Different mechanisms for persistent and residual left-to-right shunt after transcatheter sinus venosus defect closure and their management

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ABSTRACT

Transcatheter closure of superior vena cava (SVC) form of sinus venosus defects (SVDs) using covered stents is emerging as an alternative to surgery in the current decade. A covered stent placed in the cavoatrial junction creates a roof for the right upper pulmonary vein (RUPV) that stops the left-to-right shunt and redirects the vein to the left atrium. While surgical literature has clearly documented the incidence of stenosis of SVC and RUPV, sinus nodal dysfunction, and persistent residual shunts following surgical correction, it is imperative to have similar data after this new transcatheter intervention on the incidence of complications and follow-up outcomes. Since patients with pretricuspid shunts are often clinically asymptomatic, correction is primarily performed to prevent a persistent right heart volume overload and allow remodeling of the heart chambers. Any residual left-to-right shunt after a correction will result in persistent right heart dilatation. Residual flows can result from various mechanisms, including lack of apposition of the covered stent to the free edge of the SVD, fabric breach, and persistent anomalous drainage of additional right-sided pulmonary veins that drain very high in the SVC or can be due to a coexistent defect in the oval fossa. This review analyzes the different mechanisms, explains the transesophageal and angiographic images for each one, and offers solutions tailored for various reasons. Different mechanisms warrant different treatment principles. A solution for residual shunt from one mechanism may not be appropriate for residual flow through another mechanism. A thorough understanding would aid the operator in effective interventions for these SVDs.

Keywords: Anomalous pulmonary venous drainage, covered stent, right heart dilatation, right upper pulmonary vein, superior vena cava, transesophageal echocardiography

INTRODUCTION

Surgical correction remains the gold standard for correcting sinus venosus defects (SVDs).[1] Even though various modifications evolved in the surgical techniques, including Warden anastomosis, superior vena cava (SVC) or right upper pulmonary vein (RUPV) stenosis, sinus nodal injury, and missed additional pulmonary veins are seen in few patients on follow-up.[1] Transcatheter closure using balloon-mounted covered stent(s) is emerging as an alternative to surgery with growing experience in the current decade.[2-4] The covered stent creates a wall between the SVC and the RUPV, thus roofing the latter and redirecting its flows to the left atrium. Balloon interrogation determines the appropriate diameter and length of the covered stent, whose outer wall will appose against the free edge of the SVD to eliminate the left-to-right shunt completely.[2,5] Any residual flow may potentially lead to persistent right heart dilatation even after the intervention.

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Normalization of right heart dilatation remains the primary goal of intervention in most pretricuspid shunts, as they are often asymptomatic. The prevalence of residual defect after transcatheter closure varies depending on the imaging modality. Transesophageal echocardiography (TEE) is very sensitive in identifying even insignificant leaks due to its high spatial and temporal resolution. A residual flow with a color Doppler width of <5 mm was identified in 7% of 115 patients after surgical SVD closure, which was considered clinically insignificant.[1] Analysis of residual shunt on magnetic resonance either by velocity-encoded phase contrast imaging or volumetric analysis is not sensitive to detect small left-to-right flows.[6] Similarly, small residual defects may remain undetected on computed tomography. This review article explains the different mechanisms responsible for persistent right heart dilatation after transcatheter SVD closure and discusses their management.

MECHANISMS OF RESIDUAL LEFT-TO-RIGHT SHUNTS

The left-to-right interatrial shunt from SVD is arrested when the outer wall of the covered stent apposes against the free edge of the defect and redirects the RUPV to the left atrium. The following mechanisms may lead to persistent pretricuspid left-to-right shunt after transcatheter SVD closure.

1. Fabric breach: Major and minor
2. Lack of apposition of the outer wall of the covered stent to the free edge of the SVD
3. Upper end of RUPV draining into the SVC cranial to the covered stent
4. Additional small pulmonary vein draining high in SVC above the main RUPV
5. Residual defect caudal to the covered stent due to its inadequate length
6. Interatrial shunts at the level of the oval fossa.

