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CASE REPORT
Adult congenital heart disease

Modified approach for transcatheter correction of superior sinus venosus atrial septal defect: a case series

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Background Transcatheter correction of superior sinus venosus atrial septal defect (SVASD) with partial anomalous pulmonary venous drainage (PAPVD) of the right upper and/or right middle pulmonary vein (RUPV/RMPV) has recently been described as an alternative to surgical approach in a substantial number of patients. We describe a modified technical approach for transcatheter correction of SVASD using transoesophageal echocardiography (TOE) to confirm adequate stent landing zone and apposition and evaluate its feasibility, safety, and procedural success.

Case summary From 2019 to 2021, three consecutive patients received a transcatheter correction of SVASD with PAPVD by redirecting the superior vena cava and RUPV/RMPV to the left atrium by implantation of a custom-made covered Cheatham platinum stent (10-zig, length: 60–80 mm). Prior to stent implantation, a balloon testing was performed in the anticipated landing zone using TOE to confirm complete defect closure and unobstructed pulmonary venous drainage. Stent deployment and flaring of the interior stent portion were performed with TOE guidance to confirm adequate landing zone and apposition and to avoid residual shunt or pulmonary vein obstruction.

Conclusion Transcatheter correction of SVASD with PAPVD was performed without any complications. The follow-up period was 7.8, 13.6, and 29.8 months, respectively. During follow-up, no mortality, stent embolization, or obstruction of pulmonary venous drainage occurred. The TOE-guided modified transcatheter approach for correction of SVASD with PAPVD is safe and feasible with excellent post-procedural results and represents an alternative to surgical treatment in a pre-selected patient cohort.

Keywords Superior sinus venosus atrial septal defect • Transcatheter correction • Covered CP stent • Case series • Transoesophageal echocardiography guidance

ESC Curriculum 9.7 Adult congenital heart disease • 7.4 Percutaneous cardiovascular post-procedure

Learning points

- The modified transcatheter correction of superior sinus venosus atrial septal defect is feasible with excellent post-procedural results.
- The modified transcatheter correction of superior sinus venosus atrial septal defect is an alternative to surgical treatment in a pre-selected patient cohort.
- Transoesophageal echocardiography guidance is substantial to confirm adequate landing zone and complete defect closure after stent placement.

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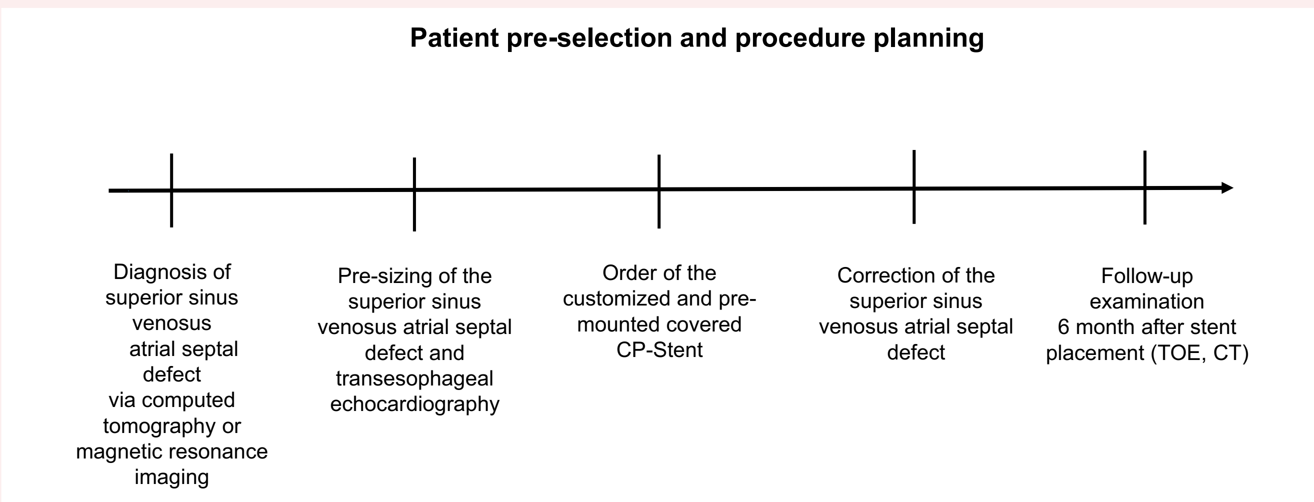
Introduction

