



Transcatheter closure of a large ruptured sinus of Valsalva aneurysm to right ventricle – technical challenges

Przecewnikowe zamknięcie pęknięcia dużego tętniaka zatoki Valsalwy do prawej komory – trudności techniczne

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Abstract

Sinus of Valsalva aneurysm, usually a congenital anomaly, almost always ruptures into the right sided cardiac chambers, causing a left-to-right shunt with profound haemodynamic consequences. With the availability of the current generation of devices and hardware, transcatheter closure has gradually replaced the surgical route. To date, most closures have been performed using an Amplatzer duct occluder (ADO). Here, we report the case of a 21 year-old male where a very large (18 mm) rupture of a sinus of Valsalva aneurysm from non-coronary cusp to right ventricle was closed by a percutaneous approach using a 20/18 mm Cocoon duct occluder (CDO; Vascular Innovations, Nonthaburi, Thailand).

Key words: sinus of Valsalva aneurysm, right ventricle, Amplatzer duct occluder, Cocoon duct occluder, transcatheter closure

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Introduction

A ruptured sinus of Valsalva aneurysm (RSOVA) is a rare cardiac abnormality, accounting for < 1% of all congenital heart defects. It forms due to a congenital weakness at the junction of the aortic media and annulus fibrosus [1]. Aneurysms arise from the right coronary sinus (RCS) in 70% of cases of RSOVA, from noncoronary sinus (NCS) in 29%, and rarely, 1%, from the left coronary sinus [2]. Its presentation varies from asymptomatic findings to the sudden onset of chest pain due to acute heart failure associated with a severe left-to-right shunt if communicating with the right-sided heart chambers, with rapid deterioration causing death, with a mean survival period for untreated RSOVA patients of 1–3.9 years [3]. Early surgical intervention is the treatment of choice. Although surgery is the gold standard, percutaneous transcatheter closure (TCC) has now become equally efficacious, and brings with it fewer complications.

Case report

A 21 year-old manual labourer with no other significant comorbidities presented with a history of palpitations and breathlessness on exertion of six weeks' duration. On admission, his functional status was New York Heart Association (NYHA) class III. On examination, he was found to have continuous murmurs in the right second and third intercostal spaces, with peripheral signs of aortic run-off. Electrocardiogram showed left ventricular hypertrophy with right ventricular enlargement, and chest X-ray showed mild cardiomegaly with right ventricular enlargement. Transthoracic echocardiography showed aneurysmally dilated right sinus of Valsalva which was communicating into right ventricle (RV) with continuous flow revealing a classical **windsock deformity**. Colour Doppler examination revealed a ruptured aneurysm of the right coronary sinus of Valsalva causing a large left-to-right

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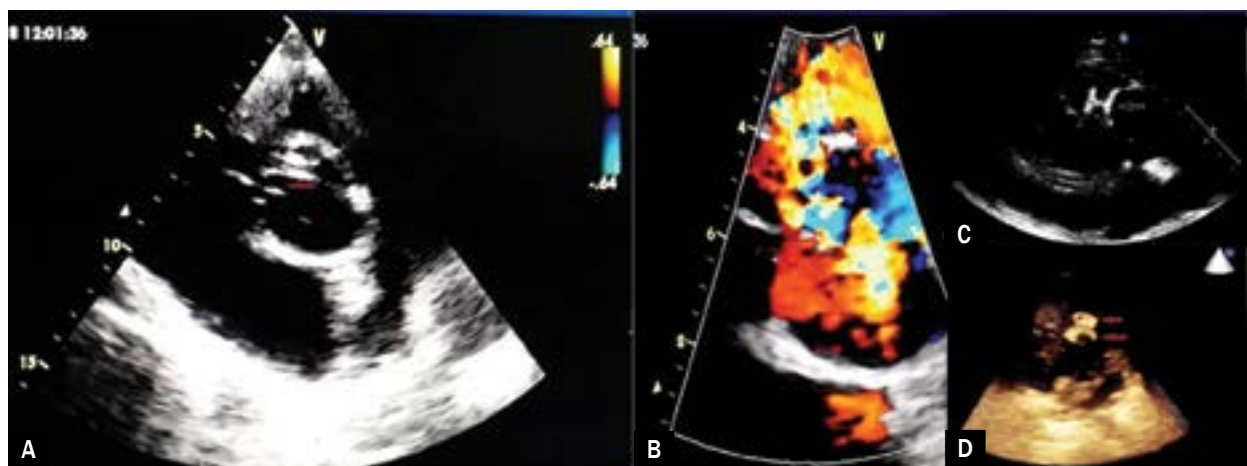


