

## 선천성 사엽성 대동맥 판막 질환

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### Congenital Quadricuspid Aortic Valve Disease

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The quadricuspid aortic valve is a rare congenital cardiac morphology. In regard to the hemodynamics of the quadricuspid aortic valve, the regurgitation is most common, the regurgitation accompanying the stenosis or pure stenosis are rare. We report here a case with quadricuspid aortic valve disease which has been known to be extremely rare.

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**Key words:** 1. Congenital heart disease  
2. Aortic valve  
3. Aortic valve, anomaly

#### CASE REPORT

A 74-year-old male with history of hypertension, diabetes and coronary artery disease (CAD) was referred to our hospital. He had undergone percutaneous coronary intervention (PCI) 10 months prior to visit. He had been hospitalized for the last one for heart failure caused by the aortic stenosis and the coronary artery stenosis.

At the time of the transfer to our unit, blood pressure was 180/80 mmHg, heart rate was 94 beats per minute, respiration rate was 22 beats per minute. On physical examination, systolic murmur and the rales were heard. BUN/Cr was 23.1/1.7 mg/dL. Echocardiography revealed severe aortic stenosis with cusp calcification. Aortic valve opening area was 0.4 cm<sup>2</sup> and the ejection fraction was 54%. Coronary angiogram demonstrated the restenosis on the proximal LAD.

Coronary artery bypass grafting and aortic valve replace-

ment was done at 35th hospital day.

The aortic valve was severely calcified with dense commissural fusion and the distinction of each cusp was not clear. After the removal of the calcified valve, the commissure of each cusp was confirmed. The aortic valve had 3 equal-sized cusps and 1 smaller cusp (Fig. 1). In Hurwitz and Roberts classification, this aortic valve was classified as type *b*. One equal sized and another small cusps had no coronary orifice and both right and left coronary arteries originated from the juxta-commissural area of two equal-sized cusps. Two coronary arteries were located beside the commissure.

The aortic valve was replaced with the 21 mm sized mechanical valve (St. Jude Medical Inc, Minneapolis, Minn).

The patient was discharged 14 days after surgery without additional problems. The patient remained well over a 15-month follow-up period.

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본 논문의 저작권 및 전자매체의 지적소유권은 대한흉부외과학회에 있다.

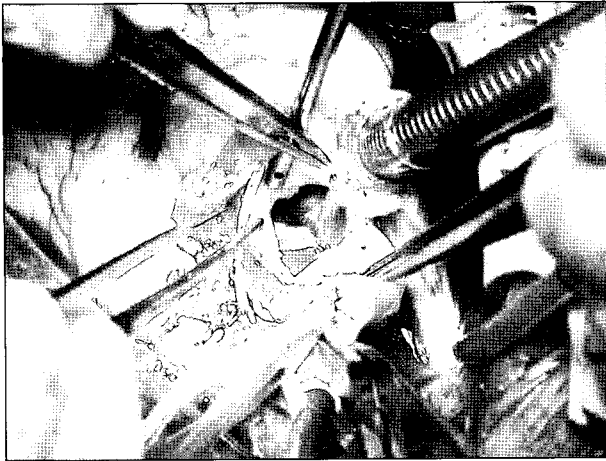


Fig. 1. Operative finding shows the quadricuspid morphology.

## DISCUSSION

Quadricuspid aortic valve is a congenital cardiac anomaly that is the rarest form of aortic valve abnormality. Simonds reported 2 cases out of their 25,666 cases of autopsy[1]. According to the literature survey by Tutarel, the mean age of patients was 50.7 years, the gender ratio of male and female was 1.6 : 1, and males were predominant citation.

It has been reported that the diagnostic method in the past was its discovery during surgery or autopsy in many cases, and recently, due to the improvement of echocardiography, the cases discovered prior to surgery are on rise[2], and it was discovered by aortogram in a small number of cases. In echocardiography, it has the characteristic that during the diastolic phase, the QAV disease shows as a X shape, and during the systolic phase, it shows as a quadrangular shape formed by 4 opened valves[3]. In regard to the hemodynamics of the QAV, it has been reported that the regurgitation was most frequent, 74.7%, the regurgitation accompanying the stenosis was 8.4%, the cases functioning as normal was 16.2%, and the case with the pure stenosis as our case has been reported to be very rare, 0.7% [4]. The regurgitation is frequent because examining the quadricuspid valve anatomically, the cases with three cusps of same size and the rest 1 cusp smaller in size were most frequent[5], and in such cases, due to the asymmetric distribution of stress and the abnormal coaptation of the cusps, it developed to the aortic regurgitation in many cases, hence, it has been reported that

in such cases, the prevention of subacute bacterial endocarditis was required[3]. In addition, the QAV accompanies other cardiac malformation such as hypertrophic cardiomyopathy, atrial septal defect, patent ductus arteriosus, ventricular septal defect, and mitral valve anomaly in many cases. And it has been reported that, particularly, the malformation of the coronary artery ostium or the coronary artery malformation was 10.2% of the entire cases[4].

The QAV disease requires the aortic valve replacement in many cases, and it requires more thorough examination prior to surgery for the detection of the presence of accompanying cardiac malformation, and particularly, since the single coronary ostium or the displacement of the origin of the left coronary artery or the right coronary artery is frequent, it is important to prevent ostial obstruction of the coronary artery when fixing the prosthetic valve ring[6].

In our case, similarly, the origin of the right and left coronary artery was higher than normal and lean toward to commissure, hence, while implanting a valve, a careful attention was paid not to obstruct the coronary ostium.

As its treatment, together with the correction of accompanying malformation, the aortic valve replacement is performed, however, in rare cases, the aortic valve repair may be performed[2].

As we experienced the congenital QAV disease that has been shown to be extremely rare.

## REFERENCES

1. Simonds JP. *Congenital malformations of the aortic and pulmonary valves*. Am J Med Sci 1923;166:584-95.
2. Kozo N, Hitoshi O, Junji Y, Kazuhiro H, Hisashi S, Yasushi N. *Aortic valve repair for quadricuspid aortic valve associated with aortic regurgitation and ascending aneurysm*. J Thorac Cardiovasc Surg 2004;128:759-60.
3. Barry JF, Bijoy KK, Carole AW, James BS, Catherine LT, Tajik AJ. *Incidence, description and functional assessment of isolated quadricuspid aortic valves*. Am J Cardiol 1990; 65:9378.
4. Oktay T. *The quadricuspid aortic valve: a comprehensive review*. J Heart Valve Dis 2004;13:534-7.
5. Hurwitz LE, Roberts WC. *Quadricuspid semilunar valve*. Am J Cardiol 1973;31:623-6.
6. Oktay T. *Quadricuspid aortic valves and anomalies of the coronary arteries*. J Thorac Cardiovasc Surg 2004;127:897.

=국문 초록=

사업성 대동맥 판막 질환은 매우 드문 선천적 심기형이다. 혈역학적으로 사업성 대동맥 판막 질환은 대부분 판막폐쇄부전을 일으키며, 드물게는 협착과 폐쇄부전을 동반하거나 협착 소견만을 보이기도 한다. 저자들은 매우 드문 것으로 알려진 사업성 대동맥 판막 질환을 경험하여 보고하는 바이다.

- 중심 단어 : 1. 선천성 심기형  
2. 대동맥 판막  
3. 대동맥 판막 기형