Bilateral Partioning of Systemic Venous Chamber in Conjuction with Atriopulmonary Anastomoses (Fontan-Kreutzer)

- A new technique -

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〈국문초록〉

체정맥환류 이상을 동반한 복잡심기형 환자에 있어 체정맥심방 양분을 이용한 Fontan씨 술식 치험 - 새로운 수술방법 -

김 진 국'ㆍ김 용 진'ㆍ서 경 필'

Fontan씨 술식이 적용증이 되는 복잡심기형에 있어 체정맥 환류이상은 수술을 어렵게 할 뿐 아니라 수술사망율을 높이는 중요한 요인중의 하나이다. 본 서울대학교 병원 흉부외과에서는 상대정맥이 좌 우에 모두 있고 하대정맥이 우측 심방으로 들어오며, 또 좌측간정맥이 좌측심방으로 들어오는, 폐동맥협 착증을 동반한 단심방 및 단심실 환아를 경험 하였다. 이 예에 대한 치료에 있어, 저자등은 좌측 상대정맥과 좌측간정맥으로 들어온 정맥혈은 좌측 폐동맥으로, 우측 상대정맥 및 하대정맥으로 들어온 정맥혈은 우축폐동맥으로 가게 하고 폐동맥을 통해 들어온 피는 단일심으로 가게하는 변형 Fontan씨 술식을 시행하였다. 이는 구조적으로는 특이하나, 혈액순환 생리상 체순환과 폐순환을 완전히 분리시켜 주는 완전교정술이며, 기존의 술식에 비해 수술이 용이하고 폐정맥 환류 장애가 생기지 않을 수 있는 장점이 있다고 사료된다.

-Abstract-

A technique applicated for physiologic correction of complex congenital cardiac disease suitable for Fontan procedure in which drainage of left superior vena cava and hepatocardiac vein to left atrium combined is described. We made one systemic venous baffle from left hepatocardiac vein to left superior vena cava and another systemic venous baffle from right inferior vena cava to the right superior vena cava with rigid prosthetic material(0.5mm thickness PTFE patch). And then we anastomosed directly between the right sided atrial appendage and right pulmonary artery, and left-sided atrial wall beneath the appendage and left pulmonary artery.

We believe that this procedure is superior to the method using intratrial tube graft to divert the left hepatocardiac venous blood to right atrium, and applicable for physiologic correction of any complex congenital cardiac disease suitable for Fontan-type procedure in which anomalies of systemic venous drainage combined.

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After the introduction of Fontan procedure for physiclogic correction of tricuspid atresia in 1971¹¹, the procedure has since been modified and extended to other more complex lesions^{2–8}). But till now, the coexistence of anomalies of systemic venous connection poses both technical difficulties at repair and an increased operative mortality. In this report, we describe a new operation in which bilateral systemic venous baffles and bilateral atriopulmonary anastomoses was created in a patient with presently physiologically–uncorrectable cardiac anomalies, including bilateral superior venae cavae, right inferior vena cava and large left hepatocardiac vein in association with single ventricle variety.

Case

A 12-year-old boy was admitted to the Seoul National University Children's Hospital for surgical repair of cyanotic congenital heart disease. He was severely cyanotic, but well-developed. A grade 2/6 ejection murmur was noted at the base of the heart. A Chest X-ray film showed slightly decreased pulmonary vasculature. Electrocardiography demonstrated a normal sinus rhythm and right axis deviation. Echocardiography disclosed physiologically single ventricle with ventricular D-loop and right ventricular main chamber associated with large single atrioventricular valve. And it showed right and left superior vena cava (SVC), inferior vena cava(IVC) and left hepatocardiac vein drained to an left-sided atrium with no discernible atrial septum. By cardiac catheterization and angiography, a catheter from IVC could be passed into the left hepatocardiac vein through atrium, and also into the left SVC due to due to devoid of atrial septum and roof of coronary sinus-(Fig. 1).

