Approach to sinus of Valsalva aneurysms: a review of 53 cases

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Abstract

Objective: The reported experience with sinus of Valsalva aneurysms (SVAs) is limited. Our approach to this subset of patients and an algorithm-dependent classification are presented. **Methods**: Between 1985 and 2000, 53 patients (mean age: 24 ± 12; range 4–60) underwent repair for ruptured (64%) or non-ruptured (36%) SVA. Associated lesions were present in 21 patients; VSD in 18, moderate to severe aortic insufficiency in five, aortic stenosis in four (two subaortic membrane and one bicuspid valve), PDA in two, mitral insufficiency in one, tetralogy of Fallot in one and endocarditis in one. Operative procedures included simple or Teflon pledgetted direct suturing (31 cases; 58%), patch repair (21 cases; 40%), and stentless porcine bioprosthetic aortic root replacement in a case with extensive involvement and aortic root distortion (2%). Concomitant procedures were VSD repair in 18 patients, aortic valve replacement in four, aortic valve resuspension in three, subaortic membrane resection in two, PDA ligation in two, mitral annuloplasty in one and total correction in one. **Results**: Early mortality was 1.9%. A permanent pacemaker was inserted in one patient due to complete heart block. The survivors were followed up for 8.2 ± 5 years (range: 21 days to 15 years). There were three reoperations due to suture dehiscence; patch repair was undertaken in these patients with no further unfavorable consequences. All patients were in NYHA Class I or II as of their last follow-up. **Conclusions**: Repair of SVA can be performed with an acceptably low operative risk and a good symptom-free long-term outcome expectation. Echocardiography provides all the necessary details for diagnosis. Dual exposure/patch repair strategy is advocated in the ruptured cases. © 2001 Elsevier Science B.V. All rights reserved.

Keywords: Sinus of Valsalva; Aneurysm; Repair; Rupture

1. Introduction

Although first reports of sinus of Valsalva aneurysms (SVAs) appeared in the literature as early as in the nineteenth century [1], few large or long-term series are currently available. The first successful surgical repair by means of cardiopulmonary bypass was reported in 1957 by Lillehei et al. [2]. SVAs are repaired with low risk and good results in today's cardiovascular surgery practice with the techniques has been considerably refined; however, precise guidelines for the optimal approach for varying presentations are yet to be defined due to relatively limited reported experience, both in size and number.

In this report, we present our experience with 53 patients throughout 15 years and propose a classification system purely depending on the algorithm to be followed according to the clinical presentation.

2. Materials and methods

Between February 1985 and February 2000, 53 patients underwent repair for SVA. Our patient population consisted of 31 male (58%) and 22 (42%) female patients with a mean age of 24 ± 12 (range 4–60).

2.1. Clinical presentation

Presenting symptom was dyspnea (32 patients, 60%), chest pain (12 patients, 23%), syncope attacks (three patients, 6%), congestive heart failure (two patients, 4%); four patients (8%) were asymptomatic and diagnosis had been made during routine examinations or investigations for other diseases. A detailed trans-thoracic echocardiographic examination was sufficient for definitive diagnosis in all cases. Cardiac catheterization was seldom performed (only in 10 patients; 19%) and those were only to confirm the echocardiographic diagnosis in the early era of our experience. There were 34 ruptured (64%) and 19 non-ruptured (36%) cases (Table 1). Associated lesions (Table 2) were present in 21 patients (40%), VSD in 18 (34%), moderate to severe aortic insufficiency in five (9%), aortic

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Table 1
Patient data and anatomical features

	Whole group (53)	Ruptured cases (34 (64%))	Unruptured cases (19 (36%))
Involved sinus of Valsalva			
Right coronary sinus	35 (66%)	24	11
Non-coronary sinus	15 (28%)	10	5
Left coronary sinus	3 (6%)	0	3
Sex (female/male)	22/31	14/20	8/11
Mean age	24 ± 12	18 ± 7	30 ± 9
Presenting symptom			
Dyspnea	32 (60%)	26	6
Chest pain	12 (23%)	6	6
Syncope	3 (6%)	0	3
Congestive failure	2 (4%)	2	0
Asymptomatic	4 (8%)	0	4
Related cardiac chamber (in ruptured cases only)			
Right ventricle		20 (59%)	
Right atrium		13 (38%)	
Right ventricle and right atrium		1 (3%)	

stenosis in four (8%; two being subaortic membrane and one being bicuspid valve), PDA in 2 (4%), mitral insufficiency in one (2%), tetralogy of Fallot in one (2%) and endocarditis in one (2%).

