대흉외지 2005;38:164-167

□ 증례보고 □

선천성 사엽성 대동맥판막

백만종* · 나찬영* · 오삼세* · 황성욱* · 이 철* · 김재현* · 서홍주*

Congenital Quadricuspid Aortic Valve

Man-Jong Baek, M.D., Ph.D.*, Chan-Young Na, M.D., Ph.D.*, Sam-Sae Oh, M.D.*
Seong Wook Whang, M.D.*, Cheul Lee, M.D.*, Jae Hyun Kim, M.D.*, Hong Ju Seo, M.D.*

Quadricuspid aortic valve is a rare congenital abnormality but it is well recognized as the cause of significant aortic regurgitation. We describe 5 patients who underwent surgery for severe aortic regurgitation associated with quadricuspid valve. In all patients, this abnormality had been incidentally detected during surgery. Two of the patients had infective endocarditis. In accordance with the Hurwitz and Roberts classification, two valves were type d, two were type a, and one was type c.

(Korean J Thorac Cardiovasc Surg 2005;38:164-167)

Key words: 1. Aortic valve, anomaly
2. Aortic valve
3. Aortic valve insufficiency
4. Endocarditis

CASE REPORT

Case 1

A 66-year-old male patient was referred with symptoms of congestive heart failure. The patient had been treated with antibiotics for endocarditis in another hospital. Echocardiography revealed severe aortic regurgitation and severe mitral regurgitation with multiple vegetations on both valves. After the antibiotics were added for 4 weeks, aortic and mitral valve replacement with On-X 21 mm and 27/29 mm, respectively, and tricuspid annuloplasty were performed. We initially tried aortic valve repair but failed. Operative findings showed that the aortic valve was quadricuspid, type *d* (one large, two intermediate, and one small cusp) of the Hurwitz and Roberts classification[1], and it had vegetations (Fig. 1). The mitral valve had a large perforation and multiple vegetations on the anterior leaflet. Ten months later, he is doing well without recurrence of endocarditis.

Case 2

A 15 year-old male was referred due to congestive heart failure. In another hospital, during the treatment for endocarditis, a sudden ventricular fibrillation developed due to an acute myocardial infarction but resuscitated. Echocardiography demonstrated severe aortic stenosis, grade IV regurgitation, and left ventricular aneurysm. Left ventricular ejection fraction (EF) measured by cine magnetic resonance imaging (MRI) was 29%. Coronary angiography and aortography

*부천세종병원 흉부외과
Department of Thoracic and Cardiovascular Surgery, Sejong General Hospital, Sejong Heart Institute, Bucheon, Korea
논문접수일 : 2004년 11월 8일, 심사통과일 : 2005년 1월 6일
책임저자 : 백만종 (422-711) 경기도 부천시 소사구 소사본 2동 91-121, 부천세종병원 흉부외과
(Tel) 032-340-1151, (Fax) 032-340-1236, E-mail: kubmj@chol.com
본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

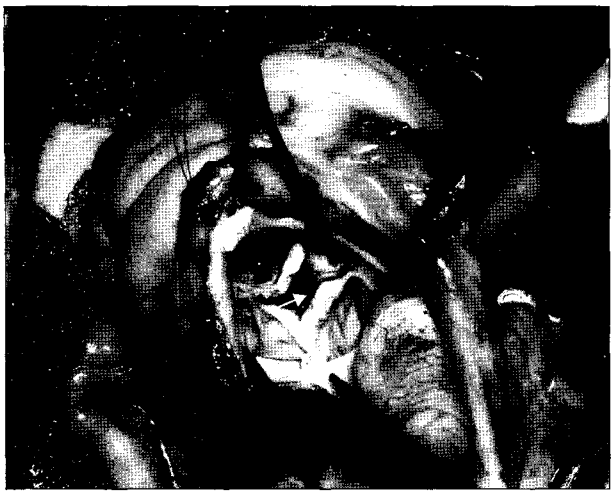

Fig. 1. Quadricuspid aortic valve with vegetations (white arrow).

showed total occlusion of the proximal left anterior descending artery (LAD) (Fig. 2) and fusiform dilatation of the ascending aorta with maximal diameter of 40 mm. Aortic valve replacement with St. Jude HP 19 mm, reduction aortoplasty for the ascending aortic dilatation, grafting of the left internal thoracic artery to the LAD, and endoventricular circular patch plasty were performed. Operative findings showed QAV with type *a* (four equal-sized cusps), multiple vegetations attached to the aortic valve. He was discharged at postoperative 30 days without complications. Follow-up MRI at postoperative 13 months showed that the left ventricular wall motion was much improved with EF of 54.1%. Sixteen months later, he is doing well without symptoms.

