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Surgery for ruptured sinus of Valsalva aneurysm: Five-year experience with 19 patients

Rüptüre sinüs Valsalva anevrizması için cerrahi: 19 hasta ile beş yıllık deneyim

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Background: In this article, we reviewed our five-year experience in the repair of ruptured sinus of Valsalva aneurysm (RSVA), and the current literature regarding the efficacy of patch closure.

Methods: A retrospective review identified 19 patients (15 males, 4 females; mean age 28±12 years; range 14 to 55 years) who underwent RSVA repair between January 2009 and October 2013. The RSVA originated from the right coronary sinus in 17 patients (89.4%), and from the noncoronary sinus in two patients (10.5%). Ruptured sinus of Valsalva aneurysm did not originate from the left coronary sinus in any of the patients. In 11 patients (57.8%), the most common occurrence was right ventricular rupture, whereas it was right atrial rupture in eight patients (42.10%). Common associated defects were subaortic ventricular septal defect in two patients (10.7%), aortic insufficiency in seven patients (36.8%), and tricuspid insufficiency in one patient (5.2%). A bicameral approach was used for repair. Ruptured sinus of Valsalva aneurysm was repaired with an expanded polytetrafluoroethylene patch in all cases. Aortic valve was replaced in seven patients, and tricuspid in one patient.

Results: The hospital mortality rate was 5.2%. Follow-up, ranging from one month to 4.5 years, was available in 94.3% of survivors (n=18). Actual survival rate was 95%. Freedom from reoperation was 100%.

Conclusion: To conclude, surgical repair appears to be the optimal choice for the treatment of RSVA. Rapid surgical intervention after diagnosis may lead to successful outcomes.

Keywords: Acyanotic congenital heart disease; patch repair; ruptured aneurysm; sinus of Valsalva aneurysm.

Amaç: Bu yazıda rüptüre sinüs Valsalva anevrizması (RSVA) tamirindeki beş yıllık deneyimimiz ve yama ile kapamanın etkinliğine ilişkin mevcut literatür incelendi.

Çalışma planı: Retrospektif bir incelemeyle Ocak 2009 ve Ekim 2013 tarihleri arasında RSVA tamiri geçirmiş 19 hasta (15 erkek, 4 kadın; ort. yaş 28±12; dağılım 14-55 yaş) belirlendi. Rüptüre sinüs Valsalva anevrizması 17 hastada (%89.4) sağ koroner sinüsten, iki hastada (%10.5) nonkoroner sinüsten kaynaklanmaktaydı. Rüptüre sinüs Valsalva anevrizması hastaların hiçbirinde sol koroner sinüsten kaynaklanmamaktaydı. On bir hastada (%57.8) en yaygın olay sağ ventrikül rüptürüyken sekiz hastada (%42.10) sağ atriyum rüptürüydü. Yaygın ilişkili defektler iki hastada (%10.7) subaortik ventriküler septal defekt, yedi hastada (%36.8) aort yetmezliği ve bir hastada (%5.2) triküspid yetmezliği idi. Onarım için bikameral yaklaşım kullanıldı. Rüptüre sinüs Valsalva anevrizması tüm olgularda genişletilmiş politetrafloroetilen yama ile onarıldı. Yedi hastada aort kapağı, bir hastada triküspid değiştirildi.

Bulgular: Hastane mortalite oranı %5.2 idi. Süresi bir ay ila 4.5 yıl arasında değişen takip, sağ kalanların %94.3'ü (n=18) ile yapıldı. Gerçek sağkalım oranı %95 idi. Tekrar ameliyattan uzak yaşam oranı %100 idi.

Sonuç: Rüptüre sinüs Valsalva anevrizması için en iyi tedavi seçeneğinin cerrahi tamir olduğu anlaşılmaktadır. Tanıdan sonra hızlı cerrahi girişim başarılı sonuçlar sağlayabilir.

Anahtar sözcükler: Asiyanotik doğuştan kalp hastalığı; yama tamiri; rüptüre anevrizma; sinüs Valsalva anevrizması.

