Surgical Treatment of a Defect Recurring 22 Years after Closure of an Inferior Sinus Venosus Defect: A Case Report

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Background: Residual shunt after closure of an inferior sinus venosus defect (ISVD) is a rare complication with a high rate of reintervention.

Case Presentation: Here, we report a rare case of a recurrent defect identified 22 years after closure of an ISVD. The defect (25 × 10 mm) was located at the inferior vena cava-right atrial junction and was closed directly when the patient was 5 years of age. No residual shunt was detected and follow-up was discontinued at age 12 years. However, a residual atrial septal defect shunt was detected incidentally at age 27 years. During the second surgery, the lower end of the original defect was opened and then closed with an expanded polytetrafluoroethylene patch.

Conclusions: Because of the high rate of reintervention for residual shunt after ISVD closure, patch closure was selected as a better option to reduce tension at the inferior-posterior border. Patients with this profile should be followed closely, at least during childhood, including by echocardiography.

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Key words: adult congenital heart disease, atrial septal defect, inferior sinus venosus defect, reoperation

Background

Residual shunt after closure of an inferior sinus venosus defect (ISVD) is a rare complication with a high rate of reintervention (approximately 10%)[1]. We present a case of a recurrent defect diagnosed and repaired 22 years after ISVD closure surgery during childhood.

Case Presentation

Written informed consent for publication of case details and images was obtained from the patient before drafting of the manuscript.

A 27-year-old Japanese woman with a surgical history of ISVD closure was referred to our hospital for a medical assessment required for scuba-diving certification. The examination revealed a coincidental finding of an 11.5-mm atrial septal defect (ASD) at the inferior vena cava-right atrial junction.

The patient had undergone surgery for ISVD at age 5 years, when she weighed 15 kg. The preoperative diagnosis at that time was secundum-type ASD. During surgery, the size of the defect was 25 × 10 mm, and no lower rim was present near the inferior vena cava (IVC). A patent foramen ovale was also observed. The defect near the IVC was closed directly by over-and-over stiches in two layers with 5-0 Prolene sutures. The patent foramen ovale was also closed directly. Ostium of the right lower pulmonary vein was not detected in the right atrium. Her postoperative course was uneventful. Transthoracic echocardiography showed no residual shunt at age 12 years, when follow-up was discontinued.

At age 27 years, a transthoracic echocardiogram during the abovementioned medical assessment revealed a recurrent defect measuring 11.5 mm, with mild-to-moderate tricuspid valve regurgitation, a left ventricular end-

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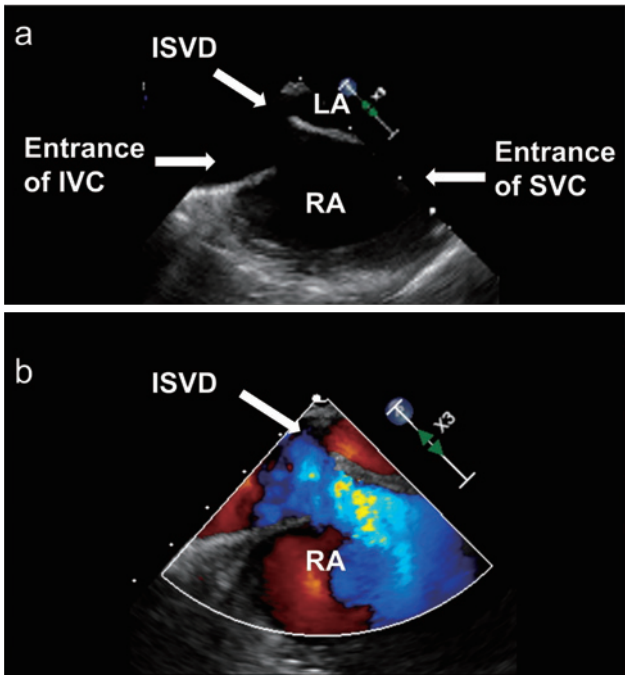

Fig. 1 **a:** Plain echocardiogram, **b:** Color Doppler transesophageal echocardiogram at the level of the atrial septum revealed the recurrent defect near the entrance of IVC. No residual atrial septum is seen inferiorly. SVC: superior vena cava, IVC: inferior vena cava, ISVD: inferior sinus venosus defect, LA: left atrium, RA: right atrium.

diastolic diameter of 41.2 mm, and a left ventricular end-systolic diameter of 26.1 mm. Transesophageal echocardiography revealed a defect near the entrance of the IVC (**Fig. 1**). Cardiac catheterization showed a pulmonary-to-systemic blood flow ratio of 2.7, pulmonary vascular resistance of 0.4 units/m^2, left ventricular end-diastolic volume that was 86.5% of normal, right ventricular end-diastolic volume that was 172% of normal, and arterial oxygen saturation in the ascending aorta of 98.6%. Magnetic resonance imaging showed a pulmonary-to-systemic blood flow ratio of 1.7 and all four pulmonary veins returning to the left atrium.

