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# Transcatheter Closure of Ruptured Sinus of Valsalva: Different Case Scenarios with Follow-up

Ahmed Moawad, MD; El-Farghaly Hala, MD; Ahmed S. Youssef, MSc

## Introduction

A sinus of Valsalva aneurysm (SOVA) is a cardiac anomaly that can be congenital or acquired.<sup>1</sup> Sinus of Valsalva aneurysms comprise 1% of all congenital heart entities.<sup>2</sup> Associated defects in congenital SOVAs include supracristal or perimembranous Ventricular Septal Defects (30%–60%), bicuspid aortic valves (15%–20%), and aortic regurgitation (44%–50%).<sup>3</sup>

A congenital SOVA is usually clinically silent, but may vary from a small, asymptomatic dilatation detected on transthoracic echocardiography (TTE) to symptoms of intracardiac shunt caused by rupture of the SOVA mostly into the right side of the heart.<sup>4</sup> When these aneurysms rupture, intracardiac shunting will lead to deterioration in cardiac function.<sup>5</sup>

Approximately 25% of reported SOVA cases are clinically asymptomatic.<sup>3</sup> A patient with a ruptured SOVA may present with acute chest pain, subacute shortness of breath on exertion or at rest (chronic heart failure), and/or acute cardiogenic shock.<sup>6</sup>

SOVA are thin-walled outpouchings of the sinus of Valsalva and may be tubular or saccular in shape.<sup>7</sup> Approximately 65%–85% of SOVAs originate from the right sinus of Valsalva, about 10%–30% from the noncoronary sinus, and those originating from the left sinuses comprise <5%.<sup>8</sup> The right sinus of Valsalva is the most commonly involved sinus and usually ruptures into right heart chambers. Less commonly, they rupture into left heart chambers (left atrium, left ventricle [LV]), pulmonary artery, interventricular septum, or the pericardial cavity.<sup>7</sup>

The first formal classification system for SOVA was proposed by Sakakibara and Konno in 1962; however, it is rarely used in clinical practice. The new modification for Sakakibara classification is illustrated in **Table 1**.<sup>5</sup>

**TABLE 1** Classification Systems for Rupture sinus of Valsalva<sup>5</sup>

Type	Sakakibara classification	Modified Sakakibara classification
I	Originating from left part of right coronary sinus; protruding into conus of right ventricle, just beneath commissure of right and left pulmonary valves	Protrusion and rupture into right ventricle just beneath pulmonary valve
II	Originating from central part of right coronary sinus; protruding into right ventricle; penetrating crista supraventricularis	Penetration and rupture into or just beneath crista supraventricularis of right ventricle
IIIv	Originating from posterior part of right coronary sinus; protruding into right ventricle, just beneath septal leaflet of tricuspid valve; penetrating membranous septum	Penetration and rupture into right ventricle adjacent to or at tricuspid annulus
IIIa	Originating from posterior part of right coronary sinus; protruding into right atrium, near commissure of septal and anterior leaflets of tricuspid valve	Penetration and rupture into right atrium adjacent to or at tricuspid annulus
IV	Originating from right part of noncoronary sinus; protruding into right atrium, near septal leaflets of tricuspid valve	Protrusion and rupture into right atrium
V		Other rare conditions (eg, rupture into left atrium, pulmonary artery, left ventricle, or other structures)

RSVA, Ruptured sinus of Valsalva aneurysm.

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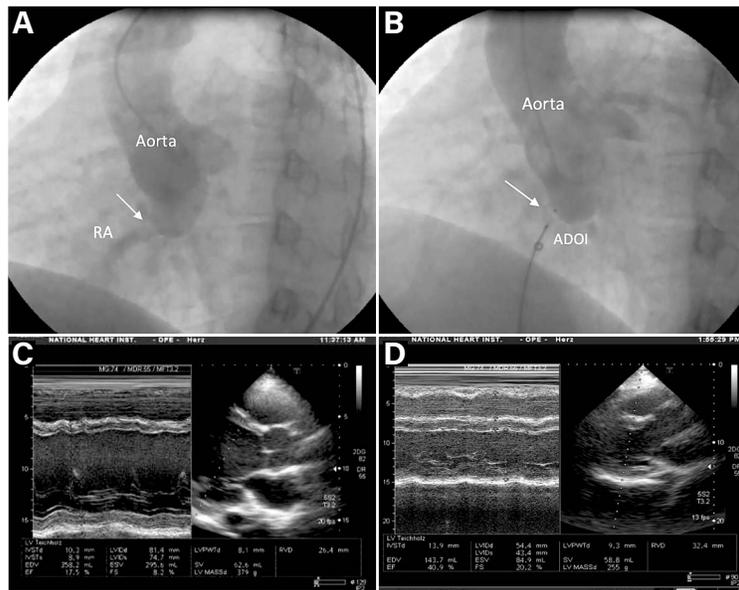
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Surgical repair via cardiopulmonary bypass is the standard management for a ruptured SOVA. However, use of a successful transcatheter closure (TCC) has been increasingly reported nowadays, mainly in single case reports.<sup>9</sup> No single report in the literature has described the different scenarios of right SOVA (RSOVA) and long-term follow-up of transcatheter closure (TCC).

## Methods

Different case scenarios of patients who had undergone TCC by members of the Pediatric Congenital Heart Disease Department in the National Heart Institute of Egypt with a follow-up period over six to 10 years are presented in this report.



**FIGURE 1** Aortography and M-mode echocardiography before and after closure of Case 1. **A)** Aortography shows rupture of right sinus of valsalva into right atrium (white arrow). **B)** Deployment of ADO1 device (white arrow). **C)** M-mode of LV in parasternal long axis view (PLAX) at presentation showing LVEF±17.5%. **D)** M-mode three years after closure showing improved LVEF to ±40.9%.

## Patient Characteristics

Ten cases with ruptured sinus of Valsalva were selected between 2007 and 2010 for TCC. The median age was 27 years (ranging from 12 to 50 years). Three of them were females, and seven were males. All patients presented with deteriorating heart failure symptoms (New York Heart Association [NYHA] Class III to IV). One patient presented with hypertensive emergency (**Case 2**), and one patient was in cardiogenic shock (**Case 6**). Comprehensive clinical examination for all cases was done and revealed bilateral basal crackles, congested neck veins, and enlarged tender livers; in addition, a continuous murmur (more accentuated in diastole) was heard along the left parasternal border with a S3 galloping rhythm. Associated defects were severe aortic coarctation (CoA) with bicuspid aortic valve and grade III/IV aortic regurgitation (**Case 2**); another two cases presented with bicuspid aortic valve, and one case had a perimembranous outlet ventricular septal defect (VSD) that was surgically closed three years before presentation (**Case 5**). One patient had previous TCC by an Occlutech® PDA occluder for a ruptured sinus of Valsalva at another center. Chest X-ray, electrocardiogram, and

