

Eisenmenger's syndrome with complete AV septal defect in a 55-year-old woman

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Introduction

Eisenmenger syndrome is characterized by an elevation of pulmonary pressure to systemic levels caused by an increase in pulmonary vascular resistance, with a severe shunt (right-left) or bidirectional shunt. This was a consequence of an uncorrected congenital defect. Its diagnosis is based on the clinical history, symptoms, echocardiography, and cardiac catheterization.

Case Description

We present the case of a 55-year-old woman who was diagnosed with an atrioventricular septal defect after a cardiorespiratory arrest at 5 months of age. Given the high probability of complications, the family did not consent to the surgical correction of the heart defect. During her adolescence, Progressive dyspnea for great effort peripheral cyanosis, weight loss, and lipothymia episodes. At 18 years of age, she was diagnosed with a complete atrioventricular septal defect based on clinical and imaging

studies (echocardiography) Eisenmenger's syndrome (Figure 1 and 2). This case included an atrial and ventricular septum defect associated with a common atrioventricular valve (Figure 3). Without surgical indications, the patient rapidly started treatment for heart failure. Sometime later, she started bosentan, associated with sildenafil, for the treatment of severe pulmonary hypertension that had already developed over time, even knowing that at the date of diagnosis, her prognosis was not favorable, and the patient was managed to reach the age of 55 years due to the specific medication and outpatient follow-up. Conclusion Physicians must be aware of the existence of rare cases such as this. It is important to be capable of recognizing the symptoms and signs to obtain a detailed medical history, choose the right complementary exams to make a diagnosis, and immediately start the specific treatment. The response to treatment was surprising, and the benefit of bosentan was remarkable. It was possible to provide an extended lifetime and more quality time throughout the years.

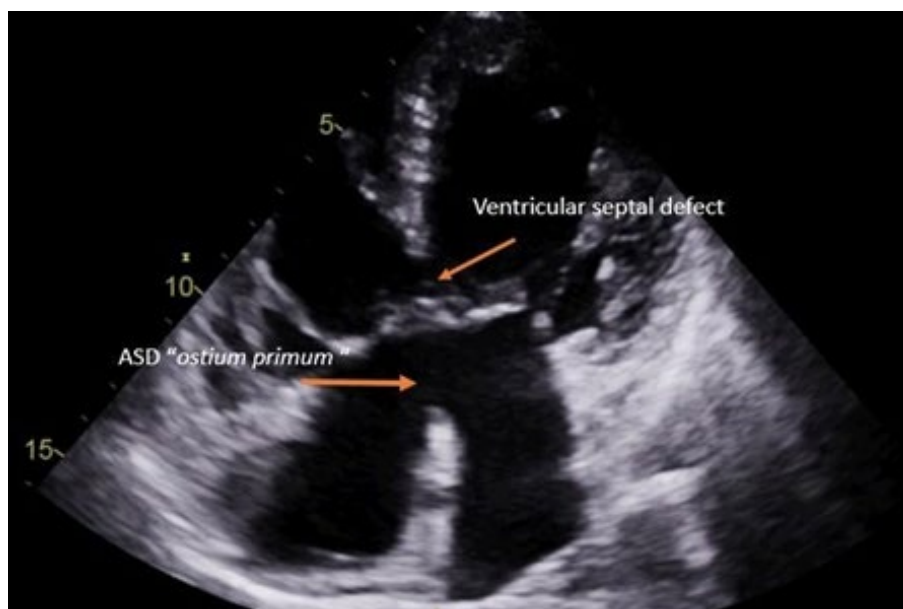


Figure 1: Atrial septal and inlet ventricular defects.

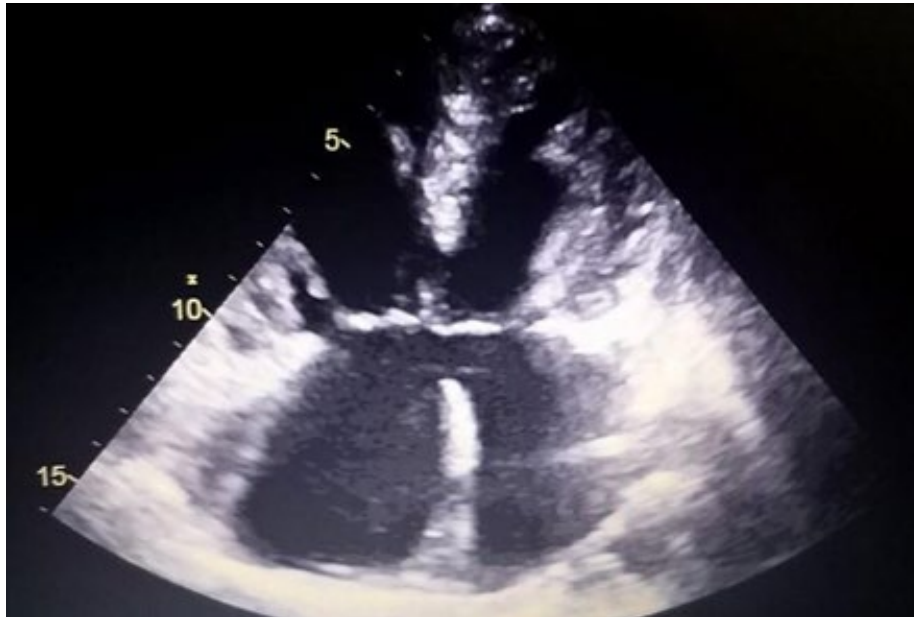


Figure 2: Atrial septal defect and inlet ventricular defect

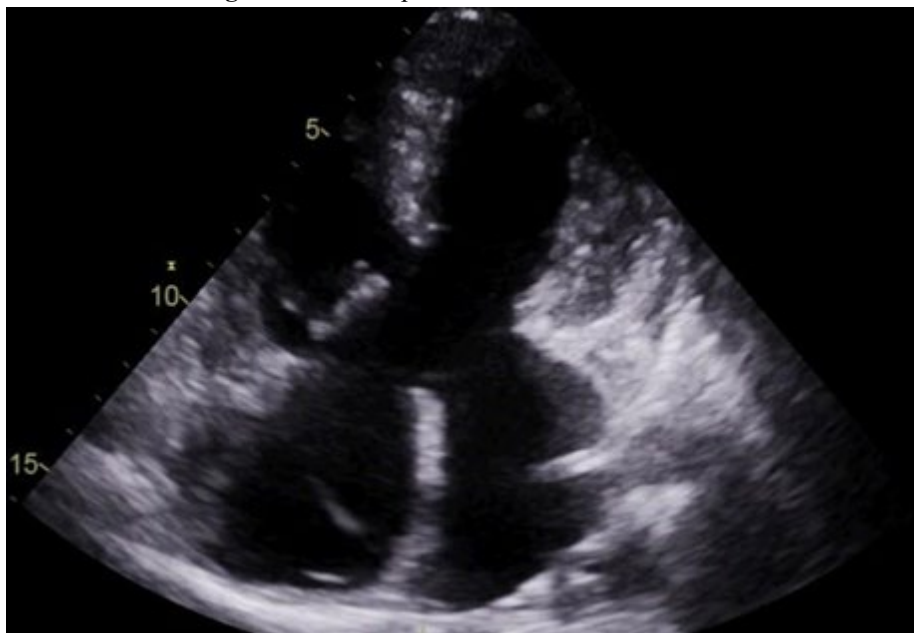


Figure 3: Complete atrioventricular septal defect with a common atrioventricular valve

Conflict of Interest

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Written informed consent was obtained from the patient for the publication of this report in accordance with the journal's patient consent policy.

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