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Percutaneous Closure of a Superior Sinus Venosus Atrial Septal Defect as an Alternative to Surgical Treatment

Superior Sinüs Venosus Atriyal Septal Defektin Cerrahi Tedaviye Alternatif Olarak Perkütan Yolla Kapatılması

ABSTRACT

A superior sinus venosus atrial septal defect (SVASD) results from a defect in the atrial wall that forms the posterior wall of the superior vena cava (SVC) and the anterior wall of the right upper pulmonary vein (RUPV), with the posterior wall of the RUPV typically connected to the left atrium. While surgery is usually recommended for SVASD, percutaneous technique may serve as an alternative to surgery in selected patients. Here, we report on the percutaneous closure of the defective posterior wall of the SVC using a covered stent, thereby closing the superior SVASD and redirecting the anomalous RUPV behind the stent into the left atrium.

Keywords: Atrial septal defect, percutaneous closure, sinus venous

ÖZET

Superior sinüs venosus atriyal septal defekt (SVASD), superior vena kava'nın (SVK) arka duvarını oluşturan atriyal duvar ile sağ üst pulmoner venin (SÜPV) ön duvarının eksikliğinden kaynaklanır. SÜPV'nin arka duvarı normalde sol atriyuma bağlıdır. SVASD için genellikle cerrahi tavsiye edilirken, SVASD'li seçilmiş hastalarda perkütan teknik cerrahiye alternatif olabilir. SVK'nın kusurlu arka duvarının greft kaplı stent ile perkütan olarak kapatılmasını bildiriyoruz. Bunun sonucunda superior SVASD kapatıldı ve anormal SÜPV stentin arkasından sol atriyuma yeniden yönlendirildi.

Anahtar Kelimeler: Atrial septal defekt, perkütan kapatma, sinüs venozus

CASE REPORT
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A superior sinus venosus atrial septal defect (SVASD) results from a defect in the atrial wall, constituting the posterior wall of the superior vena cava (SVC) and the anterior wall of the right upper pulmonary vein (RUPV), with the posterior wall of the RUPV normally connecting to the left atrium. The percutaneous closure of a superior SVASD using a covered stent was first documented by Garg et al.[1] in 2014. Since then, approximately 50 cases have been reported in the literature.[2,3]

In this case report, we detail the percutaneous closure of the defective posterior wall of the SVC using a covered stent, resulting in the closure of the superior SVASD and the redirection of the anomalous RUPV behind the stent into the left atrium (LA). Prior to the procedure, written informed consent was obtained from the patient, followed by a thorough educational session about the innovative nature of this procedure.

Case Report

A 22-year-old woman presented with increasing exertional dyspnea. Transesophageal echocardiography (TEE) revealed a 22 mm superior SVASD, without accompanying anomalous pulmonary veins. The patient declined open-heart surgery.

Pre-procedure

Eligibility for percutaneous closure was assessed through a multimodal, stepwise approach. This included an assessment using 2-dimensional and 3-dimensional TEE, cardiac computed tomography, and an ex vivo simulation of stent implantation with a 3D printed model (Videos 1, 2). In our case, diagnostic catheterization with balloon test occlusion followed by subsequent stent implantation was not performed.

