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CASE REPORT
Cardiovascular imaging

A multimodality approach to a rare case of ruptured sinus of Valsalva aneurysm with tricuspid valve involvement: a case report

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Background Ruptured sinus of Valsalva aneurysm is a rare disease entity that is potentially life-threatening if left untreated. While imaging is the mainstay of diagnosis, resultant tricuspid valve involvement may mask typical findings providing a diagnostic challenge. Disruption of the tricuspid valve during ruptured sinus of Valsalva aneurysm with consequent tricuspid regurgitation is rare and infrequently described in the literature. Description of the utility and limitations of multimodality imaging in this scenario is equally scarce.

Case summary We review the case of a young patient presenting with acute ruptured sinus of Valsalva aneurysm and involvement of the tricuspid valve on a background of severe aortic regurgitation requiring multimodality imaging for diagnostic and pre-surgical assessment.

Discussion In young patients presenting with acute decompensation and pre-existing bicuspid aortic valve regurgitation, an increased clinical suspicion of a sinus of Valsalva aneurysm rupture is imperative. Doppler and 3D transoesophageal echocardiographic assessment should be pursued to characterize abnormal flows and clarify aetiology in the context of tricuspid involvement and resultant tricuspid regurgitation. A large-volume left–right shunt in proximity to the tricuspid annulus may result in disproportionately severe tricuspid regurgitation in the absence of annular disruption due to forced systolic opening of the leaflets by shunt flow and 'windsock' prolapse. Multimodality imaging can be essential in these cases to adequately assess the extent of the ruptured sinus of Valsalva aneurysm and overcome limitations of single modality imaging.

Keywords Case report • Congenital heart defect • Echocardiography • Tricuspid valve • Multimodality imaging

ESC curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease

Learning points

- To consider ruptured sinus of Valsalva aneurysm as a cause for acute decompensation in patients with pre-existing congenital aortic valve pathology.
- To highlight limitations of isolated imaging modalities and the utility of multimodality imaging in concomitant tricuspid valve involvement and diagnostic uncertainty.
- To appreciate the various mechanisms of resultant tricuspid regurgitation with a ruptured sinus of Valsalva aneurysm in proximity to the tricuspid valve in the absence of annular disruption.

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Introduction

Ruptured sinus of Valsalva aneurysm (RSOVA) is a rare, potentially life-threatening entity.[1] Tricuspid valve (TV) involvement is infrequent but may be challenging to evaluate and obscure typical findings. Combination of newer multimodality techniques can overcome limitations of traditional imaging modalities and aid assessment in complex cases. An illustrative case is provided.

Summary figure

Time	Events
Day 0	A 26-year-old woman with a background of bicuspid aortic valve (BAV) and aortic regurgitation (AR) presents to the emergency department with congestive heart failure and diastolic and systolic murmurs. Transthoracic echocardiogram (TTE) shows severe AR, a 'thickened' TV with new, moderate–severe tricuspid regurgitation (TR) and right ventricular (RV) dilatation. Abnormal forward flow through the TV is noted.
Day 4	Transoesophageal echocardiogram appearances show an aorto-right atrial (RA) connection with abnormal continuous TV forward flow on Doppler concerning for a RSOVA with concurrent moderate–severe TR. Further imaging is perused to assess tricuspid annular involvement due to disproportionately severe TR.
Day 6	Computer tomography angiogram shows pan-cardiac contrast enhancement despite appropriate electrocardiographic gating resulting in impaired tricuspid annular assessment. A RSOVA 'windsock' prolapse through the TV is noted.
Day 8	Agitated saline 'bubble' contrast study demonstrates a RA filling defect coursing through the TV without a defect within the RV.
Day 12	Cardiac magnetic resonance (CMR) imaging confirms a RSOVA immediately basal to the tricuspid annulus with continuous, high-velocity flow along the anterior tricuspid leaflet into the RV suggesting a spared annulus. Review of TTE images demonstrates TV malcoaptation due to 'windsock' prolapse and forced systolic TV leaflet opening due to high-velocity RSOVA flow resulting in disproportionately severe TR.
Day 33	The patient is adequately diuresed and undergoes surgical repair of the RSOVA with TV repair and annuloplasty. Position of the RSOVA is confirmed immediately basal to the TV annulus.
2 months	Patient is recovering well and is near baseline on follow-up.

