Quadricuspid aortic valve (QAV) is a rare form of congenital malformation. It was first described by Balington by autopsy study in 1862. Aortic regurgitation is the predominant valvular dysfunction associated with QAV, whereas valvular stenosis is very rare. QAV may also be diagnosed as a lonely lesion without other malformation, but QAV could be associated with patent ductus arteriosus, anomalies of coronary arteries, ventricular septal defect and pulmonary artery stenosis, subaortic fibromuscular stenosis, and hypoplasia of anterior mitral leaflet.1,2)

A 56-year old male patient was admitted to our institute complaining of dyspnea. He had a history of hypertension,
diabetes mellitus, diabetic retinopathy and diabetic nephropathy. He was a chronic alcohol drinker. His vital sign was stable but initial chest X-ray showed cardiomegaly, pulmonary congestion and blunting at both costophrenic angles. Initial laboratory findings showed increased blood urea nitrogen (97 mg/dL) and creatinine level (10.6 mg/dL). With the diagnosis of aggravated chronic renal failure, he underwent emergency hemodialysis. After dialysis, he was consulted to cardiology for evaluating heart failure. Transthoracic echocardiography showed left atrial enlargement and aortic regurgitation grade II/IV at parasternal long axis view. At parasternal short axis view, aorta showed 4 cusps. Left ventricular enlargement (end-diastolic and end-systolic diameters, respectively, of 6.8 and 5.3 cm) and systolic dysfunction (ejection fraction of 44%) were also observed. Transesophageal echocardiography was performed for a detailed examination of the aortic valve and root. The examination showed a QAV (4 sinus of valsalva and 4 aortic cusp: four cusps were of equal size) with a normal systolic opening of all 4 leaflets and the presence of incomplete central leaflet coaptation (Fig. 1A and B). Color Doppler examination confirmed the existence of a central aortic regurgitation (Fig. 1C and D). No other cardiac abnormalities associated with QAV were found.

References