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Percutaneous occlusion of ruptured Valsalva sinus aneurysms into the right ventricle

Oclusão percutânea de aneurisma roto do seio de Valsalva para dentro do ventrículo direito

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ABSTRACT – The authors report on two patients whose congenital sinus of Valsalva aneurysms had ruptured into the right ventricle and were successfully occluded percutaneously. Three different devices were implanted using two different approaches. Percutaneous closure warranted that one patent ductus arteriosus occluder through a transvenous antegrade approach and that one coil plus one ventricular septal defect occluder were implanted through an arterial retrograde approach. The procedures were accomplished without complications, the devices were well positioned at angiography and echocardiography views at the end of the interventions. At 6-month follow-up patients were asymptomatic, with devices in stable position without residual shunt.

Descriptors: Sinus of Valsalva; Therapeutic occlusion; Congenital abnormalities

RESUMO – Os autores relatam dois casos de pacientes com aneurismas congênitos do seio de Valsalva, que romperam para dentro do ventrículo direito e foram ocluídos por via percutânea, com sucesso. Três dispositivos diferentes foram implantados por meio de duas abordagens diversas. O fechamento percutâneo justificou o implante de um dispositivo ocluidor de canal arterial persistente, com abordagem transvenosa anterógrada, e de um *coil* mais um ocluidor de comunicação interventricular, por via arterial retrógrada. Os procedimentos foram realizados sem complicações. A angiografia e a ecocardiografia, ao final das intervenções, demonstraram dispositivos bem posicionados. Os pacientes estavam assintomáticos, com os dispositivos em posição estável, sem *shunt* residual, após 6 meses de seguimento.

Descritores: Seio aórtico; Oclusão terapêutica; Anomalias congênitas

INTRODUCTION

Congenital sinus of Valsalva aneurysm (SVA) is a rare condition with higher prevalence in Asian compared to Caucasian populations (1.2 to 4.5% and 0.14 to 1.5%, respectively).¹ SVA may also be acquired through processes causing the walls of the aorta to weaken, such as syphilis, tuberculosis, infective endocarditis, trauma and connective tissue diseases. Congenital aneurysms are more common than acquired aneurysms and arise from defects in the sinus of Valsalva aortic media. Such defects reflect incomplete fusion of the distal bulbar septum with atrioventricular cushions, resulting in incomplete fusion of the sinus of Valsalva media and aortic annulus fibrous.^{2,3} Congenital SVA may be associated with several cardiopathies, primarily ventricular septal defect (VSD).² Ruptured SVA is a rare cause of shunt connecting the aorta to any of the cardiac chambers.

Percutaneous occlusion of a ruptured SVA was first performed by Cullen et al. in 1994, using a Rashkind® device (USCI Angiographics, Tewksbury, MA, USA).⁴ Other devices were later developed, the most widely used being Amplatzer™ prostheses (St. Jude Medical, St. Paul, MN, USA).

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CASE REPORTS

Two 49-year-old Caucasian patients presenting with ruptured right coronary SVA protruding into the right ventricle were submitted to transthoracic echocardiography and cardiac catheterization prior to percutaneous occlusion. Procedures were carried out under general anesthesia via the percutaneous femoral approach and monitored using transesophageal echocardiogram (TEE) and angiography. Patients were followed up (clinical examination and echocardiography) for 18 months after the procedure. Both had NYHA class II heart failure, left cardiac chamber volume overload and a small perimembranous VSD unrelated to ruptured aneurysm site. The first patient also presented with mild central aortic regurgitation.

Angiographic images of the left ventricle (LV) were obtained in left anterior oblique and cranial (60° and 35°, respectively) views; images of the aorta were obtained in left and right anterior oblique views, before and after occlusion device deployment. Occlusion device selection was based on prosthesis availability and angiographically determined maximum aneurysm and neck width. In both cases, aneurysms were catheterized via the aorta using a 5F

Judkins right coronary catheter and hydrophilic guide-wire, and later replaced with an 80cm-long 7F sheath.