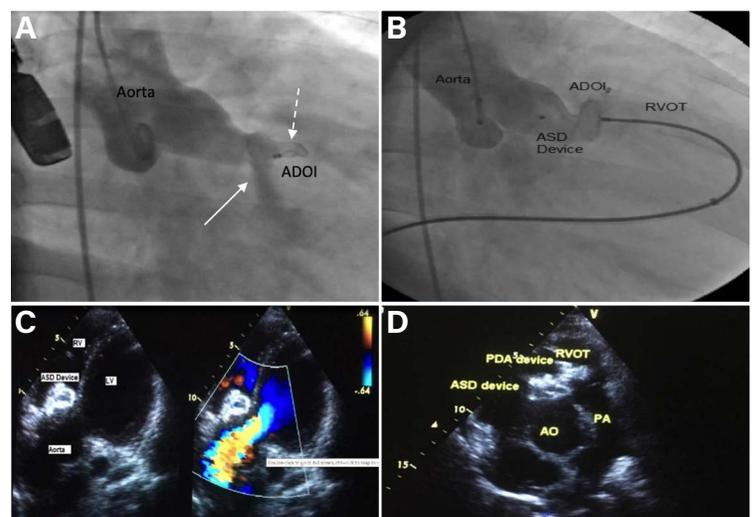
transthoracic echocardiography (TTE) with color Doppler were done and revealed left ventricular (LV) dilatation in all patients and deterioration of systolic function in five cases.

Upon obtaining informed written consent, patients were sent to the Cath Lab for further evaluation using fluoroscopy and intraoperative transesophageal echocardiography (TOE). During cardiac catheterization, their diagnoses were confirmed, and the relationship of the defect with the aortic valve and coronary arteries was revealed.

The defect was properly evaluated when the RSOVA diameter was measured at the aortic end and at the site of rupture. Echocardiography revealed RSOVA from the right coronary sinus to right atrium (RA) in three cases and right ventricular (RV) outflow in seven cases.

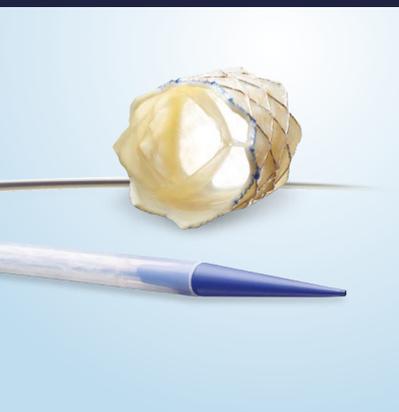
## Procedure

All procedures were performed under local anesthesia and sedation (with the exception of two cases; one was a child and the other one was in cardiogenic shock) through femoral artery and vein approach. Heparin (100 IU/ Kg), and ceftriaxone were administered. Left and right heart pressures were recorded, and aortography was taken in different projections with TOE guidance. The left anterior oblique view with cranial projection (**Figure 1A and B**) was preferred for SOVA that had ruptured into the RA, and the right anterior oblique view (**Figure 2A and B**) for SOVA that had ruptured into the RV outflow tract (RVOT). Additional views were taken if necessary. The defect was crossed by hydrophilic guiding wire (0.035 in. angled tip glide wire [Terumo Inc., Japan]) from the arterial side, followed by a multipurpose 4F catheter. A 260 cm long Amplatz super stiff J-tip wire (Boston Scientific, SCIMED, Natick, MA, USA) was then advanced, snared from the venous side (either pulmonary artery or superior vena cava) by Amplatz gooseneck snare (Microvena, White Bear Lake, MN, USA), and exteriorized out of the femoral vein to create an arteriovenous loop.



**FIGURE 2** Aortography and TTE before and after RSOVA closure of Case 4. **A)** Aortography shows large defect between the aorta and RVOT (white arrow) beside the occluder device (dotted arrow). **B)** Aortography post ASD device deployment showing no residual shunt, confirmed by color Doppler TTE. **C)** TTE apical five chamber view with color Doppler showing no residual. **D)** TTE short axis view showing both PDA and ASD devices in place.

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\*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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\*The term "stent fracture" refers to the fracturing of the Melody TPV. However, in subjects with multiple stents in the RVOT it is difficult to definitively attribute stent fractures to the Melody frame versus another stent.

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The Melody Transcatheter Pulmonary Valve and Ensemble II Transcatheter Delivery System has received CE Mark approval and is available for distribution in Europe.

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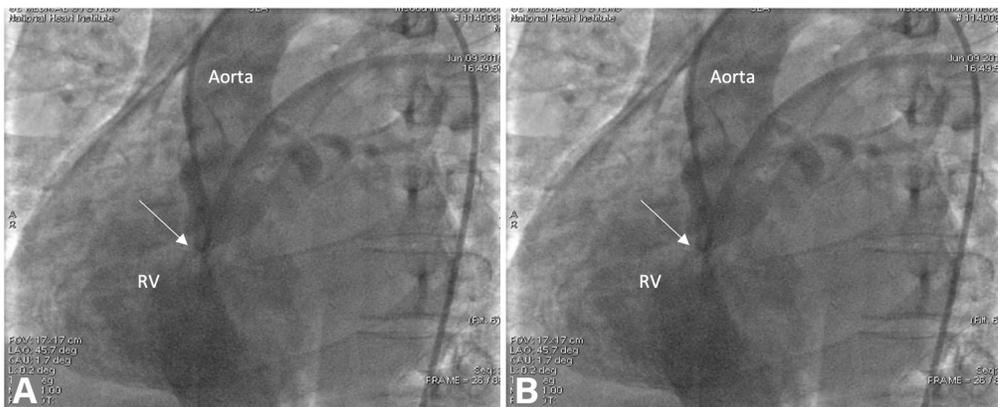
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**TABLE 2** Clinical, echocardiographic, and haemodynamic baseline variables; procedural variables; outcomes and complications.

Case no.	Age/gender	NYHA class	LVD/LVEF	Previous surgery	Associated lesions	RSOVA location	Defect size	Device used	Device size	Residual shunt/procedure-related AR at discharge	Residual shunt/procedure-related AR at follow up	NYHA class at follow up	LVD/LVEF at follow up
1. (figure 2)	36/m	IV	Dilated/depressed	None	None	RCC to RA	4	ADOI	6/8	None/none	None/none	I	improved
2. (figure 3)	12/m	IV	Dilated/depressed	None	Bicuspid AV, CoA.	RCC to RA	5	ADOI	8/10	None/none	None/none	III	Mild improve/then deteriorated
3. (figure 4)	50/f	III	Dilated/depressed	None	Bicuspid AV	RCC to RVOT	6	ADOI	8/10	Mild/none	Mild/none	I	improved
4. (figure 5)	32/m	III	Dilated/depressed	Previous RSOVA device closure	None	RCC to RVOT	10	ASDO	12	None/none	None/none	I	improved
5. (figure 6)	17/f	III	Dilated/depressed	VSD repair	None	RCC to RVOT	5	ADOI	8/10	Mild/none	None/none	I	improved
6. (figure 7)	31/m	IV (cardiogenic shock)	Dilated/depressed	None	None	RCC to RVOT	8	Coccon duct occluder	10/12	None/none	None/none	I	improved
7. (figure 8)	15/m	III	Dilated/Fair	None	None	RCC to RVOT	6	ADOI	8/10	None/none	None/none	I	improved
8	35/m	III	Dilated/Fair	None	None	RCC to RVOT	7	ADOI	10/12	None/none	None/none	I	improved
9	25/f	III	Dilated/Fair	None	None	RCC to RA	4	ADOI	6/8	None/none	None/none	I	improved
10	18/m	III	Dilated/Fair	None	Bicuspid AV	RCC to RVOT	5	ADOI	8/10	None/none	None/none	I	improved

ADO, Amplatzer duct occluder; ASD, Amplatzer septal occluder; AR, aortic regurgitation; LVD, left ventricular dimensions; LVEF, left ventricular ejection fraction; RA, right atrium; RCC, right coronary cusp; RVOT, right ventricular outflow tract; CoA, coarctation of aorta; VSD, ventricular septal defect.



