

Multislice CT imaging of ruptured left sinus of Valsalva aneurysm with fistulous track between left sinus and right atrium.

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ABSTRACT

Sinus of valsalva aneurysm is a rare condition arising from any of the three aortic sinuses. Among them, an aneurysm arising from the left coronary sinus is the rarest. Most of these cases were earlier diagnosed using echocardiography and conventional angiography. But with the availability of advanced imaging modalities like 64 slice cardiac CT and MR modalities, this condition can be accurately assessed noninvasively. We report a case of ruptured aneurysm originating from the left coronary sinus with a long windsock type of fistulous track between the aneurysm and right atrium evaluated by 64 slice cardiac CT imaging. This was later confirmed perioperatively.

CASE REPORT

CASE REPORT

We describe a case of ruptured aneurysm arising from the left coronary sinus with a long windsock type of fistulous track connecting the left sinus with the right atrium producing aorto-cardiac shunting (left to right shunting). The case was evaluated by 64 slice cardiac CT (Computed tomography) imaging, which was later confirmed by perioperative findings.

A 11 year old male presented with history of chest pain, palpitations for 3 years and breathlessness for 3 weeks. On physical examination, a continuous murmur was heard in the left parasternal region. Chest radiogram showed mild cardiomegaly with right atrial and right ventricular configuration. Echocardiography performed in the parasternal short axis view showed aneurysmal dilatation of the left coronary sinus (Fig 1) and the site of rupture (Fig 2). Modified 4-chamber view showed a track communicating between the aneurysm and the right atrium (Fig 3 and 4).

Mild enlargement of the right atrium and right ventricle was also noted (Fig 4).

The patient was then evaluated by 64 slice cardiac CT, which demonstrated diffuse aneurysmal dilatation of the left sinus of Valsalva (Fig 5 and 6). A long windsock type of tubular fistulous track was seen extending between ascending aorta and left atrium with a grossly dilated segment proximal to its opening into the posterior wall of the right atrium (Fig 7 and 8). The main left coronary artery was seen arising from the tip of the aneurysm. The site of rupture was just posterior to the origin of the left main coronary artery (Fig 7). Volume rendered images clearly depicted the aneurysm and fistulous track in relation to the adjacent structures (Fig 9 and 10).

The patient underwent open heart surgery through median sternotomy. The site of rupture of the left sinus aneurysm was closed by placing a graft. The left coronary artery was reimplanted. The opening in the right atrium was closed by direct suturing.

DISCUSSION

The sinuses of Valsalva are three subtle dilatations of the aortic root between the aortic annulus and sinotubular ridge. Each sinus is associated with corresponding right, left and non coronary valve cusps. These sinuses allow enough space for the movement of aortic valve leaflets during systole without causing occlusion of ostia of the coronary arteries.

According to TakachTJ et al, sinus of Valsalva aneurysms are more common in males compared to females (ratio being 3:1) and prevalent in eastern and Asian countries [1].

They may be congenital or acquired. Congenital causes are due to weakness or deficiency of the elastic lamina as seen in Marfan's and Ehler-Danlos syndromes. They are commonly associated with cardiac anomalies like ventricular septal defects, bicuspid aortic valve, aortic regurgitation and coronary artery anomalies. Acquired causes include infective endocarditis, syphilis, tuberculosis, atherosclerosis and deceleration trauma.

According to Meier JH et al, 65-85% of sinus of Valsalva aneurysms originate from the right sinus, 10-30% from non coronary sinus and < 5% from the left sinus [2]. These aneurysms usually arise from the right coronary and non coronary sinuses and commonly rupture into the adjacent right atrium, right ventricle, left ventricle or into the interventricular septum. Aneurysms arising from the left coronary sinus are very rare. The rare left side aneurysms have a propensity for protrusion and rupture into the ipsilateral left atrium, left ventricle, pulmonary artery, myocardium or epicardium. When they rupture into the epicardium, they may compress the left coronary artery resulting in myocardial ischemia.

