

# SPONTANEOUSLY HEALED MEMBRANOUS TYPE VENTRICULAR SEPTAL DEFECT WITH MALALIGNED INTERVENTRICULAR SEPTAL WALL AND DOUBLE-CHAMBERED RIGHT VENTRICLE IN A 56-YEAR-OLD PATIENT

JUNG SUN CHO, MD, PHD, HO-JOONG YOUN, MD, PHD, SUNG-HO HER, MD, PHD,  
SOE HEE AHN, MD, MAHN-WON PARK, MD, MIN SUK CHOI, MD, JAE BUM LEE, MD,  
JEONG U BAEG, MD, CHAN SEOK PARK, MD, PHD AND MI-JEONG KIM, MD, PHD

DEPARTMENT OF INTERNAL MEDICINE, SCHOOL OF MEDICINE, THE CATHOLIC UNIVERSITY OF KOREA, SEOUL, KOREA

A 56-year-old male presented with resting dyspnea and chest discomfort for several years. During transthoracic and transesophageal echocardiography, a spontaneously healed membranous type ventricular septal defect (VSD) with malaligned interventricular septal wall, aneurysmal changes, a subaortic ridge and a double-chambered right ventricle (DCRV) was observed. When combined with DCRV, VSD with malalignment between the outlet and trabecular septa was associated with tetralogy of Fallot. The subaortic ridge was due to turbulent flow caused by the malalignment-type VSD. The VSD with malaligned interventricular septal wall can be developed after aneurysmal changes of a perimembranous VSD. We report here in the unusual case of a 56-year-old patient who had a pathology complex comprising DCRV, subaortic ridge, spontaneously healed membranous type VSD with malaligned interventricular septal wall, and survived with surgical treatment.

**KEY WORDS:** Membranous type ventricular septal defect · Double-chambered right ventricle · Subaortic ridge · Malaligned septal wall.

## INTRODUCTION

Double-chambered right ventricle (DCRV) is a cardiac disease of the right ventricular outflow tract, where anomalous muscle bundles or a fibrous band cause obstruction.<sup>1-3)</sup>

A malaligned interventricular septal wall can develop in patients with aneurysmal changes of ventricular septal defect (VSD), the degree of which usually increases with time.<sup>4)</sup> Furthermore, when combined with DCRV, VSD with malalignment between the outlet and trabecular septa was associated with tetralogy of Fallot. A subaortic ridge in the malalignment-type VSD is caused by turbulent flow.<sup>5-7)</sup> We describe here in the unusual case of a 56-year-old man who had a spontaneously healed membranous type VSD with malaligned interventricular septal wall, DCRV, subaortic ridge, and survived with surgical treatment.

