Long-term functional results of the one and one half ventricular repair for the spectrum of patients with pulmonary atresia/stenosis with intact ventricular septum

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Received 24 September 2002; received in revised form 2 May 2003; accepted 20 May 2003

Abstract

Objective: To determine whether one and one half ventricular repair (1.5VR) is definitely superior to the Fontan procedure in patients having hypoplastic right ventricle (RV) in the setting of pulmonary atresia with intact ventricular septum (PA/IVS) or its relatives, in terms of results in the longer term. Method: Since 1987, 1.5VR has been chosen in seven patients with PA/IVS and in six having PS with hypoplastic RV. On preoperative catheterization, right ventricular end-diastolic volume (RVEDV) was 47 ± 23% of the anticipated normal value, and annular diameter of the tricuspid valve (TVD) 72 ± 22% of normal (Z value being −2.4 ± 2.1). Follow-up term was 3–15 (10 ± 4) years. Results: All patients survived 1.5VR, but one patient died of arrhythmia 9 years later. Freedom from arrhythmia was 80 and 20% at 10 and 12 years, respectively. Two patients have undergone conversion to the Fontan circulation, but none to true biventricular physiology. Consecutive catheterization (1, 5, and 10 years after 1.5VR) demonstrated no changes in %RVEDV or %TVD. Cardiac index was 2.4 ± 0.6 l/min per m² at either 5 or 10 years. RA pressure was 9 ± 3 and 12 ± 2 mmHg at 5 and 10 years, respectively. Smaller %RVEDV and %TVD were associated with episodes of atrial arrhythmia and higher RA pressure. Exercise testing showed anaerobic threshold of 16.6 ± 3.4 ml/kg per min at either 5 or 10 years. RA pressure was 9 ± 3 and 12 ± 2 mmHg at 5 and 10 years, respectively. These values were equivalent to those in patients with classical tricuspid atresia or PA/IVS undergoing the Fontan procedure, and unequivocally inferior to those in patients having PA/IVS, who could have undergone biventricular repair. Conclusion: Although we previously expected reasonable functional results after 1.5VR between the Fontan circulation and biventricular physiology, this was not always the case in patients having pulmonary atresia or stenosis with intact ventricular septum.

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Keywords: One and one half ventricular repair; Hypoplastic right ventricle; Cavopulmonary anastomosis; Pulmonary atresia; Exercise testing

1. Introduction

One and one half ventricular repair (1.5VR) has been alternatively employed as a definitive repair in patients with a hypoplastic right ventricle in the hope that the 1.5VR physiology is either superior to the Fontan circulation or beneficial so as to minimize right heart failure after biventricular repair. This is because, in terms of the former hypothesis, a small right ventricle likely provides better capacity of the pulmonary circulation than seen in the Fontan circumstance, and thus its contraction contributes to maintenance of more efficient overall circulation and functional results. As for the purpose of rescue from right heart failure after biventricular repair, some investigators may anticipate positively that the small right ventricle would become greater in its size and might function better allowing the surgeon to promote conversion from the 1.5VR circumstance to the truly biventricular physiology in the longer term. To determine whether these expectations are reasonably realistic or not, we investigated and herein describe our results in the longer term after 1.5VR.

2. Patients and methods

Our surgical strategy has principally been based on severity of right ventricular hypoplasia in patients with...
the spectrum of pulmonary atresia/stenosis with intact ventricular septum. In those with end-diastolic volume of the right ventricle (RVEDV) greater than 50% as well as annular diameter of the tricuspid valve (TVD) greater than 70% of the anticipated normal values [1], we have promoted biventricular repair. When a patient had RVEDV no greater than 20% or annular TVD no greater than 40% of the anticipated normal values, the Fontan strategy was chosen aiming towards definitive repair. Between these cut-off criteria, 1.5VR was a surgical option of consideration. Another factor taken into account for choosing 1.5VR was absence of abnormal coronary circulation depending on the right ventricle because of severe sinusoidal communications.

Thus, between 1987 and 1999, seven patients having pulmonary atresia with intact ventricular septum and six patients with critical pulmonary stenosis (including three with Ebstein’s malformation) underwent 1.5VR at our institution. Of the 13 patients undergoing 1.5VR, 12 patients had previously undergone palliative procedures; construction of a systemic-to-pulmonary shunt in ten, reconstruction of the right ventricular outflow tract on cardiopulmonary bypass in three, and the so-called Brock procedure in five. Age at 1.5VR ranged from 10 months to 9 years old, with a mean of 4 ± 3 years. Postoperative follow-up term is 3–15 years (10 ± 4 years).

