Double-orifice Mitral Valve in an Elderly Patient with Tetralogy of Fallot

Taishi SASAOKA, MD, Haruhiko OHHURI, MD, Yoshiyuki MAKITA, MD, Shingo KUROKAWA, MD, and Tohru IZUMI, MD

SUMMARY

The patient was a 67-year-old man receiving medical treatment for tetralogy of Fallot at our hospital. A routine follow-up echocardiogram revealed a double orifice mitral valve (DOMV) associated with his tetralogy of Fallot. DOMV is often associated with endocardial cushion defect, however, DOMV associated with tetralogy of Fallot is extremely rare in elderly patients. (Jpn Heart J 1996; 37: 503–507)

Key words: Double-orifice mitral valve Tetralogy of Fallot Echocardiogram

A double-orifice mitral valve is occasionally seen in patients with the endocardial cushion defect. However, the combination of two congenital anomalies, DOMV and tetralogy of Fallot, has not been described previously in elderly patients. A routine echocardiographic study of a 67-year-old Japanese man who had received medical intervention for tetralogy of Fallot at our hospital since 1988 revealed a DOMV.

CASE REPORT

The patient had been diagnosed with congenital heart disease, probably an atrial septal defect, at age 37 years. He exhibited moderate cardiomegaly on chest X-ray (cardio-thoracic ratio 79%) and electrocardiographic findings of complete right bundle-branch block. Treatment over the next 22 years was limited to the control of hypertension.

Seven years ago, the patient was admitted to hospital with refractory congestive heart failure. The diagnosis of tetralogy of Fallot was established during this admission using echocardiography. A high-grade heart block was also noted.
A type of VVI permanent pacemaker was implanted through the endocardial leads (Intermedics, No. 254–20, VVI, USA) to prevent recurrent heart failure.

Following his discharge from the hospital, the patient's cardiac status was regularly checked by echocardiography. Surprisingly, one echocardiogram revealed the presence of the combined anomaly of DOMV and tetralogy of Fallot. The DOMV completely bridged two orifices of the mitral valve that were equal

**Figure 1.** Echocardiographic short-axis view, showing double orifice of mitral valve, right ventricular (RV) hypertrophy, and pericardial effusion (PE). ALO = anterolateral orifice; PMO = Posteromedial orifice.

**Figure 2.** Apical two-chamber view, showing mitral regurgitation (MR) of the anterolateral orifice (ALO) and posteromedial orifice (PMO).
in size (Figure 1). The valve area was calculated as 1.0 cm\(^2\) in the anterolateral orifice and 1.1 cm\(^2\) in the posteromedial orifice. In both mitral orifices, the stream of backward flow was moderate (Figure 2). A small pericardial effusion was recognized (Figure 1). In addition, the typical findings of tetralogy of Fallot were confirmed, namely, stenosis of the pulmonary valve, a membranous type of ventricular septal defect (Figure 3), right ventricular hypertrophy (Figure 1), and overriding of the aorta (Figure 4).
Figure 5. Electrocardiography before pace-maker implantation, showing right axis deviation, ectopic atrial rhythm, complete right bundle-branch block and a first-degree atrioventricular block.

Figure 6. Chest X-ray, showing severe cardiomegaly but neither pleural effusion nor pulmonary congestion.

On physical examination, a systolic murmur was audible over the apex of the heart. Jugular vein dilatation and hepatomegaly were negligible. Hypocarbia (pCO₂ 29.6 mmHg, pO₂ 76.0 mmHg), liver dysfunction (glutamic oxaloacetic transaminase 33 IU/l, glutamic pyruvic transaminase 17 IU/l, lactic dehydrogenase 451 IU/l, alkaline phosphatase 622 IU/l), and renal disorder (creatinine 2.1 mg/dl, blood urea nitrogen 34 mg/dl) were noticed. Electrocardiography re-
revealed right axis deviation (100° in the mean QRS frontal vector), ectopic atrial rhythm, complete right bundle-branch block, and a high-degree atrioventricular block (Figure 5). Chest X-ray showed severe cardiomegaly (cardiothoracic ratio 82.5%) but neither pleural effusion nor pulmonary congestion (Figure 6).

**DISCUSSION**

According to Bano-Rodrigo et al., the appearance of equal-sized orifices is limited to 15% of patients with the DOMV anomaly. Other cases have shown unequal orifices in the two valves. In 48% of patients with double orifices, the mitral valves function normally, while mitral stenosis is documented in 26% and regurgitation is also seen in 26%. The framework that composes the DOMV can be divided into three forms: a complete bridge between the two valves, an incomplete bridge, and a hole formed by the congenitally abnormal valves. A complete bridge is the structure that is least commonly seen.

In the report of Bano-Rodrigo et al., tetralogy of Fallot was recorded in only 4 of the 27 cases, combined anomaly of DOMV. The patients’ ages and other clinical data were not given, but considering the natural course of tetralogy of Fallot, the cases probably involved infants or children. We have been unable to find any articles referring to elderly patients with tetralogy of Fallot combined with DOMV. Our patient appears to be the first such case.

The striking finding of DOMV was observed with high-resolution echocardiography (Hewlett Packard, SONOS1500, 3.5 Hz Probe, Andover, MASS, USA). The structural disorders seemed to provoke moderate-grade mitral regurgitation. Recent advances in echocardiographic resolution power enable us to recognize this structural disorder on a routine examination.

**REFERENCES**