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CASE REPORT

Ross-Konno and Endocardial Fibroelastosis Resection After Hybrid Stage I Palliation in Infancy: Successful Staged Left-Ventricular Rehabilitation and Conversion to Biventricular Circulation After Fetal Diagnosis of Aortic Stenosis

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Abstract We report a patient who presented during fetal life with severe aortic stenosis, left-ventricular dysfunction, and endocardial fibroelastosis (evolving hypoplastic left heart syndrome). Management involved in utero and postnatal balloon aortic valvuloplasty for partial relief of obstruction and early postnatal hybrid stage I palliation until recovery of left-ventricular systolic function had occurred. The infant subsequently had successful conversion to a biventricular circulation by combining resection of endocardial fibroelastosis with single-stage Ross-Konno, aortic arch reconstruction, hybrid takedown, and pulmonary artery reconstruction.

Keywords Aortic stenosis · Fetal balloon aortic valvuloplasty · Fetal intervention · Hypoplastic left heart syndrome · Resection of endocardial fibroelastosis · Stage 1 hybrid

Introduction

The morphologic and functional spectrum of the left ventricle in severe aortic stenosis ranges from the smaller, hypertrophied "borderline" left ventricle to the dilated and thin-walled left ventricle. In addition, several studies have demonstrated that aortic stenosis with left-ventricular dysfunction presenting in fetal life may lead to hypoplastic left heart syndrome at birth, with varying degrees of chamber hypoplasia and endocardial fibroelastosis (EFE) [6, 8, 10]. Neonates with severe aortic stenosis generally require either surgical valvotomy or transcatheter balloon valvuloplasty [7], whereas those with hypoplastic left heart are generally managed with univentricular palliation (Norwood or hybrid transcatheter-surgical techniques) or cardiac transplantation.

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Case Report

A 3.48-kg male infant was delivered by way of planned repeat Cesarean section at 39 2/7 weeks' gestation to a 36 year-old gravida 3, para 2 mother. The pregnancy was complicated by a diagnosis of fetal aortic stenosis made at 22 3/7 weeks' gestation (Fig. 1). At 23 4/7 weeks' gestation, fetal balloon aortic valvuloplasty was performed. After the procedure, as demonstrated by color Doppler, aortic arch flow had gone from retrograde to antegrade across the aortic valve and in the ascending and transverse aortic arch; there was mild aortic insufficiency. Postnatally the left ventricle and mitral valve were of normal size; the aortic valve was mildly hypoplastic at 5 mm with a peak Doppler-derived gradient of 60 mmHg; and there was significant left-ventricular hypertrophy (diastolic left ventricular posterior wall dimension, LVPWd 6 mm) with poor systolic function (LVEF% $< 20\%$). Doppler flow in the transverse arch and isthmus was antegrade. Cardiac catheterization showed increased left atrial pressure of 25 mm Hg. The aortic valve was dilated by way of a retrograde approach with a 5-mm Slalom balloon catheter (Cordis Corporation, Miami, FL, USA) with a decrease in peak-to-peak gradient from 60 to 45 mm Hg. After valvuloplasty, a 5-mm stent

