Changing trends in the management of pulmonary atresia with intact ventricular septum: the Melbourne experience

Matthew Liava’a, b, c, Paul Brooks b, Igor Konstantinov a, Christian Brizard a, Yves d’Udekem a, c, 1, 1, a

1 Department of Cardiac Surgery, Royal Children’s Hospital and the Murdoch Children’s Research Institute, Melbourne, Australia
2 Department of Cardiology, Royal Children’s Hospital and the Murdoch Children’s Research Institute, Melbourne, Australia
3 Department of Pediatrics, University of Melbourne and the Murdoch Children’s Research Institute, Melbourne, Australia

Received 13 September 2010; received in revised form 11 February 2011; accepted 16 February 2011; Available online 10 May 2011

Abstract

Objective: Management of pulmonary atresia with intact ventricular septum (PAIVS) can be directed to either biventricular repair or univentricular palliation. The optimal management strategy has yet to be defined. Methods: All patients operated at the Royal Children’s Hospital, Melbourne for PAIVS between 1990 and 2006 (n = 81) were reviewed. Patients were retrospectively stratified into a simple three-tiered classification based on right ventricle (RV) size. Multivariate logistic regression analysis was performed to identify risk factors of mortality. Results: The distribution of RV sizes was normal in 11 (14%), moderate hypoplasia in 45 (56%), and severe hypoplasia in 25 (31%) patients. RV-to-coronary-artery connections were present in 33 (41%) and RV coronary dependence in six patients (7%). Sixteen patients died (20%). The end-status of the remaining patients was biventricular repair in 31/81 (38%), 1-ventricle repair in 10/81 (12%), Fontan circulation in 14/81 (17%), transplantation in 1/81 (1%), and still awaiting repair in 9/81 (11%). Ten-year survival was 80% (95% confidence interval (CI): 71–87%). Independent predictors of mortality were lower tricuspid valve (TV) annulus size Z-score and the presence of RV-to-coronary-artery connections. Conclusions: A simple three-tiered classification based on RV size may allow initial stratification into biventricular or univentricular repair for patients with normal RV size and severe RV hypoplasia. In patients with moderate RV hypoplasia, the presence of RV-to-coronary-artery connections or a TV Z-score < –2 should caution one against attempting biventricular repair.

Keywords: Congenital heart disease; Pediatrics; Pulmonary valve

1. Introduction

The past decade has seen a marked improvement in the surgical outcomes of patients with pulmonary atresia with intact ventricular septum (PAIVS) [1–3]. The prospective multicentric study by the Congenital Heart Surgeons’ Society predicts a 5-year overall survival of 79% for those enrolled in 1997, compared with 49% survival if enrolled in 1989 [4]. This improvement has occurred despite profound differences in the approaches advocated by the different institutions involved. Initially, centers reporting success with the treatment of this condition advocated a single approach, some favoring a biventricular pathway, others a univentricular pathway. It is now becoming clear that a strategy excessively favoring a univentricular palliation will deprive some patients with relatively normal right-ventricular cavities of the benefits of a biventricular repair and the potential for a longer life expectancy. On the other hand, a strategy pushing for biventricular repair in all cases will result in increased mortality [4]. Today, a more balanced approach seems preferable, and preliminary success has been reported [1]. Unfortunately, there are yet no clear criteria to decide which patients should be orientated to a univentricular or biventricular repair.

In Melbourne, our strategy has gradually evolved over the years. Initially, we adopted a policy aimed at growing the patient’s right-ventricular cavity and pushed indications for a biventricular repair. Our decision to attempt biventricular repair was based on the presence or absence of a right-ventricular infundibulum [5]. Those patients with a patent infundibulum headed toward a biventricular repair, sometimes requiring a ‘right ventricle (RV) overhaul’ procedure consisting of resection of intraventricular obstructive muscle bundles to enlarge the right-ventricular cavity [6]. Only patients without an infundibulum were placed on the univentricular heart pathway. In the patients with only

* Corresponding author. Address: Department of Cardiac Surgery, Royal Children’s Hospital, Flemington Road, Parkville, Melbourne, Victoria 3052, Australia. Tel.: +61 3 9345 5200; fax: +61 3 9345 6386.
E-mail address: yves.dudekem@rch.org.au (Y. d’Udekem).