The superior sinus venosus atrial septal defect (SVASD) is located above the superior rim of the oval fossa and allows the superior vena cava to override the atrial septum and to drain into the right and left atrial chambers.[1] SVASD is frequently associated with partial anomalous venous drainage (PAPVD) of the right upper and/or middle pulmonary veins (RUPV/RMPV).[1] Until recently, surgical correction has been the therapeutical gold standard. The first approach for transcatheter correction of SVASD was proposed by Garg *et al.*[2] and has been adapted and modified in various pediatric cardiology centres.[3–6] In the following, we describe our modified approach for transcatheter SVASD correction using transesophageal echocardiography (TOE) guidance to facilitate pre-procedural planning and adequate stent placement.

Timeline



Case presentation

From 2019 to 2021, transcatheter closure of SVASD with PAPVD of RUVP/RMPV was performed in three consecutive patients with a median age of 59.9 years. Detailed patient characteristics and haemodynamics are displayed in *Table 1*. Patient selection and detailed procedure planning were performed in a multi-disciplinary approach including paediatric cardiologists, congenital heart surgeons, and cardiac imaging experts. Indications for treatment were right heart volume overload and a pulmonary-to-systemic blood flow ratio >1.5:1. Informed consent to use custom-made covered Cheatham platinum (CP) stents was obtained from all patients prior to the procedure. Additionally, all patients were indicated to the novelty of this interventional technique.

All three patients received a computed tomography scan prior to interventional treatment to visualize the anatomic relationship of SVASD and pulmonary venous drainage. Diagnostic catheterization with balloon test occlusion using a 24 or 34 mm Amplatzer™ sizing balloon (Abbott, Plymouth, MN, USA) was performed in all patients. The balloon was inflated in the participated stent landing zone until TOE confirmed complete elimination of the interatrial shunt and excluded pulmonary venous obstruction during maximal balloon inflation. The optimal stent length for custom-made covered CP stents was chosen from measurements during balloon testing as described by Hansen *et al.*[3]

The main steps of the interventional procedure are presented in *Figure 1*. All interventional procedures were performed under general anaesthesia with TOE guidance. Vascular access was obtained in the right femoral and right internal jugular vein (RIJV). Heparin at 100 units/kg body weight was applied to avoid catheter-induced thrombus formation. Thirty minutes prior to stent placement patients received an antibiotic prophylaxis with 2000 mg cefazolin. A 0.035 inch Amplatz Extra Stiff™ (AES) wire (Cook Medical, Bloomington, IN, USA) was introduced as a veno-venous guide rail from the right femoral to the RIJV. All patients received custom-made, 10-zig covered CP stents (NuMed Inc, Sheffield, UK) with a stent length of 60–80 mm, which were pre-mounted on BiB® catheters (PFM Medical, Carlsbad, CA, USA). The stents were placed through long delivery sheaths (16-F, Check-Flo; Cook Medical) from the right femoral vein. Securing of the stent position in the predefined landing zone was achieved by snaring the cranial end of the AES wire with a 20 mm Multi-Snare® catheter (PFM Medical) from RIJV access in one patient. In two patients, the CP stent was extracted from the RJIV access and a 2-0 Ethicon® surgical silk suture (length: 75 cm; JnJ Medtech, New Brunswick, NJ, USA) was inserted through the superior zig (*Figure 1E*). After fixation of the surgical silk, the stent was retrieved to the landing zone while the silk suture was extracted from RIJV access. Secure anchoring of the stent without dislocation to the inferior vena cava was achieved by manual traction of the multi-snare catheter or the surgical silk during stent deployment. After stent placement flaring of the middle or distal stent section was performed using an Amplatzer™ sizing balloon (Abbott) or a Z med™ II balloon (PFM Medical) for optimal stent configuration and abolishment of residual interatrial shunt under simultaneous

