

Bidirectional Cavopulmonary Anastomosis

M L Wong,**FAMS, MBBS, M Med (Paed)*, E K W Sim,***FAMS, MBBS, FRCS (Edin, Glas)*, J J Goh,***,
S C Quek,**FAMS, M Med (Paed), DCH (Lond)*, J C L Wong,**FAMS, MBBS, M Med (Paed)*,
W C L Yip,†*FAMS, MRCP, MD (S'pore)*, C N Lee,‡*FAMS, FRCS (Edin, Glas), FRACS*

Abstract

The Fontan operation has been used to palliate patients with a functional single ventricle. In many such patients the operative risk for a Fontan procedure is high. The bidirectional cavopulmonary anastomosis (BCPA) is a useful intermediate palliative procedure before Fontan correction. It may reduce the deleterious sequelae of chronic hypoxaemia and long-term ventricular overload, thus yielding a more suitable Fontan candidate.

We reviewed our experience of 9 patients with univentricular hearts who were at the time of the BCPA not considered suitable candidates for conventional Fontan operation. Two patients had double inlet right ventricle (DIRV) with pulmonary stenosis (PS) and ventricular septal defect (VSD), 2 had univentricular heart, 4 had tricuspid atresia (TA) and hypoplastic right ventricle (RV), 1 patient had atrioventricular septal defect (AVSD) with double outlet right ventricle (DORV) with hypoplastic R V. Two of the 9 patients had bilateral superior vena cavae. All except one had undergone prior palliative operations. The mean age at BCPA was 4.7 years (range 1.5 years to 6 years) and the mean weight at surgery was 15 kg (range 7 to 22 kg). There were no operative deaths. The mean hospitalization stay was 8.5 days (range 5 to 13 days) with a mean follow up of 14 months. Median oxygen saturation improved from $78.7 \pm 6.3\%$ to $85.9 \pm 3.9\%$. The BCPA is a useful palliative procedure to stage Fontan type reconstruction in selected patients with univentricular hearts.

Ann Acad Med Singapore 1999; 28:237-40

Key words: Cardiac surgery, Congenital heart disease, Fontan operation, Paediatrics, Palliative procedure

Introduction

Children with complex congenital heart disease and a functional single ventricle may not be ideal candidates for a Fontan procedure.¹⁻⁴ A staged approach to Fontan's operation has been undertaken in an effort to reduce the volume load of a single ventricle as early as possible and to minimise the impact of potential changes in the ventricular geometry and diastolic function that can accompany a primary Fontan operation.⁵⁻⁹ Bidirectional cavopulmonary anastomosis (BCPA) is a staging procedure that diverts the systemic venous flow from the superior vena cavae (SVC) to both pulmonary arteries via an end-to-side anastomosis between the SVC and the ipsilateral pulmonary artery. It improves systemic arterial oxygen saturation without increasing ventricular work or pulmonary resistance. The procedure has gained wide acceptance as an intermediate-stage palliation pro-

cedure prior to a Fontan operation.⁵⁻¹³ We reviewed our experience in patients who had BCPA performed as a staged approach to a Fontan correction.

Materials and Methods

Between November 1994 and December 1996, 9 patients with a functional single ventricle underwent BCPA. We reviewed the medical and surgical records of these patients and the data included:

- 1) anatomic diagnoses and prior palliative procedures,
- 2) demographic data,
- 3) preoperative echocardiographic and cardiac catheterisation data,
- 4) surgical methods of the BCPA and concomitant procedures,
- 5) postoperative data including systemic arterial satu-

* Senior Lecturer

Department of Paediatrics

** Consultant and Acting Head

Division of Cardiothoracic Surgery

*** Research Assistant

Cardiac Department

National University Hospital

† Consultant Cardiologist

‡ Consultant Cardiac Surgery

Gleneagles Medical Centre, Singapore

Address for Reprints: Dr Eugene Sim, Cardiac Department, National University Hospital, 5 Lower Kent Ridge Road, Singapore 119074.

ration, complications, and total days of postoperative intensive care unit stay, and hospitalisation stay, and

- 6) follow-up data including clinical status, oxygen saturation, and cardiac catheterisation data.

BCPA was used as an intermediate-stage palliation procedure for patients who would be undergoing a Fontan procedure. These patients did not undergo a direct Fontan procedure due to various risk factors. Five patients had distorted and small pulmonary arteries (PA), 2 patients had moderately raised pulmonary resistance, and in 2 patients there was atrioventricular valve regurgitation (AVVR).

