Partial Anomalous Pulmonary Venous Connection to the Superior Vena Cava

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Background: Surgical correction of partial anomalous pulmonary venous connection to the superior vena cava has been associated with postoperative venous obstruction and sinus node dysfunction. In this paper we describe our current approach and its short-term results. Material and Method: Between April 1999 and January 2000, 5 consecutive patients, ranging from 2 months to 66 years old, underwent corrective operation for partial anomalous pulmonary venous connection to the superior vena cava at Sejong General Hospital and Daegu Catholic University Medical Center. Surgical correction involved diversion of the pulmonary venous drainage to the left atrium using a right atrial flap(2 patients) or prosthetic patch(3 patients) with division of the superior vena cava superior to the entrance site of the pulmonary veins and reimplantation on the right atrial appendage to restore systemic venous drainage. Result: All patients were discharged between postoperative day 9 and 15 without complications. One Russian boy returned to his country, therefore, he was lost to follow-up after discharge. Remaining 4 patients were asymptomatic and in normal regular sinus rhythm at a mean follow-up of 17.75 ± 4.27 months. Follow-up echocardiographic study (range, 12 to 24 months) revealed no incidence of narrowing of the venous pathways or of residual shunt. Conclusion: Our current approach is relatively simple and reproducible in achieving unobstructive pulmonary venous and SVC pathways. By avoiding incision across the cavoatrial junction, surgical injury to the sinus node and its artery may be minimized. The presented surgical technique can be safely and effectively applied to the selected patients.

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Key word: 1. Pulmonary vein, partial anomalous return  
2. Vena cava, superior  
3. Anastomosis, surgical
BACKGROUND

Partial anomalous pulmonary venous connection (PAPVC) to the superior vena cava (SVC) is most commonly associated with either a sinus venosus defect or secundum atrial septal defect (ASD), although the atrial septum may be intact\(^1\). Successful operative repair demands redirection of the anomalous pulmonary veins to the left atrium through the naturally occurring or surgically created interatrial communication with unobstructed pulmonary venous or SVC flow and without injury to the sinus node and its blood supply. A variety of ingenious surgical technique have been suggested to combat major postoperative complications including SVC or pulmonary venous obstruction, sinus node dysfunction, and persistent shunts.

As an alternative method to the conventional approach through the SVC-RA junction, Warden and associates\(^2\) repaired PAPVC to the high SVC (1 cm or more above the cavaatrial junction) by coaptation of the inferior margin of the septal defect to the anterior and lateral margin of the intracardiac SVC orifice, thus directing anomalous pulmonary venous flow through the stump of the SVC and across the septal defect into the left atrium. The SVC was divided above the anomalous veins and SVC-RA flow was reconstituted by the cavoatrial anastomosis. Reported series of similar techniques have documented a low incidence of postoperative complications\(^3\)\(^-\)\(^9\). In this paper we present our surgical technique of modified Warden procedure and its short-term results.

MATERIAL AND METHOD

Five consecutive patients underwent reconstructive operation for PAPVC to the SVC at Sejong General Hospital and Daegu Catholic Medical Center during the ten months between April 1999 and January 2000. Patients with PAPVC to the right atrium and patients with inferior sinus venosus defect was excluded. Their ages ranged from 2 months to 66 years (Table 1).

Preoperative congestive heart failure was present in 2 patients (Patient 3, 5).

Echocardiographic study confirmed the diagnosis in 3 patients (Fig. 1). Patient 4 required MR angiography (Fig. 2). Cardiac catheterization and angiographic study was performed in Patient 5 with moderate pulmonary hypertension (Fig. 3). A sinus venosus defect was present in 2 patients, a sinus venosus defect and a patent foramen ovale in 1 patient, and a secundum ASD in 1 patient. The atrial septum was intact in 1 patient.

The origin of the anomalous pulmonary veins was from the right upper lobe in 3 patients, from the right upper and middle lobes in 1 patient, and from the entire right lung in 1 patient with an intact atrial septum.

A 2 month old infant (Patient 3) in intractable heart failure and bronchiectasis had a large PDA. He had a left SVC entering the coronary sinus presented with a small right SVC.

All patients were in normal sinus rhythm preoperatively.

