



## Sinus of Valsalva Aneurysm – A Possible Association with Infection of Lymphatic Mode of Spread: Report of 15 Cases

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### Abstract

**Objective:** Sinus of Valsalva Aneurysm (SoVA) is a rare cardiac defect. Both congenital and acquired aetiology of SoVA have been proposed. It was the aim of this study to describe the clinical presentation of SoVA and to investigate the possible association with infectious disease in these patients.

**Methods:** Fifteen patients with SoVA were identified. Patient case notes were retrieved and reviewed. Patients were 7 females and 8 males with a mean age of 31 years (range 10-56). Seven patients were South African Bantu, six were Chinese and two were Caucasians.

**Results:** Chronic chest infections not responding to routine antibiotics treatment were diagnosed in seven patients, tuberculosis and syphilis were each diagnosed in two patients and one patient had pharyngitis. Three patients had no significant medical history of infection. Intrathoracic lymphadenopathy was observed in 10 of 13 patients with recorded findings.

**Conclusion:** Sinus of Valsalva Aneurysm seems to have strong association with infections of lymphatic mode of spread. These infections have developed prior diagnosis of SoVA was made in majority of patients.

### Introduction

Sinus of Valsalva Aneurysm (SoVA) is a rare cardiac defect. The first report of SoVA is attributed to Hope who reported a case of ruptured SoVA in 1839 [1]. A comprehensive review by Nowicki in 1977 identified only 175 reported cases [2]. Unruptured SoVA is usually asymptomatic although they can produce aortic valve incompetence, coronary artery compression or fistulae, conduction disturbances or pulmonary outflow obstruction due to compression of adjacent structures [3]. Rupture of SoVA is usually intracardiac and leads to development of congestive cardiac failure and arrhythmias. Both congenital and acquired aetiology of SoVA have been proposed. Deficiency of elastic tissue at the site of congenital SoVA has been histologically demonstrated [3]. Acquired aneurysms have been described secondary to arteriosclerosis, syphilis, infective endocarditis, cystic median necrosis and trauma [3].

### Objective

It was the aim of this study to describe the clinical presentation of SoVA and to investigate the possible association with infectious disease in these patients.

### Patients and Method

15 patients with Sinus of Valsalva Aneurysm were identified. Nine patients were treated at Groote Schuur Hospital and the Red Cross War Memorial Children's Hospital, (Cape Town, South Africa) and six at National Heart Centre (Singapore, Figure 1).

Patient case notes were retrieved and reviewed. Patient demographic features (gender, race, age, occupation and region), clinical presentation including evidence of infectious disease, results of investigations (chest X-Rays, blood tests) and surgical or autopsy findings were studied. Syphilis was diagnosed where TPHA and VDRL tests were both positive. Tuberculous lymphadenitis was diagnosed from tissue samples. Lymphadenopathy was detected by means of clinical appearance (palpation, hoarseness), chest X-ray criteria (stretching or compression of the trachea, unilateral elevation of the diaphragm, atelectasis, [4] Figure 2). Cardiac lesions were diagnosed on echocardiography and/or angiography.

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**Table 1:** Clinical presentation.

Patient	Presentation (months)	Associated infection	Lymphadenopathy	CCF	Associate lesion
1	12	TB lymphadenitis	Intrathoracic	yes	AI, MI
2	12	TB lymphadenitis	Intrathoracic, Cervical	yes	AI, MI
3	6	CCI	None	yes	AI
4	12	Syphilis	Intrathoracic	yes	AI
5	7	CCI	Intrathoracic	yes	AI
6	18	Syphilis	None	yes	AI
7	24	CCI	Intrathoracic	yes	AI, MI
8	60	CCI	Intrathoracic	yes	AI, MI
9	24	?	Not reported	no	None
10	120	CCI	None	yes	MI, AI
11	1	CCI	Intrathoracic	yes	None
12	2	?	Intrathoracic	yes	None
13	84	Pharyngitis	Intrathoracic	yes	AI, TI
14	180	?	Not reported	no	MI, TI
15	2	CCI	Intrathoracic	yes	MI, TI

TB: Tuberculosis; CCI: Chronic Chest Infection; AI: Aortic Valve Incompetence; MI: Mitral Valve Incompetence; CCF: Congestive Cardiac Failure; TI: Tricuspid Valve Incompetence

**Table 2:** Surgery- findings, outcome.

Patient	Site of aneurysm	Fistula connection	Associated lesion	Operation	'Windsock' aneurysm	Outcome
1	NCS, LCS	LV	Bantu Aneurysm	Repair	Bantu Aneurysm	Good
2	NCS, LCS	RA, LV	Bantu Aneurysm	Repair		Good
3	NCS, RCS	RA		Repair		Good
4	NCS	RA, LV		Repair, AVR		Good
5	RCS	RA, RV	VSD	Repair		Good
6	NCS, LCS	LV		Repair, AVR		Good
7	NCS	LV	Bantu Aneurysm	Repair, AVR		Good
8	NCS, LCS	LV	Bantu Aneurysm	Repair		Good
9	RCS	RA	RCA-RA Fistula	Fistulae ligation		Good
10	RCS	RV	VSD	Repair	SoVA	Good
11	NCS	RV	VSD	Repair	SoVA	Good
12	RCS	RA		Repair	SoVA	Good
13	RCS	RV	VSD	Repair	SoVA	Good
14	NCS	RA		Repair	SoVA	Good
15	RCS	RA		awaiting	SoVA	

NCS: Non-Coronary Sinus; LCS: Left Coronary Sinus; RCS: Right Coronary Sinus; LV: Left Ventricle; RA: Right Atrium; RV: Right Ventricle; LV: Left Ventricle; VSD: Ventricular Septal Defect; RCA-LA fistula: Right Coronary Artery- Left Atrial Fistula; AVR: Aortic Valve Replacement; SoVA: Sinus of Valsalva Aneurysm

## Results

### Demographics

Between 1967 and 1996, twelve cases of Sinus of Valsalva were identified. Patients were four females and 8 males with a mean age of 26.9 years (range 10-48). 4 patients were of Negroid race, 5 were Caucasians and 3 were Coloured. All patients were coming from high TB prevalence areas (Table 1).