Under general anesthesia, a re-median sternotomy was performed with the patient in supine position. Cardiopulmonary bypass was performed with ascending aortic perfusion and two venous drainage routes. The 10-mm residual defect was detected between the lower rim of the defect that was closed in the previous surgery and the mouth of the IVC. The defect extended to the inferoposterior border of the left atrium, without residual atrial septal tissue at the inferior margin (**Fig. 2**). The defect was closed with a 0.4-mm polytetrafluoroethylene patch using interrupted sutures at the inferior margin and continuous sutures at the anterior, superior, and posterior margins. The IVC cannula was temporarily removed to ensure clear visualization of the inferoposterior margin.

The patient was discharged, without complications, at 4 days after the operation. At 3 years after the operation, the patient remains asymptomatic, with a peripheral oxygen saturation of 98% in room air. Transthoracic echocardiography at the same time showed a left ventricular end-diastolic diameter of 44.4 mm, left ventricular end-systolic diameter of 27.9 mm, no residual shunt, and no IVC stenosis.

Discussion

A sinus venosus defect, first described by Ross in 1956[2], is an incomplete absorption of the sinus venosus into the right atrium and not a true ASD. ISVD is a rare anomaly in which interatrial communication extends into the entrance of the IVC, near the confines of the atrial septum[3]. An anatomical characteristic of ISVD is that the defect originates in the entrance of IVC and then extends directly into the inferoposterior border of the left atrium; there is no residual atrial septal tissue at the inferior margin[1,4].

One report noted that only 36% of patients with ISVD receive a correct diagnosis preoperatively[1], that patients with an incorrect ISVD diagnosis have poor technical outcomes, and that the frequency of reintervention is high (15%). With correct preoperative diagnosis of ISVD, the IVC can be cannulated more caudally without placement of caval occluding tape, thereby further improving the visualization of the proper inferior border of the defect.

The preoperative diagnosis for our patient was secundum-type ASD, and the ISVD was closed directly. Snarr et al. demonstrated that absence of the posterior rim in the parasternal short axis views is a consistent finding in patients with ISVD and distinguishes ISVD from a large secundum ASD with inferior extension. They reported a significant increase in the accuracy of ISVD diagnoses when this criterion was used[5].

To avoid poor visualization of the proper inferior border of the defect, we used drainage from the IVC using suction, without placing caval occluding tape. Another option is to use drainage from the IVC with cannulation at the femoral vein with a puncture and no caval occluding tape. Even for young patients, this is an acceptable technique that ensures safety and a good cosmetic outcome.

Residual shunts of ISVD are frequently detected at the entrance of the IVC. Banka et al. reported that a residual

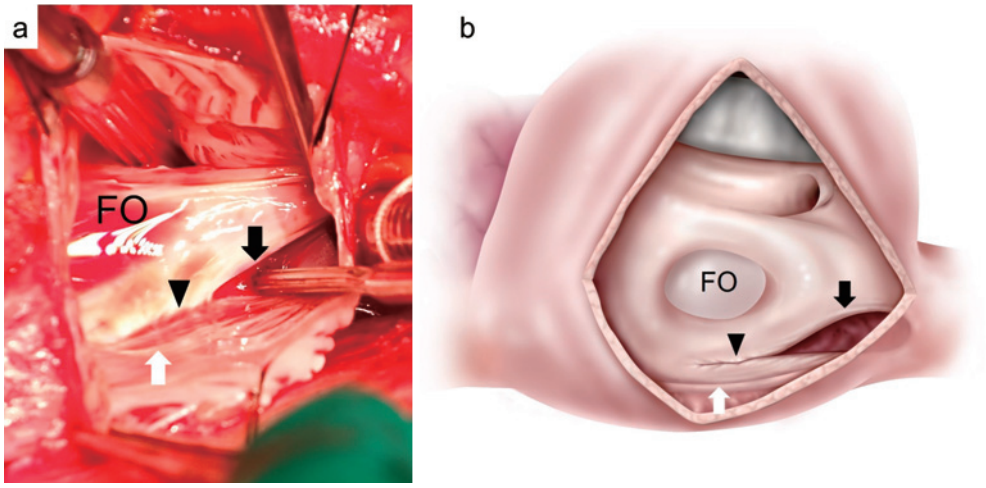

Fig. 2 **a:** Intraoperative image. **b:** Illustration. The residual shunt (10 mm; black arrow) was detected between the lower rim of the closed defect (black arrowhead) from the previous operation and the mouth of the IVC. White arrow: crista terminalis. FO: fossa ovale.

shunt of ISVD was present in five of 45 cases, and that three of those five cases had shunt flow near the entrance of the IVC[1]. Similarly, Fangan et al. reported that residual shunts detected 2-10 years after ISVD closure were at the entrance of the IVC[6]. There are two reasons for this finding. First, the ISVD lies outside the area of the true septum and thus lacks a complete muscular border at the inferoposterior rim[7]. Second, the ISVD, which has no inferior rim, is difficult to distinguish from an oval fossa defect that extends toward the inferior caval vein[7]. Poor visibility of this region may therefore have contributed to the incomplete repair, as in the cases reported by Fagan et al.[6].

