

# How to do it? Transcatheter correction of superior sinus venosus defects

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## ABSTRACT

Superior sinus venosus defect is characterized by varying degrees of override of the atrial septum by the superior vena cava and anomalous right upper pulmonary vein (RUPV) drainage. The defect is seen posterosuperior to the oval fossa and is traditionally corrected by a surgical patch that routes the anomalously draining RUPV to the left atrium. A few postoperative patients develop stenosis or occlusion of the caval or pulmonary vein and/or sick sinus syndrome secondary to vascular injury. This has kindled a recent interest in nonsurgical correction of these defects by the use of a covered stent to reroof the defect. The complex anatomy of this defect often necessitates the use of advanced imaging tools and postprocessing. Countries with restricted resources do not have easy access to technologies such as three-dimensional printed models and advanced processing like virtual reality or holography. This "How to do it?" article, applicable in these countries, explains the current understanding of this intervention, patient selection, step-by-step explanation of the evaluation, and stent implantation.

**Keywords:** Anomalous pulmonary vein drainage, balloon interrogation, covered stent, nonsurgical closure, stent exclusion, superior vena cava

## INTRODUCTION

Superior sinus venosus defects (SVDs) account for 10% of all interatrial communications. An unroofing of the right upper pulmonary vein (RUPV) leads to its anomalous drainage to either the superior vena cava (SVC) or the right atrium and a biatrial connection of the SVC.<sup>[1]</sup> Surgeons use a pericardial patch that separates a posterior pulmonary venous pathway from an anterior systemic venous pathway, which rarely leads to venous occlusions or sinus nodal injury.<sup>[2]</sup> The recent interventional technique originally reported from India, performs a similar reroofing using a covered stent that allows SVC to flow through its lumen toward the right atrium and redirects RUPV flows outside its covered surface toward the left atrium.<sup>[3,4]</sup> The outer surface of

the covered stent snug fits against the caudal edge of the SVD and closes the interatrial communication.<sup>[5]</sup>

## PREPROCEDURAL SCREENING

The essential prerequisites for closing any interatrial communication are a clinical, radiological, and electrocardiographic confirmation of significant left-to-right shunt and exclusion of patients with pulmonary vascular disease. The diagnosis of the SVD is confirmed on transthoracic or transesophageal echocardiography when the defect identified near the SVC orifice is located posterosuperior to the oval fossa and is accompanied by anomalous drainage of the RUPV [Figure 1]. In case of doubt, the anatomy

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is confirmed on computed tomography or magnetic resonance imaging.

## PRINCIPLES OF THE INTERVENTION

The three principal aims of the correction of SVD are (i) closure of the interatrial communication, (ii) directing the SVC flows toward the right atrium, and (iii) redirecting the anomalous RUPV toward the left atrium. The proof of concept of this intervention is tested in a balloon interrogation. If a long semi-compliant balloon, 2–4 mm larger than the SVC, is inflated across the superior cavoatrial junction till it occludes the SVC flows, the posterior surface of the balloon should snug fit against the caudal edge of the SVD and close the

interatrial communication [Figure 2]. While doing so, a simultaneous angiography from the RUPV should demonstrate its unobstructed flow toward the left atrium. To achieve this efficiently, the balloon should be long enough to straddle across on either side of the RUPV orifice. If the balloon stops the interatrial flows, the correction of the SVD can be made permanent using a covered stent of the same diameter as the balloon.<sup>[1,3,4]</sup>

## IDEAL ECHOCARDIOGRAPHIC ANATOMY

In patients with SVD, the lower end of SVC that receives the drainage of the anomalous RUPV is dilated to provide a roomy buffer space around the covered stent. In most patients, the defect is anatomically localized to the cavoatrial junction. For covered stent exclusion of SVD, ideal anatomy can be defined as a defect that does not have a caudal extension toward the oval fossa and drainage of RUPV is confined to the cavoatrial junction. A significant caudal extension of SVD will not allow a snug fit between edges of the defect to the covered stent.<sup>[6]</sup> Similarly, a very low draining RUPV to the right atrium and a very high draining vein to the superior end of SVC will not get rerouted to the left atrium after a covered stent [Figure 3]. Hence, a transesophageal echocardiographic identification of (i) significant caudal extension, defined as the superoinferior defect dimension of the SVD far exceeding the transverse SVC diameter; (ii) low drainage of RUPV into the right atrium; and (iii) very high drainage into the high SVC are exclusions from transcatheter therapy.