Asymptomatic aneurysms may present with a continuous murmur. They may be symptomatic when the aneurysms are large, producing pressure effects on the adjacent structures or due to rupture.

In case of rupture, they may present with chest pain, dyspnoea or cardiac failure depending upon the rapidity with which they rupture and the chamber with which they communicate [3]. Usually, the aneurysms rupture into the ipsilateral cardiac chamber resulting in aorto-cardiac shunting [4]. But in our case, the aneurysm had ruptured into and was communicating with the right atrium through a long windsock like fistulous track. The left coronary artery was seen arising from the tip of the aneurysm and the point of rupture was just posterior to the left coronary ostium.

Earlier, sinus of Valsalva aneurysms were diagnosed using colour Doppler echocardiography followed by conventional angiogram. But with the advent of new imaging modalities like cardiac CT and MRI (Magnetic resonance imaging), the condition can be non-invasively diagnosed. According to Aliye OB et al, ECG-gated (Electrocardiogram) contrast enhanced multislice CT provides much better spatial resolution of cardiac structures compared to other imaging methods and provides detailed anatomic depiction of Valsalva sinus aneurysms and the surrounding cardiac structures [5]. The

surgeon can also be provided with high resolution pre-operative 3D representations which are invaluable in planning the surgical approach and provide important anatomical information [as in our case where the pre operative information from the 3D images helped the surgeon to preserve the left coronary ostium]. However, the disadvantage of CT is the use of ionizing radiation (7.6-31.8 mSv in retrospective ECG gating and 2.1-9.2mSv in prospective ECG gating) [6].

The advantages of performing MR imaging in the setting of a known or suspected Valsalva sinus aneurysm include the ability to evaluate the left ventricular hemodynamic pattern, identify aortic regurgitation and quantify aorto-cardiac shunt or fistulous blood flow. But CT is less time consuming and the preferred investigation compared to MRI in case of acute setting of aneurysmal rupture. Conventional angiography is the gold standard and can be used for both diagnostic and therapeutic purposes.

The differential diagnoses for ruptured sinus of Valsalva aneurysm include aortic root / ascending aortic aneurysms, coronary arterio-venous fistulas and prolapsed aortic cusps. Sinus of Valsalva aneurysms occur between the aortic annulus and sinotubular ridge. Prolapsed aortic cusps occur below the aortic annulus whereas aortic root aneurysms occur above the sinotubular ridge. This differentiation can be made out either by using echocardiography or cross sectional imaging (CT or MRI). Coronary arterio-venous fistula can be diagnosed using cardiac CT, since it depicts coronary artery anatomy better. Conventional angiography can be used to differentiate the above mentioned conditions and for therapeutic purposes.

These aneurysms are closed with a pericardial or polyester patch or with sutures.

TEACHING POINT

Cardiac CT plays an important role in the diagnosis of sinus of Valsalva aneurysm and its complications like rupture. It gives important anatomical information which will help in planning the surgical approach.

