JOURNAL OF THE AMERICAN COLLEGE OF CARDIOLOGY VOL. 75, NO. 11, 2020
© 2020 BY THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION
PUBLISHED BY ELSEVIER

Transcatheter Correction of Superior Sinus Venosus Atrial Septal Defects as an Alternative to Surgical Treatment

Jan Hinnerk Hansen, MD, PhD,[a,b] Phuoc Duong, MD,[a,c] Salim G.M. Jivanji, MD,[a] Matthew Jones, MBBS,[a] Saleha Kabir, PhD,[a] Gianfranco Butera, MD, PhD,[a,c] Shakeel A. Qureshi, MD,[a] Eric Rosenthal, MD[a]

ABSTRACT

BACKGROUND The superior sinus venosus atrial septal defect (SVASD) is characterized by deficiency of the common wall between the superior vena cava (SVC) and the right upper pulmonary vein (RUPV), which is no longer committed to the left atrium.

OBJECTIVES This study sought to evaluate the potential for redirecting the SVC and RUPV flow to the right and left atria, respectively, by implantation of a covered stent in the SVC.

METHODS Review of 48 consecutive adult SVASD patients undergoing assessment for correction. Pre-procedural evaluation included cross-sectional imaging and ex vivo simulation using printed or virtual 3-dimensional models.

RESULTS Transcatheter correction was performed in 25 patients, with a further 6 awaiting stent implantation. Only 8 patients were deemed technically unsuitable. The procedure involved balloon test inflation in the anticipated stent landing zone with simultaneous transesophageal echocardiography and pulmonary venography to confirm defect closure and unobstructed pulmonary venous drainage, followed by deployment of a 10-zig covered Cheatham platinum stent. Stents of lengths between 5 and 8 cm were implanted. A second, uncovered stent was used for anchoring in 9 patients. The RUPV was protected with a high-pressure balloon during stent implantation to prevent pulmonary venous obstruction in 4 patients. The median follow-up period was 1.4 (interquartile range: 0.8 to 1.7) years, with no mortality. Stent embolization occurred in 1 patient; another required drainage of hemopericardium. Cardiac computed tomography after 3 months confirmed unobstructed pulmonary venous return. At latest follow-up, a residual shunt was present in 1 patient.

CONCLUSIONS Transcatheter correction of SVASD may be considered as an alternative to surgery in a substantial proportion of patients. (J Am Coll Cardiol 2020;75:1266–78) © 2020 by the American College of Cardiology Foundation.

The superior sinus venosus atrial septal defect (SVASD) is situated immediately inferior to the junction of the superior vena cava (SVC) and the right atrium (RA). It is usually associated with partial anomalous pulmonary venous drainage (PAPVD) of the right upper and/or right middle pulmonary veins (RUPV/RMPV). The defect results from deficient in-folding of the atrial wall that forms the posterior wall of the SVC and the anterior wall of the RUPV. The RUPV is no longer committed to the left atrium (LA) and drains into the RA at the mouth of the SVC (1,2). Surgical correction is the standard of care and, until recently, was the only treatment option (3,4). A novel transcatheter approach using a covered stent deployed in the SVC-RA junction was first published by Garg et al. (5) in 2014, having originally been presented during a conference in 2013 by Abdullah et al. (6). Since then, this technique has

Listen to this manuscript's audio summary by Editor-in-Chief Dr. Valentin Fuster on JACC.org.

From the [a]Department of Paediatric and Adult Congenital Heart Disease, Evelina London Children's Hospital, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom; [b]Department of Congenital Heart Disease and Paediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany; and the [c]School of Biomedical Engineering and Imaging Sciences, Kings College London, United Kingdom. Dr. Duong has been funded by/supported by the National Institute for Health Research Cardiovascular MedTech Co-operative. Dr. Qureshi is a consultant for NuMED. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Manuscript received October 21, 2019; revised manuscript received December 9, 2019, accepted December 30, 2019.

ISSN 0735-1097/$36.00 https://doi.org/10.1016/j.jacc.2019.12.070

been adapted and modified by others (7–11). The covered stent replaces the deficient posterior wall of the SVC, thereby closing the SVASD and redirecting the anomalous pulmonary veins into the LA behind the stent (**Central Illustration**).

We report a single-center experience of transcatheter correction of SVASDs and describe the evolution of an effective and reproducible transcatheter SVASD closure technique, including our early experience and subsequent developments (8,9).

PATIENTS AND METHODS

Transcatheter correction of SVASD and PAPVD by implantation of a covered stent was accepted as a new procedure by the Guy's and St Thomas' Trust Risk and Quality Assurance Committee in 2015. After the first procedure in March 2016, all adults (≥16 years of age) with an indication for surgical SVASD and PAPVD repair were also considered for transcatheter correction. All patients were formally discussed in the Adult Congenital Heart Disease multidisciplinary meeting. The indications for treatment included right heart volume overload and a pulmonary-to-systemic blood flow ratio >1.5:1, determined by cardiac magnetic resonance imaging (MRI). Other indications were considered on a case-by-case basis. Authorization to use custom-made stents in the patients was obtained through the unlicensed medicinal products program of the UK Medicines and Healthcare Products Regulatory Agency. When consent was obtained, all patients were made aware of the novel nature of this procedure.

SEE PAGE 1279

PRE-PROCEDURE EVALUATION AND PATIENT SELECTION. Suitability for transcatheter SVASD closure was assessed by a multimodality stepwise approach, including evaluation with 2-dimensional and 3-dimensional (3D) cross-sectional imaging and ex vivo simulation of stent implantation with printed or virtual 3D models. Diagnostic catheterization with balloon test occlusion, without proceeding to stent implantation, was performed in 3 patients early in the series.

CROSS-SECTIONAL IMAGING. Cardiac MRI or computed tomography (CT) images were reviewed to delineate the anatomy of the pulmonary venous return, and their anatomic relationship with the SVASD (**Figure 1A**). Smaller RUPV branches draining into the SVC remotely above the right atrial junction were not considered as a contraindication for transcatheter treatment, if the distance was thought to be too far for the vein(s) to be surgically redirected and the anticipated shunt volume based on the vein size was deemed to be hemodynamically insignificant.

PRINTED AND VIRTUAL 3D MODELS. Both cardiac CT and MRI datasets were used for 3D model segmentation and reconstruction (using the modality with the best image quality) to derive patient-specific 3D-printed models, as described in detail previously (8,9). The 3D models were printed in a hollow fashion with soft transparent material to allow simulation of the procedure and direct inspection of the results (**Figures 1B and 1C**).

For virtual simulation, the segmentation process was identical to the workflow of the physical 3D model but stopped at the production of the stereolithographic file. Predesigned stent prototypes with different shapes were used for virtual implantation (Rhino 6, Robert McNeel & Associates, Seattle, Washington). To plan transcatheter treatment, various stent dimensions and shapes could be fitted into the patient-specific hollow heart model (Meshmixer 3.3, Autodesk Inc., San Rafael, California). After virtual stent placement, potential areas of residual shunt (e.g., an SVC-RA junction larger than the achievable maximum diameter of the covered stent) or obstruction to the pulmonary venous pathway behind the stent were assessed by direct visualization or 3D cropping (**Figure 1D**).

