

# Zero-contrast transcatheter closure of sinus venosus defect in advanced renal failure

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## ABSTRACT

Transcatheter closure of sinus venosus defect (SVD) is an emerging intervention that utilizes a covered stent to redirect the right upper pulmonary vein to the left atrium. Preprocedural computed tomography analysis, as well as the interventional procedure, necessitates the use of radiographic contrast media. Contrast use is prohibited in patients with advanced kidney disease, who also carry high surgical risks of cardiopulmonary bypass. Transesophageal echocardiography-guided catheter intervention with zero contrast use is presented in this report, along with technical details about planning the procedure. Covered stent exclusion of SVD without contrast use has not been reported in the literature so far.

**Keywords:** Anomalous drainage, chronic kidney disease, contrast-induced nephropathy, left to right shunt, radiographic contrast media, right upper pulmonary vein

## INTRODUCTION

Sinus venosus defects (SVDs) caused by deficiency of the wall separating the superior vena cava (SVC) and right upper pulmonary vein (RUPV) lead to the anomalous drainage of the latter into the former. Surgery redirects the RUPV to the left atrium by creating a roof for the vein. Transcatheter correction using covered stents to create this roof is emerging as an alternative to surgery.<sup>[1,2]</sup> Preprocedural computed tomography (CT), as well as procedural angiography, necessitate the use of radiographic contrast media. Open-heart surgery, as well as contrast use during catheter interventions, worsen renal function in patients with chronic kidney diseases (CKD).<sup>[3,4]</sup> Advanced imaging tools allow the performance of catheter interventions without the use of contrast.<sup>[5,6]</sup> Transesophageal echocardiography (TEE) guided successful transcatheter correction of SVD in an adult with advanced CKD without the use of radiographic contrast is presented.

## CASE REPORT

A 56-year-old lady with nonoliguric CKD and systemic hypertension presented with exertional dyspnea and palpitations of 3-year duration. Clinical evaluation showed irregular heart rate from atrial fibrillation, elevated jugular venous pressures, pedal edema, cardiomegaly, and outflow ejection murmurs with wide fixed splitting of second sound. Echocardiography showed a large SVD with anomalous RUPV drainage to the SVC, right heart volume overload, moderate pulmonary hypertension, and right ventricular systolic dysfunction. Renal parameters showed urea of 127 mg/dL and creatinine of 4.1 mg/dL. The estimated glomerular filtration rate was 12 mL/min/1.73 m<sup>2</sup>. Advanced age and CKD increased surgical risks. The heart team decided on transcatheter closure after informed consent. A preprocedural CT pulmonary angiogram could not be performed due to CKD.

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**How to cite this article:** Thejaswi P, Sagar P, Sivakumar K. Zero-contrast transcatheter closure of sinus venosus defect in advanced renal failure. *Ann Pediatr Card* 2024;17:141-5.

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#### DOI:

10.4103/apc.apc\_29\_24

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Submitted: 20-Feb-2024 Accepted: 03-Apr-2024 Published: 20-Jul-2024

A large SVD with bilateral SVC was identified on TEE [Figure 1]. Left SVC drained to the right atrium through a roofed coronary sinus. The RUPV anomalously drained to the right SVC immediately above the cavoatrial junction, where the right SVC measured 22 mm. The vertical dimension of the SVD in the bicaval view was 19 mm. There were no additional oval fossa defects. On pulling the probe cranially, there were no demonstrable additional pulmonary veins draining high in SVC. Dopamine and diuretic infusion for 48 h optimized heart failure. Amiodarone was loaded to prepare for postprocedural electrical cardioversion of atrial fibrillation.