1 Yves d’Udekem is a Career Development Fellow of The National Heart Foundation of Australia Research Program (CR10M5339).
moderately reduced right-ventricular size, this approach allowed us to delay the final decision between a univentricular or biventricular repair. During the past 15 years, we have progressively stepped back from this methodology and adopted a more balanced approach. Patients with the smallest RVs or tricuspid valves (TVs), and coronary anomalies were directed toward palliative univentricular procedures. In the remaining patients, the right-ventricular outflow tract was opened surgically or with a catheter intervention and targeted for biventricular repair. Up to now, however, the decision to aim toward a biventricular or univentricular repair was made after the review of each individual case, without any specific guidelines.

We therefore reviewed our experience of patients presenting with pulmonary atresia and intact interventricular septum to determine risk factors for mortality, hoping that the identification of these risk factors may guide decision making toward either a univentricular or a biventricular pathway.

2. Methods

The study was approved by the Royal Children’s Hospital Ethics committee. All patients at the Royal Children’s Hospital admitted between 1990 and 2006 with the diagnosis of pulmonary atresia and intact interventricular septum were identified from the hospital database (n = 93). Patients with clear Ebstein’s anomaly or severely dysplastic TVs with associated pulmonary atresia were then excluded from the study (n = 12), and the remaining 81 patients constitute the core of this study. All preoperative echocardiographic examinations were reviewed by a single cardiologist blinded to the outcome to assess RV size, RV-to-coronary-artery connections and TV annulus diameter. RV size was subjectively categorized into three groups: normal, moderate, and severely hypoplastic (Fig. 1). This was based on the size of the neonatal RV in comparison with the left ventricle, the partite nature of the RV, and the extent to which muscular hypertrophy obliterated the RV cavity.

Angiography was reviewed for all patients with RV-to-coronary-artery connections to determine whether their coronary circulation was dependent on the RV. RV-dependent coronary circulation was defined by the presence of RV connections to the coronary arteries with either severe stenosis of two or more major coronary arteries (right coronary artery, left main, left anterior descending artery, circumflex artery, and posterior descending artery), or complete aortocoronary atresia [7].

The following variables were analyzed: birth weight, gestational age at birth, antenatal diagnosis, age at first intervention, TV Z-score, presence of RV-to-coronary-artery connections, RV-dependent coronary circulation, and RV size.

Primary intervention was classified as ‘shunt only’ for a systemic-to-pulmonary shunt, ‘RV outflow tract procedure only’ if a procedure was performed that allowed RV-to-pulmonary-artery flow (including surgical valvotomy, transannular patching, and successful catheter valvotomy), or both. Any patient who had significant resection of right-ventricular muscle bundles beyond the infundibular area at any stage was classified as having had an RV overhaul.

Fig. 1. Examples of (A) severe and (B) moderate hypoplasia, and (C) near-normal-sized right ventricle.
Patient follow-up was extracted from the hospital database or sought from the referring cardiologist. Each patient was ascribed to one of six end-statuses: biventricular repair, 1-ventricle repair, Fontan, death, heart transplantation, and awaiting complete repair. The biventricular repair group included patients with an atrial septal defect (ASD) left open, and the 1-ventricle repair group included patients with a bidirectional cavopulmonary shunt and RV-to-pulmonary-artery forward flow, regardless of ASD presence. The ‘awaiting complete repair’ category included patients with bidirectional cavopulmonary anastomoses on the univentricular pathway and patients with systemic-to-pulmonary shunts where right-ventricular growth was being assessed over time prior to determining suitability for univentricular or biventricular repair.

3. Statistical analysis

Data were described as mean ± standard deviation for normally distributed variables and median and interquartile range for non-normally distributed variables. Categorical data were analyzed with Fisher’s exact test. Continuous data were analyzed with the Student’s unpaired t-test. A p-value < 0.05 was considered as statistically significant with all p-values based on two-tailed tests. Actuarial survival was estimated by means of the Kaplan–Meier method. Factors showing a trend toward or significant association with death on univariate analysis were evaluated by multivariate logistic regression analysis, with exclusion of those with significant interaction. Statistical analysis was performed using STATA 10.0 (Stata Corp, TX, USA).