Table 1 Patient characteristics

	Patient 1	Patient 2	Patient 3
Patient age (years)	54	67	59
Sex	Male	Female	Male
PAPVD	RUVP	RUPV/RMPV	RUPV/RMPV
Symptoms	Palpitations	Diminished cardiopulmonary performance	Dyspnoea
Concomitant diagnosis	Paroxysmal atrial fibrillation	None	Arterial hypertension
Medical history	Two electric cardioversions	Surgical correction of an oesophageal diverticulum	None
Physical examination	Unremarkable	Unremarkable	Unremarkable
Native diameter of SVASD (mm)	15	17	10–12
Stretched diameter of SVASD (mm)	26–28	25	24–27
Shunt direction	Left-to-right	Left-to-right	Left-to-right
Pre-procedural haemodynamic parameters			
CVP (mmHg)	10	12	10
RSVP (mmHg)	35	53	50
sPAP (mmHg)	28	53	49
mPAP (mmHg)	15	31	29
PVR (WU)	1.2	4.2	3.4
Qp:Qs	1.2:1	2.5:1	2.8:1
RVEDVi (mL/m^2)	163.0	212.0	202.7
RV-EF (%)	64	69	44
Post-procedural haemodynamic parameters			
RVSP (mmHg)	27	43	40
sPAP (mmHg)	22	50	34

CVP, central venous pressure; PAPVD, partial anomalous pulmonary venous drainage; PVR, pulmonary vascular resistance; Qp:Qs, pulmonary-to-systemic flow ratio; RMPV, right middle pulmonary vein; RUPV, right upper pulmonary vein; RVEDVi, right ventricular end-diastolic volume index; RV-EF, right ventricular ejection fraction; RVSP, right ventricular systolic pressure; sPAP, systolic pulmonary artery pressure; SVASD, superior sinus venosus atrial septal defect; WU, Wood units.

TOE guidance to allow optimal stent apposition. Pulmonary vein obstruction was excluded using TOE during and after stent placement.

All patients received a chest X-ray, an echocardiogram, and an echocardiography prior to discharge. Anticoagulatory treatment consisted of aspirin, apixaban, or phenprocoumon for a period of 6 months, respectively. Residual shunt and pulmonary venous obstruction were excluded 6 months after initial procedure with diagnostic catheterization ($n = 1$) or TOE ($n = 2$).

Discussion

Transcatheter correction of SVASD with PAPVD is a complex and challenging interventional procedure, which requires a detailed, comprehensive, and multi-disciplinary pre-procedural planning. Various preconditions need to be fulfilled for a successful interventional treatment (*Table 2*). First, the SVC needs to be unobstructed and of regular size. Second, the RUPV/RMPV needs to be in direct continuity with the left atrium (LA). Therefore, the use of 3D-printed models might be helpful for anatomic visualization and procedural planning.[7] Third, the number, size, and location of anomalously draining pulmonary veins should be considered carefully, since in patients with a small junction between the SVC and the pulmonary venous pathway might be narrowed during stent deployment. A high or accessorily draining RUPV might not be connected to the LA and could remain draining into the SVC.[6] Fourth, a precise calculation of the width and length of the CP stent is indispensable for a successful procedure. Due to the flaring of the inferior stent end the shortening of the CP stent also needs to be taken into consideration when choosing the adequate stent length.[3] The cranial stent end was fixated with an Ethilon® stich in two of three patients, which was extracted from the RIJV access and sufficiently provided stent movement during balloon inflation by manual traction. Compared to a Multi-Snare® catheter, the use of the Ethilon® stich bears the advantage of a more simplified handling during the procedure. However, adequate sufficiency to anchor the stent during balloon inflation was proven for both methods.[3,6] Using TOE guidance for constant pulmonary vein monitoring during the procedure transseptal puncture and continuous LA and RUPV pressure measurement could be avoided. Additionally, TOE was indispensable to define the optimal stent landing zone and apposition during flaring of the middle and distal stent end and to obviate residual interatrial shunt.

To conclude, our case series underlines the feasibility, safety, and procedural success of transcatheter SVASD correction using our TOE-guided approach in a pre-selected patient cohort.

