



Diagnosis and surgical treatment for complete atrioventricular septal defect with intact atrial septum

Brief Report

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Abstract

A six-month-old boy with Down syndrome, complete atrioventricular septal defect with intact atrial septum, and moderate left-sided atrioventricular valve regurgitation underwent surgery. The presence of a common atrioventricular junction could not be diagnosed preoperatively. Postoperative left-sided atrioventricular valve regurgitation remained mild by complete cleft closure, direct closure of incised atrial septum, and patch augmentation of the underside of septum primum.

Introduction

Intact atrial septum, or absence of septum primum atrial septal defect, can be rarely associated with atrioventricular septal defect.¹ Diagnosis is difficult due to lack of typical ventricular deformity as atrioventricular septal defect and post-surgical left-sided atrioventricular valve outcome was poor.²⁻³ Herein, we report a surgical case who was not preoperatively diagnosed.

Case

A 6-month-old male infant with Down syndrome, a perimembranous ventricular septal defect with inlet extension, and moderate mitral valve regurgitation was admitted for surgery. In the apical 4-chamber view, there appeared to be an offset between the tricuspid and mitral valves. No ostium primum atrial septal defect was found (Fig. 1A, B). In the subxiphoid short-axis view, the annulus was divided into two parts, left and right, rather than forming a common atrioventricular valve, and the mitral regurgitation appeared to originate from a cleft (Fig. 1C, D). Cardiac catheterization showed pulmonary over-circulation with a pulmonary to systemic blood flow ratio of 2.5 without evidence of pulmonary hypertension.

The operation was performed through a median full sternotomy. Thorough inspection of inside of the heart from right atriotomy, the diagnosis was confirmed as a common atrioventricular junction without an ostium primum atrial septal defect (Fig. 2A, B). Following the incision of the primum atrial septum, the zone of apposition between both bridging leaflets was completely closed (Fig. 2C). Subsequently, the ventricular component, only under the superior bridging leaflet, was closed using an expanded polytetrafluoroethylene patch. A series of mattress stitches secured the patch to the superior bridging leaflet along the septation line between the right and left components. The superior edge of a fresh autologous pericardial patch was also incorporated, with the inferior edge sutured to the surface of the inferior bridging leaflet. Finally, the created gap between the underside of the septum primum and the surface of the septation line was augmented (Fig. 2D). Weaning from cardiopulmonary bypass was smooth. Postoperative left-sided atrioventricular valve regurgitation remains mild.

Comment

Atrioventricular septal defect can be classified into 3 types by presence or absence of common atrioventricular valve and inlet ventricular septal defect, without considering the presence of a septum primum atrial septal defect. Indeed, partial atrioventricular septal defect with inlet ventricular septal defect and intact atrial septum is reported to exist.⁴ In the presented case, the

