Interventricular Septal Hematoma Following Correction of Ebstein Anomaly

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INTRODUCTION

Interventricular septal hematoma (IVSH) is defined as hemorrhagic dissection between the muscle bundles of the interventricular septum (IVS), which occurs in adults after myocardial infarction, cardiac trauma, or various surgical procedures.[1,2] It is an extremely rare complication of congenital heart surgery, and cases reported are usually associated with patch closure of ventricular septal defect (VSD).[3] Here we present a case of IVSH following correction of Ebstein anomaly (EA).

CASE PRESENTATION

A 34-year-old woman was admitted to the hospital with a diagnosis of EA on routine prenatal examination more than 1 year ago. A review of symptoms was negative for palpitations or shortness of breath at rest and slight limitation of physical activity. A systolic blowing murmur was heard in the tricuspid region. The patient had blood pressure of 133/83 mm Hg, heart rate of 109 beats per minute, body surface area of 1.5 m^2, and 100% arterial O_2 saturation by oximetry. Preoperative transthoracic echocardiography (TTE) demonstrated a delaminated endocardium-like structure beside the true IVS, appearing like a double IVS (Figure 1A). The septal and posterior leaflets of the tricuspid valve (TV) were displaced apically from the true tricuspid annulus, both of which were attached to the delaminated endocardium-like structure. Displacement was assessed by measuring the distance between the septal leaflet of the TV and mitral hinge points. A displacement index of 8.1 mm/m^2 was recorded, supporting the diagnosis of EA. Restricted motion of septal and posterior leaflets as well as a coaptation gap was observed, resulting in severe tricuspid regurgitation. The anterior leaflet was large and sail-like without significant restriction of motion. Right atrium enlargement was seen, and right ventricular (RV) systolic pressure was estimated at 30 mm Hg. A tunnel-like color-flow Doppler pattern was seen crossing the interatrial septum, suggesting the presence of a patent foramen ovale. The myocardium was compacted with normal left ventricular (LV) systolic function (LV ejection fraction [EF] 60%). Cardiovascular magnetic resonance (CMR) was done to further evaluate the cardiac structure and function. It also revealed an additional delaminated endocardium-like structure with septal and posterior tricuspid leaflets anchored to it (Figure 1B), which was consistent with the TTE findings. The functional RV (end-diastolic volume [EDV] 113 mL, end-systolic volume [ESV] 48 mL, stroke volume [SV] 65 mL, EF 58%) was larger than the atrialized RV (EDV 97 mL, ESV 55 ml, SV 47 mL, EF 44%), consistent with Carpentier's type A. Normal LV EF was also recorded (EDV 90 mL, ESV 34 mL, SV 56 mL, EF 62%).

Due to the symptoms of severe tricuspid regurgitation and impaired RV systolic function, tricuspid valvuloplasty was recommended.[4] Cone procedure was used as this patient was classified as Carpentier's type A with large and mobile anterior leaflet. The operation was performed utilizing cardiopulmonary bypass and via aortobicaval cannulation. After right atriotomy, the TV was inspected. The septal and posterior leaflets of the TV were attached to a delaminated endocardium tissue, which was parallel to the IVS (Figure 2A). The atrial septum was intact. After plication of the atrialized RV in a continuous manner, the delaminated endocardium was anchored to the true tricuspid annulus, and thus a dead space was formed between the delaminated endocardium and pervious IVS. Out of concern that bleeding from the coronary venule would accumulate in this space, a 3 mm opening was left on the right atrial side for decompression (Figure 2B), serving as an outlet for the blood. Artificial tendinous cords were also introduced to shorten the septal and posterior leaflets. Saline test revealed satisfactory coaptation of the tricuspid leaflets without obvious regurgitation. Intraoperative transesophageal echocardiography (TEE) did not reveal the presence of IVSH. The patient was admitted to the intensive care unit in stable condition after surgery and transferred to the general ward on postoperative day (POD) 1.