Figure 1A-D. Transthoracic echocardiography showing right sinus of Valsalva communicating into right ventricle (red arrow; **A**); colour Doppler showing the ruptured sinus of Valsalva aneurysm (RSOVA) in right ventricle (**B**); well apposed device with both the discs (2D echo – white arrow; **C**) (3D; **D**)

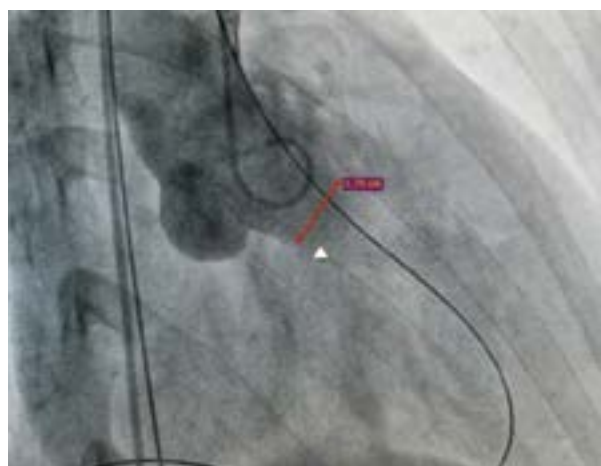


Figure 2. Aortic root angiogram showing aneurysmal sac (arrow-head) of right coronary sinus rupturing into right ventricle. The largest diameter of defect was 17 mm

shunt into the right ventricle (Figure 1). There were no associated defects such as ventricular septum defect or aortic regurgitation. Subsequently, it was confirmed on aortic root angiogram. This also revealed a mean pulmonary artery pressure of 58 mmHg and the Qp/Qs was 2.2. The communication was profiled in different views, and the largest diameter was 16–17 mm as measured by quantitative coronary analysis (Figure 2). Percutaneous device closure was planned with informed consent. The right femoral vein and artery and left femoral artery were all accessed with 6 F sheaths. Intravenous heparin (100 IU/kg) and ceftriaxone-1 gm were injected. The defect was crossed from the aortic side using a straight tip, 0.035 inch, 190 cm terumo wire (Terumo, Japan) over a 6 F Multipurpose diagnostic catheter (MP-1) (Figure 3A). Once the wire had reached into the right ventricle (RV), the catheter was further advanced into the RV. With the

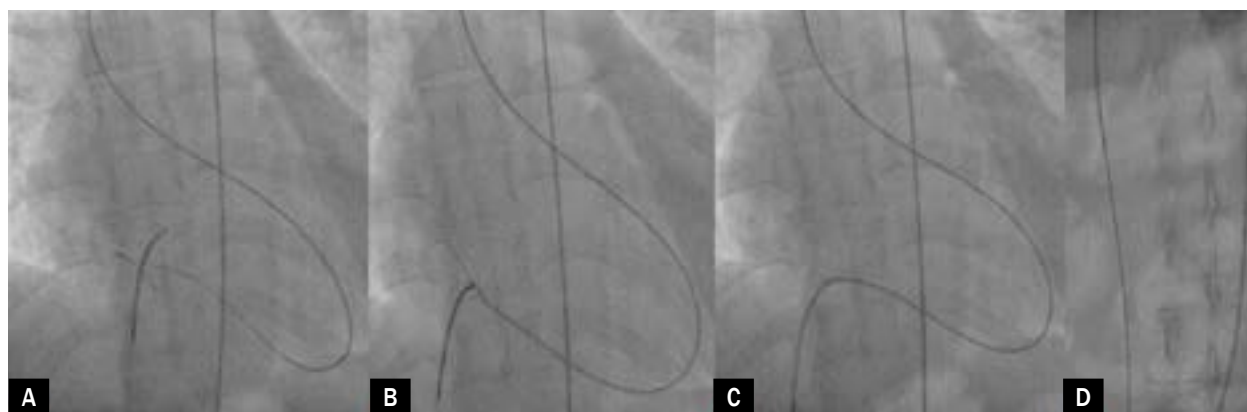


Figure 3A-D. The defect was crossed from the aortic side using straight tip terumo wire (**A**); the wire was exchanged for along terumo wire, which was pushed into right atrium where it was snared using a 2 cm EnSnare (**B-D**)