**FIGURE 3** Aortography before and after RSOV closure of Case 6. **A)** Aortography shows rupture right SOV aneurysm into RV (white arrow). **B)** Post occlusion aortography shows Coccon duct occluder effectively closing the RSOV with no residual shunt (white arrow).

The RSOVA was measured at its aortic end and at the rupture site using both TOE and angiography. The larger of the two measurements was considered for device selection. In all cases, an Amplatzer duct occluder I ([ADOI] AGA Medical Corporation, Plymouth, MN, USA) was used to close the defect, except in Case 4 (Figure 2), in whom an Amplatzer Septal Occluder (ASDO, HyperionTM, Comed BV, Netherlands) device was used and Case 6 (Figure 3) in whom a Coccon PDA occluder was used. The chosen ADOI devices were 2 mm larger than the defect size, while the Atrial Septal Defect (ASD) device diameter was 12 mm.

The Amplatzer delivery sheath (St Jude Medical, Cardiovascular Division, St Paul, MN) was advanced from the femoral vein over the stable arteriovenous loop and stabilized into the ascending aorta across the defect. An appropriately sized ADOI with its attached delivery cable was then placed through the delivery system, and its aortic disk was deployed into the ascending aorta. The whole assembly was pulled back until the aortic disk impeded the inlet (aortic end) of the SOVA as visualized on TOE.

After confirming precise placement, the rest of the ADOI was deployed on the right side across the defect. During this maneuver,

gentle traction was exerted on the delivery cable, but special care was taken to ensure aortic disk seating on the aortic side without slippage into the aneurysm. The device was then released from the delivery cable only after angiography and TTE were repeated to confirm closure of the defect and device stability, view any impingement upon the aorta or coronary arteries, and make certain that there was no significant aortic or tricuspid regurgitation (AR or TR).

All patients except Case 6 were discharged 48 hours after the procedure. They were studied clinically and by TTE at discharge and then one, three, and six months and annually thereafter. All patients received both aspirin 150 mg once daily for six months and clopidogrel 75 mg/day for one month.

## Results

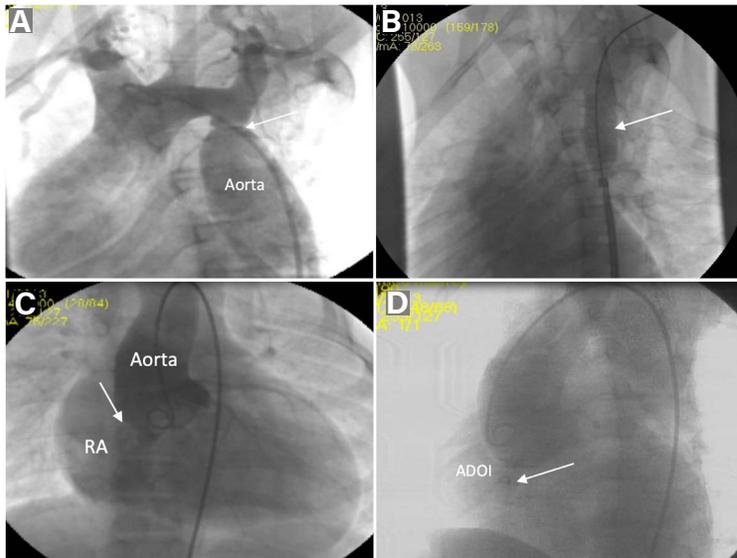
Patients characteristics are summarized in Table 2. The Qp/Qs range was 1.6–3.7, and the diameter at the rupture site was 4–8 mm. The mean fluoroscopy time was 16.3 ± 8.8 min, and the mean procedure time was 41.6 ± 20.7 min. Only two cases had mild residual shunt after deployment. All cases showed dramatic response upon closure within 48 hours; even the patient who presented with cardiogenic shock was weaned from cardiac support and discharged safely from hospital. Follow-ups over six to 10 years showed maintenance of a symptom-free period and functional class improvement.

## Deployment Success and Failure

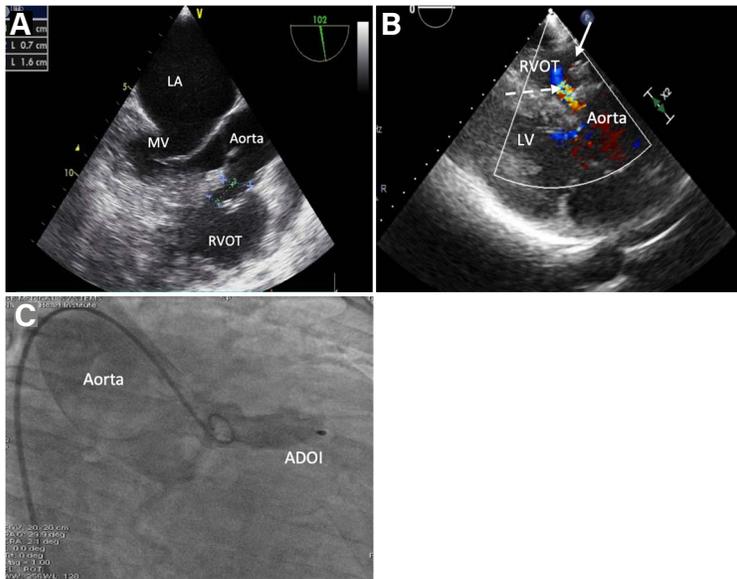
The ADOI was successfully deployed in all cases, except in Case 3, in which a trial to close the defect at the inlet was attempted. The device was slipped into the mid-portion of the defect, yet the disc was large enough to be stable at the middle of the defect, and the defect was successfully closed by the device in the middle of the aneurysm with mild residual flow.

## Complications

Residual shunt after deployment was encountered in two cases (Cases 3 and 5). No patient had procedure related AR. No other complications were reported (no TR, RVOT, obstruction, or device embolization).



**FIGURE 4** Aortography of Coarctation of aorta (Co.A) and Ruptured sinus of Valsalva (RSOV). **A)** Aortography shows severe Co.A (white arrow). **B)** Deployment of stent to dilate the Co.A. **C)** Opening of RSOV into right atrium (RA). **D)** After deployment of ADO1 device.



