Cardiol Young 2004; 14 (Suppl. 3): 57–61

ISSN 1047-9511

Original Article

Should we attempt functionally univentricular or partial biventricular repair for patients with complex congenital cardiac anomalies and hypoplasia of the subpulmonary ventricle?

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Keywords: Fontan procedure; one and a half ventricle repair; functionally univentricular heart

HYPOPLASIA, WITH OR WITHOUT DYSPLASIA, OF the subpulmonary ventricle is found in association with a wide spectrum of complex congenital cardiac anomalies.[1–7] In these cardiac defects, the systemic ventricle is usually normal, while the subpulmonary one, usually of right morphology, is incapable of supporting the entire flow of blood to the lungs.[1] There is now an alternative to the more classical functionally univentricular type of repair, which can be achieved by means of the total cavopulmonary connection, namely the partial biventricular, or the so called "one and a half ventricle" repair. In selected cases, by reducing the preload on the subpulmonary ventricle by construction of a bi-directional cavo-pulmonary shunt, it is possible to achieve complete separation of the pulmonary and systemic circulations, whilst still maintaining pulsatile flow of blood to the lungs.[2–5] In this review, we describe our experience with the "one-and-a-half" ventricular option, analysing the role of preoperative evaluation of the subpulmonary ventricle, and describing our results over the short and intermediate term.

Material and methods

Since August 1993, we have constructed a bi-directional cavopulmonary shunt in 19 patients with hypoplasia and or dysplasia of the subpulmonary ventricle. Their median age was 8 months, with a range from 4 months to 35 years. All the patients were studied preoperatively using cross-sectional echocardiography and Doppler interrogation. Cardiac catheterization was also performed in all prior to construction of the shunt, with the main purpose of measuring the mean right atrial and pulmonary arterial pressures, and calculating the pulmonary vascular resistances (Table 1). The diagnoses included pulmonary atresia and intact ventricular septum in 8 patients, Ebstein's malformation of the tricuspid valve in 3 patients, left isomerism with an inlet ventricular septal defect, and straddling of the tricuspid valve, in 2 each, and congenitally corrected transposition, atrioventricular septal defect with

Table 1. Functionally univentricular repair.

Patients	Age (year)	Diagnosis	TV Z-value	RV volume (%)	PVR
#1	4.5	PA-IVS	−7.8	32	1.7
#2	3.5	PA-IVS	−8.2	29	1.9
#3	1.5	PA-IVS	−7.1	34	2.1
#4	2	PA-IVS	−9.0	25	1.9
#5	3	PA-IVS	−7.4	35	1.9
#6	3	right hysomerism + CAVC	−7.8	29	1.7

Abbreviations: AVSD: atrio-ventricular septal defect; PA-IVS: pulmonary atresia-intact ventricular septum; PVR: pulmonary vascular resistance; RV: right ventricle; TV: tricuspid valve

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Accepted for publication January 2004

Table 2. Partial biventricular repair.

Patients	Age	Diagnosis	TV Z-value	RV volume (%)	PVR
#1	7 m	CAVC + TOF	−6.2	44	1.8
#2	3.5 y	PA-IVS	−4.3	52	1.4
#3	17 y	Ebstein anomaly	–	47	1.6
#4	11 m	Heterotaxy sdr. + VSD + anomalous systemic venous return	−2.2	67	1.9
#5	35 y	Ebstein anomaly	–	43	1.8
#6	1.1 y	VSD + TV straddling and overriding	−2.6	60	2.5
#7	3 y	Truncus arteriosus A1 (Van Praagh)	−3.3	56	3.1
#8	3.5 y	VSD + TV straddling	−4.3	50	2.2
#9	6 y	Ebstein anomaly	–	49	2.1
#10	9 m	TAPVD	−3.2	55	1.8
#11	4 y	PA-IVS	−4.2	48	1.9

Abbreviations: AVSD: atrio-ventricular septal defect; m: months; PA-IVS: pulmonary atresia-intact ventricular septum; PVR: pulmonary vascular resistance; RV: right ventricle; TAPVC: total anomalous venous pulmonary connection; TOF: tetralogy of Fallot; TV: tricuspid valve; VSD: ventricular septal defect; y: years

Table 3. Patients still waiting for repair.

