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# TEE Guidance During Transcatheter Treatment of Superior Sinus Venosus ASDs With Partial Anomalous Pulmonary Venous Drainage

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**A NOVEL TRANSCATHETER APPROACH ADOPTED AND MODIFIED BY OUR GROUP HAS EMERGED AS AN** alternative to surgical closure of hemodynamically significant sinus venosus atrial septal defect (SVASD) with partial anomalous venous drainage (PAPVD) (1,2). A covered stent placed into the superior vena cava separates its connection to the left atrium and simultaneously redirects flow from the anomalous pulmonary veins into the left atrium, thereby eliminating the atrial shunt and correcting the PAPVD (**Figure 1**). Transesophageal echocardiography (TEE) is invaluable during the procedure and is now used routinely to document both the defect and the connections of the right pulmonary veins to the superior vena cava-right atrium junction (**Figures 2 and 3**, Videos 1, 2, 3, and 4), monitor patency of the right pulmonary veins during balloon assessment of the SVC (**Figure 4**) and during stent deployment (**Figure 5**, Videos 5, 6, 7, and 8), and assess the position of the fully deployed stent and residual shunts (**Figure 6**). This paper illustrates our approach to TEE imaging during closure of superior SVASDs with PAPVD using covered stents.

The use of covered stents for transcatheter closure of SVASDs is a viable alternative to surgery in suitable patients. TEE guidance is invaluable and involves consistent imaging of pulmonary venous flow during the procedure, which is key to reducing repeated contrast injections thus shortening fluoroscopy and procedure times.

