Case Reports

Complete Atrioventricular Septal Defect and Pulmonary Stenosis

Diagnosed in a 49-Year-Old Woman after 10 Uneventful Births

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Atrioventricular septal defects constitute 4% of all congenital cardiac malformations. Patients with complete atrioventricular septal defect rarely survive for decades without surgical treatment. Pulmonary stenosis can provide a delicate balance between the pulmonary and systemic circulations and thereby increase longevity. We present the case of a 49-year-old woman whose complete atrioventricular septal defect and associated pulmonary stenosis were diagnosed only after she had given birth to 10 live children through uneventful spontaneous delivery. We discuss her successful surgical treatment in terms of the available medical literature. ***(Tex Heart Inst J 2015;42(2):166-8)***

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Atrioventricular septal defect (AVSD) can be partial, intermediate, or complete. Life expectancy varies according to the type of disease. Patients with complete AVSD rarely survive for decades without surgical treatment. Associated pulmonary stenosis is usually present in 2% of pediatric patients and 5.8% of adults with complete AVSD.[1] Pulmonary stenosis reduces pulmonary blood flow and protects the lungs from irreversible vascular changes. Patients with complex congenital cardiac anomalies and pulmonary stenosis might have balanced pulmonary and systemic circulations and thus live life with minimal or no symptoms.

Few published reports of AVSD involve pregnancy and delivery.[2] We report the case of a 49-year-old woman whose complete AVSD and pulmonary stenosis were diagnosed only after she had given birth to 10 live children through uneventful spontaneous delivery. We discuss her surgical treatment in view of the available medical literature.

Case Report

In January 2013, a 49-year-old woman was admitted to our hospital with a 6-month history of dyspnea during light exercise. She had given birth to 10 live children through normal spontaneous delivery, without sequelae. Her last child had been born 3 years earlier. She was in New York Heart Association (NYHA) functional class II–III. At presentation, she had a grade 3/6 or 4/6 systolic murmur, audible on the left parasternal border and radiating to the axilla. Her blood pressure was 125/60 mmHg, her pulse rate was 85 beats/min, and her peripheral oxygen saturation was 87%. An electrocardiogram showed sinus rhythm. A chest radiograph revealed a cardiothoracic ratio of 70%. A transthoracic echocardiogram showed a large ostium primum atrial septal defect, a large interventricular septal defect, and a common atrioventricular (AV) orifice (Fig. 1). Doppler studies revealed severe regurgitation of the left AV valve, and low-lying pulmonary stenosis with a 105-mmHg pressure gradient (Fig. 2). Cardiac catheterization revealed a normal pulmonary artery pressure (19/17 mmHg) and a large, nonrestrictive ventricular septal defect with equal right ventricular (RV) and left ventricular pressures. The Qp/Qs ratio was 1.79. No coronary artery disease was detected.

The patient underwent surgery. Transatrial resection was performed to relieve the RV outflow tract (RVOT) obstruction. A modified single-patch technique involved the use of glutaraldehyde-treated autologous pericardium. The cleft of the left AV valve was closed with use of interrupted sutures. A 31-mm valvuloplasty ring was implanted in the left AV valve for improved coaptation. After the patient was weaned from cardiopulmonary bypass, transesophageal echocardiograms showed competent AV valves and

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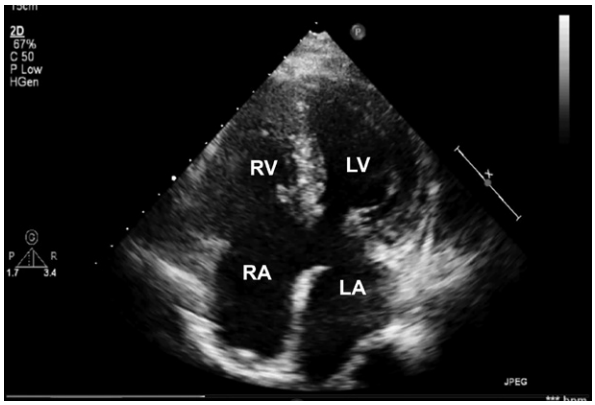

Fig. 1 *Preoperative transthoracic echocardiogram (4-chamber view) shows a large ostium primum atrial septal defect, a large interventricular septal defect, and a common atrioventricular orifice.*

LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle

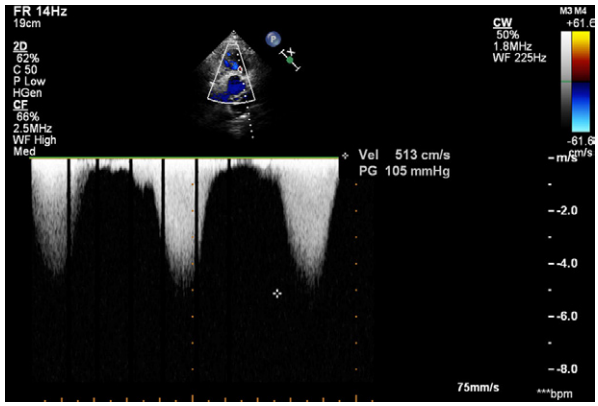

Fig. 2 *Preoperative 2-dimensional echocardiogram (color-flow Doppler mode) shows severe regurgitation of the atrioventricular valve and low-lying pulmonary stenosis with a 105-mmHg pressure gradient.*