### Clinical presentation

Clinical findings are summarised in Table 2. Two patients clinical data (9,12 Table 2) were incomplete. Mean length of history of presentation was 21.8 months (range 3-60) at operation. Two patients had tuberculous lymphadenitis and TB could be suspected in another

8 patients on the basis of reported clinical findings. Two of the patients had diagnosed syphilis. Intrathoracic lymphadenopathy was observed in 8 patients, two of whom also had cervical lymphadenopathy. Two had no record of lymphadenopathy in patient case notes, and in another two patients no lymphadenopathy was observed.

Sinus of Valsalva Aneurysm was associated with aortic valve incompetence in 8 patients, with 4 patients also having mitral valve incompetence. One patient had also coarctation of the aorta. Ten patients had signs and symptoms of congestive cardiac failure. ECG demonstrated conduction delay abnormalities in ten patients and six of the patients had Q waves on ECG despite unobstructed coronary flow on angiography (T Figure 3).



**Figure 1:** Chest X-Ray. (Left Phrenic nerve palsy probably due to mediastinal lymphadenopathy).



**Figure 2:** "Windsock" Sinus of Valsalva Aneurysm held by forceps.

### Surgical findings

Eleven patients underwent surgical repair of aneurysm. In three cases „windsock” form of aneurysm was reported among SoVA patients (Figure 1). One of the patients had aneurysm left untouched (patient 9) due to high risk of RCA damage during closure of the RCA-RA cameral fistula. Three patients had aortic valve replacement. Four patients also had repair of associated Annular Subvalvular Left Ventricular Aneurysm (ASLVA). The Sinus of Valsalva Aneurysm was cited in the non coronary sinus in 8 cases, in the right coronary sinus in 6 cases and in the left coronary sinus in 4 cases. In 6 cases there were multiple aneurysms. In one case it was unruptured aneurysm of "windsock" type communicating with right atrium via cameral RCA-RA fistula. Fistulae communicating between Sinus of Valsalva Aneurysm and the heart chambers were observed in all cases, with multiple fistulae in three patients. In 6 cases this was to the left ventricle, and in 7 cases to the right atrium, and in 2 cases to the right ventricle.

### Histological findings

Aneurysmal wall was taken for histopathology evaluation in five and aortic valve in two SoVA cases. Specimens from left atrial appendage and right atrial appendage were each taken in one case. Three patients had no tissue taken for histopathology. The wall of Sinus of Valsalva Aneurysm was composed of collagenous connective tissue lined by endothelium in five specimens. Hyalinisation was the finding in two SoVA wall specimens. Inflammatory infiltrate was observed in three and calcification was observed in two specimens. Besides features mentioned above, two of the wall specimens were consisted of three basic vessel's layers, namely intima - endothelium, media - muscle fibers and adventitia - epithelium (Figure 2). The pathological changes mostly affect the media layer, where the muscle fibres were replaced by fibrous tissue. Tissue of left atrial appendage showed vessels endothelial thickening and fibrosis in one case, and in

one case vessels' thickening was observed within tissue of right atrial appendage. Aortic valve was fibrosed in two cases and hyalinisation was observed in one.

### Complications of surgery:

- Conduction disturbances – 14
- Q waves/ST abn – 12
- Chylopericardium – 1
- Chylothorax – 1

### Discussion

The first description of aneurysmal enlargement of Sinus of Valsalva with intracardiac rupture has been attributed to Hope (1839). Thurnam (1840) published a series of six cases. Abbot in 1919 proposed and Edwards in 1957 established concept of congenital SoVA, who histologically demonstrated the deficiency of elastic tissue at the sight of congenital SoVA [1]. A comprehensive review by Nowicki in 1977 only identified 175 reported cases [2].

The high incidence of infections, developed prior diagnosis of SoVA was made, in patients from this study with the associated lymphadenopathy supports an infective origin of SoVA and lymphatic spread as the mode of spread of these infections. Syphilis as a cause of SoVA was initially emphasised by Smith in 1914 [3], reported by others [1,5,11], it has been diagnosed in two patients from this study. Tuberculous lymphadenitis has been diagnosed in two patients from this study, finding not reported in the literature previously. Sometimes infective endocarditis can associate with SoVA [6,7]. Mediastinal lymphadenopathy seems to play important role in the pathology of SoVA. It has been diagnosed in ten of thirteen patients with recorded finding, including all four patients with associated Bantu aneurysm. The similarity in the aetiology of Sinus of Valsalva Aneurysm and Bantu Aneurysm was first noted by Chesler [8]. Association of SoVA and Bantu Aneurysm was observed in four cases from this study and has been reported by others [9,10]. The similar morphology of some SoVA and Bantu Aneurysm, „finger-like” or „windsock” type of morphology (Figure 3), and the similar clinical presentation of these patients' further supports Chesler's concept of the same aetiology of these two rare cardiac lesions. Lymphatic spread of infections with associated post inflammatory destruction of mediastinal lymph nodes in both - SoVA and Bantu aneurysm, seems to play significant role in the development of these rare cardiac aneurysms.

### Conclusion

- Sinus of Valsalva Aneurysm has strong association with infections of lymphatic mode of spread.
- Sinus of Valsalva Aneurysm can associate with Bantu Aneurysm and both may have similar infective ethiology.

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