## CASE

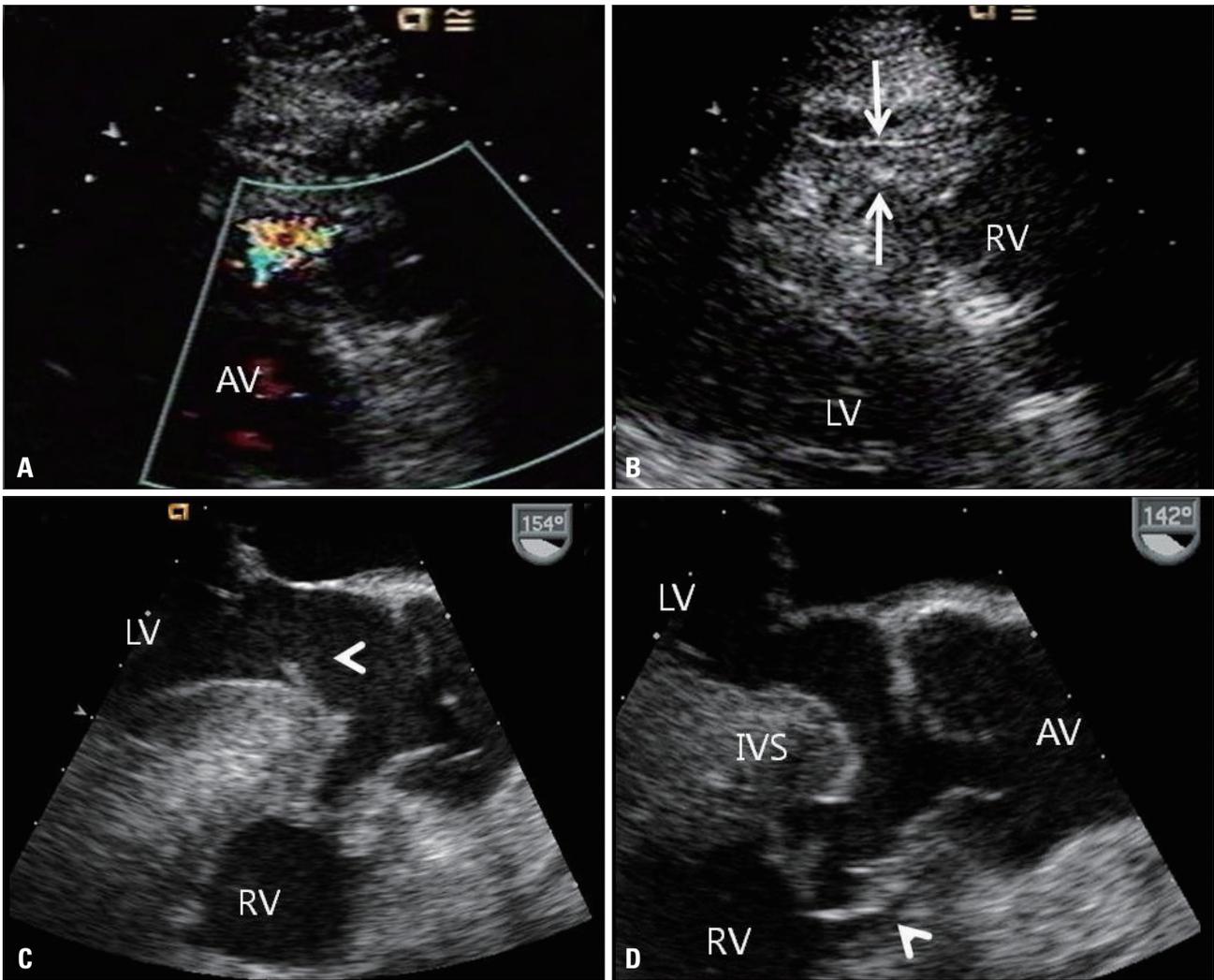
A 56-year-old male presented with resting dyspnea and chest discomfort for several years. He had no history of any surgical or catheter-based cardiovascular interventions. Physical examination showed a cardiac systolic murmur (grade 4/6) on the left upper sternal border, a systemic blood pressure of 140/75 mmHg, and a heart rate of 110 bpm. Electrocardiography showed sinus tachycardia, right axis deviation and frequent premature ventricular contraction with right ventricular bundle branch block morphology.

Transthoracic and transesophageal echocardiography revealed a membranous type VSD with anteriorly malaligned interventricular septum, aneurysmal changes, subaortic ridge and DCRV with right ventricular hypertrophy (Fig. 1). Measurement of pressure gradients using a Doppler image was not

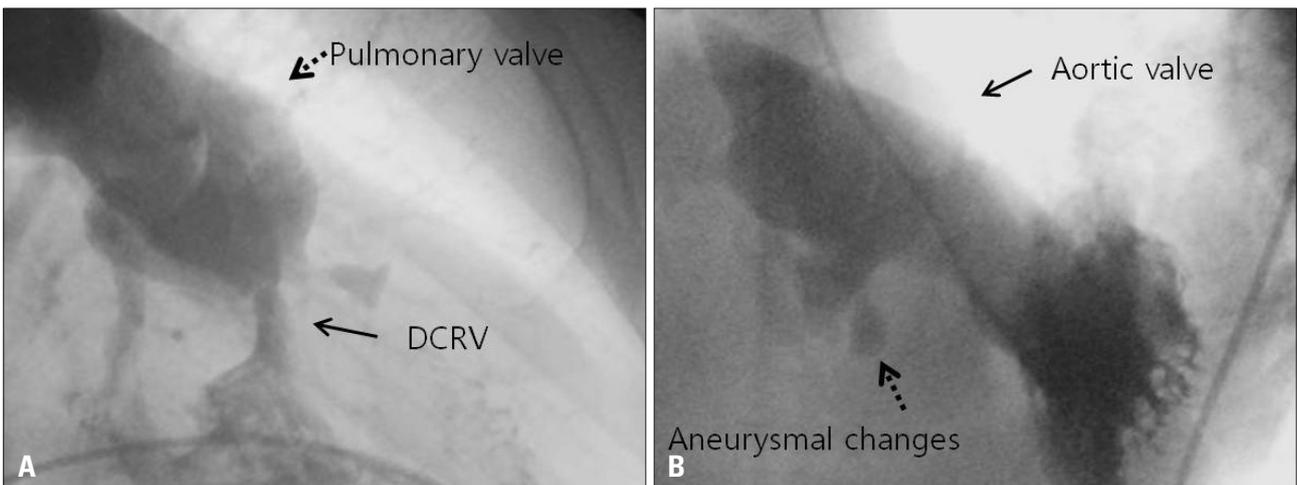
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• Address for Correspondence: Ho-Joong Youn, Department of Internal Medicine, School of Medicine, The Catholic University of Korea, 505 Banpo-dong, Seocho-gu, Seoul 137-701, Korea Tel: +82-2-3779-1066, Fax: +82-2-1588-1511, E-mail: younhj@catholic.ac.kr

