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Title
Double-Outlet Right Ventricle With Intact Ventricular Septum and Left Atrioventricular Valve Regurgitation in a Patient With Right Atrial Isomerism

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Declaration of conflicting interests

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Fetal echocardiography revealed HLHS, aortic atresia, mitral regurgitation, and a small ventricular septal defect (VSD). A baby boy (weight, 2620 g) was delivered by Cesarean section at gestational week 37. Postnatal echocardiography and multidetector-row computed tomography confirmed RAI, DORV, incomplete atrioventricular septal defect, single atrium, valvular pulmonary stenosis, patent ductus arteriosus (PDA), ductal-associated pulmonary artery coarctation, right aortic arch, bilateral superior vena cava (SVC) and separate hepatic venous drainage. No vessel

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The PDA was maintained by continuous lipo-prostaglandin E1 infusion. LAVVR gradually decreased from mild-to-moderate to mild with body weight gain (Fig. 3A). At 3 months of age (weight, 4.96 kg), the patient underwent systemic-to-pulmonary shunt, pulmonary angioplasty with total resection of PDA tissue, direct end-to-end anastomosis using his pulmonary arterial wall and division of main pulmonary artery. At 7 month of age, cardiac catheterization showed that the systolic pressure of RV and LV was 93 and 140 mmHg, respectively. The ejection fraction of RV and LV was 54.5% and 20.3%, respectively. The end-diastolic volume of RV and LV was 223.0% of normal and 39.0% of normal, respectively.

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HLHS with aortic atresia and mitral stenosis is associated with "blind-ended" LV. Pulmonary atresia with IVS is also associated with "blind-ended" RV. The pressure in such a "blind-ended" ventricle sometimes exceeds systemic pressure. Consequently, it sometimes compresses the other ventricle [1, 2] and is associated with ventriculo-coronary connection. However, other than these two anatomies, a "blind-ended" ventricle is extremely rare. To the best of our knowledge, DORV with IVS in a patient with RAI has not been reported previously.

