Case Report

Closure of Sinus Venosus Atrial Septal Defects with Transcatheter Balloon-Expandable Stents: A Single-Center Case Series

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ABSTRACT

Background: Sinus venosus atrial septal defect (SVASD) is a congenital heart defect involving an opening between the superior vena cava (SVC) and the right upper pulmonary vein (RUPV). Surgical closure is the standard treatment. Nonetheless, it is more complex than secundum ASD reconstruction and is associated with higher complication rates. Transcatheter closure is an emerging alternative.

Methods: We report 5 cases of transcatheter SVASD closure and 1 case of failure requiring surgical intervention. All patients underwent preprocedural computed tomography angiography to assess defect size and location and optimal stent dimensions. SVC stenting was performed using balloon-expandable stents, followed by RUPV angioplasty if necessary.

Results: Final angiograms and pressure measurements in the SVC, RUPV, and right atrium confirmed the absence of residual shunt and pulmonary venous obstruction. Stent migration to the pulmonary artery occurred in 1 patient, necessitating surgical retrieval and defect closure.

Conclusions: Balloon expansion testing is not mandatory before stent implantation. If RUPV obstruction occurs, flow can be reestablished via ballooning or stent implantation within the RUPV, and associated mild residual shunts may resolve spontaneously.

Keywords: Sinus Venosus Atrial Septal Defect; Sinus Venosus Atrial Septal Defect (SVASD); Case Series; Partial Anomalous Pulmonary Venous Connection; Transcatheter Closure

Introduction

Transcatheter interventions for congenital heart defects have evolved since the first transcatheter closure of an atrial septal defect (ASD) in a dog in 1972.[1]

Significant progress has since been made in these interventions.[2] Sinus venosus atrial septal defect (SVASD), first described by Peacock[3] in 1858, represents 5% to 10% of ASDs. This defect may manifest with a wide spectrum of symptoms depending on characteristics such as size and shunt volume.[4]

Surgical repair of SVASD is more complex than secundum ASD repair. Nearby structures, including the sinus node, are susceptible to injury during surgical repair.[5,6] Abdullah et al[5] described a technique to replace the deficient posterior wall of the ASD and redirect flow from the right pulmonary vein (RPV) to the left atrium (LA) via transcatheter implantation of balloon-expandable stents. Since the introduction of this method, several case reports and small series have been published.[7,8] Most studies have employed this technique in adult patients.[9,10]

Prior studies have suggested a balloon expansion test to evaluate whether the stent could obstruct flow from the right upper pulmonary vein (RUPV) to the LA, with the procedure aborted if such a finding was evident.[9,10]

We describe employing this technique to close these defects in a pediatric cardiac catheterization laboratory without a balloon expansion test.

Methods

From January through March 2023, 6 patients aged 12 to 20 years with large SVASDs and anomalous RUPV drainage underwent transcatheter defect closure. Informed consent was obtained from all patients or legal guardians, and the novelty of the procedure was explained. Preprocedural evaluation—including clinical examination, blood investigations, chest radiography (CXR), electrocardiography (ECG), and 2D transthoracic echocardiography (TTE)—was performed to determine defect anatomy.

Previous studies have advised using a balloon occlusion test during angiography to specify RUPV drainage, with patient selection for stent closure occurring after this test.[5,9,10] In the present study, cases were selected based on computed tomography angiography (CTA). RUPV stenosis and proximity of the RUPV junction to the superior vena cava (SVC) were considered limiting factors for stenting. Patients were ineligible if the RUPV-SVC junction was superior to the azygos vein entry point into the SVC, as the implanted stent could obstruct azygos flow. For a high partial anomalous pulmonary venous connection (PAPVC), implantation of a bare stent within the upper SVC and a covered stent within the lower SVC was planned to improve stability for both stents.

The procedure was performed under general anesthesia; patients received intravenous cefazolin (30 mg/kg) preoperatively. Vascular access was obtained via the right internal jugular vein or the left or right femoral vein. An arterial catheter was introduced into the left femoral artery for blood pressure monitoring. A 5-Fr short valved sheath was introduced via femoral venous access. Heparin (100 IU/kg) was administered intravenously after sheath placement. With the aid of a 5-Fr A2 multipurpose catheter, a pulmonary artery angiogram was performed to confirm contrast return to the right atrium (RA) through the RUPV during the levo phase. The catheter was parked in the RUPV for serial venography and pressure monitoring. If the RUPV was occluded or pressure was elevated, RUPV balloon angioplasty was performed to establish patency.

