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# Transcatheter Covered Stent Exclusion of Superior Sinus Venosus Defects

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## ABSTRACT

**BACKGROUND** Transcatheter correction of sinus venosus defects (SVDs) using balloon-mounted covered stents provides an attractive surgical alternative. Surgery may be complicated by superior vena caval or right upper pulmonary vein (RUPV) stenosis, sinus nodal dysfunction, and residual additional pulmonary veins.

**OBJECTIVES** Being a new intervention, technical modifications would simplify the procedure, improve universal applicability, and reduce or tackle complications.

**METHODS** Patients were included if balloon interrogation of cavoatrial junction confirmed closure of SVD and redirected RUPV to the left atrium. A single-center experience was analyzed to summarize the procedural modifications over 8 years. Transesophageal echocardiogram (TEE) on follow-up was done to identify residual shunt, RUPV flows, and stent thrombosis.

**RESULTS** A total of 100 patients including 9 children with a median age of 35 years (range, 4-69 years) underwent SVD closure after balloon interrogation. Among 57 patients balloon interrogated in the first 5 years, 70% underwent transcatheter closure, with 2 failures. RUPV occlusion caused the exclusions. Inclusions improved to 94% among the subsequent 65 balloon interrogations when RUPV protection was implemented, with 1 failure. Stent embolization caused the 3 failures warranting surgery. Recent modifications included limited transesophageal echocardiogram without anesthesia, avoiding venovenous circuit, interrogation with semicompliant balloons, trans-septal RUPV protection, overlapping stents to permit additional vein drainage to superior vena cava and tackle embolizations. There were no deaths. Minor complications included stent embolizations stabilized in catheterization laboratory in 2 patients, left innominate vein jailing in 2 patients, insignificant residual flows, and nonocclusive asymptomatic stent thrombosis in 4 patients.

**CONCLUSIONS** Procedural success was 97%. Recent modifications increased patient inclusions, decreased complications, and simplified the intervention. (J Am Coll Cardiol 2024;83:2179-2192) © 2024 by the American College of Cardiology Foundation.


Complications following surgical sinus venosus defect (SVD) repair included superior vena caval (SVC) stenosis in 4% to 5%, right upper pulmonary vein (RUPV) stenosis in 1%, sick sinus syndrome in 4%, and extremely rare redirection of SVC to the left atrium.[1] Transcatheter closure using covered stents after balloon interrogation of cavoatrial junction provided an alternative solution.[2-5] Early attempts relied on 3-dimensional printed heart models and virtual reality.[5,6] Less expensive tools such as transesophageal echocardiography (TEE) and hemodynamic information during balloon interrogation simplified the evaluation.[4,7,8] Being a new procedure, imaging modalities, techniques, types of stents, and follow-up protocols varied widely between institutions.[9-11] Long-term outcomes and complications should be documented by a reliable imaging modality before accepting this intervention as a surgical alternative. Our initial experience, procedural outcomes, and short-term follow-up have been reported elsewhere.[4] This study highlighted the procedural evolutions over the years and late

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**ABBREVIATIONS AND ACRONYMS**

**ASD** = atrial septal defect

**CT** = computed tomography

**MRI** = magnetic resonance imaging

**RUPV** = right upper pulmonary vein

**SVC** = superior vena cava

**SVD** = sinus venosus defect

**TEE** = transesophageal echocardiography

outcomes of the largest cohort of patients from a single institution.

## METHODS

This single-center study analyzed all transcatheter SVD interventions using covered stent(s) from May 2015 to October 2023. All patients consented for the procedure after counseling regarding surgical and transcatheter options. The institutional ethics committee approved the procedure and anonymized reporting.

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**PREPROCEDURAL EVALUATION AND IMAGING.** Transthoracic echocardiography assessed right heart enlargement, pulmonary hypertension and associated anomalies. Additional imaging with TEE determined the caudal extension of the SVD toward the oval fossa. If the vertical dimension of SVD on a bicaval TEE view was larger than the transverse cavoatrial diameter, it was defined as significant caudal extension. Computed tomography (CT) scan located the RUPV in relation to the cavoatrial junction and identified high-draining veins with minimal patient discomfort compared with TEE imaging. Patients without a significant caudal extension and those who did not have large high-draining additional veins were considered for balloon interrogation.

**BALLOON INTERROGATION PROCEDURE.** A stiff Lunderquist wire (Cook Medical) was placed from the right femoral vein across the cavoatrial junction with its tip parked in the contralateral brachial vein or ipsilateral jugular bulb for a stable position before introducing a semicompliant long balloon with a diameter ≥4 mm larger than the SVC (**Figure 1**). The RUPV was cannulated either antegrade through a left femoral trans-septal sheath or retrograde through a left femoral arterial access. Caudal melon seeding of the balloon during inflation due to the bell-bottom configuration of the cavoatrial junction was prevented by the upper end of the long sheath placed in the right femoral vein. Any coexisting secundum atrial septal defect (ASD) was additionally interrogated with a sizing balloon (**Figure 2**).

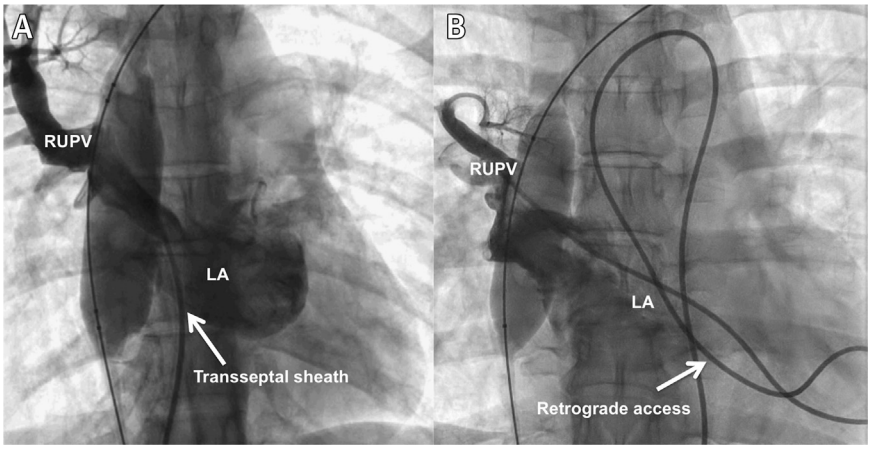
**OBSERVATIONS DURING BALLOON INTERROGATION.** Feasibility for interventional correction was confirmed when an angiogram showed the inflated balloon redirecting the RUPV to the left atrium without filling the right atrium and without increasing the RUPV pressures by >2 mm Hg. If there was a residual shunt, the balloon was upsized by 2 mm. The balloon was downsized by 2 mm if it obstructed the RUPV. In recent years, patients with gradients of 2 to 8 mm Hg between the RUPV and the left atrium were still included for transcatheter closure using RUPV protection technique.[5] Children were included only when the diameter of the interrogating balloon was ≥16 mm.

