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Original article

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 selected. 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.

Introduction

Ruptured sinus of Valsalva aneurysm (RSVA) is a rare cardiac abnormality, usually caused by a congenital deficiency of muscular and elastic tissue in the aortic wall of the sinus of Valsalva [1]. The incidence is higher in Eastern than Western populations [2–4]. The majority of aneurysms originate in the right coronary sinus (RCS) or non-coronary sinus (NCS) and rupture into the right ventricle (RV) or right atrium (RA) [5–7]. Surgical repair has been the mainstay of therapy with relatively low mortality [8,9]. However, some problems limit the application of surgical repair, for instance, severe hemodynamic instability and patch leak require a second operation [10]. Successful percutaneous closure has been increasingly reported, mainly as case reports or small series [11–14].

To date, the formal classification system for RSVA, which was proposed by Sakakibara and Konno [15] has been widely used in clinical surgery. It describes 4 types of RSVA according to the coronary sinus affected and the site where they rupture. Another important classification system proposed by Ring [16] is a hierarchical scheme based on location and acuity (ruptured or

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non-ruptured) with a modifier added for the etiological factor. With emphasis on the origin and rupture site, Sakakibara's and Ring's classifications are anatomical classifications and not practical for percutaneous closure.

Therefore, this study was designed to introduce a new classification system according to the shape of left to right shunt jet by aortic root angiography, which we believed would help toward the better understanding of angiographic morphology of RSVA and facilitate the selection of occluders for percutaneous closure.

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, RA, 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 1
Classification systems for RSVA.

Sakakibara classification	Angiographic classification
I: RCS to the RVOT below the pulmonary valve	I: Window-like type
II: RCS to RV infundibulum in the supraventricularis crest	II: Aneurysmal type
III:	III: Tubular type
IIIa: RCS to the RA	
IIIv: RCS to the RV at membranous ventricular septum	
IV: NCS to the RA	IV: Other rare conditions

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.