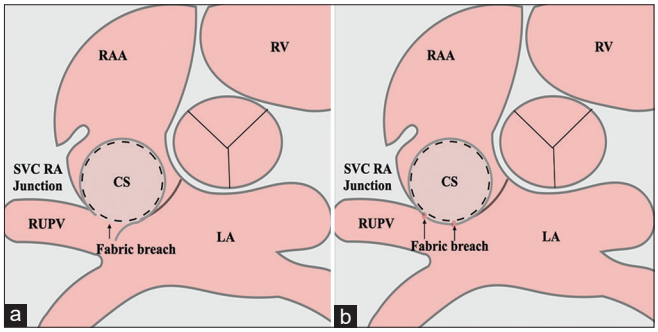

Figure 1: Fabric tear can be major (a), where the expanded polytetrafluoroethylene cover of the stent peels off, resulting in large residual flows, or minor (b), where there are small perforations in the fabric, leading to minimal shunt. RUPV: Right upper pulmonary vein, SVC: Superior vena cava, RA: Right atrium, CS: Covered stent, RAA: Right atrial appendage, LA: Left atrium, RV: Right ventricle

Fabric breach

The integrity of the expanded polytetrafluoroethylene fabric in the covered stent that creates a roof for the RUPV to close the shunt is one of the essential prerequisites for this intervention [Figure 1]. The fabric of the covered CP stent (Numed Inc., Hopkinton, NY) is glued only to its two ends, leaving the unattached mid-part vulnerable to breach during expansion. In a trial using covered CP stents in aortic coarctation, 7 out of 83 patients had endoleak and needed additional overlapping covered stents.[7] Other stents, such as covered Zephyr (Sahajanand Laser Technology Limited, Gandhinagar, India), are characterized by the fixation of fabric to all struts by sutureless lamination, where stretch can cause small pores.[8]

Major fabric breach

This manifests as a significant contrast leak into the right atrium on an RUPV angiogram after stent deployment. A repeat RUPV angiogram can confirm it after re-inflation of the balloon, which will seal the fabric breach and stop the contrast leak [Figure 2]. Intraprocedural TEE will confirm color Doppler flows through the struts of the stent that disappear on the re-inflation of the balloon [Figure 3]. This must be addressed by overlapping an additional covered stent to seal the fabric tear.[9]

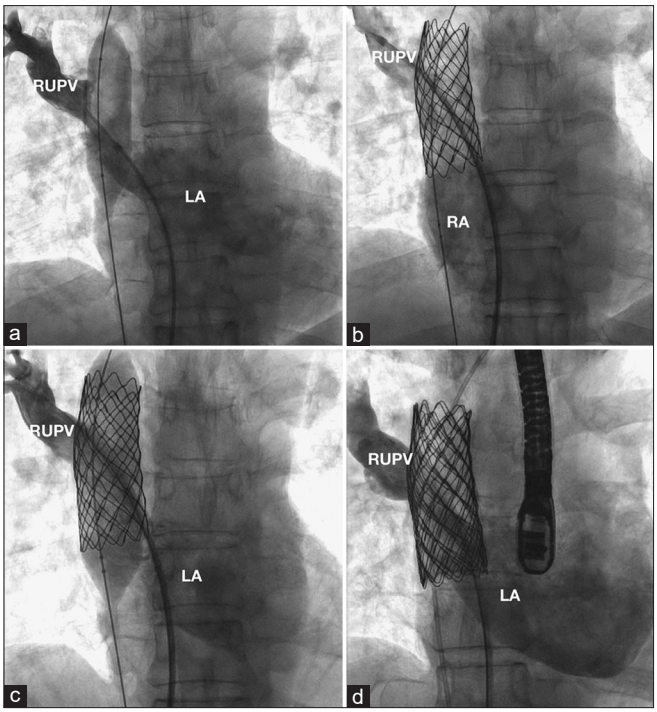

Figure 2: Following balloon interrogation (a) showing closure of left-to-right shunt and redirection of the right upper pulmonary vein to the left atrium, a long-covered CP stent (b) was deployed. RUPV angiogram shows filling of the right atrium, indicating residual flow. On reinflation of the balloon, the residual flow stops (c), indicating a possibility of fabric tear. A second overlapping covered stent (d) closes the residual shunt completely. RUPV: Right upper pulmonary vein, RA: Right atrium, LA: Left atrium

Minor fabric breach

Small perforations in the fabric may not be identified on a RUPV angiogram after covered stent deployment but are identified by TEE due to its high sensitivity. As the shunt is insignificant in minor leaks, they can be left alone [Figure 4].