**PROCEDURAL PLANNING AND VASCULAR ACCESS.** The patient was either awake or under conscious sedation during the procedure. Intubation anesthesia was restricted to children and apprehensive patients. The right femoral vein was used for advancing the interrogating balloon and covered stent. Left femoral venous access allowed RUPV cannulation through a preexisting ASD, patent foramen ovale, or a trans-septal puncture using Brockenbrough needle. Femoral arterial access allowed coronary angiogram and retrograde RUPV cannulation through the mitral valve. A venovenous circuit created in the past after right jugular access was not considered mandatory; a stiff wire from the right femoral vein parked in the brachial vein or jugular bulb provided adequate support.

**INTRAPROCEDURAL TEE MONITORING.** Intraprocedural continuous TEE monitoring was limited to young apprehensive mechanically ventilated patients, those with additional secundum ASD, those with advanced renal disease that precluded radiographic contrast use, but was not considered mandatory in all. In awake patients, a limited TEE was performed after expanding the covered stent to analyze a residual leak on RUPV angiogram (**Figure 3**).

**DEPLOYING THE COVERED STENT.** A large sheath adequate for the balloon-mounted covered stent was inserted in the right femoral vein over the stiff guidewire. Stent length was determined by adding an anchor zone ≥2 cm in the SVC cranial to the RUPV and further adding a caudal protrusion of ≥1 to 2 cm into the right atrium.[4,7] Foreshortening on expansion was considered during stent selection. The covered stents were either 10-zig covered CP (Numed Inc), covered Andrastent (Andramed GmbH), covered ZephyrXL (Sahajanand Laser Technology Limited), or covered OptimusXL (AndraTec GmbH).[12,13] In patients with a high-draining additional vein, an uncovered stent was overlapped at the cranial end of the long balloon above and within the covered stent to improve the anchor and permit the additional vein to drain into the SVC (**Figure 2**). If the SVC anchor below the additional vein seemed adequate, a single long covered stent was deployed caudal to the vein.[7] If the additional vein draining high in the SVC measured <8 mm, the patients were included for this strategy.

**FIGURE 1** Monitoring the RUPV

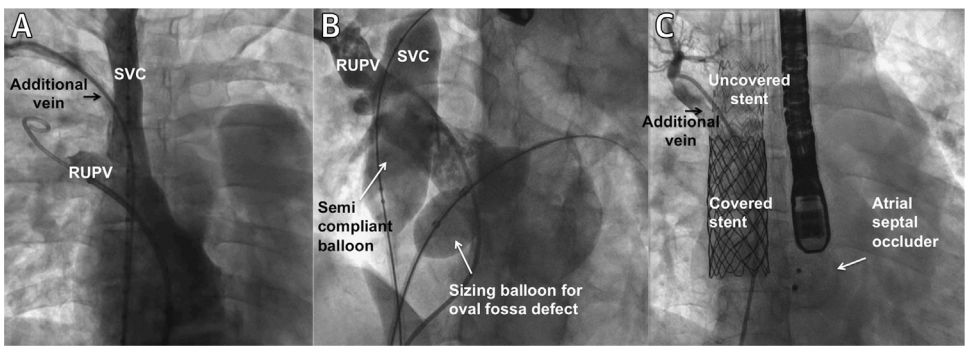



During balloon interrogation, a catheter is placed in the right upper pulmonary vein, either antegradely (A) through a venous trans-septal sheath or an arterial retrograde (B) catheter. LA = left atrium; RUPV = right upper pulmonary vein; SVC = superior vena cava.

**PULMONARY VEIN MONITORING.** RUPV cannulation and monitoring was considered mandatory both during balloon interrogation and after covered stent expansion. Our criteria for procedural success were complete closure of SVD, redirection of the RUPV to the left atrium, and a lack of elevation of the RUPV pressures beyond 2 mm Hg above the left atrium. Monitoring RUPV throughout the procedure assessed all these parameters. In our early experience, a direct RUPV cannulation through the SVD during balloon interrogation and observing the levo-phase of right pulmonary arterial angiography after stent deployment confirmed procedural success. Retrograde cannulation with an arterial catheter or antegrade cannulation through a trans-septal sheath was adopted later. In patients needing trans-septal pulmonary vein protection, additional retrograde cannulation allowed simultaneous RUPV pressure monitoring.

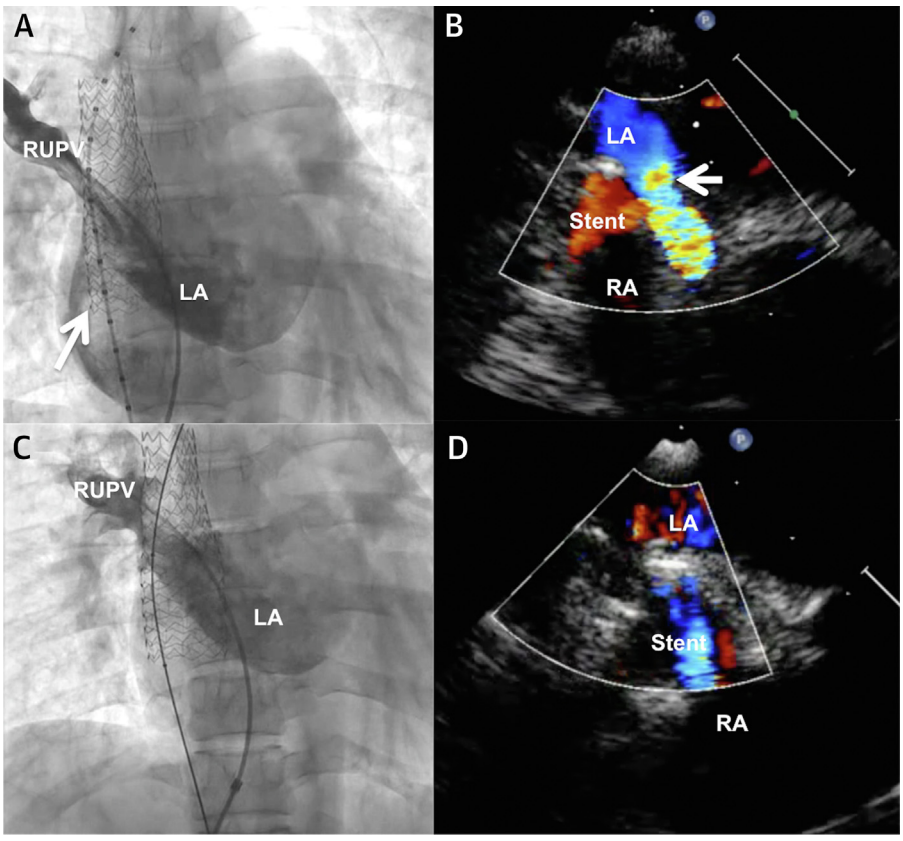
**PULMONARY VEIN PROTECTION.** Pulmonary vein protection was considered if the RUPV pressures increased by >2 mm Hg above the left atrium and RUPV angiography showed a hold-up of contrast.[5]

**FIGURE 2** Additional Pulmonary Vein and Oval Fossa Defect




Additional pulmonary veins (A) and the main RUPV are separately cannulated. An additional oval fossa defect is simultaneously occluded by a sizing balloon (B) during balloon interrogation of the cavoatrial junction using a semicompliant balloon. An uncovered stent mounted above the covered stent (C) allows the additional pulmonary vein to drain into the SVC, and oval fossa defect is closed by an occluder. Abbreviations as in Figure 1.