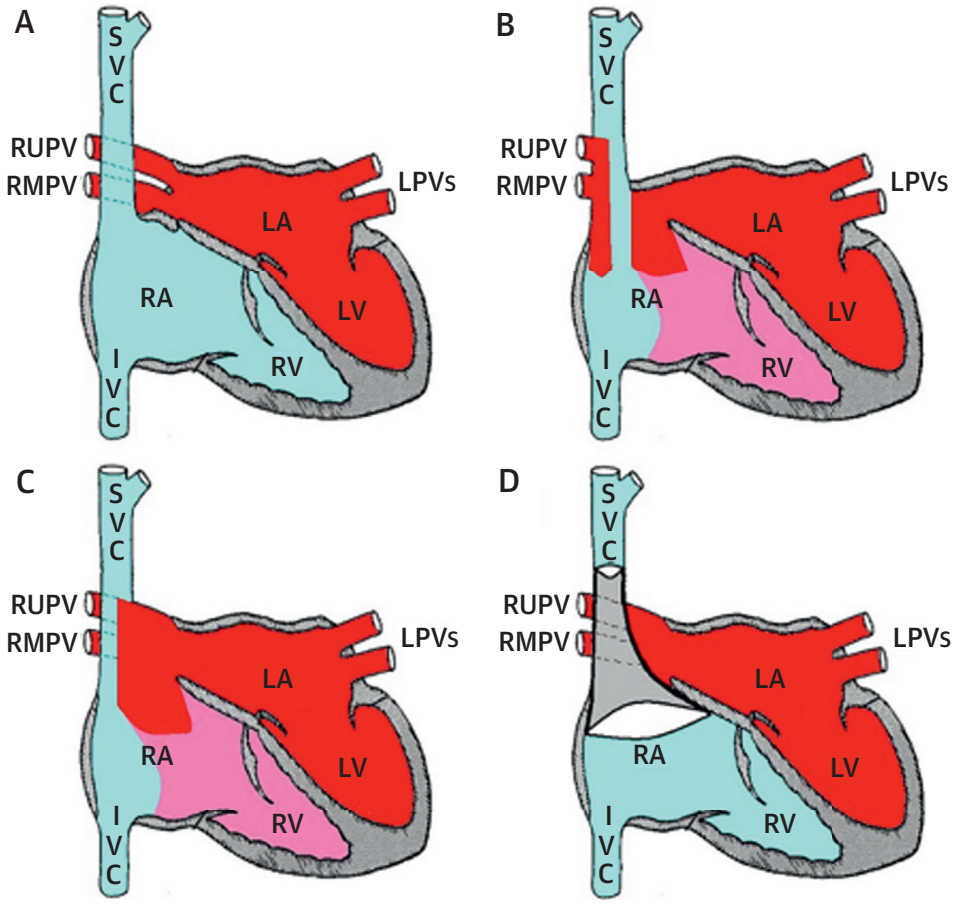
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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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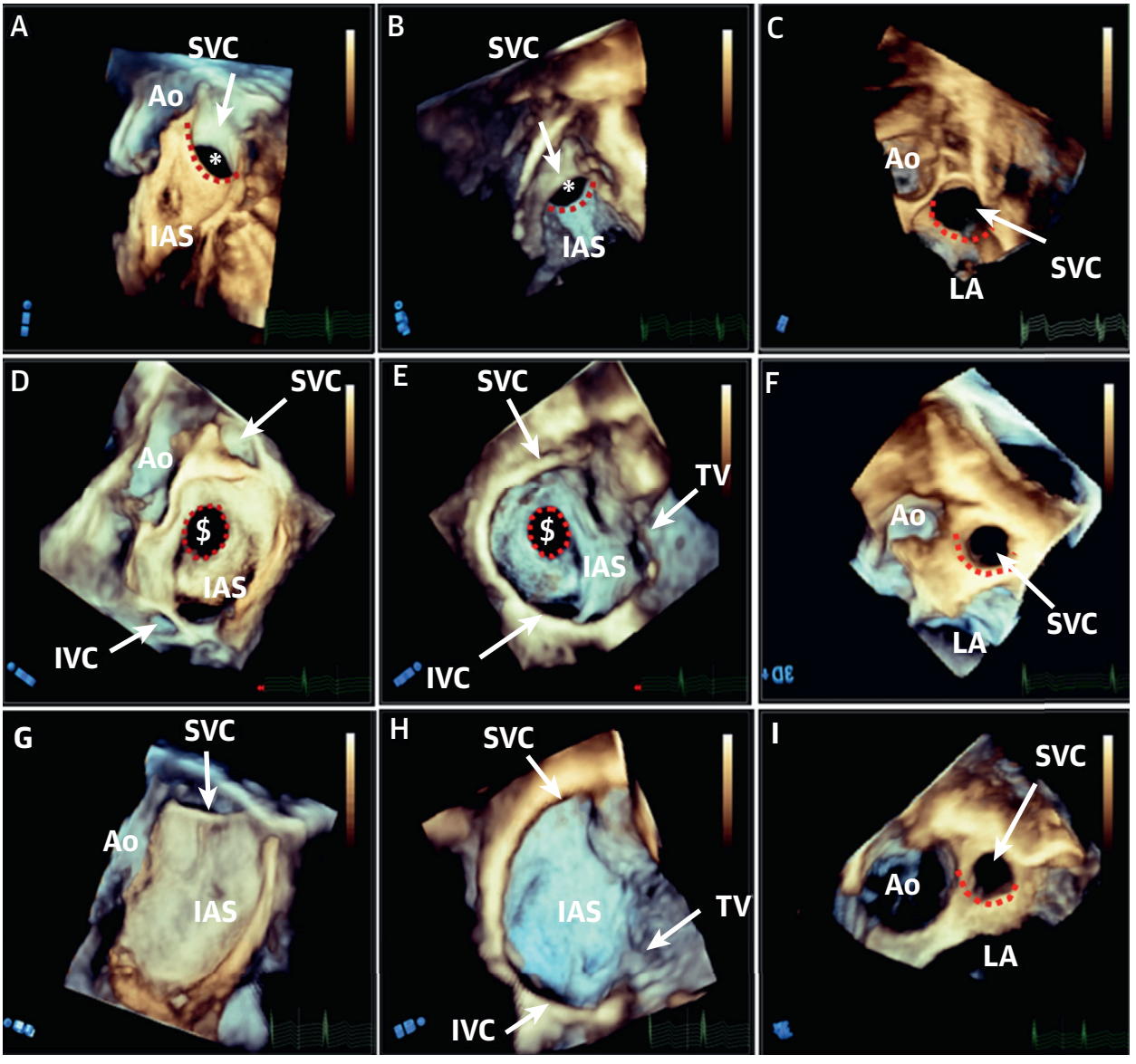
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**FIGURE 1** **Illustration of the Concept**