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FIGURES

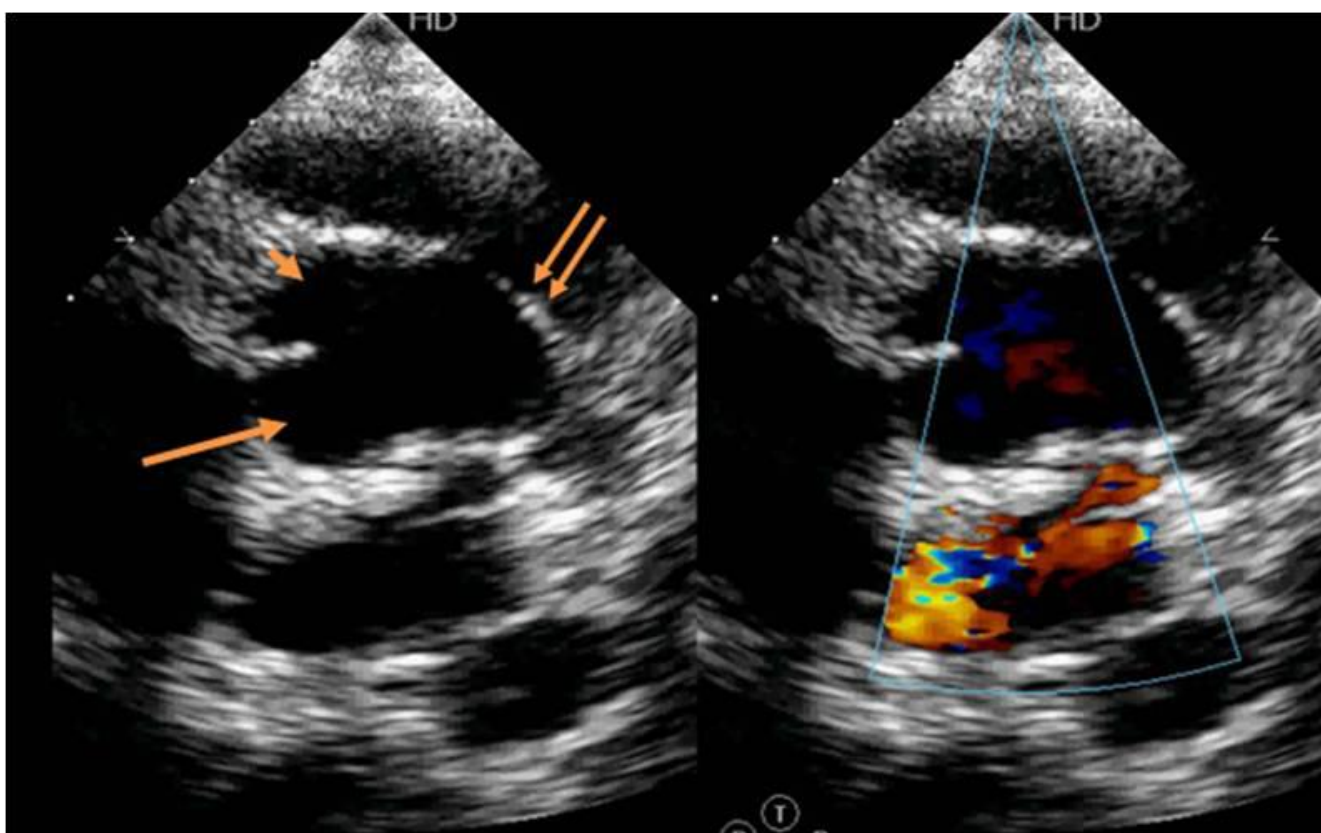


Figure 1: 11year old male with ruptured aneurysm arising from the left sinus of Valsalva. Parasternal short axis echocardiographic view (dual mode with both gray and colour Doppler imaging) shows aneurysmal dilatation of the left aortic sinus (double arrows). Also note the normal right aortic sinus (short single arrow) and non coronary aortic sinus (long single arrow). (Philips Envisor ultrasound machine using S4 probe with 2-4 MHz frequency)

