－Originals－

Modified Fontan operation

Considerations for the determination of the appropriate procedure

Hitoshi Yamauchi[1)], Yosuke Ishii[1)], Hiroya Omori[1)], Yukiko Takakusaki[1)], Ken-ichi Yamada[1)], Toshimi Yajima[1)], Takashi Nitta[1)], Masami Ochi[1)], Daichi Fukumi[2)], Yukiho Kuramochi[2)], Takashi Okubo[2)] Shun-ichi Ogawa[2)] and Shigeo Tanaka

[1)]Department of Surgery II, Division of Cardiovascular Surgery
and [2)]Department of Pediatrics, Nippon Medical School

Abstract

Background；Although the surgical results of the modified Fontan operation continues to improve, there are various advantages and disadvantages in terms of the post operative condition associated with the Fontan modifications. Late morbidity and mortality are mainly due to arrhythmias, thromboembolic complications, systemic venous hypertension and infective endocarditis. We reported our experience of the modified Fontan operation to determine an appropriate procedure for each patient.

Methods and Results；Seven patients (ranging from the age 1～14 years) underwent a modified Fontan operation including a lateral tunnel (n=1), extracardiac conduit (n=2) and autogenous atrial tunnel (n=4). There was one hospital death due to sepsis in which the patient underwent lateral tunnel procedure. The mean follow up of another six patients was 20 months (ranging from 1～39 months) and all patients were classified as NYHA class I, and remained in normal sinus rhythm without any thromboembolic complications.

Conclusions；When using the autogenous atrial tunnel, there are potential advantages；it is not associated with thromboembolism or endocarditis and has growth potential. However, in high-risk patients with increased pulmonary vascular resistance, impaired ventricular function and pre-operative atrial arrhythmias, it appears reasonable to use an extracardiac conduit. (J Nippon Med Sch 1999；66：28—32)

Key words：modified Fontan operation, extracardiac conduit, autogenous atrial tunnel

Introduction

The Fontan operation was first introduced in 1971 by Fontan and Baudet as a surgical therapy for tricuspid atresia[1], and it has evolved as the definitive palliative surgical treatment for most heart lesions when biventricular circulation is not feasible. Since the short and long-term complications of the surgery have been recognized[2], successive modifications of the original procedure have been introduced. Total cavopulmonary connection was reported in 1988 by de Leval and colleagues[3]. However, this form of right heart bypass requires extensive atrial suture lines which may induce an arrhythmogenic configuration. Thus, repairs using an extracardiac conduit may be considerably important clinically and theoretically. Recently, there are several reports of thrombus formation after the modified Fontan procedure. Considering the matter of postoperative complication, since 1995, we have applied a technique for constructing an autogenous atrial tunnel for small children. Since this technique made it possible to avoid the use of prosthetic or growth-restricted material, it decreased thromboem-

Correspondence to Hitoshi Yamauchi, Department of Surgery II, Division of Cardiovascular Surgery, Nippon Medical School, 1–1–5 Sendagei, Bunkyo-ku, Tokyo 113–8603, Japan

Table 1 Patient characteristics

Case	Age (yr)	Weight (Kg)	Diagnosis	Palliation	Operation	Status
1	6	18	DIRV PS dextrocardia	PA plasty BCPS	AT	alive
2	4	15	DORV hypoplastic RV	(−)	AT	alive
3	2.3	10	DORV noncommited VSD	PA banding BCPS	AT	alive
4	1.5	10	DORV hypoplastic RV	PA banding	AT	alive
5	1.7	12	DIRV PS dextrocardia	BCPS	LT	dead
6	14	43	TA (Ib)	Lt BT shunt original Glenn	EC	alive
7	2.3	9.6	DORV hypoplastic LV	PA banding BCPS	EC	alive

DIRV : double inlet right ventricle, PS : pulmonary stenosis, PA : pulmonary artery, BCPS : bidirectional cavopulmonary shunt, DORV : double outlet right ventricle, TA : tricuspid atresia, BT shunt : Blalock-Taussig shunt, AT : autogenous atrial tunnel, LT : lateral tunnel, EX : extracardiac conduit.