A super-stiff wire was passed through the multipurpose catheter and placed in the SVC via the right internal jugular vein to create a rail. Unobstructed RUPV and right inferior pulmonary vein flow into the LA were confirmed before closure. Stent length was selected based on the distance between the upper border of the RUPV-SVC connection and the inferior border of the SVASD. A balloon 4 mm larger than the SVC diameter was selected. Contrast TTE was performed after stent placement to confirm RUPV drainage to the LA without obstruction or residual shunt. CXR and 12-lead ECG were obtained 2 hours after stent implantation.

Case 1

CTA and angiography revealed that the RUPV adjoined the SVC-RA junction. SVC stenting was successfully performed using a Bentley covered stent (58 × 16 mm) and flared using 22 × 50–mm and 25 × 40–mm balloons. Final injection showed patent SVC draining into the RA, while the RUPV demonstrated a mild residual shunt to the RA (Figure 1). TEE confirmed these results. At the 3-month follow-up visit, CTA and TTE showed that the residual shunt had resolved.

Case 2

In patient 2, the RUPV and right lower pulmonary vein connected to the SVC-RA junction. SVC injection showed significant tapering and RUPV narrowing. A left SVC adjoining the coronary sinus was evident on CTA and angiography. SVC stenting was performed using two 12 × 59–mm Bentley stents inflated with 3 balloons (24 × 40 mm, 25 × 40 mm, and 37 × 20 mm) to control type 1 endoleak and residual shunt from the RUPV to the RA (Figure 2). After the dilation of both stents, RUPV origin angioplasty was performed using a 9 × 20–mm balloon; an additional RUPV stent was unnecessary. Final angiograms and pressure measurements in the RUPV and the RA showed a mild residual shunt from the RUPV to the RA without pulmonary venous compression or occlusion. CTA and TTE at 3-month follow-up showed that the residual shunt had resolved.

Case 3

In patient 3, CTA and selective RUPV angiography demonstrated a PAPVC to the superior SVC-RA junction and a secundum ASD. An A2 catheter was inserted into the RUPV. An 18 × 48–mm Bentley covered stent was implanted in the lower SVC segment from the azygos connection to the mid-RA. Stent flaring was performed with a 24 × 30–mm Alto's balloon, and RUPV injection exhibited no residual shunt. The secundum ASD was then closed using a 16-mm Amplatzer Cera Lifetech device (Figure 3). At 3-month follow-up, TTE and CTA showed no residual shunt.

Case 4

Anatomic evaluation of patient 4 demonstrated RUPV and right middle pulmonary vein (RMPV) connections to the SVC. One 57-mm bare Optimus stent and two 57-mm covered Optimus stents were successfully placed within the SVC using a 30 × 50–mm VASC II balloon and a 24 × 50–mm BIB catheter. After flaring, right pulmonary artery dye injection and TTE demonstrated adequate flow of the RUPV and the RMPV to the LA with a mild residual shunt to the RA (Figure 4). CTA and TTE at 3-month follow-up showed no residual shunt.

Case 5

In patient 5, the RUPV and the RMPV drained into the SVC. Similar to CASE 2, this patient had a left SVC draining into the coronary sinus. Two 57-mm Optimus covered stents were introduced via 24 × 50–mm BIB and 30 × 50–mm VASC II balloons and flared (Figure 5). The procedure was performed with a mild residual shunt demonstrated by RPV angiography and postprocedural TTE.

Case 6

Evaluations showed RUPV and RMPV connections to the SVC. One 57-mm Optimus stent was flared using a 30 × 50–mm VASC II balloon. Final angiography and TTE revealed no shunt or obstruction (Figure 6). Nevertheless, CXR 2 hours postprocedurally showed stent dislocation and embolization to the main pulmonary artery. The stent was extracted via emergent surgery, and the defect was corrected surgically. Retrospective reevaluation showed that embolization had occurred because of the short stent length. High RUPV insertion into the SVC mandated a short-covered stent to avoid the occlusion of the innominate or azygos veins. Theoretically, installing a bare stent in the upper SVC before covered stent implantation might improve stability by elongating the stent.

