Two-stage biventricular rehabilitation for critical aortic stenosis with severe left ventricular dysfunction

James M. Hammel, Kim F. Duncan, David A. Danford and Shelby Kutty

INTRODUCTION

Aortic stenosis in the neonate presenting with ductus arteriosus-dependent systemic blood flow is termed critical aortic stenosis (CAS) [1]. Poor outcomes in neonatal CAS with left ventricular (LV) dysfunction has prompted an effort to intervene in utero. The potential benefits of in utero intervention for aortic stenosis (AS) are reduction of LV afterload and promotion of flow through the left heart, which may help prevent progression of LV dysfunction and hypoplasia over the subsequent course of gestation [2, 3]. In utero intervention appears to induce beneficial changes in left heart physiology and aortic and mitral valve growth in AS and evolving hypoplastic left heart syndrome, eventually allowing a biventricular outcome after birth in some patients [3-5]. There are five essential components of in utero intervention in AS that include (1) early relief of aortic valve stenosis, (2) ongoing ductal patency, (3) ongoing foramen ovale patency, (4) ongoing high pulmonary vascular resistance and (5) passage of time. A subset of neonates with CAS presents earlier in the sequence of LV dilation-fibrosis-hypoplasia, and is analogous to foetuses with valvar AS and a dilated LV who may undergo foetal intervention in the second trimester. We present a post-natal approach to this problem in the neonate with CAS and LV dysfunction by mimicking the foetal intervention that has been associated with some success.

This report comprises a case series describing a clinical approach to neonates with CAS and LV dysfunction, analogous to the treatment that might be offered in utero. The series was designed not as a comparison with standard management, but as a feasibility study for proof of the following concept. In severe cases of CAS with LV dilation and dysfunction, the relief of LV distention with an intermediate stage operation including atrial septectomy would result in improved recovery of LV dimensions and contractility, and improved right ventricular (RV) haemodynamics during the temporary RV-dependent systemic circulation. With this rationale, a staged surgical palliation was...
undertaken consisting of initial surgical aortic valvotomy, bilateral pulmonary artery banding and atrial septectomy (henceforth referred to as stage 1 procedure, S1). This was followed by stage 2 procedure (S2) consisting of surgical patch closure of the atrial septal defect, ligation of the ductus arteriosus and removal of branch pulmonary artery bands. Prostaglandin E1 infusion was continued between S1 and S2 to maintain RV contribution to systemic perfusion via the ductus arteriosus.

MATERIALS AND METHODS

This was a retrospective study of patients with CAS at a tertiary paediatric referral centre. The Institutional Review Board at the University of Nebraska Medical Center and Children’s Hospital and Medical Center approved the study protocol. Patients had been selected for the two-stage surgical management strategy as opposed to conventional isolated aortic valvotomy because of very poor LV function and severe LV dilation, after interdisciplinary care conference discussions within our cardiology and cardiovascular surgery programme.

Surgical technique

S1 was performed with cardiopulmonary bypass and cold blood cardioplegia. S1 consisted of a complete inter-atrial septectomy, surgical aortic valvotomy and bilateral branch pulmonary artery banding. If there was any question of mitral valve adequacy, the valve was directly examined before proceeding. The complete contents of the fossa ovalis were excised. An aortic valvotomy was performed through an oblique aortotomy using a corneal/scleral knife (Alcon Medical, Fort Worth, TX, USA). Branch pulmonary artery bands were rings cut from 3 mm expanded polytetrafluoroethylene tubing. After S1, ductal patency was maintained with prostaglandin infusion to provide a period of RV-dependent systemic perfusion until LV function improved. LV dimensions and function were closely observed with serial transthoracic echocardiography every week for 2–4 weeks.

