

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.

(Korean Thorac Cardiovasc Surg 2001;34:672-9)

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

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논문접수일 : 2001년 6월 5일 심사통과일 : 2001년 9월 6일

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본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

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^{1,2)}. 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 techniques 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 associates²⁾ repaired PAPVC to the high SVC(1cm or more above the cavoatrial junction) by coaptation of the inferior margin of the septal defect to the anterior and lateral margins 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 complications³⁻⁵⁾. 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 Taegu 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 bronchiolitis 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 1. Patients

Patient No.	Age/Sex	Types of interatrial communication	Origin of pulmonary veins	Associated cardiac anomalies	Follow-up(months)
1	8yr/F	Sinus venosus defect	RUL		24
2	4yr/M	Sinus venosus defect, PFO	RUL		*
3	2m/M	Secundum ASD	RUL	PDA, left SVC	17
4	28yr/F	Intact atrial septum	RUL, RML, RLL		15
5	66yr/F	Sinus venosus defect	RUL, RML		15

*, 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; PDA, patent ductus arteriosus; SVC, superior vena cava

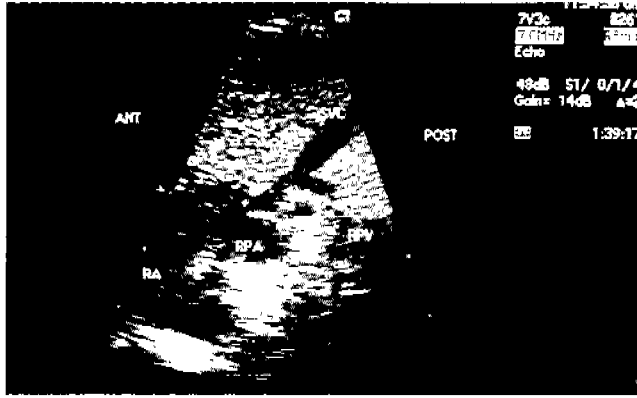


Fig. 1. Preoperative transthoracic echocardiography in Patient 3. Anomalous pulmonary vein from right upper lobe is connected to the small right SVC. SVC, superior vena cava



Fig. 2. Preoperative MR angiography in Patient 4. Pulmonary veins from the entire right lung are drained into the SVC. SVC, superior vena cava

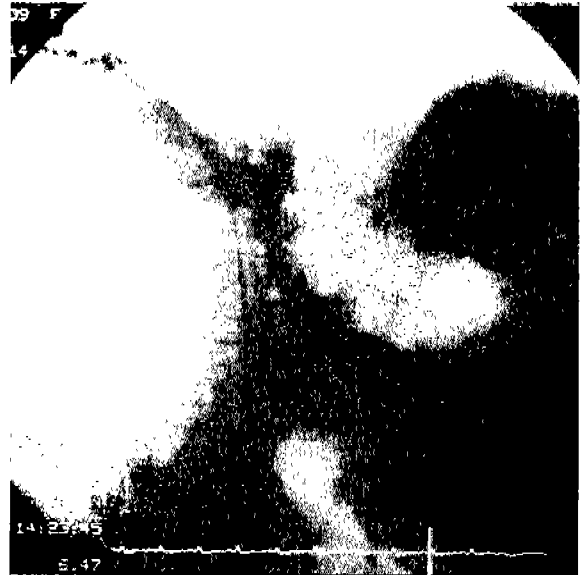


Fig. 3. Preoperative angiography in Patient 5. Anomalous right pulmonary vein is directly connected to the SVC above the cavoatrial junction. SVC, superior vena cava

