

## CASE REPORT

# Incidentally detected atrioventricular septal defect in an adult

Risa Shimbori<sup>1</sup>  | Jun Takaki<sup>1</sup> | Yasuhito Hosoda<sup>1</sup> | Ken Okamoto<sup>1</sup> | Koji Fukae<sup>2</sup> | Toshihiro Fukui<sup>1</sup> 

<sup>1</sup>Department of Cardiovascular Surgery, Kumamoto University Hospital, Kumamoto, Japan

<sup>2</sup>Department of Pediatric Cardiac Surgery, Kumamoto City Hospital, Kumamoto, Japan

## Correspondence

Risa Shimbori, Department of Cardiovascular Surgery, Kumamoto University Hospital, Seirei Hamamatsu General Hospital, 2-12-12 Sumiyoshi, Hamamatsu 430-8558, Japan.  
Email: k.hchd.1991@gmail.com

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## Abstract

A 34-year-old woman, a liver transplantation donor for her son, was referred to our hospital for preoperative evaluation. She was diagnosed with an incomplete atrioventricular septal defect (AVSD). She underwent double valve repair and patch closure of the defect. Incidental discovery of an asymptomatic AVSD in an adult is rare.

## KEYWORDS

adult, adult congenital heart disease, atrioventricular septal defect

## 1 | INTRODUCTION

The term “atrioventricular septal defect” (AVSD) covers a group of diverse malformations that have the same characteristic morphology of their atrioventricular septal junction. AVSDs account for about 7%–17% of all congenital heart diseases.<sup>1</sup> Most patients undergo surgery during childhood, with favorable long-term survival, although a few patients with incomplete AVSDs are not diagnosed until adulthood. It has been suggested that delayed surgery increases the incidence of preoperative complications and influences cardiac function.<sup>2</sup> We report a rare case of an asymptomatic AVSD that was incidentally detected in an adult patient.