Table 2 Preoperative cardiac catheterization data

Case	RAP(mmHg)	PAP(mmHg)	RpI	PA index
1	9/5	18/10	1.5	170
2	8/4	24/10	1.3	253
3	11/4	17/16	2.1	580
4	10/6	20/10	0.7	550
5	12/10	13/12	0.8	230
6	11/8	Rt 13/11 Lt 22/16	2.5	308
7	8/6	11/9	1.5	294

RAP : RA pressure, PAP : PA pressure, RpI : pulmonary resistance index (unit m^2), PA index : cross-sectional area of the right and left PA/body surface area (mm^2/m^2).

Table 3 Postoperative cardiac catheterization data

Case	SVC	IVC	ICP	PAP mmHg
1	11/9(10)	12/9(10)	12/9(10)	10/6(9)
2	8/6(7)	8/7(7)	10/4(7)	9/4(7)
3	16/15(16)	17/16(17)	17/16(16)	16/15(16)
4	20/17(19)	17/16(16)	17/16(15)	17/16(15)
6	19/18(17)	21/18(19)	21/18(19)	19/17(18)
7	14/13(13)	14/13(13)	14/13(13)	13/11(11)

SVC : superior vena cava, IVC : inferior vena cava, ICP : intra cocnduit pressue, PAP : pulmonary artery pressure, case 5 ; died.

bolism and helped us to avoid the use of anticoagulant therapy. The purpose of this study is to report our experience of modified Fontan operation and to seek for an appropriate procedure for each patient.

Materials and Methods

Patients : From June 1995 to September 1998, seven children (ranging from the age 1 to 14 years) underwent modified Fontan operation. The procedure included a lateral tunnel (n=1), extracardiac conduit (n=2) and autogenous atrial tunnel (n=4). The patient's characteristics are summarized in **Table 1**. Two patients underwent one palliative procedure, and three others underwent a palliative procedure twice prior to the modified Fontan operation. The first surgery was a pulmonary artery banding and the second a bidirectional cavopulmonary shunt performed to protect from progression of the pulmonary hypertension. The original Glenn shunt procedure was performed in one patient. Only one patient did not undergo palliative procedure. The preoperative cardiac catheterization data are listed in **Table 2**.

Surgical techniques : The procedures were performed through a standard median sternotomy incision. After the institution of cardiopulmonary bypass by means of aortic cannulation and individual caval cannulation using angled metal cannulas, cardioplegic standstill was achieved. The operation of the total cavopulmonary connection consists of three parts ; (1) end-to-side anastomosis of the SVC to the undivided right pulmonary artery ; (2) construction of a lateral