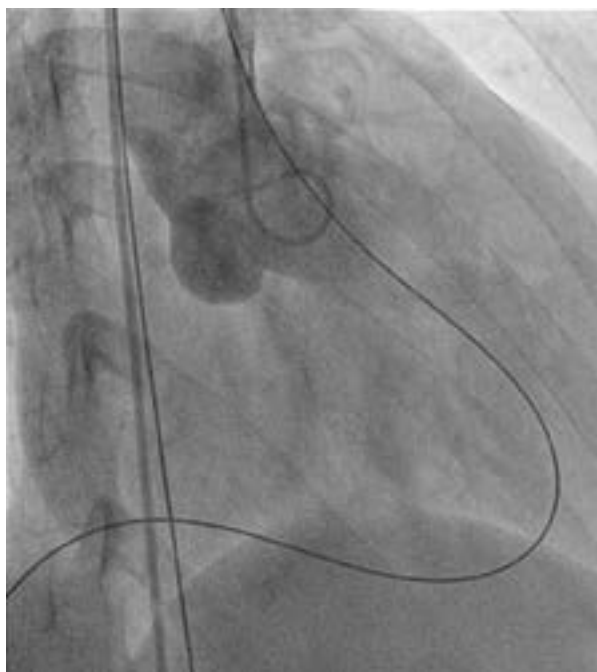


Figure 4. Arteriovenous wire loop was established

catheter kept within the RV, the wire was exchanged for a long (330 cm, 0.035 inch) terumo wire, which was pushed into the right atrium (Figure 3B). It was snared using a 2 cm EnSnare (Merit Inc, USA), and brought out

of the right femoral vein, thereby establishing a stable arteriovenous wire loop (Figures 3C, D, 4). As the largest diameter of defect was 17 mm, we chose a duct occluder device (Cocoon duct occluder, CDO; Vascular Innovations, Thailand) of 20/18 mm, meaning that its aortic segment was 2 mm larger than the diameter of the defect. As the device was compatible with a 12 F sheath, a 6 F venous sheath was pulled out and a 12 F long delivery sheath was introduced over the terumo exchange length wire from the venous side into ascending aorta across the defect. As the defect was opening into RV, and sheath being 12 F, we encountered great difficulty while negotiating it beyond the RV into ascending aorta as it tended to prolapse back to RV. The long sheath was pulled out and another 6 F MP catheter was pushed from venous side into descending aorta over the terumo wire. The terumo wire was exchanged for a 0.035 inch superstiff Amplatz wire (St Jude Med, Germany). This was pushed down to descending thoracic aorta where it was snared and brought out of the femoral artery, thereby creating another arterio-venous loop, but this time over the superstiff Amplatz wire. The long sheath was pushed over the Amplatz wire, but again the same difficulty was encountered as it tended to prolapse back to RV. Both the ends of Amplatz wire were grasped with artery forceps to keep the wire taut, and the delivery sheath was pushed with a gentle clockwise twist to facilitate its smooth delivery. With the help of this manoeuvre, it was successfully pushed beyond the ascending aorta to proximal descending thoracic