2.2. Approach and management

A simple and functional classification system can be used as a guideline for the management of these lesions (Fig. 1). Surgical indication was rupture of the SVA and/or symptomatic SVA without rupture, documented by echocardiography (Fig. 2) and/or cardiac catheterization. Operative procedures included simple or Teflon pledgetted direct suturing (31 cases; 58%), patch repair (21 cases; 40%), and stentless porcine bioprosthetic aortic root replacement

Table 2
Associated lesions and procedures

Associated lesion	Number of patients (%)	Associated procedure
Ventricular septal defect	18 (34%)	VSD repair
Aortic insufficiency	5 (9%)	Valve resuspension in 3
		Valve replacement in 1
		Root replacement in 1
Aortic stenosis	4 (8%)	
Subaortic membrane	2 (4%)	Membrane resection
Bicuspid valve	1 (2%)	Valve replacement
Fibrotic	1 (2%)	Valve replacement
PDA	2 (4%)	PDA ligation
Mitral insufficiency	1 (2%)	Mitral annuloplasty
Tetralogy of Fallot	1 (2%)	Total correction
Endocarditis	1 (2%)	Valve replacement
Total	21 (40%)	

in a case with extensive involvement and aortic root distortion (2%). Subannular ventricular septal defects in two cases were closed with a common patch with additional fixation in the middle at the aortic annulus to prevent aortic leak into the left ventricle. Concomitant procedures were VSD repair in 18 (34%) patients, aortic valve replacement in four, aortic valve resuspension in three, subaortic membrane resection in two, PDA ligation in two, mitral annuloplasty in one and total correction with a trans-annular monocusp patch in one. All patients were assessed clinically and echocardiographically at 6-month intervals in the postoperative follow-up period.

2.3. Our current surgical strategy

Currently, we prefer 'dual exposure/repair' technique in the ruptured cases (Figs. 3 and 4), in which both the aorta and the related cardiac chamber are opened and both the ends of the defect are separately repaired, preferentially using a patch at the aortic end. Following the excision of the aneurismal sac at its base in the low-pressure cardiac chamber (right atrium in Fig. 4), the resultant defect is repaired either by direct suturing or patch closure, according to the size and location of the defect. In the early era of our practice, we used gluteraldehyde-fixed autologous pericardium or Dacron patches, but our current preference is polytetrafluoroethylene patch material.

