

Biventricular Repair in Unbalanced Atrioventricular Septal Defect

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We present the case of a 2.5 year-old child with unbalanced atrioventricular septal defect due to a small left ventricle (LV) (mitral annulus of 10mm and a 0.4 ratio in relation to the tricuspid annulus, LVDD: 17 mm, LV Vd2: 15 ml/m² and LV/RV long-axis ratio of 0.71); he had a favorable outcome after biventricular surgical repair. Normal LV development was observed three months after the operation (mitral annulus of 22 mm, with a 0.84 ratio in relation to the tricuspid annulus, and LVDD of 30 mm). Current parameters for utilization of the hypoplastic ventricle are discussed.

Introduction

When surgical repair of unbalanced atrioventricular septal defect is performed, the question that arises is whether or not to use the smallest ventricle as a useful element for further ensuring effective biventricular circulation¹⁻⁴. This challenge becomes even greater when the left ventricle is the smallest one, especially in the absence of an interventricular septal defect^{2,4}.

There is controversy regarding the adequate management for such cases, with no guiding elements available to establish the best approach. The motivation for and interest in reporting this case stemmed from the favorable outcome achieved with biventricular repair of an unbalanced atrioventricular septal defect with small left ventricle and minimal interventricular septal defect.

Case report

A 25-month-old boy without Down syndrome had previously undergone pulmonary artery banding at two months of age and had worsening dyspnea for four months. On physical examination he had no respiratory distress, his mucous membranes were pink and his pulses were normal. His weight was 9600g, and height 77 cm. Oxygen saturation was 94% and blood pressure was 92/56 mmHg. Chest examination revealed mild systolic impulses, accentuated heart sounds, and a holosystolic murmur along the left sternal border. The

liver was palpable 5cm below the right costal margin and xiphoid process. The electrocardiogram showed left anterior hemiblock and right ventricular systolic overload, with qR complex in V1 and negative T waves from V1 to V4 (Figure 1). Chest radiography revealed enlarged cardiac silhouette due to enlarged right chambers, straightened pulmonary artery shadow and increased pulmonary vasculature. Echocardiography showed a large ostium-primum atrial septal defect, small inlet ventricular septal defect (VSD), and moderate tricuspid regurgitation. The left chambers were small; the left atrioventricular valve ring had 10mm, with a 0.4 ratio in relation to the 25-mm ring of the right atrioventricular valve. The Z-values of the left and right atrioventricular valves were - 3.5 and +1.95, respectively. The left ventricular (LV) diastolic diameter and LV end-diastolic volume were 17mm and 15 mL/m², respectively. The right ventricle (RV) was dilated, with 26mm (Figure 2). Cardiac catheterization showed elevated pressures and pulmonary flow greater than systemic flow (RA=LA: 17, RV=LV: 100/17, PT: 50/25-33 mmHg; QP: 6.2, QS: 2.5 and RP: 1.6 U). Angiography showed the left ventricle displaced by the dilated right ventricle; the long-axis ratio, as measured from the ventricular apex to the semilunar valve between the LV (60mm) and RV (84mm) was 0.71 (Figure 2).

During operation, a large ostium-primum atrial septal defect (ASD) and a small VSD were closed with bovine pericardial patches. Suture of the left atrioventricular valve fissure, right atrioventricular valvuloplasty with ring reduction, suture of the anterior and posterior valve commissure, and removal of the pulmonary band with end-to-end anastomosis were also performed. The left atrioventricular valve and LV were small. The immediate postoperative period was complicated by low cardiac output, acute renal failure, and infection. Ten days later, the patient was stabilized thanks to the use of vasoactive drugs at variable doses (epinephrine 0.5 to 2.5 mcg/Kg/min, dobutamine 5 to 20 mcg/Kg/min, and milrinone 0.4 to 1 mcg/Kg/min), peritoneal dialysis, and pump infusion of furosemide at high doses (8 mg/Kg/day), in addition to broad-spectrum antibiotic therapy. The patient was discharged after 16 days, receiving carvedilol and furosemide. During this period, the left atrioventricular valve ring increased to 14mm, as did the LV cavity. Echocardiography performed three months later revealed slightly dilated ventricular cavities (RV: 21mm, LV: 30mm), left atrioventricular valve ring of 22mm, and right atrioventricular valve ring of 26mm, moderate left atrioventricular valve regurgitation, mild right atrioventricular valve regurgitation, with favorable clinical conditions despite a mild systolic murmur in the mitral area and liver 2cm below the right costal margin, with the patient still on furosemide and carvedilol. ECG showed new left potentials with QRS complex in V5 and V6, which did not exist before (Figure 1).