There were six boys and three girls. Two patients had double inlet right ventricle (DIRV) with pulmonary stenosis (PS) and ventricular septal defect (VSD). Two others had univentricular heart, 4 had tricuspid atresia (TA) and hypoplastic right ventricle (RV), 1 patient had atrioventricular septal defect (AVSD) with double outlet right ventricle (DORV) with hypoplastic RV (Table I). Two of the 9 patients had bilateral superior vena cavae (SVC). All except 1 had undergone prior palliative operations. Five patients had unilateral Blalock-Taussig (BT) shunts and 2 patients had bilateral BT shunts. Three patients had atrial septectomy.

The mean age and weight at BCPA were 4.7 years (range 1.8 to 6 years) and 15 kg (range 7 to 22 kg), respectively. Median haemoglobin and haematocrit level

before the BCPA were 16.7 ± 1.59 g/dl and $48.6 \pm 11\%$, respectively. The median oxygen saturation was $78.7 \pm 6.26\%$.

Surgical Techniques

Median sternotomy was utilised and cavopulmonary anastomosis operation was performed under cardiopulmonary bypass (CPB) in all patients. The SVC was cannulated at its junction with the innominate vein. Mobilisation of the SVC and division of the azygos vein were performed. The pulmonary arteries were freed from behind the aorta and a longitudinal incision was made in the anterior aspect of the right and/or left pulmonary artery(ies). Care was taken to avoid twisting the SVC during the anastomosis. The anastomosis was performed using a single layer of running 7/0 prolene for the posterior layer and interrupted sutures for the anterior layer. In patients with bilateral SVC both SVC were cannulated and the anastomosis was constructed from the left SVC to left PA, again avoiding any twisting of the left SVC. In the patient who underwent concomitant atrial septectomy, aortic root cardioplegia was delivered and the septectomy was performed through a right atriotomy prior to the construction of the BCPA.

During the operation, additional procedures were performed in 7 of them. They included 4 right and 2 left Blalock-Taussig (BT) shunt ligation, 2 patent ductus arteriosus (PDA) ligation, one pulmonary arterioplasty and one atrial septectomy (Table I).

TABLE I: SUMMARY OF PATIENTS' DATA AT THE TIME OF BIDIRECTIONAL CAVOPULMONARY SHUNT

Patient no.	Age (years)	Anatomic diagnosis	Prior operations	Date of BCPS	Concomitant operation
1	5 4/12	DIRV, PS, VSD, hypoplastic LV, atrial situs solitus	R BTS, L BTS	10/2/95	R BTS ligation
2	5 6/12	DIRV, PS, PDA, ASD, VSD, situs solitus	Atrial septectomy	10/3/95	L BTS ligation, PDA ligation
3	4 8/12	ASD, DOV, PS, PDA, dextrocardia, situs inversus RA isomerism, bilateral SVC	R BTS	13/12/95	R BTS ligation, atrial septectomy
4	4 1/12	AVSD, DORV, PS, hypoplastic RV, PDA with collaterals, atrial situs solitus, levocardia		10/7/95	PDA ligation
5	4 8/12	ASD, VSD, TA, situs solitus, hypoplastic RV, PFO, closing PDA	BAS, R BTS	27/12/96	R BTS ligation
6	1 10/12	AVSD, RA isomerism, bilateral SVC	R BTS	16/10/96	R BTS ligation
7	5 9/12	DOLV, P'S, TA, hypoplastic RV	BAS, L BTS, dental extraction	6/12/96	L BTS ligation, L pulmonary arterioplasty, ligation of pulmonary artery
8	3 9/12	PA, VSD, TA, hypoplastic RV, levocardia	R BTS, atrial septectomy, dental extraction	28/9/95	R BTS ligation
9	4 2/12	IVS, PFO, TA, hypoplastic RV, hypoplastic PA	Atrial septectomy, R BTS, L BTS	26/12/94	R BTS ligation

Age: age at BCPS operation; ASD: atrial septal defect; AVSD: atrioventricular septal defect; BAS: balloon atrial septostomy; DIRV: double inlet right ventricle; DORV: double outlet right ventricle; DOLV: double outlet left ventricle; IVS: intact ventricular septum; PA: pulmonary atresia; PDA: patent ductus arteriosus; PFO: patent foramen ovale; PS: pulmonary stenosis; RV: right ventricle; LV: left ventricle; R BTS: right modified Blalock-Taussig shunt; L BTS: left modified Blalock-Taussig shunt; TA: tricuspid atresia; VSD: ventricular septal defect

