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CASE REPORT

Surgery for Residual Inferior Left-to-Right Atrial Shunt

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ABSTRACT

We report the case of three female patients who were scheduled for surgical correction of residual left-to-right shunt after initial repair of sinus venosus atrial septal defect (SV-ASD) during childhood. After excluding the possibility of an hemodynamic intervention, all three patients underwent a successful surgical closure through a right mini sub-axillary approach by using total peripheral cannulation for cardiopulmonary bypass and leaving the inferior vena cava completely un-snared allowing for an optimal visualization of the residual atrial septal communication and avoiding extensive dissection of mediastinal structures.

KEYWORDS

Residual sinus venosus-ASD; un-snared vena cavae; minimally invasive cardiac surgery; inferior ASD

1 Introduction

Atrial septal defects (ASD) can be found in different sites of the interatrial septum depending on their embryological origin. Sinus venosus ASD (SV-ASD) are interatrial defects morphologically adjacent to the outlet of the superior (SVC) or inferior vena cava (IVC), and they are usually associated with the anomalous drainage of one or more pulmonary veins into the right atrium, the atrio-caval junction or the venae cavae [1]. Surgical closure of this type of defects is sometimes challenging due to a difficult surgical exposure (e.g., low extension of the defect into the IVC or restrict view of the IVC) or misidentification of contiguous structures (e.g., an enlarged Eustachian valve). For this reason, the need for reoperation for a residual shunt is not a rare event [2].

Over the past decades, surgical correction of less severe congenital heart disease (CHD), such as ASDs and partial anomalous pulmonary venous drainage (PAPVD), has been increasingly performed through minimally invasive less traumatic surgical approaches [3–5] and with the aid of remote cardiopulmonary bypass (CPB).

2 Cases Presentation

In the current study, we report the cases of three females (20, 32 and 14-year-old, respectively) who underwent surgical re-intervention for residual left-to-right shunt after initial surgical repair of SV-ASD in childhood.

Intraoperative records at the time of first surgery reported the presence of a single large inferior SV-ASD in one case (Patient 1), the association of a large ostium secundum ASD type with a separate SV-ASD in the second case (Patient 2) and the association of a superior SV-ASD with a PAPVD of the right superior pulmonary artery in the SVC in the last case (Patient 3). All procedures at that moment were performed with central CPB through aortic and bi-caval cannulation, two through an anterior right thoracotomy and the other through a mid-line sternotomy. The postoperative course was uneventful and all patients were discharged in good clinical conditions with no suspicion of residual intra-cardiac defects on 2D-echocardiography.

At follow-up, all patients were asymptomatic. However, a persistent dilatation of right heart chambers required a more accurate examination since this findings are compatible with a residual left-to-right shunt at the atrial level, which was found to be present and significant (Qp:Qs > 1.5) in all three patients. The first two patients showed a residual SV-ASD with extension into the orifice of the IVC, while patient 3 showed a PAPVD of a previously unknown accessory right inferior pulmonary vein into the IVC. All patients were scheduled for elective surgery since the possibility of a percutaneous approach was not deemed feasible in the first two patient, while in the third patient it was not even an option [6].

At the time of surgery, percutaneous ultrasound-guided cannulation of the internal jugular vein and surgical isolation and cannulation of the femoral vessels were used for establishing a remote normothermic CPB. We decided to perform the correction by using a sub-axillary right thoracotomy in all three of them, entering the chest in the fifth intercostal space. Right lung exclusion was achieved in all patients.

After minimal dissection of mediastinal structures to achieve the visualization of the lateral side of the right atrium, with the aid of induced ventricular fibrillation (IVF), a right atriotomy was performed without snaring the IVC, thus enabling a better visualization of the residual intra-cardiac shunt (Fig. 1, Panels A–A1).