**FIGURE 5** TEE, TTE and Aortography before and after RSOV closure of **Case 3**. **A)** TEE Mid-esophageal long axis view at 102° shows dimensions of aneurysm of right SOV that opens into Right ventricular outflow tract (RVOT). **B)** TTE of modified Parasternal long axis (PLAX) view showing ADO1 device (white arrow) in position with mild residual flow across the (RSOV) into RVOT (dotted arrow). **C)** Aortography after deployment of ADO1 device.

**Follow-Up**

All patients were followed for six to 10 years. At time of the last follow-up, all patients remained nearly asymptomatic or in NYHA Class I. Echocardiography showed that residual shunt disappeared in patient 5. However, after four years, **Case 2** (patient with bicuspid aortic valve and moderate AR at baseline) exhibited worsening dyspnea again. TTE showed severe eccentric AR at that time, so patient was referred for aortic valve replacement, which was successful.

No late postoperative deaths, infective endocarditis, hemolysis, thrombus formation, device malposition, or significant arrhythmias were reported.

**Discussion**

A ruptured sinus of Valsalva aneurysm is a critical clinical entity that almost always ruptures into the right side of the heart causing a left-to-right shunt with overwhelming hemodynamic consequences. Patients either present acutely (such as with cardiogenic shock) or with chronic heart failure. As we are in the era of TCC for other congenital left-to-right shunts, it is expected that this rare anomaly is also flexible with respect to TCC. To our knowledge, the study of the present case scenarios is the only one of patients with this rare defect undergoing TCC with long-term follow-up (10 years).

A study by Kerkar et al.<sup>9</sup> was conducted with 20 patients with RSOVA between 2004 and 2009 with documentation of only immediate- and mid-term follow-up results (median period 24 months).

**Different Case Scenarios**

**Case 1:** A thirty-six old male presented with deteriorating heart failure symptoms. He was diagnosed provisionally as idiopathic dilated cardiomyopathy. He had a rupture of the right SOVA into the RA that was closed with an ADO1 device at the inlet of the defect (**Figure 1**). The patient’s symptomatology dramatically improved after a short period, and LV dimensions and systolic function improved after three years. Long-term follow-up (10 years until present) reports that the patient has a healthy life.

**Case 2:** A twelve-year-old cachectic (weight 22 kg) boy presented with severe heart failure symptoms (NYHA Class IV) and severely elevated arterial blood pressure. He was found to have severe CoA with bicuspid aortic valve and grade III/IV aortic regurgitation; in addition, a ruptured right SOVA in the RA was detected with LV dilatation and depressed systolic function. At first, stenting of the CoA was performed and then closure of the defect was successfully done via an ADO1 device from the inlet of the defect (**Figure 4**). His symptoms improved markedly; however, echocardiography revealed a mild decline in LV dimensions and mild improvement in LV function.

Despite an improvement of general condition and normal growth after four years, he had shortness of breath and NYHA class III/IV. Echocardiography revealed severe AR that necessitated cardiac catheterization for further evaluation. An aortogram showed severe AR and no residual shunt through the device. The progressive AR was attributed to the natural history of bicuspid aortic valve, the patient was sent for aortic valve replacement. Which was successful.

**Case 3:** A fifty-year-old female had worsening heart failure with a large aneurysm of the right coronary cusp that ruptured into the RVOT with moderately dilated LV dimensions and good contractility. She also was found to have bicuspid aortic valve. The defect was successfully closed with the ADO1 device in the middle of the aneurysm with mild residual flow (**Figure 5**) with marked symptom improvement.

**Case 4:** A 32-year-old male presented with ongoing dyspnea and orthopnea, deteriorating general condition, pulmonary congestion,



## Pediatric Cardiac Intensivist Faculty Positions

The Divisions of Pediatric Critical Care and Pediatric Cardiology in the Department of Pediatrics at Washington University School of Medicine seek applicants for faculty positions in the Cardiac Intensive Care Unit (CICU) at Saint Louis Children's Hospital (SLCH). The positions include an appointment at appropriate rank in the Washington University School of Medicine on the Clinician Educator faculty track. The ideal candidate should be board certified/eligible (BC/BE) in Pediatric Critical Care Medicine, Pediatric Cardiology, or Pediatric Anesthesia and have completed advanced training in pediatric cardiovascular intensive care (either via dual fellowships in pediatric cardiology and critical care medicine or via completion of an advanced cardiovascular intensive care training). The successful candidate will serve as an attending cardiac intensivist along with an extremely collaborative group of highly qualified dedicated Cardiac Critical Care faculty comprised of dually BC/BE Critical Care and Cardiology faculty, Critical Care faculty with advanced Cardiac Critical Care training, and senior Critical Care faculty with ample Cardiac Critical Care background. Participation in house-staff and fellow education, as well as clinical, translational or laboratory-based research will be expected. Extensive opportunities exist for scholarly collaborations with investigators at the School of Medicine and other departments throughout Washington University.

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Our CICU is an integral part of an acuity adjustable 38 beds Heart Center. Each patient room has been designed to accommodate either a critically ill or step-down patient. Hence, the number of critically ill patients is flexible and can increase up to 20 patients if necessary. The CICU is a state-of-the-art dedicated unit that provides all forms of cutting-edge cardiovascular intensive care for children and adults with congenital and acquired cardiovascular diseases. The CICU is staffed by 2 Cardiac Critical Care faculty during the day, and a dedicated in-house Cardiac Critical Care faculty at night.

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**Interested candidates should send a letter of intent and curriculum vitae to:**

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## Pediatric Electrophysiologist Faculty Position

The Department of Pediatrics of Washington University School of Medicine in St. Louis, is seeking a pediatric cardiologist to join our team of 19 clinicians and basic scientists, based at St. Louis Children's Hospital.

We seek to recruit a Pediatric Electrophysiologist to join two full time faculty members (Drs. Jennifer N. Avari Silva [Director, EP] and George F Van Hare) in our arrhythmia program. The program offers a full Electrophysiology Service, including interventional, inpatient and outpatient services. The interventional electrophysiology laboratory is a biplane cineangiography laboratory outfitted with an Ensite electroanatomic mapping system. The program has continued to grow and is currently on track to perform approximately 140 ablations, 40 pacemakers/ICDs, and 40 implantable loop recorders per year. We also offer a multidisciplinary inherited arrhythmia clinic, and Washington University offers a clinical genomic program, which we access for genotyping. Opportunities for basic, translational and clinical research are robust with close connections with researchers in the Department of Biomedical Engineering, the Department of Cell Biology and Physiology, and the Department of Developmental Biology at Washington University.

The ideal candidate must be eligible for licensure in Missouri, be board certified (or eligible) in pediatric cardiology, and have had advanced training in pediatric cardiac electrophysiology. Candidates will be considered at the Assistant or Associate Professor level.

Washington University School of Medicine is consistently ranked as one of the best medical schools in the country, and is a longstanding leader in funding for pediatric research. St. Louis Children's Hospital is a 400 bed free-standing children's hospital established in 1879. The St. Louis Children's and Washington University Heart Center includes an active surgical program, a 16-bed Cardiac Intensive Care Unit, and one of the nation's largest pediatric heart failure and heart transplantation programs.