The systemic oxygen saturation was 67%. Pulmonary arteriography revealed valvular and subvalvular stenosis and mean pulmonary artery pressure was 13mmHg. The right and left pulmon-

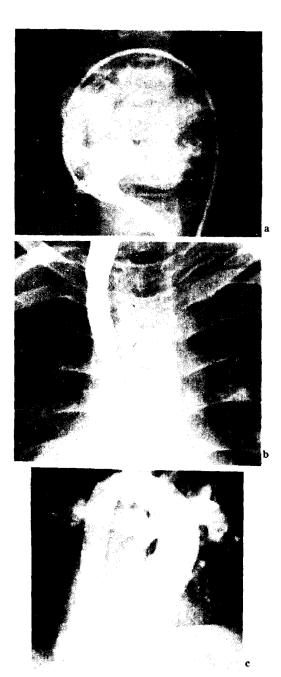


Fig. 1. Preoperative angiocardiograms

- a) Anteroposterior view shows opacification of atrium and inferior vena cava. The catheter is introduced to heart via left hepatocardiac vein.
- Anteroposterior view of showing right superior vena cava.
- c) Anteroposterior view of showing relation of great arteries. Both pulmonary arteries are confluent and good-sized

ary arteries were confluent and good-sized(PA index=256).

All findings were fitted for Fontan-variety operation. Surgical correction was undertaken with the preoperative diagnosis of single atrium, single ventricle and pulmnic stenosis associated with systemic venous anomalous drainage. On pericardiotomy, the cardiac apex was toward to left side. The Aorta and pulmonary artery was normal but somewhat side-by-side relationship, and arised from single ventricle. The coronary arteries had their usual cources. The right-sided atrium had large, long and downwardly hooked appendage akin normal left atrial appendage, and the left--sided atrium had rather small but prominently constricted appendage with its end upward. The right SVC(15 mm), left SVC(15 mm), IVC(20 mm) and left hepatocardiac vein(25 mm) were noted. So the right and left SVC were cannulated with right-angled venous cannulas, and hepatocardiac vein directly and IVC via lower right atrium with straight cannulas. With the patient on cardiopulmonary bypass, the right atrium was opened obliquely paralled to the atrioventricular groove.

Left SVC opening was located between left upper pulmonary venous opening and left atrial appendage orifice, and hepatocardiac venous opening was anterior and inferior to left lower pulmonary venous opening. Pulmonary venous drainage was normal. The AV Valve had single large annulus and more than three(but not easily defined) leaflet which suggested endocardial cushion defect, but there was no discernible rudimentary chamber with the naked eyes. Because of the fear of obstruction to pulmonary venous drainage when using intraatrial rerouting of abnormal systemic venous drainage, it was decided to partition the atrium into right and left systemic venous chamber. Thus a 0.5 mm-thickness PTFE patch was selected and tailored appropriately. The patch was sutured around the orifice of the left SVC and hepatocardiac vein. The posterior longitudinal edge was sutured anterior to the left pulmonary veins, and the anterior longitadinal edge was sutured to the left-sided atrial wall to make a patch a baffle. Another PTFE patch was selected and sutured around the orifice of right SVC and IVC, anterior to the right pulmonary veins, and posterior edge of the right-sided atriotomy incision. The remaining strip of atrial free wall, which made up part of the circumferences of right and left systemic venous chamber, allowed growth potential. The original atriotomy incision closed by direct suture to complete the new pulmonary venous(middle) chamber.

And then direct atriopulmonary anastomoses without prostheses between right sided atrial appendage and right pulmonary artery, and left--sided atrial wall beneath the appendage and the left pulmonary artery were done after main pulmonary artery division and ligation. So now blood drained via right SVC and IVC flowed into right pulmonary artery, and blood from left SVC and hepatocardiac vein into left pulmonary artery. After cessation of cardiopulmonary bypass, each systemic venous chambers and pulmonary arteries were cannulated and checked pressures. Comparing the pressures, we found significant pressure gradient between right-sided systemic venous chamber and right pulmonary artery(3 cm H₂O), So added modified Glenn operation on that side(Fig. 2).

The postoperative course was uneventful, except moderate amount of plerual effusion on both side for 8 days. Cardiac catheterization and angiopraphy 2 weeks after operation revealed very small amount of right to left shunt between left-sided systemic venous chamber and pulmonary venous chamber, but acceptably increased systemic oxygen saturation(86%) and near normal atrial pressures(mean pressure of systemic venous chamber, right; 14mmHg, left: 13 mmHg) and no restriction of systemic venous drainages to pulmonary arteries at both sides(fig. 3).

He had been followed up for 4 months and the systemic oxygen saturation at 3 months postopera-

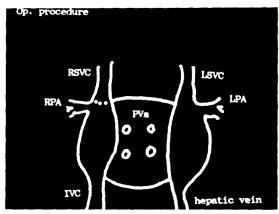


Fig. 2. Diagram illustrating method of bilateral partioning of systemic venous chamber. There is no restriction of pulmonary venous drainage.

The dotted line shows modified Glenn procedure.

tively showed more improvement than previous study(88%). He is now receiving digoxin and diuretics, but enjoying his school life without difficulty.