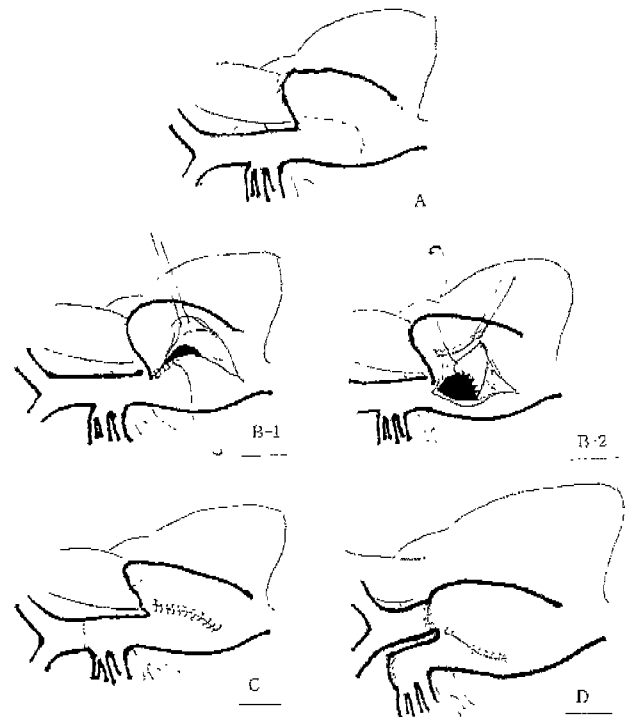


Fig. 4. Operative procedure. A. An incision is made in the right atrial appendage. B. The SVC and anomalous pulmonary veins are baffled with right atrial flap(B-1) or prosthetic patch(B-2). C. The SVC is transected just above the highest anomalous pulmonary vein. The tip of RA appendage is divided and muscular trabeculations inside the appendage are completely excised. D. The proximal of the SVC is closed. End to end cavoatrial anastomosis completes the procedure. SVC, superior vena cava; RA, right atrium

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 limb 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 atriocavoplasties. In 1965 Kirklin, Ellis, and Wood¹⁾ reported their procedure, which involves covering both defect and veins with a patch that directs blood flow from the anomalous

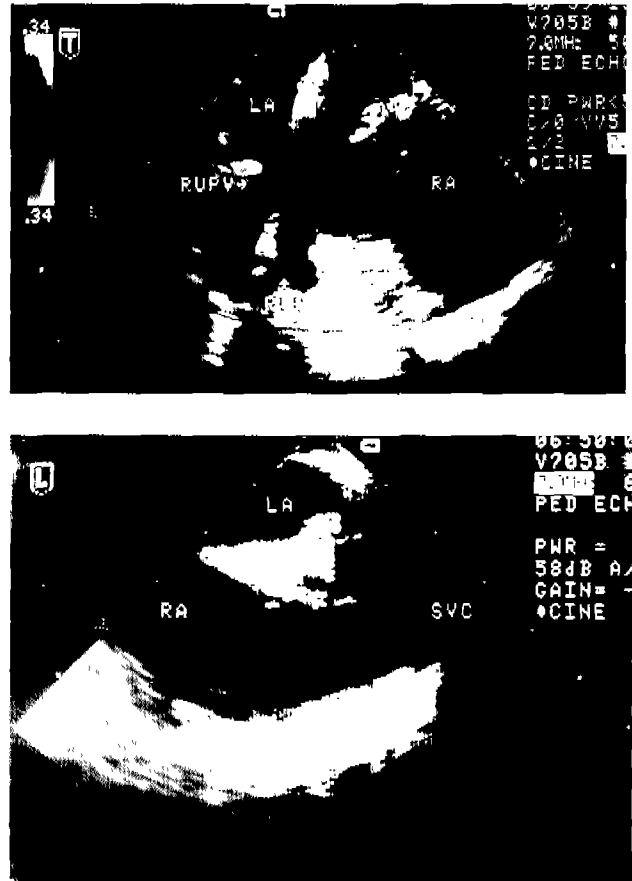


Fig. 5. Postoperative transesophageal echocardiography in Patient 1. A. Anomalous pulmonary vein is redirected through intra-atrial baffle into LA. B. Widely patent reconstructed cavaatrial pathway.