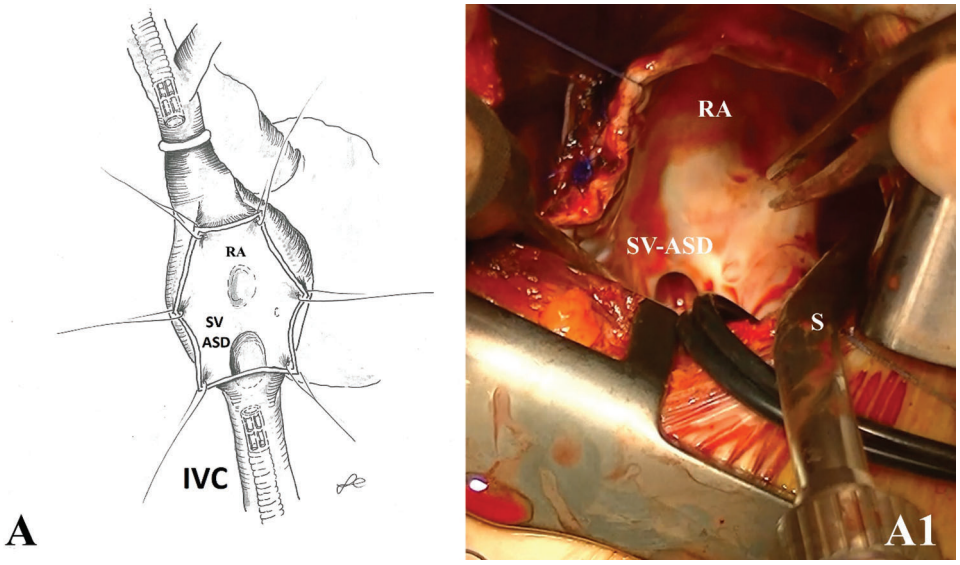

Figure 1: Graphic representations (A) and matched intraoperative view (A1) showing a right atriotomy used to identify and close a residual SV-ASD. Black cable: Fibrilator; S: Sucker of the coronary sinus; IVC: Inferior vena cava; RA: Right atrium; SV-ASD: Sinus venosus atrial septal defect

The residual SV-ASD was identified and closed by using a heterologous pericardial patch (Fig. 2, Panels B–B1) in the first two patient, while in the third patient the previously implanted patch was partially demolished and a new one was used to tunnel the anomalous pulmonary vein into the newly-created ASD.

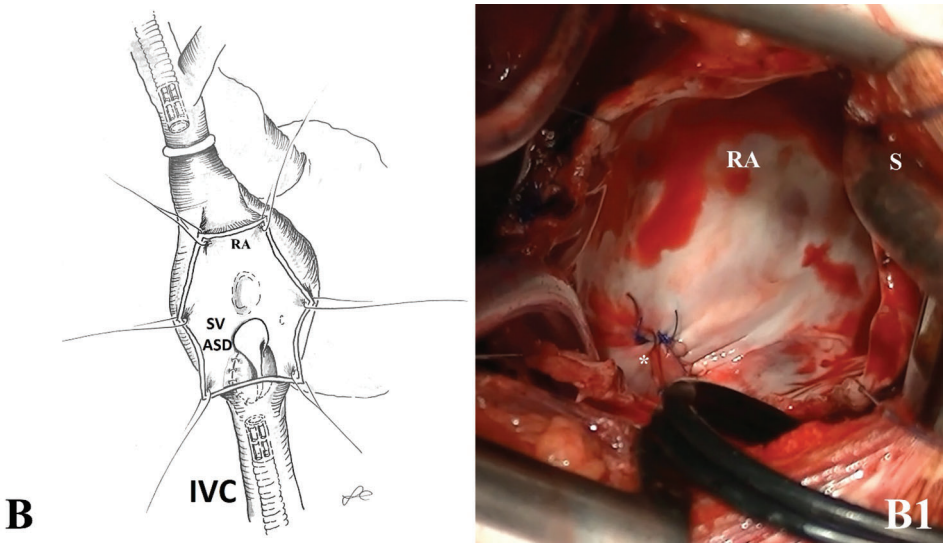

Figure 2: Graphic representations (B) and matched intraoperative view (B1) showing the surgical closure of the residual SV-ASD. *: Heterologous patch used for SV-ASD closure; Black cable: Fibrilator; S: Sucker of the coronary sinus; IVC: Inferior vena cava; RA: Right atrium; SV-ASD: Sinus venosus atrial septal defect