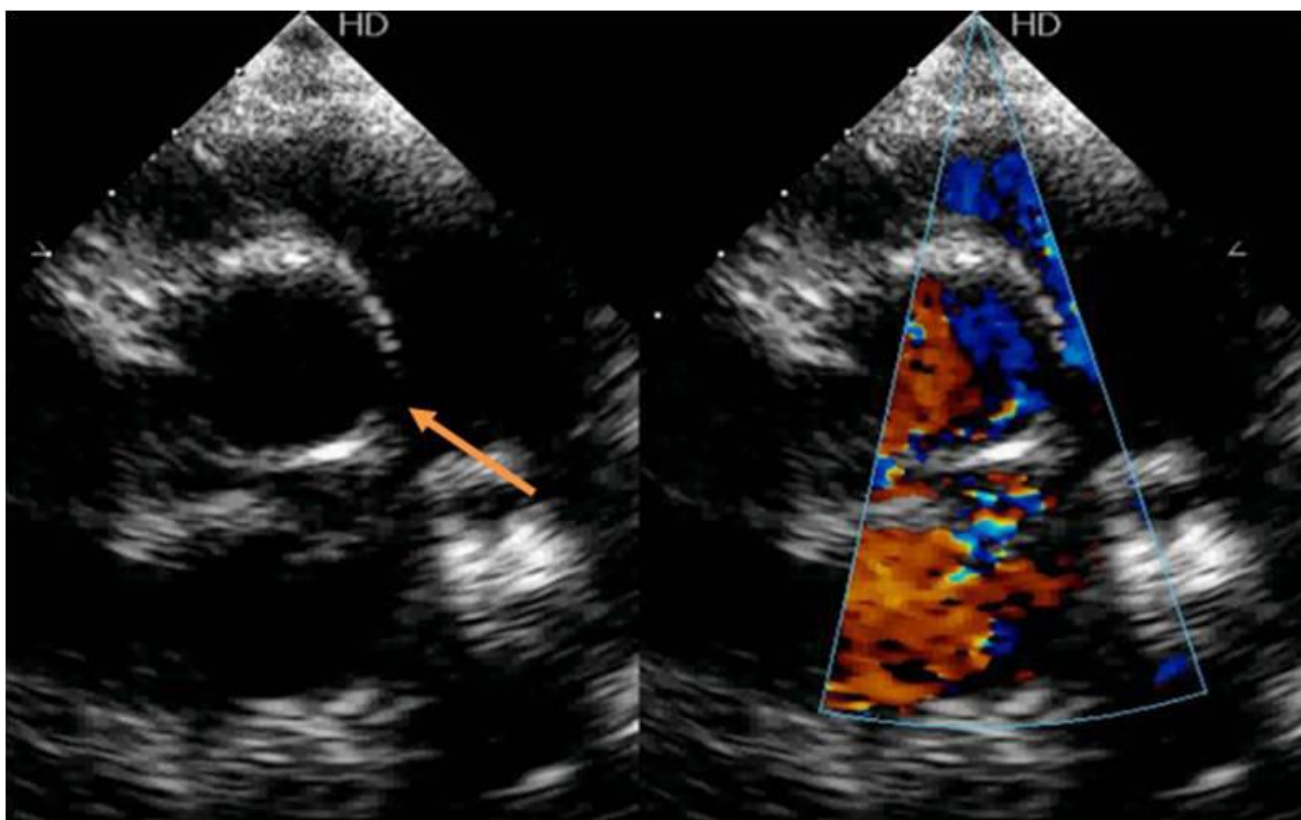


Figure 2: 11year old male with ruptured aneurysm arising from left sinus of Valsalva. Parasternal short axis echocardiographic view (dual mode with both gray and colour Doppler imaging) shows aneurysmal dilatation of the left aortic sinus with site of rupture (single arrow). (Philips Envisor ultrasound machine using S4 probe with 2-4 MHz frequency)

