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Congenital Aortic Stenosis in a 40 Day-Old Infant*

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Congenital aortic stenosis may show supravalvular, valvular or subvalvular forms and constitutes about 3-5 % of all congenital cardiac malformations.[1] Emergency surgical intervention is required in severe forms. The object of this paper is to present an infant with severe congenital valvular stenosis undergone emergency open heart surgery.

Case Report

B.Ç. (file number 1030023), a 40 day-old baby girl, was referred to Hacettepe Children's Medical Center with complaints of peroral cyanosis and breathing difficulty. The patient was afebrile, weighed 3800 gr. 3/6 degree pansystolic murmur was heard along the left sternal border, loudest on the 3rd and 4th intercostal space. Auscultation of the lungs revealed rales on both side. The liver was enlarged 4 cms. below the right costal margin. The rest of the physical examination was noncontributory. Chest X-ray revealed an enlarged cardiac shadow with mild pneumonic

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infiltration in both lung fields. ECG showed right axis deviation and right ventricular hypertrophy. He was digitalized. Pulmonary rales and peroral cyanosis disappeared. However, the patient had two episodes of cardiac arrest. He was resuscitated without any neurologic sequel. Valvular aortic stenosis with thickened aortic leaflets and poststenotic aortic dilatation was shown on heart catheterization. No shunt at any level was detected. The aortic valvular gradient was not determined because the general condition of the patient during the procedure did not permit to continue on left heart catheterization.

The patient underwent an emergency open heart surgery on February 8, 1979, using heart-lung machine, Bentley infant oxygenerator and DeBakey roller pumps under normothermic conditions. The aorta was clamped and a transverse aortotomy was performed. The aortic valvular orifice was 3 mm. in diameter, the leaflets were thickened and the valve was bicuspid. An aortic valvular commissurotomy was performed. The immediate postoperative left ventricular pressure was 100 mmHg, aortic pressure 88 mmHg and left atrial pressure 14 mmHg. The duration of the bypass was 16 minutes.

The postoperative course was uneventful and he was discharged in good condition. Follow-up examination one year later revealed satisfactory growth and development. 1/6 degree aortic systolic murmur was present.

Discussion

Congenital aortic stenosis which constitutes about 3-5 % of all congenital cardiac malformations, is associated with additional anomalies in 8-30 % of the cases.[1-4] These additional malformations lead to early operative interventions especially in infants. The additional cardiovascular anomalies in congenital aortic stenosis are: aortic coarctation, patent ductus arteriosus, ventricular septal defect, mitral stenosis and/or regurgitation, pulmonary stenosis, atrial septal defect, and endocardial fibroelastosis.[2-5]

Congenital aortic stenosis, with or without any additional cardiovascular malformations, is a life threatening pathology, causing sudden death in 6-19 % of the cases.[6] Patients with congenital aortic stenosis usually show symptoms and signs of cardiac failure and anginal chest pain. X-rays and ECG may display signs of left ventricular strain, or hypertrophy. Serial cardiac catheterizations proved that congenital aortic stenosis becomes worse with age and the valvular orifice gets relatively smaller in size in parallel to the growth of the individual.[7,8]

The accepted indications for surgery in congenital aortic stenosis are: The emergence of the symptoms, appearance of left ventricular strain in ECG, a valvular area of 0.5 cm^2 or less per m^2 of body surface, and/or a gradient of 50 mmHg or more across the left ventricular outflow tract.[3]

Emergency open heart surgery in infancy consists of two techniques: 1-Hypothermia and inflow occlusion,[10] 2-Extracorporeal circulation using heart-lung machine which is the technique used in the presented case.

Most of the patients do well and improve symptomatically and hemodynamically after valvular commissurotomy, and about 50 % of the patients show a gradient of 25 mmHg or slightly more across the aortic valve, or some regurgitation.[9] It is difficult to forecast for how long the benefit from this surgical intervention will last; it varies from patient to patient since most of these patients require an additional surgical intervention in the future as they grow up. This may involve the replacement of the valve or placement of an apical left ventricular-abdominal aortic conduit.[11,12] Valve replacement in these patients is not easy, owing to the small and narrow aortic annulus. In this connection, some operations to enlarge the aortic root and annulus before replacing the valve have been described.[13-15] Recently, left ventricular apical-aortic conduits containing tissue valves have been used in these cases, thus creating a double outlet left ventricle, and avoiding valve replacement.[11, 12]

Summary

The case of a 40 day-old baby girl with severe congenital valvular aortic stenosis who was subjected to open heart surgery using heart-lung machine is presented. The patient had two episodes of cardiac arrest during the preoperative period and was resuscitated. She did well during her postoperative course and was discharged in good condition. Clinical examination one year after the discharge was satisfactory. Congenital aortic stenosis and related problems are briefly discussed.

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