Transthoracic echocardiography on POD 4 demonstrated a hypoechoic mass measuring 50 mm × 42 mm in the IVS (Figure 3A, Video 1). The mass was ovoid with homogenous echogenic signal and was surrounded by echo-bright IVS. Due to the compression effect of the mass, right-to-left septal shift can be seen, which was marked during diastole since intracavity pressure increased within the LV during systole and thus coped with the compression effect. The septal shift starting from early diastole had a significant impact on LV filling, resulting in underfilled LV (EDV 16 mL) while EF was preserved. Blood flow in the LV outflow tract was slightly accelerated (Vmax 1.67 m/sec) also due to the compression effect of the mass. Ultrasound-enhancing agents (UEAs) were used to further characterize the mass. Myocardial contrast echocardiography revealed that there were sparse microbubbles in the hypoechoic area after several cardiac cycles, which suggested the formation of IVSH (Figure 3B, Video 2). No communication between the hematoma and cardiac cavity was detected. In addition, there was no communication between the cavity and the pericardial space. Considering the patient was hemodynamically stable, conservative treatment was carried out under intensive supervision. Follow-up TTE on PODs 7 and 9 showed no progress of the hematoma. The patient was asymptomatic and hemodynamically stable and therefore was discharged on POD 11.

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VIDEO HIGHLIGHTS

Video 1: Echocardiography on POD 4. Two-dimensional TTE, parasternal short-axis view, demonstrates a hypoechoic mass in the IVS.

Video 2: Myocardial contrast echocardiography on POD 4. Myocardial contrast echocardiography, parasternal short-axis view, demonstrates sparse microbubbles in the hypoechoic area after several cardiac cycles. No communication between the hematoma and cardiac cavity was detected. In addition, there was no communication between the cavity and the pericardial space. Flattening of the IVS (mostly in diastole) was observed due to the compression effect of the mass.

Video 3: Follow-up TTE of the IVSH. Two-dimensional TTE, parasternal short-axis view, 6 weeks postoperatively, demonstrates significant resolution of the IVSH. Normal biventricular systolic function was appreciated.

Video 4: Follow-up TTE of the IVSH. Two-dimensional TTE, parasternal short-axis view, 6 months postoperatively, demonstrates complete resolution of the IVSH.

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Follow-up TTE 6 weeks postoperatively demonstrated a smaller size of the IVSH (Figure 4A, Video 3). At the follow-up 6 months after surgery, the patient was clinically stable with significant resolution of the IVSH (Figure 4B, Video 4).

DISCUSSION

Interventricular septal hematoma is overall a rare complication and is more commonly associated with myocardial infarction or cardiac trauma in adult cardiac patients.[1] When related to congenital heart surgery, the reported cases are usually associated with repair of perimembranous VSD.[3] There is currently no consensus on the etiology of IVSH. The leading theory is that injury of the septal perforating artery contributes to the development of IVSH.[5] This vessel is at risk during suturing since it is close to the anterior-superior margin of the VSD. The use of anticoagulation therapy and cardiac compression during operation may also contribute to the hematoma formation.[6,7] Significantly elevated preoperative RV pressure and compromised RV myocardial perfusion may also serve as contributing factors.[8] The limited literature on postoperative IVSH is mostly related to VSD repair, and the IVSH was always detected by TTE or TEE soon after the surgery, which can be explained by the septal perforating artery injury.[3,5,9] The theory does not hold true in our patient since there was no VSD closure and intraoperative TEE did not reveal any hematoma. In our patient, a cavity was formed after anchoring the delaminated endocardium to the true tricuspid annulus. To prevent IVSH formation, a 3 mm opening was left on the right atrial side after suturing for decompression. We guessed that bleeding from the coronary venules on the IVS surface exceeded the decompression effect of the opening, resulting in accumulated blood within the cavity, forming a hematoma.

Complications of IVSH include lethal arrhythmia, ventricular outflow obstruction, and cardiac tamponade. Hemodynamic stability is the decisive factor for the management of IVSH.[1,6] In most of the reported cases, IVSH spontaneously resolved without further intervention.[3,5,9] Treatment options include incisional drainage or needle aspiration of the hematoma. Extracorporeal membranous oxygenation has recently been described as a potential successful management option for patients in unstable condition until the hematoma resolves.[10] Our patient was hemodynamically stable and therefore was treated conservatively with intensive monitoring. Transthoracic echocardiography demonstrated complete resolution of the IVSH 6 months after the surgery.