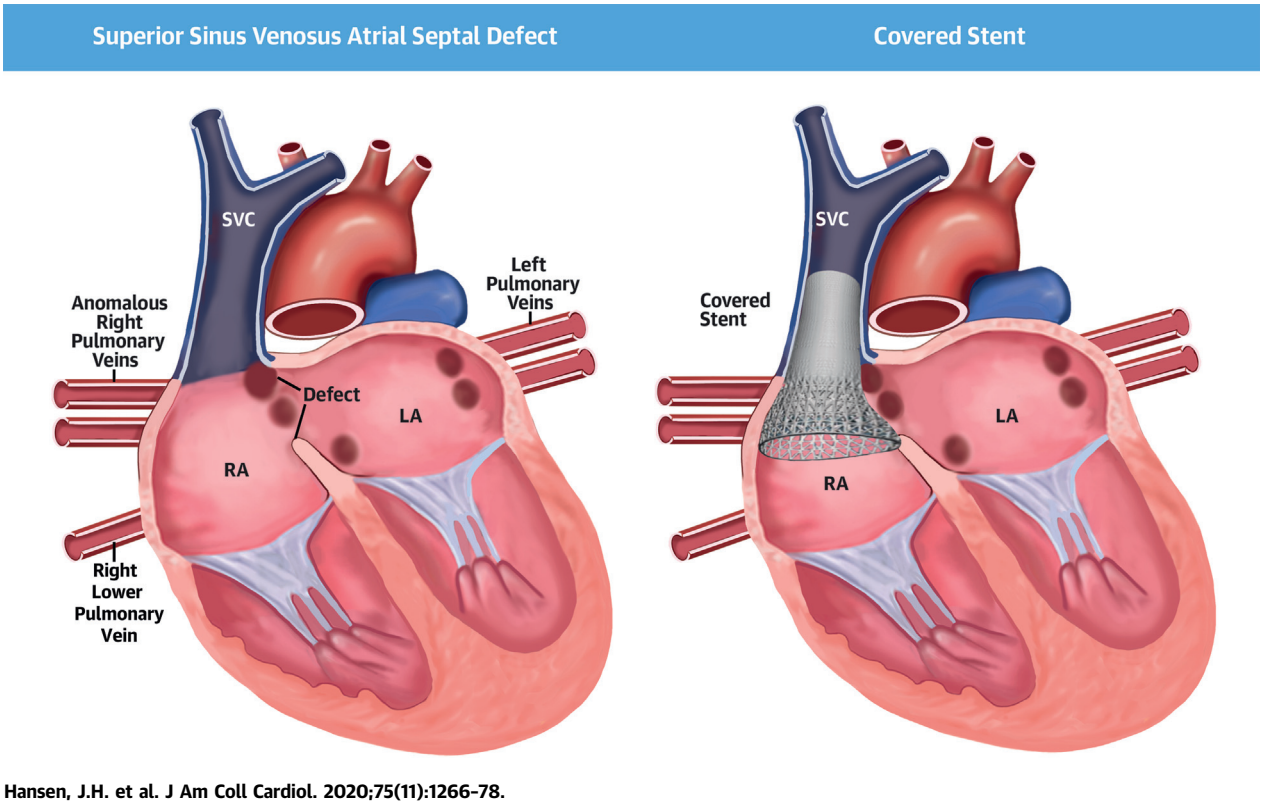
CATHETER PROCEDURE TECHNIQUE. All procedures were performed under general anesthesia with transesophageal echocardiographic (TOE) guidance. Vascular access was obtained in the right internal jugular vein (RIJV), the left and right femoral veins, and the left femoral artery. Heparin was given to maintain an activated clotting time of >250 s. Flucloxacillin was administered for antibiotic prophylaxis.

A 5-F pigtail angiographic catheter was placed in the RUPV to allow serial venography and pressure monitoring. In our earlier experience, the RUPV was entered retrogradely from the left ventricle (8,9). Subsequently, a transseptal puncture was performed using a Brockenbrough transseptal needle (St. Jude Medical, St. Paul, Minnesota) in an 8-F SRO Sheath (St. Jude Medical), allowing simultaneous LA and RUPV monitoring via the transseptal sheath and a 5-F pigtail catheter passed through the sheath into the RUPV. A veno-venous guidewire circuit was established, with an 0.035-inch Amplatz Extra Stiff wire (Cook Medical, Bloomington, Indiana) from the right femoral vein to the RIJV,

ABBREVIATIONS AND ACRONYMS

3D = 3 dimensional
BiB = balloon-in-balloon
CP = Cheatham platinum
CT = computed tomography
IQR = interquartile range
LA = left atrium
MRI = magnetic resonance imaging
PAPVD = partial anomalous pulmonary venous drainage
RA = right atrium
RIJV = right internal jugular vein
RMPV = right middle pulmonary vein
RUPV = right upper pulmonary vein
SVASD = sinus venosus atrial septal defect
SVC = superior vena cava
TOE = transesophageal echocardiography

CENTRAL ILLUSTRATION **Transcatheter Correction of Superior Sinus Venosus Atrial Septal Defects**



Hansen, J.H. et al. J Am Coll Cardiol. 2020;75(11):1266–78.

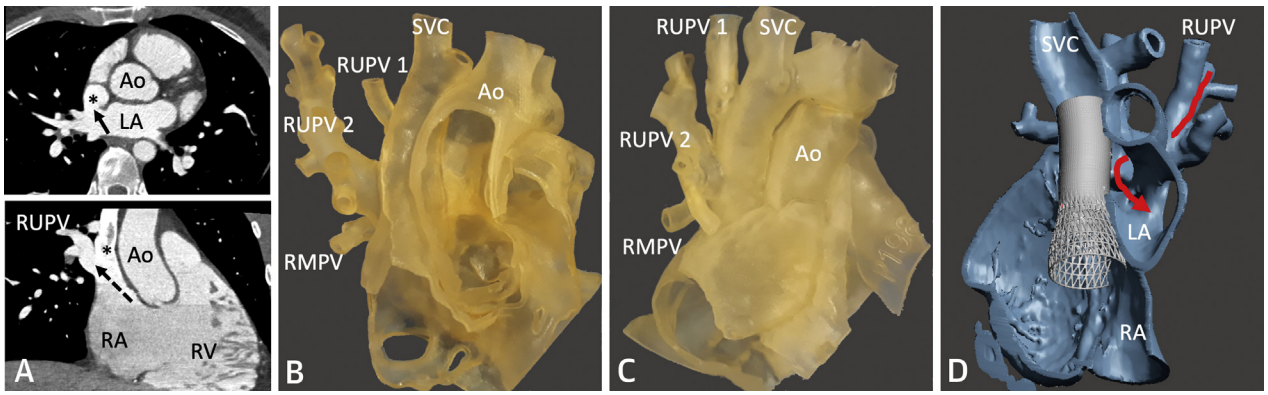
The diagram demonstrates how the covered stent is able to simultaneously close the atrial septal defect and redirect the pulmonary venous drainage to the left atrium (LA) behind the covered stent. The flared stent portion in the right atrium (RA) ensures occlusion of the interatrial communication. SVC = superior vena cava.

to facilitate balloon test occlusion and stent implantation.

Balloon testing was performed in all patients. A 24-mm or 34-mm Amplatzer sizing balloon (St. Jude Medical) or 6-cm-long AltoSa-XL PTA balloons (AndraTec GmbH, Koblenz, Germany) of 16- to 30-mm diameter were used. The initial balloon size was chosen to be at least 2 to 4 mm larger than the SVC diameter. The balloon was inflated in the anticipated stent landing zone, until TOE confirmed elimination of the interatrial shunt. An SVC angiogram was obtained to document SVC flow occlusion. During balloon inflation, RUPV flow was assessed by TOE, angiography from the RUPV catheter, and, simultaneous, RUPV and LA pressure measurements.

