Bailout shunt/banding for backward left heart failure after adequate neonatal coarctectomy in borderline left hearts

Stephen C. Browna,b, Benedicte Eyskensa, Derize Boshoffc, Bjorn Coolsa, Ruth Heyinga, Filip Rega, Bart Meynsc and Marc Gewilliga,*

a Fetal and Pediatric Cardiology, University Hospitals Leuven, Leuven, Belgium
b Pediatric Cardiology, University of the Free State, Bloemfontein, South Africa
c Congenital Cardiac Surgery, University Hospitals Leuven, Leuven, Belgium

* Corresponding author. Fetal and Pediatric Cardiology, University Hospital Gasthuisberg, Herestraat 49, 3000 Leuven, Belgium. Tel: +32-16-343865; fax: +32-16-343981; e-mail: marc.gewillig@uzleuven.be (M. Gewillig).

Received 1 April 2016; received in revised form 10 June 2016; accepted 27 June 2016

Abstract

OBJECTIVES: To determine the outcome of a bailout procedure using the right ventricle to re-assist the left ventricle in neonates after technically adequate coarctectomy but a failing borderline left heart.

METHODS: The surgical procedure was performed on bypass. A ‘reversed’ 6-mm surgical shunt was inserted between the pulmonary trunk and the descending aorta together with bilateral branch pulmonary artery banding.

RESULTS: Over a 10-year period, 89 neonates presented with coarctation and small left hearts. In 9 neonates, a hybrid procedure was performed at the outset. The remaining 80 underwent extended end-to-end coarctectomy. Two of these, despite adequate coarctectomy, developed retrograde cardiac failure with supra-systemic pulmonary hypertension, dilating right ventricles and progressive cardiogenic shock. The progressively dilating right ventricles inhibited left ventricular filling. Reversed surgical shunts were performed at 9 and 7 days post-coarctectomy. Both infants recovered rapidly and could be extubated after 4 and 7 days, respectively. Patient 1 proceeded to a univentricular repair and Patient 2 to a biventricular repair.

CONCLUSIONS: Reversed surgical shunt with bilateral banding of the branch pulmonary arteries after neonatal coarctectomy can be successfully employed as a bailout procedure in cases where a borderline left heart with growth potential cannot tolerate a biventricular circulation. It may act as an acute life-saving measure as well as a bridge to later repair. If high risk for backward failure exists in a borderline left heart with catch-up growth potential, a primary hybrid procedure is probably a more elegant and predictable strategy.

Keywords: Reversed shunt • Bilateral banding branch pulmonary arteries • Hypoplastic left heart syndrome

INTRODUCTION

Isolated coarctation of the aorta (coA) in newborns is often associated with underdevelopment of several left heart structures which may have a considerable impact on early and late outcomes. The term borderline left heart has been put forward but is an imprecise concept and essentially refers to a situation where biventricular repair is contemplated in the face of increased risk for morbidity and mortality. In the majority of patients the decision is relatively easy: many neonates will have a small but adequate left ventricle and can proceed to a successful coarctectomy with adequate catch-up growth of the left ventricle and aortic arch in the following months. At the other end of the spectrum, infants with a hypoplastic left heart without catch-up growth potential will proceed to univentricular repair via a Norwood or hybrid approach (bilateral pulmonary artery banding and stented duct) [1]. Some patients will have a very small left ventricle with potential for catch-up growth if, after birth, adequate preload to the left ventricle is provided; these patients typically can be managed with a hybrid procedure allowing the ventricle to grow for some months, after which a biventricular repair can be achieved [2].

The key question in the neonatal period is therefore: when is a left ventricle too small to sustain systemic output early after coarctectomy? Insufficient criteria exist to sort this out with complete accuracy [3–5]. However, if the clinician judged the ventricle to be big enough, but early after an adequate coarctectomy, the patient suffers from low output as a result of backward failure, and a situation with high risk of mortality is created. This article reviews our experience with a bailout procedure using the right ventricle to assist the left ventricle in neonates after a technically adequate coarctectomy but with a failing borderline left heart.

