QUADRICUSPID AORTIC VALVE PERFORATION RESULTING FROM BACTERIAL ENDOCARDITIS
—2-D Echo- and Angiographic Diagnosis and its Surgical Treatment—

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A 75-year-old man with quadricuspid aortic valve regurgitation affected by bacterial endocarditis is reported. The aortic valve consisted of 4 equal-sized cusps (type a) and a supernumerary cusp located between the right and noncoronary cusps. A right coronary ostium was close to the accessory commissure, but there was no displacement. A few small fenestrations were found at the 4 commissures and a large perforation resulting from endocarditis was observed in the noncoronary cusp. 2-D echocardiogram and angiogram suggested these findings, and they were confirmed at surgery. Successful aortic valve replacement was achieved.

AN isolated 4-cusped (quadricuspid) aortic valve is an extremely rare congenital anomaly. Although this condition is considered to be well tolerated without marked clinical evidence, there is a tendency for it to become the focus of infective endocarditis! Spontaneous aortic regurgitation also occurs frequently in later life.2—3 The condition is usually diagnosed unexpectedly at autopsy or at aortic valve surgery, and rarely by aortography4—6 or by 2-dimensional (2-D) echocardiography?

We present the preoperative (2-D) echocardiographic and angiographic diagnosis of this rare anomaly affected by bacterial endocarditis and its successful surgical management.

CASE REPORT
A 75-year-old man was admitted to a hospital on August 22, 1986, after suffering for a week from a high fever of 39°C. Although no murmur, related to the aortic or mitral valve, was present on auscultation, routine echocardiographic examination revealed the vegetative aortic valve. On the fifth hospital day, congestive heart failure developed, and increased cardiomegaly with a cardiothoracic ratio of 56% and concomitant aortic regurgitant murmur appeared. At the same time, β-streptococcus was found in blood culture.

The patient, who was diagnosed as having acute aortic regurgitation resulting from bacterial endocarditis, was referred to the Yamanashi Medical College Hospital for his exact examination on September 2. The heart rate was 86 per minute and regular; blood pressure was 100/40 mmHg. Auscultation disclosed a grade 3/6 diastolic blowing murmur through the left third intercostal sternal border to apex and a grade 1/6 early systolic murmur at apex. Electrocardiogram showed sinus rhythm with left axis deviation, incomplete right bundle branch block and multifocal premature ventricular beats.
Plain X-ray demonstrated increased cardiomegaly, with a cardiothoracic ratio of 58%, and a small amount of bilateral pleural effusion. By simultaneous 2-D echocardiography, the 4 aortic cusps with perforation of the noncoronary cusp and verrucous changes in each cusp were strongly suggested in parasternal short axis projection (Fig. 1). The concomitant vegetation on the pulmonary valve was observed to consist of 3 cusps. Collor doppler echocardiogram demonstrated severe aortic regurgitation (grade III) simultaneously with mild mitral, tricuspid and pulmonary regurgitation. The left ventricular cavity was dilated, but its function was relatively good. Radioisotope pulmonary perfusion scanning suggested left regional pulmonary infarction. The patient was functionally categorized as NYHA III. Initial medical treatment, performed with digitalis, diuretics and antibiotics (penicillin G 800–1200 x 10⁴/day, I.V.) for 2 months, was successful. Thereafter, the active endocarditis was cured clinically.

Repeat echocardiographic study and initial cardiac catheterization were carried out to determine the surgical indications. Echocardiographic data on left ventricular function were as follows: left ventricular diastolic dimension (LVDd) 53 mm, left ventricular systolic dimension (LVDs) 32 mm, end-diastolic volume index (EDVI) 104 ml/m², endsystolic volume index (ESVI) 26 ml/m², stroke volume index (SVI) 80 ml/m²/beat, ejection fraction (EF) 77%, mean velocity of circumferential fiber shortening (mVcf) 1.2 unc/sec and mean systolic ejection ratio (MSER) 356 ml/sec. The vegetative changes in the aortic and pulmonary valves, disappeared, and pulmonary regurgitation were no longer observed echocardiographically.