## CRITERIA FOR “PASS” DURING THE BALLOON TESTING

The following criteria should be satisfied. (i) Angiogram from RUPV during balloon interrogation should demonstrate the unobstructed flow of contrast

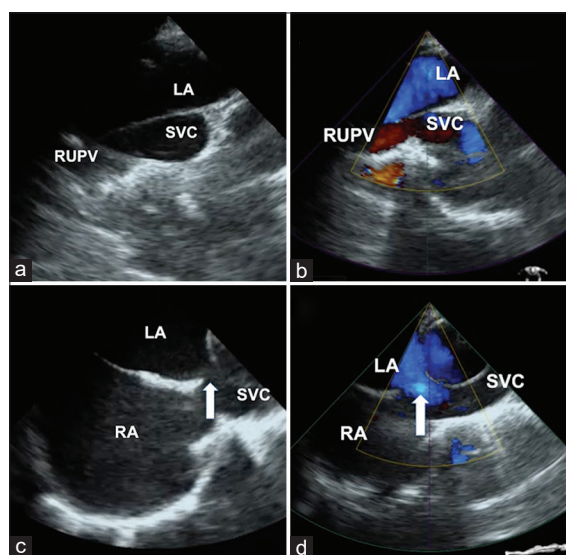


Figure 1: Transesophageal short-axis view (a) with color flow imaging (b) shows anomalous right upper pulmonary vein drainage to the superior vena cava. Long-axis view (c) with color flow imaging (d) shows interatrial communication shown in arrow between the left atrium and the right atrium

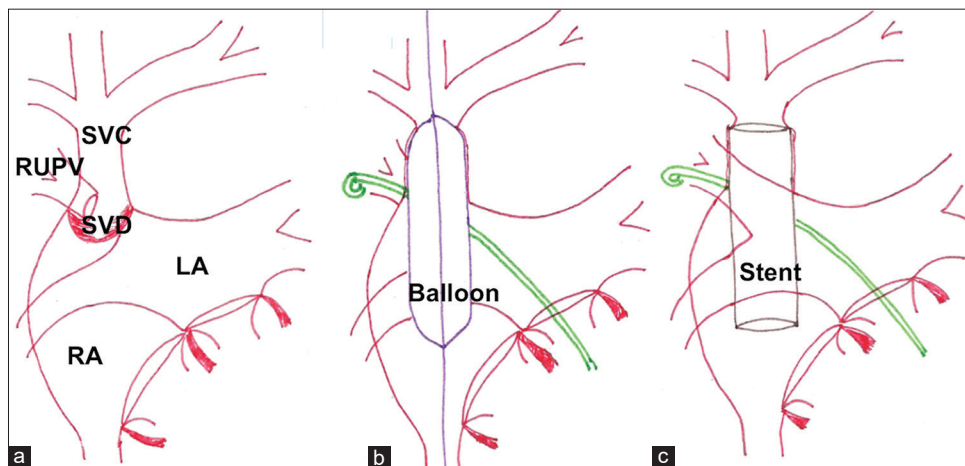
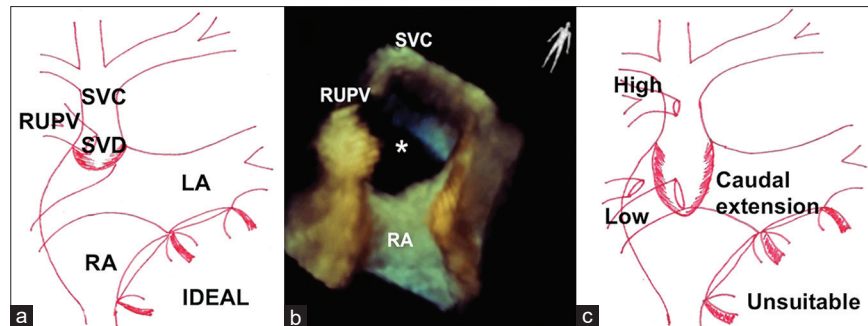
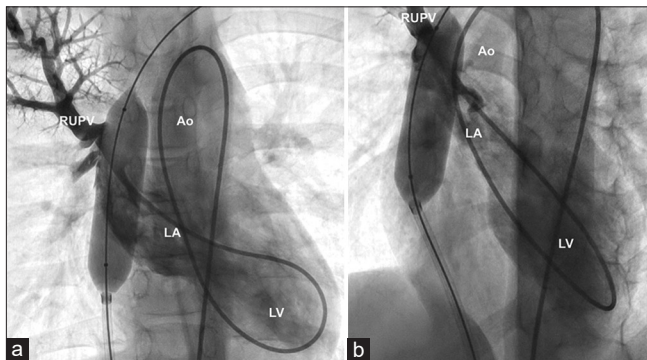