Case 3

A 57 year-old male was transferred due to congestive heart failure. Echocardiography revealed severe aortic regurgitation and stenosis. Also, he had fusiform dilatation of the ascending aorta with a diameter of 46 mm. Operative findings showed that the aortic valve was quadricuspid type *a*, and it had thickening and calcification of leaflet. Aortic valve replacement with St. Jude HP 23 mm and reduction aortoplasty with external wrapping were performed. Twenty-six months later, he is doing well.

Case 4

A 53 year-old female was referred due to symptoms of

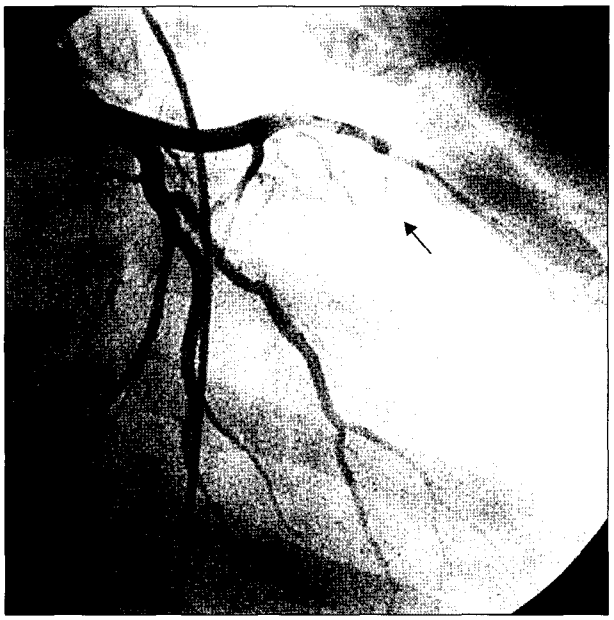

Fig. 2. Coronary angiography shows total occlusion of the proximal left anterior descending artery (black arrow).

congestive heart failure. Echocardiogram showed severe aortic regurgitation of grade IV. She had aneurysmal dilatation of the ascending aorta with a diameter of 38 mm. Aortic valve replacement with Omniscience 23 mm and external wrapping of the ascending aorta using a vascular graft were done. In the operative findings, the aortic valve was quadricuspid, type *c* (two equal larger and two equal smaller cusps) and prolapsed. Four years after operation, huge aneurysm involving the aortic root and dysfunction of prosthetic aortic valve developed. She underwent replacement of the aortic root using a composite valve graft. At three years after the reoperation, she is doing well.

Case 5

A 15 year-old female patient was referred with a diagnosis of small ventricular septal defect (VSD) and aortic regurgitation. During the follow-up, echocardiography demonstrated spontaneous closure of VSD and severe aortic regurgitation of grade IV. Ross operation with semilunar valve switch was performed after the reduction annuloplasty of the aortic valve. Aortic valve was quadricuspid type *d*. Follow-up echocardiography of postoperative 4 years showed minimal aortic regurgitation and mild pulmonic valve stenosis. Five

