Right ventricular outflow obstruction caused by cocoon duct occluder used for closure of ruptured sinus of valsalva aneurysm

Rashmi Soori, Aanchal Dixit, Prabhat Tewari, Surendra K. Agarwal

Department of Anaesthesiology, K.S. Hegde Medical Academy, Mangalore, Karnataka, Departments of Anaesthesiology and Cardiovascular and Thoracic Surgery, SGPGIMS, Lucknow, Uttar Pradesh, India

ABSTRACT
Hemolytic anemia and right ventricular outflow tract obstruction following device closure of ruptured sinus of Valsalva have seldom been reported in isolated case reports, and exact incidence is not known. A gentleman presented with severe delayed hemolytic anemia following the use of cocoon duct occluder for ruptured sinus of Valsalva. Right ventricular outflow tract obstruction of unclear etiology was also reported on transthoracic echocardiography, necessitating retrieval of the device and surgical closure of the defect. Intraoperative transesophageal echocardiography (TEE) showed right ventricular outflow obstruction by the cocoon device itself with a normal pulmonary valve. In this report, we emphasize that improper device selection for closure of ruptured sinus of Valsalva aneurysm, may lead to delayed leaks across the device, which can gradually progress causing hemolytic anemia and high gradient across the right ventricular outflow tract. Intraoperative TEE helped to delineate the cause of right ventricular outflow tract obstruction.

Keywords: Anemia, cocoon, hemolytic, occluder, ruptured, sinus, Valsalva

Case Report

INTRODUCTION
Hemolytic anemia following prosthetic valve replacement has been studied extensively. However, its occurrence after device closure of ruptured sinus of Valsalva has been seldom reported in isolated case reports, and exact incidence is not known. We report a case of severe delayed hemolytic anemia following the use of a cocoon duct occluder (CDO) for ruptured sinus of Valsalva aneurysm (SOVA) with right ventricular outflow tract (RVOT) obstruction of unclear etiology on transthoracic echocardiography (TTE). It necessitated retrieval of the device and surgical closure of the defect. Intraoperative transesophageal echocardiography (TEE) showed RVOT obstruction by the cocoon device itself with a normal pulmonary valve.

CASE REPORT
An 18-year-old male with dyspnea on exertion underwent TTE and was diagnosed with ruptured SOVA with a defect of 7 mm that opened into the RVOT with a normally functioning aortic valve (AV). He underwent device closure with a CDO size 18/16 mm (Vascular Innovations Co., Ltd, Nonthaburi, Thailand) by retrograde approach, and the device was deployed at the aortic end of the defect. Immediate TTE revealed the device in situ with no residual obstruction.
leak. However, follow-up TTE after 3 months revealed a mild leak across the device, and the patient was kept in rigorous follow-up.

One year after device placement, the patient presented with dyspnea and palpitation. Blood reports revealed hemoglobin - 6.8 gm/dL, hematocrit -26%, reticulocyte count of 6% with total bilirubin - 3 mg/dL, unconjugated bilirubin - 2.7 mg/dL, SGOT- 129 IU/mL, LDH – 2859 IU/mL. Urine microscopy revealed hemoglobinuria and albuminuria. TTE demonstrated a 4 mm left-to-right shunt across the closure device with a peak systolic gradient of 72 mmHg. In addition, TTE revealed moderate aortic regurgitation (AR) as well as flow acceleration in the RVOT on color flow and spectral Doppler, (mid-systolic peak gradient 58 mmHg) of unclear etiology. The patient was diagnosed with hemolytic anemia with the probable cause being residual shunt across the device. The patient was taken up for surgical retrieval of the device, closure of ruptured SOVA with AV replacement, relook and proceed for RVOT obstruction on an urgent basis. A written consent was taken from the patient for publishing his case as a report for furthering the knowledge.

In operation theatre, after standard anesthetic management, comprehensive TEE with real-time 3D enabled probe (X7-2t with ie33, Philips Ultrasound, Bothell WA, 98041 USA) was done. It showed ruptured SOVA arising from the right coronary sinus opening into the RVOT with residual shunt across the device in situ [Figure 1, Loop 1]. The pulmonary valve appeared normal in morphology, and the cause for RVOT obstruction was found to be the cocoon device itself [Figure 2, Loop 2]. Two units of packed red blood cells and two units of fresh frozen plasma were used for priming the cardiopulmonary bypass (CPB) circuit. After heparinization, CPB institution, and cardioplegic heart arrest, the device was retrieved by aortotomy. The minimum hematocrit on CPB was 20. The ruptured SOVA was closed with a Dacron patch, and native AV was replaced with a mechanical prosthesis (ATS size - 24). The immediate postoperative TEE on color flow Doppler revealed reduced turbulence in the RVOT [Figure 3], and the measured mid-systolic peak gradient was 12 mmHg. Follow-up hemogram showed Hb of 8.6% after 1 week of surgery and improvement in liver function test. The postoperative course was uneventful, and follow-up TTE at 1 year revealed no residual shunt, normal prosthetic AV function, and normal flow in the RVOT.