**Interested candidates should provide a curriculum vitae and contact:**

**Janet Scheel, MD, FACC, Interim Co-Director, Pediatric Cardiology**

1 Children's Place, St. Louis, MO 63110

E. [JScheel@wustl.edu](mailto:JScheel@wustl.edu), P. 314.454.4561

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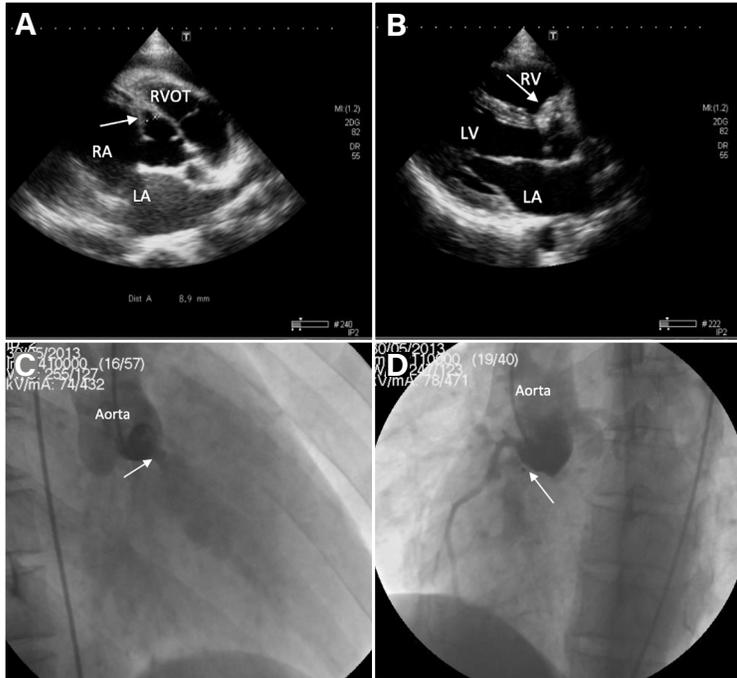


and generalized anasarca despite a previous TCC with an Occlutech® PDA occluder at another center for a ruptured sinus of Valsalva. There was a large defect between the aorta and RVOT next to the occluder device. The decision was to close the inlet of Valsalva aneurysm with an ADO1 device. The procedure was successful without any residual flow and no obstruction to the RVOT (**Figure 4**). The patient's condition improved notably soon after the procedure.

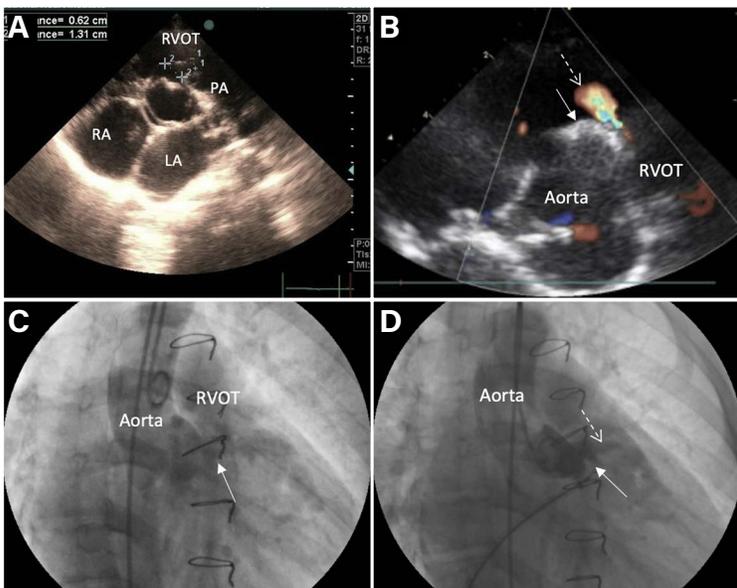
**Case 5:** A 17-year-old female, who had a history of perimembranous outlet VSD that was surgically closed three years ago presented with worsening heart failure symptoms. She had right coronary sinus dilatation and rupture into the RVOT (intraconal/subpulmonic). An ADO1 device was used to successfully close the RSOV (**Figure 7**). The device was deployed at the exit end of the aneurysm because the aortic end was too large and prevented use of a properly-sized device, which may have caused impingement on the aortic valve.

**Case 6:** A 31-year-old male presented with acute cardiogenic shock (wet and cold). A Cocoon duct occluder was used to close the RSOV (**Figure 3**). Post-occlusion aortography and echocardiography showed proper positioning of the device with good functioning of the aortic valve and patency of coronary arteries. Dramatic improvement after device deployment within 24 hours after cessation of inotropes and mechanical ventilation was seen.

**Cases 7-10:** Four classic cases of ruptured sinus of Valsalva were presented with worsening heart failure (NYHA class III) and right sinus of Valsalva aneurysm that had ruptured into the RV. ADO1 devices were successfully used for closure of the defects with marked improvement in symptoms (**Figure 6**).



**FIGURE 6** TTE and Aortography before and after RSOV closure of cases 7-10. **A)** TTE short axis shows large defect through the right SOV into RV (white arrow). **B)** ADO I device is seen closing the RSOV (white arrow). **C)** Aortography shows RSOV opens into RV (white arrow). **D)** After device closure with no residual shunt (white arrow).



**FIGURE 7** TTE and Aortography before and after RSOV closure of Case 5. **A)** TTE short axis view showing RSOV into RVOT. **B)** Color flow doppler on short axis view showing mild residual flow after deployment of device to close RSOV. **C)** Aortography shows large defect between the aorta and RVOT (white arrow). **D)** Post ADO I device deployment showing mild residual shunt (dotted arrow).

## Technical Aspects

The technique was similar to TCC of perimembranous VSD; however, we did not use balloon sizing of the defect since we were able to size the defect well using TOE. Our plan was to close the ruptured SOVA at the aortic end since closure at the rupture site (outlet) would leave behind an aneurysmal sac exposed to arterial pressure. In one patient (**Case 5**), we preferred to close the defect at the exit site, as previously mentioned, aiming to avoid use of a larger device that may affect the aortic valve. In another patient (**Case 3**), the device was slipped into middle of aneurysm but was stable with mild residual flow and no recurrence on follow-up. This event was similar to what happened to three patients in a study by Kerkar et al.,<sup>9</sup> in which the aortic disks were tilted slightly into the aneurysms with no recurrence at mid-term follow-ups.

## Complications

### Failure to Deploy

Unlike Chang et al.,<sup>10</sup> who were compelled to use a Gianturco coil instead of an ADO to overcome tortuosity of the ruptured SOVA tract, we did not face technical difficulties in introducing the delivery system or the ADO1 in any of our patients due to suitability of the device configuration to the defect shape and availability of large sizes.

On the other hand, Kerkar et al.<sup>9</sup> mentioned that two cases exhibited difficulties related to the large size of the defect and the occurrence of significant AR-related procedure before release due to encroachment of the aortic disk onto the ADO located on the aortic leaflet. We expected the same problem in patient 5, and we closed the defect at the exit point with successful avoidance of aortic valve encroachment.

