

## INTRODUCTION

- Sinus of Valsalva aneurysms (SVA) is a relatively uncommon cardiac condition characterized by an abnormal dilation of one or more of the three aortic sinuses.
- The prevalence of SVA has been estimated to be approximately 0.1% based on autopsy studies, and 0.2% in patients undergoing open cardiac surgeries, primarily in Western countries.
- Although an unruptured SVA may remain asymptomatic, a ruptured SVA can present in a serious cardiac decompensated state, as seen in the patient described in this report.
- Despite its rarity, a high degree of suspicion is crucial for an adequate diagnosis of SVA given its high mortality if left untreated.

## CASE HISTORY

- A 54-year-old male with a medical history of rheumatoid arthritis presented with non-radiating left chest pain for two weeks and worsening dyspnea, orthopnea and fatigue
- Vital signs showed a BP 134/61, HR 102, RR 18, and oxygen saturation of 98% on room air
- Cardio-pulmonary exam showed a continuous machinery murmur which was loudest in systole (5/6) and 3/6 in diastole.
- There was a palpable thrill across the precordium, diminished bilateral breath sounds, crackles in the lung bases, bilateral 2+ pitting edema, and raised jugular venous pressure
- CXR showed signs of pulmonary edema and bilateral pleural effusions
- Patient was preliminarily diagnosed with acute congestive heart failure and Lasix was given
- Transthoracic echocardiography revealed severe pulmonary hypertension and an echo density of 1.3 X 0.8 cm near the base of the septal leaflet of the tricuspid valve
- Further workup with transesophageal echocardiography revealed an outpouching from the right coronary sinus, suggestive of a ruptured right sinus of Valsalva (Figure 1)
- The ruptured SVA had created a jet hitting the tricuspid valve continuously during both diastole and systole (Figure 2)
- Cardiac catheterization with root injection showed angiographically normal coronary arteries and an aneurysmal fistula of the right coronary cusp communicating with the right side of the heart (Figure 3)
- The surgical repair of the SVA aneurysm was performed without any operative events, and the patient recovered well

