

## Letter to the Editor

## Giant Unruptured Left Sinus of Valsalva Aneurysm as an Unusual Cause of Ischemic Heart Failure

STYLIANOS KARVOUNARIS<sup>1\*</sup>, GEORGE MICHAS<sup>1\*</sup>, VASILIS KARAMPETOS<sup>1</sup>, MARINOS SOTERIOU<sup>2</sup>, STEFANOS FOUSSAS<sup>3</sup>, PETROS MAVROMMATIS<sup>1</sup>

<sup>1</sup>Department of Cardiology, Paphos General Hospital, Paphos, <sup>2</sup>Department of Cardiothoracic Surgery, American Medical Center, Nicosia, Cyprus; <sup>3</sup>Department of Cardiology, Tzaneio General Hospital of Piraeus, Piraeus, Greece

\*These authors contributed equally to this paper.

Key words: **Left sinus of Valsalva aneurysm, ischaemic heart failure, Bentall procedure.**

Manuscript received:  
May 29, 2014;  
Accepted:  
November 26, 2014.

Address:  
Petros Mavrommatis

Department of  
Cardiology  
Paphos General Hospital  
Paphos, Cyprus  
[p.mavrommatis@gmail.com](mailto:p.mavrommatis@gmail.com)

**T**he Valsalva sinuses are three small dilatations in the aortic wall immediately above each cusp of the aortic valve. Sinus of Valsalva aneurysms (SVAs) are rare anomalies, usually caused by the congenital absence of muscular and elastic tissue in the aortic wall behind the sinus of Valsalva.<sup>1</sup> Acquired aneurysms are caused by conditions affecting the aortic wall, such as infections (syphilis, endocarditis, and tuberculosis), trauma, or connective tissue disorders.<sup>2</sup> The incidence of SVAs ranges from 0.1% to 3.5% of all congenital heart defects (higher in Asian populations) and has a reported prevalence of 0.09% in an autopsy series.<sup>2</sup> Of all SVAs, 65-85% stem from the right sinus of Valsalva and 10-30% from a non-coronary sinus, while SVAs originating from the left sinus (<5%) are extremely rare.<sup>3</sup> Although an unruptured SVA is mostly asymptomatic, fatal complications can occur, such as myocardial ischaemia due to impairment of coronary flow. This report describes the management of a female patient who presented with cardiogenic shock due to overstretching of the left main coronary stem by a large left SVA.

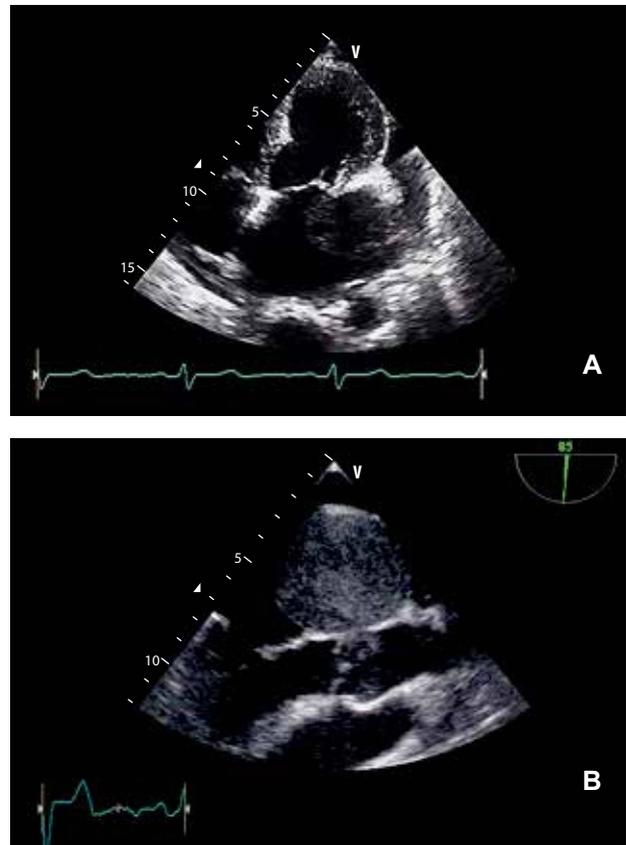
A healthy 63-year-old female presented with exertional dyspnoea of four weeks' duration, with acute worsening of her symptoms 48 hours prior to hospital admission. The patient was admitted to the coronary care unit with congestive heart failure and cardiogenic shock. The physical examination revealed a blood pressure of 70/50 mmHg, elevated jugular venous pressure, and a 3/6 systolic murmur. A 12-lead electrocardiogram showed sinus tachycardia with left bundle branch block. The chest X-ray showed cardiomegaly, bilateral pleural effusion, and pulmonary venous congestion. Initial laboratory findings showed increased total creatinine phosphokinase of 256 IU/L (26-145 IU/L), an MB fraction of 53.4 IU/L (0.0-25.0 IU/L), and troponin T levels of 0.51 ng/mL.

