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Fontan-like Operation of Seven Cases

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Abstract

We report seven patients who underwent a Fontan-like operation. Three of them died from arrhythmia, low cardiac output and ischemia. Only one of the survivors enjoys unrestricted activity. The remaining three have residual shunt at the atrial level; two of them underwent reoperation in order to close this shunt, but in one case high venous pressure in the inferior vena cava persisted resulting in protein losing syndrome, and in another case the residual shunt decreased. The remaining patient is a possible candidate for reoperation. Careful considerations of the limitations of indication for Fontan-like operation, surgical procedures and post-operative management are important to avoid the poor results.

Introduction

The operation developed by FONTAN and BAUDET⁹⁾ was first applied to patients with tricuspid atresia. At present, the principle of this operation is defined as a procedure in which the right ventricle is bypassed in order to convey desaturated venous blood from the right atrium to the lungs. The procedure also has wider application for different types of congenital heart diseases.

However, the poor results observed in patients without an atrial septum point to the limited application of the Fontan principle. Postoperative management of the patients with poor results, such as residual atrial septal defect, protein losing syndrome and impaired liver function

Key words: Fontan-like operation, Tricuspid atresia, Univentricular heart, Common atrium, Residual shunt.

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resulting from high central venous pressure, and low cardiac output syndrome from increased pulmonary capillary resistance needs further analysis of a large number of patients.

We report seven patients who underwent the Fontan procedure and describe the problems encountered.

Patients (Table 1)

Case 1. A 13-year-old male with tricuspid atresia (1b) and ventricular and atrial septal defects. A Blalock-Taussig anastomosis prior to the Fontan-like operation performed at the age of 3 years and 8 months, as episodes of anoxia occurred at the frequency of once a month. The second cardiac catheterization revealed arterial saturation of 61% and suitable status for the Fontan-like operation. At the age of 13 years, the Fontan-like operation was performed. The Blalock-Taussig anastomosis was closed with double ligation. The atrial (10×20 mm) and ventricular (5×5 mm) septal defects were closed by direct suture. A Hancock valve (23 mm in diameter) was inserted near the junction between the inferior vena cava and right atrium. Next, woven Dacron (25 mm in diameter) was grafted between the right atrial appendage and the hypoplastic right ventricle, leaving the autapulmonary valve intact.

The postoperative course was uneventful, with a central venous pressure of 20 cm of water, blood pressure 125/85 mmHg, P_{O_2} 80 to 96 mmHg (under artificial ventilation). The liver was palpable 2 cm below the costal margin. Postoperative cardiac catheterization revealed the following intracardiac pressures: main pulmonary artery 15/4 mmHg (mean 10), right atrium 21/10 (mean 15) mmHg, left atrium 7/-3 (mean 2) mmHg and left ventricle 95/0 mmHg (Fig. 1).

Table 1. Fontan-like operation of seven cases

Patient	Diagnosis	Preceding Operation	Fontan-like Operation	Postop. Course				Result	Follow-up
				CVP	BP	P_{O_2}	P_{CO_2}		
1 H N 13 m	TA (1b)	3y8m B-T	App-(Graft (25.)) - small RV Hancock valve (23) between RA & IVC ASD (20×10): DC, VSD (5×2): DC	20 cmH ₂ O	125/85 mmHg	85	42	Residual VSD (17%)	10 yrs
2 K M 9 m	TA (1b)	1y2m Glenn 6y B-T	App-(Graft (22.)) - small RV ASD (14×12): PC, VSD (6×6): PC	18	105/75	38	37	Residual ASD ↓ Reop. (10M)	4.5 yrs
3 Y F 7 f	TA (1b)·PS	6y B-T	App-[Conduit with Hancock valve (22)] - small RV Dilatation of m-PA with Graft ASD (25×20): PC, VSD (12×12): PC	17	105/70	32	36	Died 20hrs postop. (Ischemia)	-
4 R T 2.9 f	TA·TGA (SDD) PS	1y7m B-T	App-[Pericardial roll SJM valve (25A)] - r·PA ASD (1&1) 20×15 : PC	24	105/65	41	42	Died 10days postop. (LOS Arrhythmia)	-
5 M O 2.1 m	SV·TGA (SLL) PS·PH	none	App-m·PA Tricuspid valve & PFO: PC m·PA: divided	32	64/40	110	40	Died 3days postop. (LOS)	-
6 A M 8 f	SV·TGA (SDL) PS	3y10m B-T	App-r·m·PA Tricuspid valve, ASD (10×10): PC m·PA: ligated	18	90/60	32	37	Residual ASD	2 yrs
7 N F 11 f	SV·TGA (SDD) PS·SA·common A-V valve	2y9m B-T 9y Glenn	RA (SVC) - r·PA Construction of RA using Dacron Patch m·PA: ligated	22	95/65	46	42	Residual ASD·I-SVC ↓ Reop. (5M)	14 mo