**FIGURE 3 Limited Intraprocedural Transesophageal Imaging**




RUPV angiogram (A) and limited brief transesophageal echocardiogram (B) shows residual shunt filling the right atrium (RA) owing to a lack of apposition. Expansion of the lower end redirects RUPV (C) to the left atrium (LA), confirmed by echocardiogram (D). Abbreviations as in Figure 1.

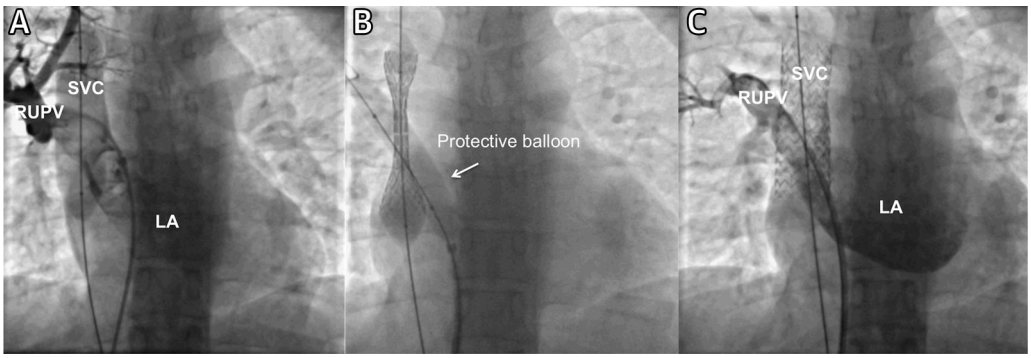
A noncompliant 12- to 14-mm balloon was kept inflated in the RUPV orifice at 12 to 14 atm through the left femoral trans-septal sheath (Figure 4). This was done before expansion of the covered stent typically at 3 to 5 atm. The caudal end of the covered stent was further dilated with a larger balloon for obliterating residual flows, while keeping the protective balloon fully inflated.

**POSTPROCEDURAL STEPS.** After confirming lack of pressure gradient between the RUPV and left atrium, an angiogram of RUPV was done to confirm complete closure of the SVD. Venous hemostasis was achieved by a figure-of-8 stitch. A daily dose of 150 mg of aspirin and 75 mg of clopidogrel was continued for 6 to 12 months in most adults. In children, aspirin was administered at a daily dose of 5 mg/kg and clopidogrel was administered at a dose of 1 mg/kg. Anticoagulation was restricted to those with atrial fibrillation.

**FOLLOW-UP.** Imaging with TEE was performed after 1 to 3 months in all patients to study residual shunt, color Doppler flows in the RUPV and identify any thrombus. As the metallic stent artifacts during CT scan and ferromagnetic artifacts during magnetic resonance imaging (MRI) obscured the blood pool imaging, TEE with high temporospatial resolution was preferred. A CT scan and MRI were used in children as outpatient TEE was impractical. Residual leak of >3 mm color Doppler width was considered a mild leak and those ≤3 mm were considered insignificant. Antiplatelet therapy duration was extended in the presence of residual leaks or associated conditions like coronary artery disease and cerebral strokes.

**SPECIAL CONSIDERATIONS IN PATIENTS <18 YEARS OF AGE.** Younger patients were included only when the interrogating balloon diameter was ≥16 mm. A caval diameter of 16 mm was considered to be adequate for upper body venous drainage in adult

**FIGURE 4** Pulmonary Vein Protection

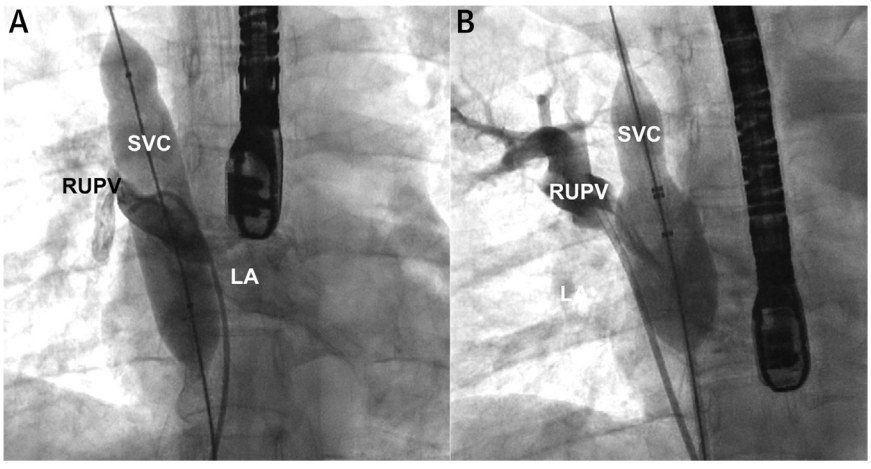



If the RUPV angiogram during balloon interrogation shows contrast stasis (A) despite complete shunt closure, the RUPV is protected by inflating a noncompliant balloon (B) during stent expansion in the SVC. A RUPV angiogram (C) confirms complete sinus venosus defect closure. Abbreviations as in Figure 1.

life. TEE monitoring under general anesthesia was always used in these young apprehensive patients. A CT scan or MRI was preferred to TEE during follow-up for assessing the residual flows and RUPV redirection.

