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**CASE REPORT**
*Cardiovascular imaging*

# Scimitar syndrome with large atrial septal defect and a rare partial anomalous venous drainage in an adult: a case report

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**Background** Scimitar syndrome is a very rare congenital cardio-pulmonary disease with anomalous right pulmonary vein draining either partially or completely into the inferior vena cava. It is called Scimitar syndrome due to the classical appearance in the chest X-ray, which resembles the curved blade of Turkish sword 'Scimitar'. It commonly associates with atrial septal defect (ASD), hypoplasia of the right lung, dextroposition of the heart, and pulmonary hypertension (PHT).

**Case summary** A 67-year-old lady, diagnosed with atrial fibrillation and moderate PHT 3 years ago, presented with worsening bilateral ankle oedema and New York Heart Association class III shortness of breath. Chest X-ray showed the Scimitar appearance. The trans-thoracic and trans-oesophageal echocardiograms revealed a 46 mm ASD and a partial anomalous pulmonary venous drainage (PAPVD) of the right upper pulmonary vein (RUPV) into the right atrium at the junction of the atria. Three-dimensional reconstruction of the computed tomographic pulmonary angiogram confirmed Scimitar syndrome of the right lower pulmonary vein (RLPV). We managed her conservatively on her wish. After 13 months, she succumbed due to a massive stroke.

**Discussion** We describe a very rare case of an elderly lady who has Scimitar syndrome with an ASD and evidence of PAPVD of the RUPV; thus, we intend to provide an antecedent for further cases, for prompt and accurate diagnosis and timely interventions in order to prevent life-threatening complications.

**Keywords** Case report • Scimitar syndrome • Atrial septal defect • Partial anomalous pulmonary venous drainage • 3D reconstruction CTPA

**ESC curriculum** 9.6 Pulmonary hypertension • 9.7 Adult congenital heart disease • 2.4 Cardiac computed tomography • 2.1 Imaging modalities

## Learning points

- In a patient with pulmonary hypertension (PHT), it is important to look for all the possible secondary causes even if some causes are rare.
- Scimitar syndrome should be suspected when a classical chest X-ray appearance is seen.
- Reconstructed three-dimensional (3D) computed tomographic pulmonary angiogram (CTPA) is at least equivalent if not superior to right heart catheterization (RHC) and pulmonary venogram in diagnosing Scimitar syndrome.
- RHC is still mandatory to precisely measure the PHT and Qp:Qs for management decisions.
- Different associations could exist with Scimitar vein; hence, it is important to look for all the possibilities.

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# Introduction

Scimitar syndrome is extremely rare in adults as it is either compensated well or manifest early due to complex associations. It is important to have a high degree of suspicion of this condition simply because it could go unnoticed in compensated adults.

It associates commonly with atrial septal defect (ASD), pulmonary hypoplasia, and patent ductus arteriosus (PDA).[1–3] There are numerous other associations as well, but we did not come across a partial anomalous pulmonary venous drainage (PAPVD) of the right upper pulmonary vein (RUPV) causing an extracardiac chamber which enters to the right atrium (RA) in the literature. Scimitar is best managed surgically, especially if the total pulmonary to total systemic shunt (Qp:Qs) is >1.5 or if it associates with other complex anomalies.[4] Otherwise, medical management would be the best for adults as the chances of surgical failure due to chronic pulmonary modifications are high.

# Summary figure

| | |
|---|---|
| 3 years prior to admission | First presentation with palpitations. Diagnosis of atrial septal defect (ASD) and atrial fibrillation |
| Admission | Worsening shortness of breath (SOB) and ankle oedema. Commencement of heart failure medication |
| Day 1 | Trans-thoracic echocardiogram (TTE) and chest X-ray (CXR): Large ASD/?Appearance of curvilinear Scimitar vein on right heart border in CXR |
| Day 4 | Trans-oesophageal echocardiogram (TOE): partial anomalous pulmonary venous drainage (PAPVD) in the right upper pulmonary vein (RUPV), large ASD, and absent right lower pulmonary vein (RLPV) |
| Day 10 | Discharged on warfarin, furosemide, digoxin, and bisoprolol |
| 2 weeks after discharge | Computed tomographic pulmonary angiogram (CTPA) confirms the Scimitar vein. Patient declined further intervention |
| 13 months after discharge | Massive right middle cerebral artery (MCA) infarct after missing warfarin for 1 week |
| 2 days after the second admission | Dies due to complications of stroke. |

## Patient presentation

A 67-year-old lady who was diagnosed with atrial fibrillation and moderate pulmonary hypertension (PHT) after an echocardiogram 3 years ago presented to us with worsening bilateral ankle oedema, New York Heart Association class III shortness of breath, and palpitations over 3 months. She did not have a past history or a family history of pulmonary disease, thyroid disease, or any other cardiac disease. She also denied any history of recurrent lung infections.

