

Case report

Percutaneous Atrial Septal Defect (ASD) closure technique in case of association with an azygos continuation of the inferior vena cava “case report”

Nassime Zaoui*, Amina Boukabous, Nadhir Bachir, Ali Terki and Nabil Irid

Cardiology Department, Omar Yacef Draa Ben Khedda Hospital, Tizi-Ouzou Medical University, Algeria

Abstract

Introduction: Atrial Septal Defect (ASD) is the most common congenital heart disease, accessible to percutaneous closure in 90% of cases. The closure procedure is performed usually under local anesthesia and TTE by femoral access. The association of OS-ASD with an azygos continuation of the inferior vena cava is very rare (< 0.1/1000 births) making femoral access impossible. Only a few cases are mentioned in the literature, here we describe the procedure as faithfully as possible.

Important clinical finding: We present a case of a 32-years-old female candidate for percutaneous closure of OS-ASD with right cavity dilatation who present during her procedure an unusual guidewire path suspecting an azygos continuation of the inferior vena cava, confirmed by CT angiography, making impossible the closure via the femoral approach.

Therapeutic intervention: After being confronted with the categorical patient refusal of the surgery, we performed successfully the procedure; one month later; under general sedation by internal jugular approach. We finished with manual compression before extubating the patient.

Outcomes: The follow-up was favorable at the cost of a hematoma at the puncture site and brachial plexus compression, which regressed after 3 days.

Conclusion: We opted for general anesthesia and intubation to guide the procedure by TEE. We placed it in the aorta, which gave us good stability to continue successfully the procedure. We underestimated the risk of complication at the puncture site, which could have been avoided by using a vascular suture device or more prolonged compression.

Main takeaway lesson: Percutaneous closure is the reference treatment for OS-ASD. In case of is associated with an azygos continuation of the inferior vena cava, the right internal jugular vein remains a reasonable approach; it requires discussion and rigorous preparation by the whole team. The management of the puncture site in this situation remains delicate and requires great concentration.

Abbreviations

ASD: Atrial Septal Defect; LA: Left Atrium; LV: Left Ventricle; M: Month; MPA: Multipurpose A; OS: Ostium Secundum; RV: Right Ventricle; TEE: Transesophageal Echocardiography; TTE: Transthoracic Echocardiography

More Information

***Address for correspondence:** Nassime Zaoui, Cardiology Department, Omar Yacef Draa Ben Khedda Hospital, Tizi-Ouzou Medical University, Algeria, Email: nassime.zaoui@chu-brugmann.be

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Ostium secundum ASD is accessible to percutaneous closure in 90% of cases; with better results than surgery with a lower complication rate [3].

According to ESC guidelines [4,5], ASD closure is indicated (class I) in case of right ventricle overload and without pulmonary hypertension or in case of paradoxical embolism (class IIa) and contraindicated in Eisenmenger physiology (class III).

The procedure is performed under local anesthesia and TTE or general anesthesia and TEE (for the first procedures and child) through the femoral vein. A stiff guidewire is placed in a pulmonary vein through the ASD, using an MPA catheter, and allows the consecutive rise of the calibration balloon and the sheath that brings the prosthesis until the interatrial septum that it will sandwich under echocardiographic control [6-9].

After the procedure, Aspirin is maintained for 6 months with clinical and echocardiographic control at M1, M3, M6, and M12 [10].

Association of OS-ASD with an azygos continuation of the inferior vena cava, is very rare (<0.1/1000 births) [11] making femoral access impossible [12-14].

Patient information

De-identified patient-specific information: We describe the case of a 32-year-old patient with no history in whom a routine medical examination for asthenia 2 years before the current episode objectified a heart murmur motivating her orientation in cardiology for echocardiography which objectified an ASD OS of 20 mm -21 mm with right cavitory repercussions (RV at 40 mm and RV/LV ratio at 0.9, the right atrium at 20 cm²) and a systolic pulmonary artery pressure at 40 mmHg.

Faced with the desire for pregnancy and with this right cavitory impact and the existence of satisfactory edges (retro-aortic edge 9 mm, posteroinferior edge 10mm, atrioventricular edge 10 mm, upper edge 9 mm, lower cave edge 10 mm, upper cave edge 12 mm) her treating cardiologist decides to entrust her to us for percutaneous closure of this defect (Figure 1).