Transthoracic echocardiography is key in the diagnosis and follow-up of IVSH.[3,5,9] The characteristic feature is the presence of a hypoechoic or anechoic area within the thick and echo-bright IVS. The acoustic characteristics of hematoma change over time, appearing as anechoic during active bleeding and hypoechoic when thrombosis is formed. Differential diagnoses include intracavity thrombi or ventricular trabeculations. Preoperative and postoperative image comparisons help rule out the likelihood of ventricular trabeculations. An intracavitary thrombi may have well-defined margins that are distinct from the endocardium, while IVSH is a hypoechoic or anechoic area with an overlying endocardial layer. Transthoracic echocardiography

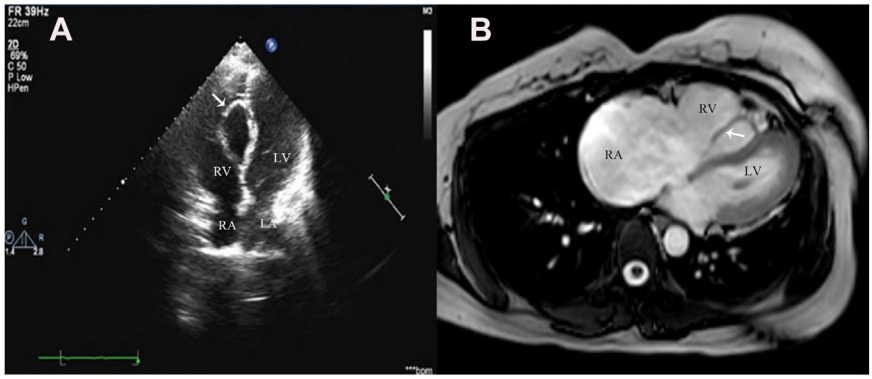

Figure 1 Preoperative TTE and CMR. **(A)** Two-dimensional TTE, apical 4-chamber view, end-diastole phase, demonstrates a delaminated endocardium-like structure (*arrow*) beside the true IVS. **(B)** Cardiovascular magnetic resonance, cine sequence, apical 4-chamber display, end-systole phase, demonstrates an additional delaminated endocardium-like structure (*arrow*). *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

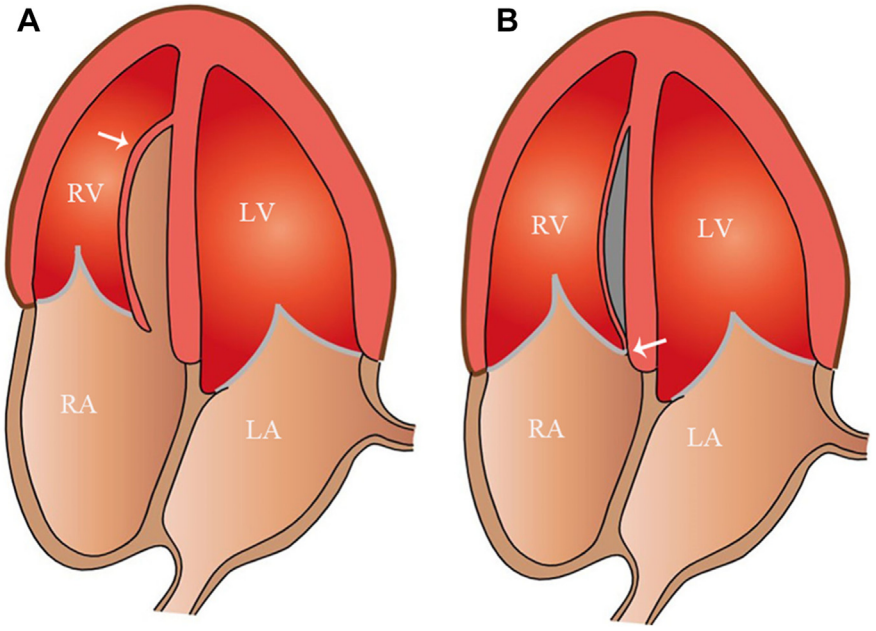

Figure 2 Schematic illustration of the operation. A delaminated endocardium was seen in the surgery (**A**, *arrow*), which was then anchored to the true tricuspid annulus with a 3 mm hole (**B**, *arrow*) that remained on the right atrial side. *LA*, Left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle.