### Residual Shunt

Residual shunt was seen in two cases out of ten at time of discharge compared with five cases out of 18 in study held by Kerkar et al.<sup>9</sup> In our



study, residual shunt was reported in two patients, including patient 5, in whom the residual shunt was small and had disappeared upon follow-up. Also, in **Case 3**, the residual shunt was mild and persistent, yet the symptoms were markedly improved.

## Conclusion

Although the number of cases is small, TCC of a ruptured sinus of Valsalva seems to be a feasible and effective alternative treatment modality. It could be a live-saving procedure for patients with cardiogenic shock due to acute RSOV.

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# AngioAtlas – An Angiographic Library of Congenital Heart Disease

Renelle S. George, MD & Martin Bocks, MD

Early in her fellowship training, then pediatric cardiology fellow Renelle George, MD, identified a need for a resource in which trainees could learn congenital angiography. As a first-year fellow, she had little exposure to angiograms and searched for examples while she prepared for cases during her cath rotation. "I mostly found low-quality, still images in books," she reports. What she found most useful, however, was looking at angiograms from previous cases. "I realized that we actually had a wonderful repository of angiograms to learn from, only that it required some work to get to," she remarks. Dr. George then approached director of pediatric interventional cardiology, Martin Bocks, MD, and together they launched AngioAtlas, an angiographic library of Congenital Heart Disease. "We are extremely pleased with how AngioAtlas has turned out," Dr. Bocks states. "It really seems to have filled a niche in our congenital community, improving training and providing a reliable platform to profile interesting angiography during interventional cases."

AngioAtlas is available online at [angioatlas.org](http://angioatlas.org) and contains angiograms depicting normal anatomy, congenital heart defects, post-surgical anatomy, and interventions. The site was launched in September 2019 and content is being added on a rolling basis. A new section featuring interesting cases was added in May 2020, with the goal to promote information-sharing among the interventional community and to showcase less frequently seen diagnoses and interventions. "Fantastic things are happening in cath labs around the world every day, and AngioAtlas offers a unique opportunity to tell everyone about them," Dr. Bocks remarks. Persons interested in submitting a case should visit [angioatlas.org/submit-case](http://angioatlas.org/submit-case) and refer to the submission guidelines for additional information, <https://angioatlas.org/defects/cases/submission-guidelines/>.

"It's been a lot of work, but a lot of fun as well," Dr. George adds. "AngioAtlas has grown from a little idea to help trainees prepare for their cath rotations to a resource that's used by nearly 1,000 users across the world. We've received a great deal of positive feedback from trainees, cath lab staff, and interventionalists, who have also been using the site for parental education. It's been really exciting to watch the project take on a life of its own and expand beyond the goals we originally intended. We've also partnered with several organizations who share our passion for education and our mission to advance the education of learners of congenital cardiology everywhere. We are incredibly grateful to everyone who has supported us on this journey, particularly our content contributors and reviewers. We look forward to continuing to develop the site into a valuable resource for learners and interventionalists alike."

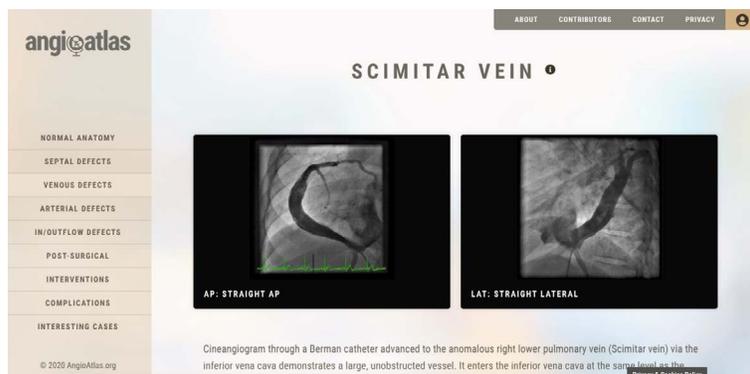
Visit [angioatlas.org](http://angioatlas.org) for free access to all content (registration required).

To contact Dr. George and Dr. Bocks directly, email [info@angioatlas.org](mailto:info@angioatlas.org).



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Sample defect page showing AP and lateral angiograms of a Scimitar Vein.



## MARTIN BOCKS, MD

*Director, Pediatric Interventional Cardiology*  
UH Rainbow Babies & Children's Hospital  
Case Western Reserve University  
School of Medicine  
Cleveland, OH, USA



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- These benefits are for full time employees, employees in other types of employment classifications may be eligible for some of these benefits.

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# Midwest Fetal Care Center Publishes Novel Case Study of Fetal Unguarded Mitral Valve Orifice

Clinicians from the Midwest Fetal Care Center, a collaboration between Allina Health and Children’s Minnesota, recently published a first-of-its-kind case study of a baby with an unguarded mitral valve orifice, an extremely rare disease with only seven described cases in the literature. The article in the *Journals of the American College of Cardiology* highlights the team’s experience and learnings with the diagnosis and treatment of this rare cardiac anomaly, <https://www.jacc.org/doi/pdf/10.1016/j.jaccas.2020.11.020>.

The unborn baby was identified as having congenital absence of the mitral valve, severe enlargement of the left heart chambers, marked underdevelopment of the aorta and a persistent heart arrhythmia. The baby required antiarrhythmic medications during pregnancy and was delivered via Cesarean section at 37 weeks when he became clinically unstable. After delivery, the baby was immediately intubated, started on intravenous prostaglandin and antiarrhythmic medications, and transferred to the cardiovascular intensive care unit. Within 12 hours of delivery, the neonate developed hemodynamic instability and he was electively taken to the cardiovascular operating room and placed on central venoarterial extracorporeal membrane oxygenation (ECMO) and was listed for heart transplant. Once on ECMO, baby was successfully converted to a normal heart rhythm and he maintained clinical stability until Day of Life 26 when he had an acute episode of bradycardia, hypotension, and eventual asystole. The family elected to withdraw support and consented to an autopsy.

This is the first known case of this combination of heart findings, and required care by a multidisciplinary team across the perinatal spectrum, including experts in: fetal cardiology, electrophysiology, heart failure/transplant, pediatric cardiac surgery, perinatology and neonatology.

## More About the Midwest Fetal Care Center

The Midwest Fetal Care Center (MWFCC), a collaboration between Allina Health and Children’s Minnesota, brings together a multi-disciplinary team of highly trained maternal-fetal medicine experts from Allina Health and pediatric and neonatal specialists from Children’s Minnesota. Open since 2008, the MWFCC is a national referral center and regional leader in fetal diagnosis, fetal intervention and comprehensive fetal care for unborn babies with complex conditions.