4. Results

Between 1990 and 2006, 81 patients with pulmonary atresia with intact interventricular septum were identified in our institutional database. Their mean birth weight was 3.8 ± 0.8 kg, and median age at initial intervention was 4.5 (2–11) days. Thirty-two patients (40%) were born with an accurate antenatal diagnosis. Twelve patients received their initial intervention beyond the neonatal period (>1 month) either because of prematurity and low birth weight (n = 9) or delayed presentation due to the presence of aortopulmonary collaterals (n = 3).

By retrospective review, right-ventricular size was classified as being normal in 11 patients (14%), moderately hypoplastic in 45 (56%), and severely hypoplastic in 25 (31%). The median TV Z-score of the patients with normal size RV was −0.6 (−0.9 to 0.9), moderate hypoplasia −1.3 (−1.9 to −0.9), and severe hypoplasia −2.7 (−3.2 to −2.4).

Mean overall TV Z-score was −1.46 ± 1.39 and RV-to-coronary-artery connections were present in 33 (41%) patients. RV coronary dependence was identified in six patients (7%). Mean follow-up was 7.1 ± 5.6 years.

4.1. Death prior to intervention

Three patients died before any procedure. The first was born at term with a birth weight of 3.27 kg. His TV Z-score was −3.4, RV-to-coronary-artery connections were present, and the coronary circulation was considered RV dependent. Active treatment was withdrawn after parental request. The other two patients were born prematurely at 32 and 34 weeks’ gestation, with birth weights of 1.42 and 1.91 kg. Their management involved an assisted feeding regime to allow growth prior to operative intervention. Both died before this could occur.

4.2. Diagnostic and interventional catheterization

From 1990 to 2006, 71 of 81 patients underwent diagnostic catheterization.

The primary intervention in 17 patients was an interventional cardiac catheter with wire perforation and balloon pulmonary valvuloplasty. Trans-catheter laser or radio frequency-assisted techniques were not used in our institution during this time period. The characteristics of these patients are displayed in Table 1.

RV-dependent coronary circulation was not present in any of these patients and no patient had a severely hypoplastic RV.

In four patients (24%), catheter valvotomy was considered a technical failure. These patients underwent subsequent surgical intervention at a median of 1 (0–2) day, involving: a left modified Blalock–Taussig (BT) shunt and pulmonary valvotomy (n = 2), pericardial effusion drainage, right modified BT shunt and pulmonary valvotomy (n = 1), and pulmonary valvotomy (n = 1). Follow-up data showed that two patients ended up with a biventricular repair and two with a 1-ventricle repair.

In 13 patients, trans-catheter valvotomy was considered a technical success. However, 9/13 required further surgery at a median of 9 days (1–25) involving: a modified BT shunt (n = 8) including two with concomitant pericardial effusion drainage, and a pulmonary valvotomy with arch repair (n = 1). The single death (1/17) occurred late and was due to poor myocardial function in the one patient of the group, who had RV-to-coronary-artery connections. The final outcomes of the 17 patients undergoing primary management with interventional catheterization are detailed in Fig. 2.

4.3. RV-dependent coronary circulation

Six patients were identified to have RV-dependent coronary circulation. One patient died before any interven-

Table 1. Patient characteristics for primary catheter intervention.

<table>
<thead>
<tr>
<th>Patient characteristics</th>
<th>No. of patients</th>
<th>Birth weight (kg)</th>
<th>Gestation (weeks)</th>
<th>Antenatal diagnosis</th>
<th>Age at first intervention (days)</th>
<th>TV Z-score</th>
<th>Presence of RVCAC</th>
<th>RV size: normal</th>
<th>RV size: moderate hypoplasia</th>
<th>RV size: severe hypoplasia</th>
<th>RVDCO</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of patients</td>
<td>17</td>
<td>3.4 ± 0.6</td>
<td>40 (38–40)</td>
<td>5 (29%)</td>
<td>2 (1–6)</td>
<td>−1.0</td>
<td>1 (65%)</td>
<td>5 (29%)</td>
<td>12 (71%)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Birth weight (kg)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gestation (weeks)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Antenatal diagnosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at first intervention (days)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>TV Z-score</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Presence of RVCAC</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV size: normal</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV size: moderate hypoplasia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RV size: severe hypoplasia</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RVDCO</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

