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CASE REPORT

Anatomical correction of atrioventricular discordance using three-dimensional replica

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Abstract
Surgical experience with {S,L,D} segmental anatomy of atrioventricular discordance with double-outlet right ventricle is extremely rare. In addition to ordinary cardiac examination, we reviewed electrophysiological studies and a three-dimensional cardiac replica (crossMedical, Inc., Kyoto, Japan). Consequently, we preoperatively confirmed the intracardiac rerouting line and the appropriate right ventricle incision line. A Senning procedure, intracardiac rerouting, and subaortic stenosis resection were performed in a 2.6-year-old patient (weight, 10.6 kg). The three-dimensional cardiac replica contributed definitively to the anatomical correction.

Keywords Atrioventricular discordance · Double-outlet right ventricle · Anatomical correction · Senning · Intracardiac rerouting

Introduction

Other than the surgical reports that describe the corrected transposition of the great arteries, few surgical reports describing atrioventricular discordance (AVd) have also been published. Particularly, reports on surgical experience in {S,L,D} segmental anatomy of AVd with double-outlet right ventricle (DORV) are extremely rare; the anatomical characterization, too, remains unclear.

Case

A female neonate, born after a 38-week 1-day gestation (weight 2.340 kg), had been misdiagnosed as having false Taussig–Bing anomaly and aortic coarctation during the neonatal period. Extended aortic arch anastomosis and atrial septal defect enlargement with pulmonary artery banding (circumference 18 mm) and repeat pulmonary artery banding (unfastening to a circumference of 20 mm) were performed at 4 days and 1 month of age, respectively. Subsequently, {S,L,D} segmental anatomy of AVd, DORV, and subaortic stenosis could be diagnosed. Large subaortic and small muscular ventricular septal defects (VSDs) were detected. At 3 months of age, cardiac catheterization revealed left ventricular (LV) and right ventricular (RV) end-diastolic volumes that were 145 and 99% of normal, respectively. A three-dimensional cardiac replica (3DCR; crossMedical, Inc., Kyoto, Japan) was constructed using computed tomography data. Considering the intraventricular rerouting space, higher RV end-diastolic volume and body weight were preferable for anatomical correction. At 7 months of age, electrophysiological studies revealed posterior atrioventricular bundle because of electric potential at the posteroinferior side of VSD. A right modified Blalock–Taussig shunt was performed at the age of 10 months using a 5-mm expanded polytetrafluoroethylene (ePTFE) graft (Gore & Associates, Arizona, USA).

At 2.5 years of age, cardiac catheterization revealed LV and RV end-diastolic volumes that were 94 and 178% of normal, respectively [LV/RV/ascending aorta (Ao)/pulmonary artery (PA) systolic pressures, 94/95/76/15 mmHg; LV/RV/Ao/PA oxygen saturations, 83/76/80/80%; pulmonary vascular resistance, 0.89 u m^2; and PA index, 167 mm^2/

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m^2 (Fig. 1a, b)]. Coronary artery location was confirmed (Fig. 1c). A new 3DCR providing real-time images of intraventricular rerouting and ventricular incision line was constructed (Fig. 2a).

At 2.6 years of age (weight, 10.6 kg), a Senning procedure, intraventricular rerouting, subaortic stenosis resection, and PA patch angioplasty were performed. Cardiopulmonary bypass with Ao and bicaval cannulations was initiated. The Blalock–Taussig shunt was removed, and antegrade blood cardioplegia was performed. The lateral side of the RV at a 10-mm distance from the pulmonary valve was incised 30 mm in diameter in a longitudinal fashion. The intraventricular rerouting suture comprised a 5-0 polypropylene pledgeted (4-mm spaghetti) suture (Fig. 2b–d). Subaortic stenosis was resected, and VSD was closed using a 0.4-mm ePTFE patch. The Senning procedure for the systemic venous route floor was performed using a 0.4-mm ePTFE patch, because little atrial septal tissue was preserved. The small muscular VSD at the apex portion was not seen from the initial RV incision. Because of the small VSD shunt, the operation was continued using the previously reported beating-heart technique with continuous coronary perfusion [1]. PA patch angioplasty from the banding proximal to the right PA distal