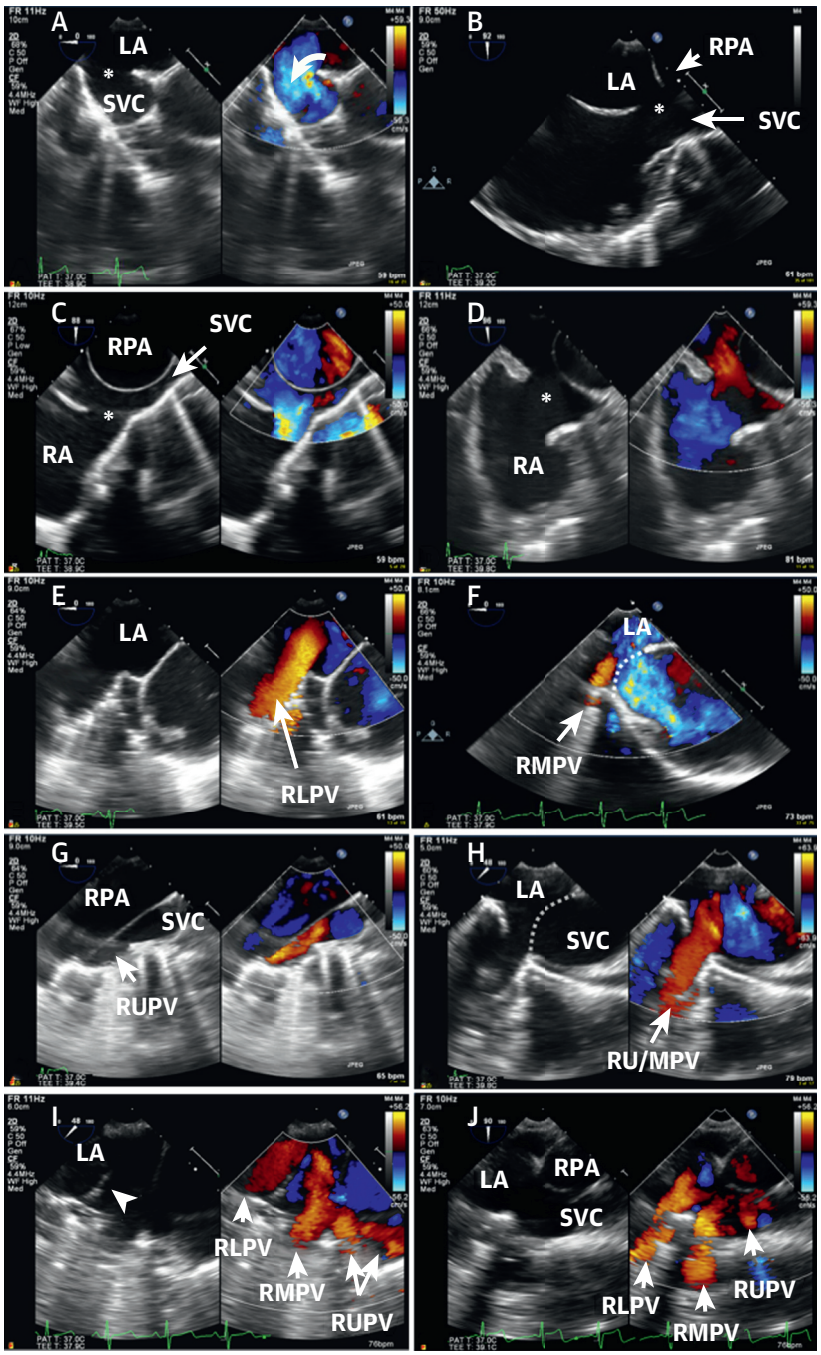

**(A)** In the normal heart, the right pulmonary veins (right upper pulmonary vein [RUPV] and right middle pulmonary vein [RMPV]) drain behind the superior vena cava (SVC) into the left atrium (LA). **(B)** The RUPV and RMPV drain directly into the SVC without continuity to the LA, while the RMPV may still drain behind a stent into the LA, the RUPV would be occluded by the stent—this type is not suited to stent correction. **(C)** In the commonest form of sinus venosus atrial septal defect with partial anomalous venous drainage, the wall between the RUPV and RMPV and the SVC is absent and the RUPV and RMPV drain into the back of the SVC-right atrium (RA) junction in continuity with the LA. **(D)** Stent correction is possible with the anatomy in **C**, where the covered stent **(gray)** closes off the sinus venosus atrial septal defect and facilitates redirection of the RUPV and RMPV into the LA. IVC = inferior vena cava; LPV = left pulmonary vein.