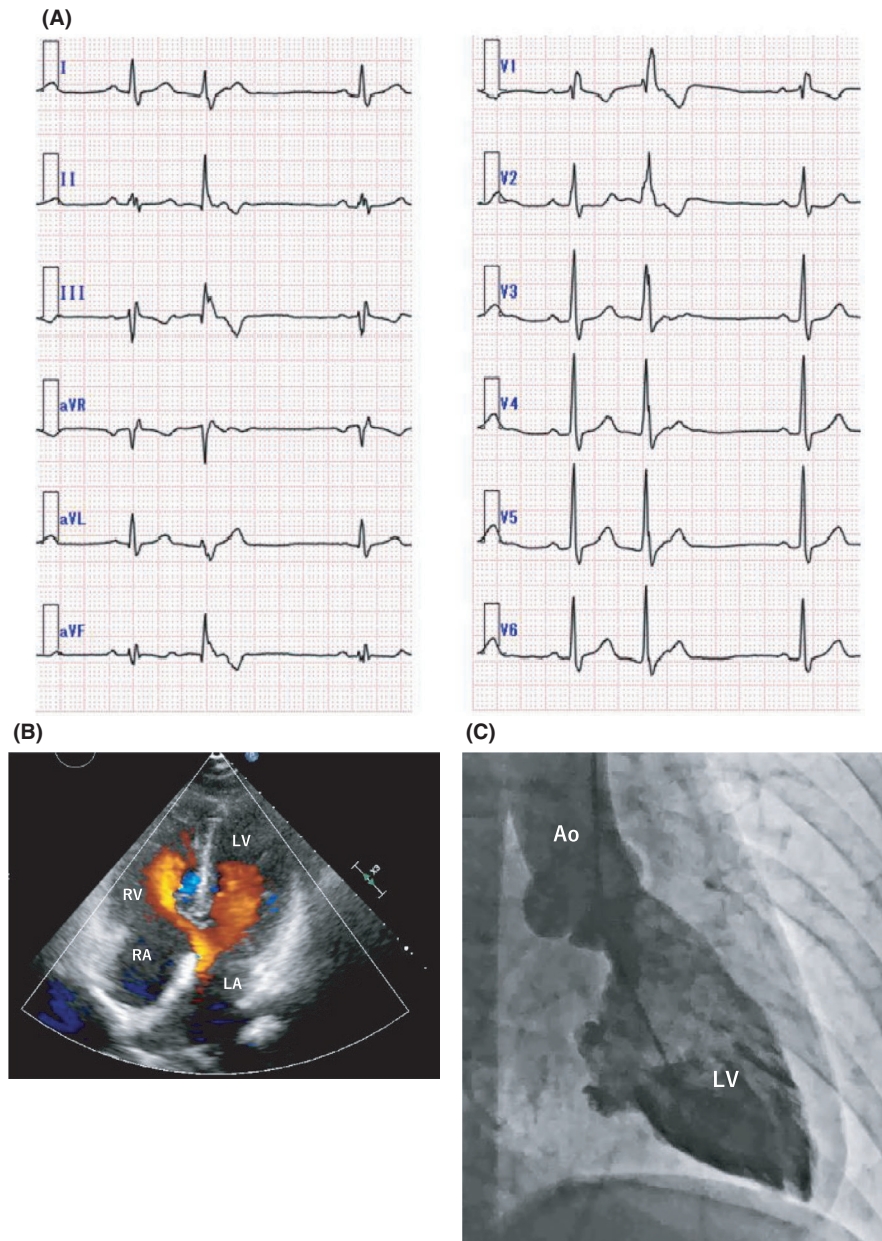
## 2 | CASE REPORT

A 34-year-old woman who had planned to be a liver transplantation donor for her son was referred to our hospital

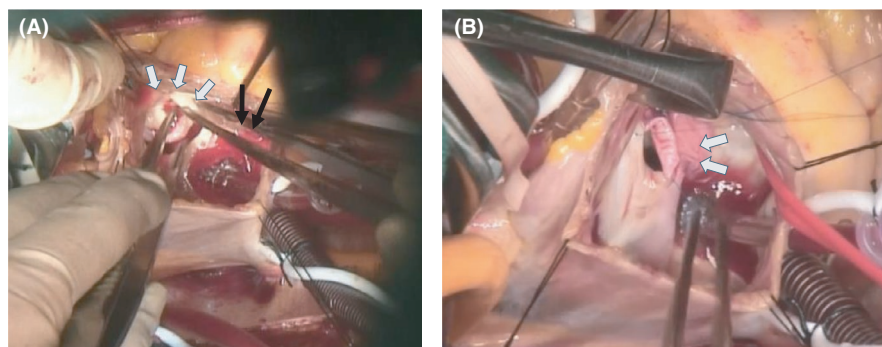
for preoperative evaluation. She had no symptoms and had an unremarkable medical history. She had undergone a successful vaginal delivery. Her vital signs were stable upon admission. Cardiac auscultation revealed a grade 3/6 systolic ejection murmur at the second left sternal border and fixed splitting of the second heart sound. Twelve-lead electrocardiography showed sinus rhythm, left axis deviation, and an incomplete right bundle branch block pattern (Figure 1A). Two-dimensional echocardiography revealed preserved left ventricular contractility, although both atria and right ventricle were dilated. Moreover, there was a defect in the lower part of the atrial septum and a restrictive ventricular septal defect (VSD). Color Doppler echocardiography showed a left-to-right atrial shunt, mild-to-moderate left atrioventricular valve (LAVV) regurgitation, and moderate right atrioventricular valve (RAVV) regurgitation (Figure 1B). Cardiac catheterization revealed a gooseneck deformity (Figure 1C). Right-heart catheterization showed a mean pulmonary

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**FIGURE 1** (A) Twelve-lead electrocardiography showed sinus rhythm, left axis deviation, incomplete right bundle branch block pattern. (B) Four chamber view of transthoracic echocardiogram shows a left atrial to right atrial shunt. (C) Cardiac catheterization shows gooseneck deformity. (LV, left ventricle; LA, left atrium; RV, right ventricle; RA, right atrium)



**FIGURE 2** Operation. (A) White and black arrows indicate the cleft of left atrioventricular valve and ventricular septal defect, respectively. (B) Pericardial patch is sutured in place to close atrial component of defect (white arrows). Coronary sinus remains draining to right atrium

artery pressure of 16 mmHg and a mean pulmonary capillary wedge pressure of 10 mmHg. The pulmonary-to-systemic flow ratio was 2.08.

She was diagnosed with an atrioventricular septal defect. Even though she had no symptoms, she was deemed

unfit to be a liver transplant donor, and she was advised to undergo AVSD repair. Median sternotomy, cardiopulmonary bypass, and aortic cross-clamping were performed as usual. We observed a defect in the lower portion of the atrial septum, a small VSD surrounded by fibrous tissue,

and a cleft of the LAVV and RAVV (Figure 2A; white and black arrows indicate a cleft of the LAVV and VSD, respectively). It was diagnosed as an intermediate-type AVSD. We first sutured the pericardium (white arrows) to the junction of the RAVV and LAVV annuli (Figure 2B). Suturing at the area of the atrioventricular node and coronary sinus was performed very close to the LAVV. Next, the LAVV cleft was closed with interrupted sutures, and edge-to-edge repair was performed to address the RAVV regurgitation.