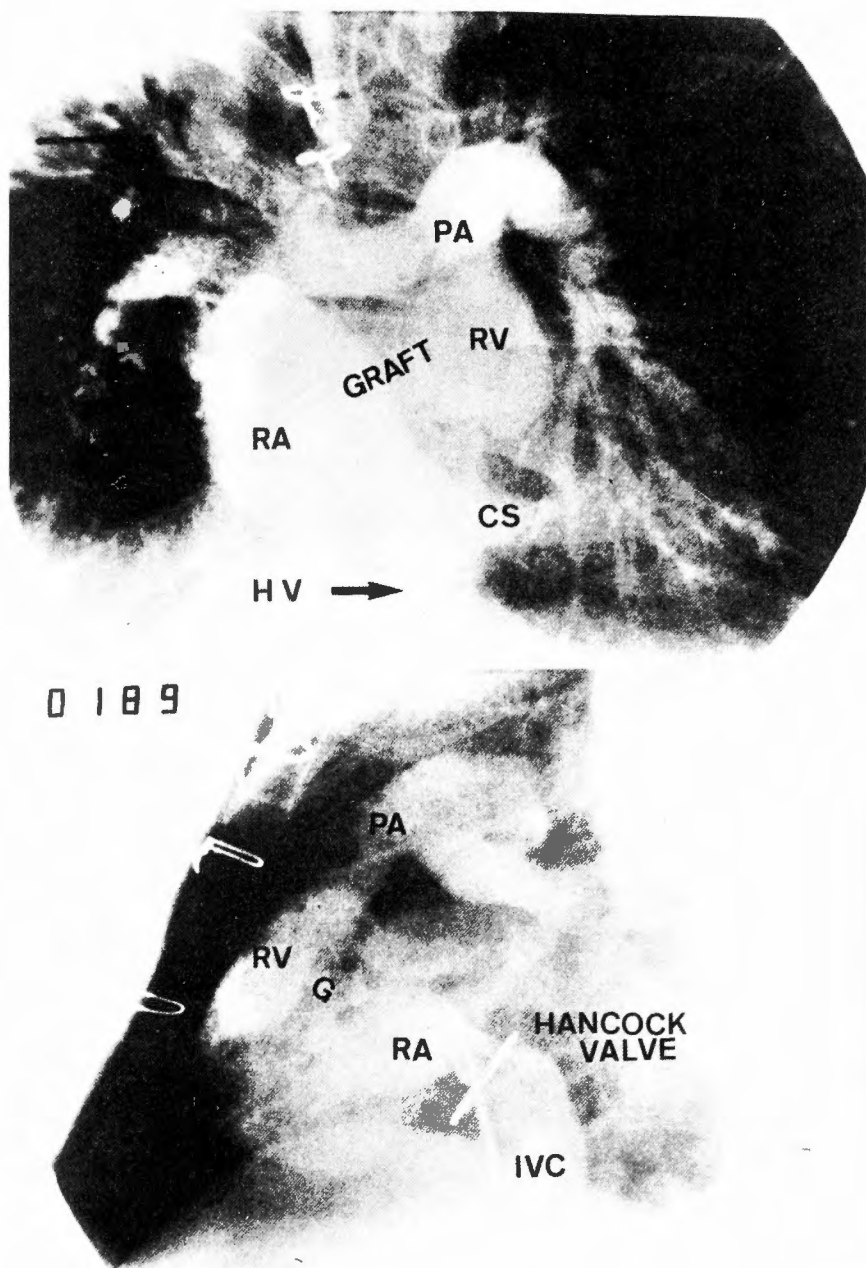


Fig. 1. Cardioangiography performed nine years postoperatively (Patient 1)
PA: pulmonary artery, RV: right ventricle, G: graft, RA: right atrium, HV: hepatic vein, CS: coronary sinus

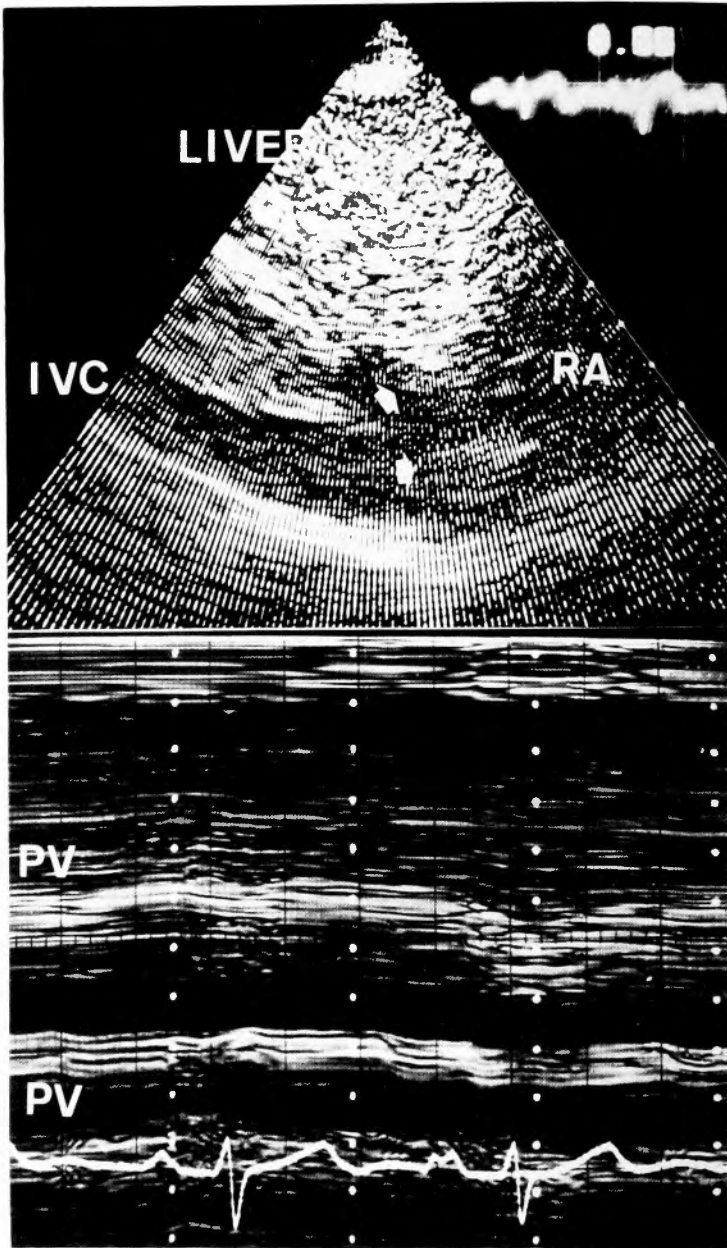


Fig. 2. Echocardiography recorded nine years postoperatively (Patient 1)
 RA: right atrium, IVC: inferior vena cava, PV: frame of prosthetic valve,
 Arrows show a frame of the prosthetic valve.

Echocardiography showed a non-functioning Hancock valve without calcification (Fig. 2). At present the patient is well without limitation of physical activity.

Case 2. A 9-year-old male with tricuspid atresia (Ib) underwent a Glenn anastomosis at the age of 1 year and 2 months, and a Blalock-Taussig anastomosis at the age of 6 years in order to

alleviate severe cyanosis. At the age of 9 years, the Fontan-like operation was performed. The atrial (12×14 mm) and ventricular (6×6 mm) septal defects were closed with Dacron patches. The Blalock-Taussig anastomosis was ligated. The pulmonary valve was normal. A woven Dacron graft (22 mm in diameter) was used as a bypass between the right atrium and hypoplastic right ventricle.