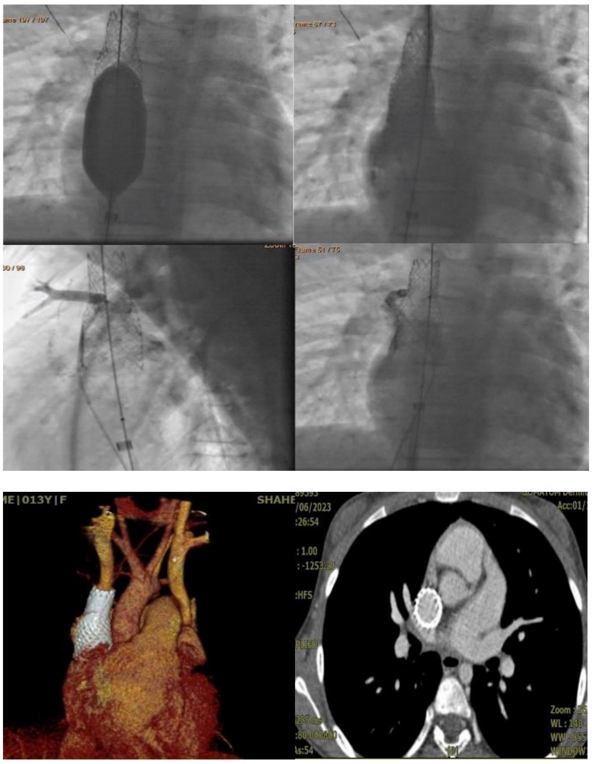

Figure 1. Covered stent in the superior vena cava crossing the defect to form a secondary conduit for right upper pulmonary vein drainage into the left atrium. Follow-up computed tomography angiography shows stent position and patency. No postprocedural narrowing was observed from the right upper pulmonary vein to the right atrium.

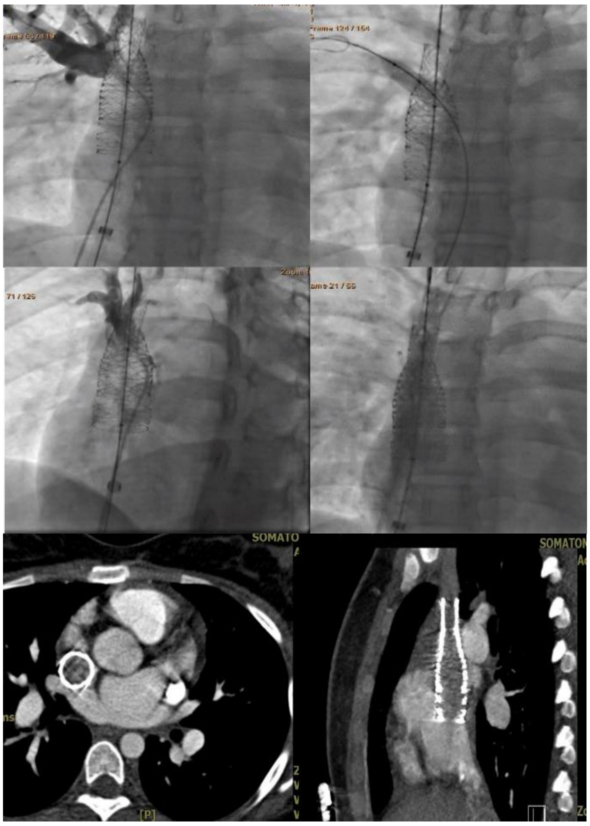

Figure 2. Angiography shows 2 stents in the superior vena cava used to resolve right upper pulmonary vein narrowing via balloon angioplasty. The right upper pulmonary vein pathway remains patent. Follow-up computed tomography angiography confirms right upper pulmonary vein patency without stenosis.

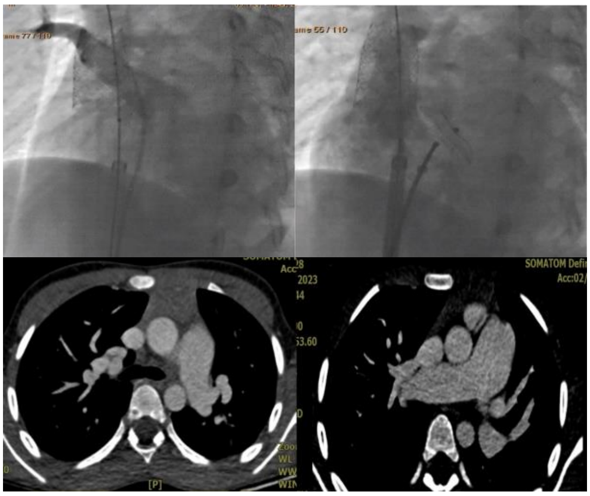

Figure 3. After superior vena cava stent placement and patency confirmation, the secundum atrial septal defect was simultaneously closed. Computed tomography angiography demonstrates a connection between the right upper pulmonary vein and the right middle pulmonary vein, which remained patent after stent placement. The secundum atrial septal defect was closed with a device, and no leakage from either abnormality was observed during follow-up.