S2 was scheduled after LV function improved. Even though the triggers for proceeding to S2 were not prospectively defined, in each case the LV shortening fraction had doubled from baseline, and a prominent prograde colour Doppler flow signal reached the transverse aortic arch before S2 was attempted. Prior to the initiation of a cardiopulmonary bypass, a test occlusion of the ductus arteriosus was performed to confirm preparedness of the LV alone to provide systemic perfusion. After the initiation of the cardiopulmonary bypass, the ductus was ligated and the branch pulmonary artery bands were removed. Patch repair or angioplasty of the branch pulmonary arteries was not required in any patient. The atrial septal defect was closed with a patch of fixed autologous pericardium. Although none of the patients in this series had aortic arch hypoplasia, if they had, S2 would be the point at which the aortic arch could be surgically augmented. S1 and S2 are schematically illustrated in Fig. 1.

RESULTS

Between January 2008 and April 2009, four patients with CAS underwent the 2-stage biventricular rehabilitation strategy. The most recent clinical follow-up data in survivors were obtained in November 2011. All four patients had severe LV dilation and dysfunction at neonatal presentation, were mechanically ventilated and received inotropic infusions immediately after birth for haemodynamic stabilization. Patient demographics and clinical outcomes are summarized in Table 1. Patients 1 and 4 had the most severe LV dilation (LV internal dimension Z-scores of 6.9 and 7.7) at presentation, and both were born prematurely (32 and 35 weeks). Serial echocardiographic data of the four patients are shown in Table 2.

The clinical course in patient 2 was complicated by arrhythmia, renal dysfunction, sepsis and profuse thoracostomy drainage with anasarca, despite decrease in LV dimensions and improvement in LV fractional shortening. On the seventh postoperative day, the branch pulmonary artery bands were loosened to manage haemodynamic instability. Clinical status slowly improved and by the 14th postoperative day, LV function had improved adequately to allow conversion to biventricular circulation. The patient did have residual mild aortic valve stenosis with a peak gradient of 40 mmHg and a mean gradient of 20 mmHg, and mild aortic insufficiency. The clinical status was not stable.

Figure 1: Schematic illustration of the components of S1 (A) and S2 (B). See text for details.
enough to undergo arch reconstruction. LV systolic function remained good; however, there was continued and worsening biventricular diastolic dysfunction, which contraindicated conversion to a single-ventricle pathway. Difficulties with fluid balance, renal failure and infection worsened, the patient was unsuitable for cardiac transplantation and died 41 days after S2.

Patient 3 presented within hours after birth in extremis. This patient had CAS with a less dilated but severely dysfunctional LV, and completely intact atrial septum. The patient was taken directly to the operating room for S1, but haemodynamics remained marginal after surgery. LV contractility did not improve after S1, and there was evidence of biventricular diastolic dysfunction on transthoracic echocardiography. At 70 days of age, persistent biventricular failure contraindicated single-ventricle management. The parents did not wish to pursue cardiac transplantation, and support was withdrawn. Post-mortem examination showed severe biventricular hypertrophy and biventricular endocardial fibroelastosis (EFE).

The fourth patient was diagnosed by foetal echocardiography at 32 weeks of gestation and presented with severe non-immune hydrops and polyhydramnios. The RV was markedly compressed by rightward displacement of the ventricular septum, accounting