Pre-procedural imaging data and measurements obtained during balloon testing were used to choose the stent length and the appropriate balloon diameter for implantation. The stent length was selected based on having at least 2 cm of the unexpanded stent in the SVC and 2 cm protruding into the RA below the entrance of the pulmonary veins. The balloon diameter was chosen to be 2 to 4 mm larger than the balloon interrogated SVC diameter. For all patients, 10-zig covered Cheatham platinum (CP) stents in 5- to 8-cm lengths mounted on balloon-in-balloon (BiB) catheters with outer balloon diameters between 18 and 30 mm (NuMED, Hopkinton, New York) were used. The stents were introduced through a long delivery sheath (18-F Check-Flo Performer, Cook Medical, Bloomington, Indiana, or 20-F DrySeal, Gore, Flagstaff, Arizona) from the right femoral vein. The stents were deployed using simultaneous TOE imaging and angiography from the RIJV and RUPV. The cranial end was intended to sit sufficiently high in the SVC to allow secure anchoring while the caudal end of the stent was well below the inferior rim of the defect on TOE. Flaring of the RA portion of the stent to ensure complete coverage of the interatrial communication was performed with the outer balloon of the BiB, which did

FIGURE 1 Pre-Procedural Assessment With Cardiac CT and 3D Printed or Virtual Models



(A) Cardiac CT showing the SVASD **(solid arrow, upper panel)** at the posterior wall of the SVC **(*)** and the anterior wall of the right-sided pulmonary veins. The RUPV drains into the SVC **(dotted arrow, lower panel)** at the SVC-RA junction. **(B)** Printed 3D model showing a small RUPV 1 draining remotely into the high SVC and a large RUPV 2 potentially redirectable with a stent. The patient proceeded to stent implantation, leaving RUPV 1 draining into the SVC. **(C)** Printed 3D model showing a large RUPV 1, which is unable to be redirected with a stent while it is amenable to surgical redirection. The patient proceeded to surgical repair. **(D)** Virtual 3D model (sagittal view, en face to the sinus venosus defect) visualizing redirection of the RUPV toward the LA behind the stent **(red arrow)**. The flared RA portion ensures occlusion of the interatrial communication. 3D = 3 dimensional; Ao = aorta; CT = computed tomography; LA = left atrium; RA = right atrium; RMPV = right middle pulmonary vein; RUPV = right upper pulmonary vein; SVASD = superior sinus venosus atrial septal defect; SVC = superior vena cava.

not necessarily achieve its maximum diameter in the SVC. A Coda Balloon (Cook Medical, Bloomington, Indiana) was used if additional flaring was required.

A high-pressure balloon (Atlas PTA Balloon, Bard Peripheral Vascular, Tempe, Arizona) was inflated in the RUPV to prevent obstruction of the pulmonary venous return behind the stent when this was considered to be at risk of compression during deployment or flaring of the stent.

If apposition of the stent to the SVC appeared to be insufficient (length of <1.5 cm or instability noted), an additional bare stent was placed overlapping the cranial end of the covered stent. Additional covered stents were implanted for residual shunts only.

Final angiograms from the SVC and RUPV; pressure measurements in the RUPV, LA, and RA; and TOE were used to assess for residual shunting and absence of pulmonary venous compression.

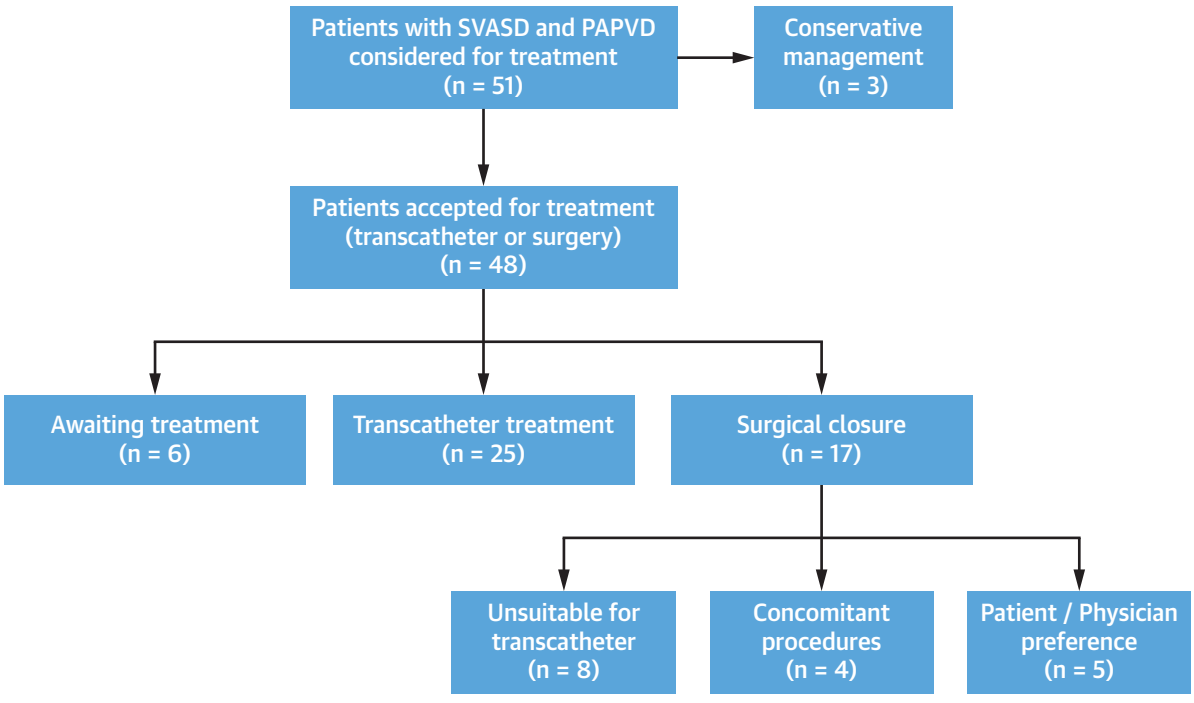
POST-PROCEDURAL MANAGEMENT AND FOLLOW-UP. Dual antiplatelet therapy was commenced with clopidogrel 75 mg and aspirin 75 mg for 2 months, followed by aspirin alone for a further 4 months. Patients receiving anticoagulant agents continued receiving them (without additional antiplatelets) for at least 6 months. Routine post-procedural investigations included posterior-anterior and lateral chest x-ray and transthoracic echocardiography the day after the procedure. Clinical follow-up with transthoracic echocardiography and 12-lead electrocardiography was scheduled at 3 and 12 months and at yearly intervals thereafter. A cardiac CT was performed after 2 to 3 months to confirm stent position and exclude pulmonary vein obstruction. A cardiac MRI after 1 year was used to assess right heart remodeling and to quantify any residual shunting.

DATA COLLECTION AND STATISTICAL ANALYSIS. Data were collected from the institution's Adult Congenital Heart Disease multidisciplinary meeting records and patients' medical records. Continuous variables are expressed as median (interquartile range [IQR]) and categorical data as counts and percentages. Continuous variables were compared with the Mann-Whitney *U* test or with the Wilcoxon signed rank test for repeated measurements. All statistical analyses were performed with the statistical software package SPSS version 25.0 (IBM Corp., Armonk, New York). A value of $p < 0.05$ was considered statistically significant.

RESULTS

PATIENT POPULATION. Between March 2016 and September 2019, 48 consecutive patients with SVASD and PAPVD were accepted for treatment after review in the Adult Congenital Heart Disease multidisciplinary meeting (Figure 2). The anatomy was considered suitable for a transcatheter approach in 40 (83.0%) patients, based on evaluation of cross-sectional imaging and ex vivo simulation with 3D printed or virtual models (Figure 1). Factors

FIGURE 2 Patients Assessed for SVASD Treatment



Transcatheter treatment was performed in 25 patients, and 6 were awaiting stent implantation. A total of 17 patients were referred to surgery—only 8 were deemed unsuitable for a transcatheter approach. PAPVD = partial anomalous pulmonary venous drainage; SVASD = superior sinus venosus atrial septal defect.

precluding transcatheter correction included pulmonary veins draining directly into the SVC high above the right atrial junction that could not be redirected by the stent but would be amenable to surgical redirection (Figure 1C). In 2 patients initially thought to be suitable, pulmonary vein obstruction occurred during balloon testing, and stenting was not performed (Figure 3).