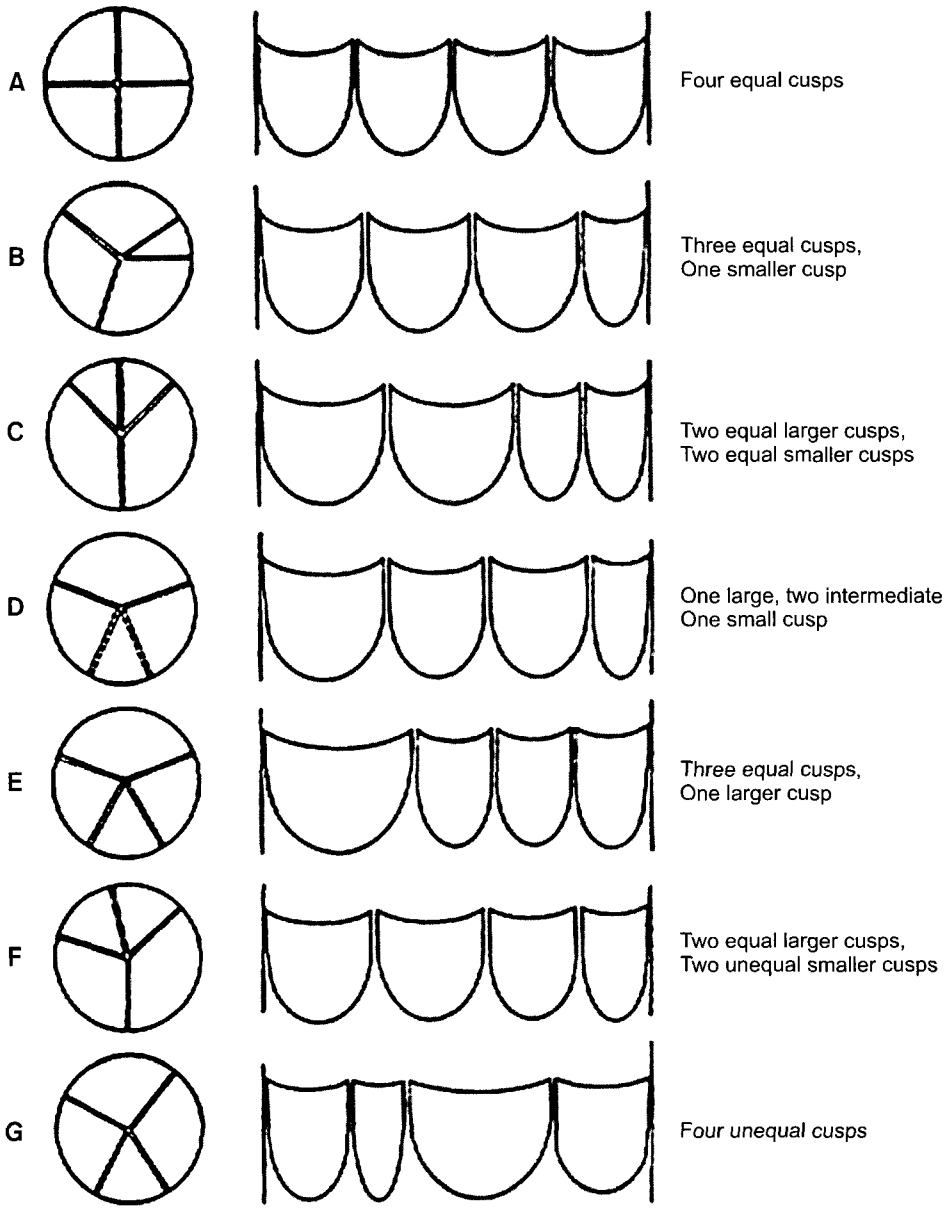

Fig. 3. Seven described anatomic variations of quadricuspid valves. The most common variation in either aortic or pulmonic valve consisted of 3 equal-sized cusps and 1 smaller cusp.

years later, she is doing well.

DISCUSSION

Quadricuspid aortic valve (QAV) is a rare congenital abnormality (Fig. 3) but it is well recognized as the cause of significant aortic regurgitation. The incidence of this abnormality has varied greatly between 0.008% and 0.013%, with more than 100 cases reported in the literatures[1-5]. The quadricuspid semilunar valves are reported to occur five times more frequently in pulmonic valve than aortic valve[6]. The quadricuspid aortic valve (QAV) is very rare, far more than unicuspid or bicuspid valve. These abnormalities have rarely been reported in autopsy and echocardiography studies, and surgical results[1-5]. Since 1990, we performed aortic valve replacements in 625 patients, and among them, 5 patients had QAV - an incidence of 0.8%.

