Cardiol Young 2006; 16: 92–94

ISSN 1047-9511
doi: 10.1017/S1047951105002180

Brief Report

Atrioventricular septal defect with intact septal structures presenting as left atrioventricular valvar insufficiency

Ulisses Alexandre Croti,[1] Carlos Henrique De Marchi,[1] Vera Demarchi Aiello[2]

[1]Pediatric Cardiac Surgery, Hospital de Base da Faculdade de Medicina de São José do Rio Preto; [2]Laboratory of Pathology, Heart Institute (InCor), University of São Paulo Medical School, São Paulo, Brazil

Abstract We describe the findings in a six-year-old girl who presented with signs of left atrioventricular valvar insufficiency. The echocardiogram showed a common atrioventricular junction, intact atrial and ventricular septal structures. At surgery, the left-sided atrioventricular valve was found to be tri-foliate, and corrected by valvoplasty. To the best of our knowledge, this is the first case of atrioventricular septal defect with common atrioventricular junction and intact septal structures diagnosed during life.

Keywords: Atrioventricular canal malformation; congenital heart defect; valvoplasty

The anatomical hallmarks of hearts with atrioventricular septal defects are well known. In most instances, the bridging leaflets of the common valve are attached so as to separate the septal defect into atrial or ventricular components, or else they float, permitting atrial and ventricular shunting. Rarely, however, the septal structures can be intact even in the setting of a common atrioventricular junction. To date, as far as we are aware, such cases have been diagnosed only at autopsy.[1,2] We describe here such a patient diagnosed during life.

Case report

A six-year-old white girl presented with recurrent respiratory infections and failure to thrive during the first year of life. The parents reported that she remained asymptomatic up to the age of 5 years, when she became dyspnoeic on exertion, and a cardiac murmur was heard. On physical examination, the patient was in good general health, eupnoeic, and acyanotic. Cardiac auscultation revealed a pansystolic murmur in the mitral area, and a protosystolic click. On examination the lungs were normal. The liver was palpable at the right costal margin. The peripheral pulses were all present and symmetrical.

The electrocardiogram demonstrated a junctional rhythm, a heart rate of 75 beats per minute, and the QRS frontal axis at minus 30 degrees.

The chest X-Ray showed a cardiothoracic index equal to 0.55. The right heart border was prominent. There was also enlargement of the left atrial appendage, with a prominent pulmonary trunk. The pulmonary parenchyma showed mildly increased vascular markings.

The echocardiogram demonstrated usual atrial arrangement with a left-sided heart, normally connected systemic and pulmonary veins, and concordant atrioventricular and ventriculo-arterial connections. The arterial duct was closed. The interatrial septum was intact, but appeared thin in the juxtavalvar area. There was a common atrioventricular junction with two valvar orifices, but the ventricular septum was also intact. The left atrioventricular valve showed a trifoliate configuration, and remarkable regurgitation, with moderate dilation of the left-sided chambers. The aorta was unwedged relative to the atrioventricular valves, and there was disproportion between the inlet and outlet dimensions of the ventricular septum (Fig. 1).

The child was prepared for surgery under hypothermic cardiopulmonary bypass and cardioplegia at 28 degrees Celsius, approaching through a median sternotomy. The integrity of the atrial septum was

Correspondence to: Vera D. Aiello MD, Laboratory of Pathology, Heart Institute (InCor), University of São Paulo Medical School, Av. Dr. Enéas C. Aguiar, 44, São Paulo, SP, 05403-000, Brazil. Tel: +55 11 3069 5251; Fax: +55 11 3069 5279; E-mail: vera.aiello@incor.usp.br