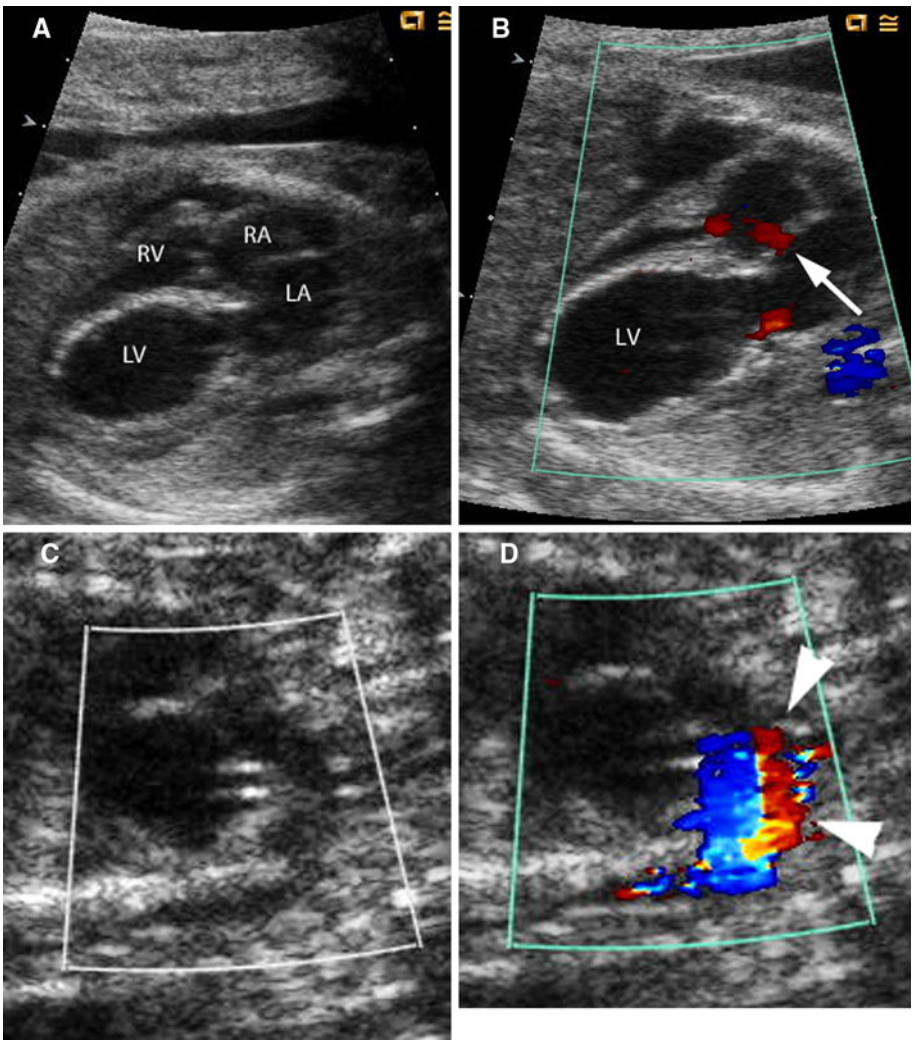

Fig. 1 Fetal echocardiogram at 22 4/7 weeks' gestation. The left ventricle is dilated with echo-bright endocardium suggestive of endocardial fibroelastosis (**a**), and there is left-to-right flow across the atrial septum (**b**). Sagittal two-dimensional (**c**) and color Doppler (**d**) images demonstrate retrograde perfusion of the aortic arch in *red* (*arrowheads*), and antegrade flow in the ductal arch is also apparent (*blue*). *LA* left atrium, *LV* left ventricle, *RA* right atrium, *RV* right ventricle

was placed in the interatrial septum. The neonate was maintained on prostaglandin infusion.

On day of life 14, with no change in clinical status or LVEF%, the child underwent stage I hybrid palliation with placement of an 18-mm ductal stent and bilateral pulmonary artery banding. Repeat dilation of the aortic valve was performed at that time, resulting in a decrease in peak-to-peak gradient to 11 mm Hg and mild to moderate aortic insufficiency.

The LVEF% improved gradually to 60% during the subsequent months. Cardiac catheterization at 9 months of age showed a 38 mm Hg peak-to-peak gradient across the aortic valve and mild to moderate aortic insufficiency. Left atrial pressure was 11 mm Hg; left ventricular end diastolic pressure was 14 mm Hg; and estimated pulmonary vascular resistance was 4 Woods units. After recovery from a severe intercurrent pulmonary illness, the child underwent a Ross-Konno procedure, placement of a 16-mm right ventricle-to-pulmonary artery homograft, ductus arteriosus and atrial stent removal, aortic arch reconstruction, pulmonary artery debanding, resection of extensive EFE (Fig. 2), and closure of an atrial septal defect. He was successfully extubated on postoperative day 7 and weaned from inotropic and vasodilatory support. Follow-up echocardiography showed no evidence of mitral inflow obstruction or regurgitation, normal left-ventricular systolic function, and a well-functioning autograft with no aortic arch obstruction. At catheterization 1 month later, the cardiac index was 3.3 ml/min/M2 by thermodilution; wedge pressure mean was 10 mm Hg; and pulmonary vascular resistance was 6 Woods units (decreasing to 2.6 Woods units in response to inhaled nitric oxide).

At 1 year of life, left heart dimensions are normal (mitral valve 13.7 mm [Z score +0.7], LV end diastolic diameter 24.9 mm [Z score +0.3], and LVPWd 6 mm [Z score +2.4]), and his systolic function is normal with an LVEF% of 69% by Simpson's biplane method (Fig. 3).