2.4. Statistical analysis

This study does not include any comparative statistics. Data were presented as mean ± standard deviation. Event-free survival curve was estimated using the Kaplan–Meier
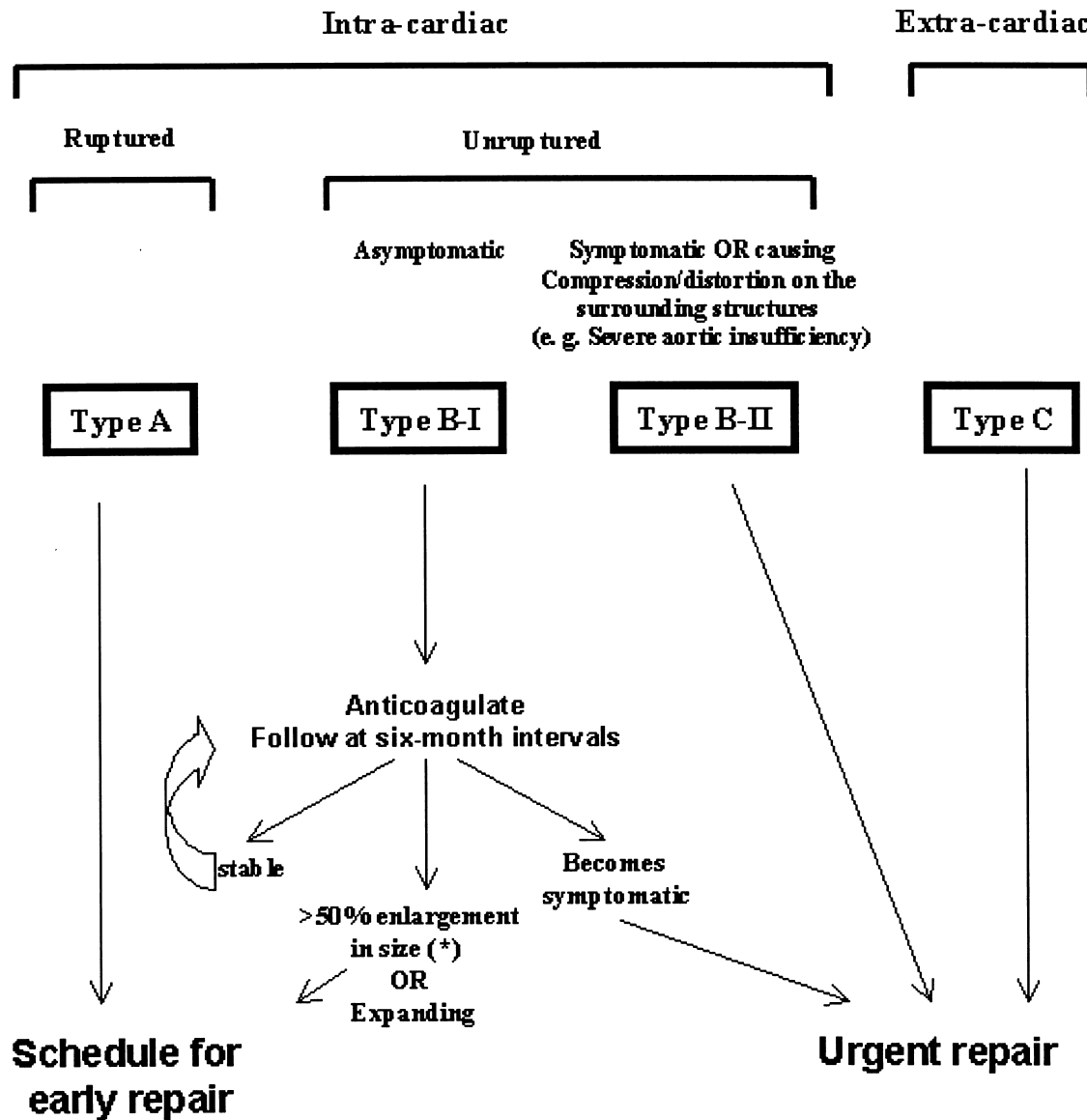
Fig. 1. The algorithm-dependent classification for the approach to SVAs. *'Normal' size of a sinus can be estimated by the average of the other uninvolved sinuses on echocardiographic or angiographic examination.

method by using SPSS software (release 6.0, SPSS Inc. Chicago, IL).

3. Results

3.1. Early mortality and postoperative complications

Early mortality was 1.9% (one patient) due to low cardiac output developed following the procedure. The average aortic cross-clamping time was 71 ± 28 min (22–139 min). Intravenous inotropic drug support was needed in eight patients (15%); no mechanical support was necessary. A complete atrio-ventricular heart block developed in one patient after resection of a heavily calcified bicuspid aortic valve. A permanent pacemaker was inserted due to persistence of the heart block. Mean duration of postoperative hospital stay was 8 ± 4 days, ranging between 4 and 21 days. Except for the abovementioned permanent pacemaker insertion, postoperative recovery period was uneventful for the remaining patients.

3.2. Postoperative follow-up

The survivors were followed up for 8.2 ± 5 years (range 21 days to 15 years). Three late dehiscences occurred in the cases repaired, and secondary repair was done using patch. These three reoperations were performed at the 67th, 123rd

Fig. 2. Color doppler-echocardiographic image of a right SVA ruptured into the right atrium. Excellent information can be obtained by trans-esophageal echocardiography. RA, right atrium; RV, right ventricle; PA, pulmonary artery; AN, aneurysm sac.

and 218th postoperative days. In all the three, the repair had been done with direct closure using simple suturing at the initial operation. At the reoperations, patch repair was undertaken in these cases with no further unfavorable consequences. One anticoagulation-related bleeding at the 41st postoperative day necessitated hospitalization and was treated medically. All patients were in NYHA Class I or II as of their last follow-up. The Kaplan–Meier event-free survival estimate (Fig. 5) revealed an $88 \pm 5\%$ event-free survival at 14 years, including the operative mortality and postoperative complications (six events: one early postoperative death, one permanent pacemaker insertion, three late suture dehiscence and one anticoagulation-related bleeding).