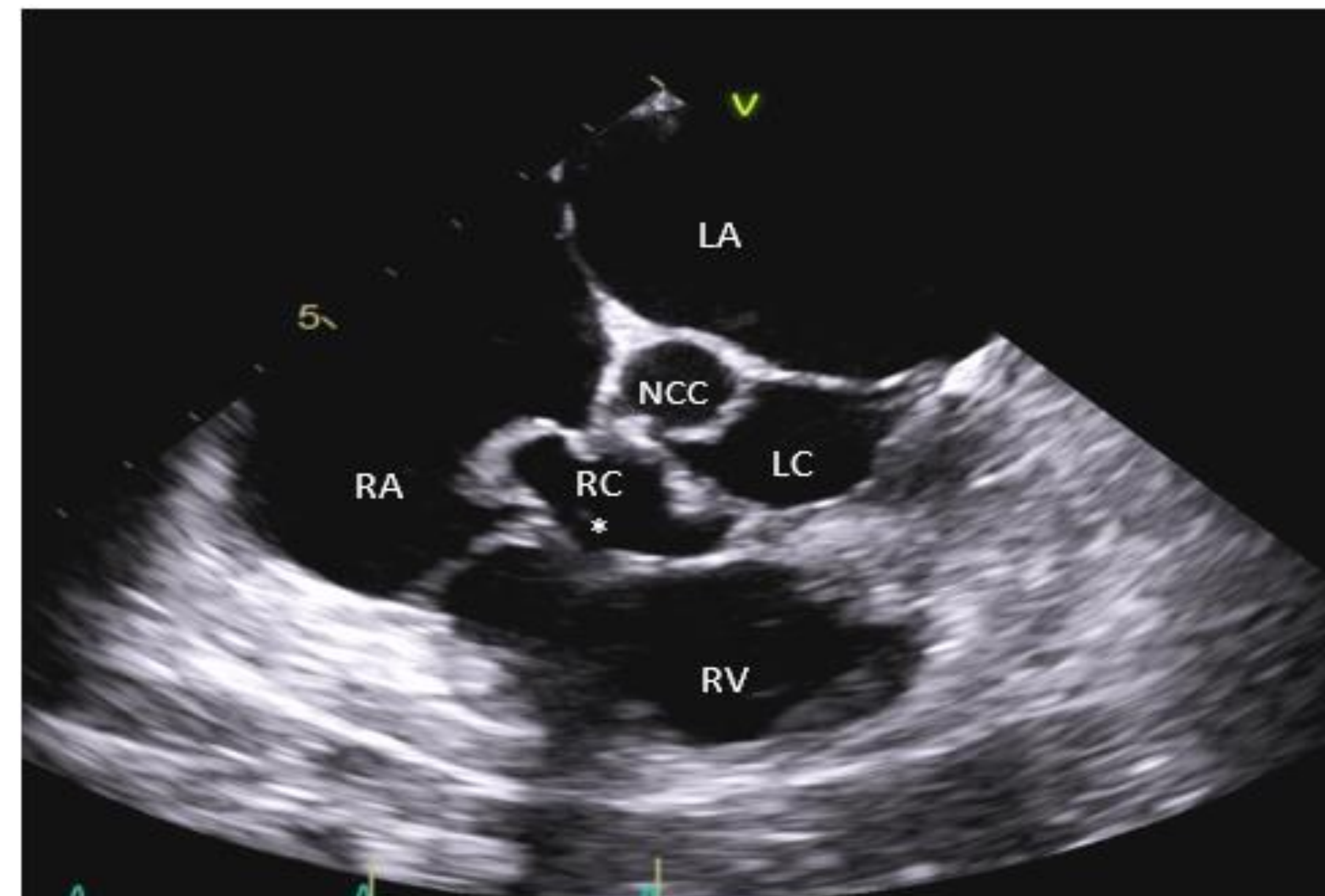


Figure 1: Transesophageal echocardiography (TEE) short axis view showing outpouching (labeled with an asterisk \*) from the right coronary sinus. Right coronary sinus (RC), left coronary sinus (LC), non-coronary sinus (NCC), right atrium (RA), left atrium (LA), right ventricle (RV)