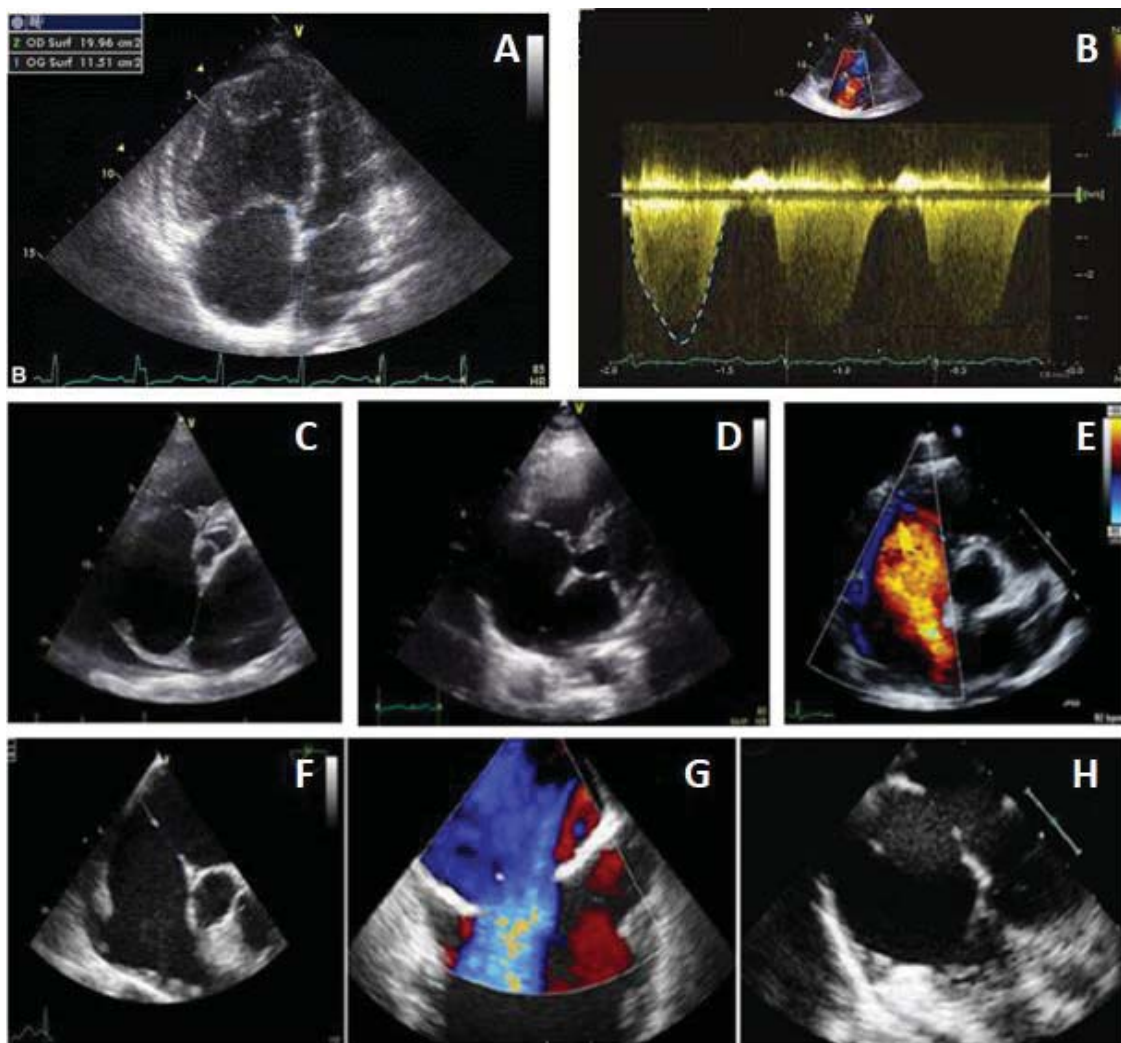


Figure 1: ASD evaluation before the percutaneous procedure. (A: Right cavities dilatation, B: Tricuspid regurgitation, C-D and E: ASD and edges in TTE, F-G, and H: ASD and edges in TEE).



Clinical findings

We started the procedure under local anesthesia and femoral vein 6Fr access, we introduced an MPA catheter, which drew an unusual path with a loop behind the right atrium to join the superior vena cava and then the right atrium.

Diagnostic assessment

Faced with this trajectory, we suspected an association with an azygos continuation of the inferior vena cava. We interrupted the procedure and requested a CT angiography that confirmed the diagnosis (Figure 2).

We discussed a surgical closure, which was categorically refused by our patient. We, therefore, performed an ultrasound of the jugular vein in the Trendelenburg position confirming its good caliber.

Therapeutic intervention

We returned to Cath Lab 1 month after, the patient was put under general sedation, intubated, and ventilated, to be able to perform TEE.

