Angiographic features of ruptured sinus of Valsalva aneurysm: New classification

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ABSTRACT

Background: The formal classification system for ruptured sinus of Valsalva aneurysm (RSVA) is from a surgical aspect and is seldom utilized for percutaneous closure. This study was undertaken to introduce a new classification for RSVA according to the angiographic features of patients.

Methods: We retrospectively studied 30 cases of RSVA undergoing percutaneous closure between July 2005 and September 2013. The data of patients' angiographic features, management, and outcomes were collected and analyzed.

Results: The patients included 18 males and 12 females with a median age of 42.5 years (range, 24–74 years). According to the shape of left to right shunt jet, patients were divided into four types: type I, window-like, 56.7% (\(n = 17\); type II, aneurysmal, 16.7% (\(n = 5\)); type III, tubular, 16.7% (\(n = 5\)); and type IV, other rare conditions, 10.0% (\(n = 3\)). One patient in type IV had a giant RSVA and the other 2 in type IV presented with angiographic features of long and funnel shape. Total occlusion rate was 93.3% (28 out of 30 patients) at discharge and during a median follow-up of 18.5 months (1–96 months). In patients with types I and II, small-waist double-disk ventricular septal defect (VSD) occluders were chosen. In patients with type III, muscular VSD occluders were chosen. We failed in 2 out of 3 patients in type IV for serious hemolysis and occluders were retrieved finally. The proportion of patients in New York Heart Association class III/IV was reduced from 73.3% at baseline to 10% at the time of last follow-up (p < 0.001).

Conclusion: According to the shape of left to right shunt jet, we propose a new and simple classification for RSVA. It could help toward the better understanding of angiographic morphology of RSVA and facilitate the selection of occluders for percutaneous closure.

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was performed using a 6 French pigtail catheter to observe the scopic guidance, as described before[18]. Aortic root angiography

Percutaneous device implantation

Materials and methods

Patient selection

Patients would be considered candidates for percutaneous treatment if they had the European system for cardiac operative risk evaluation II (EuroSCORE II) >20%, or if surgery was deemed to be of excessive risk. The EuroSCORE II calculator is online at http://www.euroscore.org/calc.html. From July 2005 to September 2013, a total of 30 patients with RSVA in 5 institutions were enrolled in our study for attempted percutaneous closure. The Ethics Committee of each hospital approved the study, and the decision to proceed with percutaneous closure was discussed by a dedicated heart team.

Closure device and follow-up

The occluders used were modified double-disk ventricular septal defect (VSD) occluders (Shanghai Shape Memory Alloy Ltd, Shanghai, China), which were designed based on the Amplatzer occluders and had been widely used for selected VSD in China[17]. There are 3 types of modified VSD occluders used in the study: small-waist double-disk occluders, muscular occluders, and asymmetric occluders. The diameter of the small-waist double-disk occluder is different in the left (aortic) and right (RA or RV) side disk[10]. The above occluders were approved by the State Food and Drug Administration (SFDA) of China in 2003, and received CE mark in 2008.

Percutaneous device implantation

Percutaneous device implantation was performed under fluoroscopic guidance, as described before[18]. Aortic root angiography was performed using a 6 French pigtail catheter to observe the size, shape, and opening of RSVA. An arterial-venous wire loop was established from right femoral artery, ascending aorta, RSVA, RV, inferior vena cava, and right femoral vein. A 9–10 French delivery sheath was placed into the ascending aorta across the lesion over the loop.

We chose an appropriate occluder that measured 2–4 mm larger than the entrance diameter of RSVA. The selected occluder with its attached delivery cable was then inserted through the delivery sheath from the venous route, and its aortic disk was deployed in the ascending aorta. A gentle traction was exerted on the delivery cable to confirm seating of the left disk on the aortic side without slippage into the RSVA. Then the rest of the occluder was deployed on the right side across the lesion. The device was released after making certain that there was no significant aortic regurgitation (AR), tricuspid regurgitation, or encroachment on coronary arteries on the basis of aortic root angiography, transthoracic echocardiography (TTE), or transesophageal echocardiography (TEE) evaluation.

TTE and electrocardiography were performed 1 week later and routinely assessed at 1, 6, and 12 months after the procedure and yearly thereafter. The residual shunt was defined as trivial (<1 mm color jet width), small (1–2 mm color jet width), moderate (3–4 mm color jet width), or large (>4 mm color jet width) by TTE[19]. Valve regurgitation was evaluated by color Doppler flow imaging in a standard way.

