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Transapical Closure of Left Ventricular to Right Atrial Shunt Following Endocarditis

The communication between the left ventricle (LV) and the right atrium (RA) (Gerbode defect) is a very rare congenital or acquired defect of the septum dividing the right heart from the left heart. In recent years, percutaneous closure with a device has become a therapeutic option to traditional surgical repair. It can be performed using the transfemoral approach and a second option is the transapical approach.

We report the case of a 37-year-old female patient, with a history of two cardiac surgeries: closure of an ostium primum atrial septal defect at 4 years of age, and prosthetic mitral valve replacement (disc valve) for heart failure due to eft, at the age of 6 years. She presented with late prosthetic valve endocarditis due to S. viridans during her first pregnancy, which was medically treated. At the age of 32 years, the patient was diagnosed with asymptomatic acquired Gerbode defect. She presented dyspnea on moderate exertion at the age of 37. Physical examination revealed a grade 3/6 systolic murmur at the left parasternal edge that radiated to the right of the sternum. It also revealed c-v wave in the jugular venous pulse and pulsating liver as seen in tricuspid regurgitation. The ECG revealed biventricular hypertrophy and right atrial enlargement, whereas the chest x-ray showed cardiomegaly and mechanical prosthesis in mitral position.

The echocardiography showed marked biventricular enlargement, and the color Doppler revealed systolic flow from the LV to the RA, with improved visualization in the tranesophageal echocardiography (TEE) showing the defect at the level of the anterior membranous septum below the mitral prosthesis, with flow towards the RA. Three-dimensional (3D) echocardiography images showed that defect directly, and allowed to estimate its borders (Figure 1).

Percutaneous closure of the LV to RA shunt from the femoral artery was decided. In the catheterization laboratory, under general anesthesia, with TEE guidance and accessing via both femoral arteries, a guidewire could be introduced with difficulty through the defect but the catheter with the device could not be positioned. The device had to advance from the femoral artery through the aorta, the arch, and from the LV apex make a countercurve to get through the orifice between the ventricular septal defect (VSD) patch and the atrial septum. Since it was more rigid than the guidewire, it could not be positioned properly. The procedure was ended without complications.

In that situation, the transapical approach was chosen. A submammary left anterior thoracotomy through the sixth intercostal space was performed under general anesthesia, and a purse string suture was performed for LV puncture. A 7 Fr introducer sheath...
was guided by transepicardial echocardiography to define the puncture site and with 3D TEE for guidance during the procedure. Right femoral vein puncture to access the Gerbode defect was also performed, forming a loop with the transapical access. A pigtail catheter was advanced through the left femoral artery into the LV for contrast injections. An angled Glide guidewire was used in the transapical approach. Once the 0.035 Glide guidewire was passed through the LV to the RA defect, the guidewire was tunneled in the RA, and extracted forming a loop.

The Glide guidewire was advanced into the LV through the shunt for the implantation of the 12-mm Amplatzer Vascular Plug II device from the RA, deploying the retention disc in the LV without interfering with the aortic and mitral valves or the course of the shunt. Follow-up angiography revealed absence of flow through the device (Figure 2).

The outcome was asymptomatic and the patient leads a normal life.

The septum dividing the right heart from the left heart has an interatrial portion, an interventricular portion, and a small segment between the RA and the LV due to the more apical insertion of the tricuspid valve compared to the mitral valve.

The defect in question was described by Gerbode in 1958. (1) Two types of defect have been identified: congenital (usually associated with mitral valve defects) and acquired (secondary to valve surgery or post-endocarditis). (1) Acquired causes secondary to trauma, myocardial infarction and repair of VSD have also been described. In the case of the patient we report here, it was interpreted as sequelae of prosthetic valve endocarditis.