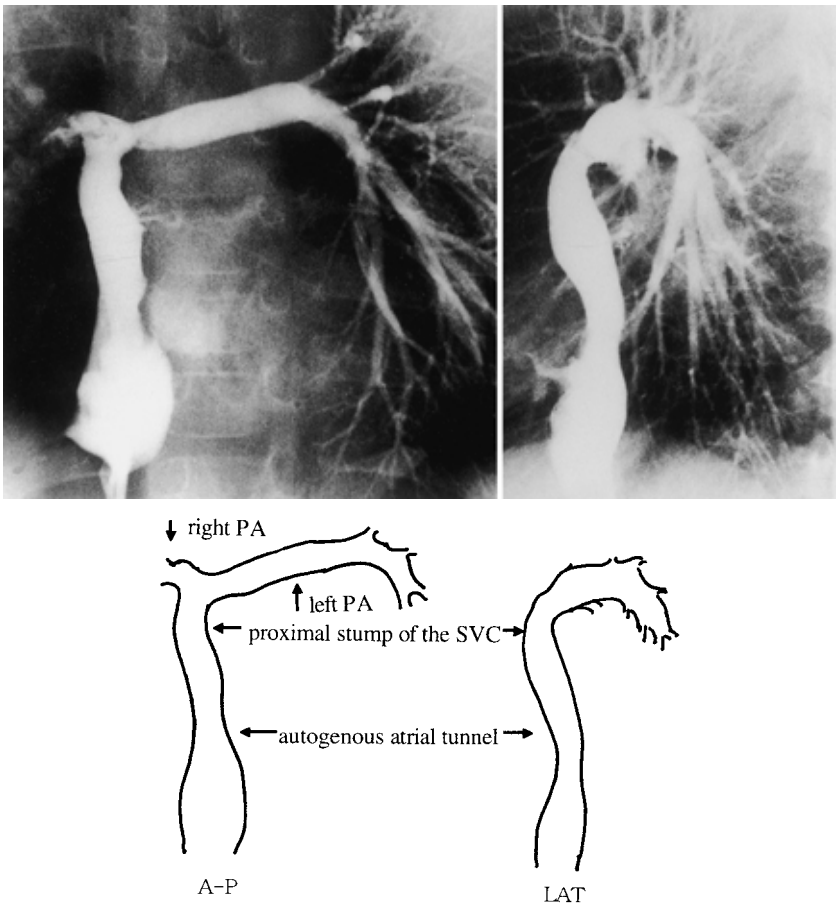

Fig. 1 Postoperative angiograms of case 2
Intra-atrial tunnel is constructed by autogenous atrial flap

tunnel with the use of the posterior wall of the right atrium ; (3) use of a prosthetic patch to channel the IVC to the transected SVC that is anastomosed to the main pulmonary artery. The lateral tunnel suture line originated at the limbus of the fossa ovalis, traveled inferiorly between the coronary sinus and the IVC orifice, around the IVC, laterally up to the crista terminalis, around the SVC orifice, and back to the fossa ovalis. An extracardiac conduit was constructed by an end to end anastomosis between the distal end of the transected IVC and the pulmonary artery with a PTFE graft running suture, and the proximal end of the conduit was then sutured to the bilateral pulmonary arteries with end to side anastomosis. An autogenous atrial tunnel was constructed by suturing the atrial flap around the orifice of the IVC through the base of the coronary sinus, and then to the incised septal edges. There was one case with a small right atrium and counterclockwise rotation that could not construct an autogenous atrial tunnel. Therefore, a heterogenic pericardial patch for intra-atrial tunnel from the IVC to the pulmonary artery was used. In three cases the proximal stump of the SVC was anastomosed to the pulmonary artery, however in two cases after the bidirectional cavopulmonary shunt, we used the right atrial appendage for anastomosis, because it was not feasible to use the adherent proximal stump of the SVC. Weaning from cardiopulmonary bypass was accomplished in the usual manner with minimal inotropic support and use of infusion of vasodilators in all cases.

Cardiac catheterization : Cardiac catheterization was done 1～12 months after operation. The pressures in the atrial tunnel and pulmonary arteries are listed in **Table 3**. Flow in the autogenous atrial tunnel was somewhat pulsatile. The unobstructed flow from the IVC to the pulmonary artery is demonstrated in **Fig. 1** and **2**.

Results : There was one hospital death due to sepsis 40 days after operation which underwent a lateral tunnel procedure using a heterogenic pericardial patch. The mean follow up of the remaining six pa-