Lack of apposition of the outer wall of the covered stent to the free edge of sinus venosus defect

The diameter of the SVC is invariably smaller than the diameter of the cavoatrial junction and the upper part of the right atrium. This leads to a nonuniform "bell-bottom" or a "funnel-shaped" configuration for the covered stent. Even though the semi-compliant balloon apposes against the free edge of SVD, leading to its complete closure during balloon interrogation, minimal recoil of the expanded covered stent at its caudal end may reduce the apposition. The residual flows are identified on the edges of the covered stent on RUPV angiography [Figure 5].

In these cases, the lower end of the stent needs to be flared with a larger balloon than the one used for balloon interrogation and initial stenting [Figure 6]. A shorter balloon would be preferred to prevent the expansion of the stent at the level of RUPV to prevent its narrowing. If residual flow from lack of apposition persists even after postdilatation with a large balloon, a small low-profile occluder such as Amplatzer Duct Occluder II or Cribriform Occluder (Abbott, Plymouth, MN) or Konar-MF device (LifeTech Scientific, Shenzhen, PRC) may be used [Figure 7] to plug the shunt.[10,11] Insignificant residual flows may disappear on follow-up as the reduction in the size of the right atrium from a favorable remodeling increases the approximation of the atrial septum to the outer surface of the covered stent.[3]

Upper end of right upper pulmonary vein draining into the superior vena cava cranial to the covered stent

After balloon interrogation, when a covered stent is deployed across the cavoatrial junction, the cranial end of the stent should be positioned well above the drainage of the RUPV. Otherwise, the RUPV may continue to drain into the SVC, leading to a persistent shunt. A caudal migration of the stent balloon assembly during its expansion due to the bell-bottom shape of the cavoatrial junction may lead to the cranial end of the covered stent falling below the RUPV [Figure 8].

If this occurs, it not only leads to a residual flow from RUPV but also reduces the anchor of the stent in the SVC, thereby risking stent embolization. This can be addressed by implanting another covered stent of similar diameter overlapped cranial to the pulmonary vein to provide an adequate anchor zone [Figure 9]. The expansion of this second stent should be done with utmost care to prevent a second caudal migration.

Additional small pulmonary vein draining high in superior vena cava

In our cohort of 105 cases, an additional small RUPV draining into the SVC was identified in 14% of patients. These high-draining veins were not considered amenable for surgical redirection into the left atrium, as they would need placement of a baffle using a long patch running through the entire length of the SVC. Warden

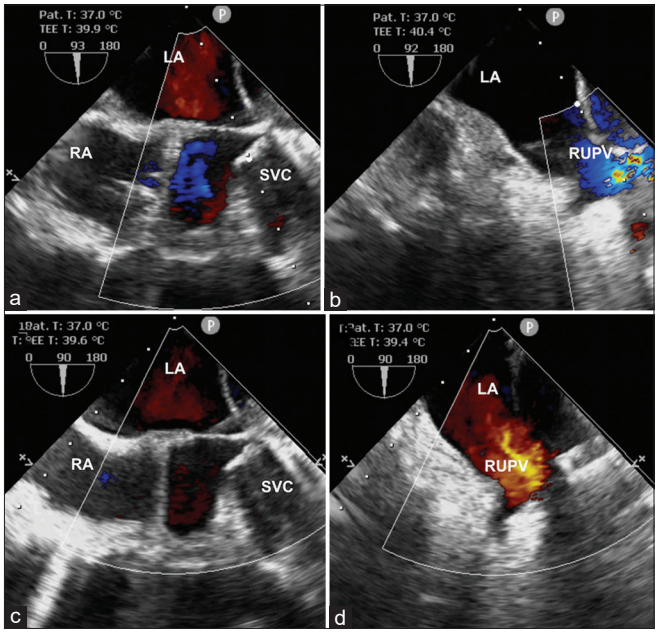

Figure 3: Bicaval view of transesophageal echocardiogram (a) following covered stent implantation in the superior vena cava (SVC) shows blue flows on color Doppler from the left atrium (LA) to the right atrium indicating residual shunt. The right upper pulmonary vein (RUPV) shows blue color flows (b), indicating flow directed toward the SVC. After deploying a second overlapping stent, the left-to-right shunt disappears (c), and RUPV flows (d) are red, directed toward the LA. RUPV: Right upper pulmonary vein, SVC: Superior vena cava, RA: Right atrium, LA: Left atrium