Statistical analysis

All continuous variables are expressed as mean values and standard deviation or median with range as appropriate, and discrete variables are presented as percentages. Univariate analysis was performed by the Student’s t-test and chi-square test or Fisher’s exact test. A p-value <0.05 was considered statistically significant. The data were analyzed with statistical software SPSS 17.0 (Chicago, IL, USA).

Results

Baseline characteristics

The patients included 18 males and 12 females with a median age of 42.5 years (range, 24–74 years). Major symptoms were chest pain in 17 (56.7%) and dyspnea in 21 patients (70.0%). Nine patients (30.0%) had coronary artery disease and 11 patients (36.7%) had significant renal dysfunction (estimated glomerular filtration rate <60 ml/min). New York Heart Association (NYHA) functional class III/IV was prevalent in 22 patients (73.3%). The mean logistic EuroSCORE II was 25.2 ± 8.4.

Angiographic classification system

Following a detailed analysis of the 30 cases, we proposed a novel classification system for RSVA according to the shape of left to right shunt jet by aortic root angiography (Table 1). Patients were divided into 4 types: type I, window-like, the shunt jet is scattered immediately after crossing the ruptured site (Fig. 1A); type II, aneurysmal, the shunt jet is aneurysmal shape (Fig. 1B); type III, tubular, the shunt jet is a tubular shape with a long waist (Fig. 1C); and type IV, other rare conditions.

The baseline characteristics of patients in each type are summarized in Table 2. The patients were divided into: window-like, 56.7% (n = 17); aneurysmal, 16.7% (n = 5); and tubular, 16.7% (n = 5). Three patients with other rare angiographic shape were identified in type IV. 10.0% (n = 3) (Fig. 2). One of them presented with a giant RSVA from RCS to RA adjacent to the tricuspid valve. The other 2 presented with angiographic features of long and funnel shape. Aneurysms originated from the RCS in 19 patients (63.3%) and most of them (33.3%) were in type I. The aneurysm terminated into the RA in 16 patients (53.3%) and the majority of them (33.3%) were in type I. Coexistent perimembranous VSD was found in 6 patients. Four of them were in type I and the other 2 were in type IV. Three patients (10.0%) combined with AR, and 2 of them were in type I.

<table>
<thead>
<tr>
<th>Classification systems for RSVA.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sakakibara classification</td>
</tr>
<tr>
<td>I: RCS to the RVOT below the pulmonary valve</td>
</tr>
<tr>
<td>II: RCS to RV infundibulum in the supraventricularis crest</td>
</tr>
<tr>
<td>III: RCS to the RA</td>
</tr>
<tr>
<td>IV: RCS to the RV at membranous ventricular septum</td>
</tr>
</tbody>
</table>

NCS, non coronary sinus; RA, right atrium; RCS, right coronary sinus; RSVA, ruptured sinus of Valsalva aneurysm; RV, right ventricle; RVOT, right ventricular outflow tract.
Fig. 1. Types I, II, and III in the angiographic classification system for ruptured sinus of Valsalva aneurysm. (A) In the type I, window-like, the shunt jet is scattered immediately after crossing the ruptured site; (B) In the type II, aneurysmal, the shunt jet is aneurysmal shape after crossing the ruptured site; (C) In the type III, tubular, the shunt jet is long and the diameter in the left and right sides of the ruptured site is the same.

Table 2
Baseline characteristics of patients in each type.

<table>
<thead>
<tr>
<th>Total group</th>
<th>Window-like</th>
<th>Aneurysmal</th>
<th>Tubular</th>
<th>Other rare conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients (n)</td>
<td>30 (100)</td>
<td>17 (56.7)</td>
<td>5 (16.7)</td>
<td>5 (16.7)</td>
</tr>
<tr>
<td>Involved sinus of Valsalva</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RCS [n (%)]</td>
<td>19 (63.3)</td>
<td>10 (33.3)</td>
<td>3 (10.0)</td>
<td>4 (13.3)</td>
</tr>
<tr>
<td>NCS [n (%)]</td>
<td>11 (36.7)</td>
<td>7 (23.3)</td>
<td>2 (6.7)</td>
<td>1 (3.3)</td>
</tr>
<tr>
<td>Related cardiac chamber</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RA [n (%)]</td>
<td>16 (53.3)</td>
<td>10 (33.3)</td>
<td>2 (6.7)</td>
<td>3 (10.0)</td>
</tr>
<tr>
<td>RV [n (%)]</td>
<td>14 (46.7)</td>
<td>7 (23.3)</td>
<td>3 (10.0)</td>
<td>2 (6.7)</td>
</tr>
<tr>
<td>Associated VSD [n (%)]</td>
<td>6 (20.0)</td>
<td>4 (13.3)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Associated AR [n (%)]</td>
<td>3 (10.0)</td>
<td>2 (6.7)</td>
<td>0</td>
<td>1 (3.3)</td>
</tr>
</tbody>
</table>

Data are expressed as number (percentage). NCS, non coronary sinus; RA, right atrium; RCS, right coronary sinus; RV, right ventricle; AR, aortic regurgitation; VSD, ventricular septal defect.