Patients	Age (month)	Diagnosis	TV Z-value	RV volume (%)	PVR
#1	7	PA-IVS	−6.9	40	1.9
#2	13	c-TGA	−3.2	59	2.3

Abbreviations: c-TGA: corrected transposition of the great arteries; PA-IVS: pulmonary atresia-intact ventricular septum; PVR: pulmonary vascular resistance; RV: right ventricle; TV: tricuspid valve

common atrioventricular valvar orifice and tetralogy of Fallot, common arterial trunk, and totally anomalous pulmonary venous connection in the final four patients (Tables 1–3). Of the patients, 14 (67%) had required surgical manoeuvres prior to construction of the bi-directional cavopulmonary shunt, involving construction of a modified Blalock–Taussig shunt in 9 patients, banding of the pulmonary trunk in 4, reconstruction of the right ventricular outflow tract with an autologous pericardial patch in 2, pulmonary valvotomy in 1, construction of a modified right Blalock–Taussig shunt in association with separation of the pulmonary arteries from a common trunk in 1, plasty of the tricuspid valve in 1, atrial septectomy in 1, and closure of a persistently patent arterial duct in 1 (Table 4). In 1 patient with Ebstein's malformation and recurrent supraventricular tachycardia, specifically the third patient listed in Table 2, an accessory muscular atrioventricular connection had been successfully ablated percutaneously using radiofequency energy 6 years prior to the repair.

Table 4. Surgery previous to the bi-directional cavopulmonary shunt (mainly to increase or decrease pulmonary blood flow).

	Patients
• Modified B-T shunt	10
• MPA banding	6
• Atrioseptectomy	2
• RVOT reconstruction	1
• TV plasty	1
• MPA detachment from common trunk	1
• PDA closure	1
• Pulmonary valvectomy	1
• MPA closure	1

Abbreviations: B-T: Blalock–Taussig; MPA: main pulmonary artery; PDA: patent ductus arteriosus; RVOT: right ventricle outflow tract; TV: tricuspid valve

Preoperative assessment of the right ventricle

We assessed the structure of the tricuspid valve, and the volume of the subpulmonary ventricle, in 16 patients prior to repair. We excluded from assessment the 3 patients with Ebstein's malformation (Tables 1 and 2). The tricuspid valve was quantified using the largest annular diameter obtained using a standard apical four chamber echocardiographic view. This was indexed against body surface area, and then compared to the normal values collated by Rowlatt et al.,[8] thus permitting calculation of the Z-score. The end diastolic volume of the right ventricle was assessed in all prior to repair, using a modification of the method of Tomita et al.[9]

Surgical procedures performed in association with construction of the bidirectional cavopulmonary shunt

The manoeuvres included take-down of the Blalock–Taussig shunt in 10 patients, atrial septectomy in 5 patients, and closure of a ventricular septal defect in 4 patients. We reconstructed the outflow tract of the right ventricle in 4 patients, performed plasty of the tricuspid valve in 3 patients, ligated the pulmonary trunk in 3 patients, carried out plasty of the branches of the pulmonary trunk in 3 patients, constructed a bi-directional cavopulmonary shunt bilaterally in 2 patients, corrected totally anomalous pulmonary venous connection in 1, closed the tricuspid valve in 2, corrected an atrioventricular septal defect associated with tetralogy of Fallot in 1, partial closed the tricuspid valve in 1, and created a tunnel from the hepatic

https://doi.org/10.1017/S1047951104006584 Published online by Cambridge University Press

Table 5. Surgery at the time of bi-directional cavopulmonary shunt.

	Patients
• B-T take-down	10
• Atrial septectomy	5
• VSD closure	4
• RVOT reconstruction	4
• TV plasty	3
• MPA ligation	3
• PA branches plasty	3
• TV closure	2
• TAPVD repair	1
• CAVC + TOF repair	1
• Partial TV closure	1
• Hepatic veins–left sided atrium diversion	1

Abbreviations: AVSD: atrio-ventricular septal defect; B-T: Blalock–Taussig; MPA: main pulmonary artery; PA: pulmonary artresia; RVOT: right ventricle outflow tract; TOF: tetralogy of Fallot; TAVPC: total anomalous venous pulmonary connetion; TV: tricuspid valve; VSD: ventricular septal defect

veins to the left-sided atrium in the final patient (Table 5). When performing the bi-directional cavopulmonary shunt as a staging procedure towards the Fontan operation, we also interrupted any additional source of antegrade flow of blood to the lungs. In 6 patients we finally performed a functionally univentricular repair (Table 1), but we undertook a partial biventricular repair in 11 (Table 2), while 2 patients are still waiting their final repair (Table 3).