Key Words

Heart septal defects, ventricular; heart defects, congenital; heart ventricles.

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Manuscript received June 25, 2008; revised manuscript received August 27, 2008; accepted September 26, 2008.

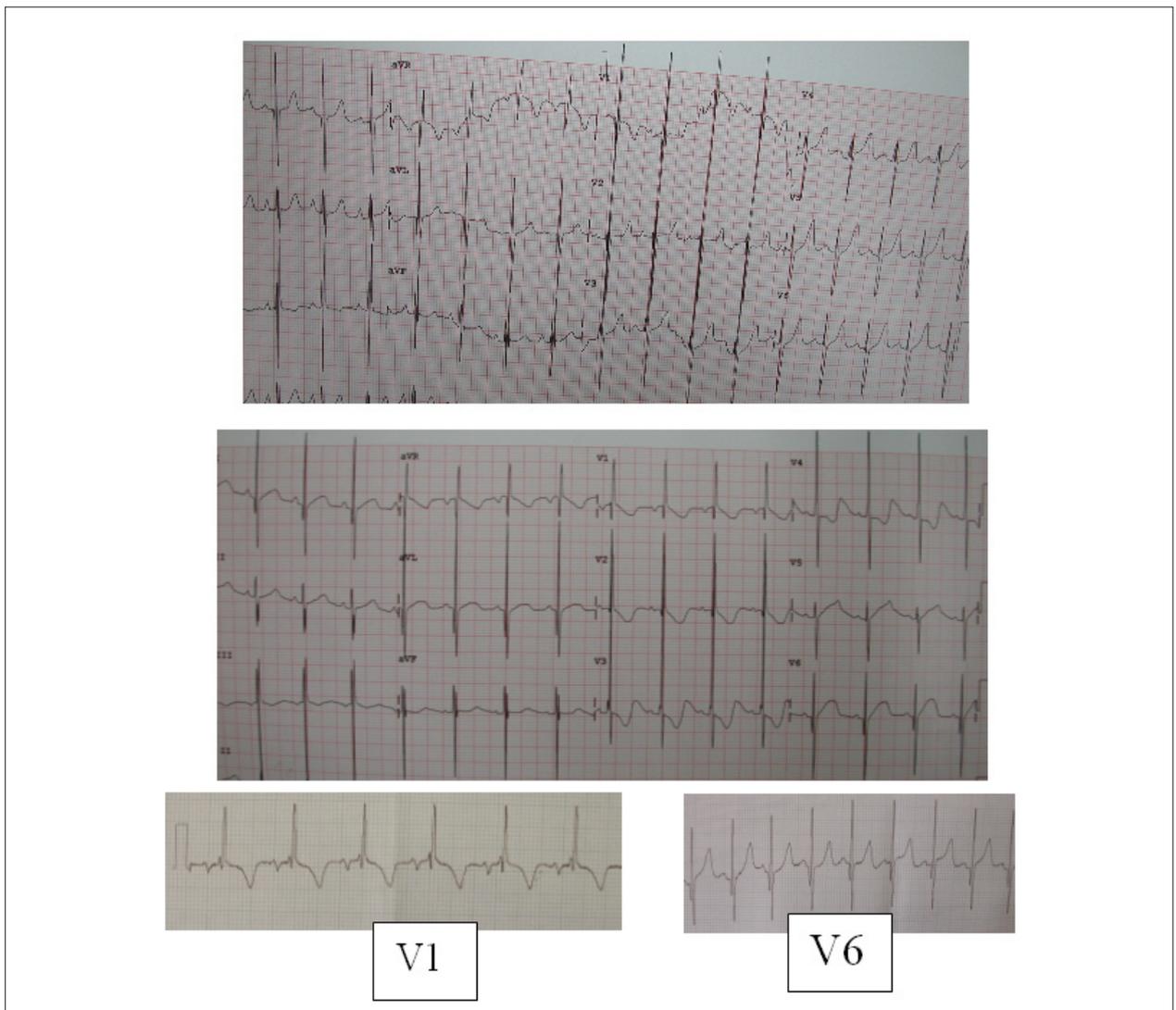


Fig. 1 - Eletrocardiograma em período pré-operatório (acima), pós-operatório imediato (meio) e três meses após a correção (abaixo) com nítido aparecimento de potenciais de ventrículo esquerdo, além da diminuição dos potenciais de ventrículo direito.