Results

The patients have been followed up for a period of 1 to 23 months (mean 14 months). The early postoperative course was smooth in all patients and congestive heart failure was not encountered in any patient. There was neither operative mortality nor major morbidity. No further arrhythmia occurred and there were no neurologic complications, stenoses or pulmonary vascular obstructive problems. Four of the patients had mild atrioventricular valve regurgitation in the early postoperative stage. Patient 5 was admitted a month after the BCPA operation for a pericardial effusion. She was treated conservatively and the problem was resolved with anti-inflammatory medications one month later. The mean duration of stay in the intensive care unit was 7 days, and the mean hospitalisation stay was 8.5 days. There was improvement in the median haemoglobin level and systemic arterial oxygen saturation level from $78.7 \pm 6.3\%$ to $85.9 \pm 3.9\%$ (Fig. 1). The median haemoglobin level post-BCPA was 14.8 ± 0.99 g/dl compared to the preoperative value of 16.7 ± 1.59 g/dl. Three patients underwent follow-up catheterisation and all the patients underwent follow-up echocardiography. These procedures confirmed the excellent bilateral pulmonary blood flow and the wide patency of the anastomosis.

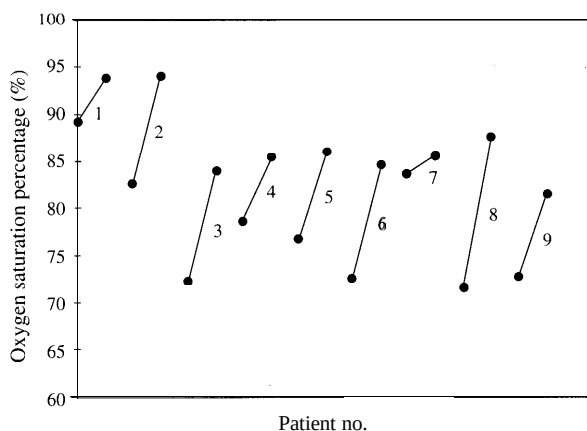


Fig 1. Preoperative oxygen saturation percentage versus postoperative oxygen saturation percentage.

Discussion

The original cavopulmonary, or Glenn shunt has been used to palliate a variety of congenital heart disease with excellent relief of cyanosis.⁹ The first successful Fontan operation was performed in 3 patients with tricuspid atresia in 1968 and reported in 1971. In the early years, operative mortality rate ranged from 11% to 15% and it has decreased to 3.7% in more recent years.^{14,15} The

Fontan operation, when applied to other forms of univentricular heart diseases, yields less encouraging results. Kirklin et al² reported a mortality rate of 13% for patients with tricuspid atresia compared to a 29% rate for patients with other forms of congenital heart diseases.

Norwood et al^{5,16} contended that mortality could be reduced by staging patients with BCPA prior to the completion of Fontan operation. They reported a mortality of 7% to 8% in 158 patients who underwent staging and this figure was substantially lower than the 16% for a comparative cohort ($P < 0.05$). Experience reported from other institutions also showed a lower mortality in patients who underwent BCPA. Nine tricuspid atresia patients underwent BCPA in 1972 and Azzolina et al¹⁷ reported a mortality rate of 33%. Another 19 had BCPA reported by Abrams with no mortality.⁴ Only one death was reported by Mazzer et al in 1989¹⁰ after performing the procedure in 18 patients.

Although it was felt initially that the BCPA should only be performed on older children, the operation has been increasingly performed on children at a much younger age. Albanese et al¹⁹ had reported their experience with 27 patients who were under two years of age. Early mortality rate was 15% and late mortality rate was 8.7%.

In a recent report, Chang et al²⁰ had also shown that the BCPA procedure can be safely performed with good results in 17 infants under six months of age. The mortality rate was 6% and there were no late deaths. Apart from the above reports, Reddy et al²¹ had also reviewed their experience in a series of 9 infants who underwent early primary BCPA for palliation in a variety of defects with a single functional ventricle. There were no early deaths but 2 patients died due to abdominal malformations of asplenia syndrome and hypoxia respectively in the medium-term follow-up.

Clinical and functional information from our experience confirms the safety BCPA and its efficacy in providing adequate oxygenation and in preparing patients with complex cardiac anomalies for a subsequent Fontan operation with minimal or no pulmonary arterial distortion. The BCPA is a useful palliative procedure in staging to an ultimate Fontan operation. The advantages of the BCPA procedure are:

- 1) because the anastomosis is formed in an end-to-side fashion, it allows the caliber of the superior vena cava, rather than that of the ipsilateral pulmonary artery (which may be hypoplastic), to define the size of the anastomosis hence it avoids volume overload of the single ventricle and prevents pulmonary distortion,
- 2) it prevents the development of pulmonary vascular obstructive disease, and

- 3) it leaves the right and left pulmonary arteries in continuity. The BCPA provides excellent palliation for selected patients with congenital heart disease and it is followed by good intermediate term palliation in complex conditions that are less ideal for a one stage Fontan procedure.