Functionally univentricular repair

In 6 patients (Table 1), having re-assessed the potential of the subpulmonary ventricle, we proceeded to a functionally univentricular correction. The median age at repair was 3 years, with a range from 1.5 to 4.5 years. Assessment had revealed severe hypoplasia or dysfunction of the subpulmonary ventricle, with a Z-score calculated at less than −7 in all. The estimated end-diastolic ventricular volume was below 40% of normal values in all. The total cavopulmonary connection was achieved using an intracardiac tunnel in 5, and an extracardiac conduit in 1. Fenestration was performed in all cases. There were no hospital deaths or reoperations, and all patients were discharged home in good hemodynamic condition, and in sinus rhythm, receiving oral anticoagulation for 6 months, maintaining the international rating between 2 and 3. As assessed clinically and echocardiographically, they are all currently in good general condition and asymptomatic, receiving acetyl salycylic acid orally at a dose of 10 milligrams per kilogram of body weight.

Partial biventricular repair

In 11 patients (Table 2), we constructed a biventricular cavopulmonary connection in addition to complete intracardiac septation and repair of the intracardiac anomalies. This was achieved as a primary procedure in 9 patients, while in the 7th and 8th patients listed in Table 2, we were unsure at the time of construction of the shunt whether to proceed to a total cavopulmonary connection or a partial biventricular repair. At a later stage, therefore, we re-examined the parameters of the subpulmonary ventricle, and then proceeded to a partial biventricular correction.

The median age at repair was 3 years, with a range from 6 months to 35 years. We judged the subpulmonary ventricle capable of coping with the reduced volume load based on Z-scores for the tricuspid valve calculated at between −2.5 and −6.2, with a mean score of −3.4, and on the end-diastolic volume of the subpulmonary ventricle being between 44% and 67% of normal, with a mean of 54%. Pulmonary resistances were calculated at less than 2 units per meter squared in 10 patients. In the seventh patient listed in Table 2, who underwent separation of the pulmonary arteries from a common arterial trunk and construction of a Blalock–Taussig shunt, the pulmonary vascular resistances were found to be 3 units per meter squared. An interatrial communication was closed in most patients at the time of the partial biventricular repair, while it was left open in the 11th patient listed in Table 2. A large atrial septal defect was reduced to 4 millimeters in diameter by inserting a fenestrated patch in the seventh patient listed in Table 2, who had elevated pulmonary arterial resistances. In all the patients, we monitored continuously the mean right atrial and superior caval venous pressures in the immediate postoperative course, for periods of between 48 and 192 hours, with a mean of 61 hours, using temporary transjugular and transthoracic catheters. The right atrial pressures ranged from 6 to 11 millimeters of mercury, with a mean of 7 millimeters of mercury, and the mean superior caval venous pressure ranged from 12 to 16 millimeters of mercury, with a mean of 14 millimeters. There were no major complications in the post-operative course, apart from the seventh patient listed in Table 2, who had increased pulmonary vascular resistances, and was treated for 8 days in the intensive care unit because of low output and superior caval venous syndromes. There were no hospital deaths, and no reinterventions. All patients were discharged home in good hemodynamic condition, and free from oral anticoagulation therapy.

We have followed all of them after discharge with periodical clinical and echocardiographic assessments for periods of 8 months to 7.3 years, with a mean of 29.8 months. There have been no late deaths and no reoperations. Of the patients, 7 were in the first grading of the classification of the New York Heart Association, with 1 in the second class, this being the seventh

https://doi.org/10.1017/S1047951104006584 Published online by Cambridge University Press

patient listed in Table 2. Echocardiographic examination has shown good left ventricular function, absent or mild tricuspid and pulmonary valvar regurgitation, with pulsatile antegrade flow across the pulmonary valve in all. Doppler evaluation of the superior caval vein showed, in all, a systolic reversal of flow simultaneous to ejection from the right ventricle, and antegrade flow during the rest of the cardiac cycle. Interrogation of the inferior caval vein disclosed normal patterns, with low amplitude reverse flow during atrial systole, and antegrade flow during the rest of the cardiac cycle. Interventional cardiac catheterization was performed in 3 patients, 4 months, 7.4 months and 5 years after correction, for treatment of residual lesions. Percutaneous balloon dilation of the pulmonary trunk was performed in the 1st and 8th patients listed in Table 2, after reconstruction of the previously ligated trunk, and the left pulmonary artery was dilated in the 7th patient listed in Table 2, the procedure being repeated 10 months and 14 months after repair, because of hypoplasia after its separation from a common arterial trunk. The mean right atrial pressure was found to be between 6 and 7 millimeters of mercury, compared to the pressure in the superior caval vein, which was 12, 14, and 24 millimeters of mercury, this last value being found in the 7th patient of Table 2, who had elevated pulmonary vascular resistances. In this patient, the atrial septal defect was closed using an Amplatzer device 35 months after surgical correction, producing complete intracardiac septation. Exercise stress testing was performed in two adults, 39 months and 19 months, respectively, after repair. They had had moderately reduced exercise tolerance preoperatively, but the tests were now within normal limits, the test being interrupted at 125 watts because of exhaustion in both of them. No arrhythmias were detected during the entire examination.