With somatic growth, the atrial septum appears to grow asymmetrically, and Gossett et al. reported that growth of the IVC rim after transcatheter closure of an ASD was considerable[8]. In our patient, the portion closed in the previous operation was intact and, over a long period of time, a residual defect was created between the lower rim of the closed defect and the mouth of the IVC.

The cause of the recurrent defect may be related to the technique that was used to close the ISVD. The defect in our patient was closed directly in two layers. At the inferior-posterior border, the atrial septum tissue was anchored to the left atrium tissue. The residual defect arose in the inferoposterior border and may have been caused by severe tension during the patient's growth. Direct closure did not seem suitable for closure of the ISVD; patch closure appeared to be a better option to reduce the tension in the inferoposterior border. Other reports have described good outcomes after similar surgical repairs using a patch[1,4].

Conclusions

This case report described successful surgical treatment of a recurrent defect after ISVD closure 22 years earlier. Absence of tension in the inferoposterior border is needed for this surgical technique, and further improvement of the visualization of the proper inferior border of the defect is desirable. In light of the high rate of reintervention for residual shunt after ISVD closure, patients with this profile should be followed closely, at least during childhood, including by echocardiography.

Availability of data and materials: The datasets supporting the conclusions of this article are included in the article.

Authors' contributions: KS is the first and corresponding author of this manuscript. TS, KO, JA, and SS participated in the second operation of this case. TS, MW, and RF treated the patient before and after the second operation. YI supervised the second operation. TS and YW supervised the editing of the manuscript. KS drafted the manuscript, and all authors read and approved the final manuscript.

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References

1. Banka P, Bacha E, Powell AJ, Benavidez OJ, Geva T. Outcomes of inferior sinus venosus defect repair. J Thorac Cardiovasc Surg [Internet]. 2011 Sep;142(3):517–22. Avail-

able from: http://www.ncbi.nlm.nih.gov/pubmed/21334015
2. Ross DN. The sinus venosus type of atrial septal defect. Guys Hosp Rep [Internet]. 1956;105(4):376–81. Available from: http://www.ncbi.nlm.nih.gov/pubmed/13375954
3. Plymale J, Kolinski K, Frommelt P, Bartz P, Tweddell J, Earing MG. Inferior sinus venosus defects: anatomic features and echocardiographic correlates. Pediatr Cardiol [Internet]. 2013 Feb;34(2):322–6. Available from: http://www.ncbi.nlm.nih.gov/pubmed/22854830
4. Crystal MA, Al Najashi K, Williams WG, Redington AN, Anderson RH. Inferior sinus venosus defect: echocardiographic diagnosis and surgical approach. J Thorac Cardiovasc Surg [Internet]. 2009 Jun;137(6):1349–55. Available from: http://www.ncbi.nlm.nih.gov/pubmed/19464447
5. Snarr BS, Liu MY, Zuckerberg JC, et al. The parasternal short-axis view improves diagnostic accuracy for inferior sinus venosus type of atrial septal defects by transthoracic echocardiography. J Am Soc Echocardiogr [Internet]. 2017 Mar;30(3):209–15. Available from: http://www.ncbi.nlm.nih.gov/pubmed/28139440
6. Fagan S, Veinot JP, Chan KL. Residual sinus venosus atrial septal defect after surgical closure of atrial septal defect. J Am Soc Echocardiogr. 2001 Jul;14(7):738–41.
7. Naqvi N, McCarthy KP, Ho SY. Anatomy of the atrial septum and interatrial communications. J Thorac Dis [Internet]. 2018 Sep;10(Suppl 24):S2837–47. Available from: http://www.ncbi.nlm.nih.gov/pubmed/30305943
8. Gossett JG, Mansfield L, Acevedo J, Lay AS, Rychlik K, Wax DF. Growth of the atrial septum after Amplatzer device closure of atrial septal defects in young children. Catheter Cardiovasc Interv [Internet]. 2015 Nov 15;86(6):1041–7. Available from: http://www.ncbi.nlm.nih.gov/pubmed/26013563

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