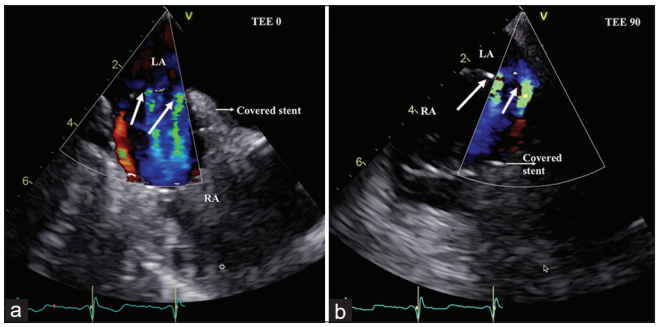

Figure 4: Transesophageal echocardiography with color Doppler in axial view (a) and bicaval view (b) shows two small fabric perforations in the covered stent measuring 2 mm each causing left-to-right shunt from the left atrium to the right atrium. TEE: Transesophageal echocardiography, RA: Right atrium, LA: Left atrium

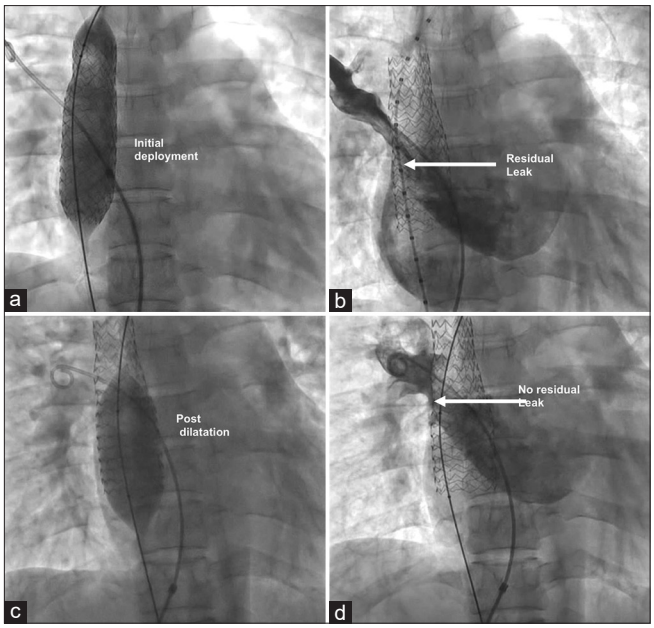

Figure 5: Following a covered stent deployment (a), a right upper pulmonary vein angiogram shows its redirection to the left atrium (b), but there is a small residual flow (arrow) along the right edge. After postdilatation of the lower end with a larger balloon (c), the residual flow disappears (d)