Discussion

Surgery for complex cardiac anomalies with two ventricles and two atrioventricular valves, when associated with hypoplasia or dysplasia of the subpulmonary ventricle, still represents a challenging situation worthy of debate amongst paediatric cardiac surgeons.[1–7] The main goal of repair is to permit the small subpulmonary ventricle to function as a pumping chamber, and thus provide antegrade and pulsatile flow of blood to the lungs. If this situation is achieved, then the normal, or close to normal, pressures in the territory drained by the inferior caval vein should avoid most of the life-threatening complications linked to the Fontan circulation, such as chronic hepatic dysfunction, protein-loosing enteropaty, plastic bronchitis, and so on.[10,11]

In order to achieve optimal surgical results, it is essential properly to select the appropriate operation for the individual patient. Thus, we believe it is essential to assess meticulously the structure and function of the subpulmonary ventricle, evaluating its capability of supporting at least two-thirds of the systemic venous return 1. The end diastolic ventricular volume and the Z-score for the tricuspid valve are, in our opinion, the most important parameters with which to assess the size of the ventricle and its potential pumping capabilities. We have found that, in the presence of a "good" volume in the subpulmonary ventricle, the Z-score for the tricuspid valve can be as low as −6, and yet still it is possible to achieve successful repair. According to our experience, nonetheless, patients having a Z-score lower than −7, and a ventricular volume lower than 40% of normal, need to be palliated using a functionally univentricular type of repair.

In 2 of our patients, the 7th and 8th as listed in Table 2, we constructed initially only a bi-directional cavopulmonary shunt, with the aim of deferring the partial biventricular repair because of the borderline nature of the indications at the time of construction of the bidirectional shunt. In 1 of these patients, the 7th in Table 2, repeated cardiac catheterization showed that the pulmonary vascular resistances were too high, at 3.1 units per meter squared, to permit completion of the Fontan circulation. As an alternative to transplantation of the heart and lungs, we successfully achieved a partial biventricular repair 6 months later. In the 8th patient as listed in Table 2, after initial banding of the pulmonary trunk because of significant straddling of the tricuspid valve, a detailed re-evaluation of the parameters measured for the subpulmonary ventricle, together with the anatomy of the straddling valve, convinced us to attempt ventricular septation. This was achieved without any residual ventricular shunting, and only mild incompetence of the tricuspid valve was found after correction.

Of our patients, 3 presented with severe Ebstein's malformation, and were severely cyanotic. Having unloaded the hypoplastic and dysplastic subpulmonary ventricle by means of a bi-directional cavopulmonary shunt, we were able to perform aggressive reconstruction of the tricuspid valve, achieving a competent valve at the expense of a reduced annular diameter for the weight of the patient. It also proved possible to close the atrial septal defect. In 1 patient with pulmonary atresia and intact ventricular septum, after an initial pulmonary valvotomy and construction of a central systemic-to-pulmonary arterial shunt, we performed a bidirectional cavopulmonary anastomosis together with reconstruction of the right ventricular outflow tract and overhaul of the right ventricle as described by Pawade et al.[12] The overhaul consisted of

commissurotomy of the tricuspid valve and splitting of the papillary muscles. This permitted better movement of the tensor apparatus, improving the excursion of the leaflets, and hence the performance of the valve.

We conclude, therefore, that partial biventricular repair represents a valid surgical therapy with which to separate the pulmonary and systemic circulations whilst maintaining pulsatile flow of blood to the lungs and, at the some time, a low pressure in the territories drained by the inferior caval vein.

We are convinced that, in dealing with complex congenital cardiac malformations where the subpulmonary ventricle is hypoplastic, it is crucial to make a meticulous assessment of the potential resources of the underdeveloped ventricle. Only in this way is it possible to offer the patient the most physiological correction possible.

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