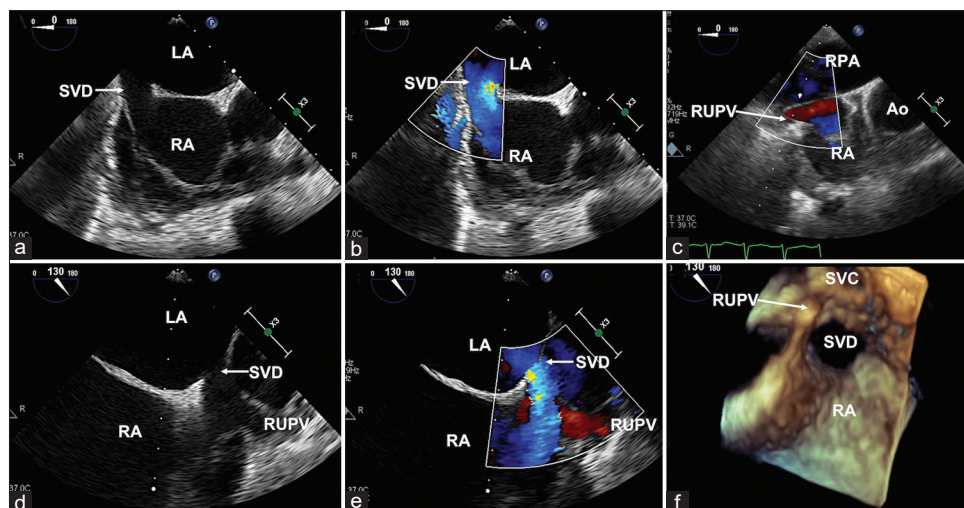
After elective intubation anesthesia for continuous TEE monitoring, bilateral femoral venous and left femoral arterial accesses were obtained. The pulmonary artery and atrial pressures were 43/20 (30) mmHg and 15 mmHg, respectively. A step up of 17% in the oximetry run indicated a shunt ratio of 2.5:1. The indexed pulmonary vascular resistance was 1.6 Wood units. After a TEE-guided transseptal puncture in the oval fossa, a pigtail catheter was placed in the RUPV through an 8F Mullins sheath advanced from the left femoral vein to simultaneously monitor left atrial and RUPV pressures. TEE confirmed the position of the RUPV catheter [Figure 2].

A Lunderquist wire (Cook Medical, Bloomington, IN) was parked in the right internal jugular bulb from the right femoral vein [Figure 3]. Balloon interrogation of the SVD was performed using a 26 mm × 8 cm BIB balloon (Numed Medical, Hopkinton, NY) selected by adding 4 mm to the transverse echocardiographic diameter of the cavoatrial junction. TEE allowed positioning the lower end of the balloon 2 cm below the caudal edge of the

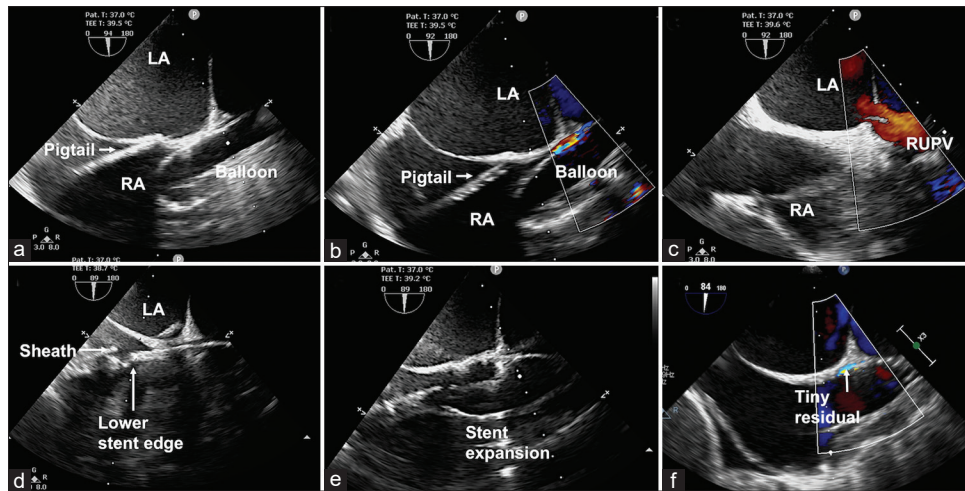
SVD. Fluoroscopy allowed centering the balloon on the pigtail in the RUPV. Upon balloon inflation, TEE showed near-total occlusion of the SVD and redirection of the RUPV to the left atrium. Simultaneous pressure recording in RUPV and left atrium showed a pressure of 21 mmHg, indicating restrictive left ventricular physiology. Advancing the tip of a large 22F Extralarge Check-Flo introducer (Cook Medical, Bloomington, IN) from the right femoral vein prevented a caudal milking down of the inflated balloon toward the right atrium. As the balloon interrogation showed the feasibility of transcatheter correction, a 79 mm non-foreshortening covered Zephyr stent (Sahajanand Laser Technology Limited, Gandhinagar, India) was crimped on the same balloon. The caudal end of the stent was positioned 2 cm below the free edge of the SVD. Deployment of the stent showed satisfactory closure of the SVD with negligible residual flows (3 mm residual color flow width). The spectral Doppler interrogation from RUPV toward the left atrium showed an unobstructed redirection of the blood flows.