Figure 2: Thematic cartoon (a) showing sinus venosus defect at the junction between the superior vena cava and the right atrium with anomalous right upper pulmonary vein drainage. After placing a long balloon in the cavoatrial junction, the right upper pulmonary vein is monitored by pressures and angiography (b) before a covered stent placement (c)



**Figure 3:** An ideal echocardiographic anatomy (a) is a sinus venosus defect at the junction of superior vena cava and right atrium and anomalous right upper pulmonary vein drainage confined to the cavoatrial junction. This is illustrated in the right atrial enface view of a three-dimensional echocardiogram (b). When there is a caudal extension of the sinus venosus defect toward the oval fossa or drainage of the right upper pulmonary vein too high or too low, (c) they are not suitable



**Figure 4:** During balloon occlusion of cavoatrial junction, right upper pulmonary vein angiogram in anteroposterior (a) and hepatoclavicular (b) views shows exclusive filling of the left atrium left ventricle and aorta indicating complete rerouting

toward the left atrium. (ii) RUPV should not show any significant rise beyond 2 mmHg from baseline during balloon interrogation. An increase more than 2 mmHg without any stasis of contrast is an indicator of a restrictive left ventricle physiology that should be confirmed by a pullback from the RUPV toward the LA. (iii) RUPV angiogram should not opacify the right atrium in an angiographic projection that separates the two atria such as the hepatoclavicular view [Figure 4]. This angiogram is performed with a high-pressure injector using at least 15–20 ml of contrast to avoid ambiguities in its interpretation. If RUPV is occluded, the balloon is downsized by 2 mm till the patient “passes” the balloon test. If there is residual left-to-right flow, the balloon is upsized by 2 mm until a “pass” is obtained.

### OPTIONAL ECHOCARDIOGRAPHIC CONFIRMATION

The angiographic and hemodynamic assessment from the RUPV catheter is a foolproof method of assessing the criteria for “pass” in balloon testing. However, it can also be confirmed by color Doppler transesophageal echocardiography that can demonstrate laminar color

flows from RUPV toward the left atrium [Figure 5]. This may need intubation anesthesia in an apprehensive patient or mild conscious sedation with midazolam and glycopyrrolate in a cooperative patient. If the patient has an additional oval fossa defect, echocardiographic imaging will differentiate between the residual flow through the SVD and the oval fossa defect.

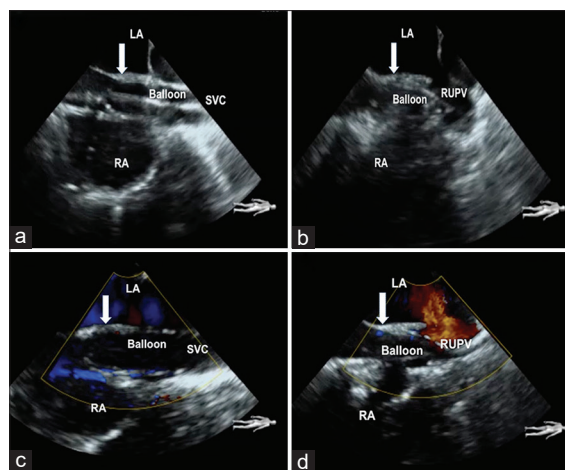
### CHOOSING THE COVERED STENT – DIAMETER

Once the balloon test is completed, the diameter of the chosen covered stent should be equal to the diameter of the balloon. While an 8-zig covered CP stent (NuMed Corporation, Hopkinton, NY) is sufficient up to a diameter of 22 mm, a 10-zig covered CP stent is chosen for larger diameters. A covered Andrastent XXL (Andramed GmbH, Reutlingen, Germany) that can be dilated up to 32 mm, covered Optimus CVS XXL (AndraTec GmbH, Koblenz, Germany) that can be dilated to 28 mm, and BeGraft aortic (Bentley Innomed GmbH, Hechingen, Germany) that can be dilated to 30 mm are the other alternative options. All stents foreshorten on larger diameters as specified by the vendors.