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A sinus of Valsalva aneurysm (SVA) is an infrequent occurrence, which may be either congenital or acquired, that has an incidence rate ranging from 0.14-3.5% for patients who undergo open heart surgery.[1] In addition, SVAs can rupture into any of the cardiac chambers to form an aorticocardiac fistula, but those on the right side are more affected. Moreover, males are three to four times more likely to have SVAs, and the incidence of a ruptured sinus of Valsalva aneurysm (RSVA) is higher in Asian (1.2%-4.94%) versus Western populations (0.5%-1.5%).[1] Once rupture has occurred, the mean survival period for untreated patients is one to two years, which demonstrates the need for early surgical intervention.[2]

The first successful surgical treatment for an SVA was performed in 1957 by Lillehei using a cardiopulmonary bypass (CPB).[2] He approached the aneurysm via the involved chamber, and closure was done with interrupted silk stitches. Since that time, various closure techniques (primary closure vs. patch closure) and surgical approaches (transaortic, dual, or involved chamber have been tried, but there is still no consensus on which technique is best.[3] Herein, we reviewed our five-year experience with RSVAs to assess the long-term outcome of surgical repair and the factors that influenced our patients' prognosis.

PATIENTS AND METHODS

This retrospective study was composed of 19 patients (15 males, 4 females; mean age 28±12 years; range 14 to 55 years) with an RSVA who underwent surgical repair at our facility between January 2009 and October 2013. At admission, 18 of the patients were symptomatic, and four presented with an acute onset of symptoms. Fourteen had dyspnea, 12 suffered from palpitations, 12 experienced fatigue, two had chest pain, two had a fever, and one experienced from syncope. In addition, 12 were categorized as having New York Heart Association (NYHA) functional Class III and IV (Table 1). Furthermore, a continuous "machinery" murmur was heard at the left sternal border in 17 patients. The preoperative systemic pulse pressure ranged from 40 to 150 mmHg (mean, 70±20 mmHg), and the chest radiography findings included an increased cardiothoracic ratio of 0.53±0.07 (range 0.45-0.79) with varying degrees of pulmonary plethora (Figure 1). We also found that the electrocardiographic findings were normal in just two of the patients. Ten others had left ventricular hypertrophy while one had right ventricular hypertrophy. The RSVA was diagnosed by echocardiography in 18 patients (Figure 2), and it originated from the right coronary sinus in 17 patients (89.4%) and the noncoronary sinus in two others (10.5%). None originated from the left coronary sinus. Rupture into the right ventricle was the most common finding in 11 patients (57.8%), but for eight others, it occurred in the right atrium (42.10%). Furthermore, we discovered a subaortic ventricular septal defect (VSD) in two patients (10.7%) and found that seven had aortic insufficiency (36.8%) while one had tricuspid insufficiency (5.2%) (Table 2). To repair the RSVA, we employed the bicameral approach, which includes the involved chamber and the aorta, and used expanded polytetrafluoroethylene (ePTFE) patches for all of the patients. Moreover, the aortic valve was replaced in seven of the patients and the tricuspid valve was replaced in one.

Table 1. Pre- and postoperative New York Heart Association class

	Preoperative		Postoperative	
	n	%	n	%
NYHA				
Class I	1	5.2	15	78.9
Class II	1	5.2	3	15.9
Class III	15	78.9	0	0
Class IV	2	10.5	0	0

NYHA: New York Heart Association.

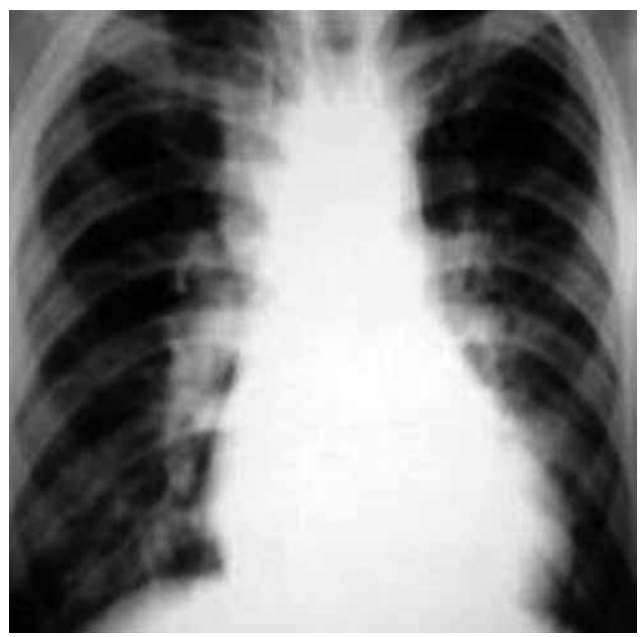

Figure 1. Preoperative chest radiograph of a patient showing the increased cardiothoracic ratio and pulmonary plethora.