During the postoperative course, he suffered from dyspnea and cyanosis, with PO_2 of 27 to 40 mmHg and P_{CO_2} 35 to 40 mmHg. Postoperative cardiac catheterization data were as follows: left pulmonary 18/9 (mean 11) mmHg, left ventricle 84/10 mmHg, right atrium 22/9 (mean 13) mmHg and left atrium 15/0 (mean 4) mmHg. Arterial saturation was 80.5% with a right-to-left shunt 0.77 l/min. Angiography of the right atrium revealed a right-to-left shunt at the atrial level.

Because of serum hepatitis, surgery was delayed for six months, at which time closure of another atrial septal defect (6.5×6.5mm) was performed using a combined extracorporeal circulation and surface hypothermia. However, reoperation was necessary due to massive hemorrhage from the hilus of the right lung. A fourth operation was performed to remove the massive Oxycel[®] used in the previous operation. The postoperative course was very complicated with pyothorax, pneumothorax, oliguria, intestinal bleeding and azotemia. The left diaphragm was elevated with low excursion. The liver was palpable 5–6 cm below the right costal margin with slight liver dysfunction. Support by diuretics and digitalis was needed to improve congestion of the lower body. One and a half years postoperatively, protein losing syndrome became remarkable, occasionally complicated with edema and ascites. Total serum protein was 4 g/dl and albumin was 2 to 2.5 g/dl. Therefore, 25% albumin 150–200 ml was administered once or twice a week, but with little effect. Intravenous hyperalimentation (400–700 Kcal a day) was performed for 36 days. However, this treatment also had little effect.

Cardiac catheterization performed 2 years postoperatively revealed that mean pressures of the superior and inferior vena cava were 15 and 19 mmHg, respectively, with no pressure gradient between the right atrium and main pulmonary artery. Angiography showed no stenosis at the graft or anastomosed sites (Fig. 3).

Case 3. A 7-year-old female with tricuspid atresia (Ib) and pulmonary stenosis. A left-sided Blalock-Taussig anastomosis was performed to alleviate severe cyanosis at the age of 6 years. Postoperative cardiac catheterization revealed: pulmonary arterial resistance of 378 dynes·sec·cm⁻⁵, main pulmonary arterial pressure 24/9 (mean 17) mmHg, cardiac output 1.87 l/min, cardiac index 2.34 l/min/M² and saturation of arterial blood 87.7%.

At operation, the size of the atrial and ventricular septal defects were 25 and 6 mm in diameter, respectively. Fontan procedure in this operation included closure of the Blalock-Taussig anastomosis, patch closure of the atrial and ventricular septal defects, and bypass, using a valved (Hancock, 22mm in diameter) conduit between the right appendage and a site extending from the hypoplastic right ventricle to the main pulmonary artery. Immediately after the operation, the central venous pressure was 17 cm of water and the systolic blood pressure ranged from 60 to 105 mmHg. However, the patient died due to sudden cardiac arrest the following morning. Autopsy

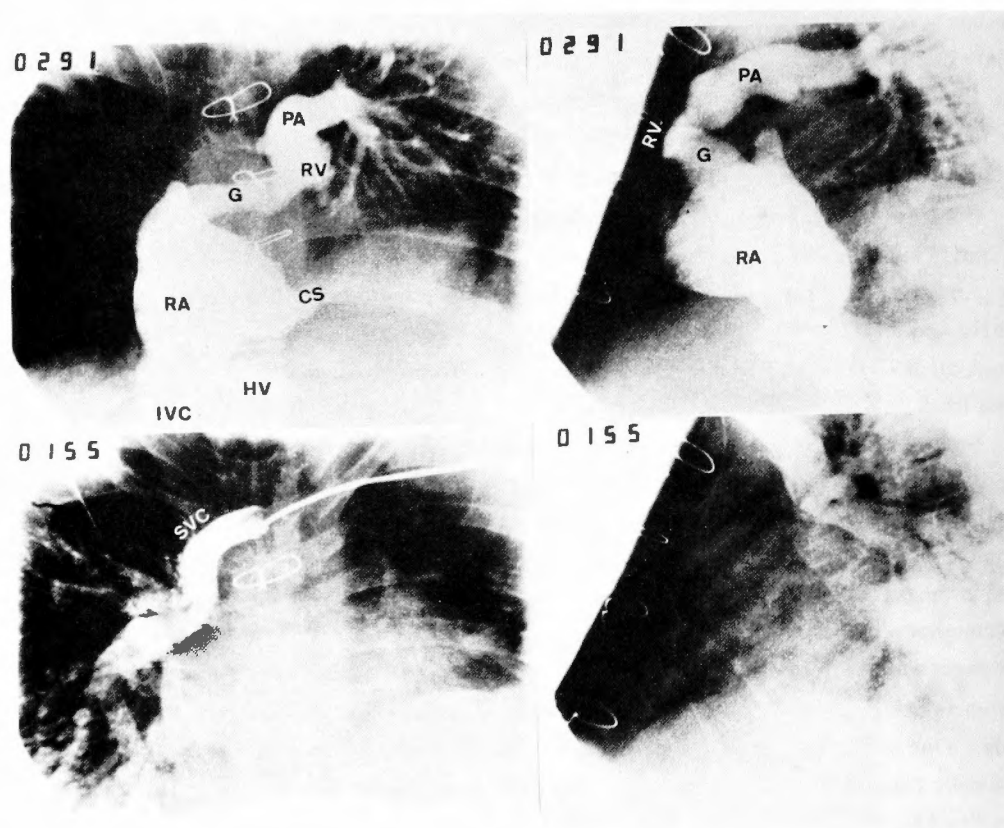


Fig. 3. Cardioangiography after closing a residual shunt (Patient 2)

PA: pulmonary artery, RV: right ventricle, G: graft, RA: right atrium, CS: coronary sinus, IVC: inferior vena cava, HV: hepatic vein

revealed residual atrial septal defect. The narrow incision through the right atrial appendage might have prevented the closure of the defect. Thus, one possible reason for the sudden cardiac arrest may have been compression of the graft as it passed under the median of the sternum.