On preoperative cardiac catheterization, TVD was measured to be 72 ± 22 (40–112)% of the anticipated normal value. This corresponds to a Z value of tricuspid diameter −2.4 ± 2.1 [2].

RVEDV was 47 ± 23 (20–100)% of the anticipated normal value, with ejection fraction of the right ventricle being 52 ± 13 (25–70)%). Right atrial pressure was 8 ± 4 (4–16) mmHg, systolic right ventricular pressure 70 ± 46 (17–159) mmHg, mean pulmonary arterial pressure 16 ± 6 (8–27) mmHg, end-diastolic volume of the left ventricle 216 ± 41 (150–264)% of the anticipated normal value with ejection fraction of the left ventricle 61 ± 10 (39–79)%. Qp/Qs value was calculated as 1.1 ± 0.6 (0.5–2.3).

Through a median sternotomy and on standard cardiopulmonary bypass, 1.5VR was established. In seven patients undergoing the definitive procedure before 1990, the superior caval vein was anastomosed to the right pulmonary artery in a side-to-end fashion just as the conventional Glenn procedure. The pulmonary arteries became, therefore, non-confluent. After 1990, in contrast, the superior caval vein was anastomosed to the pulmonary arteries in a bidirectional fashion in six patients. Atrial septal defect or small interatrial communication at the oval fossa was closed in all except for one patient. The right ventricular outflow tract was enlarged either by trans-annular patching in eight or by infundibulectomy in one. Valvotomy of the tricuspid valve was done in three patients. The annular attachment of the tricuspid valve was plicated in one patient. Three patients having Ebstein’s malformation did not undergo plasty to the tricuspid valve. Total cardiopulmonary bypass time was 152 ± 62 (58–280) min, and aortic cross-clamp time 38 ± 28 (11–98) min.

An average value together with a standard deviation was used to describe data in continuous numbers. Survival rate and freedom from arrhythmia were calculated by the Kaplan–Meier method. Statview 5.0 (SAS Institute, Cary, NC) was used as statistical software.

3. Results

3.1. Mortality

All patients survived 1.5VR. One patient died suddenly of ventricular tachyarrhythmia 9 years 8 months after the procedure. Another patient underwent conversion to total cavopulmonary connection 10 years 11 months after 1.5VR, and survived the reoperation. Four months later, however, the patient died of sepsis. Survival rate at 10 years was 88.9% (Fig. 1).

3.2. Arrhythmia

Ventricular arrhythmia was noted only in one patient who died late. Atrial arrhythmia was found in another four patients, particularly 8 years or longer after 1.5VR (Fig. 1); paroxysmal atrial or supraventricular tachycardia in three, and atrial flutter in one.

3.3. Postoperative catheterization

Cardiac catheterization was carried out 5 or 10 years, as well as 1 year after 1.5VR. There was no obvious tendency towards enlargement, or growth, of RVEDV or annular TVD (Fig. 2); RVEDV being 51 ± 31%, 60 ± 14%, and 19 ± 11%, and %TVD being 67 ± 13%, 72 ± 7%, and 53 ± 16% at 1, 5 and 10 years, respectively. Right atrial pressure remained mildly high even 10 years after 1.5VR (Fig. 3), although superior caval venous pressure was not markedly high. In three patients, there was pressure difference greater than 5 mmHg noted between the right atrium and the superior caval vein (Fig. 3). Cardiac index

ranged from 1.8 to 4.2 l/min per m² (Fig. 4). There was a weak reverse correlation present between cardiac index and right atrial pressure (Fig. 4).

3.4. Exercise testing

Exercise testing was carried out 5 years after 1.5 VR in eight patients, and 10 years in three. Anaerobic threshold was 16.6 ± 3.4 (13.1–24) and 13.1 ± 2.7 (10.9–16.1) ml/kg per min, with maximal oxygen uptake being 24.8 ± 4.9 (20.5–35.2) and 20.1 ± 2.3 (16.1–24.1) ml/kg per min, at 5 and 10 years, respectively (Fig. 5).