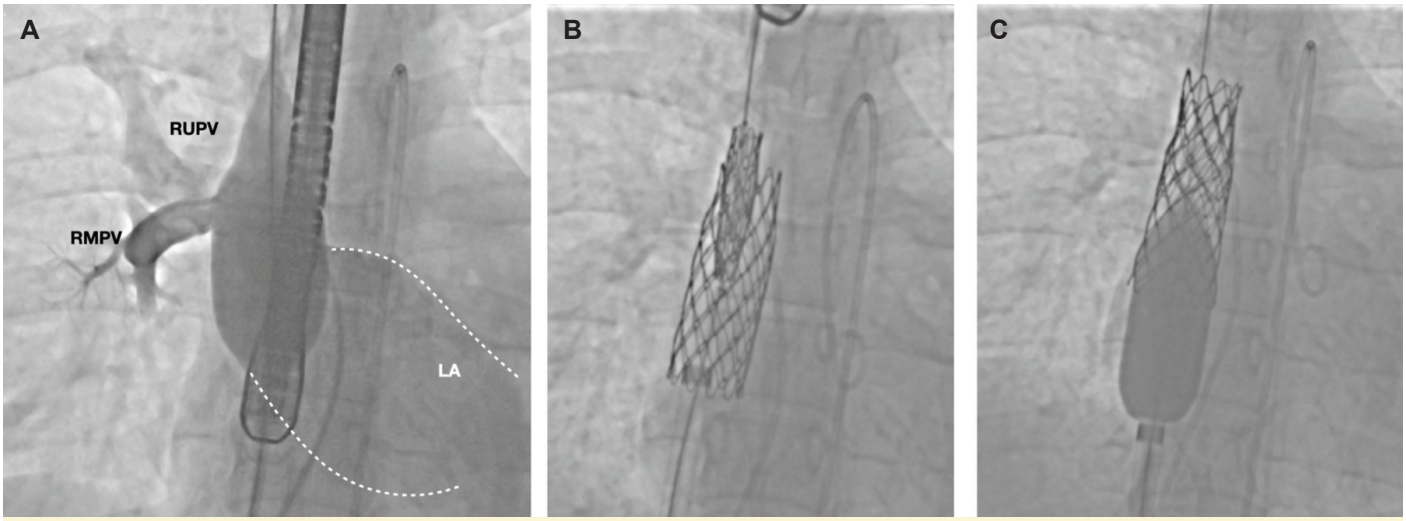

Figure 1. (A) Balloon occlusion testing with an Amplatzer sizing balloon and selective pulmonary vein angiography. (B) Anchoring of an additional stent, overlapping the covered stent, allowing for (C) stable flaring of the covered stent. LA, left atrium; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein.

Procedure

The procedure was conducted under general anesthesia with simultaneous TEE guidance. Vascular access was obtained via the right internal jugular vein (RIJV), the left and right femoral veins, and the left femoral artery. Heparin was administered to maintain an activated clotting time of >250 s.

A transseptal puncture was performed using a Brockenbrough transseptal needle (St. Jude Medical), and a 6 Fr multipurpose catheter (MPA 1) was placed in the RUPV to allow for simultaneous venography. A veno-venous guidewire circuit was established using a 0.035-in Extra Stiff wire (Cook Medical) from the right femoral vein to the RIJV to facilitate balloon occlusion and stent implantation.

A 34-mm Amplatzer sizing balloon II (St. Jude Medical) was tracked and inflated at the SVC for balloon testing, with inflation continued until TEE confirmed the elimination of the interatrial shunt (Video 3). The initial balloon size was chosen to be at least 2 to 4 mm larger than the SVC diameter. The patency of the RUPV to the LA was confirmed with angiography from the RUPV while the balloon remained inflated in the SVC (Figure 1A, Video 4). During balloon inflation, flow in the RUPV was assessed by TEE, and simultaneous pressure measurements were taken in the RUPV and LA.

The measurements obtained during balloon testing were instrumental in selecting the appropriate stent length and balloon diameter for implantation. Based on these measurements, a 10-zig covered Cheatham platinum stent (CP) measuring 60 mm in length was mounted on an 18×45 mm balloon-in-balloon (BiB) catheter (NuMED, Inc.) for use. The stent was introduced through an 18-F Check-Flo Performer (Cook Medical) via the right femoral vein. Following the inflation of the inner balloon, the position of the stent was verified. The upper end of the stent was positioned just below the azygos SVC junction, and the lower end was situated in the right atrium (RA), below the inferior rim of the atrial septal defect (ASD), as confirmed by TEE. After the inflation of the outer balloon, the stent's opposition to the wall was ascertained through simultaneous TEE imaging and angiography from both the RIJV and RUPV.

A high-pressure balloon was kept on standby in the RUPV to mitigate any potential obstruction of the pulmonary venous return behind the stent.

An additional 8-zig, 39 mm CP stent (NuMED, Inc.) was deployed, overlapping with the cranial end of the covered stent, to preclude any possibilities of stent instability or embolization (Figure 1B, Video 5). The lower end of the stent underwent redilation with a 25x40 mm Z-Med II balloon (NuMED, Inc.) ensuring a secure attachment to the interatrial septum (IAS) (Figure 1C).

Pulmonary venography of the SVC and RUPV, pressure measurements in the RUPV and LA, and TEE were utilized to assess the remaining shunt and confirm the absence of pulmonary venous compression. At the conclusion of the procedure, pulmonary venous return was unobstructed according to pulmonary venography and Doppler echocardiography results. There was no gradient from RUPV to LA, and no residual shunt was detected between the atria.

ABBREVIATIONS

ASD	Atrial septal defect
BiB	Balloon-in-balloon
CP	Cheatham platinum
CT	Computed tomography
ECG	Electrocardiogram
IAS	Interatrial septum
LA	Left atrium
MPA	Multipurpose catheter
RA	Right atrium
RIJV	Right internal jugular vein
RUPV	Right upper pulmonary vein
SND	Sinus node dysfunction
SVASD	Sinus venosus atrial septal defect
SVC	Superior vena cava
TEE	Transesophageal echocardiography