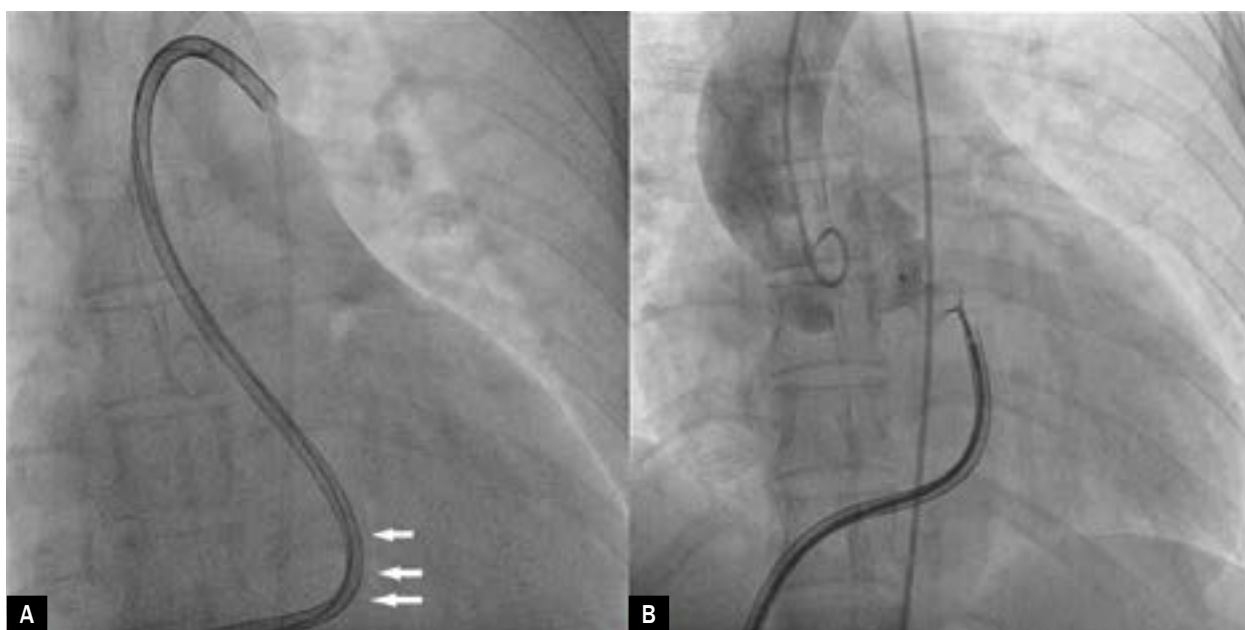


Figure 5A, B. 12 F long delivery sheath was pushed beyond the ascending aorta into descending thoracic aorta (white arrow shows the bend across right ventricular outflow tract (RVOT), **A**); aortic end of Cocoon duct occluder (CDO) was positioned to perfectly align with the aortic end of the ruptured sinus (**B**)

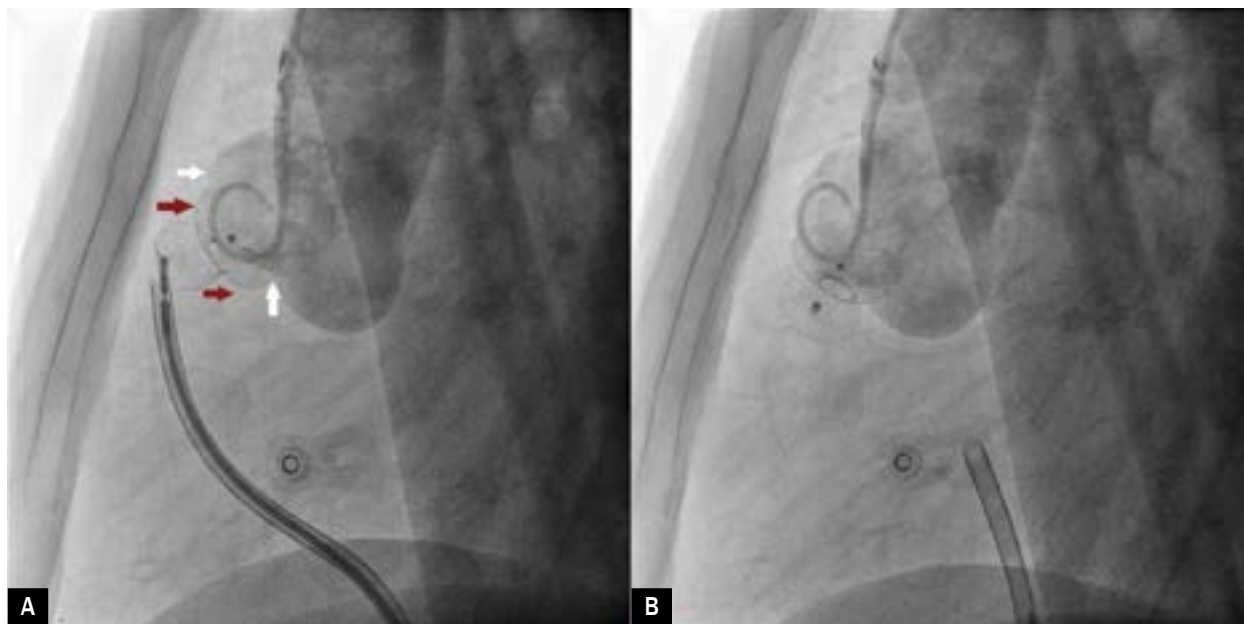


Figure 6A, B. Position of the device and complete sealing of the defect was confirmed in straight lateral position as no contrast was flowing across the device (A); device position after its deployment (B)