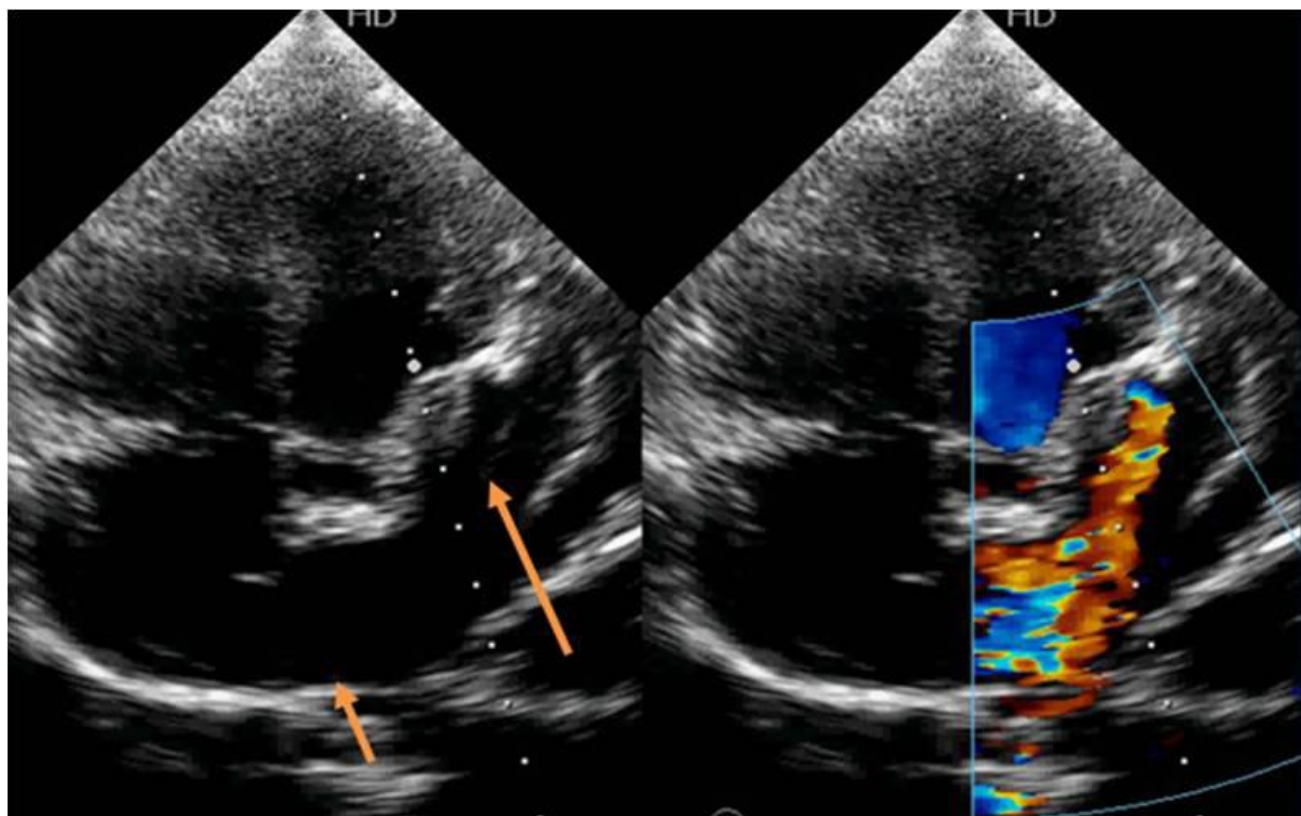


Figure 3: 11year old male with ruptured aneurysm arising from the left sinus of Valsalva. Modified 4-chamber echocardiographic view (dual mode with both gray and colour Doppler imaging) shows fistulous track (single long arrow) arising from the left aortic sinus with bulbous dilatation distally (single short arrow). (Philips Envisor ultrasound machine using S4 probe with 2-4 MHz frequency)