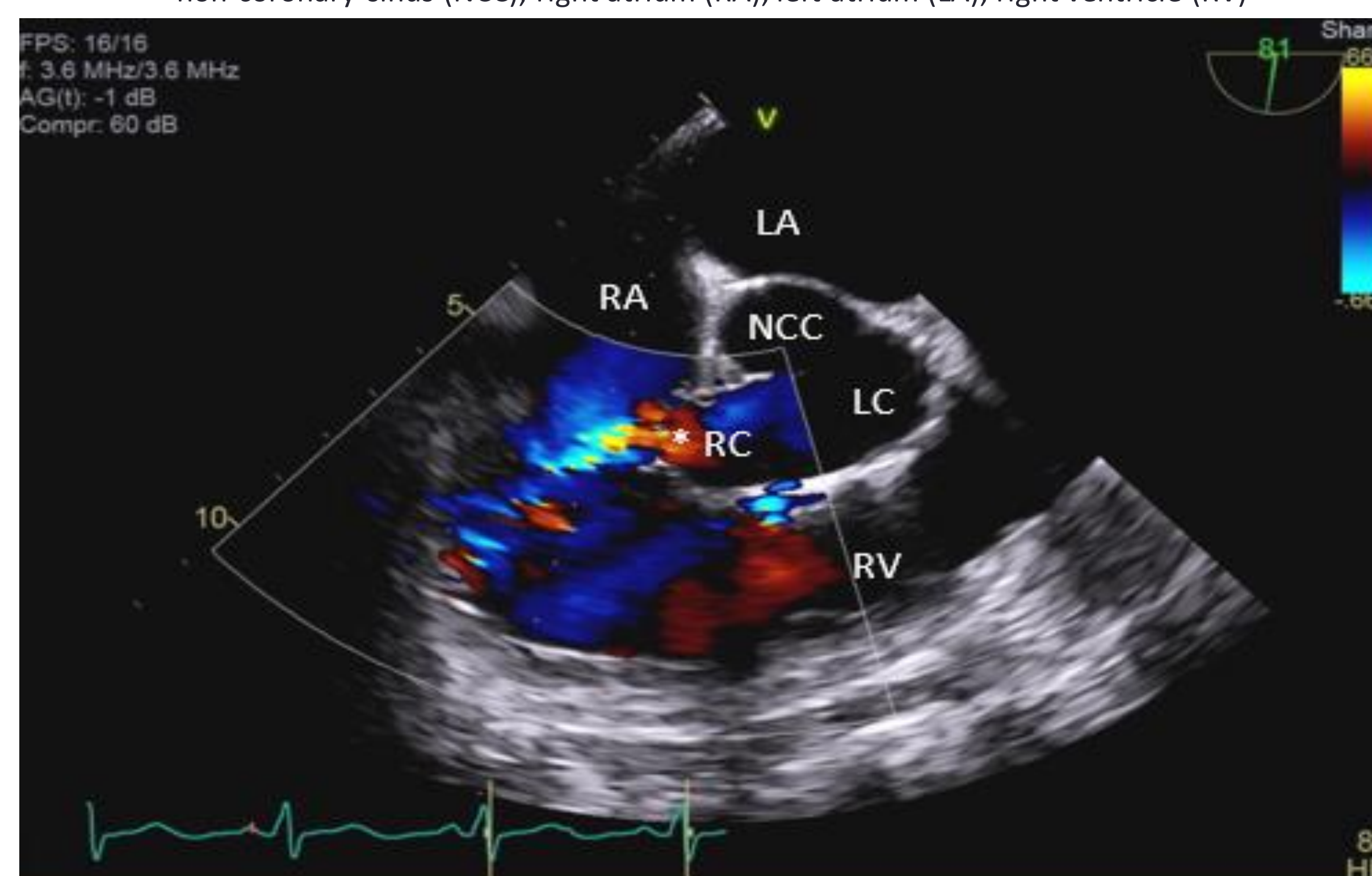


Figure 3: Transesophageal echocardiography (TEE) short axis view with color Doppler showing a jet hitting right side of the heart continuously during both diastole and systole