Operative technique. The median sternotomy was used in all patients. The thymus was resected or divided and dissection of the SVC carried to the innominate vein. The SVC and anomalous pulmonary veins were fully mobilized and azygos vein divided. Arterial perfusion was established through the ascending aorta. One venous cannula was placed in the inferior vena cava directly. Drainage of the SVC was accomplished

<table>
<thead>
<tr>
<th>Table 1. Patients</th>
<th>Patient No.</th>
<th>Age/Sex</th>
<th>Types of interatrial communication</th>
<th>Origin of pulmonary veins</th>
<th>Associated cardiac anomalies</th>
<th>Follow-up(months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8 yr/F</td>
<td>Sinus venosus defect</td>
<td>RUL</td>
<td></td>
<td></td>
<td>24</td>
</tr>
<tr>
<td>2</td>
<td>4 yr/M</td>
<td>Sinus venosus defect, PFO</td>
<td>RUL</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>2 yr/M</td>
<td>Secundum ASD</td>
<td>RUL</td>
<td>FDA, left SVC</td>
<td></td>
<td>17</td>
</tr>
<tr>
<td>4</td>
<td>28 yr/F</td>
<td>Intact atrial septum</td>
<td>RUL, RML, RLL</td>
<td></td>
<td></td>
<td>15</td>
</tr>
<tr>
<td>5</td>
<td>66 yr/F</td>
<td>Sinus venosus defect</td>
<td>RUL, RML</td>
<td></td>
<td></td>
<td>15</td>
</tr>
</tbody>
</table>

\(^a\) A Russian boy came back to his country, so he was lost to follow-up after discharge; PFO, patent foramen ovale; ASD, atrial septal defect; RUL, right upper lobe; RML, right middle lobe; RLL, right lower lobe; FDA, patent ductus arteriosus; SVC, superior vena cava.
through right atrial (Patient 1, 2, 3) or innominate vein (Patient 4, 5) cannulation. In Patient 2, the large PDA was doubly ligated.

After institution of total cardiopulmonary bypass with moderate systemic hypothermia and cardioplegic arrest, an incision was made in the right atrial appendage (Fig. 4A). The right atrial orifice of the SVC were baffled with a right atrial wall flap (Patient 1, 2), Gore-tex membrane (Patient 3, 4) or pericardial patch (Patient 5) and running nonabsorbable polypropylene suture so that the entire SVC and anomalous pulmonary veins drained across the naturally occurring sinus venosus defect (Patient 1, 2, 5) or surgically created interatrial communication (Patient 3, 4) into the left atrium (Fig. 4B). Patients with small secundum ASD (Patient 3) and intact atrial septum (Patient 4)
required complete resection of septum primum and superior limbus partial excision with endothelial suturing prior to intraatrial baffle construction. In Patient 2, the patent foramen ovale was directly closed.

The SVC was transected just above the highest anomalous pulmonary vein, and the proximal end of the cava was oversewn, allowing this part of the SVC to act as a conduit for drainage of the anomalous veins into the left atrium. The tip of the right atrial appendage was divided and all muscular trabeculations inside the appendage were completely excised to increase the length of the appendage and reduce the risk of SVC obstruction(Fig. 4C). The distal end of the cava was anastomosed end to end to the superior aspect of the right atrial appendage with absorbable polyglyconate suture(Fig. 4D). Patients were discontinued from cardiopulmonary bypass in the usual manner.

**RESULT**

Postoperative transesophageal echocardiography(4 patients) and transthoracic echocardiography(1 patient) showed unobstructed redirected pulmonary venous and superior caval flow without shunt(Fig. 5). Patients were discharged between postoperative day 9 and 15 without complications.

Because one patient(Patient 2) from Russia came back to his country, he was lost to follow-up after discharge. Remaining 4 patients are asymptomatic and in sinus rhythm at a mean follow-up of 17.75±4.27 months(range, 15 to 24 months). No patient has had a documented arrhythmia or symptoms suggestive of arrhythmia. Follow-up echocardiography in 4 patients(range, 12 to 24 months) revealed no incidence of narrowing of the venous pathways or of residual shunts. Dynamic chest computed tomography in Patient 5 confirmed free SVC-right atrial flow(Fig. 6).

**DISCUSSION**

A variety of surgical techniques have been suggested for repair of PAPVC to the SVC including simple use of a baffle to redirect the pulmonary venous drainage across an interatrial communication, partitioning of the SVC, and various types of atrioconoplasties. In 1965 Kirklin, Ellis, and Wood reported their procedure, which involves covering both defect and veins with a patch that directs blood flow from the anomalous pulmonary veins through the ASD into the LA. Schuster, Gross, and Colodny recommended a second patch to enlarge the junction between the SVC and the RA to decrease the incidence of postoperative SVC obstruction. Kyger and colleagues described early and late results in 109 patients with sinus venous defects. No instances of obstruction of the SVC were detected clinically. In 6 patients the cavoatrial junction was enlarged with a pericardial patch to avoid obstruction of the SVC. In 8 patients with PAPVC to the high SVC the anomalous veins could not be redirected to the LA(deliberately omitted or ligated in 7 patients with small anomalous veins, and right upper lobectomy in 1 patient with a large anomalous vein). Thirty percent of the patients whose cavoatrial junction was enlarged with a patch developed persistent new postoperative arrhythmias compared with an overall incidence of 14%.
patients with similar operative procedure. Agrawal, Khanna, and Tampe\cite{11} described follow-up study of 44 patients after repair of sinus venous defect with PAPVC to the SVC at a mean age of 13.3 years. Another pericardial patch was used to widen the SVC and SVC-RA junction. Echocardiographic evaluation after 6 months in 25 patients and after 2 years in 11 patients revealed no incidences of venous obstruction or residual shunt. Of 27 patients assessed 2 years after surgery, 2 patients developed sinus node dysfunction in the form of junctional rhythm. One patient required a permanent pacemaker.