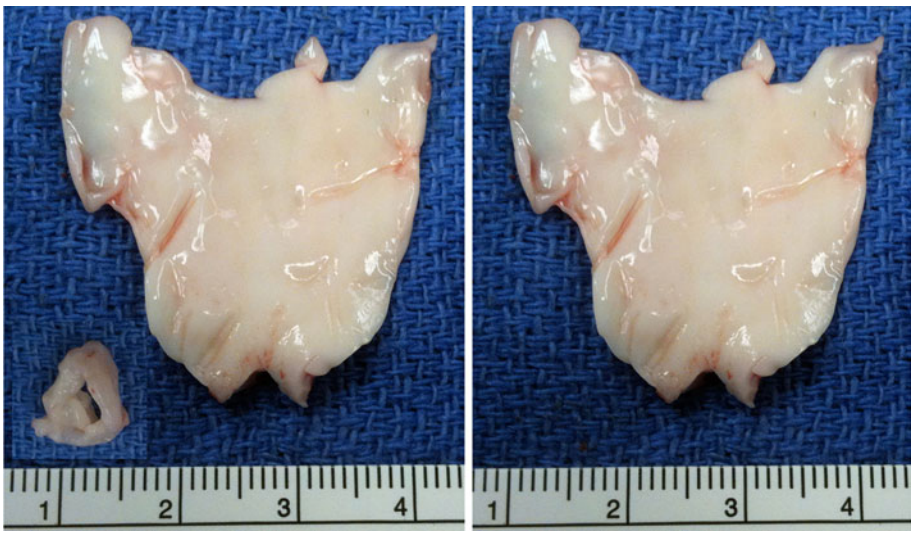

Fig. 2 Surgical specimens of the explanted aortic valve (*inset*, *left*) and left-ventricular endocardium. The aortic valve was severely dysplastic, doming, and unicuspid. At the time of biventricular repair, the valve was detached from the aortic wall. The endocardium was diffusely thickened and excised from base to apex through the left-ventricular outflow tract

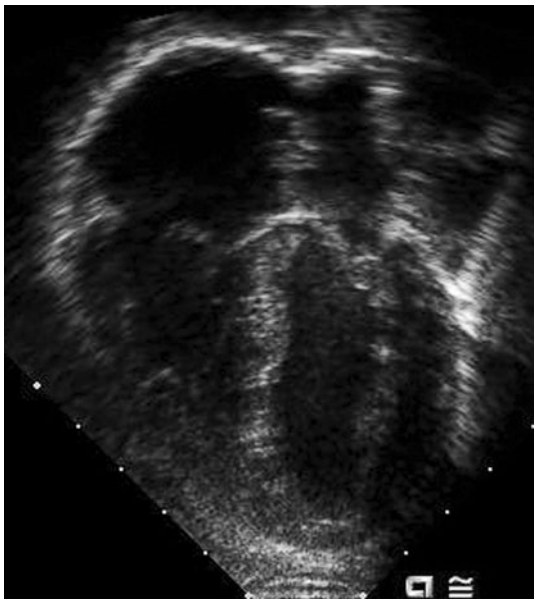

Fig. 3 Echocardiogram 2 months after biventricular repair demonstrates an apex-forming left ventricle and mild residual left-ventricular hypertrophy

Discussion

Experimental models involving surgically created aortic obstruction do not recapitulate the human phenotype [3]; therefore, study of the pathophysiology of this disease has been difficult. However, natural history studies have shown that in the presence of certain physiologic findings (specifically left-to-right atrial shunting and retrograde aortic arch perfusion in fetal life), progression to hypoplastic left heart syndrome at term can be reliably predicted [6]. The rationale for fetal aortic valvuloplasty is to relieve aortic obstruction and promote antegrade flow through the left heart and aorta. With restoration of antegrade flow, we observed continued growth of the left heart structures through the remainder of gestation; however, ventricular function did not improve appreciably. The reason for this is speculative, but it has been proposed that the fetal left ventricle responds to obstruction with increase in mass without increase in capillary density, which may result in a hypertrophied myocardium with increased susceptibility to ischemia [9, 12]. The extensive EFE in this patient as well as the observed hypertrophy might support this theory. Fetal valvuloplasty partially relieved the obstruction, and after birth we undertook a combined approach that may have further mitigated the effect of long-standing obstruction and ischemic insult through relief of obstruction (by way of additional valvuloplasties) and decrease in preload (left atrial decompression with atrial stenting). Facilitating antegrade perfusion of the aorta and coronary circulation with normally saturated blood, along with improvement in myocardial perfusion, may have contributed to the slow improvement in the patient's systolic and diastolic function.