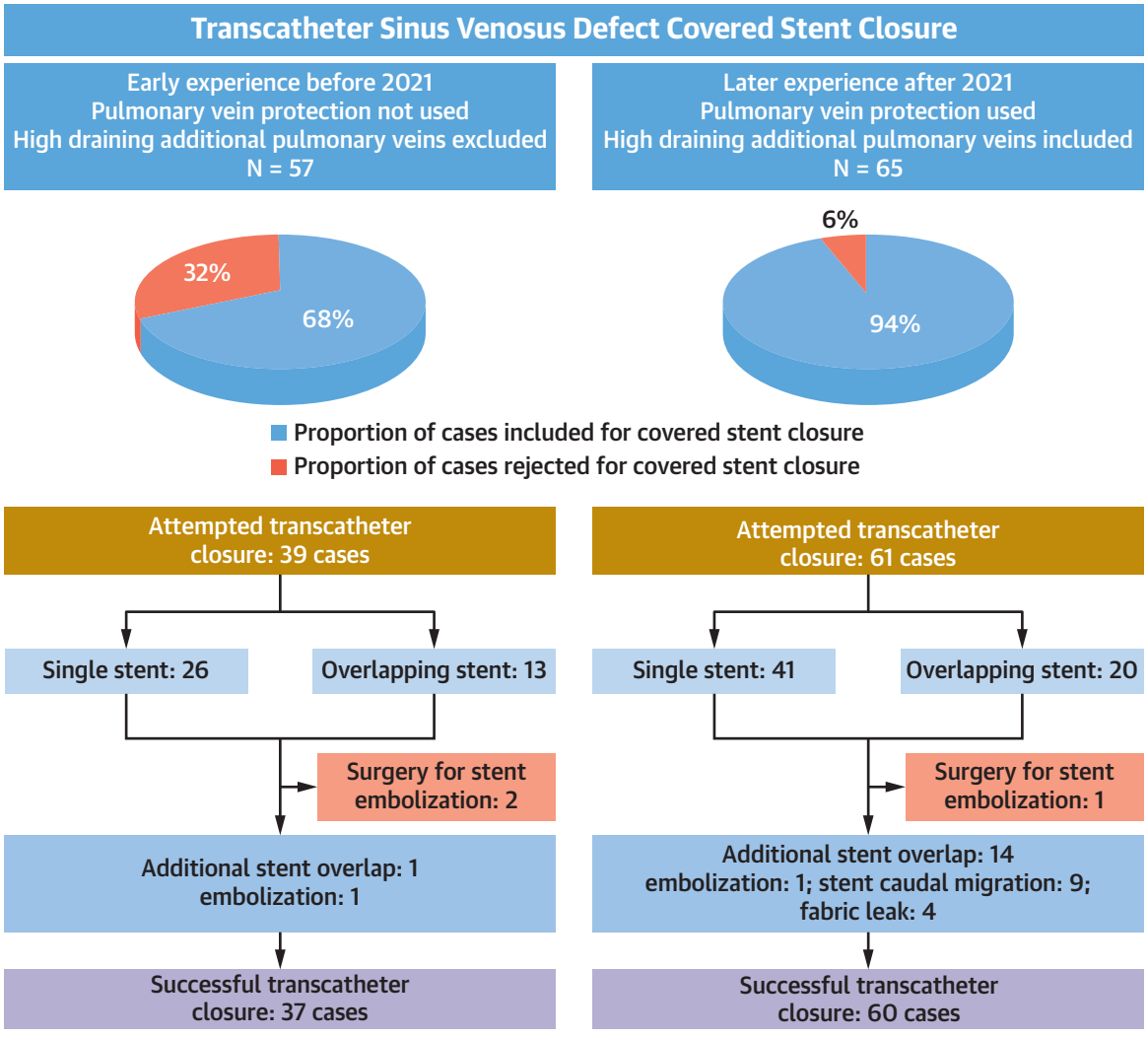
**PROCEDURAL EVOLUTION OVER TIME.** A compliant sizing balloon used for balloon interrogation earlier was replaced by semicompliant cylindrical balloons as the former bulged into the RUPV and occluded some of them (Figure 5). Stability of the guidewire obtained by venovenous circuit after jugular access was alternatively achieved by deep placement of stiff wires in brachial vein or jugular bulb, improving the workflow in catheterization laboratory. Continuous TEE guidance under anesthesia was replaced by short conscious TEE after stent expansion. This strategy avoided intubation in patients with severe pulmonary hypertension and morbid obesity. An RUPV protection strategy and accepting patients with high-draining additional veins led to a significant decrease in exclusions compared with the initial

**FIGURE 5** Difference in Interrogating Balloons




An inflated semicompliant balloon (A) in the SVC shows closure of shunt and the RUPV redirected to the LA. But a sizing balloon (B) bulges into the RUPV and causes mild obstruction. Abbreviations as in Figure 1.

**CENTRAL ILLUSTRATION** Procedural Evolution Over Time




**Sagar P, et al. J Am Coll Cardiol. 2024;83(22):2179-2192.**

Pulmonary vein protection strategy, including patients with additional high-draining veins and overlapping stents for stent embolization and migration increased patient inclusion and procedural success.

experience.[4] Stent embolizations as well as minimal migrations before guidewire removal were tackled effectively by additional overlapping covered stents.

**STATISTICAL METHODS.** Statistical analysis used IBM SPSS Statistics version 25 software (IBM). Categorical variables were expressed as a frequency or a percentage. Continuous variables were presented as median (range) or mean ± SD, as appropriate. Analysis of factors associated with stent thrombosis was tested using Fisher exact test for categorical data and Mann-Whitney *U* test for continuous data in non-normal distribution. A value for *P* value of $<0.05$ was considered statistically significant.

## RESULTS

Among a total of 122 patients who underwent balloon interrogation in the past 8 years, 100 cases fulfilled the inclusion criteria. Our first publication consisted of 42 balloon interrogations and 24 covered stent implantations.[4] Reasons for the 18 exclusions were pulmonary venous obstruction, incomplete SVD occlusion and high pulmonary vascular resistance. When pulmonary vein protection was not considered during the initial 57 balloon interrogations performed before 2021, our inclusion rate was 68.4%.[5] When pulmonary vein protection was adopted in the

subsequent 65 cases, the inclusions improved to 93.8% (Central Illustration). Twenty-eight cases underwent transcatheter closure with pulmonary vein protection.[5] Sixteen among 17 patients with high-draining additional pulmonary veins were included after 2021.

**PATIENT CHARACTERISTICS.** A total of 100 patients with a median age of 35 years (range, 4-69 years) formed the study cohort (Table 1). Atrial arrhythmias in 9 patients included atrioventricular nodal reentrant tachycardia in 3 and atrial flutter/fibrillation in the rest. All except one were medically or electrically cardioverted before the procedure. Three patients with severe pulmonary hypertension were pretreated with targeted pulmonary vasodilators before the intervention. Although TEE was the most common preprocedural imaging modality earlier, a CT scan was increasingly used later. Severe renal failure in 1 patient was managed with a zero-contrast policy under general anesthesia and TEE guidance.

**PROCEDURAL DATA.** Of the 100 patients, 81 underwent balloon interrogation and covered stent implantation in the same sitting and the rest were staged due to nonavailability of appropriate stent on shelf. Mechanical ventilation for continuous TEE monitoring was used only in 14 cases. Concurrent procedures included 6 ASD device closures, 1 balloon pulmonary valvotomy and 1 coronary stent angioplasty. Procedural details are presented in Table 2.

**NUMBER AND TYPES OF STENTS.** A total of 151 stents used in 100 patients included 126 covered and 25 bare stents. A single covered stent was the initial choice in 67 patients and 2 overlapping stents on a long balloon were chosen in the remaining 33 cases. Two covered stents were overlapped to achieve an adequate length in 11 cases where a single long stent was unavailable. An uncovered stent was overlapped as an anchoring stent in the remaining 22 patients. Additional stents were used in 18 cases to manage problems such as residual flows, caudal stent migration or stent embolization. A long covered 10-zig CP stent was the most common stent used (Table 2).

**ADDITIONAL PULMONARY VEINS.** Seventeen patients with high-draining additional pulmonary veins were considered unsuitable for surgical redirection after heart team discussion. An overlapping uncovered anchoring stent in eleven of these patients allowed their unobstructed drainage into the SVC (Figure 2). A single covered stent was precisely placed caudal to the additional vein in 5 patients where the anchor zone was initially considered to be adequate. Minimal caudal migration in 2 among these 5 patients was managed using additional anchoring stents.