Postoperatively, two-dimensional echocardiography showed no leakage around the pericardial patch. Both RAVV and LAVV regurgitations were trivial (Figure 3). The patient's postoperative course was uneventful.

### 3 | COMMENT

Atrioventricular septal defect is classified into four groups: complete, partial with an isolated primum atrial septal defect, partial with an isolated inlet VSD, and intermediate. Intermediate AVSD is anatomically characterized by the presence of an ostium primum atrial septal defect occupying the portion of the atrial septum just superior to the individually formed RAVV and LAVV, and varying degrees of splitting of the anterior leaflet of the LAVV.<sup>1,3</sup> Intermediate-type AVSD is less frequent than complete or partial AVSD and is rarely encountered in adulthood.<sup>4</sup>

Complete AVSD presents early in life, and unless treated, it expediently develops into irreversible pulmonary vascular disease. The presentation of incomplete or intermediate AVSD is variable. The optimum age for repair of incomplete AVSD is 3–5 years, but some patients

are diagnosed after presenting with congestive heart failure symptoms in adulthood.<sup>5</sup>

Gatzoulis et al. reported that in 6 of the 10 of incomplete AVSD adult patients thought to be asymptomatic before operation, the functional capacity improved postoperatively, regardless of age. They also reported that the long-term survival after repair of incomplete AVSD in adults is favorable, corresponding to 86% after 10 years. As pulmonary arterial hypertension, complete atrioventricular block, and atrial arrhythmias are risk factors for mortality and morbidity, the authors recommend early surgery for AVSD, because older patients are more likely to present with the added complications of atrial arrhythmia and moderate or severe RAVV regurgitation.<sup>2</sup> Whether routine LAVV cleft closure during primary repair in adult patients with partial AVSD decreases the reoperation rate remains unclear; previous studies repaired the cleft of the LAVV using a suture technique between bridging leaflets and an additional annuloplasty.<sup>2</sup> Zhou et al.<sup>6</sup> also reported that in adult patients with moderate/severe LAVV regurgitation, repair using an additional band implantation significantly reduced the incidence of recurrent regurgitation and improved long-term outcomes. In our case, because LAVV regurgitation was mild to moderate, an additional annuloplasty was not necessary.

Fortunately for our patient, her pathology was diagnosed incidentally before she developed a complicated preoperative condition. Although her cardiac function had been preserved without arrhythmias, she was not allowed to be an organ donor. We believe that it is important for cardiologists to recognize that it is possible, although rare, for adult patients to be diagnosed incidentally with asymptomatic AVSD.

### ACKNOWLEDGEMENT

Published with written consent of the patient.

### CONFLICT OF INTEREST

None declared.

### AUTHOR CONTRIBUTIONS

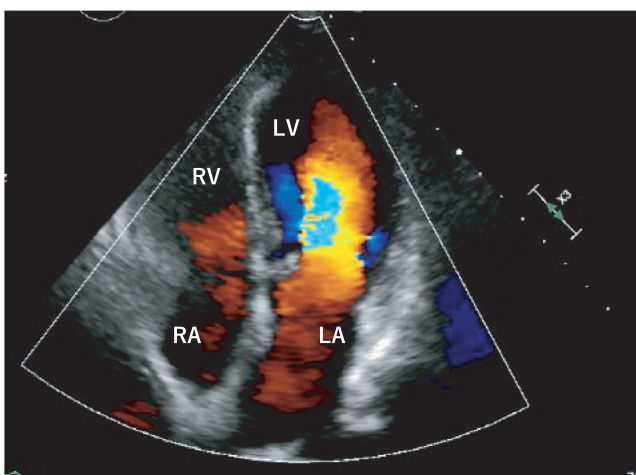
Risa Shimbori, Jun Takaki, Hosoda Yasuhito, Ken Okamoto, and Koji Fukae: Drafting article. Toshihiro Fukui: Critical revision.

### ETHICAL APPROVAL

Written informed consent was obtained from the patient.

### CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.



**FIGURE 3** Post-operative transthoracic echocardiogram, there was no leak around pericardial patch. (LV, left ventricle; LA, left atrium; RV, right ventricle; RA, right atrium)

## DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

## ORCID

Risa Shimbori  <https://orcid.org/0000-0001-6909-9860>

Toshihiro Fukui  <https://orcid.org/0000-0002-4110-6237>

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