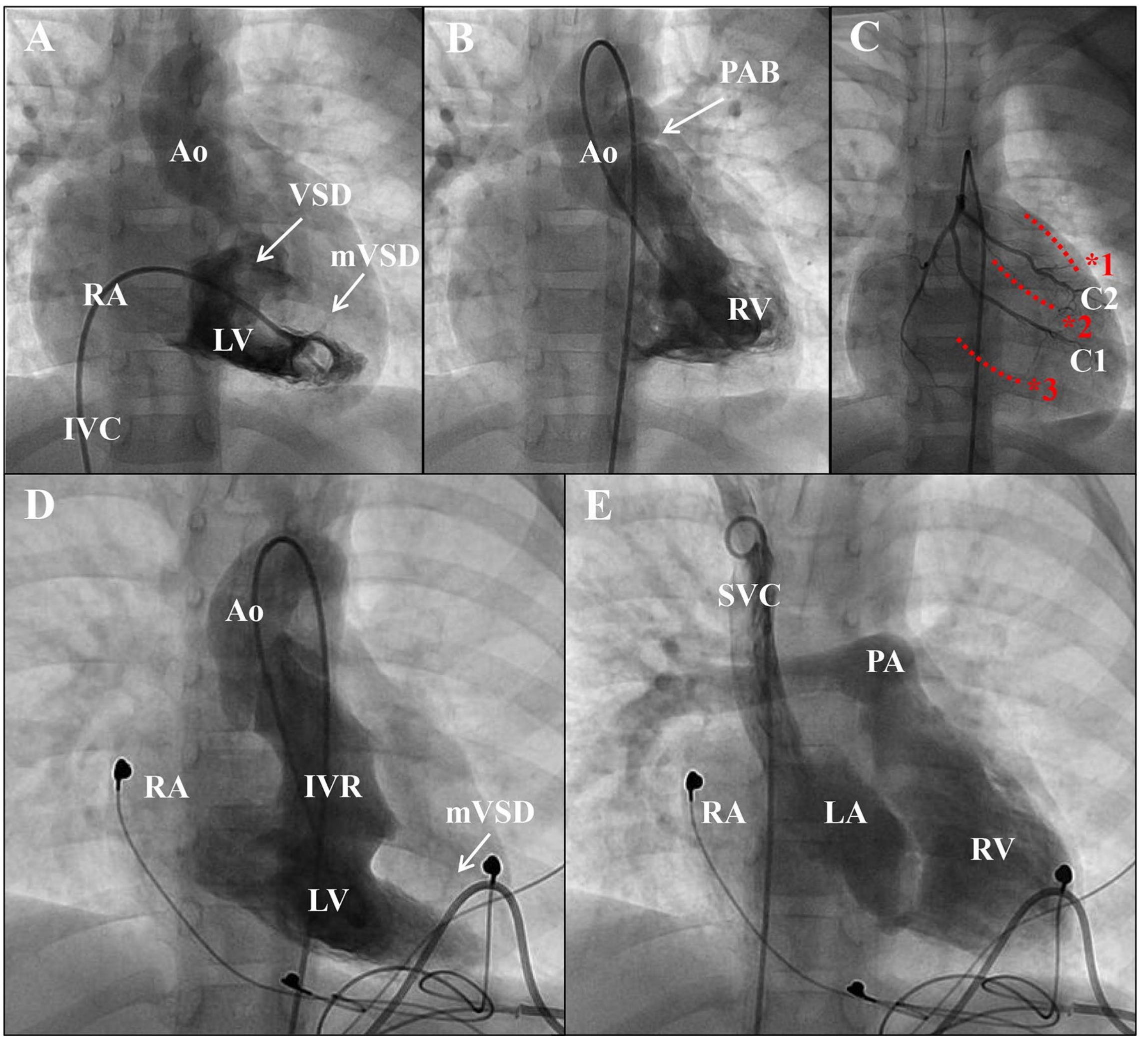

Fig. 1 Preoperative (A, B, C) and postoperative (D, E) cardiac catheterization. **a** Frontal view of the LV angiogram. The catheter was inserted into LV through RA from the IVC. Large subaortic VSD and small muscular VSD are indicated. **b** Frontal view of the RV angiogram. The catheter was inserted into RV from Ao. **c** Frontal view of the coronary angiogram. C1 indicates the coronary artery on the intraventricular septum. C2 indicates a major coronary branch on the RV free wall. The three red dotted lines indicate considerable ventricular incision lines. **d** Frontal view of the LV angiogram. The catheter was inserted into LV through the intraventricular route from Ao. There was no intraventricular route pressure gradient. Despite leaving a very small muscular VSD (mVSD), the calculated pulmonary-to-systemic blood flow ratio was 1.0. **e** Frontal view of the SVC angiogram. Senning route, LA, RV, and PA were enhanced from SVC angiography. *Ao* ascending aorta, *IVC* inferior vena cava, *LA* left atrium, *LV* left ventricle, *PA* pulmonary artery, *PAB* PA banding, *RA* right atrium, *RV* right ventricle, *VSD* ventricular septal defect