Case presentation

A 26-year-old woman with a background of a non-syndromic BAV, severe AR, and preserved left ventricular (LV) systolic function presented to the emergency department with acute congestive heart failure. Examination revealed normal observations apart from a blood pressure of 102/65 mmHg and tachycardia at 92 b.p.m. Significant bipedal oedema was noted with systolic and diastolic murmurs and clear lung fields on auscultation.

An electrocardiogram showed sinus tachycardia. Chest X-ray revealed cardiomegaly and pulmonary congestion. Laboratory tests showed normal electrolytes, renal function, and complete blood count. A clinical diagnosis of severe AR with decompensated heart failure was made and further imaging arranged.

Transthoracic echocardiography (TTE) confirmed a type 1 BAV, severe AR, and preserved LV contractility with a 40 mm ascending aorta diameter. New RV dilatation was identified with a thickened appearance of TV leaflets and moderate–severe, central TR initially presumed functional (*Figure 1A and B*). Abnormal, high-velocity, continuous forward flow through the TV was noted on colour flow mapping (CFM) resulting in bidirectional systolic TV flow in the context of TR (*Figure 1C*; Supplementary material online, *Video S1*).

Further assessment with transoesophageal echocardiography (TOE) confirmed an abnormal TV–aortic interface with high-velocity flow on CFM throughout the cardiac cycle suggestive of a RSOVA (*Figure 2A and B*; Supplementary material online, *Video S2*) Doppler assessment confirmed continuous forward flow through the region and the TV (*Figure 2B and C*). 3D rendering further differentiated anterolaterally directed RSOVA flow from central, posteriorly directed moderate–severe TR (*Figure 3*; Supplementary material online, *Video S3*). The degree of TR was felt to be disproportionate to the degree of RV dilatation. A small, high-velocity jet was additionally noted on the RV aspect of the tricuspid annulus concerning TV disruption, and further investigation was undertaken (*Figure 3C*; Supplementary material online, *Video S2*).

An agitated saline study conducted to delineate the intracardiac shunt from the right-sided flows demonstrated a large filling defect immediately superior to the TV in an otherwise bubble-filled RA without a deficit within the RV (see Supplementary material online, *Video S4*).

Computerized tomography angiography (CTA) demonstrated normal coronary ostia. Pan-cardiac enhancement due to the substantial shunt resulted in impaired visualization of tricuspid annular integrity, but the origin of a large non-coronary cusp defect was seen immediately basal to the septal TV leaflet insertion. Shunt course was similarly obscured, but RSOVA 'windsock' prolapse through the TV was observed (*Figure 4*).

Cardiac magnetic resonance imaging confirmed RSOVA origin above the tricuspid annulus coursing along the anterior TV leaflet into the RV suggestive of a spared tricuspid annulus (*Figure 5*; Supplementary material online, *Videos S5 and S6*) alongside moderate–severe TR (regurgitant volume 66 mL, regurgitant fraction 31%).

On further review, 'thickened appearance' of the TV on TTE represented the RSOVA 'windsock' through which high-velocity flow is demonstrated (*Figures 1A* and *2*; Supplementary material online, *Video S1*). Size and proximity of the 'windsock' result in prolapse into the TV with consequent malcoaptation and leaflet 'flutter' as they are further separated by high-velocity forward flow during systole resulting in significant TR in the context of mild annular dilatation (see Supplementary material online, *Video S7*).

The patient was diuresed and proceeded to surgical RSOVA repair with a CardioCel patch and aortic valve replacement. Perioperative findings confirmed a non-coronary RSOVA with a large 'windsock' in close proximity to the tricuspid septal-anterior commissure requiring repair and annuloplasty.

The patient had excellent recovery with no residual RSOVA, normalized RV size, trivial TR (see Supplementary material online, *Video S8*), and near-premorbid exercise tolerance.