aorta (Figure 5A). CDO, attached to the delivery cable, was then inserted into the delivery sheath and pushed to open its aortic disc in the ascending aorta. The whole assembly was pulled back and positioned carefully until the aortic disc perfectly aligned with the aortic end of the ruptured sinus. This was ensured after injecting contrast through the pigtail catheter which was positioned into aortic sinus from the left femoral route. The position was ensured via two views: right anterior oblique with cranial angulation (RAO cranial), and straight lateral projection (Figures 5B, 6A). After confirming the precise placement, the rest of the CDO was deployed on the right side across the defect by gently pulling the delivery sheath. During this manoeuvre, a gentle traction was exerted on the delivery cable, but taking special care to ensure that the aortic disc was seated on the aortic side, with no slippage into the aneurysm. After a 10 minute delay, aortic root angiogram was again performed to ensure proper device positioning and rule out any para-device leak. The CDO was then released from the delivery cable by turning the pin-vice anticlockwise (Figure 6B). Device position was confirmed and any embolisation was ruled out on fluoroscopy (Figure 7). Echocardiogram on the following day showed no residual flow across the device (Figure 1C, D). The patient was discharged in a stable condition the next day on aspirin 75 mg once daily for the next six months.

Discussion

Sinus of Valsalva aneurysm almost always ruptures into the right side of the heart causing a left-to-right shunt with profound haemodynamic effects, especially when the rupture is sudden. Most SVAs arise from right coronary sinus ruptures in right ventricle and those arising from non-coronary sinus ruptures into RA [4]. Since the first report of a device closure of a RSOVA in 1994, there have been various case reports of RSOVA closure using a patent ductus arteriosus (PDA) occluder, a ventricular septal defect (VSD) occluder, an atrial septal defect (ASD) occluder, and a Rashkind umbrella, for example. Therefore, the transcatheter approach has become the gold standard for RSOVA if it is amenable to device closure.

The technique is similar to device closure of a perimembranous VSD, although the defect is located just above the aortic valve instead of below. Sizing of the defect can be done by angiographic measurement, and/or periprocedural echocardiography with colour Doppler interrogation which helps in device selection (2–4 mm larger than the aortic end). However, echocardiogram also gives additional information about its neighbouring structures, namely the aortic valve, tricuspid valve, and right ventricular outflow tract, and it does the periprocedural monitoring of acute aortic, and tricuspid regurgitation, and residual shunting. Some authors have also employed the