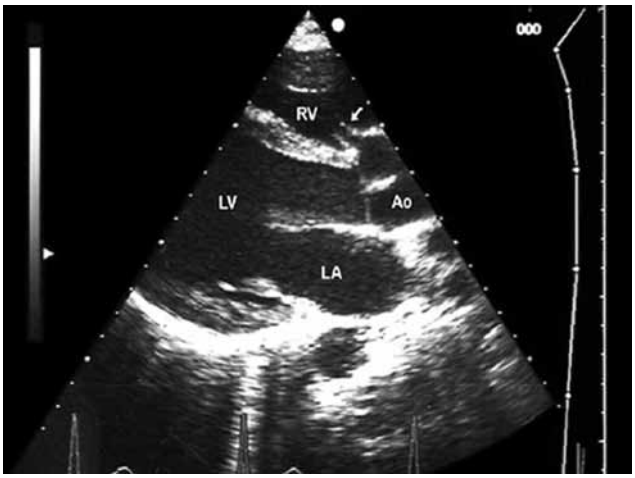

Figure 2. Transthoracic echocardiograph (parasternal long-axis view) showing the ruptured sinus of Valsalva opening into the right ventricle (RV) (arrow). LV: Left ventricle; Ao: Aorta; LA: Left atrium.

Surgical procedure

Cardiopulmonary bypass and moderate hypothermia were used in all of the cases, and direct coronary ostial antegrade hypothermic blood cardioplegia was utilized for myocardial protection. Furthermore, an oblique aortotomy was performed to check the pathology of the aneurysm, aortic cusps, and associated cardiac anomalies. For two patients with an isolated RSVA that had ruptured into the right ventricle without a VSD, an aortotomy was the preferred surgical technique, whereas the double-chamber approach was used in the remaining 17 patients with an RSVA that had ruptured into either the right atrium (n=8) or the right ventricle (n=9). This type of rupture was associated with a right (i.e., VSD) and right ventricular outflow tract obstruction (RVOTO). The RSVA was primarily repaired with a patch or aortic valve replacement (Table 3). Direct closure of the aneurysm was done in two patients (10.7%), and seven (36.84%) received a prosthetic aortic valve. Patch closure was performed on the remaining 17 patients who did not require direct closure.

The VSDs were approached through a right ventriculotomy and then closed with a patch (n=2). The surgical procedures associated with the RSVA closure are listed in Table 3.

RESULTS

The early mortality rate in our study was 5.2% (n=1) as one patient dyed because of septic shock on the 12th postoperative day. Temporary atrioventricular (AV) block developed in one patient, but none of the patients required a permanent pacemaker. In addition, no ventricular arrhythmias or sudden cardiac deaths were seen in any of the patients in this series. The mean duration of the postoperative hospital stay was 10±4 days (range 4-15 days), and all 18 survivors were NYHA Class I or II when they were discharged from the hospital (Table 1). Three were classified as NYHA class II with a history of dyspnea that may have been caused by aortic valve incompetence. The other 15 had no such complaints. Finally, we determined that the actual survival rate was 94.8%, and the freedom from reoperation for reoccurrence was 100%.

DISCUSSION

The goals of RSVA repair procedures are to close them securely, remove the aneurysmal sac, and repair any

Table 2. Origin and site of rupture of the sinus of valsalva aneurysm

Origin	Right ventricle	Right atrium	Number of patients
Right coronary sinus	11	6	17
Noncoronary sinus	0	2	2
Left coronary sinus	0	0	0
Total	11	8	19

Table 3. Coexisting lesions and corresponding procedures

Lesion	Procedure	Number of patients	
		n	%
Ventricular septal defect	Patch closure (Dacron)	2	5.2
Aortic regurgitation	Aortic valve replacement	7	36.8
Right ventricular outflow tract obstruction	Repair	1	5.2
Tricuspid insufficency	Tricuspid valve replacement	1	5.2

associated defects without causing heart block or aortic valve dysfunction, and different surgical strategies have evolved to achieve these goals. However, because of the rarity of RSVAs, there have been no clinical trials to show whether one surgical repair technique is superior to another.