### CHOOSING THE COVERED STENT – LENGTH

The stent should have a sufficient anchor in the SVC to avoid a caudal embolization. An easily identified superior landmark is the junction of the innominate veins that ensures a secure anchor of the stent in the SVC. Maintenance of patency of the azygos vein is not too vital in these patients as SVC obstruction is unlikely after large stent placement. The lower end of the stent (or inferior landmark) should extend around 2 cm below the level of the RUPV to ensure that the outer surface of the stent snug fits against the caudal edge of the SVD. The distance between the superior and the inferior landmarks is measured with a marker pigtail to estimate the length of the covered stent [Figure 6]. The longest commercially





**Figure 5:** Transesophageal echocardiogram (a) shows inflated balloon in the junction between the superior vena cava and the right atrium that obliterates the sinus venosus defect between the left atrium and right atrium. The right upper pulmonary vein is seen connecting (b) to the left atrium. Color flow imaging shows the absence of flows from left atrium to the right atrium (c) and laminar unobstructed flow from the right upper pulmonary vein to the left atrium (d)

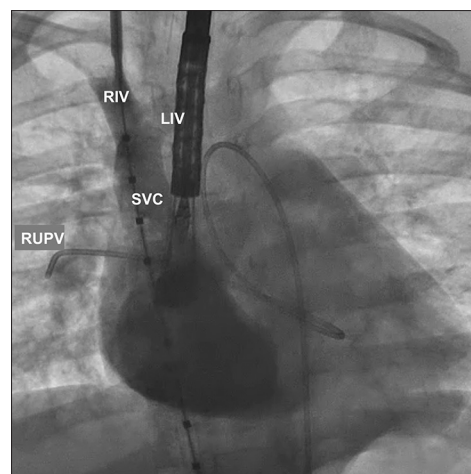
available CP stent is 60 mm while they can be custom ordered up to 80 mm. The longest commercially available Andrastent XXL and Optimus CVS is 57 mm, while the longest large vessel BeGraft aortic is 48 mm. Stent overlap should be planned when using the last three stents.

## PLACING THE RIGHT UPPER PULMONARY VEIN CATHETER FOR GUIDANCE

The key to the balloon interrogation is a stable position of an angiographic catheter with side holes (avoid wedging) in the RUPV. Our preferred method is a retrograde arterial approach. After placing a 6F femoral arterial access that provides continuous monitoring of the arterial pressures, a 5F Judkins right coronary catheter is advanced through the aortic and mitral valves into the left atrium guided by an angled exchange length Glidewire (Terumo Corporation, Tokyo, Japan). This guidewire is advanced into the RUPV and then exchanged for a 5F pigtail catheter [Figure 7]. Otherwise, a second venous catheter from the left femoral vein can be advanced along the edges of the SVD into the RUPV. A third option is to explore for a patent foramen ovale through a venous catheter or perform a septal puncture with a Brockenbrough needle.<sup>[4]</sup> This catheter monitors the pressure in the RUPV and assists repeated angiography injections.

## DEPLOYMENT OF THE STENT

The most feared complication is embolization of the stent caudally into the right atrium. Precise placement



**Figure 6:** A marker catheter guides the length of the covered stent. The superior landmark is the junction of the right and left innominate veins. The inferior landmark is 2 cm below the right upper pulmonary vein orifice

of the stent in reference to the superior and inferior landmarks explained above is the vital step. The femoral vein access is upsized to a 20F Performer sheath (Cook Medical, Bloomington, IN) or Dryseal flex (W. L. Gore, Flagstaff, AZ) to allow large profile covered stents mounted on large balloons. After obtaining a 6F right jugular venous sheath, a venovenous circuit is made between the femoral and jugular veins. The covered stent is mounted on an appropriate balloon whose diameter is chosen based on the previous “passed” balloon test. The stent should straddle across the RUPV catheter and should satisfy the superior and inferior landmarks before it is expanded fully [Figure 8]. A check angiogram from the side arm of the jugular and femoral sheath as well as a check angiogram from the RUPV catheter can guide slow-controlled inflation of the balloon-stent assembly till the stent is anchored well in the SVC. The lower end of the stent can be flared using the same balloon moved inferiorly and reinflated at higher pressures.