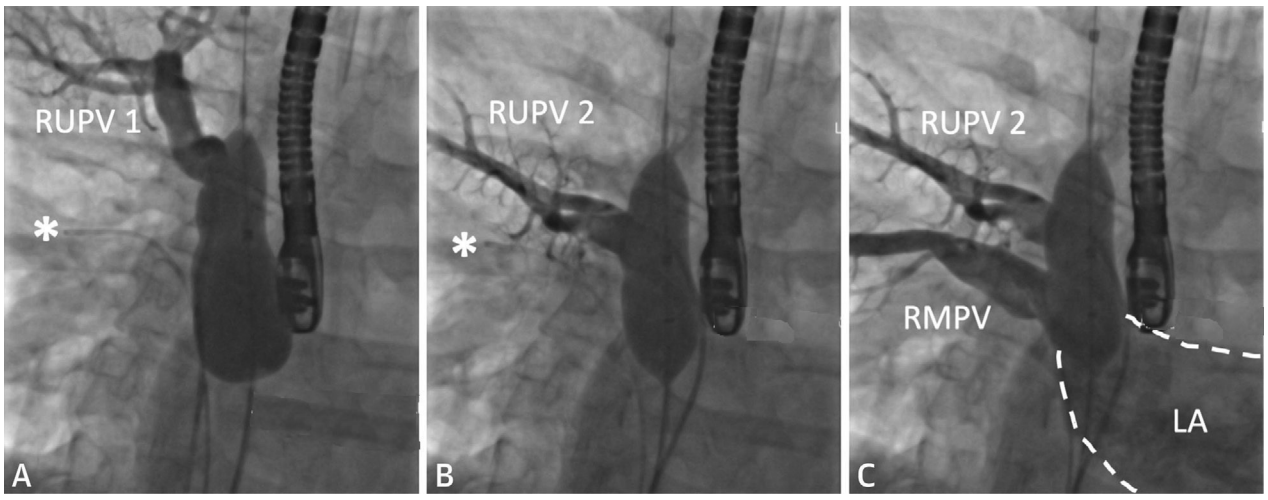
Transcatheter correction was performed in 25 patients, and a further 6 await stent implantation. The remaining 17 patients were referred for surgery. Concomitant procedures were required in 4 patients (tricuspid valve annuloplasty and maze procedure, implantable cardioverter-defibrillator lead removal from the left ventricle, placement of an epicardial pacemaker, and coronary artery bypass grafting). Although potentially suitable, these patients were not considered for transcatheter treatment. In 5 patients, surgery was the patient's or referring physician's preference (Figure 2).

PATIENT CHARACTERISTICS. Characteristics of the 25 patients who underwent transcatheter correction are summarized in Table 1. Right ventricular volume overload was the indication for closure in all but 2 patients. In 1, the need for a transvenous pacemaker was the indication—the pacing lead consistently entered the left side of the heart and revealed the diagnosis of a residual SVASD after previous surgical repair. The second patient was a professional diver presenting with recurrent episodes of decompression illness.

Bilateral SVCs were present in 8 (32.0%) patients, and smaller RUPV branches draining into the SVC high above the right atrial junction were present in 6 (24.0%).

Pre-procedural planning involved ex vivo simulation using a printed 3D model in 18 (72.0%) patients and a virtual model in 5 (20.0%). Transcatheter correction was performed relying on cross-sectional imaging alone in 4 patients. Diagnostic cardiac catheterization with balloon testing was performed in 6 patients. In 3 of these, the intention was to implant a stent. However, it was apparent that longer custom-made stents on larger BiB catheters were needed than had been anticipated. The patients subsequently returned for the definitive procedure.

FIGURE 3 Balloon Test Occlusion



Balloon testing with Amplatzer sizing balloon and selective pulmonary vein angiography. Two right-sided upper pulmonary veins (RUPV 1 and RUPV 2) are occluded with the balloon inflated **(A,B)**, whilst the RMPV (* tip of the catheter in RMPV) is diverted into the left atrium **(dotted line) (C)**. The patient underwent surgical repair. Abbreviations as in Figure 1.

PROCEDURAL OUTCOME. Procedural data are given in Table 2. The median age at the procedure was 46 years (range 18 to 66 years; IQR: 33 to 54 years). RUPV access was established with a retrograde arterial approach in the first 10 (40.0%) patients. The transseptal route after needle puncture was subsequently chosen in all except 2 patients with a patent foramen ovale. In the majority, a 6-cm-length 10-zig covered CP stent was implanted (Table 2). The caudal end of the stent was flared to a median diameter of 28 mm (IQR: 26 to 32 mm). The unflared portion in the SVC measured 19 mm (IQR: 16-20 mm).

Balloon testing was suggestive of potential pulmonary venous obstruction in 4 patients (Figure 4), and a high-pressure balloon (16-mm Atlas Gold, n = 3; 12-mm Atlas Gold, n = 1) was inflated in the RUPV to avoid compression during deployment and flaring of the stent (Figure 5).

Additional stents were placed during 13 (52.0%) procedures. In 9 patients, a bare CP stent was used to anchor the covered stent in the SVC (Figure 6). Stent migration toward the RA, especially during flaring of the caudal portion of the stent, had been noted in 5 of these 9 patients. Stent instability was associated with a shorter stent segment apposed to the SVC (distance

TABLE 1 Patient Characteristics

Male/female	14/11
Age, yrs	45 (33-54)
Weight, kg	74 (64-83)
Height, cm	169 (168-178)
cMRI Qp:Qs	2.5 (2.1-2.6)
RVEDVi, ml/m^2	153 (128-173)
Bilateral SVC	8 (32)

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

cMRI Qp:Qs = pulmonary-to-systemic blood flow ratio derived by cardiac magnetic resonance imaging; RVEDVi = indexed right ventricular end-diastolic volume; SVC = superior vena cava.