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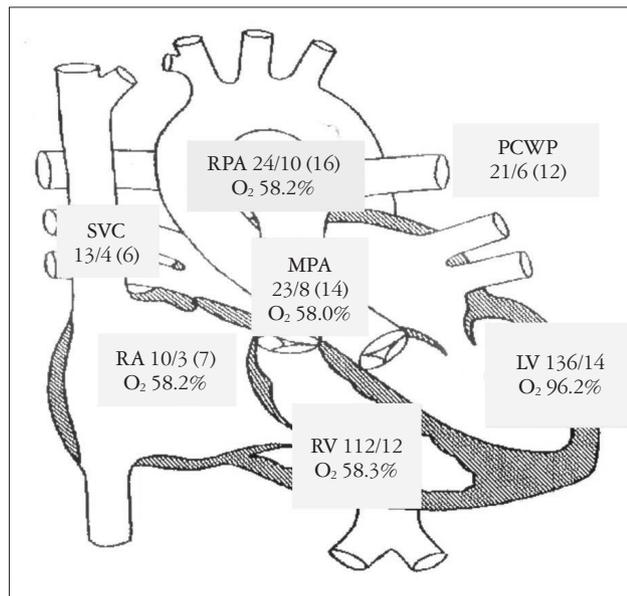


**Fig. 1.** Preoperative transthoracic and transesophageal echocardiography. (A) Transthoracic echocardiography reveals a high-velocity systolic jet arising in the mid right ventricular outflow tract. (B) Right ventricular hypertrophy is seen on the parasternal long axis view. Transesophageal echocardiography reveals a subaortic ridge (arrowhead) (C) and aneurysmal changes (arrowhead) of the ventricular septal defect with anterior deviated interventricular septum (D). AV: aortic valve, RV: right ventricle, LV: left ventricle, IVS: interventricular septum.



**Fig. 2.** Preoperative right and left ventriculography. Right ventriculography at end-systole reveals an anomalous muscle bundle dividing the cavity into 2 chambers (A), and left ventriculography shows spontaneous closed malalignment ventricular septal defect with aneurysmal changes (B). DCRV: double-chambered right ventricle.

made because the ultrasound beam could not be made parallel to the outflow tract jet. Right ventriculography confirmed an anomalous obstruction within the right ventricle. Preoperative cardiac catheterization demonstrated a right ventricle-to-pulmonary artery gradient of 89 mmHg (Fig. 2 and 3). Left ventriculography revealed a membranous VSD with anteriorly