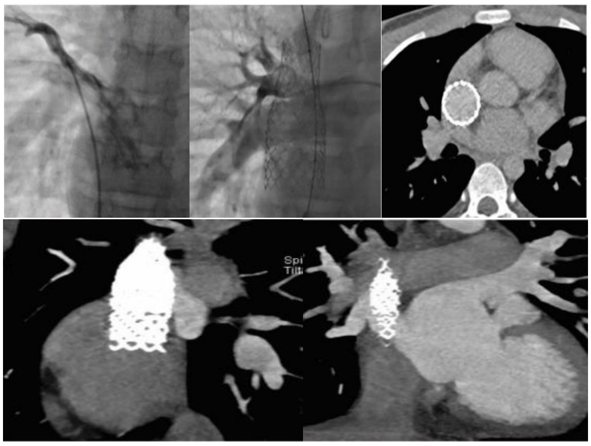

Figure 4. Anomalous right upper pulmonary vein drainage into the right atrium. After superior vena cava stent insertion, right upper pulmonary vein drainage reassessment showed no residual flow into the right atrium. Follow-up computed tomography angiography confirmed successful superior vena cava stent placement and patent right upper and right middle pulmonary vein draining into the left atrium.

Figure 5. Superior vena cava stent placement to redirect right upper and right middle pulmonary vein drainage into the left atrium. After placement, the right upper and right middle pulmonary veins were patent with no residual flow into the right atrium. Follow-up computed tomography angiography shows the superior vena cava stent position and a patent right upper pulmonary vein lumen.

Figure 6. Angiographic evaluation shows right upper and right middle pulmonary vein drainage into the right atrium. Postprocedural angiography after superior vena cava stent placement demonstrated a patent pulmonary vein without drainage into the right atrium. Follow-up chest radiogram obtained 2 hours postprocedurally depicts stent dislocation into the main pulmonary artery. Based on preprocedural computed tomography angiography and the clinical course, 2 stents may enhance stability in a long, narrow superior vena cava to prevent complications.

Results

Patient ages ranged from 12 to 20 years (Table 1). Stenting of the SVASD was successful in all patients, all of whom had symptoms consistent with New York Heart Association functional class 2 or 3 at admission.

No patient experienced procedural complications, and all stents were successfully deployed. In Case 1, postprocedural angiography revealed a mild residual shunt without pulmonary vein compression.

In Case 2, selective injection before stent flaring showed obvious RUPV narrowing. The patient underwent successful balloon angioplasty of the RUPV, after which a mild shunt remained.

Case 3 underwent stenting of a PAPVC RUPV to the high SVC-RA junction and concurrent repair of the secundum ASD with an Amplatzer device. No complications were evident.

A mild residual shunt remained in both Case 4 and Case 5. Delayed stent embolization occurred in Case 6, necessitating emergent surgical extraction. After the procedure, a heparin infusion at 10 IU/kg per hour was administered for 2 days, and dual antiplatelet therapy (aspirin [100 mg] and clopidogrel [75 mg]) was initiated, with a recommended duration of 6 months for aspirin and 1 month for clopidogrel.

Table 1. Characteristics and outcomes of the studied patients

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age, y	13	15	12	14	14	20
Sex	Female	Female	Male	Male	Female	Male
Weight	45 kg	50 kg	40 kg	70 kg	78 kg	64 kg
Qp/Qs ratio (Qp:Qs)	2.8:1	2.5:1	2.5:1	1.6:1	2/1	1.8/1
Residual Shunt	Mild	Mild	None	Mild	Mild	None
Complications	None	None	None	None	None	Stent embolization
Concomitant Structural Disorder	None	Bilateral SVC	ASD2	None	Bilateral SVC	None

SVC: superior vena cava; ASD2: secundum atrial septal defect

Follow-up

In the first case, a mild residual shunt from the RUPV to the RA was still noted on TTE 2 weeks after the procedure. During 3 months of follow-up, the shunt resolved, and no residual shunt was noted on the most recent echocardiography. Aside from the residual shunt, no other complications occurred.