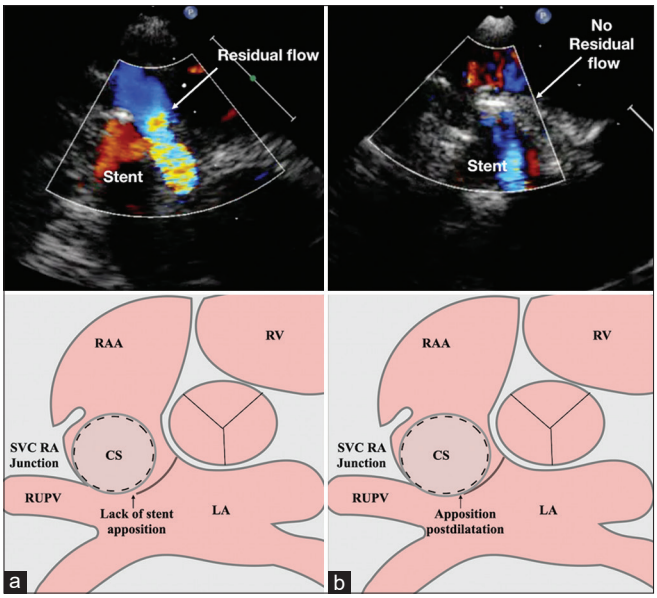

Figure 6: Transesophageal echocardiogram shows a residual flow at the sides of the stent (a) that disappears after postdilatation with a large balloon (b). The accompanying cartoon demonstrates the mechanism. RUPV: Right upper pulmonary vein, SVC: Superior vena cava, RA: Right atrium, CS: Covered stent, RAA: Right atrial appendage, LA: Left atrium, RV: Right ventricle

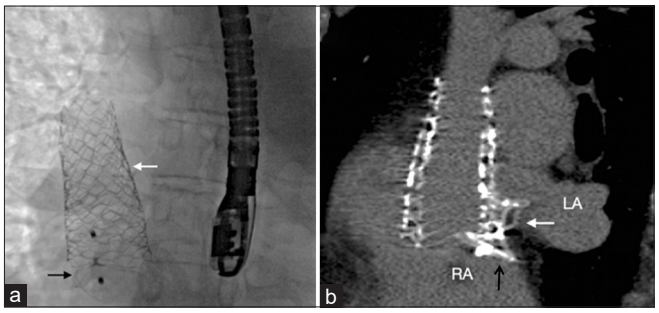

Figure 7: A small residual flow (a) across the edge of the stent (white arrow) that persists even after dilating the lower end with a larger balloon is closed by a Konar-MF device (black arrow). After 6 months, a computed tomographic image (b) shows the covered stent and the occluder. RA: Right atrium, LA: Left atrium

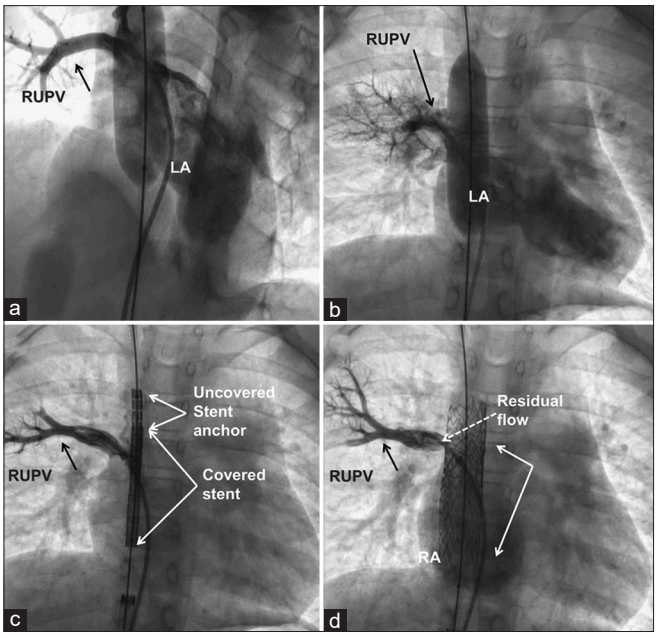

Figure 8: Balloon interrogation shows closure of interatrial shunt and redirection of the right upper pulmonary vein (RUPV) to the left atrium in left anterior oblique (a) and anteroposterior (b) projections. A covered stent overlapped above with an uncovered anchoring stent is positioned, ensuring that the upper end of the covered stent is above the RUPV (c), but caudal migration (d) leads to persistent flows from RUPV to the superior vena cava through the uncovered stent struts filling the right atrium. RUPV: Right upper pulmonary vein, RA: Right atrium, LA: Left atrium

procedure in such instances would necessitate a division of the SVC close to the innominate veins and a stretched anastomosis to the right atrial appendage. In many adults, the heart team decision often favored leaving the small additional pulmonary veins to drain to the SVC and accept the small left-to-right shunt. Furthermore, during transcatheter SVD closures, these veins were intentionally allowed to drain into the SVC.