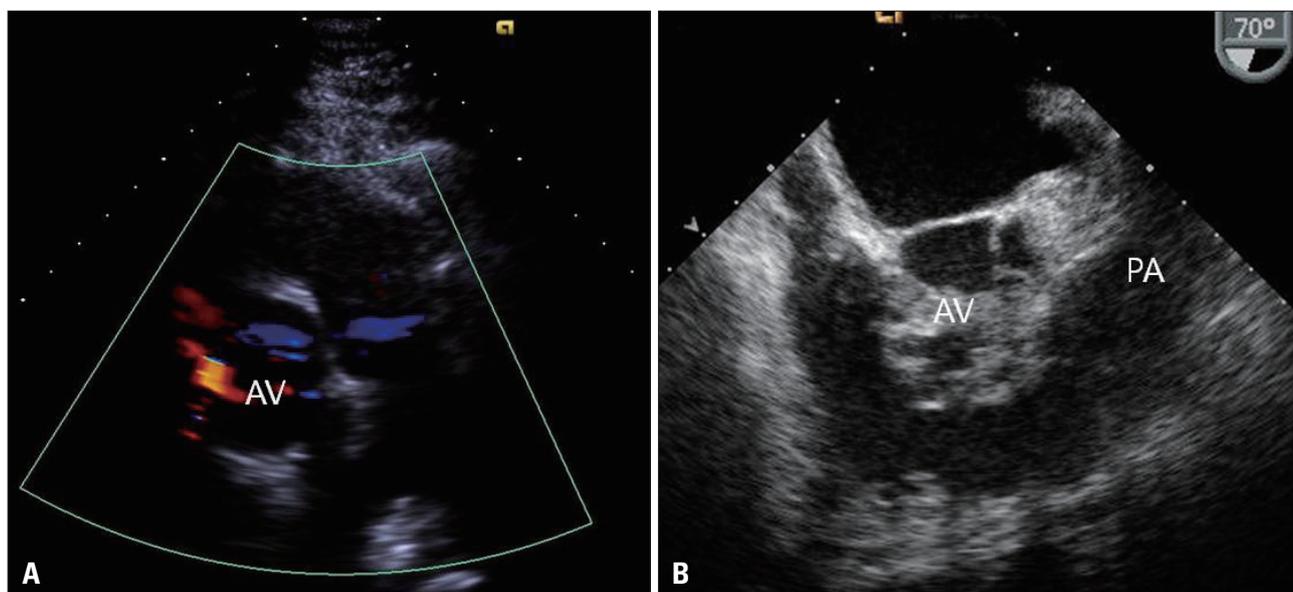
**Fig. 3.** Preoperative cardiac catheterization demonstrates a right ventricle-to-pulmonary artery gradient of 89 mmHg. There is no significant step-up of O<sub>2</sub> saturation in the right ventricular sample that is higher than the highest right atrial sample because of spontaneously closed malaligned ventricular septal defect. Systolic pressure/diastolic pressure is expressed as a mean pressure and O<sub>2</sub> saturation as a percentage. SVC: superior vena cava, RA: right atrium, RV: right ventricle, MPA: main pulmonary artery, RPA: right pulmonary artery, PCWP: pulmonary capillary wedge pressure, LV: left ventricle.

malaligned interventricular septum, aneurysmal changes. The patient underwent open-heart surgery involving resection of an encircling infundibular fibrous band through right atriotomy. Resection of the subaortic ridge was not carried out because it caused mild aortic regurgitation without pressure gradients in the left ventricular outflow tract. Dyspnea was improved at follow-up. Transthoracic echocardiography taken after surgery revealed that the high-velocity systolic jet in the mid right ventricular outflow tract disappeared and that right ventricular outflow tract diameter was much improved (Fig. 4).

### DISCUSSION

Approximately two-thirds of patients have VSD that is diminished in size by aneurysmal changes with apposition of tricuspid tissue over the VSD.<sup>4</sup> In our case, transthoracic and transesophageal echocardiography revealed a membranous type VSD with anteriorly malaligned interventricular septum, aneurysmal changes. The parasternal long axis view is useful for determining the degree of aortic overriding (Fig. 1D). DCRV is an infundibular pulmonary stenosis resulting from discrete fibromuscular narrowing or hypertrophied subvalvular muscle bundles. In many cases, VSD is combined DCRV.<sup>1-3</sup> Also, malaligned between the outlet and trabecular septa VSD with DCRV is associated with TOF.<sup>6(8)9</sup> In our patient, DCRV was not associated TOF because anteriorly malaligned interventricular septum without malalignment of infundibular septum was combined. VSD was located very close to the right aortic cusp and showed spontaneous closure with aneurysmal formation, which is rare without aortic valve prolapse.<sup>9-11</sup>

The subaortic ridge is a fibrous structure from the crest of the ventricular septum to the left ventricular outflow tract. This case had a subaortic ridge caused by increased turbulence



**Fig. 4.** Postoperative transthoracic and transesophageal echocardiography. A: Transthoracic echocardiography reveals that a high-velocity systolic jet arising in the mid right ventricular outflow tract disappeared after surgery. B: Transesophageal echocardiography reveals much improved right ventricular outflow tract diameter after surgery. AV: aortic valve, PA: pulmonary artery.

adjacent to the area of VSD and enhanced by septal malalignment.<sup>6,7)</sup> A subaortic ridge may result in progressive subaortic stenosis or aortic valve deformities, and resection is usually recommended. In our case, the patient underwent resection of an encircling infundibular fibrous band through right atriotomy. However, resection of the subaortic ridge was not carried out because it caused mild aortic regurgitation without any pressure gradients in the left ventricular outflow tract.

In summary, the malaligned interventricular septum in membranous type VSD can develop secondary to aneurysmal changes of VSD. We reported the unusual case of a patient who had a pathology complex comprising DCRV, subaortic ridge, and VSD with malaligned interventricular septal wall and survived with surgical treatment.

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