Occluder selection and outcomes

The data of patients’ management, outcomes, and complications are illustrated in Table 3. Total occlusion rate was 93.3% (28 of 30 patients) at discharge and during a median follow-up of 18.5 months (1–96 months). The diameter at the ruptured site varied from 4 to 8 mm (median 6 mm) and the size of the occluders used to close the ruptured site varied from 6 to 12 mm (median 8 mm).

For patients in types I and II, small-waist double-disk occluders were used. For patients in type III, muscular occluders were chosen (Fig. 3). Four patients in type I combined with perimembranous VSD and 2 of them were implanted with asymmetric occluders for VSD simultaneously. The other 2 patients who combined with small perimembranous VSD (less than 3 mm in diameter) were only treated for RSVA.

One patient in type IV presented with a giant RSVA from RCS to RA adjacent to the tricuspid valve, measuring 41 mm × 34 mm at the maximal echocardiographic diameters. TTE revealed that the entrance diameter was 4 mm and a 6-mm small-waist double-disk occluder was deployed across the lesion successfully.

We failed in the other two patients in type IV with angiographic features of long and funnel shape. Both of them presented with coexistent perimembranous VSD (7 mm and 11 mm in diameter). One also combined with moderate AR. Percutaneous closure of RSVA was undertaken using a muscular VSD occluder combined with a small-waist double-disk occluder and a muscular VSD occlude, respectively. We planned a second procedure for VSD closure in one week. Moderate residual shunt occurred immediately and hemolysis developed 4 h and 8 h after percutaneous closure.

Fig. 2. Type IV, two cases of rare conditions in the angiographic classification system for ruptured sinus of Valsalva aneurysm (RSVA). (A) Aortogram reveals a giant RSVA; (B) Aortogram reveals the shunt jet is long and funnel shaped after crossing the ruptured site.
Interventional information and outcomes for different types.

<table>
<thead>
<tr>
<th>Type</th>
<th>Patients (n)</th>
<th>Defect size (mm)</th>
<th>Occluder size (mm)</th>
<th>Occluders used (n)</th>
<th>Complications [n (%)]</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Small-waist double-disk</td>
<td>Muscular</td>
<td>Asymmetric</td>
</tr>
<tr>
<td>Total</td>
<td>30</td>
<td>6</td>
<td>8</td>
<td>24</td>
<td>7</td>
</tr>
<tr>
<td>Window-like</td>
<td>17</td>
<td>4</td>
<td>8</td>
<td>17</td>
<td>0</td>
</tr>
<tr>
<td>Aneurysmal</td>
<td>5</td>
<td>6</td>
<td>8</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Tubular</td>
<td>5</td>
<td>7.5</td>
<td>10</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Other rare conditions</td>
<td>3</td>
<td>6</td>
<td>11</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

Data are expressed as number (percentage).

AR, aortic regurgitation.

The 2 patients were converted to surgery after conservative treatment for one week.

Follow-up evaluation

At discharge, trivial residual shunts were noticed by TTE in 3 patients in type I. De novo mild occluder-related AR developed in 5 patients in type I. During a median follow-up of 18.5 months (1–96 months), trivial residual shunts disappeared in all patients. Mild occluder-related AR disappeared in 4 out of 5 patients and remained unchanged in 1 patient. Two patients with mild AR preoperatively remained unchanged at discharge and at the time of last follow-up.

At baseline, NYHA functional class was II in 8 patients, III in 19, and IV in 3. At most recent follow-up, 14 patients were in NYHA class I and 13 in NYHA class II. Three patients combined with coronary artery disease were still classified in NYHA class III. The proportion of patients in NYHA class III/IV was reduced from 73.3% at baseline to 10% at the time of last follow-up. A similar closure rate in RSVA with surgical closure has also been reported to be between 90.9% and 95.0% [20,21]. Excellent results of percutaneous closure indicate that it is a promising alternative to surgery in appropriately selected patients.