### Table 1: Patient demographics and clinical outcomes

<table>
<thead>
<tr>
<th>Patient</th>
<th>Prenatal diagnosis</th>
<th>GA (weeks)</th>
<th>Birth weight (kg)</th>
<th>Comorbidities (cardiac/ non-cardiac)</th>
<th>Age at S1 (days)</th>
<th>Age at S2 (days)</th>
<th>Change in LV end-diastolic dimension Z-score from S1 to S2/ death</th>
<th>Age at hospital discharge/ death (days)</th>
<th>Age and status at the most recent follow-up of survivors</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Yes</td>
<td>35</td>
<td>3.2</td>
<td>Respiratory distress syndrome</td>
<td>4</td>
<td>19</td>
<td>6.9–1.8</td>
<td>44 (died)</td>
<td>Alive and well at 45 months, echocardiogram: (peak aortic valve systolic velocity 2.8 m/s, mean gradient 19 mmHg, mild aortic regurgitation, normal LV EF 67%)</td>
</tr>
<tr>
<td>2</td>
<td>Yes</td>
<td>39</td>
<td>2.6</td>
<td>Endocardial fibroelastosis, mild arch hypoplasia</td>
<td>2</td>
<td>16</td>
<td>6.6–2.5</td>
<td>53 (died)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>No</td>
<td>38</td>
<td>2.7</td>
<td>Endocardial fibroelastosis, intact atrial septum</td>
<td>0</td>
<td>None</td>
<td>2.6–2.4</td>
<td>70 (died)</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Yes</td>
<td>32</td>
<td>2.4</td>
<td>Severe non-immune hydrops, polyhydramnios</td>
<td>8</td>
<td>41</td>
<td>7.7–1.9</td>
<td>105</td>
<td>Alive and well at 34 months, echocardiogram: (peak aortic valve systolic velocity 3.4 m/s, mean gradient 32 mmHg, mild aortic regurgitation, normal LV EF 70%)</td>
</tr>
</tbody>
</table>

GA: gestational age; LV: left ventricle; EF: ejection fraction.

### Table 2: Serial echocardiographic data in four patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Time point</th>
<th>Age at echocardiogram</th>
<th>LVIDd (cm)</th>
<th>LVIDd Z-score</th>
<th>LVIDs (cm)</th>
<th>FS (%)</th>
<th>EF (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Pre S1</td>
<td>3 days</td>
<td>3</td>
<td>6.9</td>
<td>2.86</td>
<td>4</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>Pre S2</td>
<td>16 days</td>
<td>1.9</td>
<td>1.8</td>
<td>1.3</td>
<td>32</td>
<td>62</td>
</tr>
<tr>
<td></td>
<td>Last follow-up</td>
<td>45 months</td>
<td>3.3</td>
<td>0.7</td>
<td>2.1</td>
<td>36</td>
<td>67</td>
</tr>
<tr>
<td>2*</td>
<td>Pre S1</td>
<td>1 day</td>
<td>3.0</td>
<td>6.6</td>
<td>2.6</td>
<td>14</td>
<td>31</td>
</tr>
<tr>
<td></td>
<td>Pre S2</td>
<td>15 days</td>
<td>2.1</td>
<td>2.8</td>
<td>1.5</td>
<td>28</td>
<td>51</td>
</tr>
<tr>
<td></td>
<td>Last follow-up</td>
<td>52 days</td>
<td>2.3</td>
<td>2.5</td>
<td>1.4</td>
<td>38</td>
<td>71</td>
</tr>
<tr>
<td>3*</td>
<td>Pre S1</td>
<td>0 day</td>
<td>2.2</td>
<td>2.6</td>
<td>2.0</td>
<td>8</td>
<td>20</td>
</tr>
<tr>
<td></td>
<td>Last follow-up</td>
<td>65 days</td>
<td>2.2</td>
<td>2.4</td>
<td>1.95</td>
<td>10</td>
<td>23</td>
</tr>
<tr>
<td>4</td>
<td>Pre S1</td>
<td>7 days</td>
<td>2.9</td>
<td>7.7</td>
<td>2.6</td>
<td>10</td>
<td>24</td>
</tr>
<tr>
<td></td>
<td>Pre S2</td>
<td>39 days</td>
<td>1.1</td>
<td>1.9</td>
<td>0.9</td>
<td>17</td>
<td>31</td>
</tr>
<tr>
<td></td>
<td>Last follow-up</td>
<td>34 months</td>
<td>3.1</td>
<td>0.3</td>
<td>1.9</td>
<td>39</td>
<td>69</td>
</tr>
</tbody>
</table>

LVIDd: LV end-diastolic dimension; LVIDs: LV end-systolic dimension; FS: fractional shortening; EF: ejection fraction.

