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Absent pansystolic murmur in Eisenmenger ventricular septal defect

Kazuhito Hirata *, Tomohiro Arakaki, Masahiro Tamashiro, and Masanori Kakazu

Division of Cardiology, Yuuai Medical Center, 50-5 Yone, Tomigusuku city, Okinawa 901-0224, Japan

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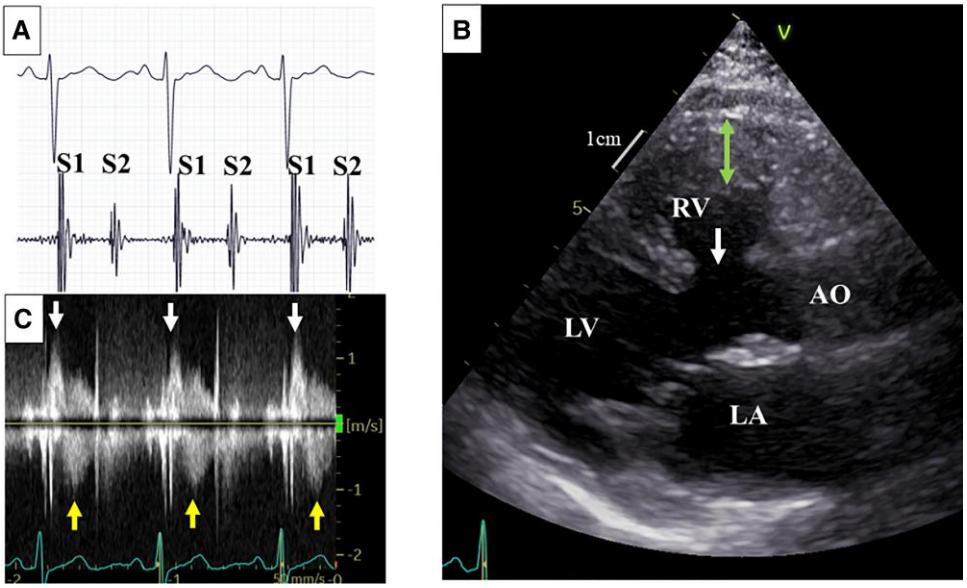

Figure 1 (*A*) A phonocardiogram recorded at the 4th left interspace shows that the pansystolic murmur of the VSD was absent. S1, the first heart sound; S2, the second heart sound. (*B*) Parasternal long-axis view of the transthoracic echocardiogram. A white arrow indicates the defect. A green arrow indicates hypertrophic right ventricular wall. RV, right ventricle; LV, left ventricle; AO, ascending aorta; LA, left atrium. (*C*): Pulse Doppler recording at the defect showing early systolic left-to-right shunt (white arrows) and mid-systolic right-to-left dominant bidirectional flow (yellow arrows) with the flow velocity of 1.0–1.4 m/s, indicating no pressure gradient.

Case description

A woman in her late 50 s presented to our hospital with NYHA Class 3–4 dyspnoea for several months.

Her history was remarkable for a large ventricular septal defect (VSD) with pulmonary hypertension, diagnosed at the age of 8 years. At that time, repair surgery was deferred due to high operative risks. The patient had been stable until 2 years prior to this admission, when she started to have NYHA Class 2 dyspnoea.

On admission, blood pressure, heart rate, and respiratory rate were 95/70 mmHg, 95/min, and 20/min, respectively. Acrocyanosis and clubbed fingers were noted. Notably, no typical pansystolic murmur

* Corresponding author. Tel: +81 98 850 3811, Fax: +81 98 850 3810, Email: kheart911@yahoo.co.jp
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was observed (*Figure 1A*). The second heart sound was single without splitting. Arterial blood gas showed a partial oxygen tension of 44 Torr, and oxygen saturation was reduced at 71%. Echocardiography revealed a large, non-restrictive peri-membranous VSD with marked right ventricular (RV) hypertrophy (*Figure 1B*). No additional congenital defects were noted. An estimated pulmonary artery systolic pressure was 80 mmHg. Pulse-wave Doppler revealed a bidirectional shunt across the defect (*Figure 1C*). Furthermore, the low flow velocity (1–1.4 m/s) was consistent with the absence of a pressure gradient across the VSD. Hence, the patient was diagnosed with Eisenmenger syndrome.

This case made us contemplate the pathophysiology of the generation of cardiac murmurs. The murmur of the VSD is generated by left-to-right shunt flow resulting from the pressure gradient between the left ventricle (LV) and RV across the defect. A high-pitch pansystolic murmur at the lower sternal border is a hallmark of VSD. In Eisenmenger syndrome, the RV pressure is elevated and is almost equal to that of the LV and the VSD murmur is not generated under these haemodynamic conditions.[1] If the systolic murmur is heard in Eisenmenger syndrome, it usually originates from systolic ejection murmurs of semilunar valves or tricuspid regurgitation murmur.[1]

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Authorship

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