pulmonary veins through the ASD into the LA. Schuster, Gross, and Colodny⁶⁾ recommended a second patch to enlarge the junction between the SVC and the RA to decrease the incidence of postoperative SVC obstruction. Kyger and colleagues⁷⁾ described early and late results in 109 patients with sinus venosus defects. No instances of obstruction of the SVC were detected clinically. In 6 patients the cavaatrial 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 cavaatrial junction was enlarged with a patch developed persistent new postoperative arrhythmias compared with an overall incidence of 14%

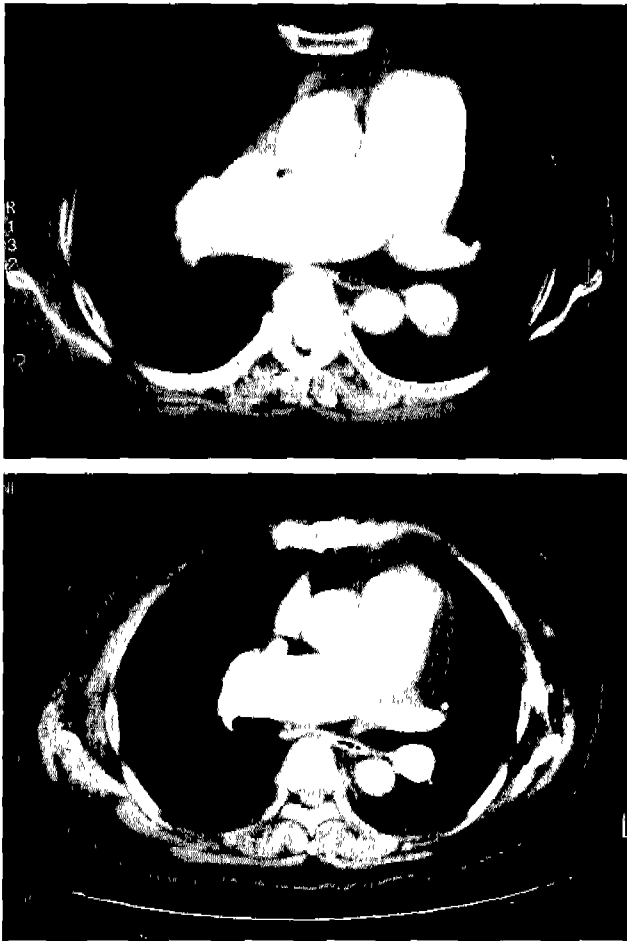


Fig. 6. Dynamic chest computed tomography in Patient 5. A. Preoperatively anomalous pulmonary vein is connected to the SVC above the markedly dilated right pulmonary artery. B. Postoperative follow-up at 15 months shows free cavoatrial flow. SVC, superior vena cava

persistent new postoperative arrhythmias. Trusler, Kazenelson, Freedom, and Williams⁸⁾ reported comprehensive follow-up of 29 children after repair of PAPVC and sinus venosus defect. Eight patients required an additional SVC patch to maintain an adequate lumen. Cardiac catheterization in 18 patients revealed trivial to slight narrowing of the SVC in 8 children and a significant SVC-RA pressure gradient in one child. In 4 patients small left-to-right shunts were detected. Holter monitoring and exercise testing detected rhythm abnormalities in 13 of them, but sinus node recovery times were normal in all 12 patients tested. Stewart, Alexson, and Manning⁹⁾ reported encouraging results of repair of PAPVC to the SVC with simple pericardial baffle without enlargement of the SVC in 15 patients. Jemielity and co-workers¹⁰⁾ recently reported 1 SVC obstruction of 25 adult