Angiographic classification system

On the basis of 30 cases of RSVA in the present study, we introduced a new classification system from the standpoint of interventional cardiologists. According to the shape of left to right shunt jet by aortic root angiography, our classification system describes 4 types of RSVA. The patients were divided into: type I, window-like, 56.7% (n = 17); type II, aneurysmal, 16.7% (n = 5); type III, tubular, 16.7% (n = 5); and type IV, other rare conditions, 10.0% (n = 3). In the new classification system, window-like shape of RSVA is the most common pattern. Three patients with rare angiographic shape were identified in type IV. One of them presented with a giant RSVA from RCS to RA and the other 2 patients with angiographic features of long and funnel shape.

Rupture of aneurysm most often originated from the RCS in each type of RSVA. Rupture into the RA was identified in 53.3% of cases and RV in 46.7%. No cases exhibited rupture into the left ventricle or pulmonary artery. The most frequent coexistence of RSVA is VSD.
which has been reported to be between 26% and 55% [22,23]. AR is the second most common associated lesion and occurs in 20–30% of RSVA patients [24]. In our study, perimembranous VSD was found in 6 patients (20%). Four of them were in type I and the other 2 were in type IV. Three patients associated with mild and moderate AR were enrolled in our study. Patients combined with subarterial VSD or severe AR should be converted to surgery.

Occluder selection in each type

Usually interventional cardiologists select occluders to match the anatomy of lesions according to their intuition. Our classification system is on the basis of the angiographic shape which could link each RSVA type to a specified occluder.

In our series, 56.7% patients were in type I and 16.7% were in type II. Patients in types I and II were treated with small-waist double-disk VSD occluders, which had been widely used in multi-hole perimembranous VSD with aneurysm in China [25]. The small-waist matches the entrance diameter of window-like and aneurysmal RSVA. Its left side can cover all the left inlets completely with little interference on aortic valve function. The shape of this occluder recovers well after release and does not occupy enough space to affect aortic valve function. In patients with tubular shape we preferred a muscular VSD occluder with a long connecting waist. In our study, 5 patients were in tubular shape (16.7%) and 5 muscular occluders were implanted successfully. No de novo residual shunts, AR, or hemolysis occurred immediately or during the follow-up period.

Moreover, there were some rare presentations of RSVA unified into type IV. In 3 cases with rare angiographic features, we selected individual occluders according to the angiographic shape. We deployed one small-waist double-disk occluder to close the giant RSVA from RCS to RA successfully [26]. Regrettfully, we failed in the other 2 patients with angiographic features of long and funnel shape. Both of them had coexistent VSD and one combined with moderate AR. Percutaneous closure of RSVA was undertaken using a muscular VSD occluder combined with a small-waist double-disk occluder and a muscular VSD occluder, respectively. Moderate residual shunt occurred immediately and hemolysis developed 4 h and 8 h after percutaneous closure. Occurrence of hemolysis might be due to the unsuitable selection of occluders and high pressure flow from the VSD. With moderate residual shunt during the procedure, we should withdraw the occluders and convert the patients to surgery immediately. Since there was no proper occluder for the long and funnel RSVA, surgical repair might be more suitable. Moreover we have been designing a special occluder shaped like a brandy wine glass that might be suitable for the long and funnel shape (Fig. 4).

An algorithm for the selection of occluders

Based on this new angiographic classification, we were able to propose a meaningful algorithm for the selection of occluders (Fig. 5). For patients with window-like and aneurysmal shape,
small-waist double-disk occluders were advised. In patients with tubular RSVA, we preferred the muscular VSD occluder with a long connecting waist. In some rare conditions, we should select individual occluders according to the angiographic shape of RSVA. If there was no proper occluder, surgical repair might be more suitable. Furthermore, in our opinion, designing individual occluders is also important for RSVA patients with some special angiographic features.

Limitations

Our study proposed a new angiographic classification and demonstrated significant benefits of percutaneous closure of RSVA, nevertheless it had some limitations. To the best of our knowledge, the present series is the largest study of patients with this rare abnormality undergoing percutaneous closure. However, the sample size of 30 cases from 5 institutions was not large enough. Besides, our classification is proposed according to the retrospective review of angiographic features. We need more cases to refine the classification system and manifest its effect on the selection of occluders in RSVA patients.

Conclusions

Based on angiographic findings of patients undergoing percutaneous closure, we proposed a new classification for RSVA. Our angiographic classification system is simple for understanding the angiographic morphology of RSVA and practical for the selection of occluders. We would like to undertake a large, prospective study to refine the classification system and better understand how this classification improves the management in individual patients.

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References

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