RVCAC: right ventricle-to-coronary-artery connections; and RVDCO: right ventricle dependent coronary circulation.
tion. The remaining five patients had a right modified BT shunt performed during the neonatal period. One of them died at the age of 9 months and four progressed to undergo a bidirectional cavopulmonary shunt (BCPS). One of the four patients who underwent a BCPS had a concomitant ligation of coronary fistula and a left main coronary artery angioplasty at the age of 5 months. He subsequently died of low-output syndrome 4 days after that procedure. One patient underwent a Fontan procedure at the age of 4 years and is still alive and well 2 years later. One is alive at 19 months and is awaiting Fontan completion. The last patient was transplanted at the age of 16 years.

4.4. Requirement for systemic-to-pulmonary shunt

The overall requirement of a systemic-to-pulmonary shunt was 69/81 (85%). All patients with a severely hypoplastic RV received a shunt except for the three patients described above, who died before any intervention (Table 2). Most patients (8/11) with normal-sized RV also required a shunt.

4.5. RV overhaul

Eighteen out of 81 patients, who had a patent outflow portion of their RV (22%) underwent an RV overhaul procedure. Fourteen resulted in biventricular repair. Thirteen were in the New York Heart Association (NYHA) class I or II at the time of last follow-up, and one was listed for heart transplantation due to limited exercise capacity and recurrent ventricular arrhythmias. This patient had a normal size non-contractile right-ventricular cavity. Four patients, who were subjectively estimated to have a right-ventricular cavity too small to be subjected to a biventricular repair, underwent a 1-ventricle repair.

4.6. Outcome

Final end-statuses were death 16/81 (19%), biventricular repair 31/81 (38%), 1-ventricle repair 10/81 (12%), Fontan 14/81 (17%), heart transplant 1/81 (1%), and awaiting final procedure 9/81 (11%). Of the nine patients awaiting completion of repair, four are on the Fontan pathway, four patients are left with shunts waiting a final management decision, and one has been lost to follow-up (Table 3).

None of the patients with a normal size RV ended up with a univentricular heart circulation, but two of the patients with a severely hypoplastic RV had a biventricular repair. These two patients had TV Z-scores of −2 and −1.7, and one required an RV overhaul procedure to enable biventricular repair.

No patient with a TV Z-score < −2 (n = 27) and only one patient (n = 33) with RV-to-coronary-artery connections have attained a biventricular repair. All deaths occurred in the first 2 years after birth. The causes of death were cardiac 12/16, unknown 2/16, respiratory failure 1/16, and withdrawal of treatment 1/16. In the 12 cardiac deaths, coronary ischemia is presumed to have played a significant role in inducing either a cardiac arrhythmia or sudden death. The three patients, who did not receive an intervention, died at days 2, 9, and 31 of life. For the other 13 patients, the median days until death after initial intervention was 145 (80–211); only two patients died within 30 postoperative days. One patient died intra-operatively on day 3 of life when undergoing a modified BT shunt. Bradycardia occurred during thoracotomy and was unresponsive to intervention. The second patient had poor biventricular function post trans-annular patching and BT shunt. She required a ventricular assist device for increasing lactic acidosis on day 1, and, on return to theater, was found to have an aneurysmal ventricular septum impeding left-ventricular function. Ventricular function did not recover and death ensued after withdrawal of the ventricular assist device.

Of the 11 patients on the univentricular pathway who died, only two had undergone BCPS prior to death. The remaining 11 patients were all dependent on a systemic–pulmonary shunt without any forward pulmonary blood flow.

Fourteen out of 16 of the patients dying in the follow-up period were known to have RV-to-coronary-artery connections and three of the six who were identified to have RV coronary dependence died. After 15 years, Kaplan–Meier actuarial survival was 80% (95% confidence interval (CI): 71–87%) (Fig. 3).