Accepted for publication 3 October 2005

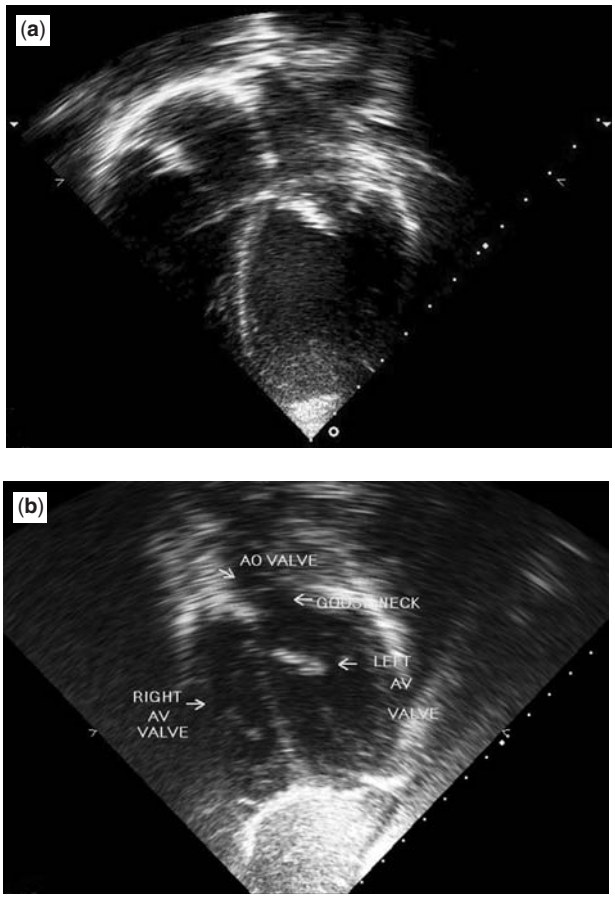

Figure 1.
*Echocardiography in the apical four-chamber view (**a**) demonstrates a common atrioventricular junction and intact septal structures. The subcostal coronal view of the left ventricle (**b**) shows the unwedged aorta and the right and left atrioventricular orifices.*

confirmed, albeit that the region close to the right atrioventricular valve was thinner than usual. A longitudinal incision in the atrial septum permitted observation of the left atrioventricular valve, which was tri-foliate. The valve was repaired by suturing closed the zone of apposition between the left ventricular components of the bridging leaflets using 5–0 polypropylene interrupted sutures (Fig. 2). The patient was discharged on the seventh postoperative day, and follow-up echocardiography revealed only mild regurgitation across the left atrioventricular valve, with intact septal structures.

Discussion

The atrioventricular septal defect, or atrioventricular canal malformation, is a frequent cardiac malformation characterized by a common atrioventricular junction and disproportion of the inlet and outlet dimensions of the ventricular septum. There is

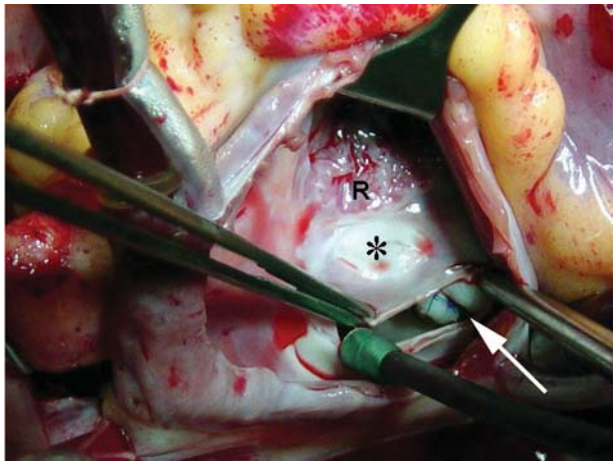

Figure 2.
*Surgical view of the right atrium, with a longitudinal incision of the atrial septum near the oval fossa, showing the right (**R**) and left atrioventricular valves. The latter was sutured in order to close the zone of apposition between the bridging leaflets (**arrow**). The atrial septum at the atrioventricular junction (*) appeared thinner than usual.*

variation concerning the number of valvar orifices within the common junction, and the presence of shunts between the atrial and ventricular chambers. The commonest forms are, the ones with a common atrioventricular valve guarding the common junction, with both so-called "primum" and ventricular defects, and the forms with separate right and left orifices and shunting confined at the atrial level, the so-called "primum defect". In the latter, the left atrioventricular valve is tri-foliate, showing two medial leaflets and one mural leaflet. The concept of fusion of the superior and inferior bridging leaflets of the common valve with each other and to the crest of the ventricular septum, made it easy to understand this so-called "partial" form of the defect.[3] Rarely, cases have been described with an intact atrial septum in the context of a common atrioventricular valve.[4–6] It is exceedingly rare, however, to encounter cases with the anatomical hallmarks of the common junction in the absence of the potential for shunting. As far as we are aware, such cases have previously been described only at autopsy.[1,2]

