CASE REPORT

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Left atrioventricular valve regurgitation repair with concomitant outflow obstruction release on repaired atrioventricular septal defect case

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Abstract

Background Left ventricular outflow tract stenosis and atrioventricular valve regurgitation are often problems encountered in adulthood after complete atrioventricular septal defect repair. The surgical approach and indications for managing long-term outcomes such as left atrioventricular valve regurgitation and left ventricular outflow tract stenosis after complete atrioventricular septal defect repair have been discussed.

Case presentation A 23-year-old woman with intellectual disability was diagnosed with complete atrioventricular septal defect and underwent two-patch repair without cleft closure in childhood. Follow-up examination in adulthood demonstrated moderate left-sided atrioventricular valve regurgitation and left ventricular outflow tract stenosis with a circumferential ridge (peak velocity, 3.7 m/s; pressure gradient, 54 mmHg). Intraoperative findings showed a circumferential ridge under the aortic valve, and we removed the ridge. In addition, a cleft was present at the anterior leaflet, and we completely closed the cleft. Anticoagulation therapy was not initiated, and no embolic complications occurred. Follow-up echocardiography demonstrated no ridge under the aortic valve and only mild-range left AVVR.

Conclusions We successfully performed surgical treatment without valve replacement or anticoagulation therapy in a patient with poor medical compliance. Delayed reoperation leads to degeneration of the valve structure and makes more difficult to repair. Atrioventricular valve regurgitation should be evaluated in combination with based on the etiology of the regurgitation especially cleft related or not, in addition to the dilatation annulus, cleft size, and depth of the leaflet coaptation depth, and associated other valve diseases.

Keywords Complete atrioventricular septal defect, Atrioventricular valve regurgitation, Left ventricular outflow tract stenosis

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Background

The surgical approach and indications for managing long-term outcomes such as left atrioventricular valve regurgitation (AVVR) and left ventricular outflow tract stenosis (LVOTS) after complete atrioventricular septal defect (AVSD) repair have been discussed [1, 2].

Case presentation

A 23-year-old woman who had undergone two-patch repair with a TISSUE-GUARD patch (Baxter, Deerfield, IL, USA) for Rastelli type A complete AVSD in childhood was referred to our department because of left AVVR and LVOTS. Asymptomatic mild AVVR and LVOTS were noted after the intracardiac repair in childhood. Follow-up echocardiography showed progression of the AVVR and LVOTS.

Electrocardiogram showed complete right bundle branch block and QRS width of 0.128 s.

Three-dimensional transesophageal echocardiography showed a completely open cleft of the left atrioventricular valve leaflet (Fig. 1A) and a moderate-range regurgitation jet (vena contracta=3.8 mm) (Fig. 1B). Additionally, a circumferential ridge was present under the aortic valve (Fig. 1C). The peak velocity through the LVOT was 3.7 m/s with a pressure gradient of 54 mmHg. Enhanced computed tomography demonstrated a ridge of the LVOT 10 mm beneath the aortic valve (Fig. 1D). No restriction of aortic valve motion or calcification was present.

Cardiopulmonary bypass was established with aortic perfusion and right internal jugular and inferior vena cava drainage. The aortic valve had no degeneration, but a circumferential ridge without calcification was found approximately 10 mm beneath the aortic valve, without continuity to the left atrioventricular valve annulus (Fig. 2A). We circumferentially removed the ridge. When



Fig. 1 Preoperative examinations. A: Completely opened cleft. B: Severe regurgitation from a cleft. C: Abnormal ridge of the LVOT. D: Ridge of the LVOT 10 mm beneath the aortic valve



Fig. 2 Intraoperative findings. A: Abnormal ridge of the LVOT was located 10 mm beneath the aortic valve. B: Completely opened cleft of left atrioventricular valve leaflet

approaching the left atrioventricular valve through the atrial septum, a large cleft at the middle of the anterior leaflet caused regurgitation from the cleft without annular dilatation (Fig. 2B). The total length of the cleft was examined by attaching a thread to the tip of the cleft and pulling it forward. We then completely closed the cleft, and a saline test showed no leakage (Video 1). Ring annuloplasty was not performed because of its potential to cause outflow stenosis.

The postoperative course was uneventful, and the patient was discharged on postoperative day 11. Anticoagulation therapy was not initiated, and no embolic complications occurred. Follow-up echocardiography demonstrated no ridge under the aortic valve and only mild-range left AVVR.

Discussion and conclusions

Left AVVR and LVOTS are common long-term problems after AVSD repair. An unclosed cleft is a well-known cause of left AVVR [1, 2]. Stulak et al. [1] reported that the most common indication for reoperation in AVSD was left AVVR, partly caused by an unclosed cleft and previous cleft repair dehiscence. They found no differences in outcomes between valve repair and replacement.

Alexi-Meskishvili et al. [2] demonstrated that valve anomalies, including dysplastic valvular tissue and fibrotic leaflets, can lead to recurrent regurgitation or valve replacement. We speculate that delayed reoperation leads to degeneration of the valve structure. However, quantitative evaluation of AVVR is difficult because of the eccentric regurgitation jet, and the optimal timing for reoperation might be missed. The guideline indicates that the only criterion for surgical intervention of AVVR is the grade of regurgitation, and do not mention about the associated disease entities. The surgical indication should not be based only on the regurgitation grade. Clinicians must evaluate AVVR in combination with the dilatation annulus, cleft size, and depth of the leaflet coaptation area. Furthermore, maybe other information such as symptoms, size of heart, cardiac contraction, and BNP levels should also be included in the evaluation.

In terms of repair techniques, complete cleft closure is required for long-term durability, and ring annuloplasty might be useful [1, 2]. When performing annuloplasty in AVVR repair, care should be taken to avoid creating postoperative LVOTS. Foker [3] reported that discrete subaortic stenosis has a circular fibromuscular rim of tissue and is an acquired lesion. The author also stated that discrete subaortic stenosis is generated by shear forces and is associated with cardiac structural abnormalities [3]. Leichter et al. [4] described 35 patients with subaortic valvular stenosis, 2 of whom had an AVSD anatomy. The authors speculated that anatomical outflow narrowing in AVSD leads to development of subaortic valvular stenosis [4]. In our case, left AVVR secondary to a large cleft could be controlled by cleft closure alone; ring annuloplasty was unnecessary.

Abbreviations

 AVSD
 Complete atrioventricular septal defect

 AVVR
 Atrioventricular valve regurgitation

 LVOTS
 Left ventricular outflow tract stenosis

Supplementary Information

The online version contains supplementary material available at https://doi. org/10.1186/s13019-024-02991-6.

 $\label{eq:supplementary} \begin{array}{l} \textbf{Supplementary Material 1: Video 1.} \\ \textbf{Surgical procedures for LVOTS and} \\ \textbf{left} \ \textbf{AVVR} \end{array}$

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Author contributions

T.K.: Data curation; Writing-original draft, K.I.: Conceptualization; Writing-review & editing, H.J.: Data curation, S.M.: Data curation, H.S.: Supervision.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Informed consent was obtained from patient using our institutional consent form and an opt-out method.

Consent for publication

Informed consent was obtained from patient using our institutional consent form and an opt-out method.

Competing interests

The authors declare no competing interests.

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