**TABLE 1 Patient Data**

| | |
|---|---|
| Age, y | 35 (4-69) |
| Age ≤12 y | 9 (9) |
| Female | 49 (49) |
| Symptoms | |
| Asymptomatic | 28 (28) |
| Exertional dyspnea | 72 (72) |
| Paradoxical embolism | 2 (2.0) |
| Arrythmias | 9 (9) |
| Weight, kg | 63 (17-119) |
| Body surface area, $m^2$ | 1.63 (0.72-2.25) |
| Bilateral superior vena cava | 24 (24) |
| Pulmonary hypertension | 72 (72) |
| Mild | 59 (59) |
| Moderate | 10 (10) |
| Severe | 3 (3) |
| Additional high-draining pulmonary vein | 17 (17) |
| Preprocedural imaging | |
| Transesophageal echocardiography | 33 (33) |
| Computed tomography scan | 64 (64) |
| Magnetic resonance imaging | 3 (3) |

Values are median (range) or n (%).

In one patient, the fabric of a long, 79-mm covered Zephyr stent was removed from 15 mm of its cranial end to allow flows from the additional vein.

**ADDITIONAL OVAL FOSSA DEFECT.** Six patients with an additional secundum ASD underwent simultaneous interrogation of the SVD and ASD using 2 balloons to evaluate feasibility. Along with transcatheter SVD closure, ASD was closed with conventional occluders ranging in size from 12 to 36 mm (Figure 2). The evaluation and procedural details have been reported elsewhere.[14]

**PROCEDURAL OUTCOMES AND COMPLICATIONS.** The procedure was successful in 97 cases (Table 3). There were no in-hospital deaths. Three procedural failures were due to intraprocedural stent embolization that required surgery. Surgery was uneventful in these 3 patients. Stent embolization occurred in 5 cases, 2 of which occurred among the first 6 cases in the early years of experience. Although sequential deployment of 3 covered, 45-mm, 8-zig covered CP stents to straddle across the defect failed in our first patient, use of a single 48-mm covered Andrastent in the sixth patient aged 5 years also led to embolization. Barring these 2 patients, none of our patients received a stent shorter than 57 mm. Embolizations were related to short stents and inadequate SVC anchor given the conical shape of the cavoatrial junction. The stent in 1 patient embolized immediately after removal of the guidewire and needed surgical referral. Two recent embolizations that occurred

**TABLE 2 Procedural Data**

| | |
|---|---|
| 2-staged procedure (balloon interrogation/covered stent) | 19 (19) |
| Mechanical ventilation | 14 (14) |
| Intraprocedural transesophageal echocardiography | 33 (33) |
| Pulmonary:systemic blood flow ratio | 2.35 (1.5-4.0) |
| Right upper pulmonary vein cannulation | |
| Direct venous | 16 (16) |
| Retrograde arterial | 22 (22) |
| Trans-septal | 62 (62) |
| Successful redirection of RUPV with first balloon | 55 (55) |
| Need for upsizing the balloon | 23 (23) |
| Need for downsizing the balloon | 17 (17) |
| Both | 5 (5) |
| Venovenous circuit from jugular vein | 21 (21) |
| Pulmonary vein protection | 28 (28) |
| RUPV monitoring during stent expansion | |
| Direct venous | 8 (8) |
| Retrograde arterial | 20 (20) |
| Trans-septal | 60 (60) |
| Both retrograde arterial and trans-septal | 4 (4) |
| Levo-phase of right pulmonary artery angiogram observation | 8 (8) |
| Additional oval fossa defect device closure | 6 (6) |
| Number of stents used per patient | |
| 1 | 60 (60) |
| 2 | 29 (29) |
| 3 | 11 (11) |
| Bare stents for anchor/to prevent additional vein occlusion | 25 (25) |
| Additional stent for bail out | 18 (18) |
| Impending stent migration | 9 |
| Complete stent embolization within the guidewire | 2 |
| Fabric leak with residual shunt | 4 |
| Inadequate cover of the SVD by the caudal end of the stent | 1 |
| Inadequate anchor for covered stent deployed below high draining additional veins | 2 |
| No. of stents used | 151 |
| Covered CP stent | 52 (34.4) |
| Covered Andrastent | 31 (20.5) |
| Covered Zephyr stent | 40 (26.5) |
| Covered Optimus CVS stent | 3 (1.9) |
| Uncovered Andra stent | 13 (8.6) |
| Uncovered Zephyr stent | 12 (7.9) |
| Flaring of lower end with larger balloon | 39 (39) |
| Procedural time, min | 105 (55-240) |
| Fluoroscopy time, min | 25.29 (9.31-82.14) |
| Dose area product, mGray.cm$^2$ | 52,584 (7,135-290,917) |

Values are n (%), median (range), or n.
RUPV = right upper pulmonary vein; SVD = sinus venosus defect.

**TABLE 3 Procedural Outcomes**

| | |
|---|---|
| Procedural success | 97 (100) |
| Complications | |
| Embolization | 5 (5) |
| Surgical retrieval | 3 (3) |
| Overlapping stent to stabilize embolization | 2 (2) |
| Caudal migration | 11 (11) |
| Left innominate vein jailing | 2 (2) |
| Fabric breach | 4 (4) |
| New-onset atrial fibrillation | 1 (1) |
| Intracranial bleed; died after 3 weeks | 1 (1) |
| Residual leak immediate after the procedure | 97 |
| No | 38 (39.2) |
| Trivial (<3 mm color Doppler width) | 42 (43.3) |
| Mild (≥3 mm color Doppler width) | 17 (17.5) |

Values are n (%) or n.

when the stent was still around the stiff guidewire were successfully managed with additional overlapping transjugular anchoring stents (Figure 6). There were no delayed stent embolizations, vascular access complications, or major arrhythmias.

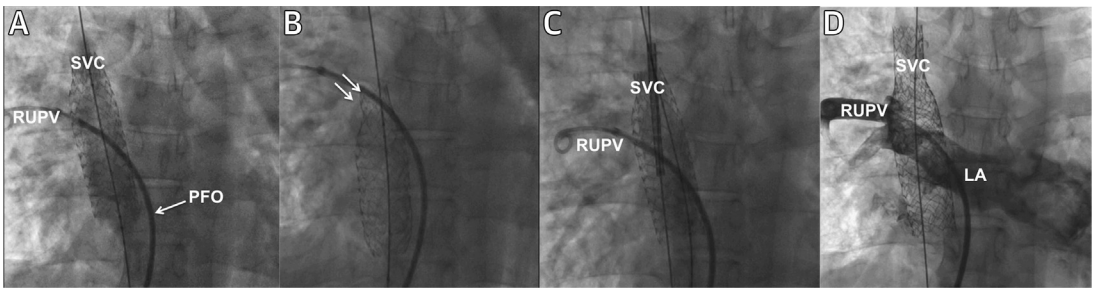
**Caudal stent migration.** The bell-bottom configuration of the cavoatrial junction predisposed to caudal milking of the stent in 9 patients, leading to a small residual flow from the RUPV into the SVC from the cranial end of the stent in 8 cases and inadequate SVC anchor in one case. Eight patients with residual flows were managed with an overlapping covered stent within the previous stent. An uncovered stent was overlapped in the patient with inadequate anchor.