This can be achieved by deploying the covered stent caudal to the drainage of the additional vein whenever the site of drainage of this vein is quite high, and the anchor zone is sufficient. If the drainage site is not very high and the anchor zone caudal to the drainage site is not sufficient, an uncovered stent is overlapped cranial to the covered stent that is mounted caudally in the same long balloon and deployed in such a way that the additional vein drains through the struts of the uncovered stent into the SVC [Figure 10].

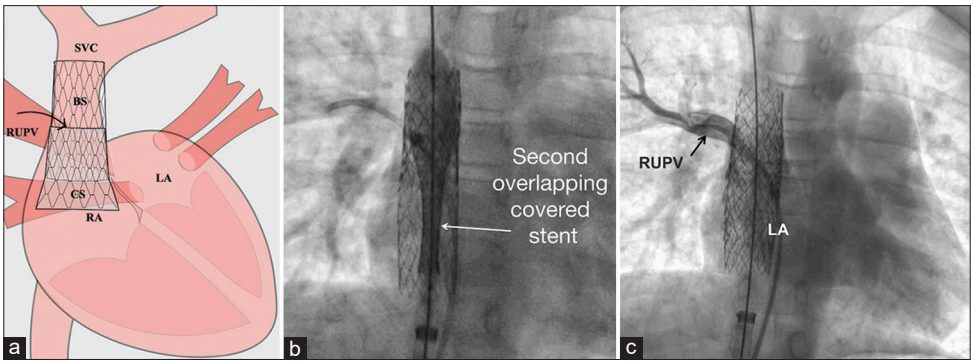

Figure 9: Cartoon (a) shows the upper end of the covered stent (CS) slipping caudally below the right upper pulmonary vein (RUPV) orifice, causing residual shunt through the side struts of the uncovered stent (BS). An overlapping CS (b) placed cranial to the RUPV stops the residual flows (c) and redirects the vein to the left atrium. RUPV: Right upper pulmonary vein, SVC: Superior vena cava, RA: Right atrium, CS: Covered stent, BS: bare (uncovered) stent, LA: Left atrium

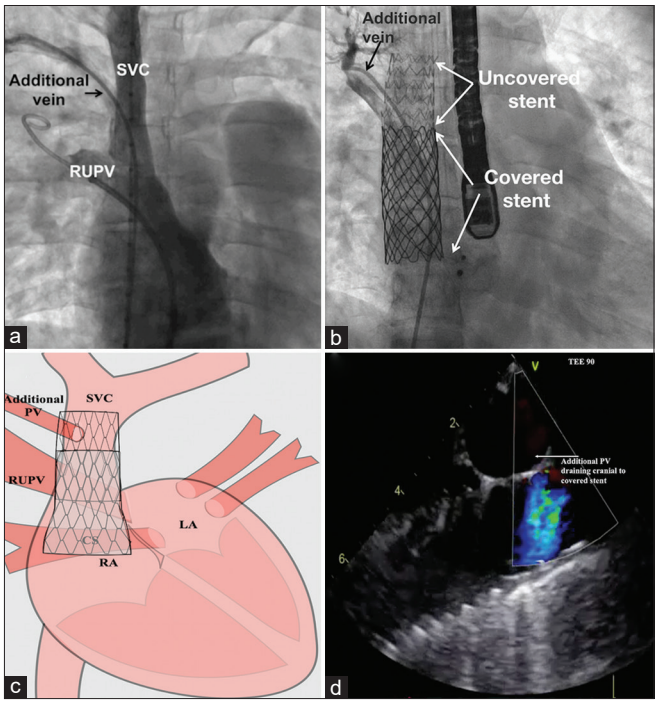

Figure 10: The main right upper pulmonary vein and additional small vein are cannulated separately (a) during an angiogram of the superior vena cava (SVC). A covered stent is deployed caudal to the additional vein to prevent its occlusion and its persistent drainage to the SVC (b), but an uncovered stent is overlapped at the upper end to provide an anchor (c). A bicaval view of the transesophageal echocardiogram (d) shows the flows from the additional pulmonary vein very high in the SVC. RUPV: Right upper pulmonary vein, SVC: Superior vena cava, RA: Right atrium, LA: Left atrium, PV: Pulmonary vein

Residual defect caudal to the covered stent due to its inadequate length

In most cases, the caudal-free edge of the SVD is separated very well from the oval fossa by a broad limbal tissue. The caudal extent of the SVD toward the oval fossa is variable. Excessive caudal extension of SVD is defined as its vertical diameter measured on a bicaval plane of TEE longer than the diameter of the cavoatrial junction measured on a horizontal plane.[2] The length of the covered stent is chosen to ensure that its lower end extends well below the free edge of the SVD. Nonuniform foreshortening of the covered CP stent leads to minimal shortening at the cranial SVC end and more in the caudal atrial end. This may predispose to a lack of coverage for the free caudal edge of the SVD.