3.5. Postoperative catheter intervention

Catheter intervention was employed in two patients after 1.5 VR. In one patient, balloon angioplasty was carried out for localized stenosis of the left pulmonary artery 4 years after definitive repair. The interventional procedure was effective. In the other patient having Ebstein’s malformation, the tricuspid valve, although its measured annular diameter was around 70% of normal (Fig. 2), proved to be hemodynamically obstructive, and congestion of the liver had been consecutively documented. The pediatrician attempted to dilate the tricuspid valvar orifice three times using balloons (5, 6, and 10 years after 1.5 VR), but the procedures were not drastically effective.

3.6. Conversion to the Fontan circulation

In the patient undergoing repeated catheter intervention, protein loosing enteropathy was eventually noted. Right atrial pressure was 14 mmHg at 10 years (Fig. 3). Aortic valvar regurgitation also progressed. Subsequently, 1.5 VR physiology in this particular patient was converted to the Fontan circulation as already mentioned in the mortality section. Conversion was carried out in an extracardiac fashion using an 18-mm polytetrafluoroethylene (PTFE) tube with placement of a 5-mm fenestration. Cryoablation of the right atrium and plasty to the aortic valve were concomitantly employed.

In another patient with pulmonary atresia with intact ventricular septum, conversion to total cavopulmonary connection was carried out 13 years 10 months after biventricular repair.

Exercise testing

Fig. 5. Exercise testing. Black dots represent data in patients undergoing one and one half ventricular repair. For comparison, similar plotting was carried out in white circles in patients with pulmonary atresia with intact ventricular septum or those with tricuspid atresia undergoing the Fontan procedure. Furthermore, asterisks show similar data 10 years after biventricular repair in patients with pulmonary atresia with intact ventricular septum.
the initial 1.5VR. Atrial tachyarrhythmia repeatedly had occurred since 12 years after the repair. The right ventricle remained to be rather rudimentary (Fig. 2). High right atrial pressure continued (Fig. 3). Conversion was carried out by the extracardiac Fontan method using a 20-mm PTFE tube, and cryoablation was concomitantly employed for the right atrial wall. Postoperatively, hemodynamics improved.

4. Discussion

Use of 1.5VR [3] has been positively documented in patients with hypoplastic right heart [4–12]. Its surgical outcome immediately after the procedure has become quite reasonable with low mortality. In the longer term, nonetheless, the surgeon should know much more about efficacy of this particular circulation. Functional results are likely correlated with right ventricular function supporting a half of the pulmonary circulation [12]. As wisely commented by Hanley [13], we should clarify policy and criteria for choosing this intermediate procedure. A borderline candidate for biventricular repair could possess right ventricular function considerably good for 1.5VR. In such a patient, RVEDV may become greater, and size of the tricuspid valve may grow [6].

In this respect, we do not think that our basic indication of 1.5VR thus far is ‘backing off’ from biventricular repair. In only one of the 13 patients of this study, biventricular repair was considered preoperatively as a surgical repair of choice. This particular patient possessed a relatively large-sized right ventricle with RVEDV being 100% of normal before definitive repair (Fig. 2), although the annular attachment of the tricuspid valve was clearly obstructive (its diameter being 60% of the anticipated normal value) and all the valvar leaflets of the valve proved to be severely dysplastic. With these findings, accordingly, we chose 1.5VR concomitantly with plasty to the tricuspid valve rather than biventricular repair. Three years have passed after the procedure, and this patient might be able to undergo conversion to biventricular repair in the future if replacement of the tricuspid valve would become feasible using a prosthetic valve, since RVEDV has not regressed postoperatively. In the other 12 patients, we regarded 1.5VR as an alternative to the Fontan procedure on the basis of volumetry of the right ventricle and morphology of the tricuspid valve before definitive repair. Of course, these angiographically measured parameters do not necessarily represent right ventricular function. They are, nonetheless, among morphologic factors which affect right ventricular function and, at the same time, which are to be measured in the form of numeric data.