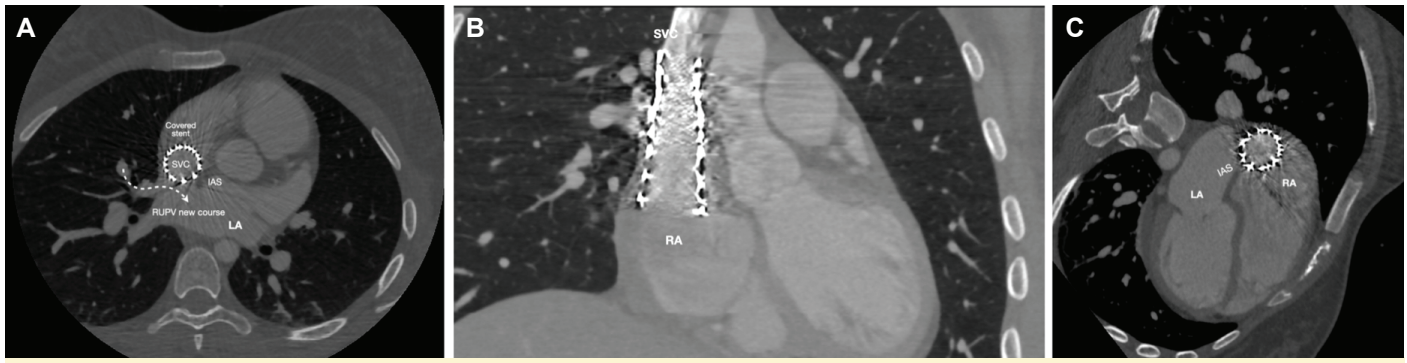

Figure 2. Cardiac CT in the (A) axial and (B) sagittal views shows a patent SVC stent and pulmonary venous return behind the stent. (C) The post-procedure CT angiogram displays good opposition of the lower end of the stent to the IAS. CT, computer tomography; IAS, interatrial septum; LA, left atrium; RA, right atrium; SVC, superior vena cava.

Post-procedure

The patient was transferred to the ward on the same day. The electrocardiogram (ECG) showed a sinus rhythm. The following day, a computed tomography (CT) angiogram was performed, revealing unobstructed RUPV and SVC flow (Figures 2A, 2B). The stent appeared to be well-opposed to the IAS (Figure 2C). The patient was subsequently discharged with a prescription for dual antiplatelet therapy, and no late complications were observed on follow-up echocardiography.

Discussion

This case represents a successful instance of percutaneous closure of a superior SVASD in an adult patient, executed without any complications. SVASDs constitute approximately 4-11% of ASDs.[4] Although surgery is generally the recommended course of treatment for SVASD, this case demonstrates that percutaneous techniques can serve as a viable alternative to surgery for selected patients with SVASD.[5]

Similar to the surgical correction of SVASD, the potential risk of sinus node dysfunction (SND) is a significant concern, particularly when a stent is placed at the junction of the SVC and the right atrium.[6,7] In the case of our patient, no instances of SND were observed during the follow-up period after the percutaneous closure. Another complication commonly associated with surgical repair is SVC stenosis; however, this is considered to be less likely to occur with the percutaneous approach.[8]

Balloon occlusion testing at the SVC-RA junction prior to stent implantation is crucial for evaluating the potential for pulmonary venous obstruction and determining the appropriate balloon size for the stent implantation procedure. To circumvent pulmonary venous compression, we meticulously monitor pulmonary venous flow via transseptal access. Furthermore, having access to the RUPV from the left atrium may facilitate bail-out balloon dilatation or even stent implantation in the event of unexpected RUPV obstruction. TEE imaging plays a vital role throughout the procedure; it guides the transseptal puncture and monitors for residual defects and pulmonary vein obstruction during balloon testing, stent implantation, and post-deployment dilation.

The primary risk associated with this therapy is stent migration, a complication that may arise in approximately 1% of cases if the stent deployment does not align perfectly with the anchoring site in the SVC.[9] The 60 mm 10-zig covered CP stent, currently the longest balloon-expandable stent approved by the CE, is utilized in such procedures. To mitigate the risk of stent embolism, we intentionally oversize the stent diameter by 2 to 4 mm relative to the SVC, aiming for an apposition zone of at least 2 cm within the SVC. Nevertheless, stent instability is a frequent occurrence, necessitating additional stabilization with an overlapping bare metal stent.

Residual shunting may manifest due to inadequate ASD coverage by the stent, significant stent shortening during expansion of its caudal portion (potentially exposing the inferior rim of the defect), or excessively high stent deployment. TEE assessments reveal that over half of the patients exhibit a small residual shunt at the conclusion of the procedure.[10] We hypothesize that the interatrial communication will eventually close completely, following stent endothelialization and subsequent volume relief, ultimately leading to a reduction in RA size.

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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Video 1. This 3D printed heart model displays the anatomical relationship between the superior vena cava (SVC) and the right upper pulmonary vein (RUPV).

Video 2. Findings from transesophageal echocardiography in a sinus venosus atrial septal defect. RA, right atrium.

Video 3. Confirmation of the defect's closure during balloon occlusion testing with transesophageal echocardiography. B: Amplatzer sizing balloon.

Video 4. Balloon occlusion testing with an Amplatzer sizing balloon; during this testing, pulmonary venography confirmed drainage to the left atrium. LA, left atrium; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein.

Video 5. An additional stent is anchored, overlapping the covered stent.

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