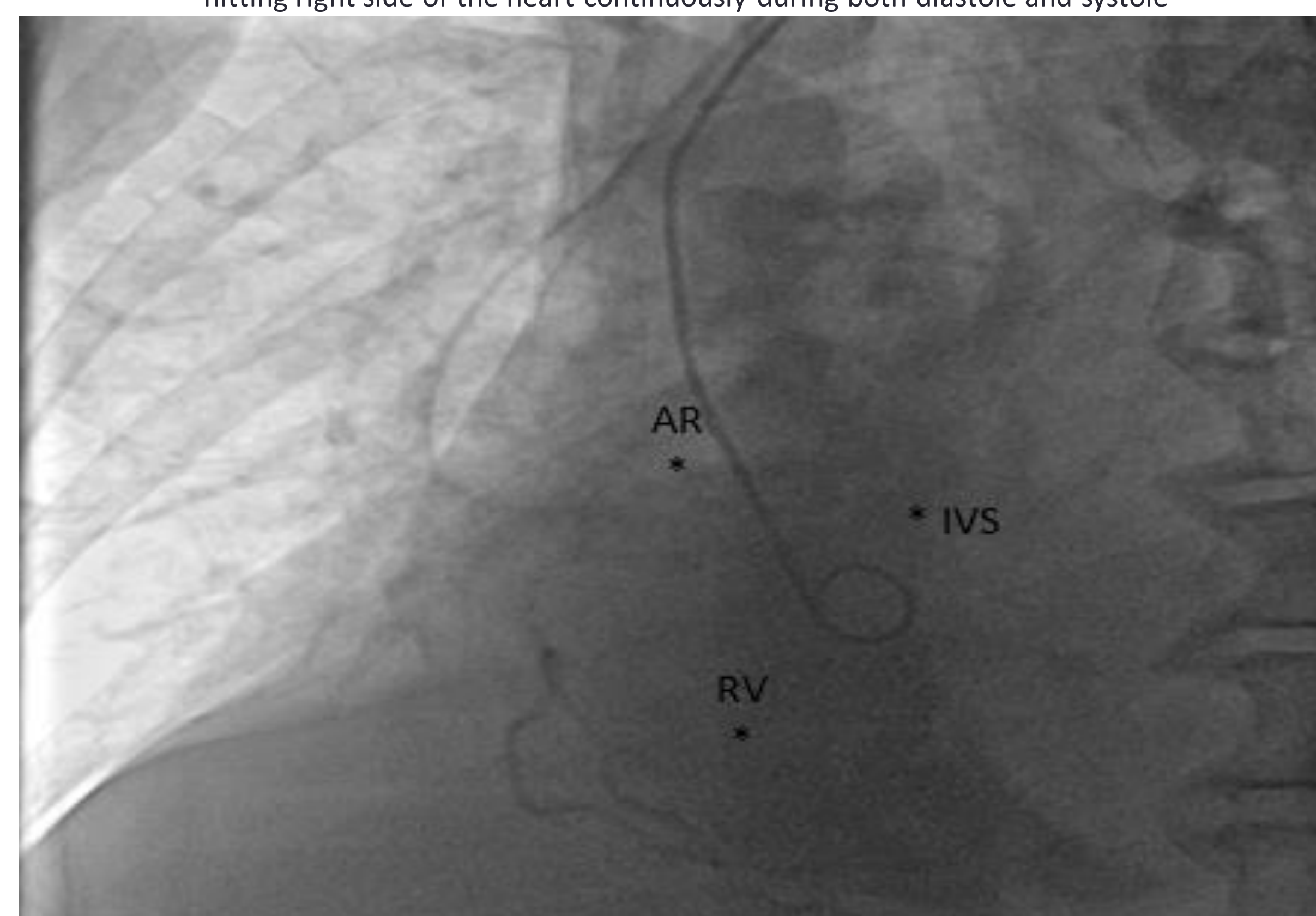


Figure 3: Cardiac catheterization with aortic root injection resulted in visualization of right heart chambers due to aneurysmal fistulization of the right coronary cusp communicating with the right side. Right ventricle (RV), interventricular septum (IVS) and aortic root (AR)

## DISCUSSION

- SVA can either be congenital or acquired in origin.
- These SVAs are associated with ventricular septal defects (VSD) in approximately 60% of patients. However, in rare cases, they can also be associated with atrial septal defects, such as a patent foramen ovale, as observed in the patient described in this report.
- A high degree of clinical suspicion is required for an accurate diagnosis of SVAs. Unruptured and small SVAs are usually asymptomatic, but as they grow in size, they can cause cardiac ischemia, cardiac conduction abnormalities, and even fatal arrhythmias like ventricular fibrillation or complete heart block.
- The first diagnostic modality in such cases is transthoracic echocardiogram. In cases where diagnosis is not clear, transesophageal echocardiogram and cardiac magnetic resonance imaging (MRI) can aid in the diagnosis, but a definitive or confirmatory diagnosis can be made by cardiac catheterization.
- Surgical intervention is recommended for patients with a SVA with a root diameter greater than 5 cm, as this size has been correlated with a higher risk of mortality.
- For patients with SVA measuring less than 5 cm, medical management can be employed. In such cases, it is crucial to maintain tight control of blood pressure, as is commonly done in patients with aortic aneurysms.

## CONCLUSION

- Sinus of Valsalva aneurysm (SVA) is a relatively uncommon cardiac condition characterized by an abnormal dilation of one or more of the three aortic sinuses.
- The patient in this case report presented with chest pain and difficulty breathing, and further testing revealed a ruptured right SVA, leading to surgical repair.
- A high degree of suspicion is crucial for an accurate diagnosis and prompt treatment to prevent potential complications.