As an alternative surgical approach SVC partitioning and various atrio cavaloplasties were advocated to construct unobstructed atrio caval channel. Chartrand et al\cite{12} described the technique of partitioning and enlargement of the SVC in 9 children with encouraging results. The partitioning was done with a longitudinal suture starting above the highest pulmonary vein directing the pulmonary venous flow through the enlarged ASD into the LA. The anterior cavo-a uricular tunnel was enlarged with a RA appendage-SVC angioplasty. Kubota and associates\cite{13} examined mid-term results of their rotation advancement flap method of repair\cite{14} in 11 patients, which consisted of atrial partitioning with a polytetrafluoroethylene sheet, enlargement of the cavo atrial junction by a right atrial flap, and protection of the sinus node by keeping their incision through the anterolateral wall of the atrio caval junction. Follow-up hemodynamic and electrophysiologic studies in 7 patients revealed successfully rerouted pulmonary venous drainage and reconstructed cavo atrial junction without stenosis and without clinically significant atrial pacemaker dysfunction. They pointed out a possibility of injuring the procaval sinus node artery originating from right coronary artery in this type of operative method. Nicholson and colleagues\cite{15} reported lateral transcaval approach in 66 patients(range, 1.5-65 years) with sinus venous syndrome. An incision was made in the lateral SVC wall at the junction with the anomalous pulmonary veins. The pericardial patch was sutured to the medial, superior, and inferior margins of the defect within the SVC. The SVC was closed by using the pericardial patch as a sandwich, thus completing the baffle and closing the defect. Mean follow-up of 4.1 years(range, 1-9 years) for 64 patients revealed no evidence of systemic or pulmonary venous obstruction with either 2-dimensional or Doppler flow studies. Holter monitoring at a mean of 7.3 years postoperatively in 30 patients showed normal sinus node function without sustained atrial arrhythmia.
Repair of anomalous pulmonary venous connection to the SVC using an atrial wall baffle with reimplantation of the SVC on the RA appendage was suggested by Lewis in 1958. Elenbaas, Thilen, and Lawrence described an atrioseptectomy and transplantation of the SVC into the RA appendage. In 1984 Warden and associates reported 15 patients with PAPVC to the high SVC. They redirected the anomalous pulmonary venous flow into the LA through the cardiac end of the SVC by coaptation of the anterior border of the septal defect to the intracardiac orifice of the SVC. The SVC was divided above the anomalous veins and anastomosed to the RA appendage. In 1995 the same group described follow-up (6 months to 30 years) of 40 patients including early experiences.

The atrial septal defect rim was coapted to the intracardiac SVC orifice in 23 patients, and intracardiac baffle was used in 17 patients. One 31-year-old woman died of severe pulmonary hypertension. One symptomatic SVC obstruction required reoperation. Sick sinus syndrome developed late in 1 patient (2.5%). No patient required a pacemaker. Williams et al. used a pericardial patch to baffle the SVC and anomalous veins to the LA. Continuity between the cephalad end of the SVC and the RA was established by direct anastomosis to the RA appendage or by creation of a pedicle conduit of RA appendage, RA free wall, and pericardium. All 6 children were in sinus rhythm without venous obstructions. Vargas and Kreutzer reported repair of total anomalous pulmonary venous connection (TAPVC) to the SVC in 3 children using a J-shaped right atriotomy to create a posterior flap of the RA wall that was sutured to the anterior border of the ASD and around the SVC orifice. The SVC was divided above the pulmonary veins, and SVC-RA continuity was reestablished by direct anastomosis. Gaynor et al. applied this technique to 11 patients with anomalous pulmonary venous connection (TAPVC in 3 patients, PAPVC in 8 patients) to the SVC. Atrial wall baffle was used in 9 patients, and pericardial baffle in 2 patients. One patient developed pulmonary venous obstruction and required reoperation at 3 months postoperatively. No patient had clinical evidence of SVC obstruction, and all were in sinus rhythm.