**Fabric leak.** The integrity of the fabric of the covered stent was vital for procedural success in this intervention. The fabric leak manifested as a residual left-to-right shunt during a pulmonary venogram after complete deflation of the balloon, while it was not observed in the angiogram done with inflated balloon. When TEE confirmed fabric breach in 4 cases, they were managed with an additional overlapping covered stent. Two fabric tears in CP stents appeared as loose mobile structure on TEE.[15] The other 2 leaks in covered Zephyr stent occurred as 3- to 4-mm perforations.

**Transient caval obstruction.** Balloon interrogation as well as stent deployment within the SVC resulted in brief caval obstruction in every patient. Two patients developed brief generalized tonic-clonic seizures from cerebral venous hypertension during balloon occlusion, which recovered immediately without any residual sequelae. Three patients developed self-limiting facial petechiae and subconjunctival hemorrhage. Even though three-fourths of patients had a single SVC, the caval obstruction manifested only in these 5 patients.

**Left innominate vein jailing.** A short SVC reduced the anchor zone between the union of the 2 innominate veins and the cavoatrial junction (Figure 7). Two patients with a short SVC had jailing of the left innominate vein. The left innominate vein was recanalized successfully with a Brockenbrough needle puncture and balloon dilatation.[4,16]

**FIGURE 6** Managing Stent Embolization



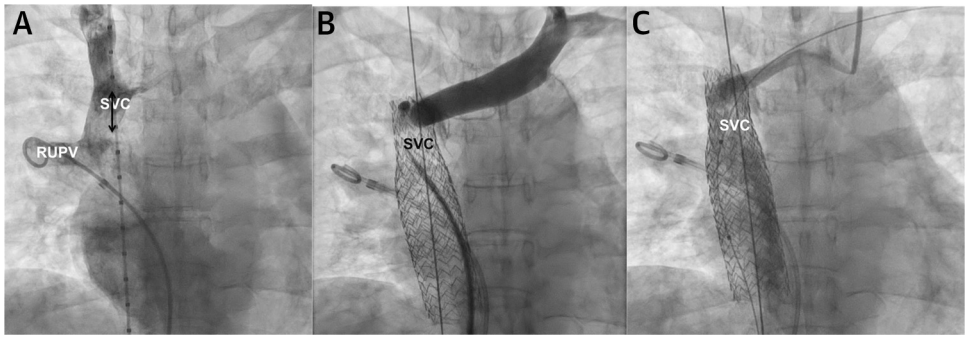

During expansion of a covered stent (A) in the SVC under RUPV monitoring, its caudal embolization (B) is identified by a lower stent position. A second covered stent from jugular vein is overlapped (C) and expanded to stabilize the embolized stent (D). A RUPV angiogram confirms successful redirection to the LA. Abbreviations as in Figure 1.

**INTRACRANIAL BLEEDING AND SHORT-TERM MORTALITY.** A 60-year-old man with a SVD had systemic hypertension, moderate hyperkinetic pulmonary hypertension, and heart failure. A fabric leak during intervention warranted an additional overlapping covered stent. A CT scan for persistent stupor and seizures after the procedure showed a subarachnoid hemorrhage. Despite gradual neurological recovery, he succumbed to pneumonia after 3 weeks in another institution.

**Follow-up.** Being a new procedure, follow-up was insisted upon and was complete in all patients (Table 4). The median follow-up duration was 19 months and the longest follow-up was 90 months. Fifty-six cases completed 1 year of follow-up. Imaging during follow-up was primarily with TEE in 80 patients, MRI in 3, CT scan in 4, and transthoracic echocardiogram in young children.

**Residual shunt.** Immediate periprocedural TEE or angiography showed residual shunt in 60.8% that was mild (>3 mm) in 17.5% and insignificant in the rest. The proportion with a residual shunt decreased at the 1- to 3-month follow-up to 42.7%, of whom 8.3% had a mild leak. After 1 year of follow-up, 30.4% had residual shunt with a mild leak in 5.4%. Residual shunt benefitted 4 patients with restrictive left ventricle physiology and 1 with significant pulmonary hypertension.[17,18] Reintervention for the largest residual shunt of 7 mm in 1 patient using a 14-12 Konar MFO (Lifetech Scientific) after 1 year was reported previously.[19] TEE was more sensitive than CT scan or MRI to identify clinically insignificant shunts. Spontaneous reduction of residual flows favor the hypothesis of right atrial remodeling resulting in improved stent apposition to the edges of the SVD.[5]

**FIGURE 7** Left Innominate Vein Jailing




When the SVC anchor zone (A) between the RUPV and the union of innominate veins is very small, the left innominate vein may become occluded (B). It is recanalized by Brockenbrough needle puncture and sidestrut dilatation (C). Abbreviations as in Figure 1.

**TABLE 4 Follow-Up Outcomes**

| | |
|---|---|
| Follow-up duration (months) | 19 (1-90) |
| Follow-up imaging modality | |
| Transesophageal echocardiography | 80 (83.3) |
| Only transthoracic echocardiography | 5 (5.2) |
| Magnetic resonance imaging | 3 (3.1) |
| Computed tomography scan | 4 (4.2) |
| Awaiting review | 4 (4.2) |
| Residual leak at 1 month follow-up | |
| No | 55 (57.3) |
| Trivial | 29 (30.2) |
| Mild | 8 (8.3) |
| Awaiting review | 4 (4.2) |
| Additional device closure for residual leak | 1 (1) |
| Residual leak at 1 year follow-up | 56 |
| No | 39 (69.6) |
| Trivial | 14 (25) |
| Mild | 3 (5.4) |
| Complications on follow-up | |
| Rhythm abnormality | 0 (0.0) |
| Pulmonary venous obstruction | 0 (0.0) |
| Thrombosis | 4 (4.2) |
| Duration of dual antiplatelet therapy (months) | 8 (6-20) |

Values are median (range), n (%), or n.

**Antiplatelet therapy.** Dual antiplatelet therapy was the routine postprocedural strategy in most patients. Novel anticoagulation was used in 3 patients, 2 after cardioversion of atrial fibrillation and another with fabric tear managed with a second covered stent. Antiplatelets were continued beyond 1 year for coronary atherosclerosis in 2 and previous stroke in 2 other patients.

**Thrombosis.** Asymptomatic thrombi were identified in 4 patients on a routine surveillance TEE. All patients had dual SVC drainage. The first one with a pedunculated nonocclusive thrombus in the atrial end of the stent at 6-month follow-up associated with a 7-mm residual shunt was reported previously.[19] Layered nonocclusive luminal thrombus in 3 other cases were located in the mid portion of the wider atrial segment and not in the narrow caval segment (Figure 8). One of the 31 patients who received covered Andrastents, 2 of the 40 patients where covered Zephyr stents were used, and 1 of the 52 patients who received covered CP stents were identified to have these asymptomatic thrombi. Although 3 of these thrombi occurred when we overlapped stents, 1 occurred in a patient with a single stent. There was no correlation with patient body surface area, stent length, or procedural time (Table 5). Factors that were significantly associated with thrombus were bilateral SVC compared with single SVC ($P < 0.001$) and use of >1 stent ($P = 0.027$). Immediate residual flow was also more often associated with thrombus, but it had a borderline significance ($P = 0.056$). None of the thrombi caused flow turbulence in the SVC and they uniformly resolved within 3 to 6 months on oral anticoagulation. The residual defect in the first patient was closed after 1 year using a Konar MFO device.

