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Aortic Atresia with Normal Left Ventricle and Intact Ventricular Septum

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We report a case of aortic atresia with biventricular hypertrophy and death due to cardiac insufficiency (at 5½ days of age). In the necropsy a normally sized left ventricle was observed, with endocardial fibroelastosis, anomalous mitral arcade, intact interventricular septum and atresia of the aortic valve. The ascending aorta was moderately hypoplastic; there was a foramen ovale and persistent ductus arteriosus. We believe that this is the first case that has been reported of this anomaly occurring with a normal left ventricle and intact ventricular septum.

Aortic atresia has been classified in two types, depending on the development of the left ventricle. Type 1 has a hypoplastic left ventricle and atresia or stenosis of the mitral valve, and type 2 has a moderately enlarged left ventricle, hypoplasia or mitral atresia, and a ventricular septal defect.

The few cases described in the literature with good development of the left ventricle showed a ventricular septal defect, with the blood volume of the ventricle discharging through the septal defect in systole. In the review we have carried out, we have not observed any case of normal size of the left ventricle together with an intact ventricular septum. The only etiopathogenic explanation, to our knowledge, is that the aortic atresia is due to a late intrauterine closure of the valvular orifice caused by the joining of the raphes of the valves when the process of formation of the left ventricular cavity and the ventricular septum has been completed.

CASE REPORT

A male infant was born as the product of a full-term pregnancy and normal vaginal delivery. The infant was hospitalized at the age of 3½ days, due to respiratory distress. The delivery was the first childbirth of a 27-year-old mother who was in good health.

Physical examination revealed a birth weight of 3,290 gm and generalized subcyanosis. The respiration rate was 100/min, and the Silverman score was 5 to 6. There were no peripheral pulsations in the arms or legs. The first heart sound was weak, and a short grade 1/2 murmur was heard at the fourth left intercostal space, radiating to the base; the second heart sound was loud and single, with a gallop rhythm. The arterial pressure (flush technique) was decreased in the arms but was not detected in the legs. The liver was palpable 4 cm below the right costal margin.

Roentgenologic examination showed marked cardiomegaly and increased pulmonary vascularity. The right atrium was enlarged.

The electrocardiogram showed sinus rhythm at a rate of 167 beats per minute. The QRS axis was 110°. Right atrial and biventricular hypertrophy was diagnosed. The right ventricular complexes were Rs, and a small wave was noted in lead Va, with a depressed ST segment and negative T wave. In spite of the administration of diuretic drugs and digoxin, the infant died 28 hours after hospitalization.

Postmortem Study

The weight of the heart was 38 gm, and the great arteries appeared to be normally related (Fig 1). A patent ductus

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FIGURE 1. Anterior view of specimen, showing arteries normally related. Note normal size of open left and right ventricles. Ascending aorta is moderately hypoplastic.
The right and left coronary arteries were normal, and the epicardial distribution of the left anterior descending coronary artery suggested a normally developed left ventricle (Fig 1). The aortic arch was normal. The left ventricular volume \((1/6\pi \times L \times M \times N = 1/6\pi \times 3.2 \times 2.2 \times 2.2 = 8.07 \text{ cu cm})\) was acceptable.

**Discussion**

Clinically, our case corresponds to the “hypoplastic left-heart syndrome,” but the ECG suggested the presence of a well-developed left ventricle with alterations in repolarization. In some cases, investigators have encountered signs of left ventricular hypertrophy with a hypoplastic ventricle, due to the thickness of its muscular mass. To date, cases have been described showing a normal left ventricle or one of large size with a ventricular septal defect. The presence of a septal defect, in decompresing the left cavities, favored better development and functioning of the structures, with both ventricles behaving like a common chamber with similar pressures. (We accept the general theory that endocardial fibroelastosis develops provided the mitral valve is open, and we believe that the hypoxia existing in these cases contributes towards this.) The belief that the “precocious” closure of the foramen ovale may contribute to the underdevelopment of the left ventricle in suppressing the right-left fetal physiologic shunt is in disagreement with our case, in which the foramen ovale was only slightly permeable.

We believe that the theory to best explain the anatomic findings in this case would be that of the late intrauterine fusion of the commissures of the aortic valve, since the two commissures of the valve can be seen perfectly with their relative joining raphes; the malformation of the mitral valve might have contributed to the fusion of the aortic valves, which would exist initially with small rings. The interventricular wall and the left ventricle would be completely developed when the process of valvular fusion commenced development. This condition is amenable to surgical aortic valvotomy. We believe that this is the first case of aortic atresia with a normal left ventricle and an intact ventricular septum to be described in the literature.

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