Following stent exclusion of the SVD, the patient was electrically cardioverted to sinus rhythm. Coronary atherosclerotic narrowing was excluded by a limited angiography using 8 mL of contrast media for both vessels. After intravenous diuretics, she was extubated uneventfully within 6 h and discharged home after 3 days on Apixaban, aspirin, amiodarone, diuretics, and antihypertensives. There was no significant change in azotemia.

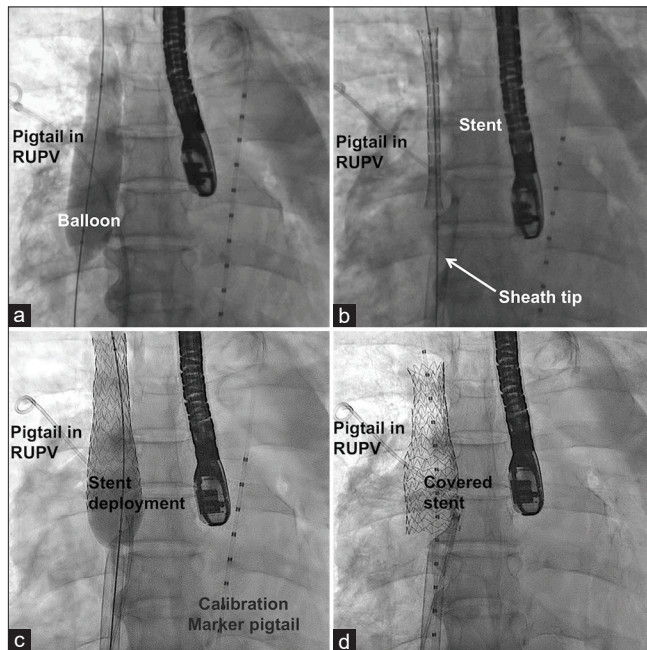
Six months following the procedure, there was significant symptom relief, and she remained in sinus rhythm. TEE showed an insignificant 3 mm residual shunt with a mean interatrial gradient of 5 mmHg [Figure 4].



**Figure 1:** Short axis view of transesophageal echocardiogram (a) with color Doppler images (b) show sinus venosus defect between left atrium (LA) and right atrium (RA). At a higher plane (c), anomalous drainage of the right upper pulmonary vein to the superior vena cava is seen. It is also demonstrated in bicaval view (d) with color flows (e). The right atrial enface view (f) of the three-dimensional echocardiogram clarifies the anatomy. SVD: Sinus venosus defect, RUPV: Right upper pulmonary vein, RPA: Right pulmonary artery