Case 4. A 2-year-old female with tricuspid atresia, transposition of the great arteries (SDD) and pulmonary stenosis underwent a right-sided Blalock-Taussig anastomosis at the age of 1 year and 7 months. Postoperative angiography revealed that the anastomosis became stenotic with increasing cyanosis. The right pulmonary vein wedge pressure was 15 mmHg; pulmonary arterial resistance was $114.1 \text{ dynes} \cdot \text{sec} \cdot \text{cm}^{-5}$, and arterial saturation 81%. Fontan-like operation through a median sternotomy was performed at the age of 2 years and 9 months. There were two atrial septal defects; primum ($15 \times 20 \text{ mm}$) and secundum ($15 \times 20 \text{ mm}$). These defects were closed with a $15 \times 30 \text{ mm}$ Dacron patch. Thereafter, a SJM valve (25A) was inserted at the right atrial appendage, which was anastomosed end-to-side to the right and main pulmonary artery, using a rolled pericardial patch. Weaning from cardiopulmonary bypass was without difficulty. Blood pressure was 105/65 mmHg; pulse rate 160/min, Po_2 41.0 mmHg and central venous pressure 21.5–24.4 cm of water.

On the second postoperative day, cardiac arrest occurred suddenly after administration of potassium chloride and digitalis for bigemia and tachycardia (190/min). Cardiac massage was performed for about 1 hour but it could not prevent the resulting neurological impairment. On the 8th postoperative day, the patient died due to bronchopneumonia.

Case 5. A 2-year-old male was diagnosed as having a single ventricle (type A), transposition of the great arteries (SLI), mild pulmonary stenosis and ventricular septal defect by cardiac catheterization performed at the age of 2 years. The intracardiac pressures were: right pulmonary artery 65/25 (mean 42) mmHg, right ventricle 80/8 mmHg, right atrium (mean 5) mmHg and ascending aorta 85/50 (mean 70) mmHg. Arterial saturation was 61.1% and pulmonary arterial resistance $250 \text{ dynes}\cdot\text{sec}\cdot\text{cm}^{-5}$. A Fontan-like operation was performed, and the right atrium was separated from the tricuspid valve and coronary sinus with a Dacron patch (20 × 25 mm). The right atrial appendage and main pulmonary artery were anastomosed end-to-end, after the main pulmonary artery was divided and the pulmonary valve closed by continuous sutures.

During the postoperative course, right heart failure resulted in low cardiac output, with a considerable amount of pleural effusion and ascites, high central venous pressure (25–37 cm of water), and low blood pressure (40–80 mmHg). However, blood gas analysis revealed a Po_2 of 61 to 160 mmHg. On the 3rd postoperative day the patient died of anuria.

Case 6. An 8-year-old female was diagnosed as having a single ventricle, transposition of the great arteries (SDL) and pulmonary stenosis at the age of 2 years and 10 months. At the age of 3 years and 10 months, she underwent a Blalock-Taussig anastomosis to alleviate cyanosis. The second cardiac catheterization was performed at the age of 8 years. Pressures of the left pulmonary artery and left pulmonary vein wedge were 21/9 (mean 16) mmHg and (mean 15) mmHg, respectively. Arterial saturation was 82.4%.

A Fontan-like operation was performed. The diameters of the ascending aorta and main pulmonary artery were 23 and 20 mm, respectively. The main pulmonary artery was situated right and posterior to the ascending aorta.

Patent foramen ovale (10 mm in diameter) was closed with a Dacron patch. The tricuspid valve was closed with a patch covering the coronary sinus situated at the side of the tricuspid valve, preventing complete A–V block. Next, anastomosis between the right atrial appendage and right pulmonary artery was performed. The main pulmonary artery was closed with double ligation near the pulmonary valve. The duration of total perfusion was 208 minutes, with an aortic cross-clamping time of 128 minutes.

A catheter was inserted into the subphrenic space to drain ascites. During the first 24 hours postoperatively, the volume of blood and ascites drained was over 3500 ml. The blood pressure was 90/60 mmHg, central venous pressure 18 cm of water, Po_2 31–36 mmHg in Fio_2 of 80%.

Intraaortic balloon pumping was performed postoperatively, but the effect was not remarkable because of tachycardia and supraventricular irregular rhythm. Thrombosis in the external iliac artery developed 48 hours postoperatively. Despite thrombectomy using a Fogarty catheter, deep ulceration in the right lower extremity subsequently developed, resulting in slight disability

of the foot joint.

Extubation was performed on the 10th postoperative day. Arrhythmia was noted for 4 weeks postoperatively, such as nodal rhythm, paroxysmal atrial tachycardia, premature ventricular beat and ventricular tachycardia.

Postoperative cardiac catheterization performed 4 months after the operation disclosed a large right-to-left shunt at the atrial level. Pressures of the superior and inferior vena cava were 17 and 14 mmHg, respectively. Arterial saturation was 72.0%. Angiography of the inferior vena cava revealed that a large amount of blood, returning to the right atrium from the inferior vena cava, flowed into the univentricle through the tricuspid valve.

Case 7. An 11-year-old female with a single ventricle, transposition of the great arteries (SDD), pulmonary stenosis, common atrium and common atrioventricular valve. The patient underwent a Blalock-Taussig anastomosis at the age of 2 years and a Glenn operation at the age of 9 years.