PATIENTS AND METHODS

This is a retrospective analysis. Data were extracted from patients’ files and captured on standard spreadsheets. Echocardiographic
measurements were performed by an independent paediatric cardiologist based on 2D images. Valves were measured from hinge point to hinge point. For all measurements, the average of five separate measurements was used. z-Values were calculated using standard available nomograms [6].

During the period from 2005 to 2015, 89 newborns presented with isolated coarctation and variable degrees of small left ventricles; clear hypoplastic left hearts were excluded from this analysis. Nine patients underwent a trans-sternal hybrid procedure because of a borderline left heart, and in the others (n = 80) an extended end-to-end coarctectomy was performed as the primary intervention. Two of these 80, despite complete relief of the coarctation and initial stable haemodynamics, developed progressive circulatory failure regardless of optimal therapy and required an urgent bailout intervention. Both these patients form the basis of this study.

Both infants were doing well initially after coarctectomy with good pulsatility of the abdominal aorta and minimal gradients across the aortic arch (<10 mmHg). Urinary output was adequate, and there was no evidence of lactic acidosis. However, both infants developed progressive and severe cardiac failure not responding to conventional medical treatment. Echocardiography showed a small but contractile left ventricle with pulsatile flow across the arch, but a progressive dilating right ventricle with worsening tricuspid valve regurgitation and supra-systemic pulmonary hypertension. The left ventricle was increasingly squashed by the right ventricle (Figs 1–3). Patient 2 developed renal failure on Day 6 and rising lactate levels. This condition was diagnosed as ‘backward left heart failure with progressive pulmonary hypertension and right heart failure’.

A bailout procedure was indicated. The surgical procedure aimed to allow the right heart to re-assist the left heart similar to the neonatal circulation with an open duct. This was performed on bypass through sternotomy. A ‘reversed’ surgical shunt (6-mm Gore-Tex® vascular graft, Gore Medical, Flagstaff, AZ, USA) was inserted between the pulmonary trunk and the descending aorta together with bilateral branch pulmonary artery banding as for a standard hybrid procedure (2-mm ring strips of a 3.5-mm Gore-Tex® graft, Gore Medical) (Fig. 4).

RESULTS

Both newborns had criteria consistent with borderline left hearts at presentation with small mitral and aortic valves prior to the initial extended end-to-end coarctectomy. Details can be viewed in Table 1. Urgent bailout surgery employing a ‘reversed’ shunt procedure was performed 9 and 7 days after initial coarctectomy. After this surgery was performed, their conditions rapidly improved. Patient 1 was ventilated for 4 days and Patient 2 for 7 days and discharged from the intensive care unit after a total stay of 7 and 13 days, respectively (Fig. 4).

Follow-up

Patient 1 has been followed up for 3 years; the mitral valve remained very small, impeding catch-up growth of the left heart; she proceeded to a Damus–Kaye–Stansel operation and total cavopulmonary connection (Fontan). Patient 2 showed progressive growth of the left ventricle; the left ventricle grew to subnormal dimensions, with nice antegrade flow across the arch and decreasing cyanosis of the lower abdomen. He proceeded to breakdown of the reversed shunt and debanding at the age of 3 months; he maintains a good and normal biventricular circulation at the 9-month follow-up.

DISCUSSION

The difficulty and unreliability of using current methods of defining borderline left hearts are illustrated by these 2 cases. There exists a broad spectrum of left heart dimensions in newborns with aortic coarctation and borderline left hearts that is not predictive of the final outcome [7–9]. It appears that the small left ventricle in especially coA retains the potential to grow depending on inflow and outflow, but remains difficult to predict [3, 10]. Scoring systems still have grey areas and no single measurement has been demonstrated to accurately predict outcome for an individual patient [7, 11–13]. den Dekker et al. [5] have also elegantly demonstrated that once z-values go beyond −2, they diverge substantially, making it more difficult to predict outcomes of small left hearts.