The second and third cases showed no pathologic findings on postprocedural or 3-month follow-up TTE.

The 3-month follow-up TTE and CTA of the fifth patient showed a patent PV without any shunt.

We anticipated that the shunt would resolve after stent endothelialization and that volume unloading would lead to a reduction in right atrial size, atrial remodeling, and SVC insertion. A qualitative reduction in right ventricular size was noted on postprocedural echocardiography, and CTA confirmed a significant reduction in right ventricular volume and appropriate device position 1 month after the procedure.

Sinus node dysfunction is a potential concern in the surgical approach and remains a consideration after SVASD correction with balloon-expandable stents when a stent is implanted at the SVC-RA junction. Accordingly, all patients underwent ECG Holter monitoring during follow-up, and no bradycardia or sinus pause was observed.

Discussion

Surgical closure of SVASD is associated with arrhythmia, residual shunts, or patch dehiscence.[11] Compared with surgery, transcatheter SVASD closure may be a cost-effective alternative in that it avoids cardiopulmonary bypass and prolonged ICU stays.

Various modified transcatheter techniques have been described. Some studies have utilized virtual and 3D printed modeling to predict treatment results.[9] Garg et al[12] performed transcatheter SVC stenting for SVASD using available resources in developing countries without advanced imaging or 3D-printed models.

Successful transcatheter SVASD intervention depends on accurate preprocedural CTA measurements and anatomical considerations. CTA-guided sizing identifies morphological variations—such as left SVC, innominate vein anomalies, or pulmonary vein abnormalities—that may preclude transcatheter intervention.[10]

While many studies have used self-expanding stents, we employed balloon-expandable stents with acceptable results. Patient selection has traditionally relied on balloon expansion testing to ensure that the SVC balloon does not occlude RUPV drainage. Studies have excluded patients if balloon occlusion testing indicated inadequate RUPV drainage.[5,9,10,13] Modern imaging, including CTA, may substitute for balloon occlusion testing.[13] We did not routinely perform balloon occlusion testing; rather, we evaluated anatomy via preprocedural CTA and employed a parked RUPV catheter to facilitate ballooning or stenting to establish RUPV-to-LA drainage, as described in CASE 2. Patients were, therefore, selected before catheterization. We sought to offer this technique to more patients because RUPV occlusion can be addressed without major complications. Still, long-term follow-up with CTA is required.

The only complication encountered was stent embolization 2 hours postimplantation, which required emergent surgical extraction. It is advisable that a surgeon be included in the clinical team for this procedure. Ideally, it should be performed in a hybrid catheterization laboratory. Except for this case, all results were satisfactory. Retrospective evaluation suggests that if angiography shows a rapidly reducing SVC diameter or a cone-shaped covered stent, a bare stent should be implanted in the upper SVC so as to improve stability and elongate the construct.

Concerns exist regarding long-term SVC or stent-site stenosis in low-weight children. Based on experiences with stent redilation for aortic coarctation in children, SVC redilation should be considered during follow-up if stenosis occurs.[14] Mild residual shunts noted postimplantation resolved within 3 months. This finding is consistent with those reported by Abdullah et al,[5] who reported successful long-term SVASD stenting, including 1 case of residual shunt eventually closed with a device and overlapping covered stents. Close follow-up via TTE and CTA is required to monitor for residual shunts following nonsurgical management. Self-expanding stents are advantageous because a single stent may be sufficient if the length is determined accurately, reducing costs. However, Muthukumaran et al[10] reported that self-expanding pins or barbs might protrude through the SVC and scrape the ascending aorta, leading to hemopericardium or cardiac tamponade. Balloon-expandable stents do not carry this risk.[10]

Conclusions

Transcatheter closure of SVASD with balloon-expandable stents, guided by imaging modalities such as CTA instead of balloon expansion testing during angiography, may be considered an alternative to surgery. Furthermore, RUPV occlusion during balloon expansion testing should not always be considered a contraindication for transcatheter closure of SVASD.

Declarations:

Ethics Approval

Informed consent was obtained from all patients and/or their legal guardians, and the novelty of the procedure was explained to all patients and their legal guardians.

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Conflict of Interest

The authors report no conflict of interest.

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