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Evolution of Staged Approach for Fontan Operation

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Abstract

Background: During the early development of the Fontan operation, a number of physiologic and anatomical limits were proposed as selection criteria, and two criteria, pulmonary vascular resistance and ventricular function, have been important in predicting surgical outcome. The use of the bidirectional cavo pulmonary shunt as a staging procedure performed to control the pulmonary blood flow adequately and reduce ventricular volume over load has resulted in marked improvements in the early and late Fontan procedure results.

Methods and results: At our hospital we perform systemic pulmonary shunt or pulmonary artery banding in patients if pulmonary blood flow can not be controlled adequately in the neonatal period and then perform bidirectional cavo pulmonary shunt six months afterwards. During this operation we also performed simultaneous surgical repair for pulmonary artery distortion, anomalies of pulmonary venous connection, restriction of bulboventricular foramen and atrioventricular valve regurgitation. To determine the efficacy of this staged approach in avoiding increases in pulmonary vascular resistance and impaired ventricular function, surgical results were investigated. From February 1995 to May 2001, eighteen patients with cardiac morphology unsuitable for biventricular repair were admitted to our hospital. Twenty-six palliative procedures, were performed including seven pulmonary artery banding, three systemic pulmonary shunt, thirteen bidirectional cavo pulmonary shunt, one original Glenn procedure, four repair of coarctation of the aorta, two total anomalous pulmonary venous connection repair, one mitral valve plasty, and two patients required Damus-Kaye-Stansel procedure to release restrictive bulboventricular foramen. Fifteen patients underwent a modified Fontan operation (total cavopulmonary connection) after these palliative procedures. The operative mortality rate for these palliative procedures was 3.8% (1/26). The operative mortality rate for Fontan operation was 7.1% (1/14). Three patients awaiting the Fontan operation were considered good candidates for a final operation and no patients in this series were considered unsuitable for Fontan completion.

Conclusion: Our strategy of staged approach for Fontan procedure offers a good prognosis.
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Key words: staged Fontan operation, bidirectional cavo pulmonary shunt, total cavopulmonary connection

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Introduction

The Fontan principle of redirecting systemic venous blood into the pulmonary arteries via a lateral tunnel[1] or an extra cardiac conduit[2] has now gained a much wider application than initially intended. Besides tricuspid atresia, the indications for the Fontan procedure have been expanded to include single ventricle and numerous complex of univentricular heart. However, during the early development of the Fontan procedure, the mortality and morbidity remained significant high (12 to 33%); nevertheless a number of physiologic and anatomical limits were proposed as selection criteria including pulmonary artery distortion, pulmonary vascular resistance, pulmonary artery pressure, anomalies of systemic and pulmonary venous connection and impaired ventricular function[3–5]. For high-risk Fontan candidates, the introduction of a staged approach may extend the indications for the Fontan procedure. Our strategies for children who are candidates for the Fontan procedure are summarized as follows. ① Control of the pulmonary blood flow within a favorable range in the neonatal period by placing a systemic-pulmonary shunt or pulmonary artery banding; ② Positive usage of Bidirectional cavo pulmonary shunt as a staging procedure with the repair of associated cardiac anomalies when necessary; and ③ Completion of the Fontan circulation at a younger age. The purpose of this study was to review our experience with the staged Fontan strategy to investigate improvements in the mortality and establishment rate of Fontan operation and in order to determine the most appropriate procedure for individual patients.

Materials and Methods

Patients: From February 1995 to May 2001, eighteen patient with cardiac morphology unsuitable for biventricular repair, underwent twenty-six palliative procedures, and Fontan circulation was established in fourteen patients after these palliative procedure. The patients' characteristics are summarized in **Tables 1 and 2**. Palliative procedure: We performed twenty-six palliative procedures for seventeen patients initially unsuitable for primary Fontan procedure, including seven pulmonary artery banding, three systemic pulmonary shunt, thirteen bidirectional cavo pulmonary shunt, one original Glenn procedure, four repair of coarctation of the aorta, two total anomalous pulmonary venous connection

Table 1 Data of Patients undergoing the Fontan procedure

No.	Sex (year)	Age	Diagnosis	Palliation	Final operation
1	M	4.6	DORV, PS, hypo RV	(−)	AT
2	F	6.0	DIRV, PS, Dextro	BCPS, PA plasty	AT
3	M	2.3	DORV, noncomVSD	PAB/ BCPS	AT
4	F	1.7	DORV, hypo RV	PAB	AT
5	M	14	TA(Ⅰb)	BTS/ original Glenn	EC
6	M	2.3	DORV, hypo RV	PAB/ BCPS	EC
7	M	2.2	TA(Ⅱc), CoA	SFA/ BCPS, DKS	EC
8	F	2.4	MA, hypo LV, CoA	SFA/PA plasty/ BCPS	EC
9	M	2.9	TA(Ⅰb), PLSVC	Bilateral BCPS	EC
10	F	1.8	DORV, MS, CoA	SFA/ re-PAB/ BCPS	EC
11	F	4.1	PS, TS, hypo RV	BCPS/ PA plasty	EC
12	F	2.7	TS, PA, hypoRV	C-shunt / BCPS	EC
13	F	10	DORV, hypo LV	BCPS	EC

