Ruptured aneurysms of the sinus of valsalva

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Summary
Aneurysms of the sinus of Valsalva are uncommon disorders and are usually congenital in origin. When these aneurysms rupture into an intracardiac chamber, they may be silent initially but later give rise to progressive heart failure due to left to right shunting and aortic regurgitation. The mortality and morbidity in untreated cases is high. We report 13 patients with ruptured aneurysms of the sinus of Valsalva who underwent surgical repair. There were seven males and six females with a mean age of 24.5 years. Three patients were asymptomatic and five were in congestive cardiac failure. The majority of patients (61.5%) had insidious onset of symptoms, only 2 cases presenting acutely. The connection was between the right aortic sinus and the right ventricle in 11 cases and the non coronary sinus and the right ventricle in 2 cases. Associated cardiac anomalies included a ventricular septal defect (8 patients) and aortic regurgitation (6 patients). There was 1 post operative death and 1 patient required re-operation three months later for a recurrence of the fistula. All 6 patients with aortic regurgitation required aortic valve replacement. All patients remained well and asymptomatic during follow up ranging from 2 to 19 years.

Key words: Aneurysms of the sinus of Valsalva, Congestive cardiac failure

Introduction
Thutnam[1], in 1840, described the first case of an aneurysm of the sinus of Valsalva which had ruptured into the right ventricle. This condition is an uncommon entity accounting for only 0.14% to 0.43%[2] of all procedures requiring cardiopulmonary bypass. The incidence has been reported to be higher among the Orientals compared to the Caucasians. [3]

Materials and Results
Between March 1969 and April 1990, 13 cases of ruptured aneurysms of the sinus of Valsalva (RASV) were seen at the University Hospital, Kuala Lumpur. The hospital records of these patients were retrospectively analysed.

Our series is similar to those of others in showing a slight male preponderance (53.4%) and a peak occurrence in the third decade of life (mean age 24.5 years).[3] (Table 1)

Dyspnoea (9 patients), chest pain (4 patients) and palpitations (1 patient) were the presenting symptoms. The functionsl class of the patients at the time of diagnosis varied widely. Five patients were in congestive cardiac failure. Only 2 of our patients presented with acute heart failure associated with sudden cramping chest pain. Both these patients had been noted to have asymptomatic cardiac murmurs when young and in both, infective endocarditis was excluded as a cause of the acute cardiac decompensation. The remaining 8 symptomatic patients had progressively deteriorating effort tolerance over periods varying from 1 month to 5 years. Three patients remained asymptomatic, the heart murmur being an incidental finding.

Table 1
Clinical findings and outcome in surgically corrected cases of RASV, UHKL (1969 – 1990)

No.	Age / Sex	Fistula	Co-existent cardiac defects	Functional Class (Pre-op)	Outcome
1[a]	27/F	R – RV	–	I	Alive Residual AI
2[a]	20/M	N – RV	–	I	Alive Residual AI
3[a]	19/F	R – RV	VSD Infundibular PS	I	Alive
4	22/M	N – RV	–	II	Alive Required re-op 3 months later
5	36/M	R – RV	–	IV	Alive Residual AI
6	24/F	R – RV	VSD	IV, acute onset	Alive
7	20/M	R – RV	VSD	IV	Alive Residual AI
8[b]	23/M	R – RV	VSD; AI	IV	Alive
9[b]	37/F	R – RV	AI	II	Alive
10[b]	18/M	R – RV	VSD; AI	II, acute onset	Alive Residual
11[b]	31/M	R – RV	VSD; AI	II	Alive
12[b]	22/F	R – RV	VSD; AI Infundibular PS	II	Alive
13[b]	20/F	R – RV	VSD; AI Infundibular PS	II	Succumbed

Key: [a]pts 1 – 3 were asymptomatic
[b]pts 8 – 13 required aortic valve replacement

Abbreviations:
(R: right coronary sinus; RV: right ventricule; VSD: ventricular septal defect; AI: aortic incompetence; PS: pulmonary stenosis)

The most common physical findings were a widened pulse pressure (13 patients), cardiomegaly (12 patients) and a continuous murmur at the left sternal edge (13 patients). The murmur was loudest in the third or fourth left intercostal spaces.

The right aortic sinus was the commonest site of the aneurysm (11 cases) and in all cases, the site of rupture was into the right ventricle. In the 2 cases involving the non coronary sinus, the fistulous tract also opened into the right ventricle. Isolated RASV was present in only 4 cases. Co-existent cardiac anomalies included a ventricular septal defect (8 cases), aortic regurgitation (6 patients) and right ventricular outflow tract obstruction due to muscle bands (3 patients).

The electrocardiogram (ECG) showed left ventricular hypertrophy with a normal electric axis in 10 cases and the heart size was increased on the chest X-ray in 12 patients. The aneurysm and the site of rupture were correctly diagnosed by two-dimensional (2-D) echocardiogram in 6 of 8 patients studied by this method. Figure 1 is a 2-D echocardiographic picture of a patient with a fistulous connection between the right aortic sinus of Valsalva and the right ventricle.