4.7. Risk factors for death

Univariate analysis revealed that lower TV Z-score (p < 0.0001), RV-to-coronary-artery connections (p < 0.001),

![Table 3. Outcomes based on right-ventricle size.](image)

<table>
<thead>
<tr>
<th>RV size</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Bi-V</td>
</tr>
<tr>
<td>Normal</td>
<td>10</td>
</tr>
<tr>
<td>Moderate hypoplasia</td>
<td>19</td>
</tr>
<tr>
<td>Severe hypoplasia</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>31</td>
</tr>
</tbody>
</table>

Bi-V: biventricular; 1.5-V: 1/2 ventricle; Uni-V: univentricular; and HTx: heart transplant.

![Table 2. Requirement for systemic-to-pulmonary shunt.](image)

<table>
<thead>
<tr>
<th>Right ventricle</th>
<th>Shunt only</th>
<th>Shunt and RVOT procedure</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>1</td>
<td>7</td>
<td>8/11 (73%)</td>
</tr>
<tr>
<td>Moderate hypoplasia</td>
<td>16</td>
<td>23</td>
<td>39/45 (87%)</td>
</tr>
<tr>
<td>Severe hypoplasia</td>
<td>20</td>
<td>2</td>
<td>22/25 (88%)</td>
</tr>
</tbody>
</table>
smaller RV size ($p = 0.005$), and RV-dependent coronary circulation ($p = 0.026$) were statistically significant risk factors for death. Multivariate logistic regression revealed that only lower TV $Z$-score and presence of RV-to-coronary-artery connections were independent risk factors for death (Table 4).

5. Discussion

Pulmonary atresia with an intact interventricular septum still carries a high risk of early mortality. In our series, 20% of patients died during follow-up, with most of the deaths occurring within the first 2 years of life. These results are comparable to other contemporary reports [1,4,8].

Mortality as low as 2% at 7.5 years has been reported [2]; however, this involved a high rate of univentricular palliation. It has now been clearly demonstrated that performing a large proportion of univentricular heart palliations is the best way to reduce mortality in this patient population [4]. This practice, however, may deny a biventricular repair and the possibility of increased longevity to patients with favorable morphology [4]. Most groups today would agree that a balanced approach is necessary in the management of these patients, reserving a univentricular pathway for patients with RVs at the smallest end of the spectrum and attempting a biventricular repair in those with near-normal size RVs. Initial management will differ according to stratification toward univentricular palliation or biventricular repair. While these decisions are easily taken for those at the extreme ends of the spectrum, clinicians still lack criteria to make decisions for the majority of those between these extremes.

We took a simple approach to retrospective stratification of right-ventricular size, based on echocardiography. Patients were subjectively characterized into three groups (normal, moderately hypoplastic, and severely hypoplastic). This approach seemed the easiest and most practical way of stratifying patients. It separated, on a subjective basis, those for whom the uni- or biventricular approach was the only reasonable approach, from those for whom this decision was much more difficult. While this classification helped us in the overall description of our patient population, it did not seem to be a predictor of outcome as accurate as the TV $Z$-score. The relationship between right-ventricular size and the TV annulus $Z$-score has been well described [9]. Our experience suggests that patients with a TV $Z$-score $> -2$ should be directed to a biventricular repair and those with a TV $Z$-score $< -2$ should have a univentricular heart palliation. In our series, none of the patients with a TV $Z$-score $< -2$ achieved a biventricular repair, and, in our institution, it seems reasonable that below this size of TV, only palliation with shunting should be offered initially. Patients with the smallest RVs are those who are the most at risk of developing RV-to-coronary-artery connections [10]. In case of RV coronary dependence, opening of the right-ventricular outflow tract may put the patient at risk of coronary ischemia and should therefore be avoided [10]. By not attempting to open the right-ventricular outflow tract of the smallest of these RVs, this risk is automatically avoided. Further, in our series, only one of the 33 patients identified to have RV-to-coronary-artery connections ultimately reached biventricular repair status, and it seems reasonable to favor univentricular palliation whenever these connections are identified.

We were not very successful in achieving definitive treatment or significant improvement with initial catheter intervention in these patients. Even though the initial failure rate of right-ventricular opening was only 24%, many of the patients with initial technical success required further surgery. Eventually, only 24% of patients, who underwent catheter valvotomy, did not undergo an operation in the same hospital stay. However, we did not use radio frequency or laser-valve perforation, both of which have been reported as having a higher technical success rate than balloon valvotomy alone [11,12]. The vast majority of our patients needed a shunting procedure in the neonatal period, and, as long as an additional form of blood supply requires a surgical intervention, the impact of interventional procedures seems to be limited to the exceptional cases of normal-sized RVs with limited hypertrophy.