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References

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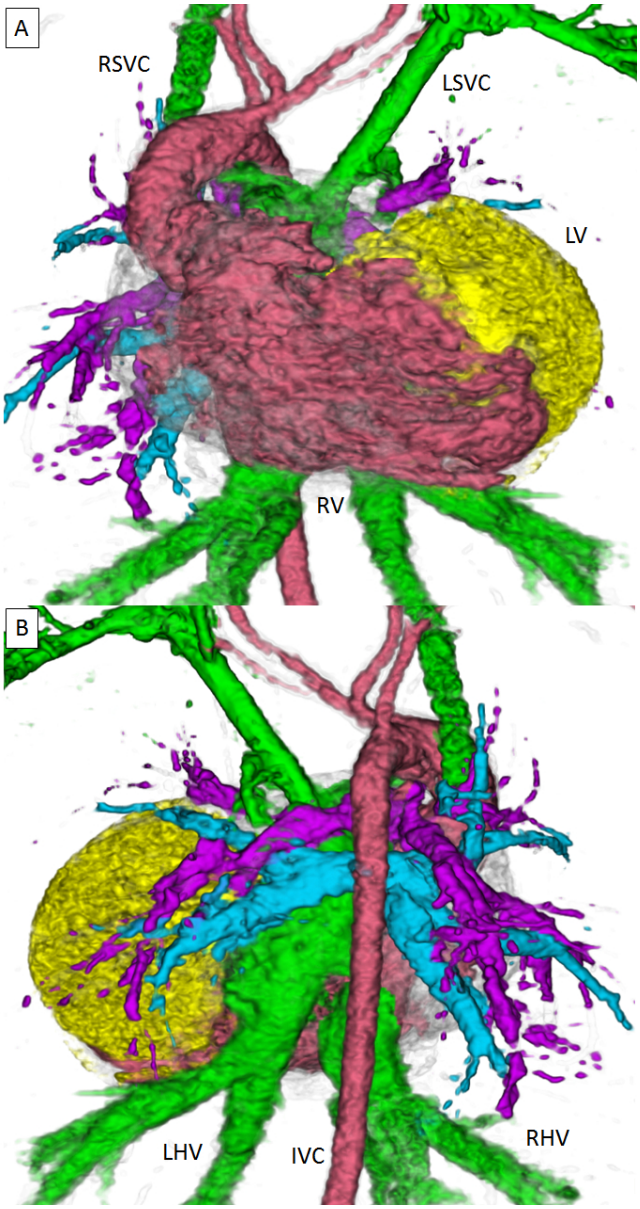
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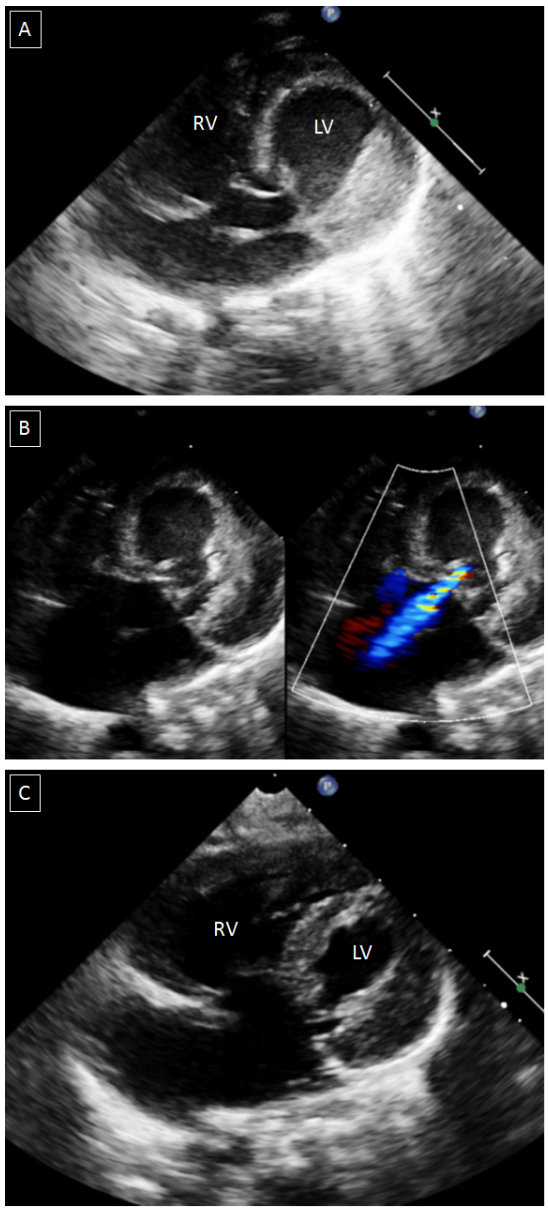
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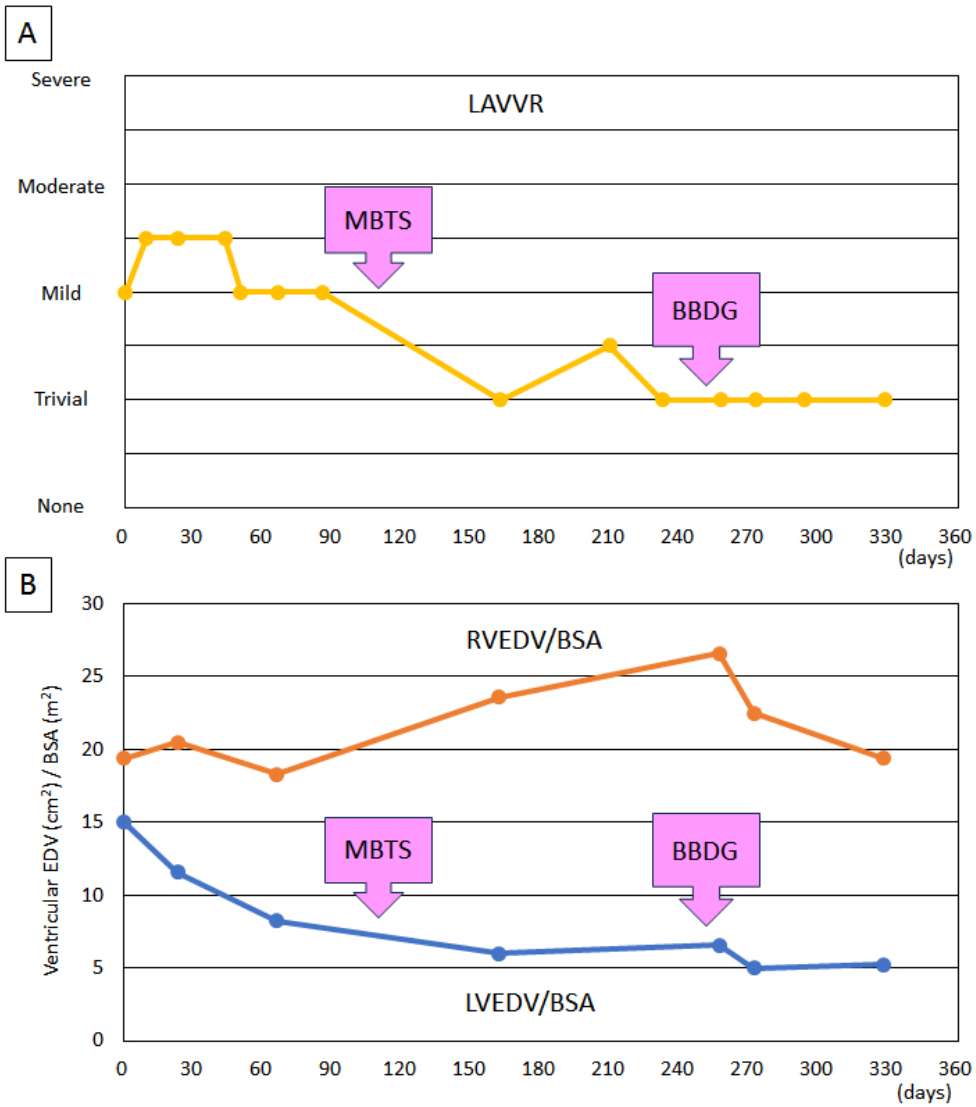
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239x450mm (72 x 72 DPI)



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254x283mm (72 x 72 DPI)