Pressures in the aorta and left ventricle were 120/40 and 120/4 mmHg, respectively, end-diastolic pressure of the left ventricle (LVEDP) was 36 mmHg, which was markedly elevated. Pulmonary wedge pressure was 15/6 (mean 8) mmHg and pulmonary arterial pressure was 28/10 (mean 17) mmHg. No pressure gradients were observed across the 2 semilunar valves. Retrograde aortography demonstrated severe aortic regurgitation (grade IV) with mild mitral regurgitation, and the expected 4-cusped aortic valve was observed (Fig. 2). The angiographic left ventricular function study showed that end-diastolic and end-systolic volume indexes were

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in valve incompetency. The aortic valve was excised and an aortic valve replacement with a Carpentier-Edwards bioprosthesis (size 21 A) was achieved. His postoperative hemodynamic state was uneventful, but a complicating mediastinal infection was successfully treated with antibiotic administration and continuous mediastinal irrigation. The patient was discharged 82 days after operation and is doing well 1 year postoperatively.

DISCUSSION

A quadricuspid aortic valve is extremely rare. Hurwitz and Roberts\(^2\) studied the anatomic cusp variations found in quadricuspid semilunar valves and described 7 different types. Of the 7 quadricuspid aortic valves reviewed, 3 had 3 equal and 1 smaller cusp (type b); 2 had 4 equal cusps (type a); 1 had 3 equal and 1 larger cusp (type e), and 1 had 4 unequal cusps (type g). Type a has been reported to be equal in incidence to type b in surgically corrected cases.\(^8\)

This anomaly is important clinically because of the possible focus of infective endocarditis and the onset of spontaneous aortic regurgitation in later life. In both Hurwitz's\(^2\) and Davia's\(^3\) series, 8 (44%) of 18 quadricuspid aortic valves had functional abnormalities: 7 had aortic regurgitation and 1 had aortic stenosis without infective endocarditis. Few cases were examined by aortography and/or 2-D echocardiography before surgery or autopsy. Peretz et al\(^7\) first reported a case of quadricuspid aortic valve with severe aortic regurgitation diagnosed by aortography in 1969. Chandrasekaran et al\(^7\) reported 2 cases observed by 2-D echocardiography in 1984. They described that the diastolic 2-D echocardiographic appearance of the quadricuspid aortic valve has an “X” rather than the “Y” configuration of the normal tricuspid aortic valve, as shown in our case. We think it most useful to identify the quadricuspid aortic valve of Hurwitz's type a.

To our knowledge, 10 or more surgically corrected cases have been reported in the world literature. From the surgical viewpoint, it is important to recognize that displacement of the right coronary arterial ostium, which is in a lower position and close to the commissure between the right and supernumerary cusps, is not rare, as described by Lanzillo.\(^6\) In fact, it was rather difficult to fix the prosthetic valve ring around the supernumerary commissure without ostial

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135 ml/m\(^2\) and 56 ml/m\(^2\), respectively. Ejection fraction ratio was 59%. The cardiomegaly was improved to a cardiothoracic ratio of 50% preoperatively.

On November 19, 1986, the patient was treated surgically through cardiopulmonary bypass with moderate general hypothermia, blood cardioplegia and topical cardiac cooling. The aortic valve was inspected through an oblique aortotomy. The valve was confirmed to consist of 4 almost equal-sized cusps with a supernumerary cusp interposed between the right and noncoronary cusp (Fig. 3). A right coronary orifice existed close to the commissure, but was not displaced. A few small fenestrations at the 4 commissures, which were thought to be congenital, were visualized. The proliferated edges of the 4 cusps were also considered to have resulted
obstruction, which was close to the right coronary orifice, although it was not displaced downward in our case.

REFERENCES

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