Combined Acute Rheumatic Fever and Congenitally Bicuspid Aortic Valve

A Hitherto Unconfirmed Combination*


An 18-year-old man is described in whom rheumatic heart disease, as evidenced by the presence of classic Aschoff bodies, occurred in combination with a congenitally bicuspid (purely incompetent) aortic valve. To our knowledge, this is the first report documenting the presence of rheumatic heart disease and congenitally bicuspid aortic valve in the same patient.

Although fusion of two of three aortic valve cusps (one of three commissures) causing “acquired” bicuspid valve is common in rheumatic heart disease, the occurrence of a congenitally bicuspid aortic valve in a patient with unequivocal rheumatic disease, i.e., with Aschoff bodies present, has not been reported. The report which follows describes this association in the same patient.

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CASE REPORT

An 18-year-old black man was well until two years before death when he developed flank pain for which he was hospitalized. Examination at that time disclosed blood pressure of 140/80 mm Hg, collapsing peripheral pulses, and grade 3/6 basal systolic and diastolic precordial murmurs consistent with aortic "stenosis" and regurgitation. Electrocardiogram (Fig 1) and chest roentgenogram (Fig 2) showed cardiomegaly. After several days, the flank pain subsided; its etiology was never determined. Infective endocarditis was ruled out by repeatedly negative blood cultures. After discharge, he remained well and active until he suddenly collapsed and died while at work.

At necropsy the heart weighed 800 gm. Both ventricular cavities were dilated and their walls hypertrophied (Fig 3). Neither atrial cavity was dilated. No foci of myocardial fibrosis or necrosis were noted grossly. The aortic valve was congenitally bicuspid. Each cusp was diffusely fibrotic and retracted but mobile, and devoid of calcium (Fig 3). The valve orifice was not stenotic, but it was incompetent. Both coronary arteries arose in front of the anterior cusp, which contained a raphe. The mitral leaflets were mildly but diffusely thickened by fibrous tissue, but the chordae tendineae were normal. The tricuspid and pulmonic valves were normal. The left ventricular endocardium was diffusely opaque, and the right ventricular endocardium was focally opaque. Histologic examination disclosed multiple typical Aschoff bodies in the endocardium of both atria and in the endocardium and interstitium of both ventricles (Fig 4).

COMMENTS

Although each is a fairly common condition, a congenitally bicuspid aortic valve and rheumatic heart disease have not previously been clearly demonstrated to be present simultaneously in the same patient. Previous studies1-3 from this laboratory have shown that congenitally bicuspid aortic valves may occur in as high as 2 percent of the population, and the incidence of rheumatic heart disease clinically has been reported to be as high as 6 percent of the population.4 Thus, it is surprising that the two conditions have not been described previously in the same patient. One of the reasons for this discrepancy may be different criteria utilized for defining the congenitally bicuspid condition of the aortic valve and for designating valvular disease as rheumatic in type. Our criteria for designating an aortic valve as congenitally bicuspid have been delineated elsewhere.5 In essence, there are only two aortic valve cusps, only two true commissures, and, in about half of the cases, a false commissure or raphe also is present. The cusps are oriented either anteriorly and posteriorly (and if a raphe is present it is always in the anterior cusp) or right and left (and if a raphe is present it is always in the right cusp). The distance circumferentially between any two true commissures is always greater than between a true and a false commissure. The raphe rarely extends as far cephalad in the aorta as do the true commissures.

The only unequivocal criterion of rheumatic heart disease is the presence of Aschoff bodies. Although a number of different stages of Aschoff bodies have been described,6 only in the granulomatus stage can an Aschoff body be unequivocally identified.7 In our labo-
ratory, we have found Aschoff bodies at necropsy or in biopsies of atrial appendages or papillary muscles obtained at operation only in patients with diffuse disease, nearly always stenosis, of the mitral valve. About 5 percent of our patients with fatal mitral valve disease studied at necropsy had Aschoff bodies. We have never observed an Aschoff body in the heart of a patient with anatomically isolated aortic valve disease, i.e., an anatomically normal mitral valve, and indeed we have been unable to find any report demonstrating an Aschoff body in the heart of a patient with isolated aortic valve disease, irrespective of the number of aortic valve cusps present. Among approximately 200 adult patients with congenitally bicuspid aortic valves studied by us at necropsy, only the patient described herein had an Aschoff body in his heart.

At least two previous authors, however, have mentioned the occurrence of Aschoff bodies in patients with bicuspid aortic valves. Gross described Aschoff bodies in 5 of 16 hearts with "so-called congenital bicuspid aortic valve." It is clear from study of his paper, however, that his criteria for Aschoff bodies were extremely loose, indeed unacceptable, and furthermore that several of his patients almost surely had acquired bicuspid aortic valves rather than congenital malformations. Hall and Ichioka found Aschoff bodies in five of eight patients with "bicuspid aortic valves." Three of these five patients were over 65 years of age and a fourth was 53. Thus, three of their patients may have had aortic valve disease of the elderly, probably degenerative in origin, rather than congenital malformations. These authors also used the loose, nonspecific criteria of Gross and Ehrlich for identification of Aschoff bodies. In addition, none of their five patients had anatomic disease of the mitral valve. To reemphasize, we have never observed an Aschoff body, using the criteria defined by Saphir, in a patient without anatomic disease of the mitral valve. Almost surely in years past there has been an "over-

![Figure 1. Electrocardiogram showing left ventricular hypertrophy and sinus rhythm.](image)

![Figure 2. Chest roentgenogram showing considerable cardiomegaly of the left ventricular type.](image)

![Figure 3. Heart. Upper: Opened aorta, aortic valve, and left ventricle. The left ventricular cavity is quite dilated, its endocardium mildly thickened, and its wall, very thick. Lower: Close-up view of the opened aortic valve. The anterior cusp, the one containing the raphe, has been severed during the opening of the valve. The posterior cusp, from which no coronary arteries arose, is intact. The anterior leaflet of the mitral valve (shown here) also is thickened by fibrous tissue. No inflammatory cells were found in histologic sections of any of the four cardiac valves.](image)
diagnosis" of Aschoff bodies at necropsy, using loose criteria which may include nonspecific inflammatory lesions of the heart.

Our patient had a classic congenitally bicuspid aortic valve and numerous classic Aschoff bodies in the heart at necropsy. To our knowledge, this combination has neither been clearly documented nor illustrated previously.

REFERENCES
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Persistence of the Third Heart Sound after Resection of the Native Mitral and Tricuspid Valves*

Evidence Against the Valvular Theory of Third Sound Origin

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A patient is described with severe rheumatic mitral and tricuspid insufficiency in whom both atrioventricular valves with their chordae tendineae and papillary muscles were resected and replaced with Hancock porcine grafts. This would appear to be the second such patient reported and the first described in detail in whom the third heart sound persisted postoperation. This occurrence documents the fact that a third heart sound can occur in the absence of native atrioventricular valve leaflets and the major portions of the subvalvular supporting apparatus, and argues against the theory that the third heart sound is generated by either the valvular leaflets or the subvalvular apparatus.

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