The mechanism for development of QAV is not fully understood. Fernandez et al[7] suggested the hypothesis that in the Syrian hamster, QAVs result from the division of one

of the three mesenchymal anlagen that give rise to normal aortic valves. The division of the anlagen is due to the invagination of the endothelial layer that starts at a very early stage of the valvulogenesis, namely, when the conotruncal ridges begin to fuse at the distal portion of the embryonic cardiac outflow tract.

Most cases of QAV have been discovered as an incidental finding at necropsy, during surgery, or aortic angiography[4]. Recently, transthoracic or transesophageal echocardiography have been used frequently to detect this abnormality[3]. In all of our patients, QAV have been incidentally detected during aortic valve surgery.

Aortic regurgitation appears to be the most predominant valvular pathology associated with QAV. All of our patients had severe aortic regurgitation of grade IV. However, severe aortic stenosis with thickening and calcification of leaflet was also combined in two patients. In a review of the international literature, aortic regurgitation occurred in 39 of 70 (56%) cases, whereas valvular stenosis was very rare and only 18 cases had a normal functional status without significant regurgitation or stenosis[4]. Our experience and these results suggest that the most prevalent hemodynamic abnormality associated with QAV is valvular regurgitation but valvular stenosis or mixed valvular lesions can occur.

In general, it has been known that QAVs with unequal sized leaflets were likely to develop significant aortic regurgitation due to unequal distribution of stress and abnormal leaflet coaptation[3]. For these reasons, patients with unequally sized QAVs may require prophylaxis against subacute bacterial endocarditis. However, in our experience, this would not appear to be the case. Our two patients had infective endocarditis invading the QAV. One patient had equally sized QAVs, and the other had unequally sized QAVs. Although QAV with equal sized leaflets had no increased risk for endocarditis, we should recognize that infective endocarditis may occur in patients with QAVs, without respect to the leaflet size.

REFERENCES

1. Hurwitz LE, Roberts WC. *Quadricuspid semilunar valve.* Am J Cardiol 1973;31:623-6.
2. Simonds JP. *Congenital malformations of the aortic and pulmonary valves.* Am J Med Sci 1923;166:584-95.
3. Feldman BJ, Khanderia BK, Warnes CA, Seward JB, Taylor CL, Tajik AJ. *Incidence, description and functional assessment of isolated quadricuspid aortic valves.* Am J Cardiol 1990;65:937-8.
4. Janssens U, Klues HG, Hanrath P. *Congenital quadricuspid aortic valve anomaly associated with hypertrophic nonobstructive cardiomyopathy: a case report and review of the literature.* Heart 1998;78:83-7.
5. Timperley J, Milner R, Marshall AJ, Gilbert TJ. *Quadricuspid aortic valves.* Clin Cardiol 2002;25:548-52.
6. Davia JE, Fenoglio JJ, DeCastro CM, McAllister HA Jr, Cheitlin MD. *Quadricuspid semilunar valves.* Chest 1977; 72:186-9.
7. Fernandez B, Duran AC, Martire A, Lopez D, Sans-Coma V. *New embryological evidence for the formation of quadricuspid aortic valves in the Syrian hamster (Mesocricetus auratus).* J Comp Pathol 1999;121:89-94.

=국문 초록=

사엽성 대동맥판막은 매우 드문 선천적 기형의 하나로써 심한 대동맥판막폐쇄부전의 한 원인으로 알려져 있다. 저자들은 사엽성 대동맥판막과 동반된 심한 대동맥판막폐쇄부전으로 수술을 받은 5명의 환자를 보고한다. 모든 환자들에서 수술 중에 진단이 가능하였다. 환자들 중 2명에서는 감염성 심내막염이 동반되었다. Hurwitz and Roberts 분류법상 2명은 d형, 2명은 a형, 그리고 1명은 c형이었다.

중심 단어 : 1. 대동맥판막 기형
2. 대동맥판막
3. 대동맥판막 폐쇄부전
4. 심내막염