Taking this background of the group of our patients into account, our present results should be discussed. We had expected better functional outcome in the longer term after 1.5VR than that after the Fontan procedure. Exercise testing, however, demonstrated that maximal oxygen uptake and anaerobic threshold were obviously below the normal standard. Our results do not disagree with the previous finding by Kreutzer and his colleagues that poor results in exercise testing had a strong correlation with poor right ventricular function [12]. Just for information, data derived from similar evaluation were shown in another series of our patients undergoing the Fontan procedure between 1987 and 1999 (Fig. 5). The comparative group includes 26 patients; those with classical tricuspid atresia and those having pulmonary atresia or severe stenosis with intact ventricular septum. The Fontan circulation had been established by atropulmonary connection in 13 and total cavopulmonary connection using an intra-atrial grafting in 13. Although statistical comparison cannot reasonably be done, results in patients undergoing 1.5VR were not worse but, at least, not better than those in patients undergoing the Fontan procedure. Furthermore, when plotting data of exercise testing 10 years after biventricular repair in six patients with pulmonary atresia or severe stenosis with intact ventricular septum, results in patients undergoing 1.5VR were obviously worse than those in patients undergoing biventricular repair (Fig. 5).

Systemic venous pressure measured and cardiac index calculated 5 or 10 years after the Fontan procedure were similarly plotted in the same Fontan group (Fig. 4). Again, results in patients undergoing 1.5VR were not worse but not obviously better than those in patients undergoing Fontan procedure. The most shocking issue was regarding arrhythmia. The curve expressing freedom from arrhythmia in the present study was quite similar to that seen in a series of patients undergoing atropulmonary connection. Occurrence of atrial arrhythmia could be related to relatively high right atrial pressure. We initially expected that right atrial pressure would be reasonably low and should be much lower than superior caval venous pressure because of cavopulmonary anastomosis. Pressure gradient between the systemic veins could produce collateral communications through veno-venous shunts [10,12]. Some of the venous drainage from the upper body may be forwarded to the inferior caval vein by such a pressure gradient and, because of that, volume load to the right ventricle could be augmented. Such a circumstance would promote growth of the initially hypoplastic right ventricle. However, this was not always the case. Right atrial pressure was almost equivalent to superior caval venous pressure in all except for three patients in whom pressure gradient between the superior caval vein and the right atrium was greater than 5 mmHg. No obvious veno-venous collaterals were identified in our series. Right ventricular volume did not become larger.

In other words, right ventricular function is fundamentally important even in the setting of 1.5VR. A right ventricle reasonably functioning would be able to forward venous blood through the inferior caval vein with low atrial pressure. In patients with such a right ventricle, that is, borderline candidates for biventricular repair, atrial arrhythmia might not occur frequently. Chowdhury et al. reported...
a lower incidence of arrhythmia than the present study [5]. In our strategy, nonetheless, such patients have been basically submitted to biventricular repair. In borderline candidates for the Fontan procedure, in contrast, it is unlikely for the small right ventricle to support very well even a half of the pulmonary circulation through the inferior caval vein in the longer term. The 1.5VR physiology in such patients is almost equivalent to the Fontan circulation composed by atrophicpulmonary connection.

As a treatment of elevated right atrial pressure and/or atrial arrhythmia subsequent to 1.5VR, we employed conversion to total cavor pulmonary connection with cryoablation of the atrial wall, following our previous experiences of similar conversion after atrophicpulmonary connection for the Fontan circulation [14,15]. Conversion from the 1.5VR circulation to total cavor pulmonary connection was also reported by Mavroudis and his colleagues [7]. Impact of surgical conversion to the Fontan circulation remains unclear. By the conversion, we cannot expect drastic improvement in hemodynamic or functional status. What we can expect would be improvement in atrial arrhythmia or severe congestion of the lower body, although these problems could remain even in the Fontan circulation. If these problems can be minimized not by way of 1.5VR but by employment of total cavor pulmonary connection from the beginning, we should bid farewell to our initial expectation that the 1.5VR might provide more efficient overall circulation and better functional status than the Fontan circulation does. In this respect, we totally agree with the opinion proposed by Hanley [13]; we can really achieve the surgical procedure of 1.5VR, but we should know exactly its advantageous indications. On the basis of our 15-year experience, 1.5VR does not possess a positive impact on functional results in the longer term as an alternative to the univentricular repair in borderline candidates for the Fontan procedure who have the spectrum of pulmonary atresia/ stenosis with intact ventricular septum.

Functional results after 1.5VR were not necessarily hopeful in the longer term. Size of the right ventricle and/or diameter of the tricuspid valve has not become sufficiently large for subsequent conversion to biventricular repair in patients in whom these structures were considerably small before definitive repair. Conversion to total cavor pulmonary connection may be needed in some patients. We must prudently determine the indication for this procedure, particularly in patients with a markedly hypoplastic right ventricle or a poor ventricular function.

References