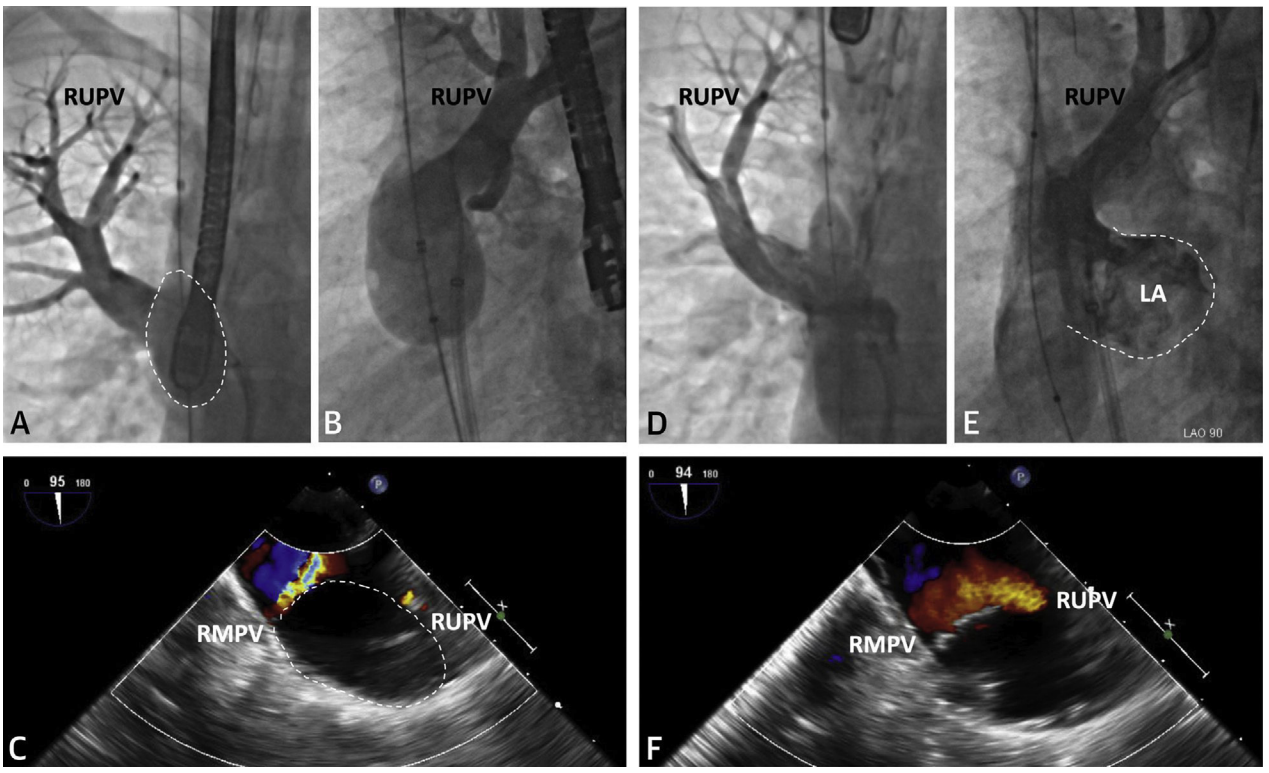
TABLE 2 Procedural Variables

Covered CP stent length, cm	
5	1 (4.0)
6	14 (56.0)
7	6 (24.0)
8	4 (16.0)
BiB diameter, mm	28 (24-28)
Additional stents, n	13 (54.2)
RA pressure pre-stent, mm Hg	9 (8-10)
RA pressure post-stent, mm Hg	8 (6-9)
RUPV pressure post-stent, mm Hg	15 (12-19)
LA pressure post-stent, mm Hg	14 (12-17)
RUPV to LA gradient post-stent, mm Hg	1 (0-2)
Procedure time, min	190 (162-222)
Fluoroscopy time, min	42 (35-54)
Dose-area product, cGy · cm^2	7,167 (3,658-11,564)

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

BiB = balloon-in-balloon; CP = Cheatham platinum; LA = left atrium; RA = right atrium; RUPV = right upper pulmonary vein.

FIGURE 4 Balloon Test Occlusion With 2 Different Types of Balloons



(A,B) Inflation of the Amplatzer sizing balloon resulted in obstruction of the RUPV. **(C)** TOE (high esophageal bicaval view with clockwise rotation) confirmed RUPV obstruction and also showed RMPV compression. **(D, E)** During repeat balloon testing with the AltoSa-XL balloon, pulmonary venography confirmed drainage to the LA **(dotted line)**. **(F)** TOE showed no compression to the RMPV but accelerated flow in the RUPV—suspicious for potential obstruction with stenting. RMPV = right middle pulmonary vein; TOE = transesophageal echocardiography; other abbreviations as in Figure 1.

from the cranial stent end to the start of the flared segment): 17 mm (IQR: 12 to 20 mm) versus 21 mm (IQR: 18 to 24 mm) ($p = 0.030$). In 1 patient with an SVC segment of 1.5 cm, the stent embolized shortly after the procedure (Figure 7). In 4 patients, a second covered CP stent was implanted due to a residual shunt. In the professional diver, the patent foramen ovale was closed with an 18-mm Figulla Flex II atrial septal defect occluder (Occlutech International AB, Helsingborg, Sweden) to ensure complete abolition of interatrial shunting.

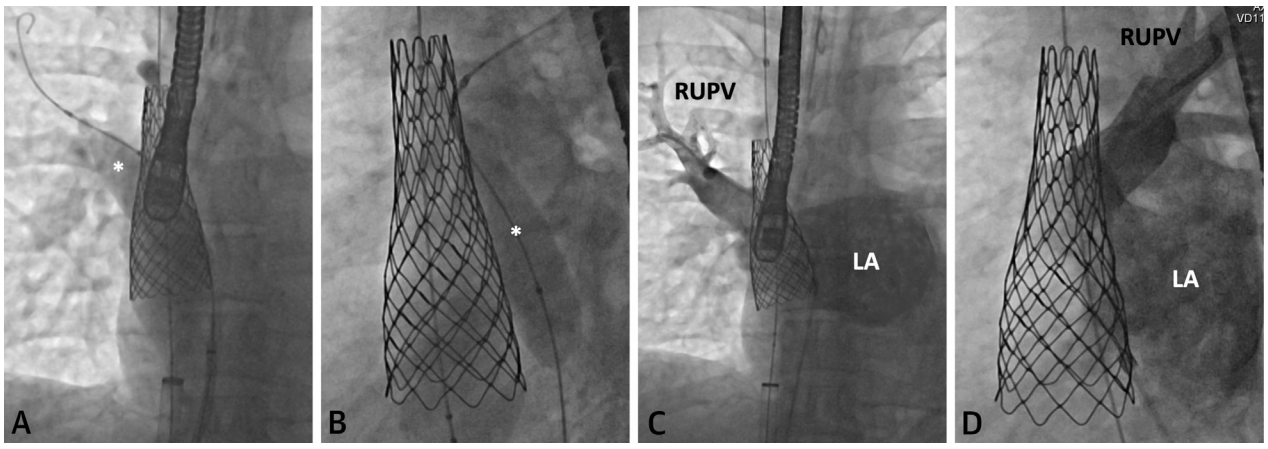
A rise in RUPV and LA pressure was noted during balloon testing and persisted in the majority of patients after defect closure. Pulmonary venous return at the end of the procedure was unobstructed in all the patients on pulmonary venography and Doppler echocardiography. There was no RUPV-to-LA gradient (Table 2).

A residual shunt was present in 11 (44.0%) patients by angiography and in 15 (60.0%) with color Doppler echocardiography. In only 1 case was the leak graded as significant, and a small residual gap at the inferior margin of the stent was noted but deemed to be acceptable. The following day, 6 (24.0%) patients still had a detectable shunt on the transthoracic echocardiogram.

POST-PROCEDURAL COMPLICATIONS. Serious adverse events occurred in 2 patients. One developed a hemopericardium 3 days after the procedure, requiring surgical drainage. At surgery, there was no perforation at the site of SVC stent. The hemopericardium was presumed to be related to the transseptal puncture. In the other, the SVC stent embolized within the first hours after the procedure (Figure 7). The patient underwent uncomplicated surgical stent removal and patch closure of the SVASD.

Other complications included a femoral artery pseudoaneurysm requiring thrombin injection after difficult retrograde access to the pulmonary veins that also raised a trabeculation flap in the left

FIGURE 5 Protection of the Pulmonary Venous Pathway to Prevent Obstruction



Same patient as in Figure 4. **(A, B)** Protection of the RUPV with Atlas Gold balloon **(*)** during stent deployment and flaring, preventing protrusion of the stent into the pulmonary venous pathway. **(C, D)** Pulmonary vein angiography confirms unobstructed flow to the LA. Abbreviations as in Figure 1.

ventricle, atrial fibrillation treated with cardioversion, and transient arm neuropraxia.

POST-PROCEDURAL FOLLOW-UP. The median follow-up period was 1.4 years (IQR: 0.8 to 1.7 years), ranging from 5 months to 3.8 years. Cardiac CT was performed in 22 patients. Another underwent angiography during pacemaker implantation 6 weeks after the procedure. The SVC stent was well positioned and patent, with unobstructed pulmonary venous drainage (Figure 8).