## HOW TO OVERLAP STENTS?

A custom 7–8-cm long covered CP stent is often used for this procedure.<sup>[7]</sup> However, the lack of availability of such a stent should not be a reason to deny this procedure. Two Andrastent XXL can be overlapped on a long 8-cm balloon and deployed. While mounting the stents, the first stent (SVC part) should be mounted on the distal part of the balloon and the proximal stent (RA part) should be overlapping the first stent [Figure 9]. When these stents are expanded, the lower right atrial stent is prevented from a caudal migration as the inner upper first SVC stent grips the outer lower second stent and pushes the latter against the walls of the SVC.<sup>[1]</sup>

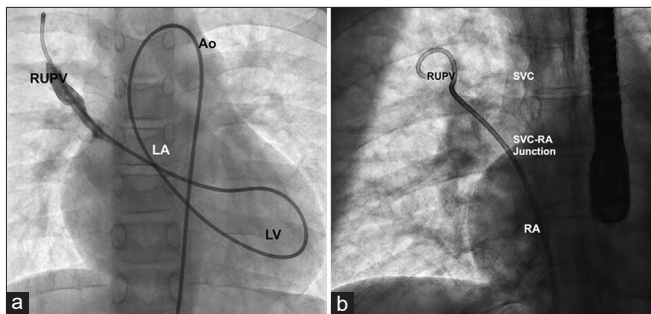
## FINAL CHECKS

Once the stent is fully expanded, an angiogram from the RUPV should demonstrate a complete seal of the SVD and redirected flows toward the left atrium. A residual flow is managed by flaring the lower end of the stent using a larger balloon. High pressures in the RUPV and left atrium indicate a restrictive left ventricular physiology, managed using furosemide and vasodilators. Hemostasis is obtained using a figure-of-8 stitch after removing the 20F large venous sheath. Postprocedural antiplatelet regimen includes aspirin and clopidogrel for 6 months. A predischarge

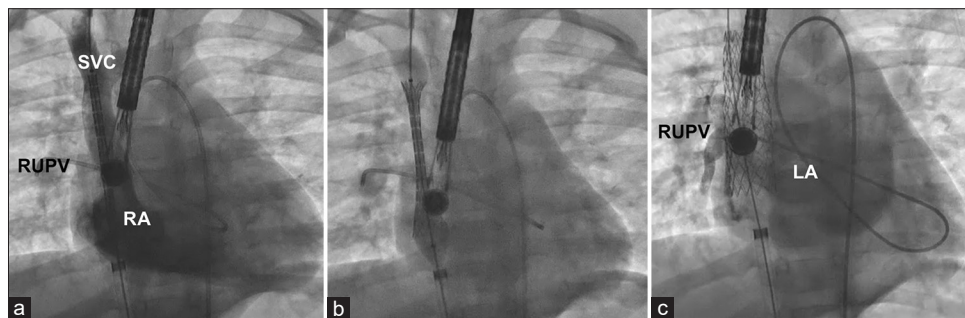
echocardiogram should confirm stable stent position, absence of residual flows, presence of normal venous Doppler pattern in the right femoral vein, and excluding femoral vein thrombosis. The patients are followed up at 3-monthly intervals for 1 year and yearly thereafter with clinical, electrocardiographic, and echocardiographic evaluations. A transesophageal study at 6 months before discontinuing antiplatelet drugs confirms the absence of residual flows and normal RUPV Doppler pattern during its flows to the left atrium [Figure 10].

## COMPLICATIONS

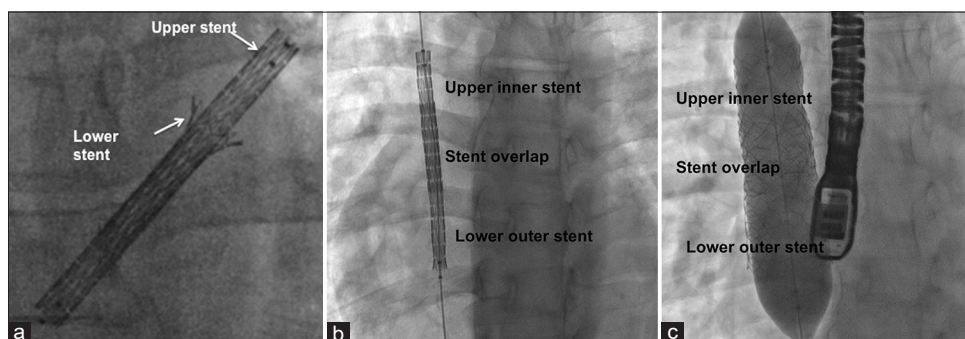
Meticulous attention to standard operating procedure should avoid complications such as vascular bleeding, cardiac injury, arrhythmias, thrombus or air embolism, guidewire injury to lungs, and infections. Stent migration is a serious complication that may need surgery but sometimes can be salvaged by an overlapping stent from the jugular vein that holds and fixes the caudally embolized stent.<sup>[1]</sup> Occlusion of the RUPV is unlikely to occur by this protocol that pretests the venous flow into the left atrium during balloon interrogation. However, such an occurrence should be dealt by advancing a guidewire followed by a 8 mm × 2 cm Mustang balloon (Boston Scientific, Boston, MA) exchanging the pigtail catheter placed in the RUPV.<sup>[4]</sup> Small residual flows may subside over time as the enlarged right atrium



