Aortic Valve Atresia with a Normally Developed Left Ventricle*

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Aortic valve atresia is usually associated with hypoplasia of the left ventricle and mitral valve. We report a patient presenting aortic atresia with ventricular septal defect and a normally sized left ventricle. The electrocardiogram showed left axis deviation and left ventricular hypertrophy. The ventricular septal defect probably favored a normal growth of the left ventricle, allowing it to unload with systole.

Congenital aortic valve atresia is usually part of a developmental anomaly most frequently called "hypoplastic left-heart syndrome." To our knowledge, all reported cases of aortic atresia were associated with hypoplasia of the ascending aorta, of the left ventricle, and of the mitral valve. We describe here a case of aortic atresia accompanied by a well-formed left atrium, mitral valve, and left ventricle. In the presence of classical aortic atresia, there is an intact ventricular septum and a competent mitral valve; this traps the left ventricle and characteristically it is hypoplastic. In contrast, in our case, aortic atresia was associated with a ventricular septal defect, which allowed the left ventricle to exercise its normal function of expelling blood and accounted for the fact that the chamber was of normal size.

Case Report

The patient was born on Dec 20, 1973, after an uneventful full-term pregnancy to a para 1, gravida 1 mother who was 23 years old. Birth weight was 3,500 gm (7 lb, 11 oz). The

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Apgar score was 8 at one minute. Because of tachypnea, dyspnea, and cyanosis, the patient was transferred to our unit at 30 hours of age. Physical examination on admission revealed a well-nourished full-term newborn male infant with minimal cyanosis but marked respiratory distress. Heart rate was 168 beats per minute; respiratory rate was 80 per minute; and arterial pressure by the "flush" technique was 60 mm Hg and 35 mm Hg at the right wrist and ankle, respectively. Fine rales were heard over both lungs. The liver was palpable about 5 cm below the right costal margin. There was a quiet precordium; the first sound was normal, and the second sound at the base of the heart was loud and single. There was a moderately loud third sound at the apex with a gallop rhythm. A grade 3/6, harsh, long systolic murmur was best heard along the left sternal border. A chest x-ray film disclosed moderate cardiomegaly and increased pulmonary vascularity.

An electrocardiogram (Fig 1) showed a probable junctional tachycardia at a rate of 120 to 130 per minute, a QRS axis of about —30°, dominant left ventricular electric forces, and marked ST-T wave changes. Despite intensive anticongestive treatment, the patient died of severe heart failure ten hours following admission.

Postmortem Examination

The postmortem examination (Fig 2) showed that there was situs solitus of the atria and viscera, with d-loop and cardiac apex pointing to the left. Two superior vena cavae were present, the left one draining into the coronary sinus. A 6 × 4-mm atrial septal defect was situated at the level of the fossa ovalis. The right ventricle was slightly enlarged. A 5 × 7-mm intracristal ventricular septal defect was present. A very large pulmonary artery emerged anteriorly from the right ventricular cavity. The right and left pulmonary arteries arose normally from the pulmonary trunk which was continuous with the descending aorta through a large patent ductus arteriosus. The pulmonary veins drained normally into a well-developed left atrium. The mitral valve was normal in size and shape. The left ventricle presented a normal architecture and was moderately enlarged. Its cavity ended blindly at the level of the aortic valve, and its only outlet was via the ventricular septal defect.

![Figure 1. Electrocardiogram showing probable junctional tachycardia at a rate of 120 to 130 per minute, QRS axis of about —30°, dominant left ventricular electric forces, and marked ST-T wave changes.](image-url)
Aortic atresia is the most severe form of the hypoplastic left heart syndrome. Clinically, our case was very similar to most cases previously described. However, the patient’s electrocardiographic pattern unexpectedly showed a left axis deviation and a left ventricular hypertrophy. This might have suggested a tricuspid atresia, even though clinical findings favored the diagnosis of a hypoplastic left heart syndrome. In retrospect, the prevalence of left ventricular electric forces suggested the presence of a functional left ventricle, which was subsequently confirmed by postmortem examination.

Aortic atresia is usually included in the hypoplastic left heart syndrome, because all the cases thus far described have been associated with underdevelopment of all of the structures on the left side of the heart. The question then arises if this case should be considered a new entity or a variant of the hypoplastic left heart syndrome. The presence of this type of aortic atresia favors the existence of an anatomic spectrum of lesions including (1) an extreme form of aortic atresia with mitral atresia and severe hypoplasia of the left ventricle; (2) aortic atresia with a small mitral valve and less marked hypoplastic left ventricle, with or without ventricular septal defect; and (3) aortic atresia with ventricular septal defect and normal left side of the heart. In this last situation the presence of a large ventricular septal defect, which decompresses the left ventricle, is most probably responsible for a normal development and function of the structures on the left side of the heart. Despite the anatomic variations, practically all of the cases reported present the same clinical picture and course.

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REFERENCES