Schuster and colleagues, who used a second patch to augment the SVC–right atrial junction to achieve unobstructed SVC drainage. However, SVC obstruction was later reported even when the SVC–right atrial junction was augmented.[1] This is more so in patients with left SVC and a smaller right SVC.[1]

The incidence of sino-atrial node dysfunction is significant (55%).[12] DeLeon and colleagues[5] demonstrated that an incision at right atrial–SVC junction was associated with higher incidence of sino-atrial node dysfunction. We used double patch technique in all patients with PAPVC to SVC. All our patients had normal sinus rhythm postoperatively and are in sinus rhythm, however, they need further investigation and follow-up to exclude sinus node dysfunction. Iyer and colleagues[13] in their series compared the results of single patch to double patch technique and concluded that double patch technique is reproducible, without any increase in complication like pulmonary venous or SVC stenosis or any rhythm problem.

In 1984, Warden and colleagues[8] reported a new technique for high PAPVC, where the SVC was transected at the point just above the highest draining pulmonary vein, the central end (towards the right atrium) was over sewn and the distal end was anastomosed to the right atrial appendage. Thus the central part of SVC formed a part of neo left atrium. Warden's group[14,15] subsequently reported further clinical experience with this technique. The main advantage of their technique is decreased manipulation of the cavo-atrial junction and avoiding the creation of conduits inside the SVC thus avoiding SA nodal dysfunction. The major concern with this technique is stenosis at the cavo-atrial anastomosis. To avoid cavo-atrial stenosis, the anastomosis must be accomplished without tension. This requires careful and extensive dissection of the brachiocephalic vein and SVC and excising all of the intra-atrial trabeculations.

Other authors[12,15–17] have also reported excellent results using Warden's technique for PAPVC to SVC. The incidence of SVC and pulmonary venous obstruction was low (0.075%) and SA nodal dysfunction was rare (0.01%).

Warden's technique was later successfully employed for repair of TAPVC to SVC (without a common chamber).[17,18]

We used the Warden's technique in two patients. In order to avoid obstruction at the cavo-atrial junction we meticulously divided all the trabeculations in the right atrial appendage and interrupted the anastomotic suture laterally and used two sutures (polypropylene 6-0) to complete the anastomosis in order to avoid purse string effect on the anastomotic segment. Care was taken for adequate mobilization of SVC for a tension free anastomosis.

On follow-up, the gradient across SVC–RAA junction was not significant (no turbulence or gradient more than 2 mmHg) in our series.

Gaynor et al[17] reported use of modified Warden's technique in 3 patients with TAPVC to SVC. They used atrial wall baffle to reroute the blood from the pulmonary veins via the ASD into the left atrium. They reported no mortality or rhythm abnormality over a mean follow-up of 2.3 ± 1.4 years. However one patient developed SVC obstruction by the pericardial patch and needed to be revised in early post-operative period. Kottayil and colleagues[18] also successfully employed Warden's technique in 4 patients with TAPVC to SVC. They also reported no mortality or rhythm disturbances over a median follow-up of 24 months.

One of our patients (patient no.1) had very high insertion of one of the pulmonary veins into the SVC, hence a tension free anastomosis could not be created even after complete mobilization of SVC. Here, we employed modified two patch technique wherein the patch for rerouting the pulmonary venous blood to OS ASD was fashioned from a Dacron tube graft to ensure unobstructed drainage. This technique is suitable for adult patients with anomalous venous drainage to SVC. Pulmonary venous and SVC obstruction is unlikely with this technique. However it is not suitable for children and patients with small right SVC and large left SVC. Furthermore, there are concerns like endothelial ingrowth and thromboembolism in the prosthetic graft. These patients require low dose anticoagulation for 6 weeks and can then be switched to antiplatelet therapy. A similar technique using Dacron patch to reroute TAPVC of mixed type was used by Berdat and colleagues.[19] Their patient was a 3 year old child in whom the right upper and middle lobe pulmonary veins were draining directly into the superior vena cava (SVC), approximately 15 and 20 mm above the superior cavo-atrial junction. Rest of the pulmonary veins formed a pulmonary venous sinus and drained into the right atrium. They resected the ridge between the pulmonary venous sinus junction and the ASD, and the caudal rim of the pulmonary venous sinus junction was sutured directly to the cranial rim of the ASD to create a continuous dorsal wall, thus considerably enlarging the ASD. They used ePTFE patch to form an intra-atrial tunnel, rerouting the blood from the pulmonary venous sinus and the two pulmonary veins connecting to the SVC across the enlarged ASD to the mitral valve.

5. Conclusion

APVC to SVC is a challenging subset of patients in the group of APVC and surgical management needs to be individualized. The detailed anatomy must be delineated using echocardiography and/or CT angiography before deciding the surgical plan. This entity can be repaired with excellent immediate and early results. However, these patients must be closely followed up for complications like systemic and pulmonary venous obstruction.

Conflicts of interest

All authors have none to declare.

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