Transthoracic echocardiography at the latest follow-up showed qualitative reduction in right ventricular size in all cases. The professional diver had no detectable shunt on contrast echocardiography 3 months after the procedure and was cleared to return to diving by the diving authorities. Cardiac MRI was available in 14 of 17 patients with a 1-year follow-up visit and confirmed a significant reduction of indexed right ventricular end-diastolic volume (Figure 9). Overall, there was no significant residual shunt, with a pulmonary-to-systemic blood flow ratio of 1.10:1 (IQR: 1.10 to 1.15:1). Only the patient with the small residual gap at the inferior margin of the stent had a discernible residual shunt on MRI. This patient also had an additional RUPV draining into the high SVC that was not redirected. The pulmonary-to-systemic blood flow ratio decreased from 3.9:1 to 2.6:1 after stent placement.

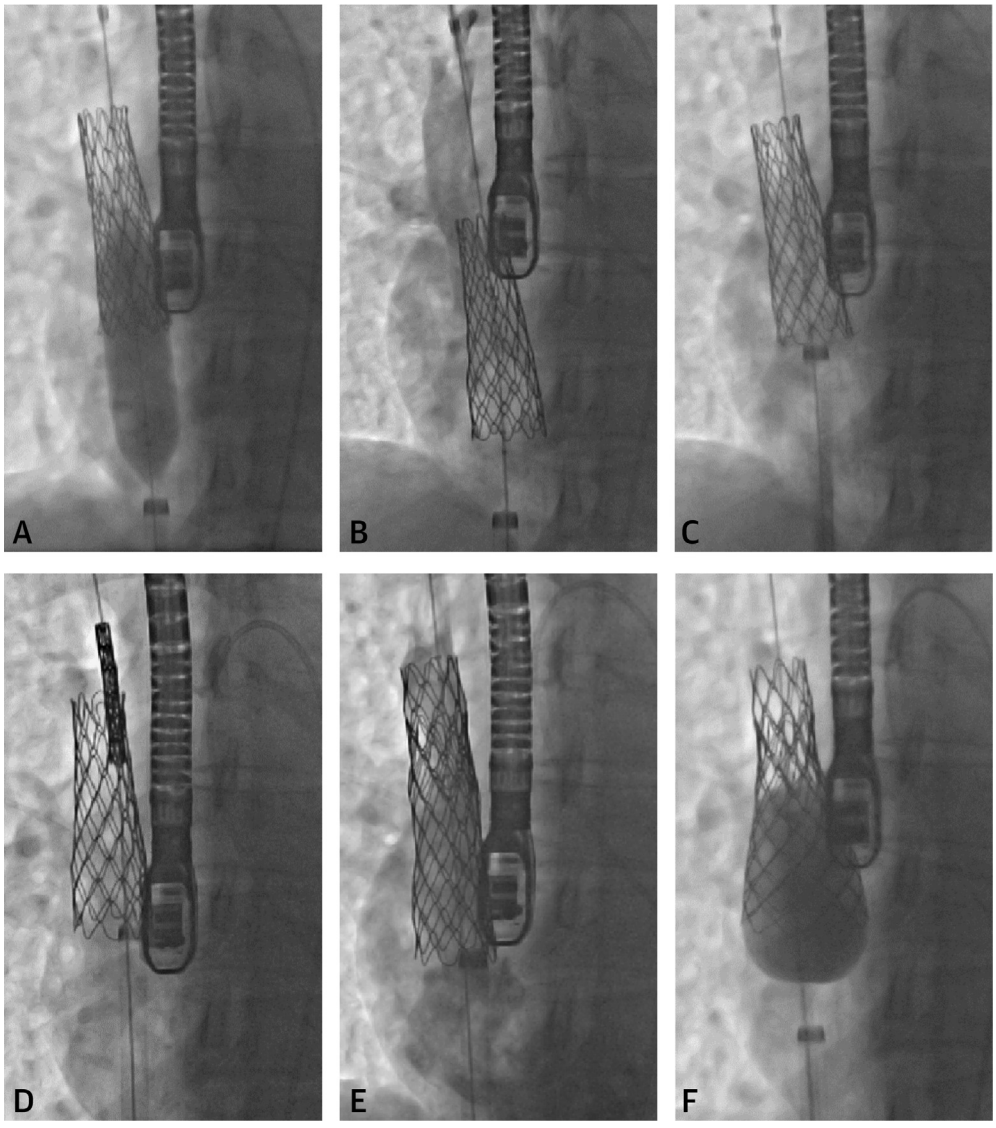
No late complications have been observed. A transvenous dual-chamber pacemaker was electively implanted through the stent 6 weeks after stent implantation in the patient in whom lead placement had failed before the procedure. At the latest follow-up, sinus rhythm was present in all remaining patients with no signs of sinus node dysfunction.

DISCUSSION

Comprehensive pre-procedure planning with advanced segmentation of cross-sectional imaging and 3D reconstruction, together with refinements in procedural techniques, have allowed transcatheter correction of SVASD to evolve into a reproducible procedure in our institution. Since the start of our experience, >75% of patients referred for SVASD treatment appear to be technically suitable for transcatheter correction. This technique may therefore become an alternative to surgery in the majority of patients with SVASD.

PRE-PROCEDURAL IMAGING AND PATIENT SELECTION. An in-depth understanding of the anatomy is essential to assess the likelihood of successful SVASD closure without compromising pulmonary venous return. A prerequisite for closing the interatrial communication and redirecting pulmonary venous flow is that the posterior wall of the anomalous pulmonary vein is either in direct continuity with the left atrium or another pulmonary vein draining into the LA. Initially, we assessed for suitability using 3D-printed models and ex vivo stent implantation, but more recently, we have simulated the procedure only in a virtual 3D model (8–11). With high-quality 3D CTs, we also have been able to proceed without creating a model.