If a significant foreshortening of the covered stent exposes the caudal edge of the SVD, TEE demonstrates a residual flow at the lower end of the stent [Figure 11]. Unlike the case of lack of apposition addressed by a further re-expansion of the lower end of the covered stent, a similar strategy here would worsen the residual flows. It is important to differentiate the lack of apposition from this residual shunt caudal to the stent due to its inadequate length. This shunt is addressed by placing an additional overlapping covered stent extending more caudally into the right atrium.

Left-to-right interatrial shunt from the oval fossa

A few patients may have a stretched patent oval foramen that may shunt from the left to the right atrium. During transcatheter SVD closure, a septal puncture is often done to monitor the RUPV pressures and perform pigtail angiograms from the vein.[3] It is also used for pulmonary vein protection techniques.[3] The orifice left after a septal puncture with an 8F sheath may persist without closure depending on the left atrial pressures, elasticity, and thickness of the atrial septum [Figure 12]. Moreover, 9%–16% of patients with SVD may have a small oval fossa defect [Figure 13]. Simultaneous closure of oval fossa defect using an occluder device during a covered stent exclusion of SVD is simultaneously carried out in such instances.[12]

THE RELATIVE FREQUENCY OF THESE DIFFERENT MECHANISMS

Publications of outcomes after covered stent exclusion

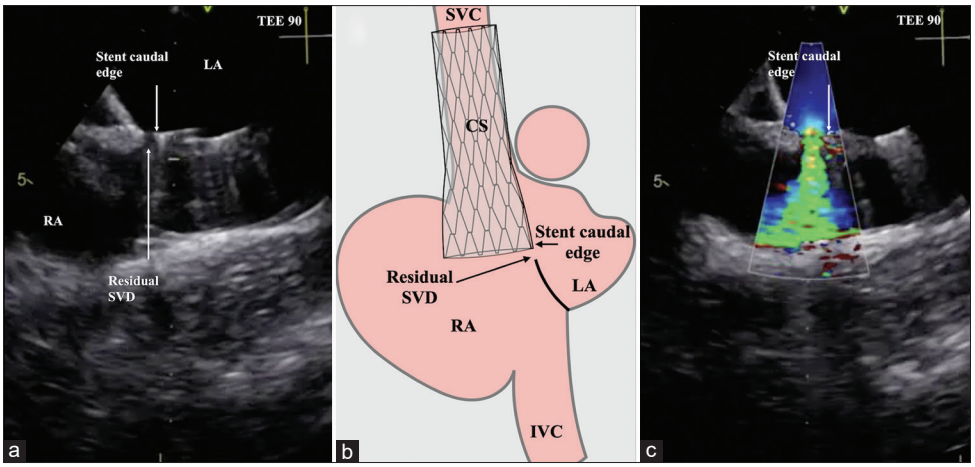

Figure 11: Bicaval view of transesophageal echocardiogram (a) shows a small gap between the free edge of the sinus venosus defect and the caudal edge of the stent, explained by a cartoon (b) with color Doppler flows (c). TEE: Transesophageal echocardiography, SVD: Sinus venosus defect, CS: Covered stent, RA: Right atrium, LA: Left atrium, IVC: Inferior vena cava