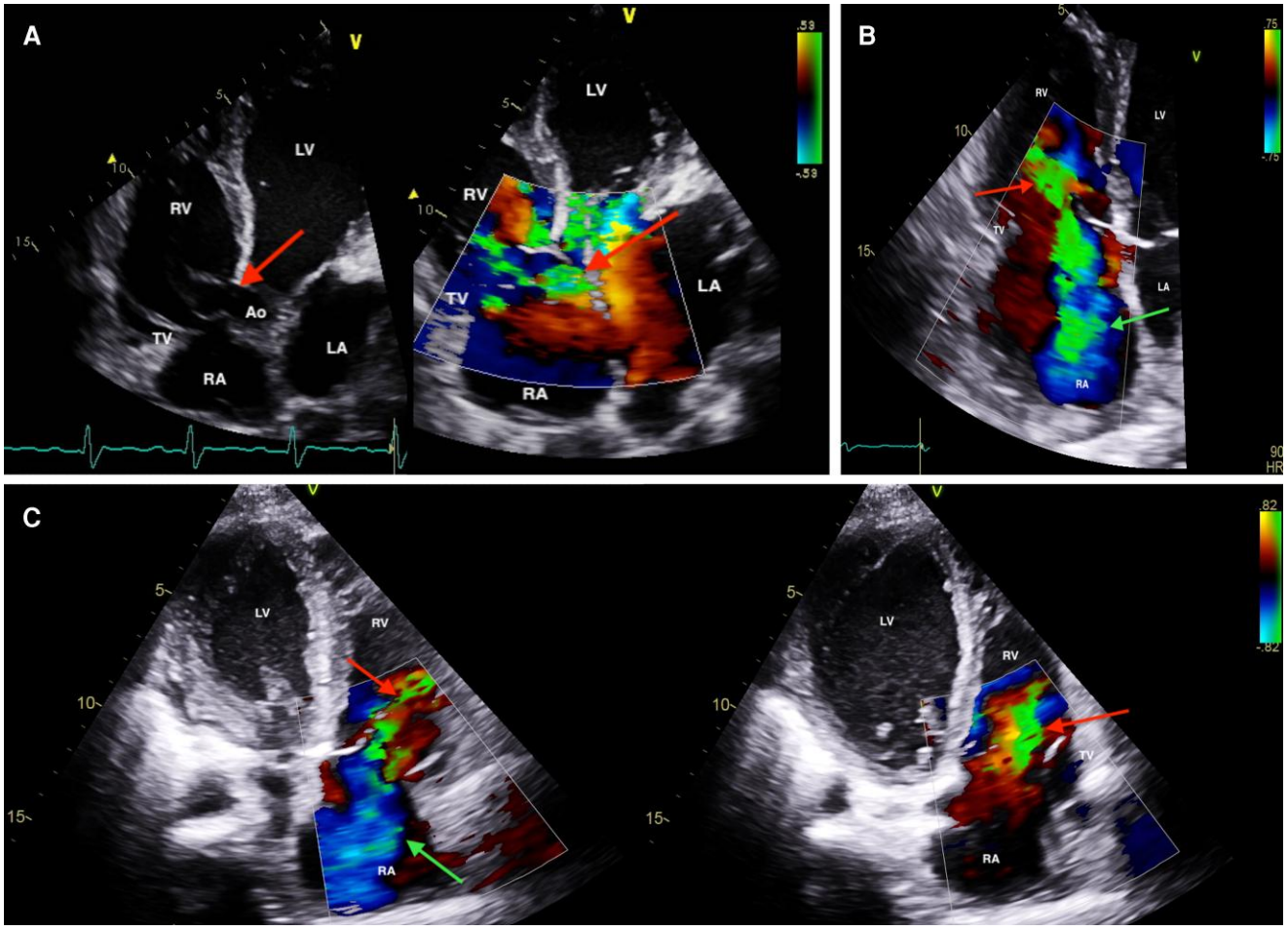


Figure 1 Transthoracic echocardiographic images. Apical five-chamber view (*A*) demonstrating a ruptured sinus of Valsalva aneurysm in close proximity to septal tricuspid valve leaflet insertion (left) with colour flow through the defect into the tricuspid valve (right). Apical four-chamber view (*B*) exhibiting bidirectional systolic flow through the tricuspid valve with moderate–severe tricuspid regurgitation. Reverse apical four-chamber view (*C*) shows bidirectional systolic flow (left) and high-velocity diastolic forward flow through the tricuspid valve (right). RV, right ventricle; LV, left ventricle; TV, tricuspid valve; Ao, aorta; RA, right atrium; LA, left atrium; red arrow, RSOVA and/or RSOVA flow; green arrow, tricuspid regurgitation; RSOVA, ruptured sinus of Valsalva aneurysm; TR, tricuspid regurgitation.