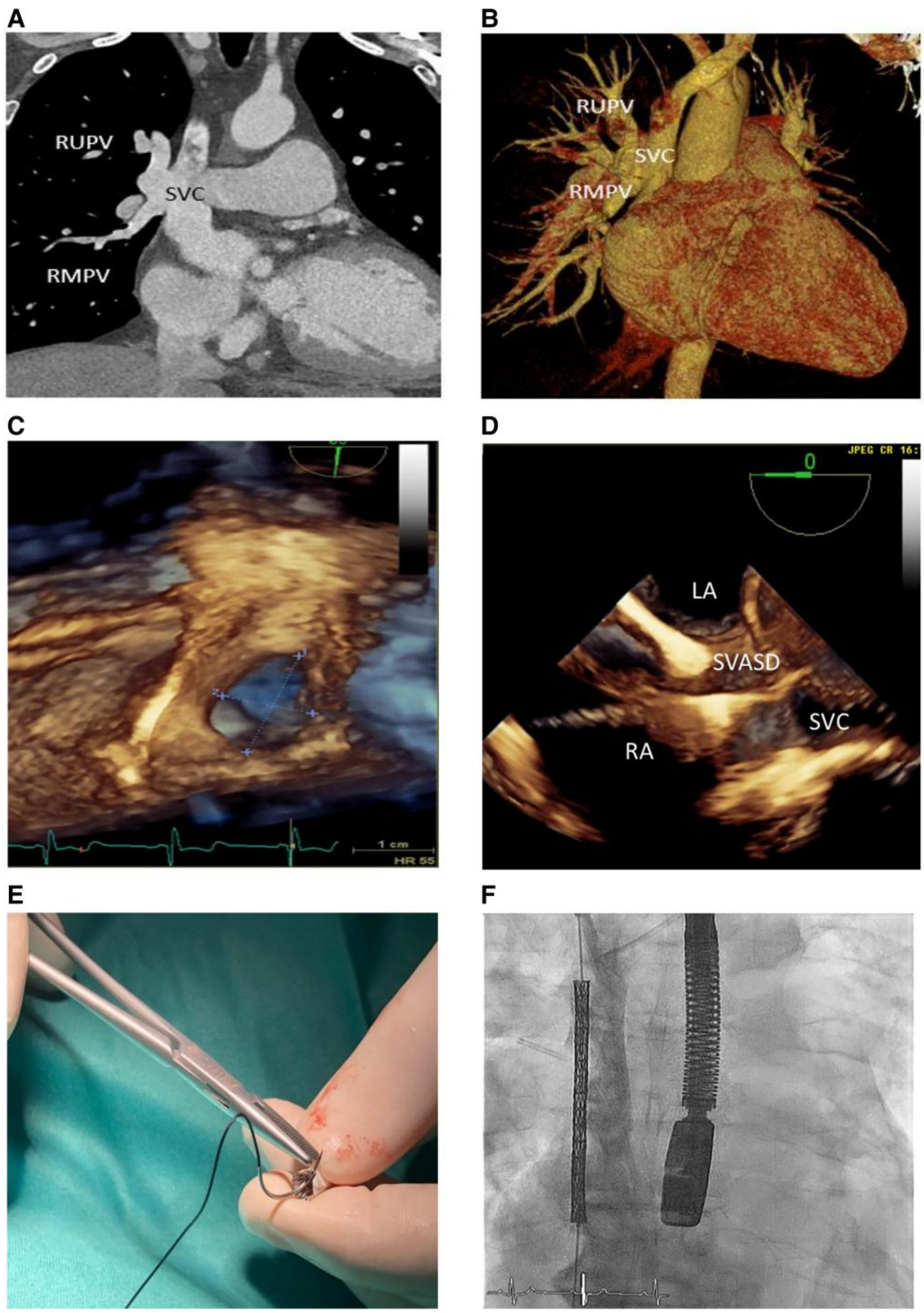


Figure 1 Stepwise modified transcatheter correction of superior sinus venosus atrial septal defect with partial anomalous pulmonary venous drainage of the right upper pulmonary vein and right middle pulmonary vein. (*A*) Computed tomography scan depicting superior sinus venosus atrial septal defect with partial anomalous drainage of the right upper pulmonary vein and right middle pulmonary vein into the superior vena cava. (*B*) 3D reconstruction of computed tomography scan images. (*C*) 3D transoesophageal echocardiography showing dimensions of the superior sinus venosus atrial septal defect (1.2 × 2.0 cm). (*D*) 3D transoesophageal echocardiography depicting the anatomical conditions between the interatrial junction, superior sinus venosus atrial septal defect and superior vena cava. (*E*) Induction of a 2-0 Ethicon® surgical silk stich, which is extracted from RIJV access to allow secure anchoring of the CP stent during balloon expansion. (*F*) Placement of the custom-made pre-mounted 10-zig/80 mm covered CP stent using a 14-F Check-Flo sheath. (*G*). Deployment of 10-zig CP stent using a 26 mm BiB® catheter. (*H*) Flaring of the inferior stent section with a 28 mm/4 cm Z med II balloon. (*I*) 3D transoesophageal echocardiography confirming adequate apposition of the covered CP stent. (*J*) 3D transoesophageal echocardiography excluding obstruction of the right pulmonary veins. (*K*) Venography of the superior vena cava revealing no residual shunt. (*L*) 3D transoesophageal echocardiography showing adequate position of the CP stent. BiB, balloon in balloon; CP, Cheatham platinum; CT, computed tomography; LA, left atrium; RIJV, right internal jugular vein; RMLP, right middle pulmonary vein; RUVP, right upper pulmonary vein; SVC, superior vena cava; TOE, transoesophageal echocardiography.