**Figure 2:** After transseptal puncture (a) from the right atrium (RA), a pigtail is advanced into the left atrium (LA). Balloon interrogation of the cavoatrial junction (b) confirms very negligible residual shunt. A rightward sweep from bicaval view (c) confirms unobstructed right upper pulmonary vein redirection to LA. Through a long sheath (d) with a tip placed in mid-RA, a covered stent crimped on a long balloon is advanced to the cavoatrial junction. The caudal end of the stent is positioned below the free edge of the sinus venosus defect. After stent expansion (e), there is a tiny insignificant residual shunt (f). RUPV: Right upper pulmonary vein



**Figure 3:** A long semi-compliant balloon (a) is inflated across the cavoatrial junction with a marker-calibrating pigtail catheter placed in the thoracic aorta. A covered stent (b) crimped on a long balloon is advanced through a long sheath whose tip is placed in the middle of the right atrium and deployed (c) across the cavoatrial junction. The calibrating pigtail assists in measuring the diameter and length of the covered stent (d) at various levels. Right upper pulmonary vein pressure is continuously monitored during the procedure. RUPV: Right upper pulmonary vein, LA: Left atrium, RA: Right atrium

Cardiac magnetic resonance showed unobstructed redirection of RUPV to the left atrium, a shunt ratio of 1:1, and a significant reduction in the size of the right heart chambers [Figure 5]. She continued to remain in nonoliguric renal failure with a creatinine level of 5 mg/dL. The patient provided written informed consent

for the procedure; the institutional review committee waived off patient consent for this anonymized report.

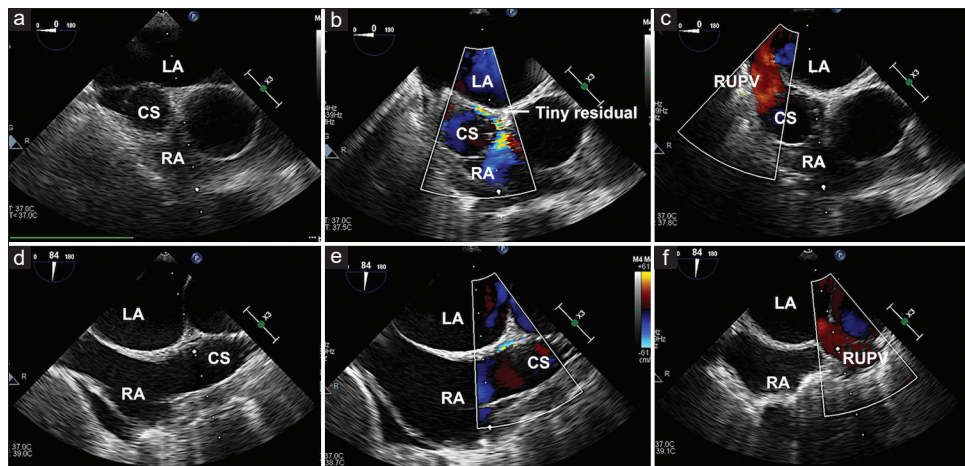
## DISCUSSION

Acute kidney injury (AKI) following contrast angiography increases morbidity, mortality, costs, and hospital stay.<sup>[7]</sup> Contrast-induced nephropathy (CIN) is caused by renal microcirculatory vasoconstriction, renal tubular cell ischemic injury, and oxidative stress leading to tubular necrosis. The severity of preexisting CKD is an incremental risk factor for the development of CIN. Other risk factors include older age, female sex, dehydration, anemia, diabetes mellitus, hypertension, heart failure, and co-administered nephrotoxic drugs. A dose of contrast media >4 mL/kg or >350 mL and contrast media exposure within the previous 72 h are risk factors for CIN.<sup>[7]</sup> When multiple risk factors for CIN coexist, as in our old hypertensive female patient with heart failure, all efforts must be made to reduce the probability and extent of further kidney injury.