We avoided sucking into the left atrium during surgery and, before closing completely the SV-ASD, care was taken to carefully de-airing the left sided chamber under echo-guidance, to avoid air embolism. The ventricular fibrillation was converted to sinus rhythm by DC shock in the first two patient, while the third patient spontaneously returned to sinus rhythm. The complete repair of the shunts was confirmed by 2D-trans-esophageal echocardiography and a bubble test. The postoperative course was uneventful, and patients were discharged home on postoperative day 7 (Patient 1), 6 (Patient 2) and 5 (Patient 3).

3 Discussion

Sinus venosus-ASD represent the 4–11% of all ASDs [1] and their closure can lead to residual ASD, due to their inferior location [2]. Similarly, associated defects such as inferior PAPVD into the IVC can be overlooked during an operation due to insufficient surgical exposure of the field and the bicaval cannulation and snaring necessary for standard central CPB.

In all three of our patients, an elective surgical correction was deemed necessary due to the significance of the left-to-right shunt and to the unfeasibility of a percutaneous closure for the anatomical location and nature of the defects. In order to avoid re-entry into the chest from the previous surgical accesses we decided to utilize a small sub-axillary incision. This was indeed a second incision of the patient's chest, but it actually allowed us to easily and safely reach the lateral surface of the right atrium and its junction with the IVC, thus avoiding an extensive dissection of mediastinal structures. The choice of using the combination of peripheral cannulation for remote CPB, induced fibrillatory arrest for intracardiac repair and unsnaring of IVC were also chosen to further limit the mediastinal dissection, since we did not need to isolate the ascending aorta and venae cavae (all structures that could have been reached in case of need).

The intra-cardiac repair without snaring of the IVC has already been described in the literature for the treatment of CHD with potentially complicated venous drainage and is mainly performed for extra-cardiac

Fontan procedures, demonstrating great feasibility with adequate venous drainage and no reduction in flow or circulatory arrest [7]. In our cases, the tip of the femoral vein cannula was held a couple of centimeters below the junction of the right atrium with the IVC, allowing us to open the right atrium without the blood flow coming from the inferior part of the body and without the risk of air-trap on the CPB circuit.

This was essential for having a complete visualization and closure of the defect, which in the first two cases extended deeply into the IVC. In fact, IVC snaring can sometimes cause potential edge distortion of an SV-ASD and this may have been the cause of these residual defects.

The third case was somehow different, insofar the SV-ASD had been properly closed in the first intervention but an anomalous accessory pulmonary vein was overlooked and was still draining at the junction between RA and IVC. Again, the bicaval cannulation probably hide the defect from the surgeons' view and was responsible for the residual shunt. Unsnaring was mandatory in this case to have proper access to the IVC and to tunnel the anomalous pulmonary vein into a re-created ASD.

An accurate diagnostic assessment is certainly fundamental both pre- and postoperatively. Our pre-operative assessment usually relies on transthoracic echocardiography (TTE), but when an unusual anatomical location of the effects if diagnosed, a transesophageal echocardiography (TEE) evaluation is always considered. At the time of surgery, the intraoperative assessment of the ASD location and dimensions is always performed by using TEE, as well as the postoperative assessment of its closure. Any residual intracardiac shunt is also ruled out with a standard bubble test performed by injecting shaken saline solution thorough a peripheral vein of the upper part of the body. In selected cases, especially in patients with ASD extended into the IVC, the bubble test is performed by injecting shaken saline solution thorough a peripheral vein of the lower part of the body (usually the femoral vein at the cannulation site).

4 Conclusion

We believe that the combination of both peripheral cannulation and un-snaring of the IVC can greatly enhance the surgical view of ASD, and with SV ASD it may even improve the surgical outcome, since it allows for a proper closure of defect extending deeply into the IVC. In our cases the standard cannulation of both venae cavae performed during the first intervention was likely the cause of the residual defects results obtained, as the suboptimal surgical exposure of the field hide some of the morphological features.

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