**FIGURE 2** **Comparison of 3-Dimensional-Rendered Transesophageal Echocardiography Images in a Superior Sinus Venosus ASD, a Secundum ASD, and an Intact IAS**



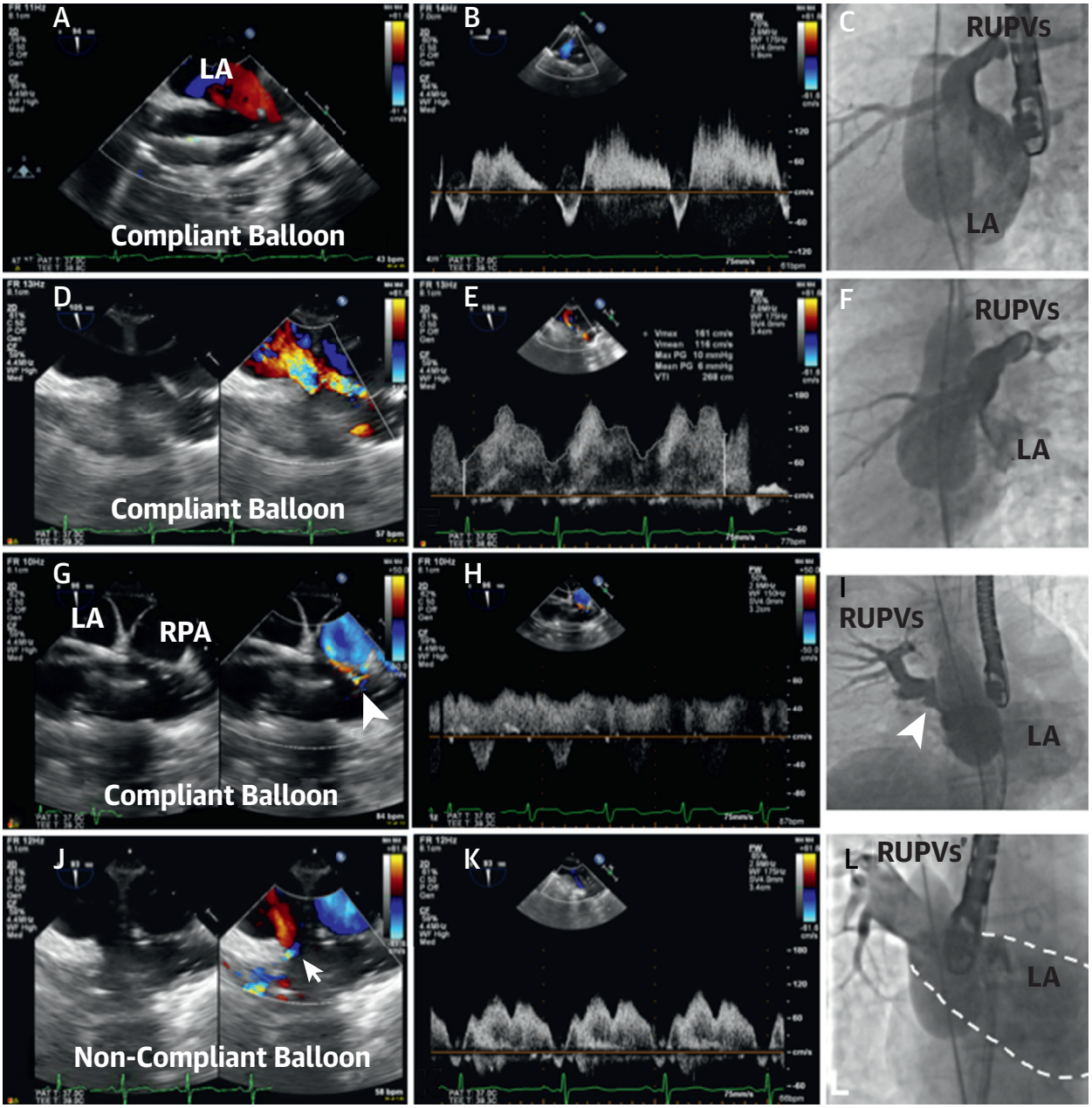

A superior sinus venosus atrial septal defect (ASD) **(*)** from **(A)** the LA aspect and **(B)** the RA aspect, in contrast to the appearance of a secundum ASD **($)** as seen from **(D)** the LA and **(E)** the RA aspects and with an intact interatrial septum (IAS) as seen from **(G)** the **left** and **(H)** the **right**. **(C)** The absence of the posterior wall of the SVC when seen from a cranial projection compared with that of **(F)** an intact SVC as it drains to the RA in a secundum ASD and **(I)** with an intact septum. Ao = aorta; TV = tricuspid valve; other abbreviations as in **Figure 1** (Video 1).