[https://www.childrensmn.org/services/care-specialties-departments/fetal-medicine/?gclid=EAlaIQobChMI4lvx9Nbi7gIVZFVCh2aOARuEAAYASAAEgL9xvD\\_BwE](https://www.childrensmn.org/services/care-specialties-departments/fetal-medicine/?gclid=EAlaIQobChMI4lvx9Nbi7gIVZFVCh2aOARuEAAYASAAEgL9xvD_BwE)



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## Echocardiography Faculty Opportunity

The Heart Institute at Cincinnati Children's invites Pediatric Cardiologist at the Assistant, Associate, or full Professor level with an interest in echocardiography and a strong track record in echocardiography research to advance the academic output and reputation of the echo lab at Cincinnati Children's.

The Heart Institute and the Advanced Imaging Service pride themselves on excellent clinical outcomes, while maintaining a strict academic focus, research productivity and exemplary professionalism. The acceptable candidate would be expected to maintain similar high standards of clinical service.

The Echo lab includes 12 imaging faculty and 22 cardiac sonographers and performs approximately 15,000 transthoracic and 500 transesophageal echocardiograms annually. The facility includes a state-of-the-art reading room as well as the necessary technology to perform all current advanced imaging techniques. Additionally, the Cardiovascular Imaging Core Research Lab provides dedicated personnel and resources for human and animal echo research.

The Heart Institute is an internationally recognized academic center of excellence for Pediatric (congenital and acquired) including Adult Congenital Cardiac Care, and clinical and basic science research. The Heart Institute incorporates the Divisions of Congenital Heart Disease, Cardiothoracic Surgery and Molecular Cardiovascular Biology to offer the full range of Pediatric Cardiac services within a free-standing not-for-profit tertiary care medical center. The Heart Institute also trains categorical Pediatric Cardiology and sub-specialty fellows in all areas of congenital heart disease practice (including 2 Advanced Imaging fellows).

### Required

M.D., D.O., or equivalent degree  
Current active medical license issued by the State of Ohio or eligible for license.  
Appropriate medical credentialing through the Medical Staff Services offices  
Completion of all required pre-employment activities  
Current Assistant or Associate Professor appointment or eligibility required.

### Preferred

Board certification Pediatric Cardiologist

### Expectations

The applicant would be expected to participate in clinical service including (but not limited to):  
Perform/interpret transthoracic and transesophageal echocardiograms.  
Perform a single out-patient clinic on a weekly basis.  
Provide limited periods of in-patient and/or consult service coverage.  
Participate in all Heart Institute clinical and management conferences.  
Perform teaching and instruction commensurate with the training mission of the Heart Institute at Cincinnati Children's  
Develop and lead new research programs within the echo lab with significant dedicated time protected for research activities.

Interested candidates should address all inquiries to:

### Andrew Redington, MD

Co-Director, The Heart Institute  
Cincinnati Children's Hospital Medical

Email a letter of intent and CV c/o [Deborah.Mancini@CCHMC.org](mailto:Deborah.Mancini@CCHMC.org)



# FDA Approves Labeling Update for Abbott's HeartMate 3™ Heart Pump for Use in Pediatric Patients

*Abbott's HeartMate 3™ Heart Pump Approved for Use for Pediatric Patients Battling Advanced Heart Failure*

*This Life-Saving Technology Provides New Treatment Option for Underserved Population*

PRNewswire -- Abbott (NYSE: ABT) announced the U.S. Food and Drug Administration (FDA) has approved updated labeling for the company's HeartMate 3™ heart pump to be used in pediatric patients with advanced refractory left ventricular heart failure. With the updated labeling, physicians now have additional options for treating this underserved population awaiting a heart transplant or for those not eligible to receive a transplant as a result of potential complications or risk related to the procedure.

The approval follows similar pediatric innovations for Abbott in recent years, including the Masters HP™, a 15mm pediatric heart valve the size of a dime, approved in 2018; and the Amplatzer Piccolo™ Occluder, a pea-sized plug approved in 2019 to help treat a potentially life-threatening opening in the heart of some premature or newborn babies.



Many children and adolescents with congestive heart failure require a heart transplant or mechanical device implant to survive. The HeartMate 3 left ventricular assist device (LVAD) – or heart pump – is an implantable device that pumps blood through the body in people whose heart is too weak to do so on its own. The HeartMate 3 pump was initially approved in the United States in 2017 for adults awaiting a heart transplant and received FDA approval for long-term use in adults in 2018. In the largest LVAD trial in the world, the HeartMate 3 pump showed a survival rate of 79% at two years – an outcome comparable to patients receiving a heart transplant.

"For families with children battling chronic diseases the future is often bleak. As physicians, we see the fear in the eyes of not only the child, but also the mothers and fathers," said Robert L. Kormos, MD, divisional vice president, global medical affairs, Abbott's heart failure business. "Imagine a child with a heart condition that does not allow them to play with friends, sing or run. Innovations, such as the HeartMate 3, can lessen the crippling effects of heart failure and allow that child to live a more normal life."

## Life-Changing Collaboration in Action

The updated labeling for HeartMate 3 to be used in pediatric patients was supported by clinical data from the Advanced Cardiac Therapies Improving Outcomes Network (ACTION Learning Collaborative), a consortium of 50+ U.S. pediatric hospitals that pooled together data to show advantageous outcomes of the HeartMate 3 in pediatric patients.

"Our mission is to improve the outcomes of children with heart failure. Historically, this has been an underfunded and understudied area in

pediatrics," said Angela Lorts, MD, MBA, and David Rosenthal, MD, co-founders of ACTION Learning Collaborative. "This technology will benefit our pediatric patients and is a leap forward for improving heart failure outcomes in children. We are honored to collaborate with Abbott on this pediatric initiative."

## Back to Living Life on Her Terms

Katrina Sellens, now 16-years-old, was an active teenager before being diagnosed with cardiomyopathy, a serious disease of the heart muscle that can lead to heart failure. She couldn't walk 10 feet without feeling exhausted and had to bend over just to breathe. In 2019, Katrina received the Abbott HeartMate 3 under a special request to treat her life-threatening disease.

"One of the most depressing aspects of heart failure is seeing your child lose the ability to do what had always come naturally," said Maria Bautista, Katrina's mother. "I didn't believe she would get as strong as she has with the HeartMate 3. We are back to living life on her terms."

Nearly two years later, Katrina is back to camping and taking care of her family's chickens. The high school sophomore recently earned her driver's permit and dreams of becoming an LVAD nurse so she can help others with heart failure.

## Resources to Learn More

Heart failure is a manageable condition, especially if it is detected early. Cardiologists and other healthcare providers can visit Abbott Cardiovascular for more resources on the latest innovations and solutions designed for patients with heart failure.

## About Abbott's HeartMate 3 Heart Pump

Abbott's HeartMate 3™ heart pump is a small, implantable mechanical circulatory support device for advanced heart failure patients who are awaiting transplantation or are not candidates for heart transplantation. It is the first commercially approved (CE Mark and FDA approved) heart pump with Full MagLev™ technology, which allows the device's rotor to be "suspended" by magnetic forces. This design aims to reduce trauma to blood passing through the pump and improve outcomes for patients.

For U.S. important safety information for the HeartMate 3, visit <https://www.cardiovascular.abbott/us/en/campaigns/heartmate-3-lvad-pediatric.html>.