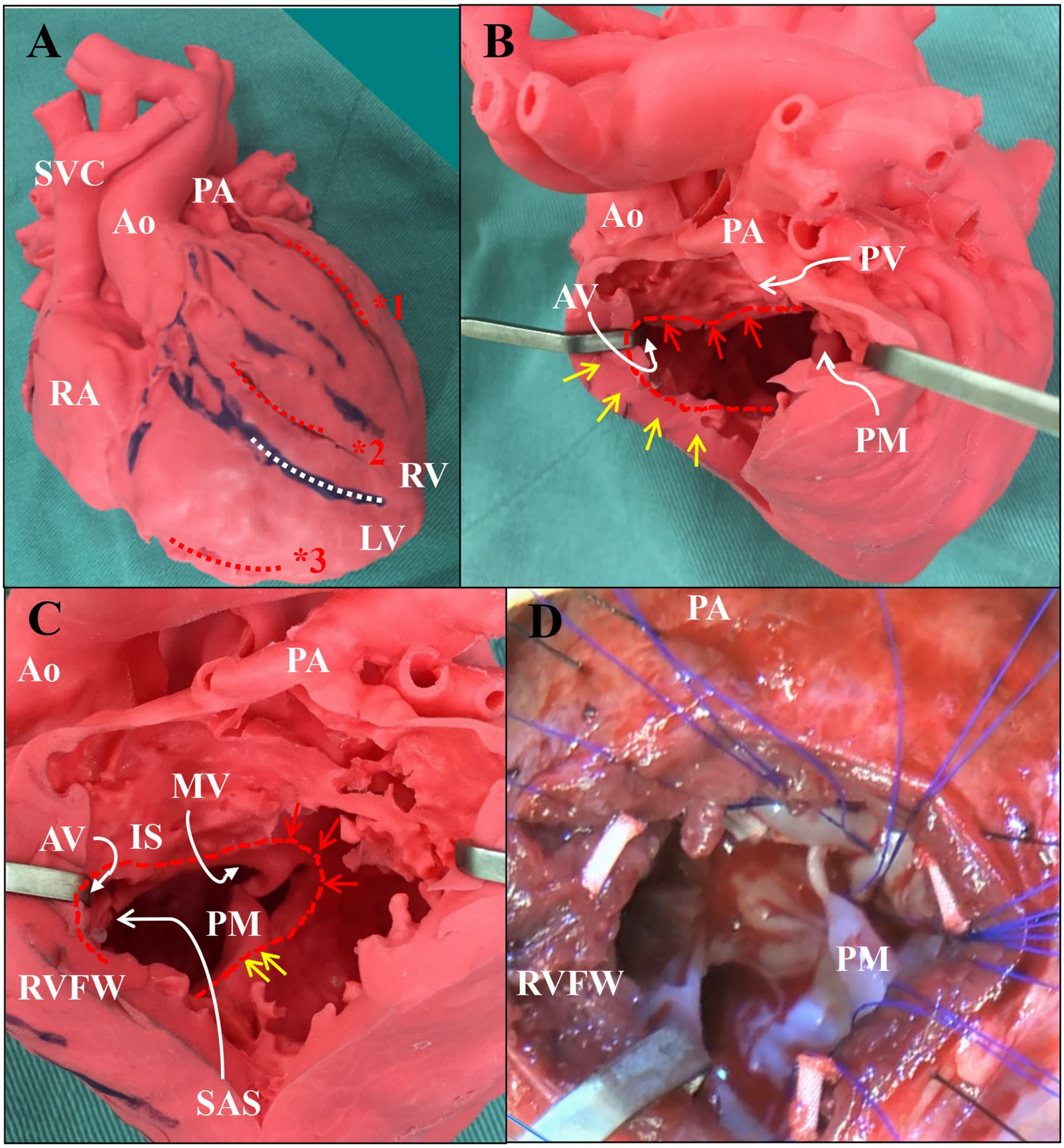

Fig. 2 Preoperative 3DCR (A, B, C) and intraoperative picture (D). **a** Complete 3DCR image. The three red dotted lines indicate considerable ventricular incision lines. Incision *1 was at the lateral side of RV, *2 was at the LV side of RV, and *3 was on LV. The white dotted line indicates the IVS. **b, c** RV lateral incision view of the 3DCR. The red dotted line indicates the suture line of the VSD patch. IS between aortic and pulmonary valve were sutured from the LV side (red arrows in **b**). The basal part of the tricuspid valve was sutured (red arrows in **c**). The inlet septum of VSD was sutured at a 5-mm distance from the crest (between red arrows and yellow arrows in C). The basal part of PM was sutured (yellow arrows in **c**). The anterior-to-superior portion of VSD was sutured from RVFW (yellow arrows in **b**). The SAS portion was made of IS of the right side of AV. **d** Intraoperative view of RV lateral incision from the ceiling camera. A total of 19 sutures were placed around VSD. *Ao* ascending aorta, *AV* aortic valve, *3DCT* three-dimensional computed tomography, *IS* infundibular septum, *IVS* intraventricular septum, *LV* left ventricle, *MV* mitral valve, *PA* pulmonary artery, *PM* papillary muscle of mitral valve, *RA* right atrium, *RV* right ventricle, *RVFW* RV free wall, *SAS* subaortic stenosis, *SVC* supra vena cava, *VSD* ventricular septal defect

site was performed using a trimmed 8-mm ePTFE graft. The RV incision was repaired with a mattress suture using felt strips. The patient was weaned from cardiopulmonary bypass with temporary pacing. Intraoperative blood sampling revealed a pulmonary-to-systemic blood flow ratio of 1.4; therefore, we estimated low influence of the residual VSD shunt.