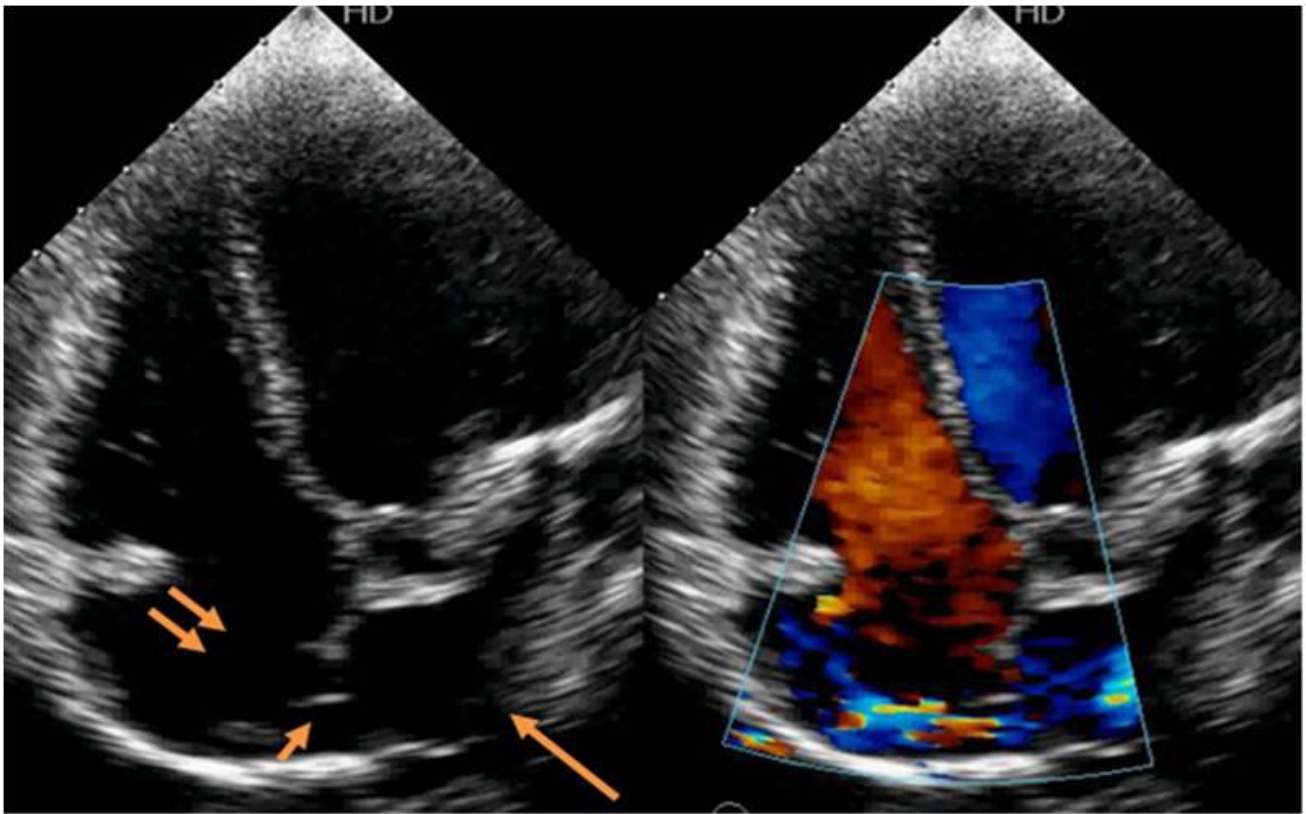


Figure 4: 11year old male with ruptured aneurysm arising from the left sinus of Valsalva. Modified 4-chamber echocardiographic view (dual mode with both gray and colour Doppler imaging) shows distal bulbous dilatation of the fistulous track (single long arrow). The track is seen opening (single short arrow) into the right atrium (double short arrows). (Philips Envisor ultrasound machine using S4 probe with 2-4 MHz frequency)



Figure 5 (left): 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. Axial MPR image shows diffuse aneurysmal dilatation of left coronary sinus (double arrows). Note origin of left main coronary artery (long single arrow) from the left sinus just anterior to the site of rupture. Also note normal right coronary artery arising from right aortic sinus (short single arrow). (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml)