We punctured the internal jugular vein in the Trendelenburg position and then 5000 unities of heparin were injected.

We positioned the patient on the catheterization table in the usual position (head up) and we placed, in addition to the usual instrument table on the patient's right, a second instrument table at the patient's head forming an L with the first table, in order to be able to handle the delivery system in complete safety.

We started by crossing the ASD via the MPA and we positioned the 0.035” stiff guidewire in the upper then lower

left pulmonary vein, however each time the angle with which the calibration balloon cross the ASD was unfavorable and brought the 0.035” wire back into the right atrium.

We decided then to put the wire in the aorta (through the ASD, then in the LA then LV and finally in the descending aorta as far as possible) in order to increase the support. From then on, the rest of the procedure became simple.

We reintroduced the calibration balloon giving a stretched ASD diameter of 23 mm (measurement by angiography and echocardiography TTE/TEE) fixing our choice on a 26 mm prosthesis which requires a septum height of 40 mm ($26 + 2 \times 7$) that was exactly our patient's septum height.

Then, we introduced the 12 Fr sheath from the internal jugular vein and prepared the prosthesis in its chamber as described earlier in this document.

The deployment and release of the prosthesis were done under angiographic and echocardiographic control (TTE/TEE).

For that, we started by putting the delivery system through the mitral valve in order to increase the support and stabilize the system, then we pushed the prosthesis till the end of its sheath and then we pulled back everything into the left atrium (to avoid trapping the prosthesis in the mitral cords) to deploy the distal disc, brought back into contact with the septum than the proximal disc to thus sandwich the septum between the two discs (Figure 3).

After confirming the correct positioning of the prosthesis and the absence of conflict with the mitral valve, the pulmonary veins, and the superior vena cava, we released successfully the Occluder by anti-clockwise rotation of its rod (Figure 4).

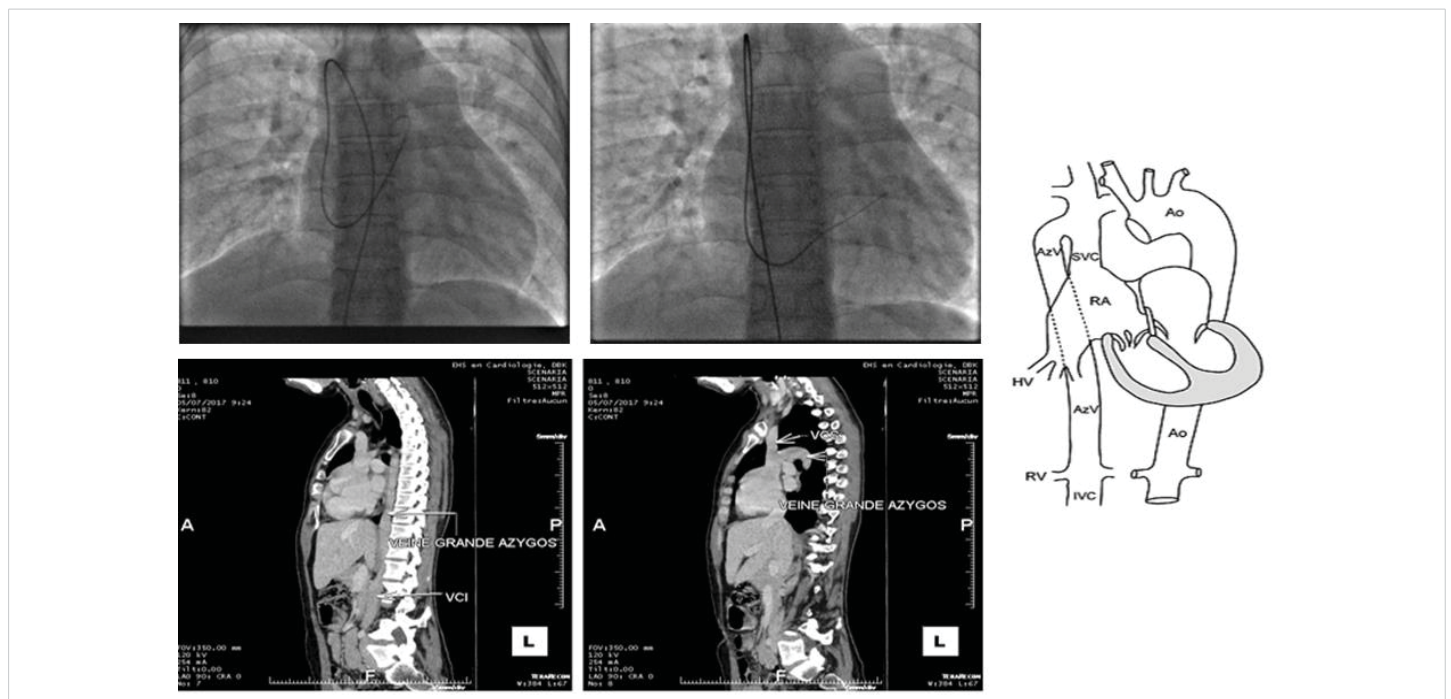


Figure 2: Azygos continuation discovered in angiography and CT.