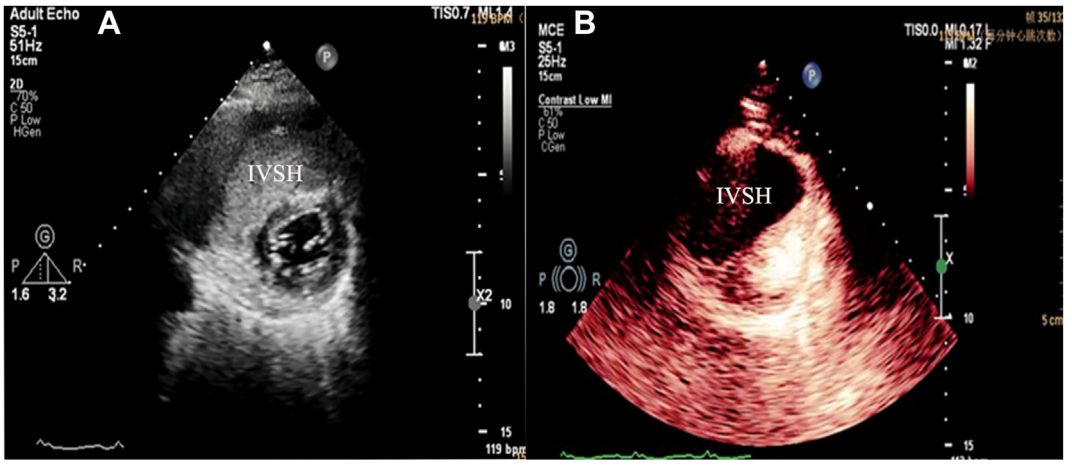

Figure 3 Transthoracic echocardiography on POD 4. **(A)** Two-dimensional TTE, parasternal short-axis view, systolic phase, demonstrates a hypoechoic mass in the IVS. **(B)** Myocardial contrast echocardiography, parasternal short-axis view, diastolic phase, revealed that there were sparse microbubbles in the hypoechoic area after several cardiac cycles. In addition, there was no communication between the cavity and the pericardial space. Flattening of the IVS was observed due to the compression effect of the mass.

can reveal the size, shape, and location of the hematoma and whether there is a fistula. In addition, it is bedside operable and noninvasive, allowing close follow-up to observe the progress of the hematoma and the patient's hemodynamic condition. The use of UEAs helps accurately delineate the endocardial boundary and emphasize the continuity of myocardial echogenicity. Contrast enhancement of the hematoma immediately second to the cardiac chamber demonstrates the existence of the communication between the hematoma and cardiac cavity.[11] Degree of enhancement of the hematoma can also provide valuable information regarding the presence of active bleeding. In our patient, there were sparse microbubbles in the hypoechoic area after several cycles that indicated the absence of continued bleeding. Hence, the use of UEAs has significant value in diagnosis and follow-up of IVSH.

CONCLUSION

We present a case of IVSH following correction of EA. The hematoma was detected by TTE and further evaluated by using UEAs. Treatment includes conservative management or surgical drainage, depending

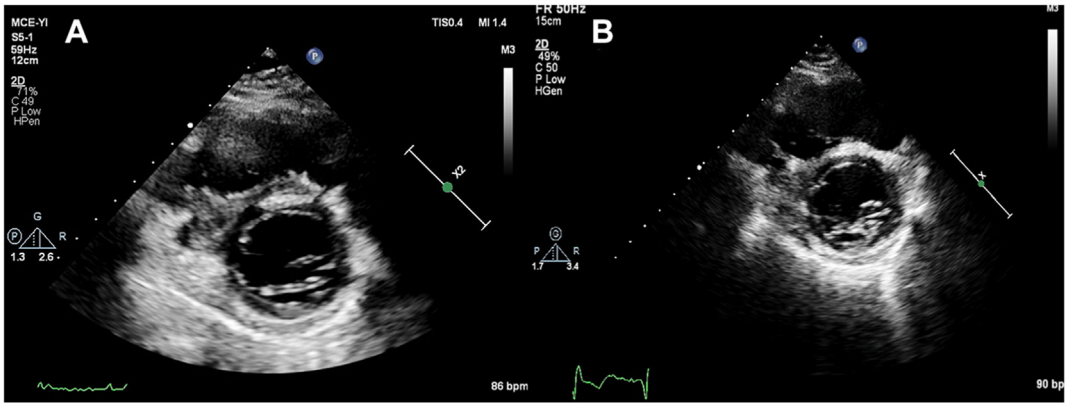

Figure 4 Follow-up TTE of the IVSH. Two-dimensional TTE, parasternal short-axis view, end-diastolic phase, 6 weeks **(A)** and 6 months **(B)** postoperatively, revealed significant resolution of the IVSH.

on the hemodynamic condition. We highlight the value of echocardiography with UEAs in the diagnosis and follow-up of IVSH to accurately assess the location, size, shunt formation, and evolution of the hematoma.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

FUNDING STATEMENT

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.005.

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