The transthoracic echocardiogram (TTE) showed a dilated left ventricle with an end-diastolic diameter of 59 mm, and severely impaired left ventricular systolic function (ejection fraction 30%) with hypo/akinetic septum and apex. Right ventricular systolic function was within normal limits. The left atrium was dilated. Surprisingly, a large pulsatile mass was seen protruding into the left

atrium, distorting the flow across the mitral valve (Figure 1A), but not suggestive of a tumour, i.e. myxoma or thrombus. Moderate mitral and tricuspid regurgitation were present. The right ventricular systolic pressure was estimated at  $\sim 49$  mmHg. No aortic regurgitation was observed. A small pericardial effusion was also noted. Urgent transoesophageal echocardiography (TOE) was performed at the bedside to better clarify the nature of this mass. In fact, this giant mass ( $59 \times 92$  mm) proved to be a typical unruptured left SVA that was partially filled with thrombotic material (Figure 1B). No intracardiac shunting or aortic regurgitation was detected. The pulmonary artery was dilated and constricted by the aneurysm. After infusion of contrast material, more precise definition of the intact aneurysmal sac was demonstrated. The infused contrast was clearly seen entering and filling the huge aneurysm, which protruded into and occupied nearly the whole left atrium.

The patient deteriorated soon after, despite inotropic support (ventricular tachycardia successfully cardioverted to sinus rhythm) and it was decided, after consultation with the surgical team, to transfer the patient to a tertiary hospital for urgent operation. Perioperative findings confirmed the presence of the large left SVA, which extended laterally to the main pulmonary artery and also protruded into the left atrium, almost completely obstructing the mitral valve orifice. The left main coronary ostium was part of the superior rim of the opening of the aneurysm, so that in the case of a patch repair, the left main coronary artery could be compromised by the suture line. Given these findings, it was decided to perform a Bentall procedure (aortic valve replacement, aortic root replacement with exclusion of the SVA, re-implantation of the coronaries; Figure 2). Unfortunately, despite vigorous support measures (including intra-aortic balloon insertion) during an overnight multi-hour operation conducted by the cardiothoracic team, the patient expired.