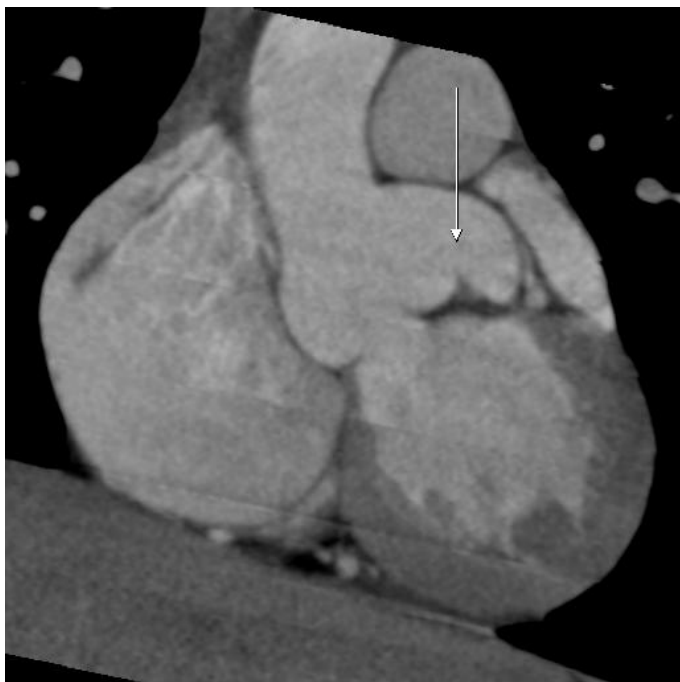


Figure 6: 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. Oblique coronal MPR image shows aneurysmal dilatation of left coronary sinus (arrow). (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml)



Figure 8: 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. Curved MPR image shows ruptured left coronary sinus aneurysm (short single arrow) with long fistulous track (short double arrows) connecting to the right atrium. Note the segment of gross dilatation (long double arrows) of fistulous track just before its opening in the right atrium (long single arrow). (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml)



Figure 7: 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. Oblique axial MPR image shows ruptured aneurysm arising from left sinus(double arrows) with long "wind sock shaped" fistulous track(short single arrow) extending between ascending aorta anteriorly and the left atrium posteriorly. Note segment of gross dilatation of the track (long single arrow) just proximal to opening in the posterior wall of right atrium. (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml)



Figure 9: 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. VRT image clearly depicts the left sinus aneurysm (long arrow) and the whole track (short arrow) from the site of rupture to the right atrium. (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml)

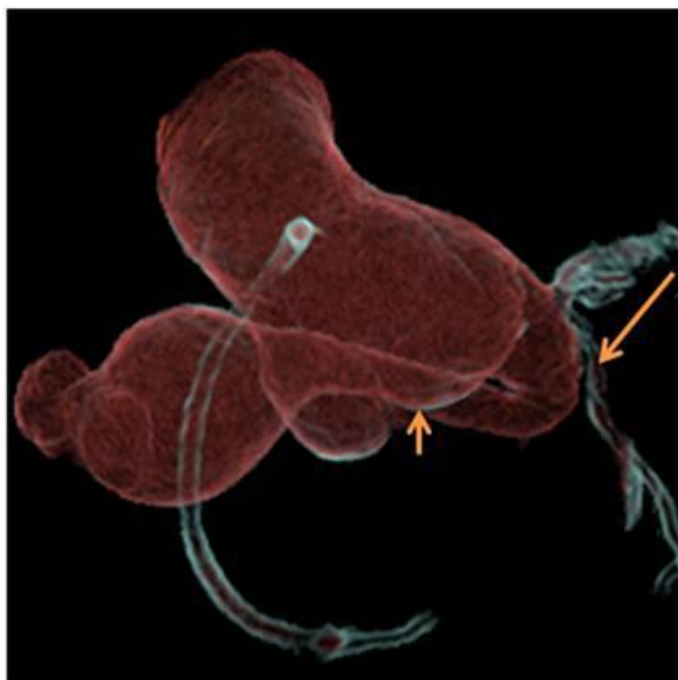


Figure 10 (left): 11 year old male with ruptured aneurysm arising from the left sinus of Valsalva. VRT image shows the left sinus aneurysm and the fistulous track (short arrow) extending from the site of rupture to the right atrium. Note the origin of left main coronary artery from the left sinus (long arrow). (GE Light speed VCT 64slice CT scanner. Protocol: 450 mAs, 120 kV, 0.625 mm slice thickness, 30ml of Omnipaque 350mg I/ml).