A Fontan-like operation was performed through a median sternotomy. The atrial cavity

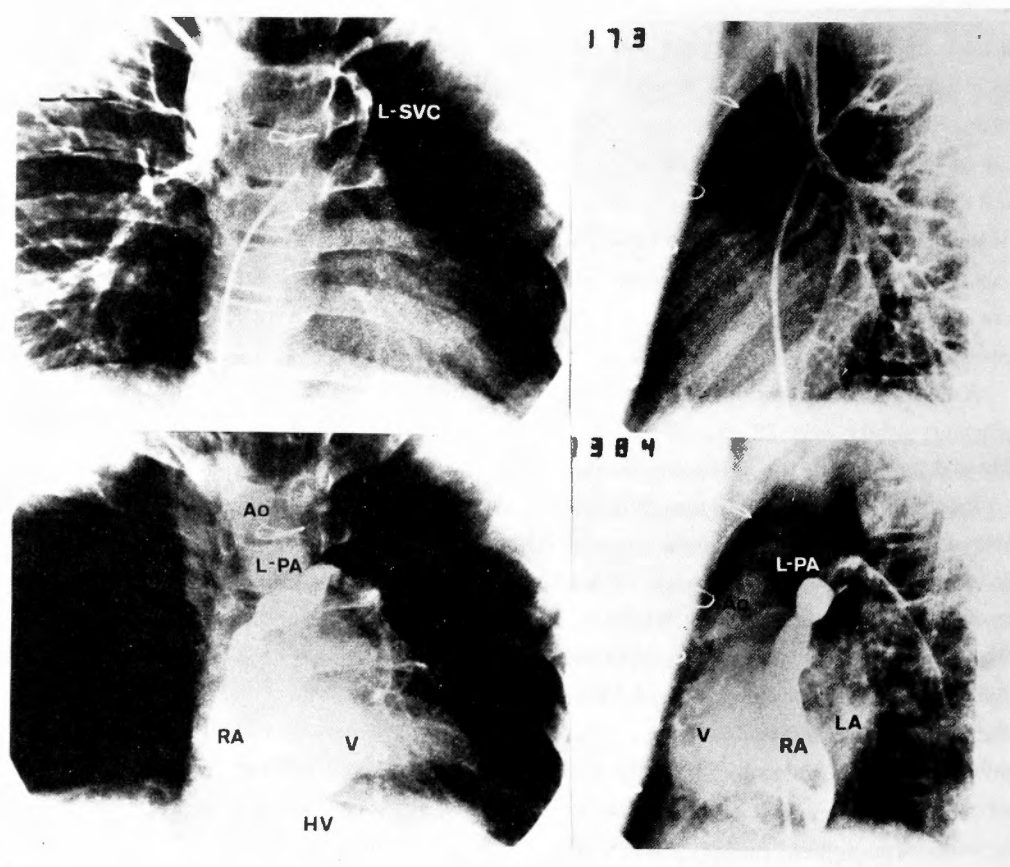


Fig. 4. Postoperative cardioangiography (Patient 7)

Ao: aorta, L-PA: left pulmonary artery, RA: right atrium, V: univentricle, HV: hepatic vein, LA: left atrium

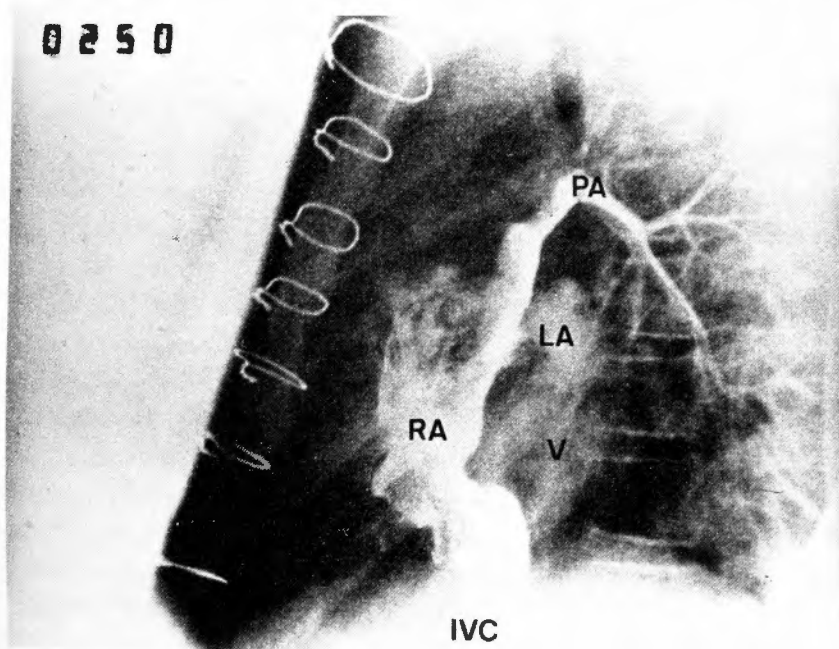
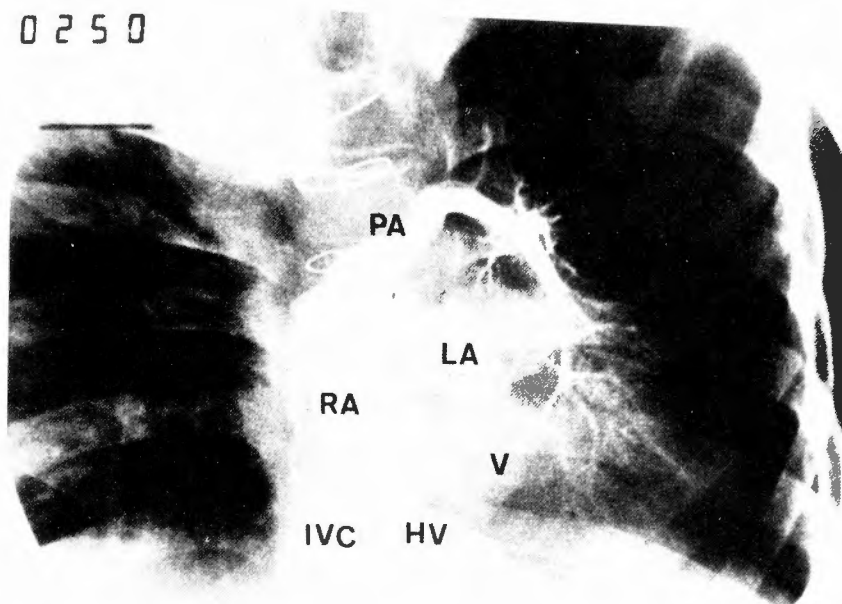


Fig. 5. Cardioangiography after reoperation (Patient 7)
 PA: pulmonary artery, LA: left atrium, RA: right atrium, HV: hepatic vein.
 IVC: inferior vena cava, V: univentricle

was divided by placing a large Dacron patch (40×45 mm) in such a way that all the pulmonary venous return was redirected through the common atrioventricular valve into the univentricle. Thus, the newly constructed right atrial chamber was completely excluded from any communication with the univentricle. The superior vena cava was also divided at a site below the ligation performed in the Glenn operation, and was anastomosed to the right pulmonary artery (20 mm in diameter), using a pericardial patch in order to enlarge the anastomosed orifice. Weaning from the cardiopulmonary bypass was without difficulty. The postoperative course was uneventful except for supraventricular arrhythmia and slight low cardiac output syndrome.