An analysis of a published case series by Chu et al.[4] found that the incidence is approximately five times higher for patients in the Far East than for those in the West, and most studies, including ours, have found that the right coronary sinus is affected the most followed by the noncoronary sinus. The aneurysm most often ruptures into the right ventricle, with the second most common site being the right atrium. However, RSVAs have also rarely been found in the left ventricle, pulmonary artery, or interventricular septum. In our study, the aneurysm ruptured most often into the right ventricle. A ruptured left SVA is seldom seen because the left coronary cusp does not usually arise from the bulbar septum in the same manner that the right and noncoronary cusps do.

Various studies have found that males are more likely to have SVAs, with figures ranging from 51% to as high as 88%, and that patients can be diagnosed anywhere from two years old to the age of 80 (mean age 31.89 years).[3,5-23] In this study, the mean age was 30.9 years, and 78% of the patients were male.

Adams et al.[24] documented a mean survival of 3.9 years in their study comprised of patients with untreated RSVAs, which suggests that early surgical intervention is needed. Surgery is also recommended in symptomatic, nonruptured aneurysms, but optimal management for asymptomatic, nonruptured SVAs is less clear. In the report by Takach et al.,[6] a patient who refused to have surgery for an asymptomatic noncoronary SVA and trace aortic regurgitation (AR) progressed to severe AR.

A VSD is the most common cardiac anomaly found in conjunction with RSVAs, with rates of between 9 and 78% having been reported.[3,5-23] In our series, the prevalence rate for patients with a VSD was 10.5% (two out of 19), and the two with the VSD were closed with a patch. In addition, no residual VSDs were subsequently identified. Aortic valve abnormalities and incompetence are common in patients with RSVAs, and aortic valve replacement may be required at the time of RSVA closure if the cusps are highly deformed and not suitable for repair. In our series, moderate-to-severe aortic insufficiency occurred in seven patients (36.8%), and the aortic valve was replaced in all of them.

Interventional closure in the catheterization laboratory is another treatment option,[7] especially since the ideal surgical approach has yet to be determined. Controversy exists among surgical centers with regard to the best closure technique (primary closure vs. patch closure) and preferred surgical approach (dual, transaortic, and involved chamber) for RSVAs. Some reports have identified an association between the primary closure technique and recurrent rupture and worsening AR.[5,7,9,19,23] Others have recommended using a patch to close SVAs in all cases because it does not deform the aortic valve and it reduces the stress on the suture line.[7,9-13,16,23]

Jung et al.[19] noted that the transaortic repair may cause postoperative AR by progressively distorting the sinus geometry. However, Liu et al.[23] found no association between this surgical approach and AR.

The involved chamber (only) technique should only be performed on those patients without significant AR. In the dual approach, both the aorta and the involved chamber are used. Althought this type of surgery has some advantages, such as being able to repair the defect from both sides, it is more time consuming.[3,7,10,12,15,18]

When an SVA ruptures into the right ventricle without associated cardiac lesions, we perform the repair through an aortotomy with the use of a patch. If an SVA ruptures into the right atrium or right ventricle and there are associated cardiac lesions, we prefer the bicameral approach because the defect can be repaired from both sides. This approach was used in 17 of our patients (89.43%) while the transaortic approach was used in just two (10.5%). We have noticed an increasing tendency to use patch repair in recent years, and most studies have also recommended the use of the dual approach[3,7,10,12,15,18] because the operative mortality rate is generally low (0.5-11%) with this procedure and the prognosis after the surgical repair of the SVA is satisfactory.[3,5-20,23] In our series, the operative mortality rate was 5.2% and the actual survival rate was 94.7% at five years, which was similar to previously published results.[2,3,5,6]

Conclusion

Surgical repair of an RSVA is associated with an acceptably low operative risk and long-term freedom from death and reoperation. When an RSVA is diagnosed, the treatment of choice should be surgical repair as soon as possible. We recommend a case-specific surgical technique beginning with an aortotomy, and a patch should be used at the aortic end to minimize aortic

leaflet distortion. The resultant defect, if opened, can be repaired either by direct suturing or patch closure, depending on the size and location.

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