DORV: double outlet right ventricle. PS: pulmonary stenosis. hypo RV: hypoplastic right ventricle. DIRV: double inlet right ventricle. Dextro: dextrocardia. BCPS: bidirectional cavopulmonary shunt. PA plasty: pulmonary artery plasty. noncomVSD: noncommitted ventricular septal defect. PAB: pulmonary artery banding. TA: tricusped atresia. BTS: Blalock-Taussig shunt. CoA: coarctation of aorta. SFA: subclavian flap aortoplasty. DKS: Damus-Kaye-Stansel procedure. MA: Mitral atresia. PLSVC: persistant left superior vena cava. TS: tricusped stenosis. AT: autogenous atrial tunnel. EC: extracardiac conduit.

Table 2 Data of Patients awaiting the Fontan operation after palliative procedure

No.	Sex	Age (months)	Diagnosis	Palliation
14	M	4	SA SV TAPVD	TAPVD repair, BCPS
15	F	8	TA PA MR	C-shunt / BCPS, MVP
16	F	38	TA (Ic) CoA	SFA/ BCPS / Lt BTS

PA: pulmonary atresia. C-shunt: central shunt. SA: single atrium. SV: single ventricle. TAPVD: total anomalous pulmonary venous drainage. MR: mitral regurgitation. MVP: Mitral valve plasty.

repair, one mitral valve plasty, and two patients required Damus-Kaye-Stansel procedure to release restrictive bulboventricular foramen. Only one patient did not undergo a palliative procedure. The flow chart for the eighteen study patients is shown in **Fig. 1**.

Final operation: Fourteen patients underwent a modified Fontan operation (total cavopulmonary connection) after palliative procedures. The Fontan procedure included a lateral tunnel (n=1), autogenous atrial tunnel (n=4) and extracardiac conduit (n=9). Postoperative systemic venous hypertension was observed in the two oldest patients (patients 5 and 13, 14 years and 10 years old, respectively). Only one patient (patient 13) required fenestration between the extracardiac conduit and atrium because of high systemic venous pressure.

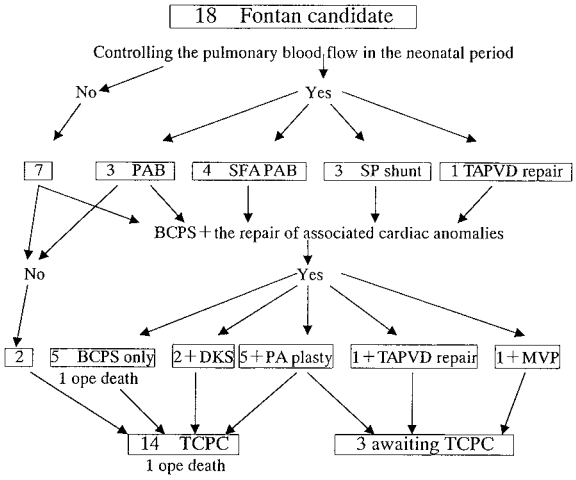

Fig. 1 Flow chart for the eighteen patients

Results

There were two hospital deaths. The patient characteristics and causes of death are summarized in **Table 3**. The establishment rate of Fontan operation was 78% (14/18). The remaining three patients, whose characteristics are summarized in **Table 2**, were considered good candidates for a final operation, and no patients in this series were considered unsuitable for Fontan completion. The operative mortality rate for these palliative procedures was 3.8% (1/26). The operative mortality rate for Fontan operation was 7.1% (1/14). No interm deaths occurred between the palliative procedures and the Fontan operation. One late death occurred four years after final operation because of supra ventricular tachcardia in a patient who underwent autogenous atrial tunnel (patient 3). The overall mortality was 16.7% (3/18)

Discussion

The Fontan operation was first described in 1971 by Fontan and Baudet[6] as a surgical therapy for tricuspid atresia, and it has evolved as the definitive palliative surgical treatment for most heart lesions when biventricular circulation is not feasible. Since its description thirteen years ago, a number of important modifications have been made and the surgical results continue to improve. These modifications can be thought of as variations in staging of the Fontan procedure and variations in the techniques used for anatomical connection of systemic venous circulation to the pulmonary arteries. The establishment of Fontan circulation in a staged fashion can be an alternative surgical strategy in patients in whom some risk factors militate against primary establishment of non-cyanotic circulation. Although there are still no universally accepted new criteria regarding the risk

Table 3 The characteristics and cause of death for patients who died during the post operative hospital stay

No.	Sex	Age (year)	Diagnosis	Palliation /Final operation	cause of death
17	F	1	SA, SV, TAPVD	TAPVD repair / BCPS	arrhythmia
18	M	1.7	DIRV, PS, Dextro	BCPS, PA plasty /LT	sepsis DIC

factors of the Fontan procedure, we consider the following to be risk factors for the completion of Fontan circulation according to previous reports in the literature: age less than one year; a mean pulmonary arterial pressure more than 20 mmHg; a distortion of pulmonary artery; a pulmonary vascular resistance more than 3 wood unit; ventricular ejection fraction less than 40% and Nakata index (cross-sectional area of pulmonary artery/body surface area) less than 200 mm/m^2. Several subsequent studies have reported that the reductions in these risk factors prior to the Fontan procedure by the staged fashion extended the operative indications and improved the mortality[7,8].