Postoperative angiography revealed a large amount of returned caval blood flowed into the

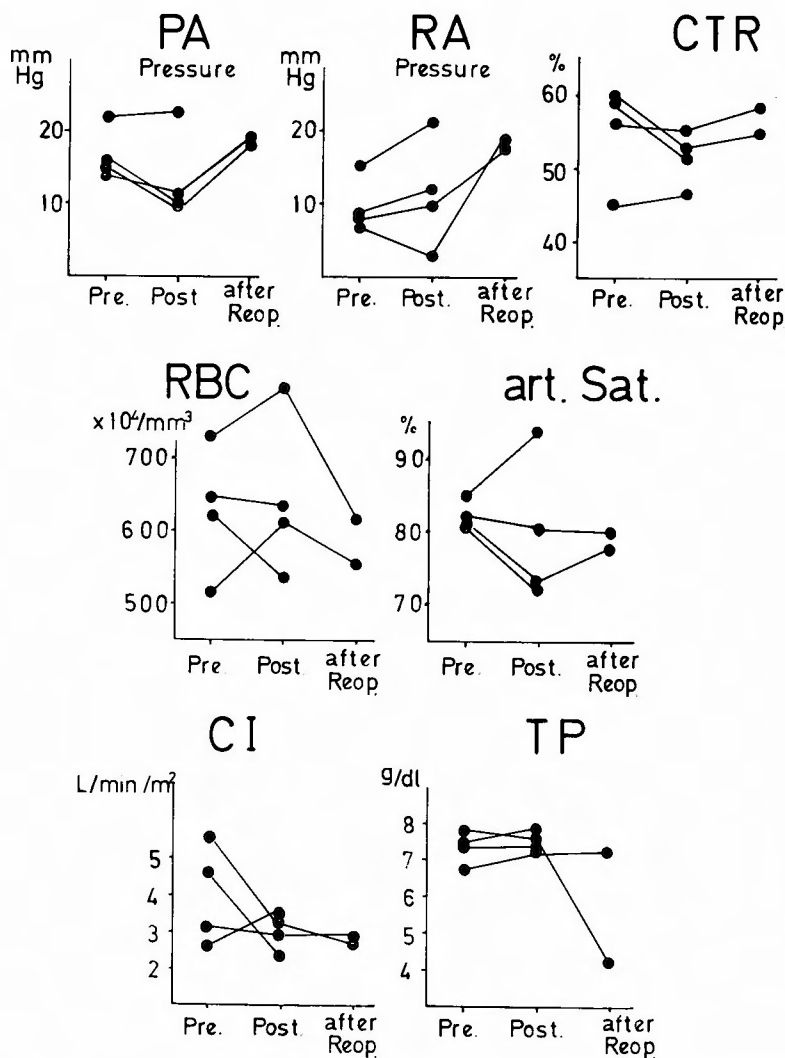


Fig. 6. Postoperative changes in clinical findings

PA: pulmonary artery, RA: right atrium, CTR: cardiothoracic ratio, RBC: red cell count, art. Sat.: arterial oxygen saturation, CI: cardiac index, TP: total protein

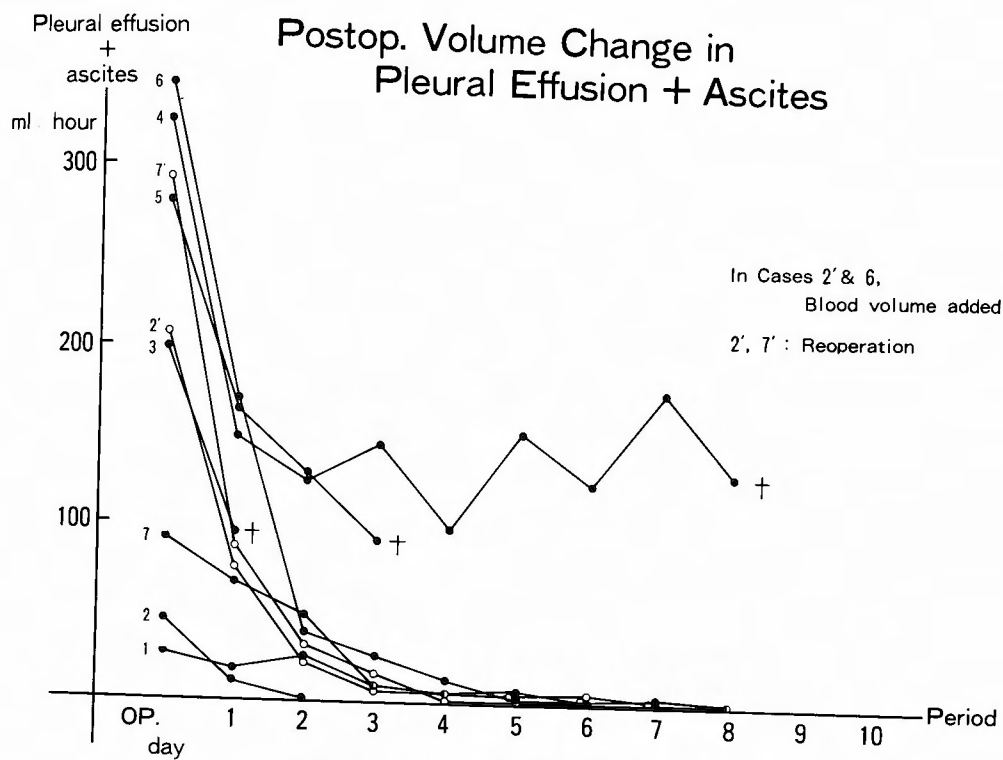


Fig. 7. Postoperative volume change in pleural effusion and ascites

left atrium through residual shunt (Fig. 4). Contrast media injected into the left pulmonary artery mainly returned to the right atrium. In addition, light physical movement developed cyanosis, and tachycardia attacks frequently occurred.