**FIGURE 3 Superior Sinus Venosus Atrial Septal Defect and Right Pulmonary Veins**



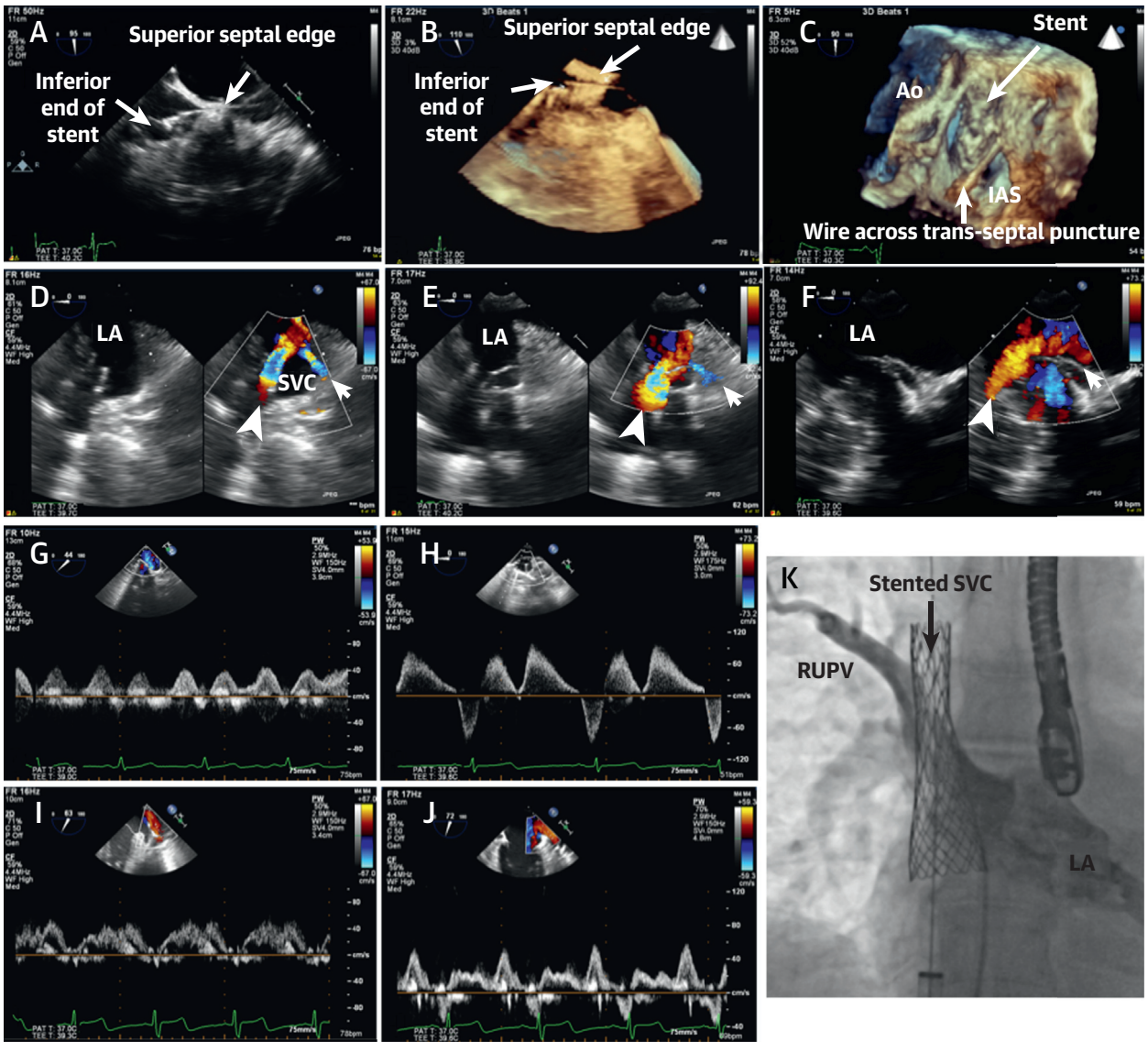

**(A)** The 0° midesophageal view demonstrating a sinus venosus atrial septal defect **(*)** with left-to-right shunting **(arrow)** and **(B)** in a bicaval view. **(C)** Bicaval view displaying the common appearance with a markedly dilated right pulmonary artery (RPA). **(D)** A large defect with deficient secundum septum. **(E)** The 0° midesophageal view demonstrating the right lower pulmonary artery (RLPV) to the LA; **(F)** the RMPV to the LA at the junction of the defect **(dotted line)** and **(G)** the anomalous RUPV to the SVC. **(H)** Short-axis high-esophageal view at 45° demonstrating a confluence of RU and RM veins entering at the SVC-LA-RA junction (**dotted line** demonstrating the deficient SVC-septal wall). **(I)** The anomalous RUPV and RMPV close to the SVC-RA junction with the RLPV draining to the LA relative to the interatrial septum **(arrowhead)**. **(J)** Bicaval midesophageal view with rightward rotation showing the pulmonary veins entering the LA (RLPV) and the SVC-RA junction (RUPV/RMPV). Abbreviations as in **Figures 1 and 2** (Videos 2 to 4).

