

Introduction

The sinus of Valsalva is located between the aortic valve annulus and the sinotubular ridge (*fig. A*). When pathologic dilation of the sinus of Valsalva occurs, this abnormality is termed sinus of Valsalva aneurysm (SOVA).¹ SOVA is estimated to occur in only 0.09% of the population and makes up 0.1% to 3.5% of all congenital heart defects. SOVA rupture is a feared complication that is associated with high morbidity and mortality. Presentation of ruptured SOVA may include acute heart failure, hemodynamic instability, and cardiac arrest.² This report highlights a case of a SOVA with right atrial fistula leading to heart failure and respiratory distress in a young man.

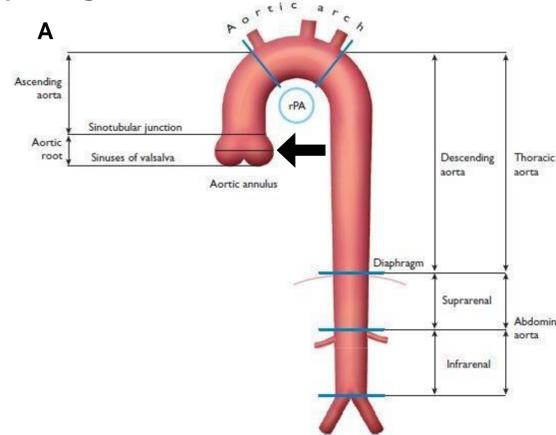


Fig. A. Aortic anatomy with sinus of Valsalva (arrow) (Image retrieved from: Anatomy of aorta, 2020).

Case Report

A 24-year-old male presented to the emergency department for nausea, vomiting, and shortness of breath. The patient had a medical history of cardiac murmur and cognitive developmental delay. On arrival, the patient was hypoxic with SpO₂ 73% and severe dyspnea. Rales and respiratory distress were noted on exam. Chest x-ray showed pulmonary edema (*fig. B*) and EKG showed sinus tachycardia with biatrial enlargement. The patient was placed on BiPAP for acute hypoxemic respiratory failure and started on intravenous diuretics. He was admitted to the intensive care unit for evaluation and monitoring.

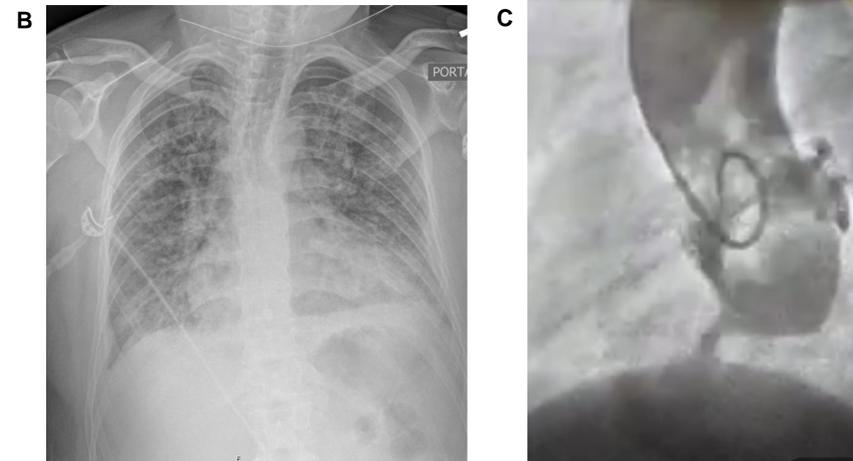


Fig. B. Initial chest x-ray showing consolidative, interstitial, and ground glass opacities with basilar predominance and small bilateral pleural effusions.

Fig. C. Cardiac catheterization with contrast injection into the aortic root demonstrating ruptured SOVA. The neck measured approximately 4.3 mm.

A transesophageal echocardiogram (TEE) showed left ventricular ejection fraction of 60%-65%, sinus of Valsalva to right atrial shunt (*fig. D*), and right ventricular systolic pressure of 60 mmHg. Agitated saline bubble study showed late appearance of bubbles on the left side. CT-angiography demonstrated ruptured SOVA with fistula to the right atrium (*fig. C*). The patient symptomatically improved with diuresis and supportive care but required transfer to a tertiary care center for percutaneous closure of the ruptured aneurysm using an Amplatzer occluder device. He was later discharged in stable condition.

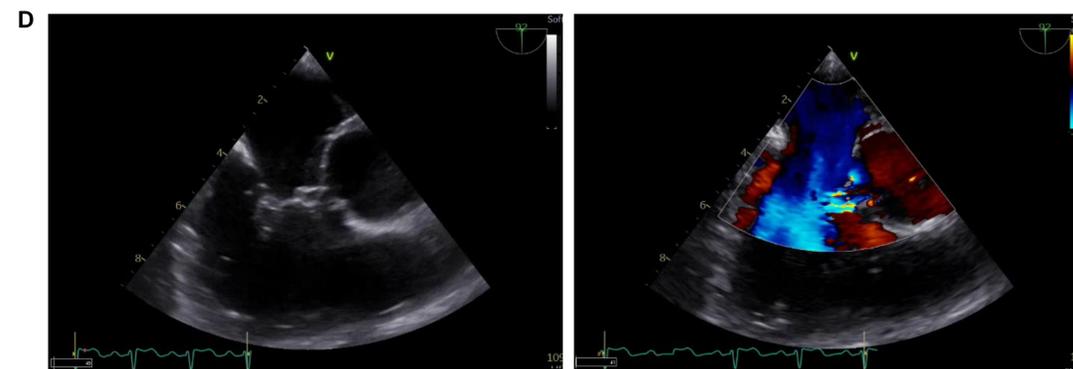


Fig. D. TEE demonstrating aneurysmal sinus of Valsalva (left) with blood flow from the aorta to the right atrium (right).

Discussion

SOVA is a rare cardiac condition, comprising a very small amount of all structural cardiac abnormalities. Patients may be asymptomatic until the SOVA is large or ruptures, such as in this case.²⁻³ This patient presented with acute heart failure and hemodynamic instability, a common presentation of this rare complication. Aneurysmal rupture is seen in only one-third of patients and can have serious consequences with estimated life expectancy of just one year.³⁻⁴ Due to high morbidity and mortality, SOVA should be recognized early and treated with urgent surgical referral.

TTE is considered the best initial diagnostic test with a sensitivity and specificity of 99% for identifying SOVA and 97% for SOVA rupture.⁷ Treatment involves surgical or percutaneous repair.⁵ Prognosis following surgical repair of SOVA rupture is excellent, with a 5 year survival rate of 83.1%.⁸ With this case report we hope to increase awareness of this rare condition and help aid in prompt recognition of such a complication.

References

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Acknowledgements

The authors would like to thank our faculty advisors for their mentorship and assistance with this poster. We would also like to thank the Kettering Health Dayton Internal Medicine Residency Program and Kettering Health Graduate Medical Education for providing us with funding and support to attend.