Many available surgical techniques for a lesion reflect some difficulties of any given operative procedure in achieving satisfactory long-term results. It can be related to the subtle but critical variations in the anatomy of these lesions. Sinus venosus defects are most commonly associated and over 90% of cases of sinus venosus defects have partial anomalous connection of the right pulmonary veins. Partial anomalous venous drainage of any of the pulmonary veins may occur occasionally in isolation either with a completely intact atrial septum or with a patent foramen ovale or secundum ASD. Based on the pathologic and anatomic findings, Van Praagh et al. postulated that in sinus venosus defects, the deficiency was in the wall that normally separates some or all of the right pulmonary veins from the SVC or the right atrium (RA), i.e., the wall between the sinus venosus part of the RA and the common venous part of the left atrium (LA). They also made a distinction between sinus venosus defects of the SVC type (right pulmonary veins unroofed into SVC) and sinus venosus defects of the RA type (right pulmonary veins unroofed into RA).

One of the principal complications after repair of PAPVC to the SVC is obstruction of the SVC or pulmonary veins. The risk of SVC obstruction is increased when a left SVC is present, and the right SVC is smaller than usual. Anomalous pulmonary veins connecting to a high SVC near the innominate vein may present a difficulty to the conventional approach. Sinus node dysfunction and atrial arrhythmia is another significant postoperative complication. The sinus node lies subepicardially in the terminal sulcus; usually in the groove to the right of the crest of the atrial appendage. On occasion the sinus node can extend over the crest of the atrial appendage into the interatrial groove ("horseshoe" position, about 10% of cases). Busquet et al. described the well-established preponderance of origin of the sinus node artery from the right coronary system (66%) as opposed to the left (30%), and infrequent a double supply (4%). Variability was noted in the course of the nodal artery relative to the cavoatrial junction - precaval (58%), retrocaval (36%) or encircling (6%). Impairment of sinus node function may occur either from placement of incision in the sinus node directly or from damage to the sinus node artery. The entire junction of the superior cava and right atrium should be treated as potential danger area. It may partly explain relatively frequent incidence of atrial dyssrhythmias following conventional repair of PAPVC to the SVC. Apart from direct surgical trauma, retraction of sinus node without vascular injury and gradual fibrosis in the area surrounding the sinus node may also contribute to postoperative sinus node dysfunction.

In our experience, our current approach is relatively simple and reproducible in achieving unobstructive pulmonary venous and SVC pathways. From the technical point of view, construction of tension-free cavatral anastomosis is the most
critical part of the procedure. The ayzyos vein is divided and all muscular trabeculations inside the right atrial appendage should be completely excised to increase the length of the appendage and reduce the risk of SVC obstruction. By avoiding incision across the cavoatrial junction, surgical injury to the sinus node and its artery may be minimized. This type of repair also can be applied to the patients with difficult anatomic subset not amenable to the more commonly applied intracaval patch technique. A thorough long-term follow-up including Holter monitoring and electrophysiologic study will be needed.

REFERENCES


=국문초록=

배경: 레정맥 및 상대정맥의 폐쇄와 동방결절의 기능장애 등의 상대정맥으로 환류되는 부분폐정맥 연결이상의 수술교정 이후 발생할 수 있는 합병증으로 실 후 장기적을 결정하는 요인이 되어왔다. 저자들은 시행하고 있는 수술방법을 기술하고 조기결과를 분석하였다. 대상 및 방법: 1999년 4월부터 2000년 1월까지 세 종병원과 대구가톨릭대학병원에서 5명의 환자가 상대정맥으로 환류되는 부분폐정맥 연결이상으로 교정수술을 받았다. 환자의 나이는 생후 2개월부터 66세까지였다. 수술방법은 우선원 피질(2예)이나 첨조(3예)를 사용하여 이상폐정맥을 심방간 통로를 통하여 좌심방으로 정상환류시키고, 상대정맥을 이상폐정맥의 상부에서 절단한 후 근위부를 폐쇄하고 상대정맥의 원위부를 우심방에 단단문합함으로써 상대정맥과 우심방의 확류를 제한하였다. 결과: 모든 환자가 합병증 없이 숨 후 9일과 15일 사이에 완화하였다. 되원 후 본국으로 돌아간 러시아 환아를 제외한 4명의 환자는 평균 17.75±4.27 개월의 추적기간 동안 증상 없이 정상활동을 보였다. 숨후 12개월에서 24개월에 시행한 심장초음파검사상 폐정맥이나 상대정맥의 혈착 및 간헐단막은 보이지 않았다. 결론: 저자들은 상대정맥으로 환류되는 부분폐정맥 연결이상 환자에서 기술한 수술방법으로 폐정맥 및 상대정맥 확류의 혈착 없이 좋은 결과를 얻을 수 있었다. 상대정맥과 우심방결합부위를 가로지르는 절개를 피함으로써 동방결절 및 그 동맥의 손상을 최소화 할 수 있을 것으로 생각한다.

중심 단어: 1. 부분폐정맥 연결이상
2. 상대정맥
3. 상대정맥-우심방 판단문합

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