For U.S. important safety information for the Masters HP Series, visit <http://abbo.tt/2taeyVL>.

For U.S. important safety information for the Amplatzer Piccolo Occluder, visit <https://www.structuralheartsolutions.com/us/piccolo-ISI>.





# Associate or Full Professor (Ladder, In-Residence, Clin X) Division Chief, Pediatric Cardiology University of California San Diego

The University of California, San Diego (<http://pediatrics.ucsd.edu>) is committed to academic excellence and diversity within the faculty, staff, and student body. The Division of Cardiology within the Department of Pediatrics at UC San Diego and Rady Children's Hospital (<https://www.rchsd.org/programs-services/heart-institute/>) is currently seeking to fill the position of Division Chief of Pediatric Cardiology at UC San Diego and Rady Children's Hospital.

UCSD is one of the leading biomedical research universities in the world with more than 80 faculty members in the National Academy of Sciences. Federal research expenditures are in the top 5 in the country and top 3 in NIH funding per faculty member. Co-located on the La Jolla Mesa with The Salk Institute, Scripps Research Institute, Sanford-Burnham-Prebys Medical Research Institute, La Jolla Institute of Immunology, the J. Craig Venter Institute and the Sanford Stem Cell/Regenerative Medicine Consortium, UCSD provides an outstanding collaborative research environment that is home to over 500 biotechnology and pharmaceutical companies. Major initiatives on campus include UCSD Altman Clinical and Translational Research Institute, Genomic Medicine, Microbiome Sciences, Diabetes and Metabolism, Big Data Sciences and Drug Discovery.

This 524-bed non-profit pediatric care facility dedicated to excellence in care, research, and teaching serves as a major regional tertiary care hospital for children. The Division of Pediatric Cardiology consists of thirty faculty with depth in the full range of cardiac subspecialties and two experienced cardiac surgeons who are board certified in pediatric cardiovascular surgery. There is a CVICU with 24/7 staffing by 7 cardiac intensivists and a cardiac inpatient unit totaling 32 beds. Annual program volumes include about 500 cardiac surgeries, over 600 cardiac catheterizations, more than 18,000 echocardiograms and outpatient visits. The Division has excelled in every aspect of its clinical mission, and strives to provide culturally competent care for children and families.

The Division Chief will oversee the management of the Division of Cardiology to include direct oversight of faculty, staff and will have direct accountability for the Division's clinical service and operational performance.

The Department is interested in candidates who have a demonstrated passion for providing outstanding care, a desire to improve systems to maximize efficiency, leadership abilities to engage physicians and hold them accountable, promote an environment that supports diversity and inclusivity among faculty, students and staff, and an enduring commitment to developing a wide range of educational opportunities.

Candidates must have an MD or equivalent and be board certified or eligible by the American Board of Pediatrics in the subspecialty of Cardiology. Candidates must have or be eligible for a California medical license or equivalent certification/permit as determined by the Medical Board of California.

Candidates must have a national reputation as a leader in academic basic, translational or clinical research, as demonstrated by high quality publication record, excellent trainee outcomes, and sustained competitive extramural funding.

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# Promising Tissue Valve Tech Blocks Calcium Buildup

## Zero Structural Valve Deterioration at Five Years in the COMMENCE Study

Nicole Lou, Staff Writer, MedPage

A tissue valve built for less structural valve deterioration showed favorable safety and hemodynamic performance at five years in surgical aortic valve replacement (SAVR), according to the COMMENCE investigators.

Surgical valves mounted with Resilia (a proprietary bovine pericardial tissue, from Edwards Lifesciences, formulated to reduce the calcification that often leads to valve dysfunction) were associated with "quite satisfactory" safety in 471 patients who had 30-day mortality and stroke rates of 1.2% and 1.6%, respectively, reported Joseph Bavaria, MD, of the Hospital of the University of Pennsylvania in Philadelphia.

By five years, patients showed 89.2% freedom from mortality and 94.5% freedom from stroke on top of zero cases of valve thrombosis or structural valve deterioration, Bavaria told the audience at this year's *Society of Thoracic Surgeons (STS)* virtual meeting, <https://www.medpagetoday.com/meetingcoverage/sts>.

The latter is significant because contemporary bioprosthetic aortic valves often begin to show structural valve deterioration around 5 years after SAVR. Bavaria estimated structural valve deterioration rates by then to be about 1.5% with the Magna device and 5% with the Trifecta.

"There continues to be a significant focus placed on tissue valve durability given the increase in life expectancy and lifestyle implications for more active patients who historically would receive mechanical valves," said Bavaria in a press release. "The

latest data from the COMMENCE study are encouraging and speak to the promise of Resilia tissue as a significant advancement in technology for patients with valve disease."

The first case of structural valve deterioration occurred at 5.2 years in the study and resulted in the patient getting a transcatheter valve-in-valve reintervention.

Additionally, the good hemodynamic performance of the Resilia valves was reflected in the "absolutely stable" echocardiography-derived mean gradients of 10.2 mm Hg at one year and 11.5 mm Hg at five years, as well as little change in average effective orifice area, from 1.7 cm<sup>2</sup> to 1.6 cm<sup>2</sup>, according to Bavaria.

Transvalvular regurgitation reached only 3.7% of patients at the mild level; there were no moderate cases observed.

"It is heartening to know that the 5-year trial data is incredibly satisfactory," Bavaria told the STS audience.

Given that valve deterioration is a concern with bioprosthetic devices, both transcatheter and surgical alike, session co-moderator Gilbert Tang, MD, MBA, of Mount Sinai Health System in New York City, asked if the manufacturer would start putting Resilia tissue on its transcatheter aortic valves.

Bavaria's guess was "probably."

COMMENCE was a single-arm FDA investigational device exemption trial that included 689 SAVR recipients at 27 clinical sites across the U.S. and Europe.

Investigators tested surgical valves with a Magna Ease design and the tissue of the Inspiris valve (albeit without the vFit feature that facilitates expansion for subsequent valve-in-valve procedures). The aldehyde capping of the Resilia tissue platform prevents calcification by blocking free aldehydes; glycerolization prevents further aldehyde exposure to the valve.

The present analysis was based on the 471 adults with sufficient five-year follow-up (mean age 66.9 years, 71.8% men). This group entered SAVR with an average STS risk of 2.0%. Roughly a quarter were classified as having New York Heart Association Class I symptoms, half Class II, and another quarter Class III.

People getting concomitant procedures represented about 40% of the cohort. The trial excluded emergency, endocarditis, and multi-valve procedures.

Session co-moderator Jessica Forcillo, MD, MSc, MPH, of Centre Hospitalier de l'Université de Montréal, commended the surgeons for choosing larger valve sizes, as 23- and 25-mm valves were the most commonly used in the study.

Bavaria acknowledged that there is insufficient data on people under age 50 in the study.

A subset of COMMENCE participants will be evaluated through 10 years, he said, adding that a small ongoing study, RESILIENCE, is assessing how calcium deposits on Resilia tissue predict long-term bioprosthetic valve durability.



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