Discussion

A RSOVA is a potentially life-threatening complication of a rare disease entity—sinus of Valsalva aneurysm (SOVA).[1] Sinus of Valsalva aneurysm is a congenital or acquired dilatation of the aortic sinuses at the aortic media–annulus fibrosis junction secondary to weakened elastic lamina.[1] With an estimated prevalence around 0.09%, SOVA most commonly affects the right sinus (65–85%), followed by, as in this case, the non-coronary (10–30%) and left sinus (<5%).[2,3] While majority of SOVA is incidental diagnosis, 33–50% present following rupture with the highest incidence between ages 20–40 years.[4] Ruptured sinus of Valsalva aneurysm results in a shunt usually from the aortic sinus to the cardiac chambers or less commonly pericardium or extra-cardiac space.[5] Other frequently associated conditions include ventricular septal defects (30–60%), AR (20–30%), BAV (10%), and less frequently coronary anomalies.[2] Tricuspid involvement is infrequent and seldom described in the literature but can complicate RSOVA diagnosis and assessment.[3]

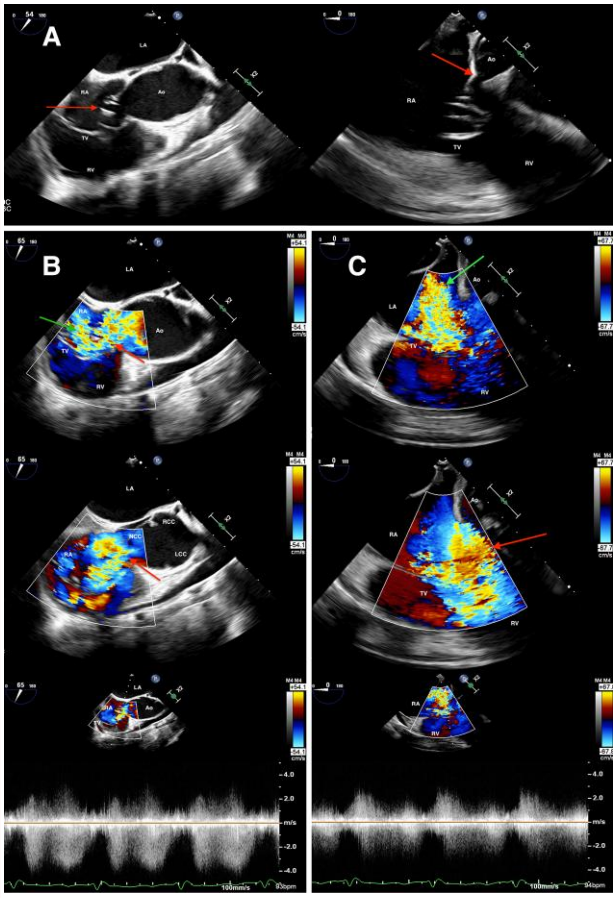


Figure 2 Transoesophageal echocardiographic images. Modified mid-oesophageal aortic valve short-axis view (*A*, left) and mid-oesophageal five-chamber view (*A*, right) revealing disruption of the aortic sinus and prolapse of the 'windsock' into the tricuspid valve. Modified mid-oesophageal aortic valve short-axis views (*B*) differentiating two individual systolic jets representing tricuspid regurgitation and ruptured sinus of Valsalva aneurysm flow (top), high-velocity diastolic ruptured sinus of Valsalva aneurysm flow (mid), and Doppler profile describing continuous flow through the ruptured sinus of Valsalva aneurysm (bottom). Mid-oesophageal five-chamber views (*C*) demonstrating moderate–severe systolic tricuspid regurgitation (top) and high-velocity diastolic ruptured sinus of Valsalva aneurysm flow through the TV (mid). Doppler profile demonstrates continuous forward flow and bidirectional systolic flow through the tricuspid valve (*C*, bottom). Red arrow, RSOVA and/or RSOVA flow; green arrow, TR. Other abbreviations as in *Figure 1* legend.