Previous reports have also documented the possibility of reversible severe left-ventricular dysfunction and ventricular growth in the neonate [1, 2, 5]. Emani et al. [4] reported a series of patients with borderline left heart disease who underwent "primary left-ventricular rehabilitation" at a median age of 5.6 months, with aortic and/or mitral valve surgery and EFE resection, with good short-term results and no deaths at a median follow-up of 25 months. Our case, however, represents the first report to our knowledge of a patient initially palliated with a hybrid transcatheter-surgical approach after fetal intervention followed by successful surgical takedown of the hybrid and conversion to biventricular circulation by adding Ross-Konno procedure and aortic arch reconstruction to the EFE resection.

Long-term outcome for patients undergoing extensive EFE resection as part of left-ventricular rehabilitation are unknown. Medium-term results from patients undergoing neonatal Ross-Konno procedures have been mixed, and particularly poor results in patients requiring concomitant aortic arch reconstruction have been noted. However, a recent large single-center experience [11] suggested that overall survival and functional status may represent improvement compared with single-ventricle palliation or neonatal transplantation. Certainly long-term follow-up data, including evaluation of functional and neurodevelopmental status and comorbidities (including pulmonary vascular disease) will be needed before there is widespread adoption of this type of approach. However, feasibility of induction of significant left heart structure growth and significant myocardial recovery allowing restoration of biventricular circulation is certainly suggested by our case and deserves further study.

References

1. Ballard G, Tibby S, Miller O, Krasemann T, Rosenthal E, Anderson D et al (2010) Growth of left heart structures following the hybrid procedure for borderline hypoplastic left heart. Eur J Echocardiogr 11:870–874
2. Brown SC, Boshoff D, Eyskens B, Gewillig M (2009) Hybrid approach as bridge to biventricular repair in a neonate with critical aortic stenosis and borderline left ventricle. Eur J Cardiothorac Surg 35:1080–1082
3. Eghtesady P, Michelfelder E, Altaye M, Ballard E, Hirsh R, Beekman RH 3rd (2007) Revisiting animal models of aortic stenosis in the early gestation fetus. Ann Thorac Surg 83:631–639
4. Emani SM, Bacha EA, McElhinney DB, Marx GR, Tworetzky W, Pigula FA et al (2009) Primary left ventricular rehabilitation is effective in maintaining two-ventricle physiology in the borderline left heart. J Thorac Cardiovasc Surg 138:1276–1282
5. Han RK, Gurofsky RC, Lee KJ, Dipchand AI, Williams WG, Smallhorn JF et al (2007) Outcome and growth potential of left heart structures after neonatal intervention for aortic valve stenosis. J Am Coll Cardiol 50:2406–2414
6. Makikallio K, McElhinney DB, Levine JC, Marx GR, Colan SD, Marshall AC et al (2006) Fetal aortic valve stenosis and the evolution of hypoplastic left heart syndrome: patient selection for fetal intervention. Circulation 113:1401–1405
7. McCrindle BW, Blackstone EH, Williams WG, Sittiwangkul R, Spray TL, Azakie A et al (2001) Are outcomes of surgical versus transcatheter balloon valvotomy equivalent in neonatal critical aortic stenosis? Circulation 104:I152–I158
8. McElhinney DB, Marshall AC, Wilkins-Haug LE, Brown DW, Benson CB, Silva V et al (2009) Predictors of technical success and postnatal biventricular outcome after in utero aortic valvuloplasty for aortic stenosis with evolving hypoplastic left heart syndrome. Circulation 120:1482–1490
9. Salih C, Sheppard MN, Ho SY (2004) Morphometry of coronary capillaries in hypoplastic left heart syndrome. Ann Thorac Surg 77:903–907
10. Sharland GK, Chita SK, Fagg NL, Anderson RH, Tynan M, Cook AC et al (1991) Left ventricular dysfunction in the fetus: relation to aortic valve anomalies and endocardial fibroelastosis. Br Heart J 66:419–424
11. Shinkawa T, Bove EL, Hirsch JC, Devaney EJ, Hye RG (2010) Intermediate-term results of the Ross procedure in neonates and infants. Ann Thorac Surg 89:1827–1832
12. Smolich JJ, Walker AM, Campbell GR, Adamson TM (1989) Left and right ventricular myocardial morphometry in fetal, neonatal, and adult sheep. Am J Physiol 257:H1–H9