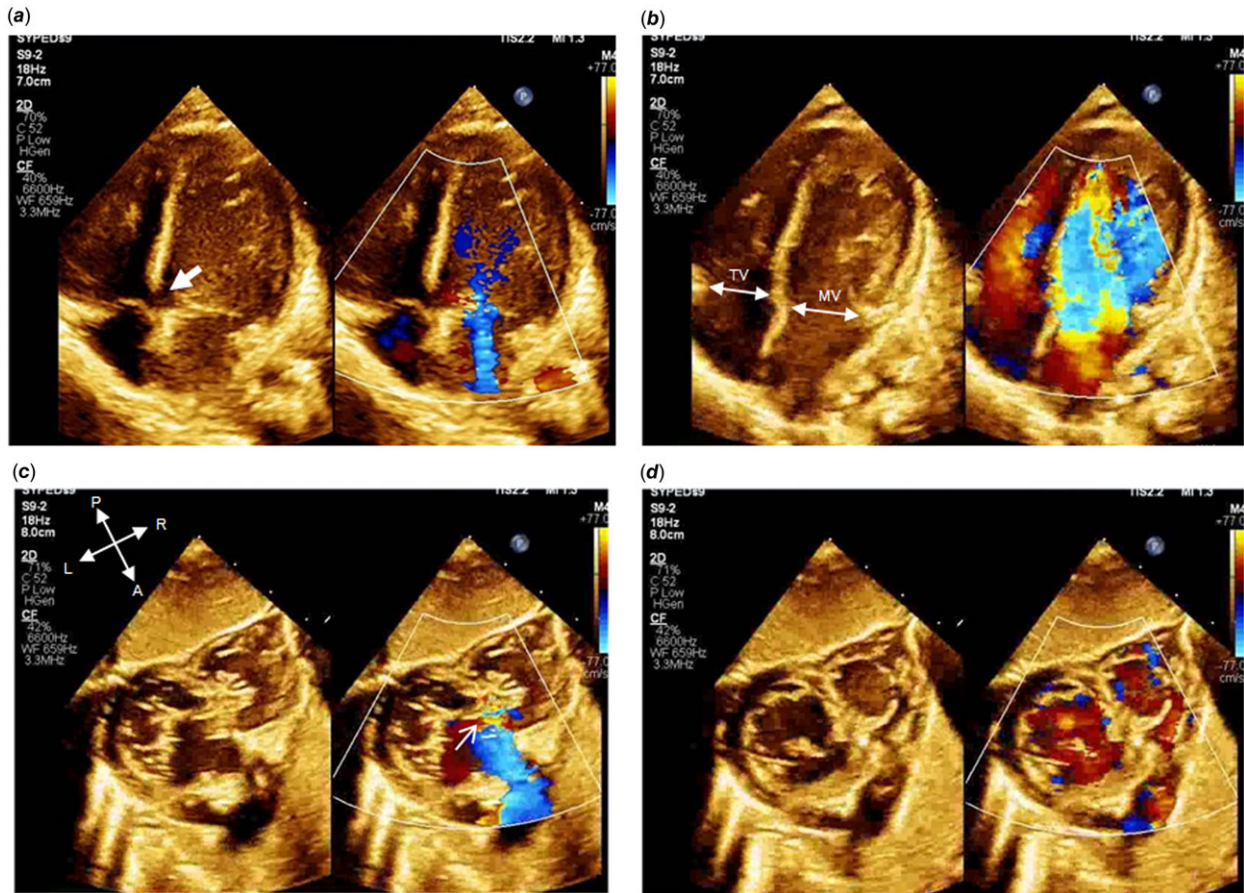


Figure 1. Preoperative echocardiography images. (a) Moderate mitral regurgitation and ventricular septal defect extending in the inlet direction (white arrow) are observed. (b) There appears to be an offset between the mitral valve (MV) and the tricuspid valve (TV). (c) The annulus seems to be divided into two parts. (d) The mitral regurgitation originates from a cleft (white arrow).

atrial septum and each bridging leaflet was connected by fibrous tissue existed at the upper surface of bridging leaflets, giving the appearance of having two valve annuli; however, we diagnosed it as complete atrioventricular septal defect rather than intermediate because the coaptation zone level, both bridging leaflets were not fused with the connecting tongue, indicating a common atrioventricular junction with a single atrioventricular valve orifice (Supplemental figure S1).

The present case had a common atrioventricular valve with intact atrial septum, which is known to be highly associated with Down syndrome.^{2,3} So, when a patient with Down syndrome has an inlet ventricular septal defect and cleft mitral valve even though not ostial primum atrial septal defect, common atrioventricular junction should be suspected.¹ Careful observation of subxiphoid short-axis view with sweeping from right atrium to left ventricular apex is reported to be able to reveal a common atrioventricular valve in diastolic frame and the absence of septal attachments of the superior bridging leaflet in systole.²

Postoperative significant left-sided atrioventricular valve regurgitation was reported to be highly associated with complete atrioventricular septal defect with an intact atrial septum.^{2,3} In this anatomy, both bridging leaflets are attached to the posterior rim of the septum primum. This results in a shallow coaptation of the cleft between the bridging leaflets, which may cause significant postoperative regurgitation. Therefore, if possible, the cleft should be completely closed. Additionally, incised atrial septum should be closed directly as well as a gap between underside of septum primum and surface of bridging leaflets at ventricular septation should be augmented with patch, not to deform left-sided atrioventricular valve, especially in cases with septal malposition.³

In summary, a common atrioventricular junction should be suspected in patients with Down syndrome, an inlet ventricular septal defect, and a cleft mitral valve, even if the atrial septum is intact. Complete cleft closure with maintaining left-sided atrioventricular valve geometry would prevent postoperative regurgitation.

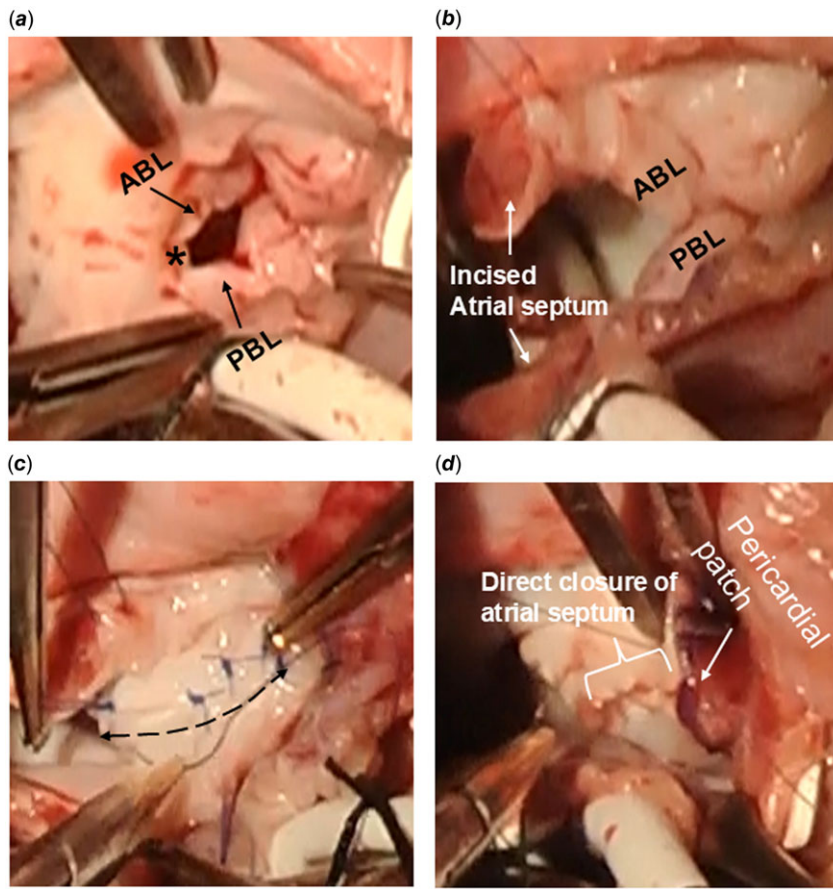


Figure 2. Intraoperative findings. **(a)** Both superior bridging leaflet (SBL) and inferior bridging leaflet (IBL) attached to the underside of interatrial septum and connected by fibrous tissue (asterisk). **(b)** After incision and retraction of interatrial septum, common atrioventricular junction could be visualized. **(c)** Cleft of left-sided atrioventricular valve (dashed line) was completely closed. **(d)** Direct closure of incised atrial septum and pericardial patch augmentation of underside of septum primum.

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Competing interests. None declared.

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