As in other studies, smaller TV size, the most objective index of right-ventricular size, and the presence of RV-to-coronary-artery connections were independent predictors of death. The exact cause of death is difficult to identify retrospectively. We are nonetheless very suspicious that the majority of the deaths encountered occurred as a consequence of coronary ischemia. Interestingly, most of the deaths seem to have happened at a distance from the initial intervention, and it seems unlikely that identifying these coronary lesions before the first surgery would have altered

<table>
<thead>
<tr>
<th>Table 4. Risk factors for death.</th>
<th>Univariate</th>
<th>Multivariate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth weight (kg)</td>
<td>0.064</td>
<td></td>
</tr>
<tr>
<td>Gestation (weeks)</td>
<td>0.085</td>
<td></td>
</tr>
<tr>
<td>Antenatal diagnosis</td>
<td>0.158</td>
<td></td>
</tr>
<tr>
<td>TV $Z$-score</td>
<td>$&lt;0.0001$</td>
<td>0.021</td>
</tr>
<tr>
<td>Presence RVCAC</td>
<td>$&lt;0.001$</td>
<td>0.019</td>
</tr>
<tr>
<td>RV size</td>
<td>0.005</td>
<td></td>
</tr>
<tr>
<td>RVDCC</td>
<td>0.026</td>
<td></td>
</tr>
</tbody>
</table>
the outcome of these patients. Therefore, we have progressively decreased the amount of diagnostic catheterization prior to initial surgery, and have reserved its use to a later stage.

We expected to find a large proportion of the 16 deaths to have occurred in patients stratified toward biventricular repair, when, in retrospect, a univentricular approach may have been more appropriate. However, this did not turn out to be the case, as only 2/16 patients had RV decompression and attempt at biventricular repair. Three out of 16 died before any operation, and the remaining 11/16 had a shunt only as initial management and were considered on the univentricular pathway.

It has recently been shown that inter-stage mortality in shunted patients with PAIVS is comparable to that for hypoplastic left heart syndrome [13]. Miyaji et al. have also shown that performing a BCPS (in patients with an ASD) increases the oxygen saturation of blood returning to the RV and thus to the coronary arteries in RV-dependent coronary circulation [14]. Theoretically, early BCPS could prevent or delay progression of myocardial ischemia in those patients reliant on RV-to-coronary-artery flow for myocardial perfusion. Whether this is also the case in patients with RV-to-coronary-artery connections without a complete RV-dependent circulation is unclear, though it should be suspected, as most long-term survivors of PAIVS with RV-to-coronary-artery connections will have areas of abnormal myocardial perfusion [15]. Early BCPS would have the additional advantage of decreasing interim mortality due to shunt thrombosis and circulatory instability with the shunted state.

Our department has initially favored an aggressive rehabilitation of the small right-ventricular cavities including the use of RV overhaul procedures. We have progressively moved away from this approach and have virtually abandoned overhaul procedures. The present review did not bring any data enabling us to estimate the long-term benefits of these procedures, but we have developed an impression from individual cases that they may not have contributed significantly to the patients’ final outcome. Like others, we have been influenced by studies showing that patients affected by this condition with biventricular repair and 1-ventricle repair did not seem to achieve better exercise capacity than those directed to Fontan palliation [16,17]. These results are undoubtedly related to policies resulting in achieving biventricular or 1-ventricular repair in patients who would have been better treated by univentricular approach, and it is likely that a more balanced approach will result in patients having a biventricular repair with a higher average exercise capacity.

In summary, patients with TV Z-score < –2 and with RV-to-coronary connections are unlikely to reach biventricular repair. It therefore seems reasonable to only perform a shunting procedure as initial intervention in these patients. Coronary ischemia seems to be the predominant cause of death in patients with pulmonary atresia and intact interventricular septum, even when placed on a univentricular pathway. However, this usually occurs beyond the initial postoperative period and neonatal diagnostic catheterization does not seem to alter outcome. Early BCPS may help to improve mortality, whereas interventional catheterization procedures should be reserved for the cases where no shunting procedure is expected.

References


