

# Aortic atresia with normally developed left ventricle in a young adult

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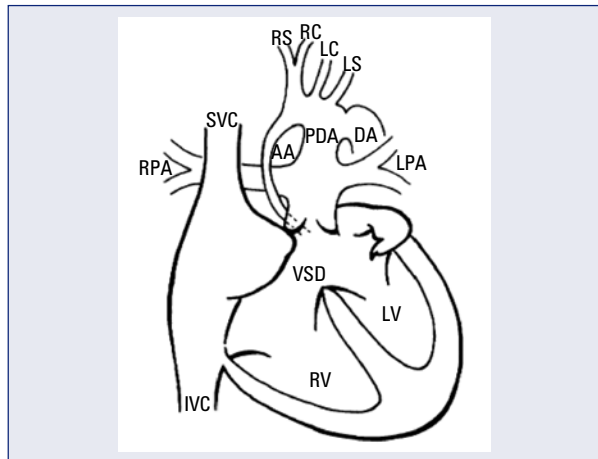
## Abstract

*Congenital aortic valve atresia is usually part of a developmental anomaly usually called 'hypoplastic left-heart syndrome'. To the best of our knowledge, most reported cases of aortic atresia have been associated with hypoplasia of the ascending aorta, of the left ventricle, and of the mitral valve. We report a patient presenting with aortic atresia with a ventricular septal defect and a normally sized left ventricle. (Cardiol J 2011; 18, 3: 304–306)*

**Key words:** aortic atresia, left ventricle, echocardiography

## Introduction

Aortic atresia is usually associated with hypoplasia of the left ventricle (LV) and mitral valve [1, 2]. However 4–7% of patients with aortic atresia have a ventricular septal defect (VSD) at the same time which may allow for the development of a normal mitral apparatus and LV (Fig. 1) [3]. In these patients, the VSD provides an 'outlet' for the developing LV and mitral valve in utero. The maintenance of flow across the mitral valve and through the LV allows for their normal growth and development [4]. Aortic atresia associated with an adequately developed LV and mitral valve has been recognized as a discrete anatomic-clinical entity [1, 2]. It was first described by Lev [5] and subsequently by Rosenquist et al. [6] in a case presenting with an aortopulmonary fenestration and interrupted aortic arch. This condition, which is relatively favorable for surgery, accounts for only a small proportion of all cases of aortic atresia. Roberts et al. [7], in reviewing 73 necropsy cases, found three hearts with normal LV and mitral valves (4%). The same frequency was reported by Freedom et al. [8] (six



**Figure 1.** Schematic diagram of a heart with aortic atresia, ventricular septal defect (VSD) and normally developed left ventricle and mitral valve; AA — ascending aorta; DA — descending aorta; IVC — inferior vena cava; SVC — superior vena cava; LS — left subclavian artery; LC — left carotid artery; RC — right coronary artery; RS — right subclavian artery; PDA — patent ductus arteriosus; LPA — left pulmonary artery; RPA — right pulmonary artery; LV — left ventricle; RV — right ventricle. Based on from [2].

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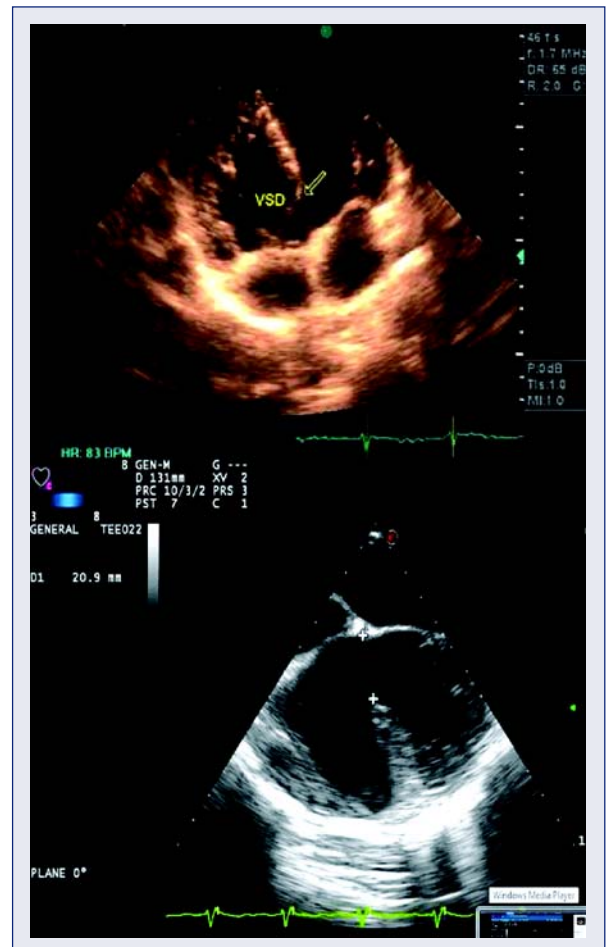
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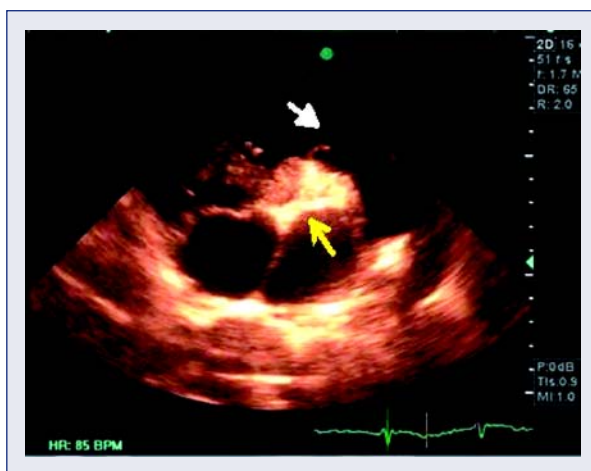
cases [4%] out of 148), Thiene et al. [2] reported a slightly higher percentage (four cases [7%] out of 58).