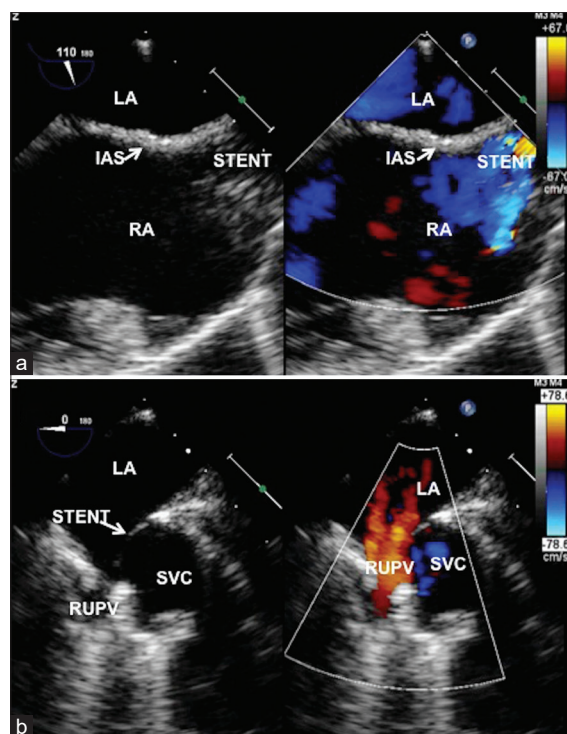
**Figure 7:** Access to the right upper pulmonary vein is obtained retrograde (a) from an arterial catheter passed from the aorta through the left ventricle and left atrium. It can be obtained antegrade (b) from a venous catheter through the cavoatrial (SVC-RA) junction also



**Figure 8:** During step-wise deployment of the stent, initial positioning (a) is guided by check injections from the jugular venous sheath that shows superior vena cava and right atrium, followed by slow inflation (b) till final full expansion of the stent (c). When fully expanded, right upper pulmonary vein injection shows exclusive filling of the left atrium



**Figure 9:** When stent overlap is needed, the upper stent is mounted first followed by the lower stent (a). This allows a zone of overlap (b) where the expanded upper stent will fix the lower stent against the walls of the cavoatrial junction (c) to prevent its caudal embolization



**Figure 10:** During follow-up transesophageal echocardiogram at 6 months, absence of flows between left atrium and right atrium indicate intact atrial septum on a long-axis view with color mapping (a) after stent placement. On a short-axis view, the right upper pulmonary vein entry to the left atrium with color flows (b) is clearly seen

remodels to a smaller size that allows the caudal edge of the SVD to approximate the covered stent. Dysfunction of the epicardial placed sinus node is unlikely and unreported after this procedure, as the stent offers an endocardial stretch. Groin bleeding and femoral vein thrombosis are dealt with in a routine manner.

## SUMMARY

Transcatheter closure of superior SVD evokes interest in recent times among pediatric interventional cardiologists as an alternative to surgery, as few postoperative patients develop venous occlusions or sick sinus syndrome. In resource-restricted nations without access to advanced three-dimensional modeling and printing, simple tools guide the closure. An initial transesophageal echocardiographic screen excludes caudal extension and veins that drain too high or too low. A “pass” in a

subsequently performed balloon test will finally identify the ideal candidate. Increasing operator experience across the world may improve the adoption of this technique in grown-up congenital heart disease patients as it shortens hospital stay and allows early recovery to employment.

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## Conflicts of interest

There are no conflicts of interest.

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