4. Discussion

SVAs are rare cardiac anomalies, which may be either acquired or congenital. A congenital lack of continuity between the aortic media and annulus fibrosis may initiate
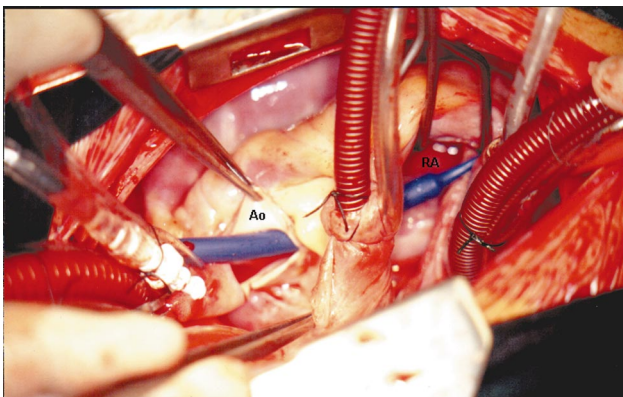

Fig. 3. Surgical exposure of the same case in Figs. 2 and 3. The blue probe passes through the fistula and indicates the communication between the right Valsalva sinus (Ao) and the right atrium (RA).
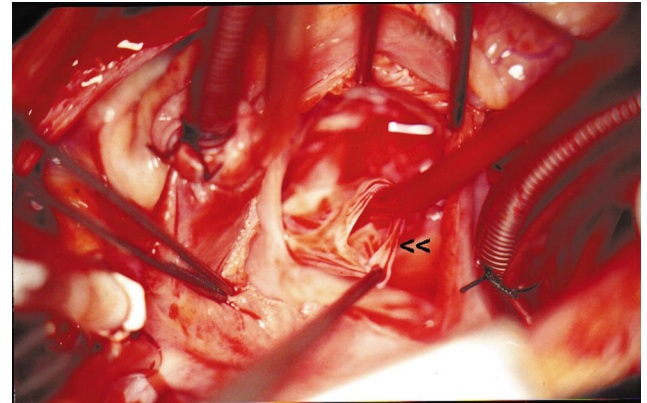

Fig. 4. Exposure of the ruptured windsock aneurysm protruding into the right atrium (arrow). This aneurysm sac was resected at its base and closed by direct suturing. The aortic end of the defect was repaired using a 4×5 mm^2 polytetrafluoroethylene patch (dual exposure and repair technique). Patch repair is preferred over direct suturing at the aortic end, in an attempt to avoid aortic leaflet distortion and late suture dehiscence. Either direct suture (for small defects) or patch repair (larger defects or, defects with a VSD) can be used in the low-pressure chamber.

aneurysm formation [3] or less frequently, infections or degenerative processes may affect the aortic wall. Acquired aneurysms may result from trauma [4], endocarditis [5], syphilis [6], Behcet [7], Marfan's syndrome, and senile-type dilatation. A frequent coexistence with VSDs suggests defective distal bulbar septum development [3].

The natural history of either ruptured or non-ruptured SVAs is difficult to determine due to rarity of these lesions. They may cause aortic regurgitation [8] and may rupture into an extra or intracardiac location. When rupture occurs into an intracardiac chamber, they may remain silent initially, but later manifest as progressive heart failure due to shunting and aortic regurgitation. By dissecting into the interventricular septum, SVAs may result in complete heart block necessitating permanent pacemaker insertion [9]. Free intrapericardial rupture is a rare but dreadful complication of these aneurysms, and carries a very high
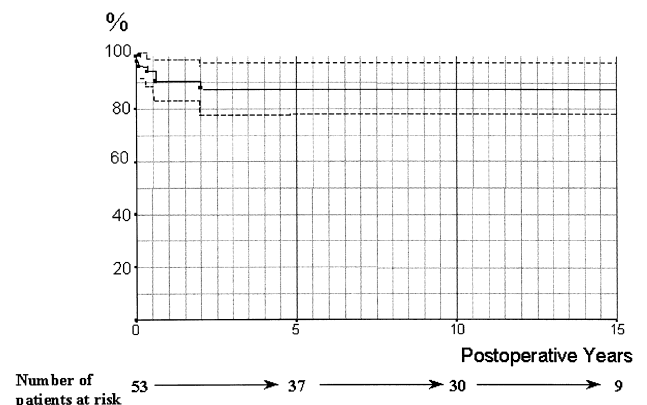