**DISCUSSION**

The potential complications of transcatheter closure of ruptured SOVA include failure to deploy the device, residual shunting, coronary ostial encroachment, procedure-related aortic regurgitation (AR), hemolysis, and RVOT obstruction.[1-3] Our patient reported severe anemia caused by hemolysis. The cause of hemolysis after placement of any engineered cardiovascular device is often residual leak across the device that leads to turbulence.[4] Our patient started with no leak across the device immediately after deployment, but it developed and progressed with time in contrast to other case reports where there were immediate leaks that receded with time.[1] The TEE views in the intraoperative period that can help in the 2D evaluation

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Figure 1: RVOT: right ventricular outflow, LVOT: left ventricular outflow. Left panel: TEE Real-time 3D image: Midesophageal (ME) aortic valve long-axis view showing CDO in situ marked with an arrow. Right panel: TEE image: ME aortic valve long-axis view showing the device in situ with color flow depicting residual shunt (in blue color) marked with an arrow.

Figure 2: LV: left ventricle, LA: left atrium. Left panel: TEE Real-time 3D image: Midesophageal (ME) aortic valve long-axis view showing CDO in situ marked with an arrow. Right panel: TEE image: ME aortic valve long-axis view showing the device in situ with color flow depicting residual shunt (in blue color) marked with an arrow.

Figure 3: TEE image: Deep transgastric view post device retrieval, color doppler showing almost laminar flow in RVOT marked with a black arrow.
of ruptured SOVA with or without closure device in situ include mid-esophageal (ME) aortic valve short- and long-axis view, ME long-axis view, ME- 5 chamber view, ME RV inflow-outflow view.[9] For Doppler assessment of flow and gradient trans gastric (TG) views are better for flow alignment and include TG long axis, TG right ventricular (RV) basal, TG RV inflow-outflow, and deep TG views.[8] The shape of continuous-wave doppler of RVOT flow in our patient is parabolic depicting a fixed variety of obstruction [Figure 2, Right panel].

A CDO is made from nitinol wires coated with platinum and has the same profile as the Amplatzer duct occluder.[6] Increasing shunt across a CDO can be due to erosion around the device, usually related to abrasive mechanical force between device and human tissue caused by the material of which it is made. It can be further worsened by the motion of the device against the tissue as is also mentioned by Huang and colleagues.[7] Another reason reported for the development of residual leaks around devices is selecting a larger size of the occluder device than appropriate for age/weight.[8] Sinha et al.[3] published a case series for closure of ruptured SOVA where the size of CDO selected was such that its aortic segment was 2 to 4 mm larger than the defect size based on the visual clue. So for defects ranging from 6 to 10 mm, CDO size varied from 10/8 to 14/12 with no residual shunt in any of their patients.[2] Another study where 30 patients were treated with percutaneous closure of ruptured SOVA by various devices, reported using occluders that were 2–7 mm larger than the narrowest diameter of the ruptured SOVA.[9] In this context, CDO size 18/16 mm used for a defect of 7 mm in our patient was an extra-large device that moved throughout the cardiac cycle [Loop 2]. The large size of CDO may also be the reason that it projected into the RVOT and gave rise to a gradient across it. As the leak across the device increased, its motion increased, and it probably projected more into RVOT resulting in cardiologists finding a severe gradient in RVOT of unclear etiology.

We emphasize that improper device selection for closure of such ruptured SOVA, may lead to delayed leaks across the device which can progress gradually causing significant hemolytic anemia and high RVOT gradient. In this case, intraoperative TEE helped to delineate the cause of RVOT obstruction which was the device itself rather than any other anatomical lesion.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Kerkar PG, Lanjewar CP, Mishra N, Nyayadhish P, Mammen I. Transcatheter closure of ruptured sinus of Valsalva aneurysm using the Amplatzer duct occluder: Immediate results and mid-term follow-up. Eur Heart J 2010;31:2881-7.
2. Sinha SC, Sujatha V, Mahapatro AK. Percutaneous transcatheter closure of ruptured sinus of Valsalva aneurysm: Immediate result and long-term follow-up. Int J Angiol 2015;24:99-104.
3. Arora R, Trehan V, Rangaswaty UM, Mukhopadhyay S, Thakur AK, Kalra GS. Transcatheter closure of ruptured sinus of Valsalva aneurysm. J Interv Cardiol 2004;17:53-8.
4. Morshed KN, Bark D Jr, Forleo M, Dasi LP. Theory to predict shear stress on cells in turbulent blood flow. PLoS One 2014;9:e105357.
5. Hahn RT, Abraham T, Adams MS, Bruce CJ, Glas KE, Lang RM, et al. Guidelines for performing a comprehensive transthoracic echocardiographic examination: Recommendations from the American Society of Echocardiography and the Society of Cardiovascular Anesthesiologists. J Am Soc Echocardiogr 2013;26:921-64.
6. Park H, Song J, Kim ES, Huh J, Kang IS. Early experiences using cocoon occluders for closure of a ventricular septal defect. J Cardiovasc Imaging 2018;26:165-74.
7. Huang Y, Kong J, Venkatraman SS. Biomaterials and design in occlusion devices for cardiac defects: A review. Acta Biomater 2014;10:1088-101.
8. McElhinney DB, Quertermun MD, Kenny D, Alboliras E, Amin Z. Relative risk factors for cardiac erosion following transcatheter closure of atrial septal defects: A case-control study. Circulation 2016;133:1738-46.
9. Xiao JW, Wang QG, Zhang DZ, Cui CS, Han X, Zhang P, et al. Clinical outcomes of percutaneous or surgical closure of ruptured sinus of Valsalva aneurysm. Congenital Heart Dis 2018;13:305-10.