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BICUSPID AORTIC VALVE AND COARCTATION OF AORTA*

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Bicuspid aortic valve (BAV) and coarctation of aorta (COA) are frequently seen together. It is believed that these malformations result from a single developmental diathesis. A case is presented of COA, aortic stenosis and aneurysm of the ascending aorta corrected by patch aortoplasty and commisurotomy. An aneurysm at the site of the coarctation repair can develop as late as 20 to 25 years after surgery. Almost all of the aneurysms described have occurred in patients undergoing patch aortoplasty. We do not recommend this technique except in special cases. *Key words: bicuspid aortic valve, coarctation of aorta, aneurysm.*

Bicuspid aortic valve has long been known to be common in patients with coarctation. It was found in 23 percent of the cases studied by Abbott[1] and in 25 percent of the series of Benkwitz and Hunter[2]. Tawes and associates[3] note in their report that among 250 living children with long-term follow-up, 32 (13%) had clinical evidence of aortic valve disease (mainly stenosis but also incompetence).

McKusick[4] suggested that in patients with coarctation of aorta (COA) and bicuspid aortic valve (BAV) cystic medial necrosis occurs due to aortic wall weakness. Aneurysm, dissection and rupture may also develop in these patients due to surgical repair[5].

We present here a patient who developed an ascending aortic aneurysm 11 years after repair of aortic coarctation with patch aortoplasty.

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Case Report

A 1.5-year-old male patient was first referred to our hospital in 1977 with tachypnea and feeding problems. On physical examination his heart rate was 145/min and his blood pressure was 140/85 mmHg on the right and 130/85 mmHg on the left side. A III/VI systolic ejection murmur was heard along the left sternal edge. Lower extremity peripheral arterial pulses were absent. His chest x-ray showed a moderate cardiomegaly (Fig. 1). Echocardiography demonstrated aortic stenosis, aortic coarctation and minimal dilatation of the ascending aorta (Fig. 2). A presumptive diagnosis of COA and BAV was made. In 1986, surgical repair for COA was performed by dacron patch aortoplasty.

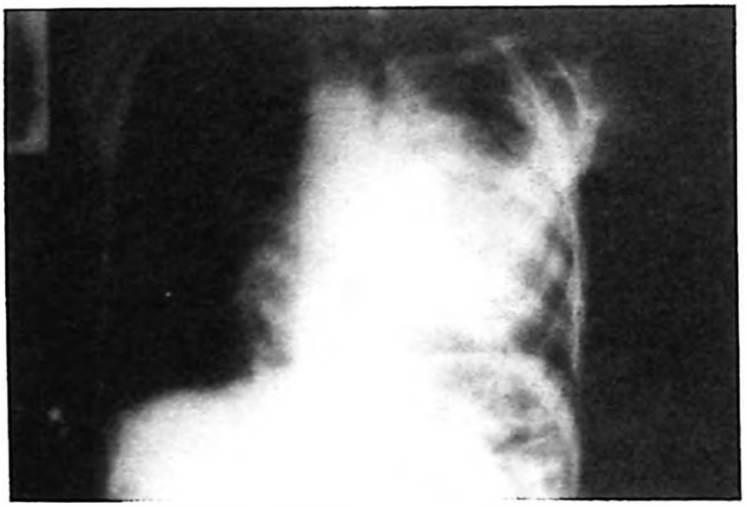

Fig. 1: Chest x-ray showed marked cardiomegaly.

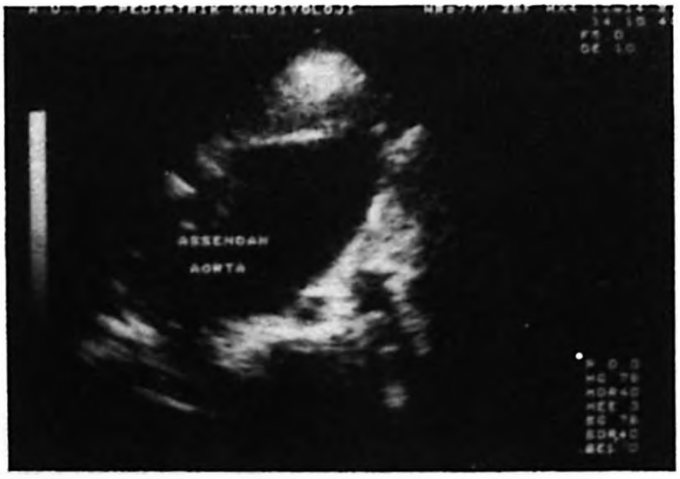

Fig. 2: Echocardiogram demonstrated aortic stenosis, aortic coarctation and minimal dilatation of the ascending aorta.