**PATIENTS AGED <12 YEARS.** Of the included patients, 9% were aged <12 years. Their median weight was 30 kg (range, 17-54 kg). The median interrogating balloon diameter was 18 mm (range, 16-22 mm). The stent length ranged from 59 to 69 mm. There was 1 intraprocedural stent embolization in our very early experience in 2015. The immediate surgical retrieval was uneventful. There were no vascular complications from the large bore access that ranged from 14-F to 18-F. There was no thrombus formation. Late follow-up imaging with MRI and CT scans after 4 or 5 years demonstrated complete closure without pulmonary vein narrowing (Figure 9).

## DISCUSSION

Surgery remained the only option for correcting SVD until a decade ago.[1] Despite different technical modifications including Warden anastomosis, SVC, or RUPV stenosis, sinus nodal injury and missed additional pulmonary veins were seen in few patients on follow-up.[1] Transcatheter closure emerged as a surgical alternative with growing experience.[2-4]

**PULMONARY VEIN MONITORING.** Complete closure of the SVD during interrogation and after stenting was shown by RUPV angiogram. It also confirmed redirection of the RUPV to the left atrium without increase of its pressure. This process removed the need for a continuous intraprocedural TEE and facilitated workflow in catheterization laboratory.[7] Earlier practice of direct RUPV cannulation through the SVD carried a risk of stent malposition during catheter removal and small residual shunt from stent nonapposition. Later, antegrade trans-septal cannulation was preferred as it additionally facilitated pulmonary vein protection.[5] Retrograde cannulation avoided the complications associated with trans-septal puncture, but provoked ventricular ectopics. Dual cannulation would permit pressure monitoring during pulmonary vein protection.

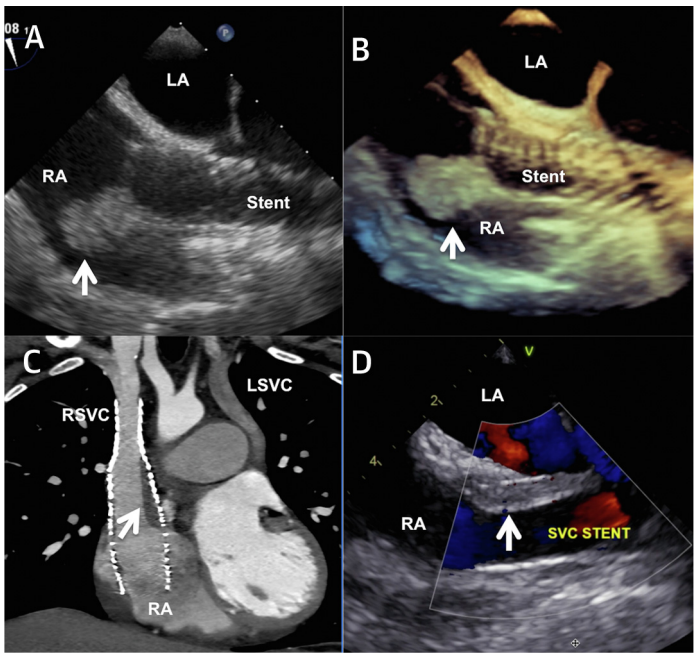
**APPLICABILITY IN YOUNG PATIENTS.** This intervention was rarely performed in children in earlier reports.[2,4,10,12] Children aged <12 years accounted for 9% of our cohort. Our smallest patient aged <5 years weighing 18 kg was the youngest reported in literature. Two major concerns were somatic SVC growth

with age and complications from large-bore vascular access. A firm circumferential contact of a stent deployed within a large vessel would arrest its further somatic growth.[20] A normal SVC diameter was 11.9 ± 3.4 mm in the pediatric age group, and adult cava measured 2.1 cm (range, 1.5-2.8 cm) and 1.7 cm (range, 1.0-2.4 cm) in the major and minor axis, respectively.[21,22] We included these children because the interrogating balloon diameter was 16 to 22 mm. This sizing provided adequate upper body venous drainage for the rest of the life. Insertion of a 14 F vascular introducer sheath to deploy a covered stent on a 16- to 20-mm balloon was technically feasible in patients aged >5 years. One stent embolization in a young child occurred early in our experience in 2015 requiring surgery. Follow-up imaging with CT scan, MRI, and transthoracic echocardiography in the subsequent children demonstrated the success of this intervention.

**REDUCING PROCEDURAL COMPLEXITY.** Procedural simplification would improve universal applicability especially in developing world.[4] Intubation anesthesia and continuous TEE monitoring was replaced by a brief conscious TEE imaging in limited patients with residual shunt on pulmonary venography. Pressure measurements and angiogram from the RUPV catheter provided similar information. Avoiding intubation benefitted high-risk patients with morbid obesity and pulmonary arterial hypertension. Instead of femoral-jugular venovenous circuit, extra stiff guidewires parked in brachial veins or jugular bulb provided adequate support. Preprocedural expensive 3-dimensional printed models and virtual reality were earlier favored instead of invasive balloon interrogation for patient selection.[23-25] However, our current experience of interrogation indicated feasibility of interventional closure in 94% of patients. Pulmonary vein protection and including patients with high-draining veins decreased the exclusions to a mere 4% in recent years.[5]

**CHOICE OF STENT AND BALLOONS.** The approximate stent length needed to completely occlude the SVD with an adequate anchor was found to be ≥7 cm in adults. A premounted long covered 10-zig CP stent was the universally available hardware for this intervention.[12] Foreshortening in the covered CP stent was unpredictable owing to variable diameters between their caval and atrial ends. A nonforeshortening long covered Zephyr stent was an attractive alternative.[13] When 2 stents were overlapped owing to nonavailability of a single long stent, the caudally placed stent was crimped outside the cranially placed stent in the same balloon making the latter grip the former after expansion.[4] An alternative cumbersome suturing of 2 stents was unsuccessful in one out of 3 patients.[26] Availability of longer semicompliant balloons ≤8 cm would make the procedure simpler and safer.