A permanent pacemaker was implanted because complete atrioventricular block had postoperatively continued for 10 days. However, the atrioventricular block soon improved to sinus rhythm with bundle block. Cardiac catheterization performed at 10 months post-operation revealed LV and RV end-diastolic volumes that were 145% and 99% of normal, respectively [LV/RV/Ao/PA systolic pressures, 78/31/78/28 mmHg; pulmonary-to-systemic blood flow ratio, 1.00 (Fig. 1d, e)]. The patients' progress at postoperative 1.5 years was satisfactory.

Discussion

AVd has been most commonly associated with ventriculoarterial discordance; however, it is occasionally associated with concordance or DORV [2]. One report described Senning and trans-MV intraventricular rerouting procedures for {S,L,D} segmental anatomy of AVd with DORV [3]; however, the position of the conduction bundle location was assumed to be akin only to that in the corrected transposition of the great arteries. We decided on intraventricular rerouting procedure based on the electrophysiological studies and 3DCR images. The three incisions are depicted in Figs. 1c and 2a. The 3DCR was actually incised, and the RV lateral incision (*1) was assumed to be the best incision for intraventricular rerouting (Fig. 2b, c). The usefulness of 3DCR for cardiac surgery has been reported [4]. 3DCR may be a very useful tool in case of unfamiliar cardiac disorders.

In conclusion, we performed anatomical correction for {S,L,D} segmental anatomy of AVd with DORV in a case that was never previously encountered. In addition to ordinary cardiac examination, we reviewed the electrophysiological studies and 3DCR images. Consequently, we preoperatively confirmed the intracardiac rerouting and appropriate RV incision lines. 3DCR contributed definitively to our surgical results.

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