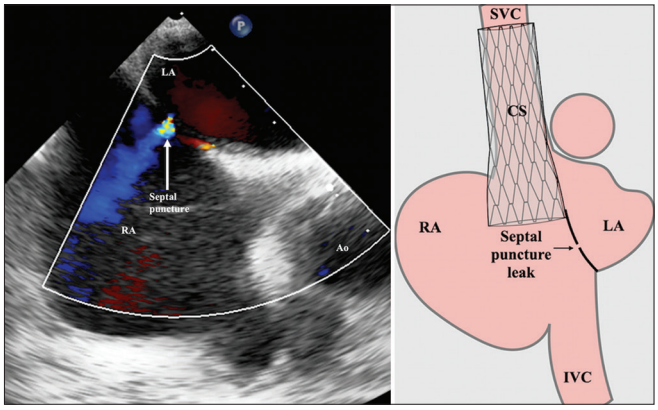

Figure 12: Transesophageal echocardiogram after a successful procedure shows a small left-to-right shunt from the left atrium to the right atrium through the septal puncture orifice, demonstrated by a cartoon. SVD: Sinus venosus defect, CS: Covered stent, RA: Right atrium, LA: Left atrium, IVC: Inferior vena cava

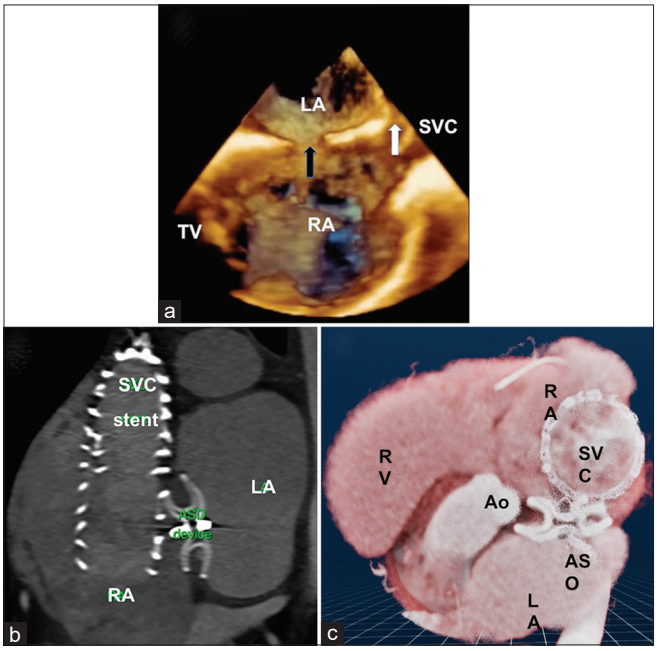

Figure 13: Bicaval view of three-dimensional transesophageal echocardiography (a) shows oval fossa defect (black arrow) and superior vena cava (SVC) straddling a sinus venosus defect (white arrow) between the left atrium and right atrium. Six months after combined intervention with a covered stent deployment in SVC and atrial septal occluder device, sagittal slice (b), and color-rendered computed tomography (c) images demonstrate the anatomy. SVC: Superior vena cava, RA: Right atrium, LA: Left atrium, RV: Right ventricle, TV: Tricuspid valve, ASD: Atrial septal defect

of SVD briefly mention about these mechanisms of residual left-to-right shunts without systematically classifying them as discussed above.[2-4] Among a total of 105 procedures of transcatheter closure of SVD in our institution over the last 7 years, a major fabric breach warranting placement of additional overlapping covered stent was observed in two patients, reported earlier.[9] A lack of apposition of the outer wall of the covered stent to the free edge of the SVD was often addressed immediately by a postdilation of the lower end of the stent with a larger balloon. Two patients had a significant flow (color Doppler width >6 mm on TEE). While a Konar-MF device addressed the shunt in one of the patients reported earlier, the other patient had hemodynamic features of restrictive left ventricular physiology that benefitted from the residual flows.[11] A caudal migration of the covered stent exposing the upper end of RUPV to allow its persistent drainage into the SVC identified in one patient was addressed by a cranially overlapping additional covered stent. Additional high-draining pulmonary veins were identified in 14% of our cohort. Additional oval fossa defect managed by a septal occluder device was reported in five of our patients.[12]

CONCLUSIONS

Transcatheter SVD closure is increasingly performed in

the current decade with a good success rate. Residual defects in transcatheter closure can be caused by various mechanisms. Awareness of the different mechanisms and their proper diagnosis by TEE imaging will help to assess the significance and address these residual shunts effectively. Each mechanism will warrant a tailored approach.

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Conflicts of interest

There are no conflicts of interest.

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