On examination, she was short-statured and cachectic with a body mass index (BMI) of 14.16 kg/m$^2$. There was mild bilateral pedal oedema but no peri-orbital oedema. Her pulse was 118 b.p.m. and irregular. Blood pressure was normal. There was a systolic murmur best heard at the pulmonary area, a loud P2, evidence of right ventricular heave, and elevated jugular venous pressure. She was tachypnoeic with reduced air entry and dullness in the right lower zone but without coarse crepitations, tracheal deviation, or bronchial breathing. Her neurological and musculoskeletal examinations were normal.

Her basic blood analysis including complete blood count, renal functions, erythrocyte sedimentation rate, lipid profile, fasting blood sugar, and liver functions was normal. However, during her stay, aspartate transaminase (AST) and alanine transaminase (ALT) rose to 228 U/L (<40 U/L) and 63 U/L (<40 U/L), respectively. Her clotting profile was normal on admission despite being on warfarin. Her C-reactive protein was elevated at 48 mg/dL (0–5 mg/dL), and thyroid-stimulating hormone (TSH) was low at 0.1 mIU/L (04–4.0 mIU/L). Interestingly, free T3 and T4 levels were normal at 3.54 pg/mL and 1.1 ng/dL, respectively. Ultrasound scan of the thyroid showed only a colloid multinodular goitre. Her N-terminal pro-B-type natriuretic peptide (NT-pro BNP) was only mildly elevated at 358 pg/mL (<125 pg/mL), even though we expected it to be more.

Her electrocardiogram showed evidence of atrial fibrillation, chamber dilatation, right axis deviation, and mild ST depressions with T-wave inversions in inferior leads. However, two samples of troponin I were negative. The trans-thoracic echocardiogram (TTE) revealed volume overload in the right heart as evidenced by moderately dilated RA, dilated right ventricle (RV) with a diameter of 57 mm at the base, moderate tricuspid regurgitation (TR), and marginally low (14 mm) tricuspid annular plane systolic excursion (TAPSE). It also showed a high probability of PHT (TR velocity >3.4 m/s; TR pressure gradient = 68 mmHg), mildly dilated left atrium (LA) with a diameter of 44 mm in parasternal long-axis view, lower normal left ventricular ejection fraction (LVEF) of 50% by Simpson's method, and a large ASD. There were widespread fluffy shadows, grossly enlarged pulmonary arteries, and the Scimitar appearance (*Figure 1*) in the chest X-ray (CXR). Of note, the classical Scimitar appearance can be easily misinterpreted as double shadowing of atria which is mainly seen in left atrial enlargement.

Trans-oesophageal echocardiogram (TOE) and the subcostal TTE views at the time of the TOE revealed a 46 mm ostium secundum