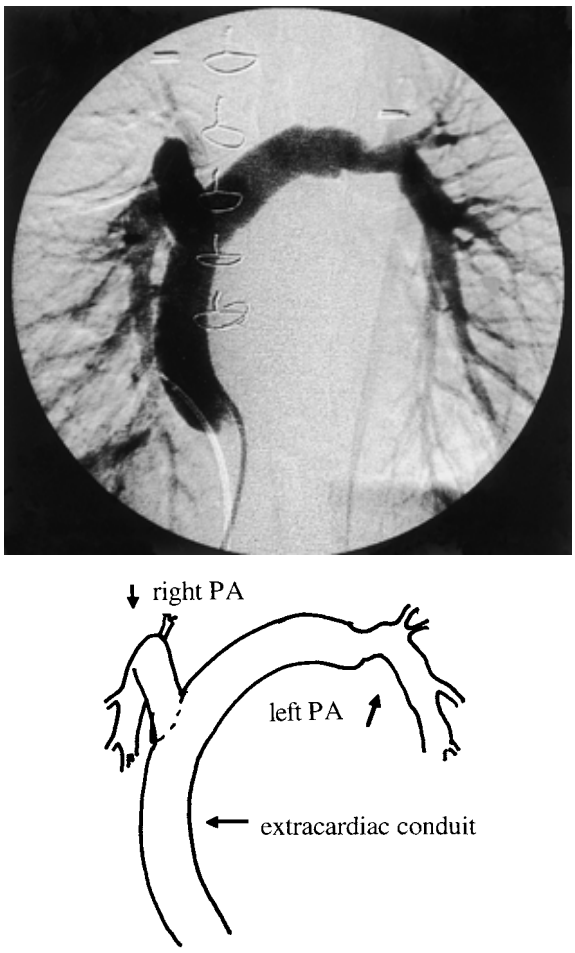

Fig. 2 A post operative angiogram of case 6
Extracardiac conduit is constructed between the distal end of the transected IVC and the bilateral pulmonary arterys with the PTFE graft.

tients was 20 months (ranging from the term 1～39 months), and all patients were classified as NYHA class I and remained in normal sinus rhythm without any thromboembolic complications.

Discussion

Total cavopulmonary connection has been introduced as a rational alternative to atriopulmonary connection for complex Fontan operations. This type of right heart bypass can be achieved by means of an intra-atrial tunnel using prosthetic materials from the inferior vena cava to the pulmonary artery with bidirectional Glenn anastomosis. In addition to the fluid dynamic advantages, according to the Laplace law, proposed by de Leval et al[3], this method may address an important emerging issue after repair of a single ventricle ; that is late atrial arrhythmias. Most forms of total right heart bypass require extensive atrial suture lines which may also induce late arrhythmias[4, 5]. Thus, repairs using extracardiac right heart bypass may involve fluid dynamic advantages and decrease the late arrhythmias by avoiding extensive atrial suture lines[6–8]. Thromboembolic complications of the Fontan operation have been reported since 1978[9]. Total cavopulmonary connection attempts to address these problems by maintaining laminar flow of the venous pathway. Thus, in theory, it decreases the risk of thrombus development within the Fontan circuit. However, Rosenthal and colleagues reported that thromboembolic complications associated with significant morbidity had occurred frequently after the Fontan operation and its modifications[10]. The rate of thrombosis appears to be similar with such modifications as total cavopulmonary connection, atriopulmonary connection and conduit interposition. Furthermore, these reports of thrombus formation suggest that all patients undergoing this procedure require long-term anticoagulant therapy[11–13]. A technique for constructing an autogenous atrial tunnel in small children requiring total cavopulmonary connection has been reported. This technique made it possible to avoid the use of prosthetic or growth-restricted materials[14–16]. The intra-atrial tunnel was constructed by suturing the atrial flap around the orifice of the IVC to the pulmonary artery. The autogenous atrial tunnel ensured a nonturbulent and somewhat pulsatile flow, and therefore this procedure would decrease the risk of the thromboembolic complications and infective endocarditis, giving it a potential benefit for pulmonary circulation in the aspect of pulsatility.

Conclusions

Autogenous atrial tunnels have a potential advantage in aspect to thromboembolic complications, growth potential and endocarditis. However for high-risk patients such as those with increased pulmonary vascular resistance, impaired ventricular function and pre-operative atrial arrhythmias, it appears to be a reasonable option to undergo the extracardiac conduit method.

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(Received, September 7, 1998)
(Accepted for publication, December 4, 1998)