The mode of failure in both our patients was unusual. It was not the result of a residual arch gradient or forward failure since good contractility with pulsatile flow could be observed in the aortic arch. Furthermore, systemic circulation was sufficient, evidenced by the absence of elevated lactate levels and adequate kidney function during the first days following coarctectomy. However, although the left ventricle demonstrated good contractility, it was

Figure 1: Echocardiography before coarctectomy. Four-chamber views before extended coarctectomy (Patient 1, left; Patient 2, right). Note the borderline features of the left ventricles. LV: left ventricle; RV, right ventricle.
clear that it could not empty the left atrium adequately. This amplified and aggravated the pre-existing pulmonary hypertension leading to supra-systemic right heart pressures and severe right heart failure. Marked dilatation of the right ventricle resulted in progressive squashing of the left ventricle, further impeding left heart inflow and output with the development of cardiogenic shock (retrograde failure).

The results of this study show that it is feasible to surgically construct a reversed shunt with simultaneous bilateral pulmonary artery banding for a failing borderline left ventricle with retrograde heart failure. Both patients survived. This procedure can act as an acute bailout and a bridge to either uni- or biventricular repair and may be life-saving in the acute setting. Both infants recovered remarkably rapidly in the intensive care units and could be discharged home. In retrospect, Patient 1 had a mild degree of mitral stenosis, which became more evident during further growth; the left heart remained too small.

When confronted with a failing circulation after coarctectomy, a Norwood procedure could have been an alternative. However, when compared with our procedure, the Norwood is much more complex, with a longer bypass run including cardioplegia, with a more hectic postoperative course, especially for patients who started with a failing circulation; a Norwood is also much more difficult to reconvert to a biventricular circulation.

At the time of the primary intervention, the clinician has to decide whether the left ventricle is large enough not only to provide adequate systemic perfusion, but also to drain the left atrium. Employment of a hybrid approach, if doubt exists, will allow time for assessment and catch-up growth for several months; biventricular repair can then be performed at a later stage on an adequate left heart, thereby avoiding early postoperative shock, early and late retrograde pulmonary hypertension. Allowing a period of assessment of left heart growth may be helpful since a borderline left heart can also result in late pulmonary hypertension after initial

Figure 2: Echocardiography at the time of retrograde failure. Four-chamber views of Patients 1 (A) and 2 (B). Note the markedly dilated RA and RV in both cases. Interaltrial and interventricular septum displaced to the left. (C) Displaced interventricular septum with squashing of the left ventricle on the short axial view. RA: right atrium; RV: right ventricle; LA: left atrium; LV: left ventricle.

Figure 3: Echocardiography after a reversed shunt. Four-chamber views of Patients 1 (A) and 2 (B). Note improvement in LV volume in Patient 2. For more details, see text. LV: left ventricular; RV: right ventricular.
improvement, with demise months or even years after a technically successful coarctectomy [14]. It would thus be a more elegant strategy to offer a primary hybrid strategy (ductal stent and bilateral pulmonary artery banding) for infants with a borderline left heart at a high risk of backward left heart failure. The absence or the presence of catch-up growth can then be evaluated prior to the next intervention.

Limitations

The study is limited from the small size of the sample and the retrospective nature. It should be emphasized that these cases are extremely rare (2/80 newborn coA with borderline left ventricles over a 10-year period).

CONCLUSIONS

Reversed surgical shunt with bilateral banding of pulmonary branch arteries after neonatal coarctectomy can be successfully employed as a bailout procedure in cases where a borderline left heart cannot tolerate a biventricular circulation. It may act as an acute life-saving measure and a bridge to later repair. If a high risk for backward failure exists, a primary hybrid procedure is probably a more elegant and predictable strategy.

ACKNOWLEDGEMENTS

We are grateful to Stephanie Philippaerts for providing the illustrations.

Funding

This work was supported by grants from Eddy Merckx Research Foundation and Sporta Monta Ventoux Research Grant 2014.

Conflict of interest: none declared.

REFERENCES