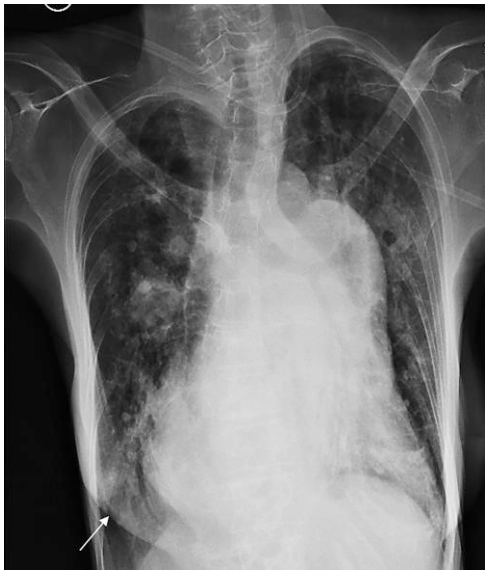

**Figure 1** Chest X-ray showing right-sided curvilinear "Scimitar vein" (arrow) and collapsed left lower lobe.

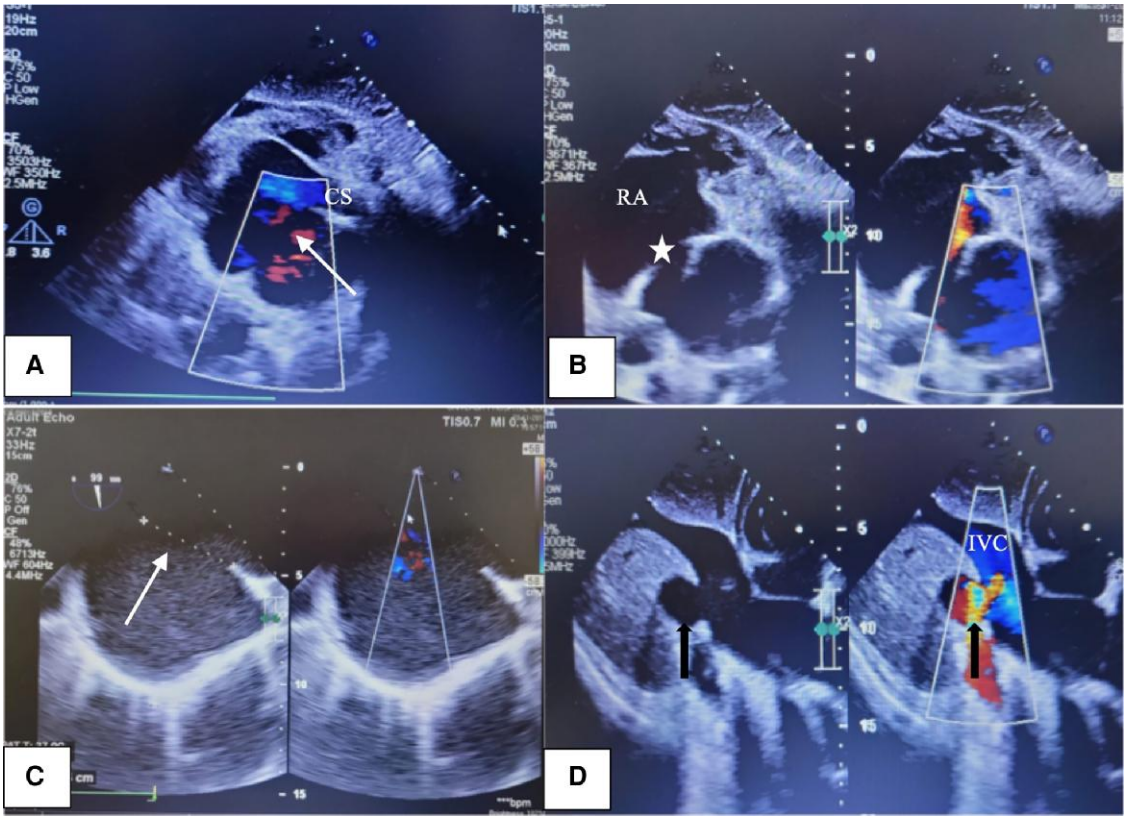


**Figure 2** TTE and TOE images. (*A*) Subcostal view of TTE shows ASD (white arrow) and dilated coronary sinus (CS). (*B*) Further superior angulation shows extracardiac chamber extending through the LA like a Cor Triatriatum to enter the grossly dilated RA (*) and disappearance of the ASD. (*C*) TOE measuring ostium secundum ASD (white arrow) at 46.4 mm. (*D*) Dilated IVC and possible drainage of Scimitar vein to the IVC (black arrow). ASD, atrial septal defect; IAS, intra-atrial septum; IVC, inferior vena cava; LA, left atrium; RA, right atrium; TOE, trans-oesophageal echocardiogram; TTE, trans-thoracic echocardiogram.

(OS) ASD, visually dilated coronary sinus, and a grossly enlarged RA [mid-level diameter = 66 mm; length = 69 mm; area of 35.77 $cm^2$ (normal < 18.5 $cm^2$)]. It also showed that the RUPV is forming an extracardiac chamber which traverses through the LA, almost creating a supero-posterior Cor Triatriatum Sinistrum, to enter the RA (*Figure 2*). Trans-oesophageal echocardiogram could not identify the right lower pulmonary vein (RLPV). A high-resolution computer tomography (HRCT) revealed moderate cardiomegaly, diffuse alveolar opacifications due to pulmonary oedema, and bilateral peri-hilar congestion (*Figure 3*). Her lung function tests showed only a mild restrictive disease which would probably be due to poor effort in execution.