**FIGURE 4** **Right Pulmonary Vein Assessment During Balloon Occlusion of the SVC**



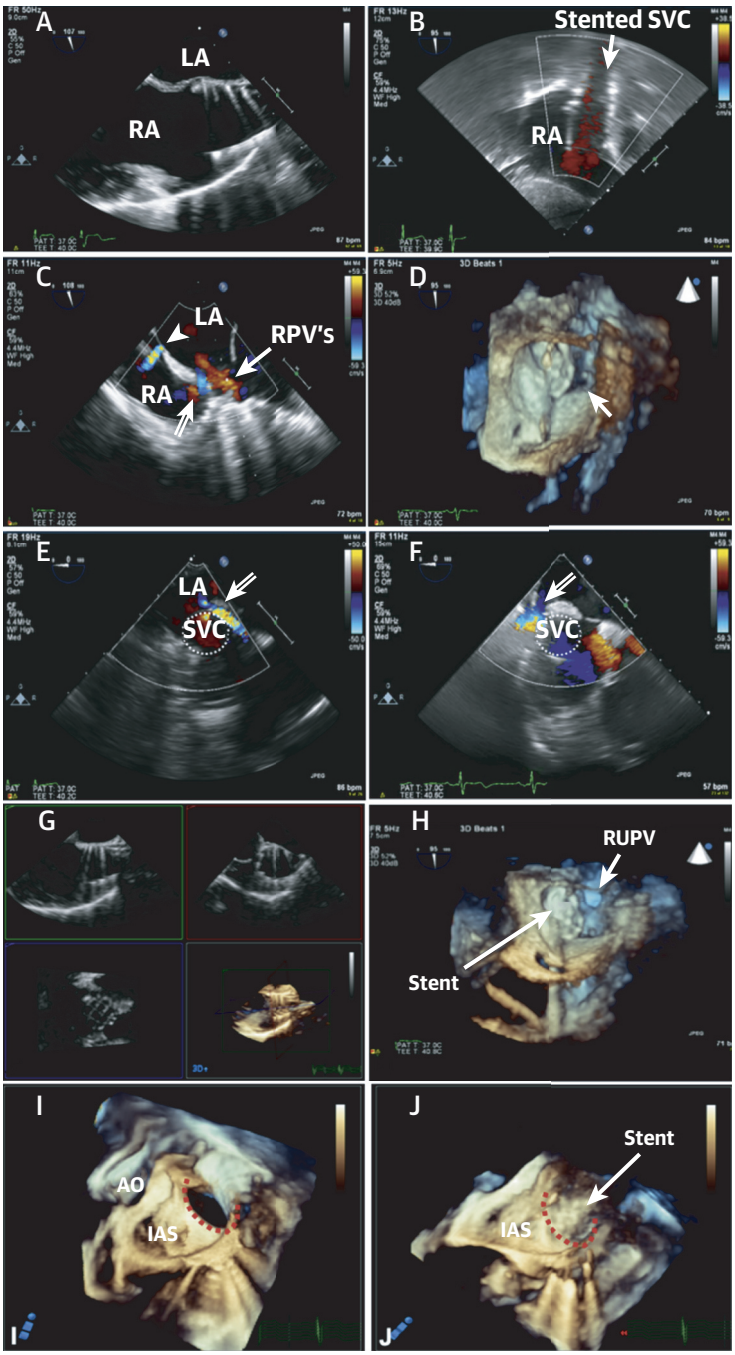

**(A to C)** Unobstructed flow. **(A)** Bicaval high-esophageal view demonstrating laminar flow in the redirected pulmonary veins. **(B)** Pulsed-wave (PW) Doppler depicts phasic flow with raised velocity and A-wave reversal due to increased LA pressures with defect occlusion. **(C)** RUPV angiography (lateral projection) shows unobstructed flow to the LA. **(D to F)** Partial obstruction. **(D)** Turbulent pulmonary venous flow with color flow aliasing. **(E)** PW Doppler demonstrates an increased mean gradient retaining the phasic wave form without return to baseline, indicating restriction. **(F)** RUPV angiogram shows contrast hold up in the RUPV and reduced flow to the LA. **(G to I)** Complete obstruction. **(G)** Limited flow seen **(arrowhead)**. **(H)** PW Doppler is faint and continuous, without returning to baseline. **(I)** RUPV angiogram (anteroposterior projection) shows limited flow to the LA and contrast hold up in the RUPV **(arrowhead)**. **(J to L)** Noncompliant balloon. **(J)** Small residual shunt **(arrow)** but **(K)** no obstruction to flow. **(L)** RUPV angiogram (anteroposterior projection) shows unobstructed flow to the LA behind the balloon **(dashed lines)**. Abbreviations as in Figures 1 and 3.

**FIGURE 5** **Deployment of the Stent**




**(A to C)** Stent position. **(A)** Bicaval 2-dimensional and **(B)** 4-dimensional views demonstrating the inferior end of the stent in relation to the superior septal edge. **(C)** Four-dimensional bicaval view showing the position of the stent seen through the defect from the LA. **(D–F)** Stent dilation. **(D)** The 0° midesophageal view demonstrating patency of the RUPV **(large arrowhead)** and residual defect **(small arrow)** as the defect is **(E)** slowly reduced and **(F)** eventually eliminated. **(G)** PW Doppler