Five months later, reoperation was performed in order to close the residual atrial shunt and patent left superior vena cava. Some dehiscences of the Dacron patch were closed, but a small shunt remained (Fig. 5).

Figure 6 shows changes in clinical findings of the 4 survivors. Postoperative pressures in the right atrium and pulmonary artery were more than 18 mmHg, and these values increased after reoperation. Postoperative low pressures of about 10 mmHg in the right atrium suggest the presence of a residual shunt. Cardiothoracic ratio slightly increased postoperatively. Red cell counts did not decrease below $5.0 \times 10^6/\text{mm}.$ even in the absence of a right-to-left shunt, and these values decreased after reoperation. Postoperative oxygen saturation of peripheral arterial blood was approximately 80% except one case. Cardiac index decreased in 3 patients. No changes were seen in total serum protein levels. However, in Patient 2 serum protein remarkably decreased due to the increased pressure in the inferior vena cava after reoperation.

Figure 7 shows the postoperative volume changes in pleural effusion and ascites. The rate of more than 50 ml/hour on the 2nd postoperative day suggests poor prognosis.

Discussion

FONTAN and BAUDET in 1971⁹⁾, reported an operation for physiological correction of tricuspid atresia in which the distal right pulmonary artery is anastomosed to the superior vena cava and the proximal right pulmonary artery to the right atrial appendage by means of an aortic homograft. Since then, numerous modifications of this operation have been established for the treatment of tricuspid atresia^{1,2,12,13,15,21,24,29,30)}, univentricular heart^{3,4,5,6,7,8,11,16,17,20,22,23,25,28,31)} and others^{11,18)}. The FONTAN principle, which DI CARLO et al.⁶⁾ defined as a procedure in which the right ventricle is bypassed in order to convey desaturated blood from the right atrium to the lungs, gives better results with respect to early mortality and morbidity when a ventricular chamber is included in the circuit.

Nevertheless, there is a tendency to extend the application of this principle to defects other than tricuspid atresia. Most cases presently reported do not always have a favorable atrial anatomy. Atrial septum and atrial wall hypertrophy¹⁶⁾, as well as normal systemic and pulmonary venous connections¹⁸⁾, were considered mandatory for a successful operation. However, atrial wall hypertrophy is no longer considered a necessary feature³¹⁾. Recently, there is an extreme opinion that the right heart is a conduit to the lungs^{14,19,23)}. The problems in cases exhibiting a common atrium have thus far been scantily reported in the literature^{6,16,18)}. DI CARLO et al.⁶⁾ reported 4 cases with common atrium, one survived the operation and 3 died. They speculated that this high risk might depend on the excess proportion of prosthetic material to the atrial myocardium resulting in excessive loss of contractility in the atrial myocardium. Thus, there is concern that present limits of the "expanded horizon"¹⁸⁾ of the FONTAN operation have been reached.

Other than the common atrium, pulmonary or systemic venous abnormalities are disorders showing high mortality following the Fontan-like operation. Two patients, who underwent a Glenn shunt prior to the Fontan-like operation, despite severe complications resulting from a large amount of residual shunt, survived. Although the Glenn shunt has been done in many subsequent patients, presently it is not considered essential for a successful FONTAN operation. However, DELEON et al.⁴⁾ speculated, from the surgical results of the GLENN shunt in patients undergoing the FONTAN operation, that an established GLENN shunt played a major role in attaining minimal postoperative hemodynamic instability, effusions, renal failure, and mortality.

The mortality may be associated with the patient's age. Two patients aged 2 years at the time of repair died due to low cardiac output and arrhythmia postoperatively. FONTAN et al.¹⁰⁾ stated that the age at the time of repair was a significant determinant of hospital mortality. Their patients³⁾ who were less than 4 years old had a mortality of 50%. Many other authors hypothesized that the optimal age for a Fontan-like operation is more than 4 to 5 years. We are also of the opinion that it is necessary to perform palliative procedures to delay the time of Fontan-like operation until the patient reaches this optimal age.

Dehiscence of the patch closing the atrial septal defect, and, if needed, a right atrioventricular valve and heart block have been two common complications of the technique; both involve severe

hemodynamic upset and often result in death in the early postoperative period. Four of our seven patients had a residual right-to-left shunt at the atrial level postoperatively, but heart block was not found. Some techniques were reported for closing the right atrioventricular valve and preventing complete atrioventricular block, such as 1) approximating the leaflets using pledgeted mattress sutures²²⁾, 2) suturing a patch to the atrial wall 1 to 2 cm above the annulus¹¹⁾.

Intraaortic balloon pumping was performed to increase the cardiac output and coronary perfusion in Patient 4, in whom supraventricular arrhythmia and tachycardia interfered with effective augmentation as shown in the diastolic pressure curve. Moreover, ulceration of the lower leg remained as a result of thrombosis. Following a Fontan-like operation, the major problem is low cardiac output resulting from right heart failure. Therefore, theoretically, it is better to apply the balloon pumping method to the right heart.

In addition, postoperative treatment is of great importance in achieving a favorable outcome. Drainage in order to remove ascites is usually performed from the Douglass' space. However, this drainage becomes ineffective after a few days because of occlusion by the omentum or fibrin.

