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Quinticuspid aortic valve causing aortic valve incompetence and stenosis

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Aortic valve dysfunction may be the consequence of congenital malformation of the aortic valve. Unicuspid,[1 2] bicuspid,[1–6] tricuspid,[2 3] and quadricuspid[6–8] aortic valves have all been described. We describe a patient who presented with cardiac enlargement caused by aortic valve incompetence and stenosis. Echocardiograms repeatedly showed multiple closing lines in the aortic valve orifice. This patient was found to have a quinticuspid aortic valve; the left coronary and non-coronary cusps were normal in size, and the remainder of the valve consisted of three small cusps divided by well-developed commissures.

Case report

The patient, a man born in 1958, was known to have had a cardiac murmur since early childhood. Cardiac catheterisation in 1971 had showed grade 2 aortic valve incompetence. At that time no stenosis was found and an abnormal number of aortic valve cusps could not be identified. There was minimal pulmonic valve stenosis with a pressure gradient of 10 mm Hg (1·3 kPa) and the patient was found to have a patent foramen ovale. Because of his youth and functional class I (NYHA) surgical treatment was postponed. Left heart catheterisation in 1978 showed grade 4 aortic valve incompetence and some aortic valve stenosis with a pressure gradient of 15 mm Hg (2·0 kPa). Again an abnormal number of aortic valve cusps could not be identified. The left ventricular end-diastolic pressure was normal, as was the left ventricular end-diastolic volume. The coronary arteries showed no abnormalities and the mitral valve was functioning properly. Functionally the patient remained in class I (NYHA) and surgery was still postponed.

At follow-up, chest radiographs showed increasing enlargement of the patient's heart. Electrocardiograms showed signs of increasing left ventricular hypertrophy, and echocardiograms confirmed enlargement of left atrium and left ventricle and also showed multiple closing lines in the aortic valve opening (fig 1). In retrospect this could be interpreted as being the result of closure of a valve with supernumerary cusps. In addition, enlargement of the aortic root was found and the mitral valve showed a high-frequency flutter.

In 1980 the patient was admitted for aortic valve replacement. Cardiac catheterisation was not repeated. On admission he had no complaints and appeared to be in good health. Physical examination showed signs of aortic valve incompetence with some aortic valve stenosis. No further abnormalities were found.

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At operation a dilatation of the aortic root was found. The aortic valve was quinticuspid. The left coronary cusp and non-coronary cusp were of normal size. Instead of one right coronary cusp there were three different cusps, divided by two well-developed commissures (fig 2). All the cusps showed signs of fibrotic thickening and retraction. There was no calcification or indication of acute or chronic inflammation or infection, either grossly or on microscopy. There was a small semilunar diaphragm over the ostium of the left coronary artery.

Discussion

Quinticuspid aortic valve seems to be a very rare cardiac malformation. To our knowledge only one other case has been reported, without any information on haemodynamic consequences.[6]

Our patient had normal-sized left coronary and non-coronary aortic valve cusps, each with normal commissures. The three small cusps divided by well-developed commissures that replaced the right coronary cusp did not reach the centre of the aortic valve opening and so caused aortic valve incompetence. We do not know whether this is the consequence of a developmental defect or a secondary abnormality. Although the fibrotic thickening and retraction were obvious, we cannot exclude the possibility that underdevelopment of the three cusps played a part in the aetiology of the aortic valve incompetence.

The origin of this quinticuspid aortic valve anomaly is not readily explained. Embryological observations in animals[9 10] and extrapolation of the explanations for quadricuspid aortic valves offer several possibilities.[6–8] Although the embryological development is a continuous process, three critical phases with regard to the supernumerary-cusped aortic valve can be recognised.[6] (1) Supernumerary prevalvular pads in the embryological truncus arteriosus evolve in supernumerary-cusped aortic or pulmonary valves after separation into aorta and pulmonary artery. (2) A normal number of prevalvular pads in the truncus arteriosus become excessively divided because of an abnormal dividing pattern of aorta and pulmonary artery, thereby producing a supernumerary-cusped aortic or pulmonary valve. (3) After normal separation of the aorta and pulmonary artery the prevalvular pads develop in an abnormal way to form a supernumerary-cusped valve.

Although only one cusp of the aortic valve seemed to be affected in our patient, the cardiac catheterisation of 1971 showed some pulmonary valve stenosis. Unfortunately we have no information about the morphology of the pulmonary valve. The anomalous right coronary cusps could possibly be related to an also anomalous right pulmonary cusp. Should the pulmonary valve ever turn out to be nor-

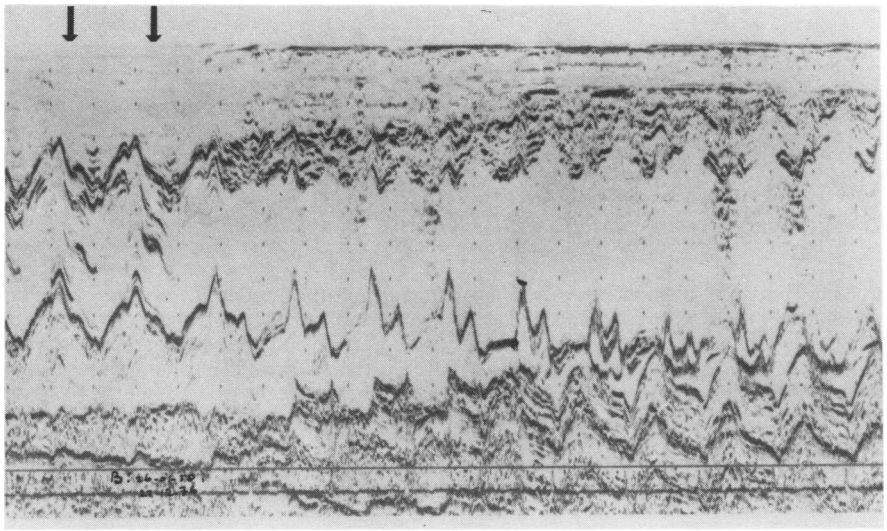

Fig 1 *Echocardiogram showing multiple closing lines in the aortic valve opening (arrows).*

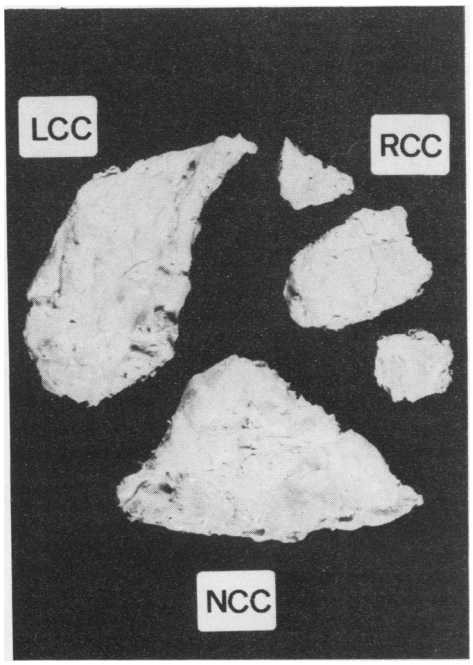

Fig 2 *Excised and rearranged cusps from quinticuspid aortic valve. LCC: left coronary cusp; NCC: non-coronary cusp; RCC: right coronary cusps.*

mal, the third explanation is the most likely one. Should the pulmonary valve be abnormal, however, the defect is likely to have happened earlier in the embryological development. In that case the first or second explanation will become more probable.

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