Due to the classical Scimitar appearance in CXR, a computed tomographic pulmonary angiogram (CTPA) was done. It revealed a Scimitar vein draining the right middle and lower lobes into the intra-hepatic inferior vena cava (IVC), the RUPV, and part of the right middle lobe veins draining into the RA via an extracardiac chamber, a grossly dilated RA, mildly dilated RV and LA, and an OS ASD (*Figure 4A–D*). It also showed an enlarged pulmonary trunk (left = 36 mm and right = 34 mm) and mild atheromatous calcification in the aorta. It was negative for chronic thromboembolism, chronic lung disease, and coronary calcifications. Three-dimensional reconstruction of the CTPA clearly displayed the abnormal venous drainage confirming Scimitar syndrome (*Figure 5*). Attempts to proceed into a right heart catheterization and pulmonary arterial-venous angiography for further confirmation failed as the

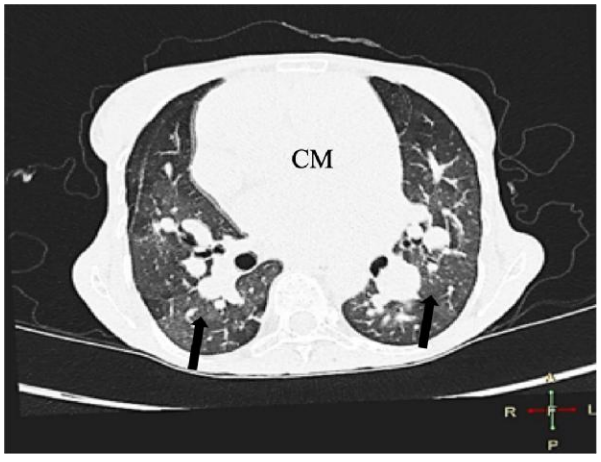


**Figure 3** High-resolution computer tomography (HRCT) of the chest revealing moderate cardiomegaly (CM), diffuse alveolar opacifications (black bold arrow) due to pulmonary oedema, and bilateral peri-hilar congestion.

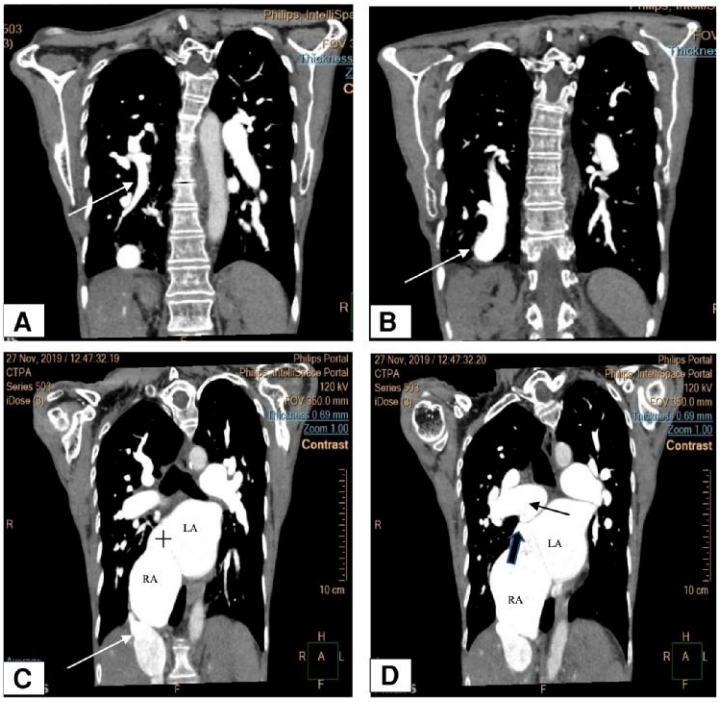


**Figure 4** Computed tomographic pulmonary angiography. (*A–B*) The course of the RLPV (white arrows) and its insertion. (*C*) The ostium secundum ASD (black cross) and the insertion point of the RLPV to the IVC (white arrow). (*D*) RUPV draining into the RA (black bold arrow) after forming an extracardiac chamber (black thin arrow). ASD, atrial septal defect; IVC, inferior vena cava; LA, left atrium; RA, right atrium; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein.