Patient 1

The ruptured SVA had anterior and posterior orifices. Following right ventricle (RV) catheterization through the 6mm-wide posterior orifice via the aorta, an arteriovenous loop was prepared with extra stiff 0.035" wire. The sheath was then advanced towards the aorta through the femoral vein, and an 8x6mm patent ductus arteriosus occluder (Cera™, Liftech Scientific CO Ltd, Shenzhen, China) deployed. Sheath advancement through the right ventricular outflow tract (first device deployment) and following RV catheterization through the 4mm-wide anterior orifice was extremely difficult; therefore, the Judkins catheter was replaced by a sheath. A 5-mm symmetric Cera™ perimembranous VSD occluder was implanted. Post-implantation aortography and TEE images revealed proper device position and lack of residual shunt. Right aortic cusp mobility was mildly reduced, with no significant increase in systolic flow velocity or aortic regurgitation severity (Figures 1 and 2).

Figure 1. Patient 1: (A) Congenital sinus of Valsalva aneurysm with two orifices (arrows); (B) two implanted occluders (OCL) (arrows); (C) color Doppler showing two implanted OCLs with no residual shunt, non-coronary sinus calcification and mild central aortic regurgitation (AR); (D) three-dimensional echocardiographic image showing two implanted OCLs. Ao: aorta; AoV: aortic valve, CALC: calcification; LA: left atrium; MV: mitral valve; LV: left ventricle; RA: right atrium; RV: right ventricle.

Figure 2. Patient 1. (A) Aortography (left anterior oblique view) showing both orifices of a ruptured congenital sinus of Valsalva aneurysm (arrows); (B) left anterior oblique view showing right ventricle opacification and the pulmonary artery during aortography; (C) right anterior oblique view showing implanted (arrow) patent ductus arteriosus occluder; (D) right anterior oblique view showing implanted patent ductus arteriosus occluder and perimembranous ventricular septal defect occluder (PMVSDO; arrow); (E) end-procedural aortography (left anterior oblique view) showing lack of residual shunt. RCSVA: right coronary sinus of Valsalva aneurysm; PA: pulmonary artery; RV: right ventricle; PDO: patent duct occluder.

Patient 2

Following catheterization of the 5mm-wide ruptured ASV orifice, the catheter was replaced by a sheath and a 9x6mm PFM® Nit-Occlud® device (Cologne, Germany) deployed via the arterial route. Angiography and TEE images revealed minimum residual shunt immediately after device deployment. Color doppler performed within 15 minutes of anticoagulation reversal revealed flow resolution. However, an undiagnosed 1.5 mm-wide VSD was noted (Figures 3 and 4).

Interventions were uneventful and both patients discharged the following day. Antiplatelet agents were prescribed for 6 months, including acetylsalicylic acid 325mg and clopidogrel 75mg; bacterial endocarditis prophylaxis was maintained indefinitely due to the presence of small VSD.

Six-month follow-up three-dimensional TEE revealed proper device position, no residual flow and cardiac chamber dimensions within normal ranges. At 18-month follow-up patients remained asymptomatic, with no echocardiographic abnormalities.

Figure 3. Patient 2. (A) right aorta/ventricle shunt (Ao-RV); (B) device implanted into ruptured congenital sinus of Valsalva aneurysm (arrows); (C) lack of residual shunt (red arrows); (D) small ventricular septal defect (white arrow); (E) three-dimensional image of a device implanted into ruptured congenital sinus of Valsalva aneurysm (arrows). LA: left atrium; LV: left ventricle.

Figure 4. Patient 2. (A and B) Aortography (left and right anterior oblique views respectively) showing ruptured congenital sinus of Valsalva aneurysm orifice (arrows); (C) Nit-Occlud® implanted into the orifice (arrow); (D) aortography showing minimal residual shunt through the device (arrow). RV: right ventricle.