patients with similar operative procedure. Agrawal, Khanna, and Tampe¹¹⁾ described follow-up study of 44 patients after repair of sinus venosus 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 atriocavoplasties were advocated to construct unobstructed atriocaval channel. Chartrand et al¹²⁾ 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-auricular tunnel was enlarged with a RA appendage-SVC angioplasty. Kubota and associates¹³⁾ examined mid-term results of their rotation-advancement flap method of repair¹⁴⁾ in 11 patients, which consisted of atrial partitioning with a polytetrafluoroethylene sheet, enlargement of the cavoatrial junction by a right atrial flap, and protection of the sinus node by keeping their incision through the anterolateral wall of the atriocaval junction. Follow-up hemodynamic and electrophysiologic studies in 7 patients revealed successfully rerouted pulmonary venous drainage and reconstructed cavoatrial junction without stenosis and without clinically significant atrial pacemaker dysfunction. They pointed out a possibility of injuring the precaval sinus node artery originating from right coronary artery in this type of operative method. Nicholson and colleagues¹⁵⁾ reported lateral transcaval approach in 66 patients (range, 1.5-65 years) with sinus venosus 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 Lewis¹⁶⁾ in 1958. Ehrenhaft, Thcilcn, and Lawrence¹⁷⁾ described an atrioseptopexy and transplantation of the SVC into the RA appendage. In 1984 Warden and associates²⁾ 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 group³⁾ 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 al⁵⁾ 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 Kreutzer¹⁸⁾ 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 al⁴⁾ 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 al¹⁹⁾ 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)²⁰⁾. Busquet et al²¹⁾ described the well-established preponderance of origin of the sinus node artery from the right coronary system(66%) as apposed 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 area²⁰⁻²²⁾. It may partly explain relatively frequent incidence of atrial dysrhythmias following conventional repair of PAPVC to the SVC. Apart from direct surgical trauma, retraction of sinus node without vascular injury¹⁵⁾ and gradual fibrosis in the area surrounding the sinus node¹¹⁾ 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 cavoatrial anastomosis is the most

critical part of the procedure. The azygos 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.

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=국문초록=

배경: 폐정맥 및 상대정맥의 폐쇄와 동방결절의 기능장애 등은 상대정맥으로 환류되는 부분폐정맥 연결이상의 수술교정 이후 발생할 수 있는 합병증으로 술 후 장기성적을 결정하는 요인이 되어왔다. 저자들이 시행하고 있는 수술방법을 기술하고 조기결과를 분석하였다. **대상 및 방법:** 1999년 4월부터 2000년 1월까지 세종병원과 대구가톨릭대학병원에서 5명의 환자가 상대정맥으로 환류되는 부분폐정맥 연결이상으로 교정수술을 받았다. 환자의 나이는 생후 2개월부터 66세까지였다. 수술방법은 우심방 피관(2례)이나 침포(3례)를 사용하여 이상폐정맥을 심방간 통로를 통하여 좌심방으로 정상환류시키고, 상대정맥을 이상폐정맥의 상부에서 절단한 후 근위부를 폐쇄하고 상대정맥의 원위부를 우심방이에 단단문합함으로써 상대정맥과 우심방의 혈류를 재건하였다. **결과:** 모든 환자가 합병증 없이 술 후 9일과 15일 사이에 퇴원하였다. 퇴원 후 본국으로 돌아간 러시아 환아를 제외한 4명의 환자는 평균 17.75 ± 4.27 개월의 추적기간 동안 증상 없이 정상동물등을 보였다. 술후 12개월에서 24개월에 시행한 심장초음파검사상 폐정맥이나 상대정맥의 협착 및 잔류단락은 보이지 않았다. **결론:** 저자들은 상대정맥으로 환류되는 부분폐정맥 연결이상 환자에서 기술한 수술방법으로 폐정맥 및 상대정맥 혈류의 협착 없이 좋은 결과를 얻을 수 있었다. 상대정맥과 우심방접합부위를 가리지 않는 절개를 피함으로써 동방결절 및 그 동맥의 손상을 최소화 할 수 있을 것으로 생각된다.

- 중심 단어:** 1. 부분폐정맥 연결이상
2. 상대정맥
3. 상대정맥-우심방이 단단문합