### Case report

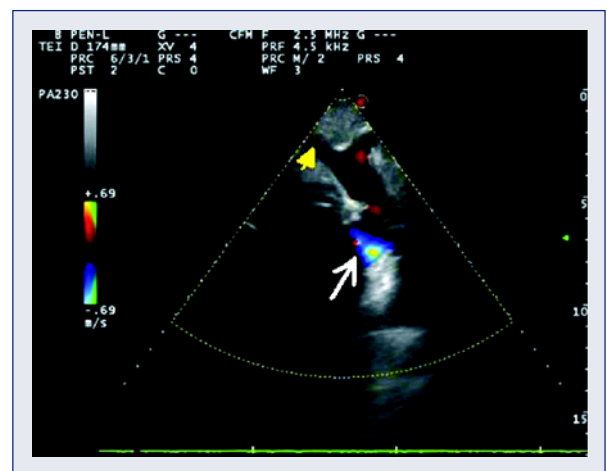
The patient was a 20 year-old man referred for echocardiography because of a history of mild fatigue and headache during exertion, but no dyspnea. On physical examination, blood pressure and heart rate were normal; pulse oxymetry showed an oxygen saturation of 85% at room air. In cardiovascular examination, the point of maximal impulse was displaced downwards and laterally; auscultation revealed a normal S1 with mildly accentuated S2 accompanied by a soft 3/6 systolic murmur in apex, left sterna border and base of heart. He had mild clubbing and no cyanosis. Other examinations were unremarkable. The electrocardiogram showed normal sinus rhythm, biatrial enlargement, extreme axis deviation and right ventricular hypertrophy. Transthoracic and transesophageal echocardiography showed situs solitus, D-loop, atrioventricular concordance, normal LV size and function, right ventricle enlargement and right ventricular hypertrophy, large outlet VSD (Fig. 2), absence of aortic valve and very small ascending aorta dilated pulmonary artery and its branches (Fig. 3), normal arch and branch vessels with retrograde flow from descending aorta, and a very large patent ductus arteriosus (Fig. 4). There was also a mild mitral regurgitation, moderate to severe tricuspid regurgitation, severe pulmonary hypertension (120 mm Hg), and small patent foramen oval.



**Figure 2.** Transthoracic (top) and transesophageal (bottom) four-chamber views revealing very large ventricular septal defect (VSD).



**Figure 3.** Parasternal short axis view shows dilated pulmonary artery (white arrow) and complete absence of aortic valve (yellow arrow).



**Figure 4.** Suprasternal long axis view shows large patent ductus arteriosus (white arrow) and distal portion of aortic arch (yellow arrow). Color Doppler flow reveals retrograde red flow through descending aorta.

## Discussion

Palliative or corrective surgery should be performed in the neonatal period, before irreversible changes in the pulmonary circulation. An increase in the pulmonary arterial diameter and muscularity may be apparent even within the first week of life [9]. The presence of the normal LV and persistent patency of ductus for early palliation pulmonary banding will be the required surgical procedure. Complete correction may be planned for a later stage. Our patient was a case of aortic atresia with normally developed left ventricle. However, considering systemic pulmonary hypertension (120 mm Hg) our patient was not a good candidate for surgical correction and at this stage only heart and lung transplantation should be considered. As with other forms of Eisenmenger syndrome, our patient has had a slowly progressive course with regard to the symptoms and signs along with electrocardiographic and hemodynamic data.

## Acknowledgements

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