*Patients who died.
for the hydropic presentation in utero. The foetus was delivered by emergent Caesarean section and transported to the catheterization laboratory immediately after birth for balloon aortic valvuloplasty. This resulted in minimal improvement in the LV function (shortening fraction 10–14%) and a mild reduction of the gradient at the native atrial septal communication (mean gradient from 10 to 6 mmHg). Owing to continued marginal haemodynamics with high fluid resuscitation requirements and lactic acidosis, the patient returned to the catheterization laboratory on Day 2 of life for static and cutting balloon dilation of the atrial septal communication. The mean gradient at the atrial level decreased to 4 mmHg, resulting in a further mild reduction in LV distention and improvement in RV filling. However, by Day 8 of life, there was still little improvement in anasarca, and RV filling required significant volume administration due to persistent LV distention. With slight improvement in LV ejection performance, the residual aortic valve gradient (peak instantaneous 40 mmHg, mean 20 mmHg), and atrial septal gradient (mean 8 mmHg) increased. S1 was undertaken at this time. Little further aortic commissurotomy was possible after the previous balloon valvuloplasty, however, significant nodular thickness could be shaved from the leaflets, reducing residual stenosis. With complete elimination of atrial septal restriction, LV dimensions decreased, anasarca improved and oedema eventually resolved. A month after S1, LV function had improved sufficiently that S2 could be performed. The remainder of convalescence was slow but uneventful and the patient was discharged after two additional months, with a corrected gestational age just past term. This patient has undergone one subsequent balloon aortic valvuloplasty at 5 months of age and remains well at 36 months of age. The echocardiographic images of this patient at the time of presentation and at the most recent follow-up are shown in Fig. 2.

**DISCUSSION**

Patients with CAS typically undergo conventional stage 1 single-ventricle (Norwood) palliation if the left-sided cardiac structures are inadequate, or balloon or surgical aortic valvotomy if the left-sided cardiac structures are otherwise adequate [6]. The critically obstructed LV in CAS may be hypertrophied, with preserved systolic function or dilated with depressed systolic function. The subset of neonates presenting with severely depressed LV systolic function is at a significant risk for early mortality [7, 8]. The two-stage surgical biventricular rehabilitation strategy described in this report was chosen for four patients with CAS. There were no specific inclusion criteria for LV dilation and systolic dysfunction; however, the intention was to include cases in which both were severe. There were no cases included with LV shortening fraction >15% or LV end-diastolic dimension Z-score <2.5. Two of the four in whom this strategy was applied had very good outcomes, and two did not. Consideration of the differences between survivors and non-survivors in this series is instructive. Most notably, better outcomes were achieved in the two premature patients with extreme LV dilation (Z-scores of 6.9 and 7.7) treated at 32 and 35 weeks of gestation.

The sequence of prenatal aortic stenosis leading to LV dilation and dysfunction, and then to LV fibrosis and hypoplasia culminating in hypoplastic left heart syndrome is known [2, 4]. Foetal aortic valvuloplasty performed for AS and an evolving hypoplastic left heart have been shown to produce beneficial alterations in left heart physiology and growth [3–5]. Although foetal aortic valvuloplasty may result in improved LV dimensions and systolic function at birth [2, 4], it is often not feasible in patients diagnosed late in gestation. When CAS is identified in the third trimester or if severe LV dilation and dysfunction have set in, elective delivery and the two-stage surgical management described herein may represent a suitable strategy.

The degree to which S1 reproduces the five accomplishments of foetal intervention is important:

1. **S1 allows optimal early relief of aortic valve stenosis.** Satisfactory relief of aortic valvar stenosis has been shown to be feasible with either surgical valvotomy or balloon valvuloplasty techniques [8, 9]. With current bypass and cardioplegia techniques, surgical valvotomy by direct visualization potentially offers the most accurate, controlled valvotomy and minimal procedure-related morbidity [8].