Our patient is an example of this rare situation. The echocardiogram was fundamental for making the morphological diagnosis, allowing the appropriate interpretation of the unusual findings. The clinical differential diagnosis would include an isolated cleft of an otherwise normal mitral valve in the absence of the anatomical hallmarks of atrioventricular septal defect, and mitral regurgitation secondary to rheumatic heart disease, the latter being frequent in children from developing countries.

The findings at cross-sectional echocardiography are fundamental in defining the number of valvar

https://doi.org/10.1017/S1047951105002180 Published online by Cambridge University Press

orifices, the pattern of leaflet insertion of the bridging leaflets, ventricular dominance, and the level of shunting in atrioventricular septal defects with common atrioventricular junction.[7–8]

In order to rule out the presence of the "primum" defect, it is advisable to take a close and cautious analysis of the images as obtained in both subcostal and apical views, as well as the Doppler pattern of flow, due to the relatively limited resolution of the method.

The operation to repair the zone of apposition between the left ventricular components of the bridging leaflets, the so-called "cleft", was undertaken with the aim of making the valve competent, improving the function and hopefully avoiding the need for future valvar replacement, which in the long-term results in higher rates of both morbidity and mortality rates.[9]

This treatment was achieved by using direct sutures alone, placing interrupted stitches close to the edge of the valve, which was possible as there was no fibrosis on the edges of the zone of apposition between the leaflets. We avoided ring annuloplasty because of the risk of valvar stenosis, as the valve was shown to be competent in the intra-operative period after filling the left ventricular filling by injection of saline.[10]

References

1. Silverman NH, Ho SY, Anderson RH, Smith A, Wilkinson JL. Atrioventricular septal defect with intact atrial and ventricular septal structures. Int J Cardiol 1984; 5: 567–573.
2. Ho SY, Russell G, Gerlis LM. Atrioventricular septal defect with intact septal structures in a 74-year old. Int J Cardiol 1990; 26: 371–373.
3. Radermecker MA, Somerville J, Li W, Anderson RH, de Leval MR. Double orifice right atrioventricular valve in atrioventricular septal defect: morphology and extension of the concept of fusion of leaflets. Ann Thorac Surg 2001; 71: 358–360.
4. Smallhorn JF, Sutherland GR, Anderson RH, Macartney FJ. Cross-sectional echocardiographic assessment of conditions with atrioventricular valve leaflets attached to the atrial septum at the same level. Br Heart J 1982; 48: 331–341.
5. Atik E, Soares AM, Aiello VD. Common arterial trunk associated with atrioventricular septal defect. Cardiol Young 1999; 9: 617–620.
6. Rowley KM, Kopf GS, Hellenbrand W, Kleinman CS, Downing SE. Atrioventricular canal with intact atrial septum. Am Heart J 1988; 115: 902–906.
7. Smallhorn JF, Tommasini G, Anderson RH, Macartney FJ. Assesment of atrioventricular septal defects by two dimensional echocardiography. Br Heart J 1982; 47: 109–121.
8. Silverman NH, Zuberbuhler JR, Anderson RH. Atrioventricular septal defects: cross-sectional echocardiography and morphological comparisons. Int J Cardiol 1986; 13: 309–331.
9. Chauvaud S, Fuzellier JF, Houel R, Berrebi A, Mihaileanu S, Carpentier A. Reconstructive surgery in congenital mitral valve insufficiency (Carpentier's techniques): long-term results. J Thorac Cardiovasc Surg 1998; 115: 84–93.
10. Perier P, Clausnizer B. Isolated cleft mitral valve: Valve reconstruction techniques. Ann Thorac Surg 1995; 59: 56–59.