FIGURE 6 Anchoring Stent

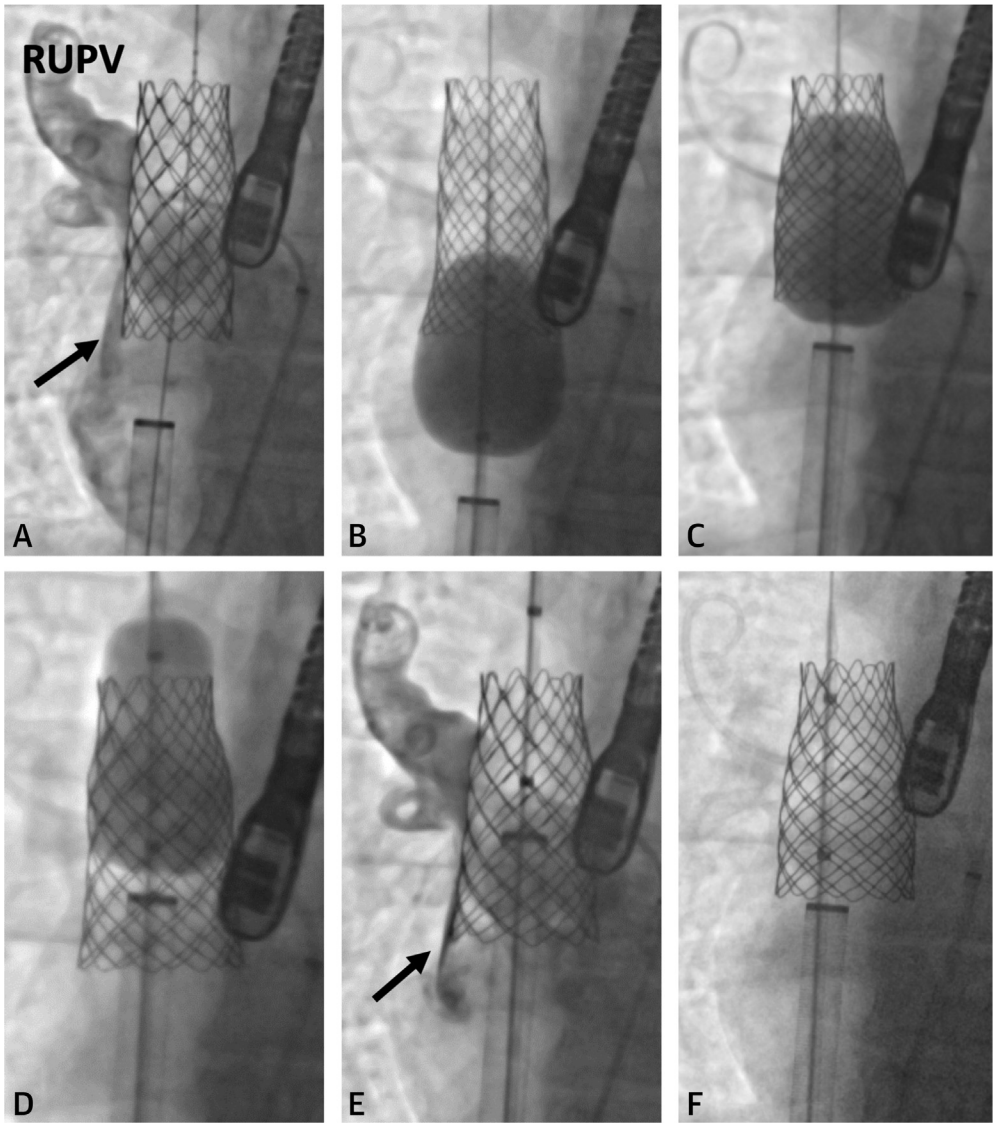


(A) Flaring of the inferior stent portion results in **(B)** caudal displacement. **(C)** The stent is readvanced into position with the delivery sheath. **(D, E)** An additional stent is passed from the jugular vein to overlap the covered stent, allowing **(F)** stable flaring of the covered stent.

In those patients referred for surgery, the underlying anatomy was unsuitable for transcatheter correction in one-half of the patients. Large pulmonary veins draining exclusively into the SVC well above the right atrial junction that were amenable to surgical redirection, or the potential for pulmonary venous compression with stenting, made the transcatheter approach unfeasible. Large defects with extension into the atrial septum may also preclude transcatheter closure (12). The need for concomitant procedures or patient or physician preference accounted for the remaining surgical cases in our cohort.

IN VIVO SIMULATION/BALLOON TEST OCCLUSION. In vivo simulation by balloon inflation in the SVC-RA junction before stent implantation is important to evaluate the potential for pulmonary venous obstruction and to guide the choice of balloon size for stent implantation. Distention of the native biological tissues is difficult to predict accurately in printed or virtual 3D models. Pulmonary venous obstruction

FIGURE 7 Insufficient Stent Apposition in the SVC

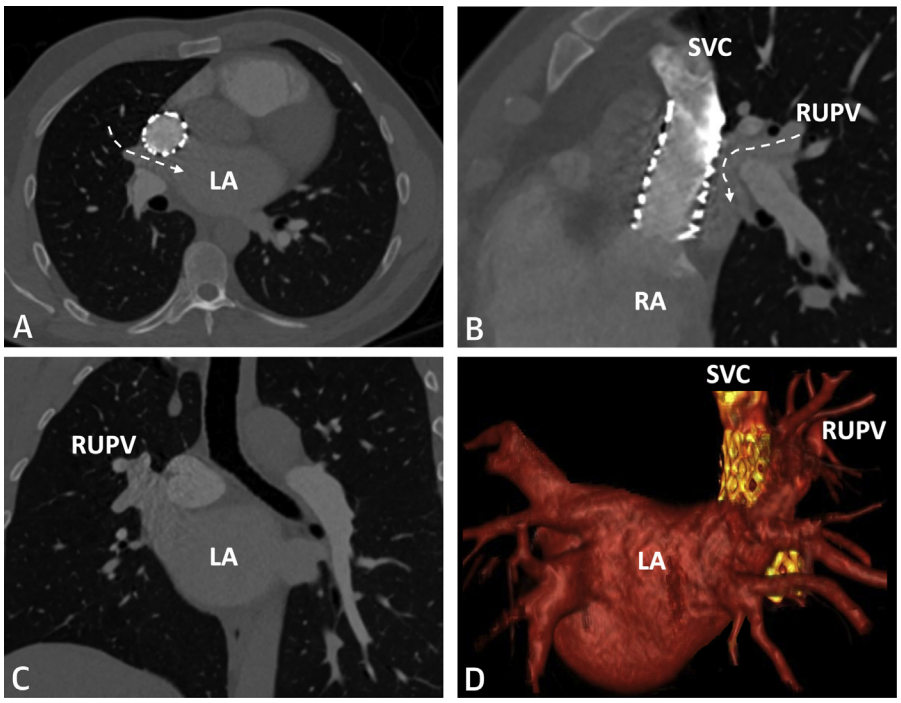


(A) Presence of a small residual leak **(black arrow)** after implantation of a 7-cm-long stent. Flaring of the **(B)** inferior, **(C)** middle, and **(D)** superior portion of the stent **(E)** reduced the leak **(black arrow)**, but this resulted in shortening of the stent to 5 cm. **(F)** The zone of apposition to the SVC was only 1.5 cm. This stent embolized a few hours after the procedure. Abbreviations as in Figure 1.

became apparent only on balloon testing in 2 patients who had been thought to be suitable based on preprocedural imaging and modeling. The choice of balloons is important. Compliant sizing balloons can simulate flaring of the RA portion of the stent to achieve complete occlusion of the defect but may also be overinflated, causing pulmonary venous obstruction (Figure 4). When this occurs, we re-evaluate pulmonary venous drainage using noncompliant balloons 2 to 4 mm larger than the SVC, accepting the likelihood of a residual shunt on imaging because the fixed-diameter balloon will not fully simulate the required flaring.

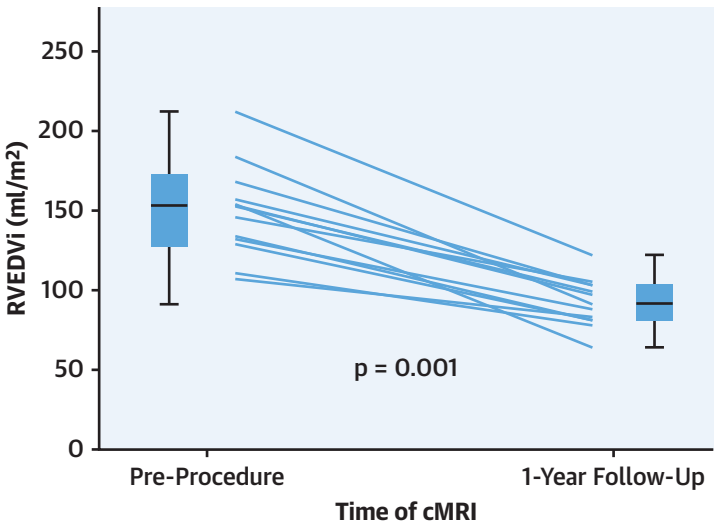
MONITORING OF PULMONARY VENOUS RETURN. To avoid pulmonary venous compression, we monitor pulmonary venous pressure and flow. Instead of entering the RUPV through the sinus venosus defect via a transvenous approach, we initially chose a retrograde arterial approach to avoid interference with stent placement. However, the retrograde arterial route may be challenging, resulting in long

FIGURE 8 Follow-Up With Cardiac CT



Cardiac CT in the **(A)** axial and **(B)** sagittal views shows patent SVC stent and unobstructed redirected RUPV **(dotted line)** draining into the LA posterior to the SVC stent. **(C)** In the coronal view, the RUPV to LA continuity is visualized with the stent out of plane. **(D)** A 3D volume rendered reconstruction (posterior coronal view) showing pulmonary venous return behind the stent. Abbreviations as in Figure 1.