Features	Sinus of Valsalva aneurysm	Aortic root & ascending aorta aneurysm	Coronary arterio-venous fistula	Prolapsed aortic cusp
Location	Above the aortic annulus and below sinotubular ridge	Above the sinotubular ridge	Between the coronary arteries and heart chamber / systemic or pulmonary circulation	Below the aortic valve annulus
Etiology	Congenital or acquired	Acquired (traumatic / infective) or Congenital	Congenital or iatrogenic / traumatic	Bicuspid aortic valve, infective endocarditis, aortic dissection, trauma
Imaging				
ECHO	Depicts site, size, and the fistulous communication with the chamber	Depicts the site above the sinotubular junction, size and presence of thrombus	Depicts the vessel involved, volume flow within dilated vessel, and drainage of the fistula.	Eccentric aortic insufficiency jet in the opposite direction, cusp below the level of the aortic annulus during diastole, diminished length of aortic leaflet coaptation
CT / MRI	Helpful in clear depiction and confirmation of ECHO findings (CT better than MRI)	Clear demonstration of the site, size of aneurysm, presence of thrombus, leak and rule out aortic dissection	MRI is good for demonstration of proximal coronary artery anomalies. CT provides excellent distal coronary artery and side branch anatomy	MRI may be helpful for assessing hemodynamic disturbance. CT is not very useful
Conventional angiogram	Gold standard. Both diagnostic and therapeutic	Gold standard. Both diagnostic and therapeutic	Gold standard. Both diagnostic and therapeutic	Gold standard. Both diagnostic and therapeutic
Complications	Rupture, Aortic / Mitral regurgitation, compression of adjacent vital structures	Aortic / Mitral regurgitation, compression of adjacent vital structures	Myocardial ischemia, papillary muscle rupture, heart failure	Aortic / mitral regurgitation

Table 1: Differential diagnoses of ruptured sinus of valsalva aneurysm.

Etiology	Congenital (Marfan's and Ehler-Danlos syndrome) or Acquired (infective endocarditis, syphilis, tuberculosis, atherosclerosis and deceleration trauma)
Incidence	Rare (Accounts for 5% of all sinus of valsalva aneurysms)
Gender ratio	Male > Female (About 3:1)
Age predilection	Can present at any age depending on size of aneurysm and complications
Risk factors	Weakness or deficiency of elastic lamina (as in Marfan's and Ehler-Danlos syndrome) and trauma
Treatment	Surgical therapy in the form of closure of aneurysm with a pericardial or polyester patch or with sutures
Prognosis	Fairly good in case of successful surgery
Imaging Findings	ECHO and cardiac CT are the main modalities used. Aneurysmal dilatation of the sinus of Valsalva will be seen projecting beyond the confines of the aortic root depending upon the size. In case of rupture, the site of rupture, the course of the fistulous track and the site of opening of fistula (type of aorto-cardiac shunting) can be localized. All these features are best depicted by cardiac CT and help the surgeons in planning surgical approach.

Table 2: Summary table of ruptured left sinus of valsalva aneurysm.

ABBREVIATIONS

AI - Aortic Insufficiency
 CT - Computed Tomography
 ECG - Electrocardiogram
 MRI - Magnetic Resonance Imaging

KEYWORDS

Sinus of Valsalva; Aneurysm; Rupture; Fistula; CT imaging

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