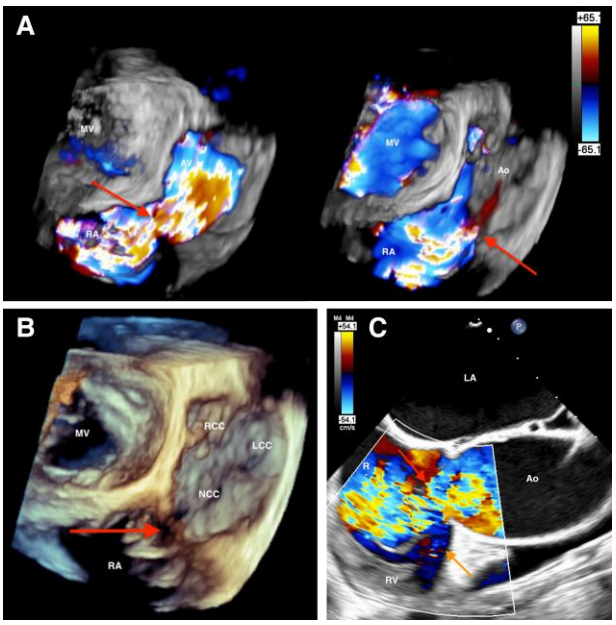


Figure 3 Transoesophageal 3D mid-oesophageal tilted enface left atrium view (*A*, *B*) demonstrating a ruptured sinus of Valsalva aneurysm with systolic (*A*, left) and diastolic (*A*, right) forward flow into the right atrium. Modified mid-oesophageal aortic valve short-axis view showing potential systolic flow apical to the septal tricuspid annulus concerning annular disruption (*C*, orange arrow). 3D, 3 dimensional.

Imaging is the mainstay of diagnosis with echocardiography frequently demonstrating continuous flow from the aortic root to the relevant cardiac chamber. An abnormal mass may be demonstrated within the cardiac chamber representing the 'windsock' of a RSOVA through which flow may be detected. Proximity of the mass to valvular structures can display an impression of endocarditis or valve thickening. While TV involvement is rare, consequent significant TR can confound assessment of RSOVA flow, particularly on TTE with more optimal visualization on TOE.[6,7] Doppler assessment and 3D TOE offer the most convincing evidence of a RSOVA with clear differentiation of continuous antegrade flow from retrograde TR. Bidirectional systolic flow through the TV was noted here.

The assessment of tricuspid annular involvement can be additionally challenging in the context of a large-volume shunt. Electrocardiography-gated contrast CTA usually provides high-resolution imaging ideal for RSOVA localization with contrast extension into the adjacent cardiac chamber.[2] Limitations of this method in the context of high-volume shunts as demonstrated include pan-cardiac enhancement obscuring contrast passage and annular structure definition. Cardiac magnetic resonance can accurately localize RSOVA origin and course, quantify valvular regurgitation, and may supply superior assessment of RSOVA extent if a large shunt is suspected.[2]

Untreated RSOVA has an average life expectancy of 1 year; thus, intervention in the form of open or transcatheter repair is essential.[1,2,8] Suggested surgical approaches include aortotomy, through the affected cardiac chamber or a dual approach. The choice of approach is dictated by aneurysm dimensions, associated valvular pathology, presence of other congenital anomalies, and chamber involved. Patch closure is utilized in large ruptures reserving primary closure for smaller lesions.[1] The extent of the RSOVA and valve involvement necessitated a dual approach and patch repair in this patient.