Discussion

The choice of the surgical technique to be used for the correction of atrioventricular septal defect with small LV is based on parameters not yet well defined and thus uncertain outcome could be presumed.

Repair using both ventricles was chosen in this case considering that the patient had survived to two years of age thanks to the output of the hypoplastic LV itself, which resulted from the pulmonary output subtracted from left-to-right blood shunt through the large ASD. This is a clinical argument for biventricular repair. This choice was feasible, although the left atrioventricular valve ring corresponded to 0.4 in relation to the right atrioventricular valve; the LV was clearly reduced and posteriorly displaced, as observed in an echocardiographic image.

Another element that made us choose this approach was the ratio between the long axes of both ventricles, from the

apex to the semilunar valves, which was 0.71 (RV: 84mm and LV: 60mm). As regards this index, Delmo Walter et al⁴ recommend the utilization of the LV in biventricular repair in cases with ventricular unbalance with long-axis ratios always higher than 0.64. Other authors have suggested this procedure for similar patients, based on this long-axis ratio^{2,3}. Following this recommendation, two deaths were observed among 19 patients with small LV operated on⁴.

LV end-diastolic volumes lower than 20 mL/m² should also be considered as prohibitive for repair, although survival in a case of LV with a volume as small as 6.8 mL/m² had been achieved².

Furthermore, based on the alignment of the atrial and ventricular septa and the larger LV volume demonstrated in the right anterior oblique view on angiography, we could also presume the probable greater volumetric LV development and adequacy after biventricular repair.