All patients underwent cardiac catheterisation and the results are summarised in Table 2. There was an increase in oxygen saturation in the right ventricle in all patients. The mean Qp/Qs ratio for the group was 3.17. The mean pulmonary arterial pressure was 28.6mmHg. Three patients had infundibular stenosis of the right ventricular outflow tract due to muscle bands with gradients of 98, 64 and 25 mmHg respectively. The fistula was well visualised by biplane supra-aortic angiography. (Figure 2)

Table 2
Hemodynamic data in surgically corrected RASV, UHKL (1969 – 1990)

No.	Pressure Measurements S/D in mm Hg)					Qp/Qs Ratio
	*RA	RV	PA	LV	Aorta	
1	5	40/8	27/18	118/0	120/70	1.74
2	4	25/4	25/10	–	90/40	2.00
3	7	70/11	42/19	103/7	107/51	2.37
4	13	47/13	47/22	100/0	100/46	3.20
5	9	55/16	55/20	–	112/60	6.70
6	4	30/3	37/14	–	124/40	2.55
7	8	50/15	50/21	100/5	100/50	3.24
8	4	57/7	55/24	120/0	116/40	4.45
9	6	29/8	13/10	94/0	95/30	1.53
10	4	56/8	50/20	–	100/40	3.30
11	5	78/1	74/30	165/0	171/74	2.80
12	4	126/10	28/10	–	150/66	1.04
13	5	94/6	30/12	–	143/99	1.00

**Mean Pressure*

The surgical approach in our patients were individualised depending on the nature of the associated cardiac anomalies. The aneurysmal sac was excised and the fistula closed by an aortotomy alone in 4 patients, a ventriculotomy alone in 1 patient and by a combined aortotomy and atriotomy in 8 patients. The aortic valve was replaced in all 6 patients with aortic regurgitation because of the incompetence and the marked deformity of the valve cusps.

The long term results as shown in Table 1 are good, the follow up ranging from 2 to 19 years. One patient required reoperation at 3 months for a recurrence of the fistula. There was only 1 death in the series, this in a patient who succumbed 2 months post operatively of sever mediastinitis. The remaining 12 patients remain well and are in New York Functional Class 1. Five patients have mild residual aortic regurgitation post operatively, which has not progressed during follow up.

Discussion

Aneurysms of the sinus of Valsalva are generally believed to be congenital in origin and this we felt to be the etiology in all our cases. Edward and Burchell[4] postulated that these aneurysms arose from a congenital lack of continuity between the aortic media and the annulus fibrosus. Acquired causes are less common and include infective endocarditis, syphilis and rheumatoid heart disease.[5] More commonly, these acquired causes tend to involve both the aortic sinus and the ascending aorta and tend to rupture into intracardiac as well as extracardiac structures such as the pericardium, mediastinum and the pleural spaces. This contrasts with the congenital aneurysms which are more likely to rupture into an intracardiac chamber due to the intracardiac location of the sinuses.[6] Although these aneurysms are congenital in origin, rupture in infancy or childhood is uncommon. It is sometimes difficult to determine if the aneurysm and subsequent rupture occured prior to or after an episode of infective endocarditis. Rarely trauma (both penetrating and closed chest injuries[7]) have resulted in aortocardiac fistulae.

It is now well recognised that the majority of patients with RASV present with insidious heart failure[3] due to chronic volume overloading and pulmonary hypertension (61.5% of our cases). This is especially so in patients with co-existent cardiac anomalies like ventricular septal defect and aortic regurgitation. Acute cardiac decompensation presenting with sudden severe chest pain and dyspnoea is an uncommon clinical presentation (15.4% of our cases). Rarely patients remain asymptomatic despite the rupture (23.1% of our cases). Rarer presentations include myocardial ischemic pain and palpitations due to the encroachment on the coronary arteries and conducting tissues by the aneurysm, especially in those cases involving the left aortic sinus.[8]

The classical auscultatory findings in isolated RASV is a continuous murmur at the left sternal edge often associated with a thrill. The character of the murmur, may however, be altered by the presence of associated cardiac defects. Occasionally, some workers have reported only a systolic murmur when the defect is small and others only a diastolic component[9] which could not be attributed to co-existing aortic regurgitation. In this case, it is postulated that little shunting occurs in systole because the pressure in the aortic sinus decreases during this phase of the cardiac cycle due to the Venturi effect.

Aneurysms of the sinus of Valsalva most commonly arise from the right aortic sinus (67.2%), followed by the non coronary sinus (25.2%) and lastly the left aortic sinus (7.6%).[10] Of the aneurysms arising from the right aortic sinus, 71% rupture into the right ventricle and 70% of those arising from the non coronary sinus rupture into the right atrium. 10 Isolated RASV are uncommon. The most common associated cardiac anomaly is a ventricular septal defect especially if the connection is between the right aortic sinus and the right ventricle (40% of cases).[10] Aortic regurgitation may also occur due to prolapse of the valve cusps and distortion of the valve ring.

The ECG and chest X-ray are generally not diagnostic. Although the aneurysm and fistula may be imaged on 2-D echocardiography, contrast studies11 or colour flow imaging is essential to diagnose rupture. In RASV, a negative contrast image or a turbulent colour jet may be seen due to blood flow from the aortic sinus to the right sided chamber.

The prognosis of RASV is grave and repair of the defect even when small, is advocated. The surgical approach varies with different workers although a double approach through both an aortotomy and a right ventriculotomy or atriotomy is preferred to prevent distortion of the aortic cusps and injury to the coronary artery during repair. Aortic valve replacement is not necessary in all cases with associated aortic regurgitation unless there is marked deformity and fibrosis of the valve cusps.[12] The long term results post repair are generally excellent. Recurrences are rare and if repaired immediately carry a low morbidity.

In the past, the majority of patients with RASV succumbed from congestive cardiac failure and infective endocarditis. Presently with increased clinical awareness and the advent of better diagnostic facilities and newer surgical techniques the prognosis of this condition has been greatly improved.

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