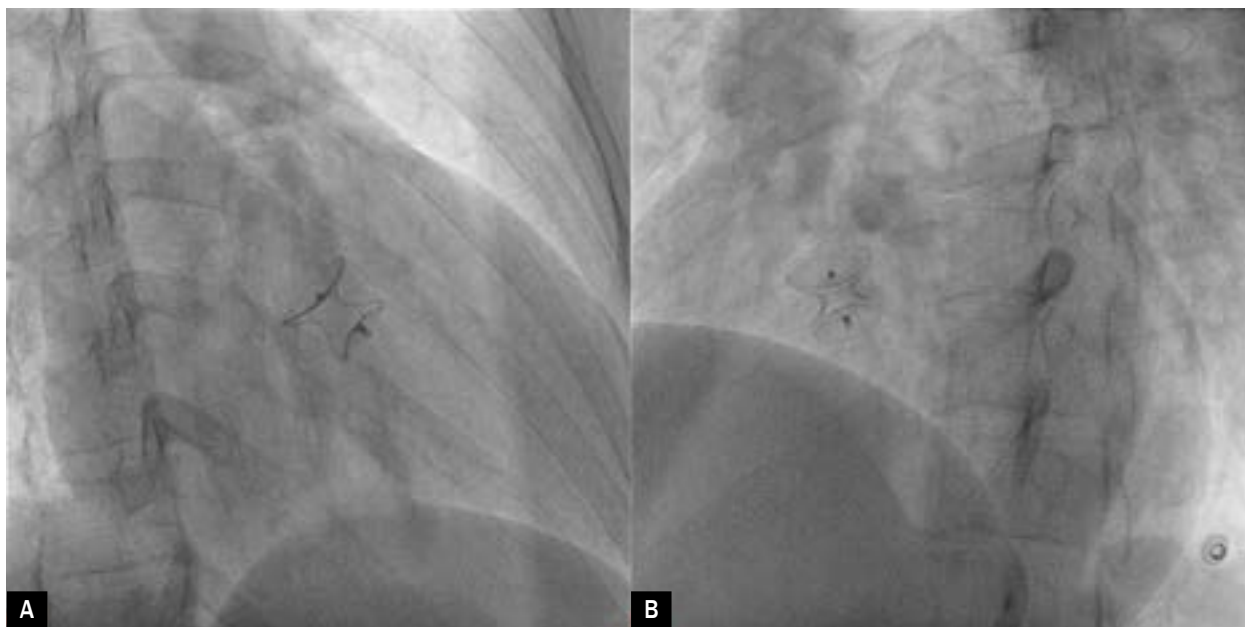


Figure 7A, B. Fluoroscopy showing stable device position (right anterior oblique view, **A**; straight lateral view, **B**)

technique of balloon sizing of the defect to choose the appropriate device [5]. In our case, the ruptured RSOVA was closed at the aortic end similarly to a 'surgeon's repair' since closure at the rupture site (exit point) would leave behind an aneurysmal sac which could rupture at another site in the long term as it remains exposed to arterial pressure [6]. Although the surgical incidence of post-procedural aortic regurgitation (AR) is 6%, most surgical incidences are based on aortography and not on the sensitive colour Doppler modality [7], it was not seen in our case. If it is moderate or more, one should not perform the device deployment as it speaks of mal-coaptation of disc with the aortic sinus. If the degree of procedure-related AR is only trivial (less than Grade I), it may not be a concern as progression of such AR, if at all, is likely to be very gradual and stable over the next 10–20 years. But it should be serially and closely monitored by transoesophageal echocardiography, and needs a longer follow-up than usual.

In our case, the sinus was rupturing into right ventricle where the antegrade delivery of the sheath (from right atrium to aorta via right ventricle) is sometimes difficult. In such a case, one should use an Amplatz superstiff wire to make a veno-arterial loop. If this does not work,

one should tightly grip both ends of the wire with artery forceps and gradually push the delivery sheath over the wire with a clockwise twist. Once it crosses the right ventricle into aorta, it should be pushed as distally as possible, at least into the proximal descending aorta, as in our case.

The Cocoon duct occluder (CDO) is a platinum-coated, self-expandable, mushroom-shaped device made from a nitinol wire mesh. Its mushroom-shaped retention skirt ensures accurate positioning at the aortic sinus of the aneurysm. Multiple poly-propylene patches sewn securely to the side of the device helps in the introduction of thrombosis, thus closes the defect. Nano platinum coating prevents nickel leeching into the bloodstream and the corrosion of the nitinol wire frame in long term implants. Platinum provides better radio-opacity, which enables easy positioning of the device in the defect.

TCC of a ruptured sinus of Valsalva can be safely and effectively carried out using CDO, and is a good alternative to surgery.

Conflict(s) of interest

The authors declare no conflict of interest.

Streszczenie

Tętniak zatoki Valsalvy, zwykle spowodowany wrodzonym defektem, prawie zawsze pęka, tworząc przetokę do prawostronnych komór serca i powodując przeciek lewo-prawy z poważnymi konsekwencjami hemodynamicznymi. Dzięki dostępności najnowszej generacji urządzeń i sprzętu przezcewnikowe zamknięcie przecieku stopniowo zastąpiło drogę chirurgiczną. Do tej pory większość zamknięć wykonywano za pomocą urządzenia zamykającego (zapinki) typu Amplatzer (ADO, *Amplatzer duct occluder*). W niniejszej pracy opisano przypadek 21-letniego mężczyzny, u którego metodą przezskórną zamknięto pęknięcie bardzo dużego (18 mm) tętniaka niewieńcowej zatoki Valsalvy do prawej komory za pomocą urządzenia *Cocoon duct occluder* 20/18 mm (CDO; Vascular Innovations, Nonhaburi, Tajlandia).

Słowa kluczowe: tętniak zatoki Valsalvy, prawa komora, *Amplatzer duct occluder*, *Cocoon duct occluder*, zamknięcie przezcewnikowe

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