REFERENCES

- Fontan F, Kirklin J W, Fernandez G, Costa F, Naftel DC, Tritto F, et al. Outcome after a "Perfect" Fontan operation. *Circulation* 1990; 81:1520-36.
- Kirklin J K, Blackstone E H, Kirklin J W, Pacifico A D, Barger L M. The Fontan operation: Ventricular hypertrophy, age and date of operation as risk factors. *J Thorac Cardiovasc Surg* 1986; 92:1049-64.
- Matsuda H, Kawashima Y, Kishimoto H, Hirose H, Nakano S, Kato H, et al. Problems in the modified Fontan operation for univentricular heart of the right ventricular type. *Circulation* 1987; 76(3 Suppl):45-52.
- Lamberti J J, Mainwaring R D, Spicer R L, Uzark K C, Moore J W. Factors influencing perioperative morbidity during palliation of the univentricular heart. *Ann Thorac Surg* 1995; 60(6 Suppl):S550-3.
- Norwood W I, Jacobs M L. Fontan's procedure in two stages. *Am J Surg* 1993; 166:548-51.
- Lamberti J J, Spicer R L, Waldman J D, Grehl T M, Thomson D, George L, et al. The bidirectional cavopulmonary shunt. *J Thorac Cardiovasc Surg* 1990; 100:22-30.
- Bridges N D, Jonas R A, Mayer J E Jr, Flanagan M F, Keane J F, Castaneda A R. Bidirectional cavopulmonary anastomosis as interim palliation for high-risk Fontan candidates: early results. *Circulation* 1990; 82(4 Suppl):170-6.
- Pridjian A K, Mendelsohn A M, Lupinetti F M, Beekman III R H, MacDonald D II, Serwer G, et al. Usefulness of the bidirectional Glenn procedure as staged reconstruction for the functional single ventricle. *Am J Cardiol* 1993; 71:959-62.
- Hopkins R A, Armstrong B E, Serwer G A, Peterson R J, Oldham N H. Physiological rationale for a bidirectional cavopulmonary shunt. *J Thorac Cardiovasc Surg* 1985; 90:391-8.
- Mazzera E, Corno A, Picardo S, Donato R D, Marino B, Costa D, et al. Bidirectional cavopulmonary shunts: clinical applications as staged or definitive palliation. *Ann Thorac Surg* 1989; 47:415-20.
- Hawkins J A, Shaddy R E, Day R W, Sturtevant J E, Orsmond G S, McGough E C. Mid-term results after bidirectional cavopulmonary shunts. *Ann Thorac Surg* 1993; 56:833-7.
- Vargas F J, Mengo G, Gallo J P, Skerl C B, Ranzini M E, Vazquez J C. Bidirectional cavopulmonary shunt in patients with multiple risk factors. *Ann Thorac Surg* 1995; 60:S558-62.
- Van Son J A M, Falk V. Developments in surgical palliation of univentricular physiology [letter]. *Ann Thorac Surg* 1997; 63:301-3.
- Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971; 26:240-8.
- Fontan F, Deville C, Quaegebeur J, Ottenkamp J, Sourdille N, Choussat A, et al. Repair of tricuspid atresia in 100 patients. *J Thorac Cardiovasc Surg* 1983; 85:647-60.
- Jacobs M L, Norwood W I. Fontan operation: Influence of modifications on morbidity and mortality. *Ann Thorac Surg* 1994; 58:945-52.
- Azzolina G, Eufraate S, Pensa P. Tricuspid atresia: Experience in surgical management with a modified cavopulmonary anastomosis. *Thorax* 1972; 27:111-5.
- Salmon A I, Sethia B, Silove E D, Goh D, Mitchell I, Alton H, et al. Cavopulmonary anastomosis as long-term palliation for patients with tricuspid atresia. *Eur J Cardiothorac Surg* 1989; 3:494-7, discussion 497-8.
- Albanese S B, Carotti A, Di Donato R M, Mazzera E, Troconis CJ, Giannico S, et al. Bidirectional cavopulmonary anastomosis in patients under two years of age. *J Thorac Cardiovasc Surg* 1992; 104:904-9.
- Chang A C, Hanley F L, Wernovsky G, Rosenfeld H M, Wessel D L, Jonas R A, et al. Early bidirectional cavopulmonary shunt in young infants. Postoperative course and early results. *Circulation* 1993; 88:II149-58.
- Reddy V M, Liddicoat J R, Hanley F L. Primary bidirectional superior cavopulmonary shunt in infants between 1 and 4 months of age. *Ann Thorac Surg* 1995; 59:1126.