Comments

In recent years, many clinical and experimental studies for technical and physiological aspects of Fontan procedure have been advocated, and so extend of the limits for that procedure has been achieved greatly²⁻⁸⁾. But still the coexistence of anomalies of systemic venous connection poses both technical difficulties at repair and an increased operative mortality. Many reports described that intraatrial baffles constructed to divert a left SVC-left atrium connection to the right side of the atrium had a significant potential to cause either systemic or pulmonary venous obstruction with consequent mortality⁷⁻⁹⁾. So some advocated that an extracardiac exclusion of the left SVC by a bidirectional end-to-side left cavopulmonary anastomosis seemed safer in this situation⁹⁾. The presence of an IVC draining into the left atrial side had another problem.

The use of an intraatrial-tube graft to connect both IVC with the right SVC, followed by an

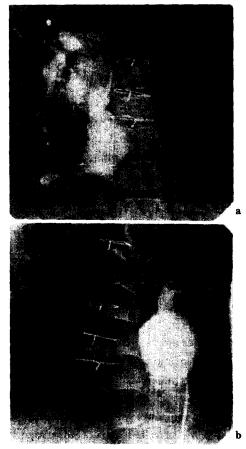


Fig. 3. Posterative angiocardiograms.

- a) Anteroposterior view shows right systemic venous chamber. The catheter is introduced via right superior vena cava.
- b) left antero-oblique view of showing left systemic venous chamber. The catheter is introduced via left hepatocardiac vein.

anastomosis of the upper part of the graft to the right pulmonary artery is an attractive alternative 10). However, the longterm outcome of such a prosthetic intraatrial tubular graft in terms of patency remains to be elucidated.

If above two condition were associated together, difficulties to repair may increase. And the rare condition in which normally drained IVC in right atrial side associated with large hepatocardiac vein draining to left atrial side coexist may be seemed to be physiologically uncorrectable. Kawashima in 1984 advocated to do total cavopulmonary shunt

operation consisting of end-to-side anastomosis between the SVC with azygos or hemiazygos continuation and the confluent pulmonary artery and division or lagation of the pulmonary artery truck-to such complex lesions¹¹.

But, as cited by himself, because that operation is virtually an extension of Glenn's idea, undesirable late sequele related to Glenn's operation may take place in the tufure and, in addition, arterial oxygen desaturation inevitably persists due to drainage of the hepatocardiac venous and coronary sinus blood into the functional left atrium. The idea for our approach was derived from two basic knowledges.

The first is that the basic concept for the Fontan procedure is not greatly different from the Glenn's, i.e., not pumping but flooding of the systemic venous blood to pulmonary circulation. And the second is that systemic venous baffle rather than pulmonary venous baffle can be constructed easy and useful to avoid systemic and pulmonary venous obstuction¹²).

Based on the above idea, we made one systemic venous baffle from left hepatocardiac vein to the left SVC and another systemic venous baffle from the right IVC to the right SVC with rigid prosthetic material (0.5 mm thickness PTFE patch) under tension. Above procedure could be done with only single right atriotomy incision 1 cm away from and paralled to the AV groove which extended from the base of the right atrial appendage to the right atrium—IVC junction. It could be done without difficulty and without the risk of obstructing neither the pulmonary venous drainage nor single AV valve orifice.

The next step was to make systemic venous to pulmonary artery continuity. To achieving this, several techniques, for example, a direct anastomosis between the roof of the atrium or the atrial appendage and the pulmonary artery, or cavopulmonary anastomosis at either or both atrial side, could be used according to the surgical facility. We used a direct end-to-side anastomosis between

right-sided atrial appendage and right pulmonary artery and another anastomosis between the roof of the left-sided atrium and the left pulmonary artery. By the above procedure we have suggested to save the sinus rhythm and atrial contraction, and so to earn some additional, but not cirtical, hemodynamic advantages¹³).

From the above our experience, we hopefully suggested that the coexistence of anomalies of systemic venous connection could poses neither technical difficalties at repair nor increased operative mortality from now. Any cases of anomalous systemic venous drainage can be managed with partitioning of atrium with bilateral systemic venous baffles and then to make systemic venous pulmonary continuity. There will be reduced risk of obstruction to systemic or pulmonary venous drainage.

In addition, there will be natural potency for growth of any chamber or orifice because of no use of prosthetic material except small portion of each venous chambers.

We believe that this procedure is applicable for physiological correction of any complex congenital cardiac disease suitable for Fontan procedure, if in which anomalies of systemic venous drainage combined.

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