2. **S1 maintains RV support of systemic perfusion through right-to-left ductus flow with prostaglandin infusion, thereby permitting an adequately balanced circulation as pulmonary vascular resistance falls during the first days and weeks of life.**

3. **The unrestrictive atrial communication created with S1 allows maximal unloading during functional recovery of the severely dilated and dysfunctional LV.** Leaving the atrial septum restrictive after a surgical or transcatheter aortic valve intervention performed alone often results in continued LV distention until (or unless) systolic function improves and LV ejection can keep up with pulmonary return. Leaving a restrictive atrial communication would not allow for sufficient unloading.
of the diseased LV, and would leave unacceptably high pulmonary venous pressures.

(4) High pulmonary resistance is maintained with this strategy. With S1, bilateral branch pulmonary artery bands are applied. An important advantage of applying bilateral bands in this setting is the facilitation of effective control of pulmonary blood flow independent of the ductus arteriosus.

(5) Time is allowed to elapse between S1 and S2, anticipating improvement of LV function, similar to the expected improvement in left heart structure and physiology from mid to late gestation after foetal aortic valvuloplasty. In retrospect, this approach may perhaps be less effective or inappropriate in patients presenting at a later gestational age (as in the case of the two non-survivors), presumably due to progression of fibrotic changes in the LV. EFE of the LV is common with left heart hypoplasia, and echocardiographic features of EFE were present in all four patients in this series. EFE is considered a marker for poor prognosis for ventricular recovery, but two in our series did well despite a degree of EFE. Higher grades of EFE have also been reported as a strong predictor of mortality following biventricular repair [7, 10].

Others have reported a hybrid approach of bilateral pulmonary artery banding and ductal stenting performed 4 weeks after balloon valvuloplasty as a bridge to biventricular repair in neonates with CAS and borderline LV [11]. Balloon valvuloplasty via an ascending aortic approach and concomitant bilateral branch pulmonary artery banding has also been reported to be feasible and efficient in a low-birth-weight (1.2 kg) neonate with CAS and severe LV dysfunction [12]. In that case report, the procedure was performed as a hybrid intervention at a gestational age of 27 weeks [12]. However, these approaches differed from ours because they did not directly address the atrial septum. We believe that maximal LV unloading with an unrestrictive interatrial communication is required during functional recovery of the severely dilated and dysfunctional LV. In theory, a widely patent ductus and non-restrictive atrial septum could divert enough flow away from the LV to cause it to involute. Over the short time course between S1 and S2 in this series, clinically relevant LV involution was not observed.

Ductal stenting by interventional catheterization has important technical challenges and procedural risks. These are often related to the variable anatomy and tortuosity of the ductus in these infants [13]. Moreover, the effectiveness of this procedure is dependent on adequate coverage of the stent at both the aortic origin and the pulmonary insertion of the ductus. Besides femoral arterial injury, there is risk for acute occlusion of a ductus manipulated at interventional catheterization, which may necessitate emergent mechanical cardiopulmonary support. A recent series on ductal stenting reported 15% procedure-related mortality [14]. While the risks of ductal stenting are mostly up front procedure-related, the risks associated with prostaglandin E1 infusion, both systemic and on the vascular tissue to be operated upon at S2, increase with time. Therefore, the choice between ductal stenting and prostaglandin E1 depends on the duration of ductal patency desired. For the short duration of up to 6 weeks used for the present indication, we judged prostaglandin E1 infusion to be better. It has been the experience of several groups including ours that patients born at term with CAS and less severe LV dysfunction and dilation, or with LV hypertrophy and preserved systolic function, can be managed using an aortic valve procedure alone with good expectation of adequate biventricular haemodynamics. Successful relief of valvar stenosis can be achieved in most cases either by surgical valvotomy [6, 8] or balloon valvuloplasty [9]. Moreover, balloon atrial septostomy can be undertaken as an additional procedure at catheterization aiming to improve RV filling and reduce LV dilation. Maintenance of ductus patency along with these measures is likely to temporarily allow the RV to support the systemic circulation and hopefully allow for LV recovery; infants can thus be managed at least for a limited time without the added control of branch pulmonary artery bands. However, all the above measures failed in the fourth patient in this series, and there was very good improvement in haemodynamics when the two-stage approach with initial branch pulmonary artery banding was undertaken. In retrospect, it is impossible to know whether conventional single-ventricle management would have resulted in a better outcome for the patients who died. Presumably because of LV hypertension and stiffness, the mitral stenosis with aortic stenosis/atresia subtypes of left heart hypoplasia with dilated LV at presentation historically have had poor outcomes with standard staged palliation. Importantly, S2 provided a second opportunity to address residual aortic valve stenosis and insufficiency. Patch repair or angioplasty of the branch pulmonary arteries was not required in any of the three patients in this series who underwent S2.