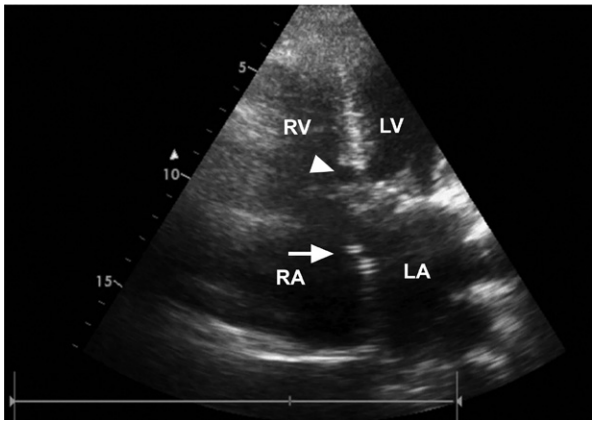

Fig. 3 *Postoperative transthoracic echocardiogram (4-chamber view) shows the pericardial patch in the atrial septum (arrow) and the closed interventricular septum (arrowhead).*

LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle

an unobstructed RVOT. Her postoperative course was uneventful except for intermittent complete AV block; a permanent pacemaker was implanted on postoperative day 15.

The patient was discharged from the hospital on the 21st postoperative day. After 3 months, echocardiograms revealed trivial left and mild right AV-valve insufficiency without residual shunting or RVOT obstruction. Figure 3 shows the results of the repair. The patient had NYHA class I functional capacity at that time and again 2 years postoperatively.

Discussion

The natural history of complete AVSD has been well documented in nonsurgically treated patients. Berger and colleagues[3] reported a survival rate of 4% in patients older than 5 years of age; the mortality rate gradually declined after a few months of life, reaching a near-plateau at 36 months. Few reports exist about the diagnosis of complete AVSD in later decades of life. In a review of 139 patients whose AVSD was diagnosed at autopsy, Tandon and co-authors[4] noted the oldest patient to be 46 years of age.

The lifespan of patients with AVSD usually depends on the balance between the pulmonary and systemic circulations, which is affected by the presence and degree of pulmonary stenosis in patients who have an unrestrictive ventricular septal defect or a single ventricle.[5]

Pregnancy itself is a circulatory burden chiefly because of the volume load, which has a substantial impact even in healthy women.[6] Pregnancy in women who have congenital heart disease (CHD) is associated with maternal cardiac sequelae and various neonatal sequelae. In a study of 49 pregnancies in 34 women who had CHD,[7] maternal cardiac sequelae were noted in 18.4% of the patients, pulmonary edema in 16.3%, symptomatic arrhythmia in 6.1%, deterioration of NYHA function by 2 classes or more in 2%, and cardiac death in 2%. Independent predictors of adverse maternal cardiac events were NYHA functional class III or IV (odds ratio [OR], 20.3), RV dilation (OR, 21.2), and pulmonary hypertension (OR, 21.8). Neonatal sequelae, which occurred in 22.4% of those pregnancies, included preterm delivery (16.3%), small birth size in terms of gestational age (12.2%), and death (2%). Independent predictors of adverse neonatal events were pulmonary hypertension (OR, 6.8) and NYHA functional class III or IV (OR, 23).[7] In a review of 2,491 pregnancies in women with structural CHD,[6] the prevalence of miscarriage was reportedly 15%; of elective abortion, 5%; and of significant cardiac sequelae, 11%. Drenthen and colleagues[2] studied 79 patients who had isolated and balanced complete AVSD and concluded that the pregnant women were prone to NYHA-class deterioration and worsening of AV regurgitation.

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We concluded that our patient's pulmonary stenosis balanced her pulmonary and systemic blood flows perfectly, enabling her to remain asymptomatic until 6 months before presentation. To our knowledge, our patient is the first with complete AVSD and pulmonary stenosis to have produced 10 live children through normal delivery without sequelae.

In patients with complex cardiac anomalies who survive into adulthood because of balanced pulmonary and systemic circulations, symptoms can develop at any time because of worsening ventricular function, increased AV valvular regurgitation, arrhythmias, or endocarditis.[5,8] Complex CHD in adult patients is usually associated with high surgical risk, and medical therapy might be preferable to surgical intervention in asymptomatic patients. However, our patient's declining functional capacity was associated with increased left AV-valve regurgitation, which necessitated reoperation. This experience leads us to recommend surgery in symptomatic patients of any age, when repair is feasible.

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