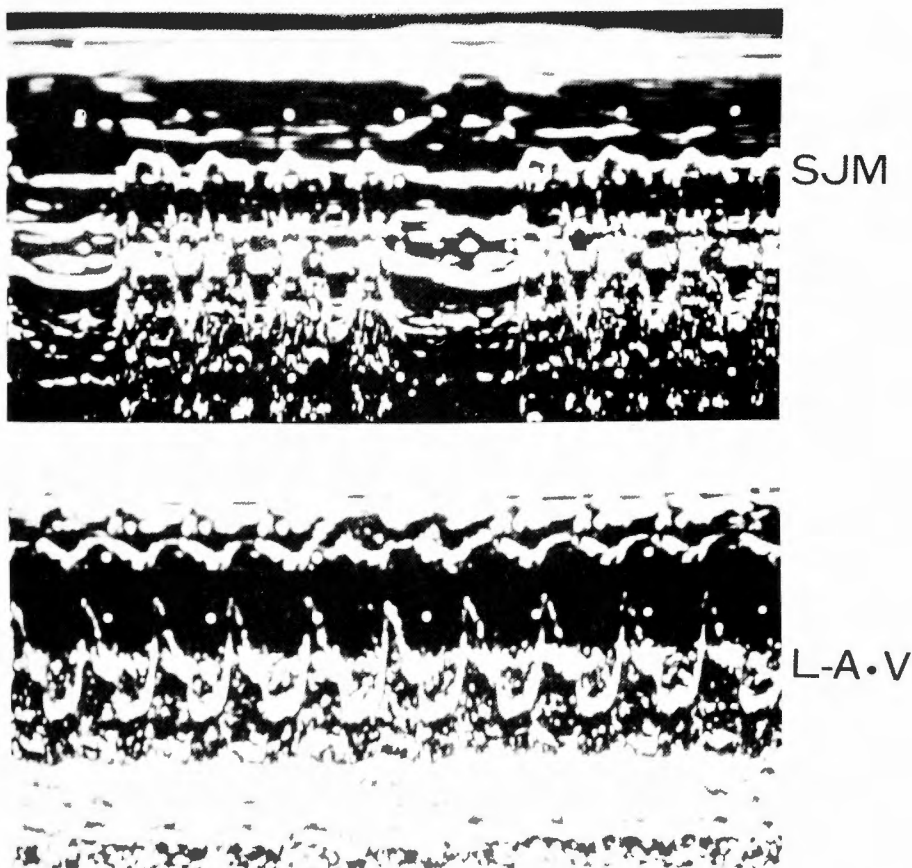


Fig. 8. Postoperative echocardiography (Patient 4)

At present, we insert a drain into the subphrenic space of the right hepatic lobe. This type of drainage is effective in removing ascites for six to seven days. In addition, loss of serum must be replaced to maintain the circulatory volume. In our patients fresh frozen plasma was infused at the same volume with pleural effusion and ascites.

A Fontan-like operation needs suitable respiratory management in the early postoperative period. As the pumping power of the right heart is reduced, negative pressure is necessary to allow the desaturated blood in the right atrium to flow into the lungs. Therefore, in the early postoperative period, positive-pressure ventilation causes a decrease in the pulmonary blood flow. In Patient 4, echocardiography showed closure of the SJM valve in site of anastomosis between the right appendage and the right pulmonary artery during inspiration (Fig. 8). PEEP had even greater adverse effects. SHULLER et al.²⁷⁾ speculated that extubation immediately after surgery, theoretically, reduces the complications caused by increased venous pressure. Also, CORNO et al.³⁾ reported that a relatively smooth postoperative course can be obtained by encouraging early extubation in order to avoid a higher venous pressure resulting from intermittent positive-pressure ventilation.

In the early postoperative period following a Fontan-like operation, overload of pressure and volume to the right atrium leads to supraventricular arrhythmia, such as nodal rhythm, junctional rhythm, paroxysmal atrial tachycardia, atrial fibrillation and premature atrial contraction; occasionally, premature ventricular contraction and ventricular tachycardia also occur. Especially, in cases in which the tricuspid valve or common atrioventricular valve is closed, there is a tendency that these arrhythmias occur. In Patient 7, premature atrial contraction, paroxysmal atrial tachycardia, nodal rhythm and atrial fibrillation were seen during the first 3 postoperative months, especially after physical exercise. Administration of digitalis and verpamil was necessary to suppress these arrhythmias.

Postoperative high venous pressure leads to protein losing syndrome resulting from portal hypertension. In Patient 2, the left phrenic nerve became paralytic, maybe due to cold injury from surface cooling during reoperation, and resulted in elevation of the left diaphragm and an increase in pulmonary arterial resistance. Thus, increase in inferior vena cava pressure might lead to protein losing syndrome. Total serum protein ranged from 3.8 to 4.6 g/dl, and serum albumin was 2.0 to 2.6 g/dl. Moreover, chronic liver dysfunction was observed, with a GOT value of 33-600 U and a GPT value of 22-500 U. An additional operation to shorten the left diaphragm will be necessary in order to decrease pulmonary arterial resistance.

Reoperation was performed in Patients 2 and 7, due to residual shunt at the atrial level. These patients underwent the GLENN operation prior to the Fontan-like operation. Therefore, cannulation into the superior vena cava was performed via the innominate vein. As complete clamping of the superior vena cava was impossible, total perfusion was avoided, and then profound hypothermia combined with surface cooling and core cooling was performed. The intracardiac procedure was performed during total circulatory arrest. In Patient 6, a residual shunt was detected, and reoperation is necessary to alleviate the present cyanosis.

We thought that our high rate of dehiscence around the Dacron patch would be decreased

by the more careful suturing in the large atriotomy. However, many problems, such as prognosis of the patients with increased venous pressure and/or congestive liver dysfunction, post-operative exercise hemodynamics^{23,28)} arteriovenous shunt following a GLENN anastomosis, progressive stenosis or thrombosis in a conduit⁴⁾ and increasing pulmonary resistance still remain to be assessed.

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和文抄録

Fontan 様手術における問題点

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Fontan 様手術を, 三尖弁閉鎖症 4 例, 単心室 3 例に施行した。その内 3 例は, 不整脈, LOS, 低酸素血症で死亡, 生存 4 例の内 3 例は遺残右→左短絡あり, 内 2 例には再手術を施行した。1 例のみ元気に過ごしている。術後の高い静脈圧, 慢性肝機能不全, 低蛋白

血症, 術後の正常下限の低心拍出量, 導管内の狭窄・血栓形成など残された問題も多い。手術適応, 手技, 術後管理上の問題点を, 7 症例の臨床経過をもとに報告する。