A bold and imaginative development, the cavopulmonary anastomosis, appeared to originate in several centers almost simultaneously. Glenn was the first in North America to perform a successful experimental cavopulmpnary shunt, which has become known as the Glenn shunt[9,10]. The original Fontan procedure included a classic superior vena cava-to-right pulmonary (Glenn) shunt. This anastomosis was not essential and was an unnecessary commitment of the larger right pulmonary circulation to the small blood volume of the superior vena caval return, and with the application of the Fontan principle, there has been a reconsideration of the possible benefits of cavopulmonary shunt[11]. Bidirectional cavo pulmonary shunt (BCPS), which connects the superior vena cava to both pulmonary arteries bidirectionaly, provides adequate arterial oxygenation without increasing the ventricular work, avoids deformity of the pulmonary arteries, and prevents an increase in pulmonary vascular resistance. Cardiac output is not limited by pulmonary blood flow, as it is after Fontan operation, because the systemic venous return from the inferior vena cava goes directly into the ventricle, bypassing the lungs[12]. Introduction of the BCPS as a staging procedure resulted in reduced mortality and morbidity for the second stage of the operation, particularly in patients with a combination of several risk factors[13,14].

Recently a high incidence of pulmonary AV malformations has been reported in patients with superior cavopulmonary anastomosis alone as their final procedure[15]. The interruption of hepatic venous return to the pulmonary circulation is considered to potentiate the development of pulmonary AV malformations. However, although BCPS is physiologically similar to the classic Glenn shunt, it has not been shown to cause pulmonary AV malformations as a common complication. One reason for this may be that it is usually performed as part of the staged approach, with most patients proceeding relatively quickly to the incorporation of inferior vena caval flow and hepatic venous flow into the pulmonary circulation, and hence the completion of the Fontan operation[16]. Reports in the literature suggest that the early usage of BCPS as a staging procedure and the completion of Fontan circulation at a younger age could prevent an increase in pulmonary vascular resistance, reduce impairing ventricular function and avoid the development of pulmonary AV malformation.

Although the surgical results of the modified Fontan operation continue to improve, there are various advantages and disadvantages in terms of the postoperative condition associated with the modifications[17]. Late morbidity and mortality are mainly due to arrhythmia, thromboembolic complications, systemic venous hypertension and impaired ventricular function. Total cavopulmonary connection was introduced in 1988 by de Leval et al.[1] as an alternative to atriopulmonary connection for complex Fontan operation, and this type of right heart bypass can be achieved by means of an intra-atrial tunnel using

prosthetic material or autogenous atrial flap from the inferior vena cava to the pulmonary artery with BCPS anastomosis. Most forms of total right heart bypass require an extensive atrial suture line, which may induce late arrhythmias[18,19]. Thus, procedures requiring the use of an extracardiac conduit may involve fluid dynamic advantages and decrease the incidence of late arrhythmia by avoiding an extensive atrial suture line[20,21]. Although autogenous atrial tunnels without the use of prosthetic material have potential advantages in regard to thromboembolic complications, the risk of infective endocarditis and growth potential, the extracardiac conduit method may be preferable in consideration of late arrhythmias[17]. Recent studies have reported that the rate of thrombosis appears to be similar with such modifications to the intra-atrial tunnel, the atriopulmonary connection and the conduit inter position[22]. Furthermore, reports on thrombus formation suggest that all patient undergoing these procedures require long-term anticoagulant therapy[23-25].

Numerous modifications in operative techniques and the evolution of the concept have all helped to improve the operative results of the Fontan procedure. Thus, our final strategy for children who are candidates for the Fontan procedure is as follows. ① Control of the pulmonary blood flow within a favorable range in the neonatal period by placing a systemic-pulmonary shunt or pulmonary artery banding; ② Positive usage of BCPS as a staging procedure with the repair of associated cardiac anomalies when necessary; ③ Completion of the Fontan circulation before the age of 3 years using extracardiac conduit; ④ Long-term anticoagulant therapy after these procedures.

In conclusion, our results suggest that a staged approach for Fontan operation can be safely performed in most patients who otherwise represent a high risk, and we consider that it contributes to a reduction in the preoperative risk for Fontan operation as well as postoperative improvements in the patients' quality of life.

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