**FIGURE 8 Thrombus Formation in Covered Stents**



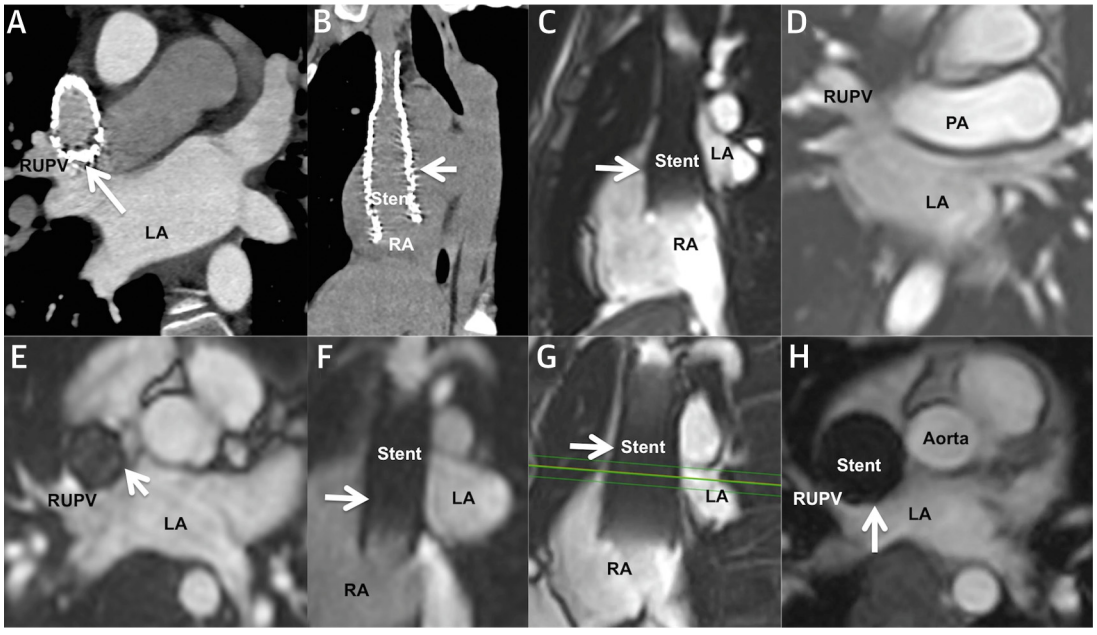

A pedunculated thrombus at the RA end of stent is seen in bicaval view (A) and 3-dimensional echocardiography (B). A layered luminal nonocclusive thrombus in another stent is visualized on a computed tomography scan (C) and transesophageal echocardiography (D).

**TABLE 5 Analysis of Factors Associated With Stent Thrombosis**

| | Total No. of Patients on Follow-Up (N = 96) | | |
|---|---|---|---|
| | No Thrombus (n = 92) | Thrombus (n = 4) | *P* Value |
| Male | 46 (50) | 3 (75) | 0.62[a] |
| Single SVC | 72 (78.3) | 0 (0) | <0.001[a] |
| Bilateral SVC | 20 (21.7) | 4 (100) | |
| BSA, $m^2$ | 1.8 (1.52-2.25) | 1.65 (0.74-2.23) | 0.277[b] |
| Stent length, mm | 75 (57-85) | 73 (57-81) | 0.35[b] |
| Stent number | | | 0.04[a] |
| 1 | 58 (63) | 1 (25) | |
| 2 | 26 (28.3) | 1 (25) | |
| 3 | 8 (8.7) | 2 (50) | |
| Immediate residual | 34 (37) | 2 (50) | 0.056[a] |
| Trivial | 45 (48.9) | 0 (0) | |
| Mild | 13 (14.1) | 2 (50) | |
| Procedural time, min | 90 (80-240) | 105 (55-210) | 0.964[b] |

Values are n (%) or median (range). [a]Fisher exact test. [b]Mann-Whitney *U* test.

**FIGURE 9 Follow-Up Imaging in Children**




Coronal (A) and modified (B) computed tomographic images 3 years after sinus venosus defect closure in a 10-year-old child; sagittal (C) and modified coronal (D) images of magnetic resonance imaging (MRI) performed after 4 years in a 5-year-old child; axial (E) and sagittal (F) MRI images performed 2 years after closure in a 10-year-old child; 3-year follow-up sagittal (G) and axial (H) MRIs of a large, 22-mm stent in an 8-year-old child. Abbreviations as in Figure 1.

**WIDENING THE HORIZON.** The feasibility of transcatheter closure improved to 94% after pulmonary vein protection and including patients with additional high-draining veins.[7] This rate was higher than in previous reports.[5] Trans-septal pulmonary vein protection was safe without complications in our experience. Protective balloon in RUPV inflated at 10 to 12 atm prevented its narrowing by the covered stent expanded at 3 to 4 atm.[5] Additional high-draining veins that were not amenable for a redirection were left to drain into the SVC during surgery. Transcatheter intervention adopted a similar strategy. Including patients with additional oval fossa defects also widened the horizon.[14]

**ACHILLES HEEL IN THIS INTERVENTION.** The caudal end of the stent failed to anchor in the wide right atrium owing to the bell-bottom shape of the cavoatrial junction, thus decreasing its stability. This might predispose to caudal migration or complete stent embolization. Five patients manifested embolization and few more had minimal caudal migration. Transjugular stabilization technique by a temporary suture involved complex procedural steps that preclude universal acceptability, but might benefit patients with a short SVC anchor.[10]

**FATE OF RESIDUAL LEAK.** Detection of a residual shunt is highly dependent on the sensitivity of the imaging tool. Transthoracic echocardiography and MRI used earlier were not sensitive to identify small residual shunts.[5] High spatiotemporal resolution of TEE identified all residual shunts on follow-up. This led to a higher detection of residual leak than other imaging modalities. A decrease in the number of patients with residual shunt as well as the quantum of shunt during follow-up owing to right atrial remodeling was documented in previous studies as well.[5] Small residual leaks would benefit adults with restrictive left ventricular physiology or pulmonary hypertension.[17,18]

**ANTIPLATELET MEDICATIONS.** Most operators advocated for dual antiplatelet therapy after this intervention.[5,9,10] Four patients with dual SVC drainage were identified on routine surveillance TEE to have asymptomatic nonocclusive thrombosis that resolved after oral anticoagulation.[19] Thrombi were identified in all types of stents, without any relationship to their length or diameter. Three of the 4 thrombi occurred in patients who received overlapping stents. None of the thrombi were seen in younger patients (<18 years of age). These thrombi would remain undetected in

the ferromagnetic artifacts of MRI and low-resolution transthoracic echocardiography, thus stressing the need for a high-sensitivity imaging tool such as TEE on follow-up.[19]

**STUDY LIMITATIONS.** Even though inclusion rates improved from 70% to 94%, a small proportion of patients might remain unsuitable for this nonsurgical intervention. The bell-bottom configuration of the cavoatrial junction decreased the stent stability, risking its caudal migration or embolization. Stent embolization remained the Achilles heel causing an element of uncertainty about the procedural success. Serial remodeling of the right atrial size after this transcatheter intervention and its impact on progressive reduction of the residual flows on follow-up should be studied in future.

## CONCLUSIONS

Transcatheter SVD closure carried a procedural success rate of 97%. Recent technical modifications decreased the procedural complexity, decreased its invasiveness, and increased patient inclusion without compromising the safety. Using long, premounted, covered stents simplified the procedure. Pulmonary vein protection and including patients with high-draining additional veins broadened the scope of this procedure. Residual shunts on follow-up were clinically insignificant and reduced over time. TEE was the ideal imaging to diagnose stent thrombosis and residual flows.

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## PERSPECTIVES

**COMPETENCY IN PATIENT CARE AND PROCEDURAL SKILLS:** Transcatheter closure of SVDs using covered stents is emerging as an alternative to surgical therapy.

**TRANSLATIONAL OUTLOOK:** Continued evolution of this technology should be coupled with research to clarify patient selection, type of stents, deployment techniques and imaging modalities to optimize long-term outcomes.

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