in the right pulmonary veins before stenting with a low-velocity biphasic flow pattern changes to reversal of the S and D waves and appearance of a prominent A-wave due to the increase in LA pressures after defect closure. An identical pattern is seen in the left PVs **(I)** before and **(J)** after stent deployment. **(K)** The RUPV angiogram in the anteroposterior projection shows unobstructed PV flow to the LA. Abbreviations as in Figures 1, 2, and 4.

**FIGURE 6 Final Evaluation of the Stent and Residual Defects**




Bicaval **(A)** midesophageal and **(B)** transgastric views show a covered stent enclosing the sinus venosus atrial septal defect flow with color Doppler demonstrating laminar stent flow. **(C)** Transeptal puncture shunt **(arrowhead)** and residual shunt at the inferior margin of the defect **(double arrow)** with redirected RUPV flow. **(D)** Bicaval 4-dimensional view (LA looking up toward the SVC stent) shows the same residual defect **(arrow)**. The 0° midesophageal view demonstrating a small residual shunt **(E)** anteriorly at the crest of the interatrial septum **(arrow)** and **(F)** posteriorly along the wall of the atria after use of a protective balloon in the pulmonary vein. **(G)** Multiplanar reformatting imaging demonstrates the stent in multiple projections. **(H)** Live 3-dimensional image (LA aspect) shows the stent baffling the SVC to the RA and the RUPV redirected to the LA. Three-dimensional rendered view (LA aspect) showing the sinus venosus atrial septal defect **(I)** before and **(J)** after stenting (Videos 5 to 8). Abbreviations as in **Figures 1 to 3**.

## AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**KEY WORDS** adult congenital heart disease, partial anomalous pulmonary venous drainage, structural intervention, superior sinus venosus ASD, transesophageal echocardiography

**APPENDIX** For supplemental videos, please see the online version of this paper.