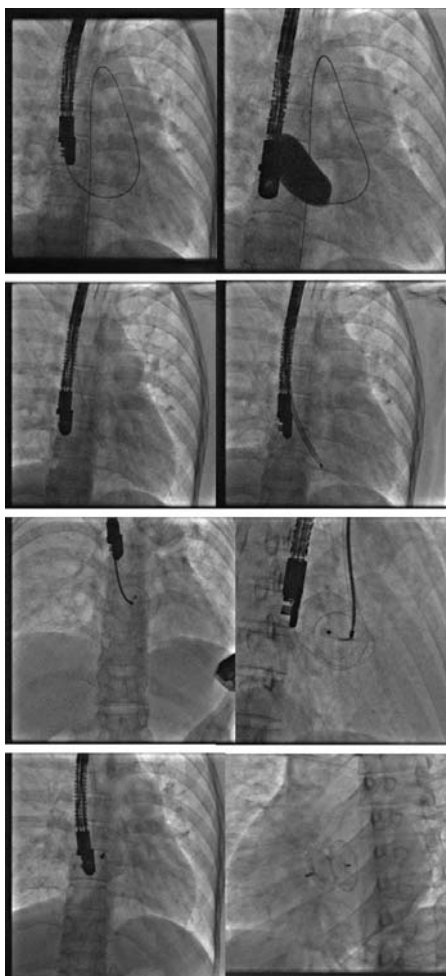


Figure 3: Procedure angiography.



Figure 4: Procedure TTE.

We considered using a femoral closure device (Femoseal©) to close the venous access after removing the delivery system, but having no experience with this device on venous access, we performed simple prolonged manual compression before waking up and extubating the patient. She was then transferred to the recovery room for 6 hours and brought back to the normal hospitalization room the same day.

Follow-up and outcomes

The follow-up was favorable in our patient at the cost of a hematoma at the puncture site and little compression on the brachial plexus with right upper limb anesthesia and paresis, which regressed after 3 days. The patient was discharged three days later under Aspirin for 6 months.

Control 7 days, 1, and 3 months after showing the total disappearance of the neurological disorders in the right upper limb, the absence of residual shunt, and the regression of the right cavity dilation.

Discussion

At first, we considered the surgical cure that is a reasonable attitude for this patient at low surgical risk, but she refused categorically, and after having considered the feasibility of percutaneous closure we changed our strategy [12-14].

We opted for general anesthesia and intubation in order to facilitate the guidance by TEE.

We were faced with a lack of support when placing the wire in the pulmonary veins as described in the classic ASD closure procedure, which is why we placed it in the aorta, and that gave us good stability to continue successfully the procedure.

We underestimated the risk of complication at the puncture site which could have been avoided by either putting a vascular suture device or more prolonged compression (but which would have required prolonging the patient's intubation).

The primary take-away lesson of this case: In case of ASD associated with an azygos continuation of the inferior vena cava, the right internal jugular vein remains a reasonable route; it requires discussion and rigorous preparation by the whole team, but also a discussion with the patient.

Learning objectives

1. To know that percutaneous management of the association “ASD and azygos continuation” is possible
2. To know how to perform this particular procedure step by step.



Patient perspective

The patient was very satisfied with the result despite the complication at the puncture site.

Informed consent

The patient consented to the sharing and publishing of her case and procedure images subject to anonymity

Ethics approval and consent to participate

The patient consented to undergo the procedure.

Consent for publication

The patient consented to the sharing and publication of data, images, and results.

Availability of data and material

The images presented during this work are available from the corresponding author upon reasonable request.

Authors' contributions

NZ was responsible for the realization of the percutaneous procedure and participated in the writing of the manuscript.

AB participated in the percutaneous procedure and the realization of echocardiography.

NB participated in the realization of the percutaneous procedure and the writing of the manuscript.

AT participated in the realization of echocardiography and the writing of the manuscript.

MB participated in the follow-up of the patient during and after hospitalization.

All authors read and approved the final manuscript.

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