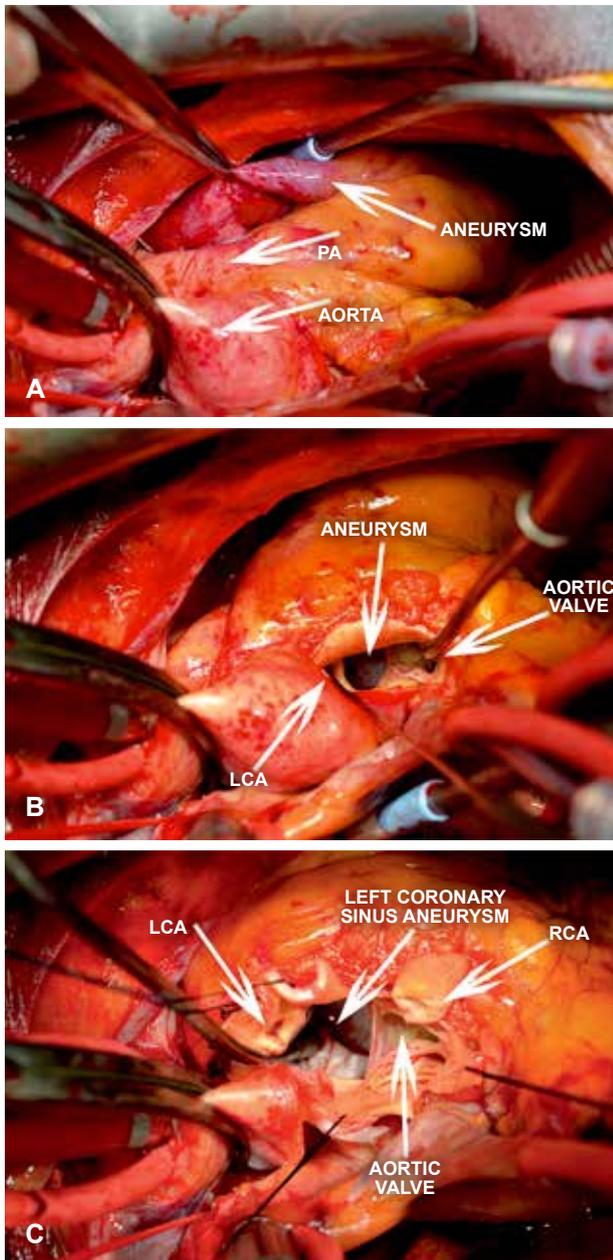
Left SVAs are extremely rare and are usually detected prior to their rupture.<sup>2</sup> Rupture of a left SVA typically occurs into the left ventricle or left atrium.<sup>4</sup> Unruptured left SVAs are usually asymptomatic. When they do become symptomatic, they often present as an acute coronary syndrome or even cardiogenic shock, by involving the left main stem, the left anterior descending, and/or left circumflex artery, either by direct compression or by spontaneous dissec-



**Figure 1.** A. Transthoracic echocardiogram showing a pulsatile mass (M) almost filling the dilated left atrium (LA). B. Transoesophageal echocardiography ( $85^\circ$ ) showing a typical unruptured left sinus of Valsalva aneurysm (SVA) ( $59 \times 92$  mm), partially filled with thrombotic material, protruding into and occupying almost the whole left atrium (LA). RA – right atrium; LV – left ventricle; Ao – aorta; RVOT – right ventricular outflow tract.

tion.<sup>5,6</sup> They can also present as aortic regurgitation, complete heart block, and resistant ventricular tachycardia.<sup>7</sup>

Coronary insufficiency due to SVA is an uncommon manifestation and can be a sign of poor prognosis.<sup>2,8</sup> There is more risk of myocardial ischaemia with a left SVA than with right or non-coronary SVAs, possibly because a left SVA can protrude between the left atrium and pulmonary trunk and compress the trunk or branches of the left coronary artery.<sup>2,8</sup> Aneurysmal dilatation can occur very rapidly, and early diagnosis and surgical treatment are mandatory.<sup>8</sup> The type of surgery depends on the anatomical constraints and ranges from a simple patch closing the aneurysmal orifice to complete reconstruction of the aortic root (the latter has a worse prognosis).<sup>8</sup> Unfortunately, in



**Figure 2.** A. The giant aneurysm is visualised just after chest opening. B. The protruding aneurysm in the left atrium. C. Anatomical preparation before Bentall procedure. PA – pulmonary artery; RCA – right coronary artery; LCA – left coronary artery

our patient the left main stem was part of the aneurysm and had been overstretched, resulting in severe obstruction of coronary flow. This severe compromise of coronary flow was the cause of the ischaemic heart failure and the ongoing myocardial infarction, which led to the patient's unfavourable outcome.

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