Fig. 5. Event-free survival curve estimated by the Kaplan–Meier method. This curve also includes operative mortality and early/late postoperative complications. Dashed lines represent 95% confidence intervals.

mortality [10]. A mean survival period of 3.9 years in patients with untreated ruptured SVAs was reported [11]. Therefore, early surgical intervention is recommended in this subset of cases. Surgical intervention is also necessary in non-ruptured but symptomatic SVAs. A non-ruptured SVA may cause right ventricular outflow tract obstruction [12], infective endocarditis [13], malignant arrhythmias [14], or myocardial ischemia/infarction due to severe distortion of coronary ostia [15] or compression of the coronary trunks [16]. Optimal management of an asymptomatic, non-ruptured aneurysm, however, is not so certain since no precise natural history is currently available. Non-ruptured SVAs, although remaining silent, may expand, causing more severe symptoms and requiring more extensive corrective procedures in the future. For this reason, some authors recommend early correction also in these types of lesions [17]. Fatal intrapericardial rupture may also happen with silent SVAs [10]. Also, thrombus formation or bacterial colonization may occur in the defective tissue and may cause serious complications such as stroke [18] and sepsis [17]. This fact calls for alertness and early intervention in this group.

The popular classification system of Sakakibara and Konno [19] defines types I–IV according to the position of the aneurysm sac relative to the right or non-coronary aortic valve cusps. This classification is commonly used today to describe the location of SVAs and indicate the cardiac chamber (right atrium or right ventricle) that is invaded by the expanding aneurysm. However, the growing sac may also protrude into the left ventricle, left atrium [14] or originate from the left coronary sinus to form an extra cardiac mass [20]. Our proposed approach (Fig. 1) depends on the cumulative experience from all those abovementioned reports and provides with an algorithm to be followed. For the unruptured, asymptomatic aneurysms (type B-I in the presented algorithm) the size of a given aneurysm may be a decision-making factor, although this is not supported by any scientific data and is open to discussion. It is our belief that, should the enlargement exceed 50% of the normal sinus size (or practically, the average of the other two uninvolved sinuses) on echocardiographic or angiographic examination, this aneurysm must be repaired, even if it remains silent. Further, if an aneurysm is expanding when compared to previous evaluations, the decision for surgical repair, again, should be undertaken regardless of its size. Similarly, symptomatic patients, and those presented with a destructive, compressive or distorting effect on the surrounding tissues and structures (e.g. severe aortic regurgitation) should be repaired immediately.

Non-invasive diagnosis of this anomaly is of particular interest. In our experience, trans-thoracic echocardiography was sufficient for the diagnosis. Cardiac catheterization and angiocardiography, although seldom performed for confirming anatomical and physiological details, was never mandatory.

Among the options for the treatment of SVAs are catheter closure [21], direct suture repair, patch closure [22], aortic root replacement [19], endoaneurysmal repair [23] and aortic valve sparing operation (remodeling). We believe that both aorta and chamber of entry should be explored for an effective correction (dual exposure and repair technique). Interventricular septum should be carefully inspected for possible association of VSD. Repair should preferentially be done with a patch rather than by simple closure, especially in large defects and at the aortic end of the fistula. This minimizes aortic leaflet distortion and late risk of dehiscence. In our experience, the three late dehiscences occurred in the cases repaired using simple suturing instead of a patch at the initial operation. At the reoperations, patch repair was undertaken in all. Similar experience was reported by others [22]. Therefore, our current tendency is toward using a patch. In contrast, it is reported by Van Son et al. that reoperation incidence was not related to location of fistula or type of repair (direct suture vs. patch) [24]. In that study, the risk of reoperation was higher with right ventricle fistulas than with those opening into the right atrium, higher with the presence of a subarterial VSD, and lower when repair was done via an aortotomy (with or without right ventriculotomy) instead of using a right ventriculotomy only.

In conclusion, surgical treatment of SVAs can be performed with an acceptably low operative risk and ensuring good long-term event-free/symptom-free survival, as emphasized by others [17,24]. This encourages early surgical intervention, which helps prevent the development of worse symptoms and more extensive disease leading to more complicated and less satisfying repairs.

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