One of the most interesting topics for discussion on this condition is its difficult diagnosis. Clinically, it is similar to a VSD with tricuspid regurgitation. Several authors refer to the Gerbode defect as an echocardiographic pitfall. It is difficult to visualize the shunt in only one echocardiographic axis, since it courses in two different planes. (3) This septal defect should be suspected when an eccentric, high-velocity systolic jet is present in the RA, simulating tricuspid regurgitation, but originating in the septum. It is usually diagnosed by TEE. In dubious cases, some authors recommend magnetic resonance imaging to determine the location and size of the shunt. (4)

Since its discovery in 1958 to the present, surgical closure has always been the solution. A bovine or autologous patch is used, requiring extracorporeal circulation. Since our patient was a young woman with two previous cardiac surgeries, the first option was the percutaneous closure with Amplatzer device via femoral artery and aortic approach. However, excessive catheter angulation prevented us from implanting the occluder device percutaneously, so another option had to be chosen. (5)

The increasing use of a transapical approach both for percutaneous valve implantation as for closure of paravalvular defects led us to consider this method. Surprisingly, the transapical approach supported by TEE (6) was very simple, as opposed to the difficulties encountered with the percutaneous approach.

The Gerbode defect is an extremely rare LV to RA shunt. It usually courses asymptotically. We consider that transapical closure of acquired Gerbode defect is an option when percutaneous closure is not possible. (5) To our knowledge, this is the first description of a patient with closure of acquired Gerbode defect using transapical approach. Our experience may be useful for the management of similar patients.

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Various Tachyarrhythmias via a Mahaim Accessory Pathway. All in One

We report the case of a 53-year-old female patient with a 10-year history of hypertension and trepidation, recurrent admissions due to wide QRS tachycardia requiring pharmacological cardioversion with amiodarone on several occasions, who was diagnosed with ventricular tachycardia. Holter monitoring showed multiple episodes of symptomatic tachyarrhythmia suggestive of nonsustained ventricular tachycardia (Figure 1 A) and the ECG revealed atrial fibrillation with wide QRS and left bundle branch block (LBBB) morphology, even under treatment with atenolol, amiodarone or flecainide. Baseline ECG exhibited sinus rhythm without preexcitation. Echocardiography and myocardial perfusion were normal.

The electrophysiological study exposed irregular episodes of wide QRS tachycardia and LBBB morphology, with nodal retrograde conduction decreasing at the bundle of His level (Figure 1 B). With incremental atrial overstimulation, progressive ventricular preexcitation with LBBB morphology was evidenced, associated with smaller increase in the A-delta interval than in the AH interval. Atrial extrastimuli at a fixed pacing train resulted in higher level of pre-excitation with inversion of the right branch of His activation sequence, consistent with a Mahaim accessory pathway (Figure 2 A). Spontaneous antidromic tachycardia was observed, induced by ectopic beats mimicking tachyarrhythmia (Figure 2 B) or by programmed stimulation, which could not be entrained due to the interruption of the arrhythmia. In some cases, antidromic tachycardia degenerated into atrial fibrillation (Figure 2 C). There was 12/12 lead electrocardiographic correlation between the ectopic beats and antidromic tachycardia. Only nodal conduction was evidenced with right ventricular stimulation.

Following diagnosis of Mahaim accessory pathway, a mapping of the accessory pathway potential was performed during atrial stimulation with a 4-mm ablation catheter in the tricuspid annulus. The catheter was placed at hour 7 of the annulus; 50 W and 60 °C radiofrequency was performed with pre-excitation disappearance. The pathway was ectopic during radiofrequency application. Stimulation maneuvers were then carried out with no connection through the accessory pathway.

The patient made good progress with no recurrence during one-year follow-up.

This case shows all the arrhythmic episodes caused by a Mahaim accessory pathway in the same patient, including repetitive, isolated extrasystoles originated in the anomalous pathway, antidromic supraventricular tachycardia, atrial fibrillation and abnormal automatism caused by radiofrequency. All these arrhythmias disappeared after successful ablation of the accessory pathway.

Mahaim fibers are unusual atrioventricular connections that exhibit decremental antegrade conduction properties, located at the tricuspid annulus and distally inserting into the right ventricle at the fascicular level in the right branch or in the myocardium near it. These pathways cause antidromic tachycardia with wide QRS and image of left bundle branch block,