A 17-year-old male patient was referred with symptoms of dyspnea. Multi-detector computerized tomography (MDCT) and echocardiography evaluation revealed quadricuspid aortic and pulmonary valves, an atrial septal defect (ASD), and pulmonary stenosis. We closed the ASD using a bovine patch and performed a commissurotomy of the pulmonary valve. Quadricuspid semilunar valves are very rare congenital abnormalities that are reported to occur nine times more frequently in the pulmonic valve than in the aortic valve. According to the Hurwitz and Roberts classification, the aortic valve was type A, and the pulmonic valve was type B. The aortic valve had normal function, but the pulmonic valve was stenotic and had abnormal function.

On physical examination, systolic ejection murmur was auscultated on the left upper parasternal border, and there were no specific findings except right ventricle hypertrophy on electrocardiography. Mild hypoxemia was detected by arterial blood gas analysis; pH 7.423, pCO₂ 44.0 mmHg, pO₂ 70.5 mmHg, and sPO₂ 94.4%. Right atrial and ventricular hypertrophy, atrial septal defect, and pulmonary stenosis were detected by echocardiography and maximal pressure gradient between right ventricle and main pulmonary artery estimated by Doppler echocardiography was 64 mmHg. Pulmonary valvular motion was restricted and the pressure gradient between right ventricle and main pulmonary artery was 60 mmHg by right atrial angiography. After diagnosing ASD and pulmo-
nary stenosis, operation under routine cardiopulmonary bypass was performed.

After cardiopulmonary bypass, incisions to the pulmonary arteries were made. Quadricuspid pulmonary valve was confirmed under direct surgeon’s visual field. Three parts of four commissures were fused. When performing commissurotomy on all three fused commissures, pulmonary regurgitation can occur. Therefore, commissurotomy was done on the left two commissures with 3 mm incision on each valve (Fig. 2). Afterwards right atriotomy and ASD closure to the verified 12 mm-size ASD was done using the bovine pericardial patch. The patient was discharged from the hospital on the seventh postoperative day without any complications. The aortic valve and pulmonary valve shows no abnormality on echocardiography at 11 months follow up after discharge.

**DISCUSSION**

Quadricuspid semilunar valves are very rare congenital malformations. To our knowledge, this is the first reported case of concomitant quadricuspid aortic and pulmonary valves. This abnormality appears to be due to abnormal process of truncus arteriosus dividing into aorta and pulmonary trunk during embryological development of the heart. Incidence of Quadricuspid pulmonary valve is 0.02~0.12% and that of quadricuspid aortic valve is 0.008~0.043%, the former occurs about 9 times more often[1-5].

Developmental mechanism of quadricuspid valves had not been understood exactly, but Fernandez et al. explained the occurrence of quadricuspid valves by a hypothesis using embryo of a Syrian hamster. Normal aortic valve originates from three mesodermal primitive cell differentiation and one of these three mesodermal cells is detached by invagination of endothelial cell at valve development period, so this process results in the formation of quadricuspid valves[4].

Quadricuspid semilunar valves were classified into 7 types by Hurwitz and Roberts in 1973. Among the 121 cases, the majority of quadricuspid pulmonary valve was type B in 72 cases (60%)[2]. In this case, 4 cusps of aortic valve were same-sized, so called type A, pulmonary valve showed 3
same-sized commissures and the remained one cusp was small-sized, so called type B.

Most quadricuspid pulmonary valve functions normally, but conversely the quadricuspid aortic valve functions abnormally. The functional disorder of quadricuspid pulmonary valves shows regurgitation in most cases, but stenosis of the valves is rare[2,5].

Aortic valve and pulmonary valves were concomitant quadricuspid valves in our case and this is extremely rare. In contrast to previously reported properties of quadricuspid valves, the function of aortic valve was normal and pulmonary valves showed stenosis. Because the four cusps of the aortic valve were the same size, classified as type A, the aortic valve seems to function normally which is probably due to regular distribution of pressure[3,5].

In operation techniques to correct the abnormality, commissurotomy and valve replacement can be done. Commissurotomy of pulmonary valve was done after considering the patient’s age and the possibility of re-operation. Post-operative echocardiography showed relieved pulmonary stenosis and no pulmonary regurgitation. Because the patient is still young and quadricuspid semilunar valves are unstable, periodical echocardiographical examination and regular follow-up to check other functional abnormalities in aortic and pulmonary valves are needed.

REFERENCES