Case Report

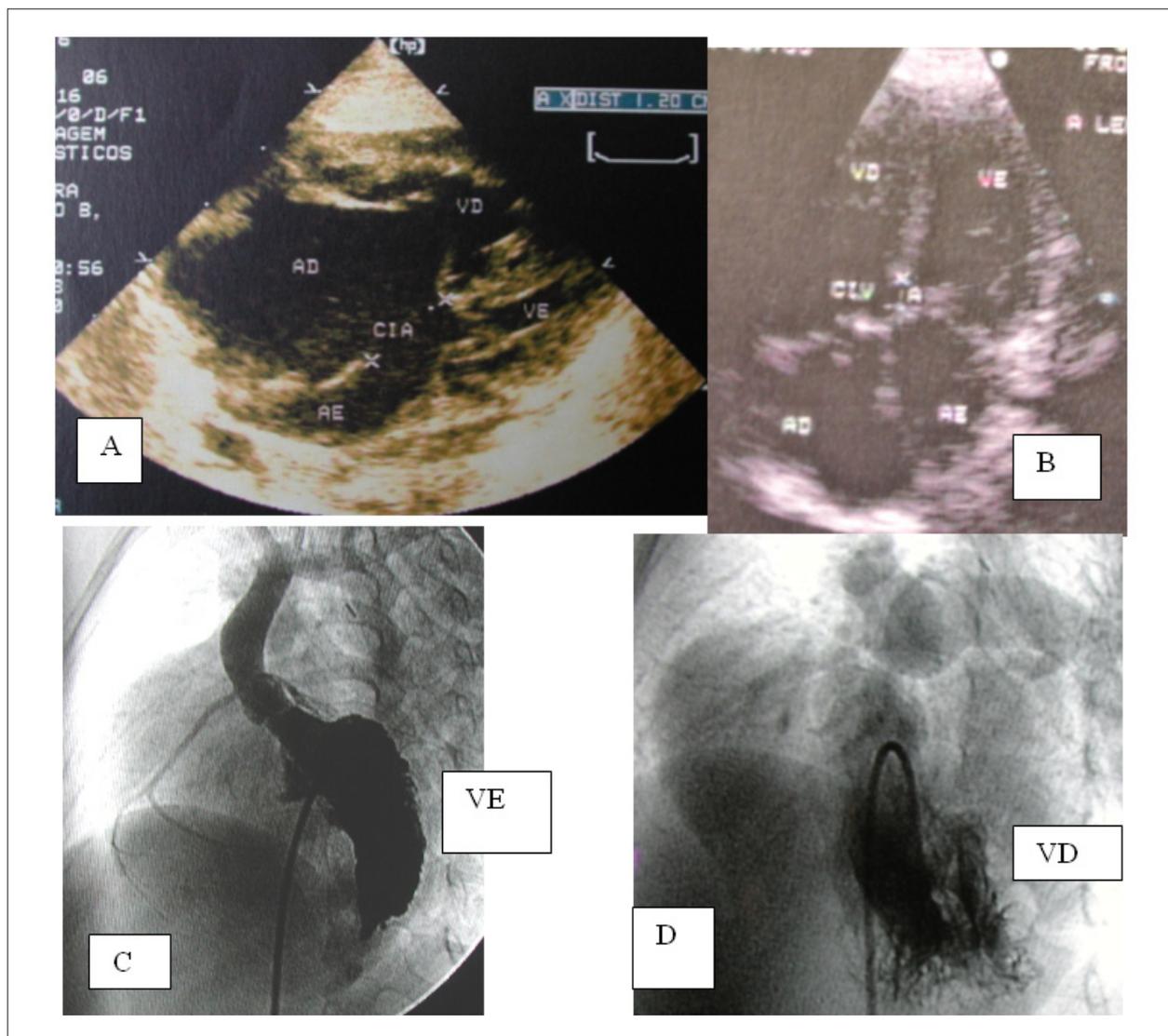


Fig. 2 - Imagens ecocardiográficas em projeção de quatro câmaras em A (pré-operatório) e em B (três meses após a correção) com contraste nítido do tamanho das cavidades esquerdas. A angiografia em projeção oblíqua anterior esquerda no pré-operatório (C e D) mostra o ventrículo esquerdo deslocado pelo direito, dilatado e mais longo.

After a difficult immediate postoperative period of LV adaptation with low output syndrome and renal failure, hemodynamic stability was achieved and the patient was discharged on day 16, in view of the increase in the LV and left atrioventricular valve ring (14mm), the latter with a 0.76 ratio in relation to the right atrioventricular valve (20mm). Further development of these left structures was observed three months after operation, with a left atrioventricular valve ring of 22mm and LV diastolic diameter of 30mm. However, no study has yet established a clear limit for the parameters used in the choice of whether or not to perform biventricular repair. Cohen MS and Spray TL³ established a ratio between the left/right atrioventricular valve rings lower than 0.67 to define unbalanced ventricles. According to Vida VL et al², biventricular repair may be adequately indicated even with a lower than 0.28 ratio between these atrioventricular structures.

Measurement of the right and left atrioventricular valve rings in atrioventricular septal defect, when correlated to body surface^{5,6}, is another parameter for the diagnosis of ventricular unbalance. Although not yet as widely established for atrioventricular septal defect as it is for many other anomalies, this ratio may also be useful for the definitive choice between univentricular and biventricular correction. In the present case, the Z-value of the left atrioventricular valve was -3.5, thus characterizing valve hypoplasia.

In order to minimize the outcome risks and to establish the most adequate management for each case of anomalies accompanied by ventricular unbalance, all these indexes should be analyzed as accurately and strictly as possible.

The efficacy of Intensive Care Units and Cardiac Recovery Units in the postoperative management helps achieve the ultimate success of similar risk procedures. For

this purpose, a well-planned, complete and experienced care structure is required for the control of fluid balance, drug therapy, mechanical ventilation, and treatment of expected complications. Likewise, a reliable laboratory diagnosis and ancillary test complex is necessary, especially when circulatory assistance with extracorporeal membrane oxygenation (ECMO) is indicated.

Acknowledgement

We thank the medical team of the pediatric intensive care unit of *Hospital Sírio-Libanês* of Sao Paulo, for the effective care and strict professionalism dedicated to this patient.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any post-graduation program.

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