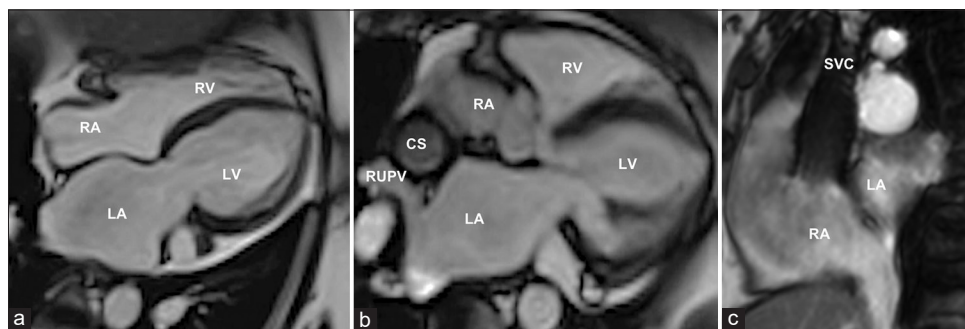
The success of transcatheter SVD closure depends on detailed preprocedural CT evaluation and clear angiographic demonstration of redirection of the RUPV into left atrium (LA) during balloon interrogation.<sup>[8]</sup> Multiple angiograms in SVC and RUPV during balloon interrogation and stent exclusion of SVD often require 150–300 mL of contrast. Preprocedural CT evaluation requires an additional 80 mL to analyze the anatomy, locate the RUPV drainage in SVC, identify additional high-draining vein(s), measure the SVC diameter to plan the balloon and stent size, and exclude other anomalies.

TEE is often used as an adjunct to CT during preprocedural planning as well as procedural angiography. However, the





**Figure 4:** Transesophageal echocardiogram after 6 months (a) shows a cross-section of the covered stent with the small residual shunt (b) between the left atrium (LA) and right atrium (RA). The right upper pulmonary vein (RUPV) is redirected (c) to LA behind the stent. Bicaaval view (d) with color flows (e) confirms the closure of the defect and redirection of RUPV (f). CS: Covered stent, RUPV: Right upper pulmonary vein



**Figure 5:** Normalization of right atrial (RA) and right ventricular size compared to left atrium (LA) and left ventricle on magnetic resonance imaging (a) after 6 months shows good remodeling. A modified view (b) shows an unobstructed right upper pulmonary vein behind the covered stent. Sagittal view (c) shows the stent in the superior vena cava sealing the atrial communication. RV: Right ventricular, LV: Left ventricle, RUPV: Right upper pulmonary vein, SVC: Superior vena cava

use of TEE as the sole imaging modality for transcatheter SVD correction has not been reported so far. Avoidance of contrast media before and during the procedure removes the biggest risk factor for the development of AKI and the worsening of CKD. In this procedure, TEE guided safe interatrial septal puncture, positioning the interrogating balloon, confirming the closure of the SVD and unobstructed redirection of RUPV to the left atrium, positioning the caudal end of the covered stent, and assessing the final outcome without any contrast media administration. When complex catheter interventions such as SVD closure increasingly utilize three-dimensional printed models or virtual reality recreations based on contrast-enhanced CT images, our report of zero-contrast interventional procedure carries practical significance in patients with advanced CKD.<sup>[9]</sup>

In elderly patients with oliguric CKD, complete SVD closure might cause pulmonary venous hypertension secondary to restrictive LV physiology. A small residual left-to-right shunt in our patient served as a fenestration to offset severe elevations in LA pressure. An adequate

response to postprocedural diuretic boluses in our nonoliguric patient allowed safe extubation without developing pulmonary edema despite the postprocedural left atrial pressure recording of 21 mmHg. Successful electrical cardioversion of atrial fibrillation to sinus rhythm also slowed the heart rate, improved the diastolic filling duration, and enabled left atrial pressure stabilization.

## CONCLUSIONS

When CKD in elderly patients with SVD leads to high surgical risks and prohibits contrast use for transcatheter correction, zero contrast intervention can be performed successfully and safely with meticulous planning and detailed intraprocedural TEE imaging.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The

patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

#### Financial support and sponsorship

Nil.

#### Conflicts of interest

There are no conflicts of interest.

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