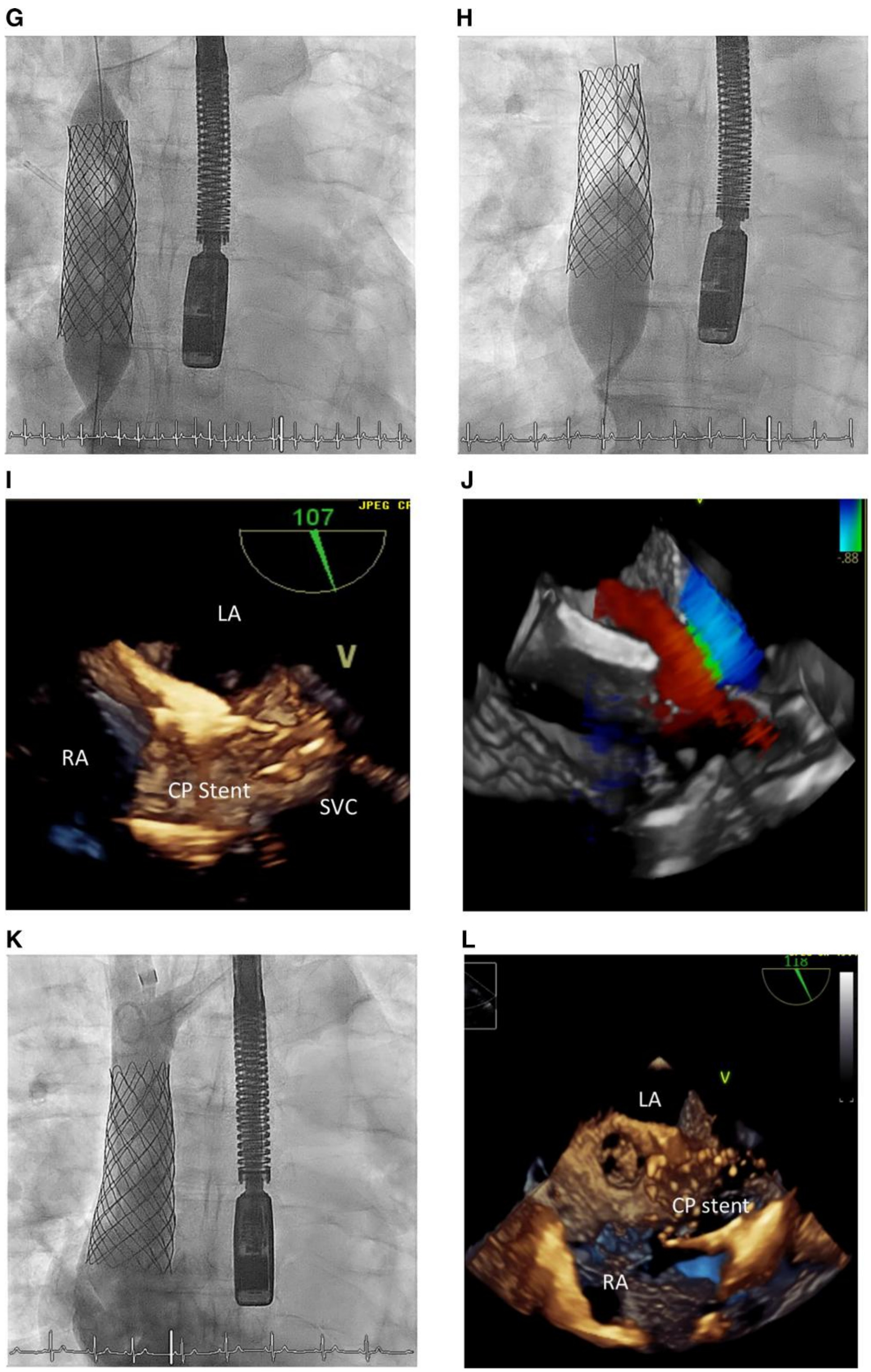


Figure 1 Continued

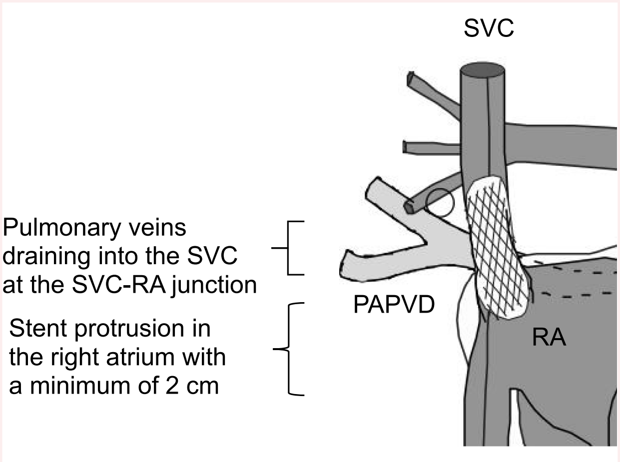


Figure 2 Schematic description of anatomic requierements for transcatheter correction of superior sinus venosus atrial septal defect. RA, right atrium; SVASD, superior sinus venosus atrial septal defect; SVC, superior vena cava.

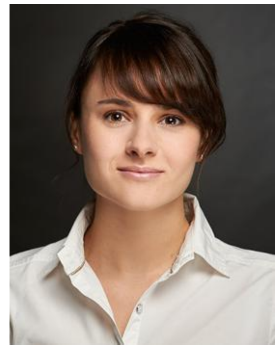
Table 2 Pre-conditions for transcatheter correction of superior sinus venosus atrial septal defect with partial anomalous pulmonary venous drainage in the adult patient cohort

Anatomic considerations	Exclusion criteria
• Regular sized and unobstructed SVC	• Large SVASD with extension to the atrial septum
• Direct continuity of the RUPV/RMPV to the left atrium	• Pulmonary veins draining into the SVC instead of the atrial junction
• Adequate number of abnormally draining pulmonary veins ($n = 1$–2)	
• Adequate size of the junction between the SVC and pulmonary vein drainage (>1.5 cm)[a]	

RMPV, right middle pulmonary vein; RUVP, right upper pulmonary vein; SVASD, superior sinus venosus atrial septal defect; SVC, superior vena cava.

[a]Adequate size of the junction between SVC and pulmonary vein drainage is determined during balloon test occlusion of SVC or can be pre-estimated using 3D printed or virtual models. In general, stent length is chosen based on the following criteria: A minimum of 2 cm of the unexpended stent in the SVC and a minimum of 2 cm protruding into the RA below the pulmonary vein entrance is necessary (*Figures 2*). Since the distal part of the stent is flared up to a diameter of 34 mm stent (until SVC is closed) shortening has to be taken into consideration. Customized CP stents are available lengthening 5–11 cm.

Lead author biography



Anastasia Schleiger is an interventional cardiology fellow at German Heart Center Charité with a special interest in interventional cardiology and univentricular heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: Informed consent was given from all patients prior to the procedure in compliance with COPE guidelines. Additionally, all patients agreed to publication of their medical data within this case series.

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

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