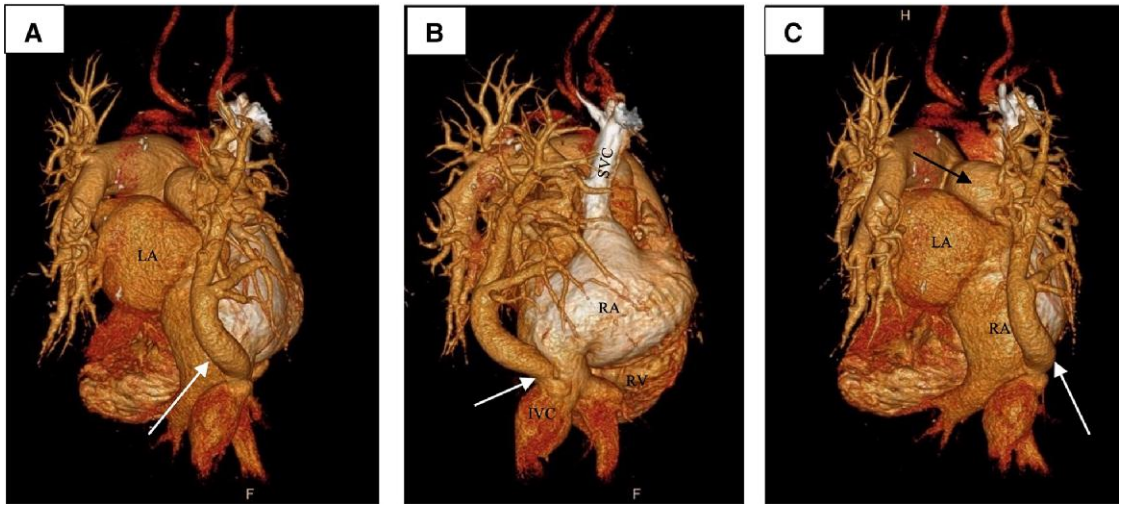


**Figure 5** Three-dimensional construction of computed tomographic pulmonary angiogram. (*A*) Scimitar vein (white arrow) formed by the right lower and middle pulmonary veins. (*B*) Insertion point (white arrow) of the Scimitar vein to the IVC. (*C*) Extracardiac chamber (black arrows) of the right upper pulmonary vein and the Scimitar vein (white arrow). IVC, inferior vena cava; LA, left atrium; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

patient declined. As per the current recommendations, the diagnosis was made on three-dimensional (3D) reconstructed CTPA and the patient was offered surgical correction due to the following reasons: echocardiographic evidence of Qp:Qs being >1.5:1, evidence of right heart congestion being present in several imaging modalities, and PHT estimated by TR pressure gradient being within correctable limits. Unfortunately, the patient adamantly declined surgical correction. Respecting the patient's autonomy, we had to resort to conservative

management and continued her warfarin to maintain an international normalized ratio (INR) of 2–3 with regular follow-ups. Digoxin 0.25 mg and bisoprolol 2.5 mg were given for rate control, and furosemide 40 mg was added to relieve her right heart failure symptoms. She was not considered for rhythm control for her atrial fibrillation because of the near-normal ejection fraction (EF), high frailty, and increased probability of recurrence due to uncorrected cardiac anomaly.

After 13 months of symptoms and complication-free period, she presented with a massive stroke due to defaulting of warfarin. On admission, the INR was 1.1 and the computed tomography (CT) brain showed a massive right middle cerebral artery (MCA) infarction. Although necessary medical management was administered, she succumbed due to complications.

## Discussion

Scimitar syndrome, also known as pulmonary venolobar syndrome, is a very rare congenital cardio-pulmonary disease with an anomalous right lower pulmonary vein draining directly into the IVC. This could be either partial or complete. Atrial septal defect is the most commonly associated congenital heart disease.[1,2] It also associates commonly with hypoplastic right lung and rarely with tetralogy of Fallot, PDA, bicuspid aortic valve, coarctation of the aorta, total anomalous pulmonary venous drainage, subaortic stenosis, ventricular septal defect, hypoplastic left heart syndrome, and dextroposition of the heart.[3,5–8] The incidence is 1–3 per 100 000 live births with a 2:1 female predominance and accounts for 3–6% of PAPVD.[8–10] The true incidence may be higher because some patients can be asymptomatic. It is mainly a neonatal or childhood disease and is extremely rare in adults. This was first identified in autopsies by Cooper[11] in London and Chassinat[12] in Paris in 1836. The first clinical diagnosis was made by Dotter[13] and his colleagues in 1949, and the name 'Scimitar' was given by Neill et al.[14] in 1960 as the pulmonary vein is narrow superolaterally and broad inferomedially, resembling the Turkish sword 'Scimitar'. This may or may not have an anomalous systemic artery supplying the affected segment of the lung.