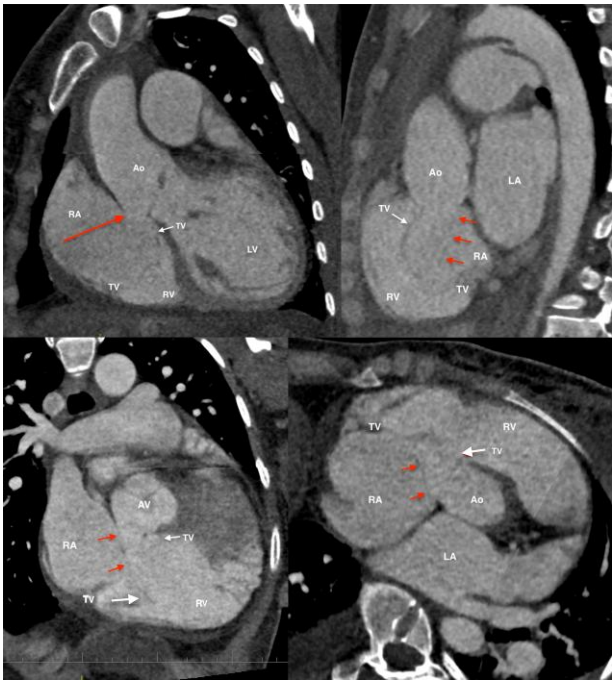


Figure 4 Contrast computed tomography angiogram images with pan-cardiac enhancement despite electrocardiographic gating. Standard coronal (top left) and sagittal (top right) reconstruction views and oblique coronal (bottom left) and oblique axial reconstruction views demonstrate the proximity of the ruptured sinus of Valsalva aneurysm to the septal tricuspid valve leaflet insertion with suboptimal annular definition impairing assessment of annular involvement. Prolapse of the 'windsock' through the tricuspid valve is demonstrated. Abbreviations as in *Figure 1* and *2* legends.