DISCUSSION

In a review of eight published series involving 418 patients with SVA, Ott et al. found different reported incidences regarding affected sinuses of Valsalva, as follows: right coronary sinus (RCS), 65.5%; non-coronary sinus (NCS), 28% and left coronary sinus (LCS), 5.5%.⁵ Aneurysm incidence differences among the three sinuses of Valsalva may reflect underlying pathogenesis. The larger the contact surface between the aortic media with infundibular septum, the more common the malformation of both structures and the higher the odds of SVA formation. Aneurysm rupture rates also differ according to affected sinus (RCS, 60%; NCS, 42% and LCS, 10%), according to a Texas Heart Institute study.⁶

Sakakibara et al. classified congenital SVA into four types, according to origin and protrusion cavity: (I) left portion of RCS, RV outflow tract just below the pulmonary cusps; (II) central portion of RCS, supraventricular crest; (III) posterior portion of RCS, RV below the septal leaflet of the tricuspid valve, (IIIv) or right atrium (RA) next to the commissure of the septal and anterior leaflets of the tricuspid valve (IIIa) and (IV) right portion of the non-coronary sinus, RA next to the septal leaflet of the tricuspid valve.⁷

In a modification of the classification proposed by Sakakibara et al., Xin-Jin et al. included only congenital aneurysms and assumed LCS aneurysms to be acquired.⁸ Authors have also observed that NCS aneurysms may protrude and/or rupture into the RA or RV and mentioned exceptional cases of SVA rupture into the pulmonary artery (PA) and left cardiac chambers or pericardium. Hence, from a protrusion or rupture site perspective, types I, II, IIIv and IIIa of the modified classification correspond to protrusion or rupture sites described in the Sakakibara et al. classification. Type IV drains into the RA wall while type V protrudes into other structures. Cases described in this study are of congenital etiology and therefore correspond to type IIIv of Sakakibara-Konno and Xin-Jin classifications. In both cases, SVA was associated with a small VSD below the crest.

Congenital SVA unrelated to other congenital cardiopathies are often asymptomatic, with clinical manifestations triggered by complications (rupture or endocarditis in particular). However, systemic embolism,⁹ left main coronary artery compression,¹⁰ and RV or LV outflow tract obstruction have been reported.¹¹ The most common complication is rupture into right cardiac chambers (RV, 60%; RA, 29%), while left cavities are seldom affected (left atrium, 6%; LV, 4%). Rupture into the PA or pericardium (1%)^{5,12} is extremely rare.

In 2014, Liu S et al. published a study comparing one group of 20 patients with ruptured SVA submitted to surgical occlusion (SO), and one group of 15 patients submitted to percutaneous occlusion with LEPU® prosthesis (Shanghai Shape Memory Alloy Ltd, Shanghai, China).¹³ Anatomical and clinical features did not differ signifi-

cantly between groups. Success rates corresponded to 95% and 93.3% (percutaneous occlusion and SO, respectively, $p=0.68$). Percutaneous occlusion was associated with one complication (persistent hemolysis requiring surgery in one patient), whereas three complications were described in the SO group (one death due to low cardiac output; one case of pericardial effusion requiring pericardiocentesis within 2 months of initial intervention and one case of residual shunt).

In a 2015 literature review, Kuriakose et al. described 12 series and 25 case reports involving 136 patients with ruptured SVA submitted to percutaneous occlusion.¹ Amplatzer® nitinol occluder or similar occlusion devices were implanted in 110 (95.6%) patients, with 95.6% cumulative success rate and no deaths. Device deployment could not be accomplished in two cases and device removal was required in another two (severe aortic regurgitation and ischemic ST-segment changes). Seven major complications were reported: prosthesis embolization (five cases; percutaneous removal and occlusion with a different device required in three) and persistent hemolysis requiring surgery (two cases). Approach and device selection were based on anatomical features and availability of materials.

Most studies to date reported the use of Amplatzer® occlusion prosthesis via venous approach.¹ Authors of this study believe the retrograde arterial approach to be often easier and faster to perform, since arteriovenous shunts are not required. Current devices such as vascular plugs and symmetric devices for IVC occlusion have the added benefit of allowing implantation via the venous or arterial route.

Authors of this study believe percutaneous occlusion of ruptured SVA to be safe and effective, and the therapeutic modality of choice under favorable anatomical conditions. Approach and device selection should be based on anatomical features and carried out by expert professionals. The retrograde arterial is preferred to the antegrade transvenous approach in most cases. Finally, occlusion device selection should conform to anatomical conditions and treatment strategy.

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None.

CONFLICT OF INTEREST

The authors declare there are no conflicts of interest.

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