Clinical features can be in a wide spectrum, from asymptomatic to severe right heart failure and PHT. Clinical features, anatomy, and pathophysiology vary vastly, according to associated anomalies and the presenting age. Symptoms are more, associations are complicated, and the prognosis is moderate in childhood disease. The prognosis is better in adults, probably because the survival up to adulthood was allowed by less grave associations and the body being accustomed to living with it. Association of a large ASD and a partial anomalous right upper pulmonary venous drainage to the RA at the junction of the atria in adults is not documented in literature as they become symptomatic and develop significant PHT in early life due to the large ASD and PAPVD. In the past, the diagnosis of Scimitar syndrome was confirmed by performing a pulmonary venous and arterial angiography. However, according to current recommendations, a 3D reconstructed CTPA is suffice for the confirmation and is at least equivalent if not superior to right heart catheterization.[7] Even though the diagnosis could be made with a 3D reconstructed CTPA, it is of utmost importance that the PHT and Qp:Qs should be precisely measured for management decisions. A right heart catheterization (RHC) was still mandatory in this regard as the PHT should not be estimated by the TR pressure gradient but be measured accurately by an RHC as per the new guidelines of PHT.[15] Qp:Qs also should ideally be assessed by an RHC. Unfortunately, we had to calculate the pulmonary pressure by TR pressure gradient and Qp:Qs by echocardiographic parameters which is no longer recommended, as we could not proceed with the RHC.

Surgery has not been relatively successful in adults due to the chronic pulmonary changes that would have already happened by the time the diagnosis is made and the high incidence of surgical complications in adulthood. Hence, it is reserved only for patients who have a notable shunt with pulmonary circulatory overload defined as a Qp:Qs >1.5:1 according to most authors.[4] Some others have suggested a cut-off Qp:Qs of at least 2:1.[16] Our patient refused surgery; therefore, we had to manage her conservatively which was not without obstacles. She had a marginal blood pressure after starting bisoprolol and furosemide, so we could not titrate these medications up or start her on mineralocorticoid receptor antagonists (MRAs). This is considered mainly to relieve the right heart congestion and to help heart failure with preserved ejection fraction (HFpEF), as evidenced by an elevated NT-proBNP and near-normal LVEF.[17] In fact, digoxin had to be added to control the rate. Sodium-glucose co-transporter-2 (SGLT-2) inhibitors could have been another option, but unfortunately, it was not available for HFpEF in Sri Lanka at that time; therefore, it was not considered. As she had no evidence of primary PHT and had a clear cardiac cause for PHT, we did not opt for pulmonary arterial hypertension specific treatment, such as phosphodiesterase-5 (PDE-5) inhibitors or endothelin receptor antagonist. For her subclinical hyperthyroidism, we referred her to the endocrine team for the commencement of carbimazole, but the team suggested surveillance only. She was referred for nutritional assessment and further management, but as she was a vegetarian and could not afford the expensive nutritional supplements, she was managed very conservatively. She was later referred for cardiac rehabilitation exercise programme as well to relieve her symptoms.

## Lead author biography

Dr Thisara Samarawickrama is an interventional cardiologist and the head of the cardiology division in General Sir John Kotelawala Defence University Hospital in Sri Lanka. He is also a senior lecturer in the Faculty of Medicine of the same university. He pioneered the CTCA, dobutamine stress echocardiography (DSE), and TOE services in the hospital and currently works at the Royal Cornwall Hospital NHS Trust in the UK.

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**Consent:** Written informed consent was obtained from the patient's next of kin (daughter) in accordance with the Committee on Publication Ethics (COPE) guidelines.

**Conflict of interest :** None declared.

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## Data availability

The data underlying this article are available in the Dryad Digital Repository, at https://datadryad.org/stash/share/R3noD550cEYTJejYKVdYinA-yU8ICcNXn68Id2LSaPg.

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