FIGURE 9 Remodeling of the Right Ventricle



Significant reduction of the indexed right ventricular end-diastolic volumes (RVEDVi) derived by cardiac magnetic resonance imaging (cMRI) confirming remodeling of the right ventricle after transcatheter treatment.

procedure and screening times. In subsequent patients, we modified the technique to access the LA and pulmonary vein by a transseptal approach. This allows continuous simultaneous monitoring of LA and RUPV pressures to detect gradients and places less reliance on repeat pulmonary vein angiograms. Furthermore, accessing the RUPV from the LA may facilitate bail-out balloon dilation or even stenting in the event of unforeseen RUPV obstruction. In 4 patients with pulmonary vein obstruction during balloon test occlusion, protection of the pulmonary vein with an Atlas Balloon during stent deployment and flaring allowed successful transcatheter correction. Transseptal puncture, however, is not without risk, and 1 patient experienced hemopericardium and cardiac tamponade. TOE imaging plays an integral part in the procedure, both for guiding transseptal puncture and for monitoring for residual defect and pulmonary vein obstruction during balloon testing, stent implantation, and post-deployment flaring.

CHOICE OF STENTS. The 6-cm 10-zig covered CP stent, used in the majority of our patients, is

currently the longest balloon-expandable stent with a CE mark. The stent can be inflated to a nominal diameter of 30 mm and up to 35 mm, which may be needed in the RA portion to close the defect, without tearing the covering. The use of self-expanding stent grafts has also been reported (7,10). However, there is more control with balloon-expandable stents, which do not have the capacity for continued expansion, potentially impinging on the pulmonary vein orifice. The balloon-expandable stent is placed in an unobstructed SVC and flared in the right atrium. To prevent stent migration, we oversize the diameter of the stent by 2 to 4 mm with respect to the SVC and aim for a minimum 2-cm zone of apposition in the SVC. Despite this, stent instability was not uncommon and required anchoring with an overlapping bare metal stent. In retrospect, anchoring with a bare CP stent may have prevented stent embolization in the 1 case observed. Significant shortening of the stent during flaring of the caudal stent part may uncover the inferior rim of the defect and lead to a residual shunt. In our most recent 9 patients, we exclusively used custom-made 7- or 8-cm-long covered 10-zig CP stents to increase the zone of apposition in the SVC and to avoid the risk of residual defects. We now only use the 6-cm stent when a small RUPV that cannot be diverted to the LA is left above the covered stent, and an uncovered anchoring stent allows continued drainage into the SVC.

FOLLOW-UP. A small residual shunt was noted on TOE in more than one-half of the patients at the end of the procedure. We anticipated that after the stent had endothelialized and volume unloading led to a reduction of RA size, the shunt would resolve. Only 1 patient had a residual shunt at late follow-up. Qualitative reduction in the size of the right ventricle was noted on post-procedure echocardiography, and cardiac MRI confirmed a significant reduction of right ventricular volume.

As with the surgical approach to the correction of SVASD, the possibility of sinus node dysfunction is a potential concern when a stent is implanted in the SVC and right atrial junction (13). There has been no short- to medium-term evidence on serial electrocardiograms of sinus node dysfunction in any of the patients during follow-up. Pulmonary venous obstruction or SVC stenosis are additional occasional complications after surgical repair (3). Although SVC stenosis is unlikely with the transcatheter approach, the potential risk of pulmonary vein stenosis is not negligible. However, careful pre-operative patient selection and intraprocedural assessment of pulmonary venous return before stent placement can identify patients at risk.

STUDY LIMITATIONS. Although covered stent implantation to correct SVASD has had a high success rate in our unit, it is only in its infancy, and our longest follow-up period is 3.8 years. We are currently less reliant on printed 3D models, but critical assessment before and during the procedure is essential to avoid pulmonary vein obstruction. Despite continued refinement of the technique (particularly using longer and anchoring stents) the stent embolized in our 20th patient, highlighting the need for surgical backup.

CONCLUSIONS

Transcatheter treatment of sinus venosus atrial septal defects is an attractive alternative to surgical treatment. The 3D reconstruction of cardiac MRI or CT imaging is essential for patient selection. Based on our single-center experience, approximately 75% of patients with an SVASD and PAPVD may be suitable for transcatheter correction. Medium-term results are promising, but longer follow-up studies are warranted.

ADDRESS FOR CORRESPONDENCE: Prof. Eric Rosenthal, Evelina London Children's Hospital, Lambeth Palace Road, London SE1 7EH, UK. E-mail: Eric.rosenthal@gstt.nhs.uk. Twitter: @erosenthal6.

PERSPECTIVES

COMPETENCY IN PATIENT CARE AND PROCEDURAL SKILLS: Transcatheter implantation of a covered stent to separate the superior vena cava and right upper pulmonary vein can redirect venous return into the right and left atria, respectively, as a potential alternative to open surgical closure.

TRANSLATIONAL OUTLOOK: Longer-term follow-up studies are needed to confirm the safety and durability of this method and guide patient selection.

REFERENCES

1. Naqvi N, McCarthy KP, Ho SY. Anatomy of the atrial septum and interatrial communications. J Thorac Dis 2018;10:S2837–47.

2. Li J, al Zaghal AM, Anderson RH. The nature of the superior sinus venosus defect. Clin Anat 1998; 11:349–52.

3. Iyer AP, Somanrema K, Pathak S, Manjunath PY, Pradhan S, Krishnan S. Comparative study of single- and double-patch techniques for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 2007;133:656–9.

4. Stewart RD, Bailliard F, Kelle AM, Backer CL, Young L, Mavroudis C. Evolving surgical strategy for sinus venosus atrial septal defect: effect on sinus node function and late venous obstruction. Ann Thorac Surg 2007;84:1651–5.

5. Garg G, Tyagi H, Radha AS. Transcatheter closure of sinus venosus atrial septal defect with anomalous drainage of right upper pulmonary vein into superior vena cava—an innovative technique. Catheter Cardiovasc Interv 2014;84: 473–7.

6. Abdullah HAM, Alsalkhi HA, Khalid KA. Transcatheter closure of sinus venosus atrial septal defect with anomalous pulmonary venous drainage: innovative technique with long-term follow-up. Catheter Cardiovasc Interv 2019 Jun 14 [E-pub ahead of print].

7. Crystal MA, Vincent JA, Gray WA. The wedding cake solution: a percutaneous correction of a form fruste superior sinus venosus atrial septal defect. Catheter Cardiovasc Interv 2015;86:1204–10.

8. Velasco Forte MN, Byrne N, Valverde I, et al. Interventional correction of sinus venosus atrial septal defect and partial anomalous pulmonary venous drainage: procedural planning using 3D printed models. J Am Coll Cardiol Img 2018;11: 275–8.

9. Riahi M, Velasco Forte MN, Byrne N, et al. Early experience of transcatheter correction of superior sinus venosus atrial septal defect with partial anomalous pulmonary venous drainage. EuroIntervention 2018;14:868–76.

10. Thakkar AN, Chinnadurai P, Breinholt JP, Lin CH. Transcatheter closure of a sinus venosus atrial septal defect using 3D printing and image fusion guidance. Catheter Cardiovasc Interv 2018; 92:353–7.

11. Butera G, Sturla F, Pluchinotta FR, Caimi A, Carminati M. Holographic augmented reality and 3D printing for advanced planning of sinus venosus ASD/partial anomalous pulmonary venous return percutaneous management. J Am Coll Cardiol Intv 2019;12:1389–91.

12. Tandon A, Burkhardt BEU, Batsis M, et al. Sinus venosus defects: anatomic variants and transcatheter closure feasibility using virtual reality planning. J Am Coll Cardiol Img 2019;12: 921–4.

13. Glatz AC, McBride MG, Paridon SM, et al. Long-term noninvasive arrhythmia assessment after surgical repair of sinus venosus atrial septal defect. Congenit Heart Dis 2010;5:141–8.

KEY WORDS adult congenital heart disease, atrial septal defect, structural intervention