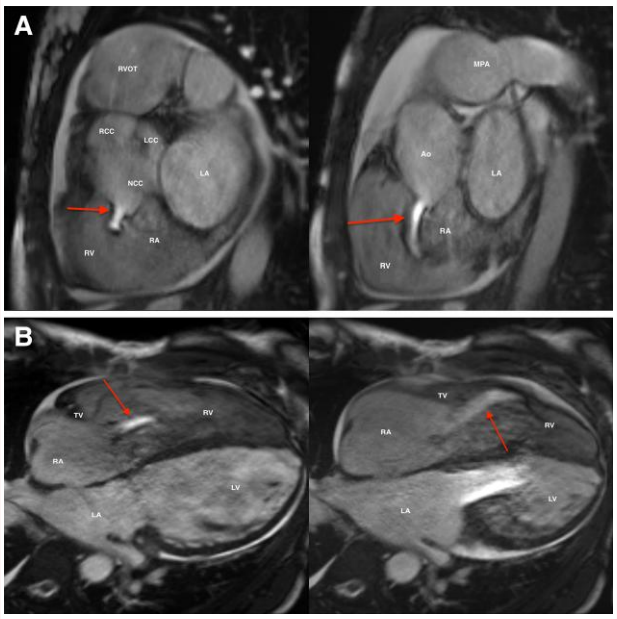


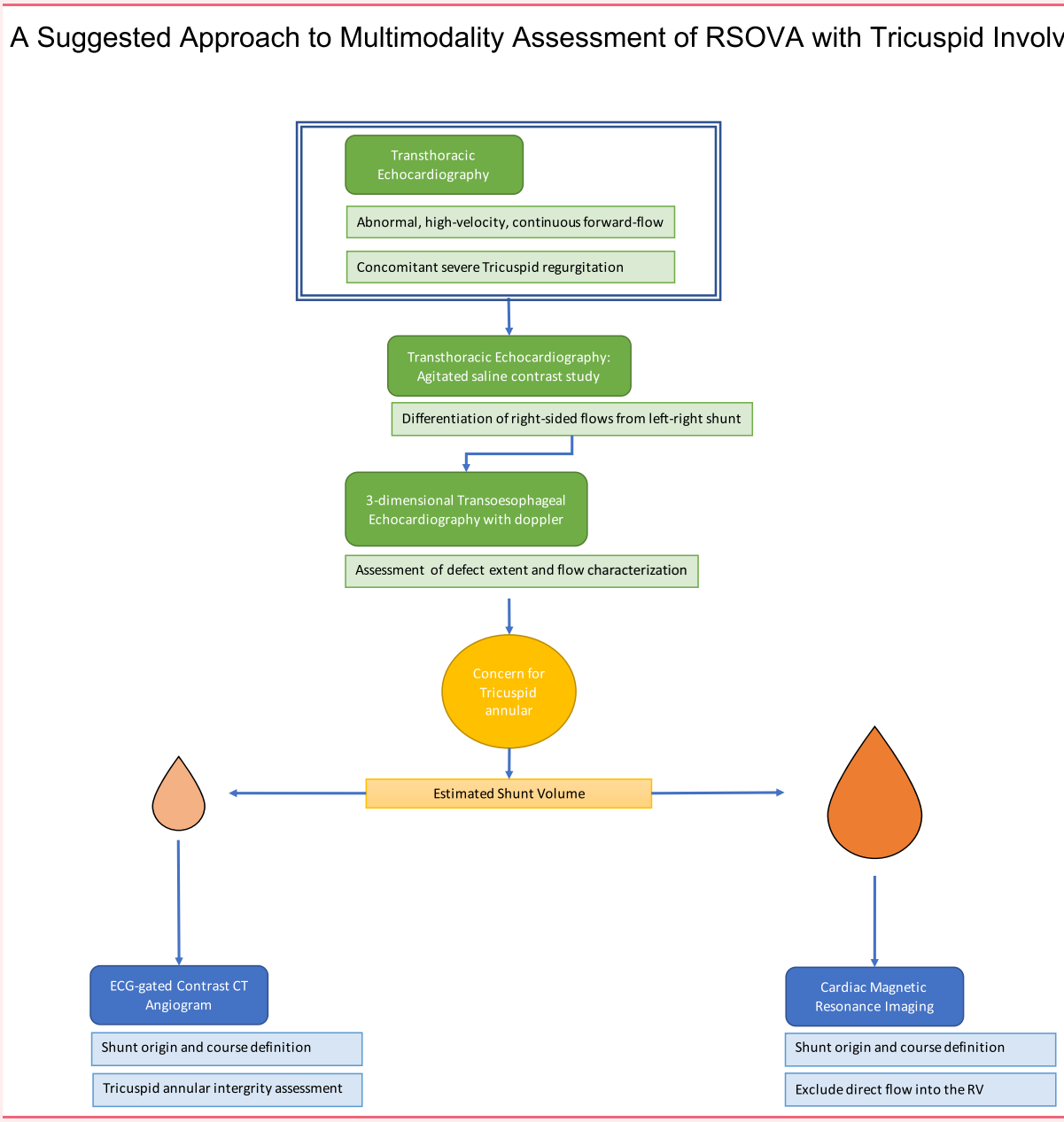
Figure 5 Cardiac magnetic resonance imaging oblique aortic sagittal views (*A*) demonstrating a non-coronary cusp ruptured sinus of Valsalva aneurysm with high-velocity flow through the right atrium, along the anterior tricuspid valve leaflet into the right ventricle. Cardiac magnetic resonance imaging TRUFI retro four-chamber view depicting systolic (left) and diastolic (right) high-velocity forward flow through the tricuspid valve. No direct flow into the right ventricle is demonstrated.

Conclusion

Ruptured sinus of Valsalva aneurysm is a rare disease entity that has a poor prognosis if left untreated due to progressive congestive heart failure.[1] While TTE is often sufficient for diagnosis, infrequent tricuspid involvement with resultant TR may mask typical findings, providing diagnostic difficulties.[5] In these scenarios, Doppler imaging and 3D TOE may be key to flow definition. Agitated saline studies may be effective at discriminating TR from RSOVA flow and are likely underutilized.

An extensive RSOVA in proximity to the TV annulus may precipitate malcoaptation of the leaflets due to 'windsock' prolapse and forced systolic opening of the leaflets due to high-velocity RSOVA flow resulting in disproportionately severe TR in the absence of tricuspid annular disruption. In such cases, multimodality imaging may hold the key to demarcation of RSOVA course and definitive tricuspid interrogation. The choice of imaging technique should be guided by the estimated magnitude of the shunt to attain optimal visualization of RSOVA extent (*Table 1*).

Table 1 A flow diagram describing a suggested approach to multimodality assessment of ruptured sinus of Valsalva aneurysm with tricuspid involvement based on estimated shunt volume



RSOVA, ruptured sinus of Valsalva aneurysm; ECG, electrocardiograph; RV, right ventricle.

Lead author biography

Dr Vindhya Wilson is a graduate of the University of Auckland in New Zealand. She is currently a final year cardiology advanced trainee at Wellington Regional Hospital in New Zealand. She has a strong interest in multimodality imaging and structural heart disease imaging as well as medical education and clinical governance.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Consent: Written informed consent was obtained from the patient for publication in accordance with COPE guidelines and documented in clinical notes.

Conflict of interest: None declared.

Data availability

The data underlying this article are available in the article and in its online supplementary material.

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