CONCLUSIONS

A novel, highly invasive approach to a small subset of infants with critical aortic obstruction and LV dilation and dysfunction is described. Our experience, albeit small, suggests that there exists a group of patients in whom the LV can be resuscitated with this approach. We speculate that they would likely do less well with conventional aortic valve intervention alone.

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REFERENCES

Critical aortic stenosis with severe left ventricular dysfunction

Viktor Hraška* and Martin Schneider

German Paediatric Heart Center, Asklepios Clinic Sankt Augustin, Sankt Augustin, Germany

* Corresponding author. German Pediatric Heart Center, Asklepios Clinic Sankt Augustin, Arnold Jansen Str. 29, 53757 Sankt Augustin, Germany. Tel: +49-2241-249603; fax: +49-2241-249602; e-mail: v.hraska@asklepios.com (V. Hraška).

Keywords: Congenital heart disease • Valve (lesions, repair, replacement) • Aortic valve repair • Aortic valve replacement • Neonate

In their study, Dr Hammel et al. [1] reported on their restricted experience with a two-stage surgical management approach designed for those infants who represent, according to the authors, an unusually high failure risk when undergoing either aortic valvotomy or conventional stage 1 single ventricle (Norwood) palliation. This risk is due to severe left ventricular dysfunction at the time of presentation. Four neonates with critical aortic stenosis and severely depressed left ventricular systolic function, as well as a dilated left ventricle, were treated using open valvotomy, bilateral pulmonary artery banding and atrial septectomy first (stage 1), with the aim of decompressing the left ventricle, while maintaining the right ventricular contribution to systemic perfusion via the ductus arteriosus. In one patient, the left ventricular function did not recover and the patient died, having received no further surgical/interventional treatment. The remaining three patients, in whom the left ventricular function recovered, subsequently underwent closure of the atrial septal defect, ligation of the ductus arteriosus, and the removal of the pulmonary artery bands (stage 2). Two patients survived and biventricular circulation was achieved.

The cohort of patients is small, the results are mixed and, in particular, the data regarding the morphology of the inflow and outflow of left ventricle, the grade of endocardial fibroelastosis, or functional parameters such as antegrade flow in the ascending aorta, which are crucial in decision-making, are missing. Based on the limited information available, it is most likely that the issue in these patients was not the borderline size of the left ventricle, but critical aortic stenosis with severe left ventricular dysfunction and dilatation. This point deserves a few comments. Dependency on the ductal circulation, despite an aggressive resuscitation, is a clear indication for semi-urgent intervention/operation.

(1) If the left ventricular function is depressed, a balloon valvotomy with a balloon not larger than 70% of the diameter of the aortic annulus might be performed to slightly increase the effective orifice area of the aortic valve, thus creating a minimal risk of regurgitation. At our institution, this so-called ‘gentle’ balloon valvotomy is used as an intermittent step to allow the left ventricle to recover and to stabilize the patient before open valvotomy [2].

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