In 1988 cardiac catheterization and echocardiography were performed in order to confirm the ascending aortic dilatation and aortic stenosis (Fig. 3) The diameter of the ascending aorta (45 mm) was also observed to be greater than the diameter of the descending aorta.

With these manifestations, surgery was performed in 1988 when the patient was 12 years old. The heart was exposed through a median sternotomy and the patient was placed on total cardiopulmonary bypass via right femoral arterial and standard bicaval cannulation.

The aorta was found to be quite dilated. Intra-operatively a vertical incision was made through the aorta. The valve was in bicuspid morphology. Valvotomy was performed by dividing the commissures with a knife to within one mm of the aortic wall. The segment that revealed aneurysmatic dilatation was resected. The aorta was then closed primarily with 4/0 polypropylene suture (Fig. 4). The postoperative period was uneventful, and the patient was discharged two weeks later.

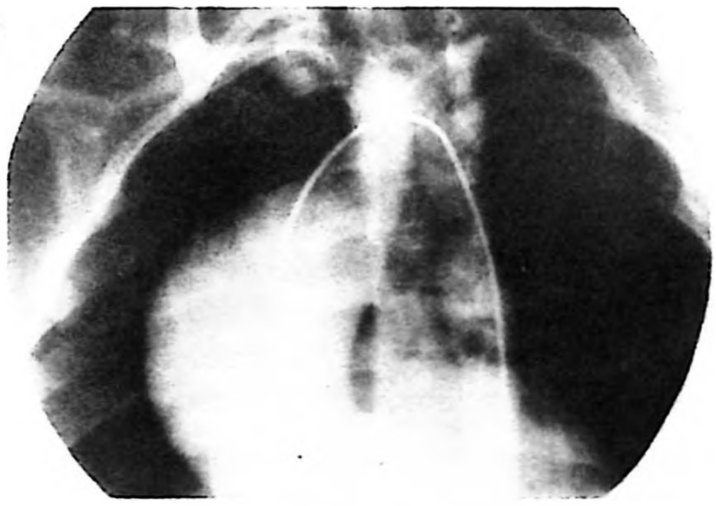

Fig. 3: Cardiac catheterization showed ascending aortic dilatation and aortic coarctation.

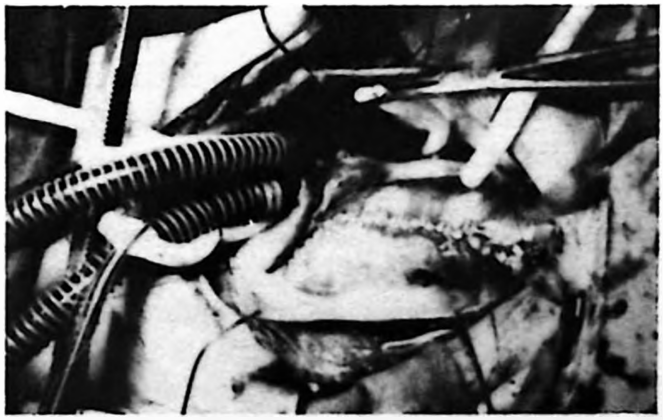

Fig 4: Aneurysmatic dilatation was resected and the aorta was closed primarily with 4/0 polypropylene suture.

Discussion

Valvar heart disease may complicate the long-term management of patients who have undergone coarctation repair, and occasionally prevents a good outcome[6]. In one study, surgery for aortic stenosis was required at three to seven years of age in seven percent of patients whose coarctation was repaired in infancy[7].

A significantly large number of patients with bicuspid valves will require surgery for calcific aortic stenosis in the fifth or sixth decade of life.

A true or false aneurysm may occur late postoperatively after repair of COA. Because the stiff patch transmits additional tension to the adjacent elastic aortic wall, a true aneurysm after a Dacron patch repair has always been associated with a false (suture line) aneurysm. In Barrat-Boynes' series[4] the incidence of aneurysm was 10 percent but increased to 25 percent in the eight patients followed 15 years or more. In the same report, aneurysm had not occurred in patients operated on before 10 years of age. Our patient was 11 years old at the time of his first operation. Dissection may occasionally occur in either the ascending or descending aorta, proximal or distal to the coarctation repair site, which may also lead to aneurysm formation.

We conclude that in cases of childhood aortic coarctation, if the patient is greater than 10 years old, patch aortoplasty can be performed instead of end-to-end anastomosis because of the low incidence of recurrent coarctation. However, because almost all aneurysms described have occurred in patients undergoing patch aortoplasty, we do not recommend this technique.

COA can be well repaired from an anterior midline approach through median sternotomy using cardiopulmonary bypass if the patient has a concomitant intracardiac defect. The advantage of this option is that reoperation may be avoided.

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