Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2022.47.007488

Percutaneous Atrial Septal Defect (ASD) Closure Technique in case of Association with an Azygos Continuation of the Inferior Vena Cava "Case Report"

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ARTICLE INFO

Received: November 23, 2022

Published: November 30, 2022

Citation: Nassime ZAOUI, Amina BOUKABOUS, Nadhir BACHIR, Ali TERKI and Nabil IRID. Percutaneous Atrial Septal Defect (ASD) Closure Technique in case of Association with an Azygos Continuation of the Inferior Vena Cava "Case Report". Biomed J Sci & Tech Res 47(2)-2022. BJSTR. MS.ID.007488.

Keywords: Case Report; ASD; Congenital Heart Disease; Percutaneous Closure; Internal Jugular Vein

Abbreviations: ASD: Atrial septal defect; LA: Left atrium; LV: Left ventricle; M: Month; MPA: Multipurpose A; OS: Ostium secundum; TEE: Transesophageal echocardiography; TTE: Transthoracic echocardiography

ABSTRACT

Introduction

Atrial septal defect (ASD) is the most common congenital heart disease, accessible to percutaneous closure in 90% of cases. 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 few cases are mentioned in the literature with few details concerning the technique, here we describe the procedure as faithfully as possible.

Important Clinical Finding

We present a case of 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 to the categorical patient refusal of the surgery, we performed successfully the procedure; one month later; under general sedation by internal jugular approach with, however, modifications of the classic technique concerning especially the guidewire positioning for a better support. We finished by 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 were faced with a lack of support when putting the guidewire in the pulmonary veins as described in classic procedure, which is why we placed it in the aorta, 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 using a vascular suture device or more prolonged compression.

Main Take-Away Lesson

Percutaneous closure is the reference treatment for OS-ASD. In case of its associated with an azygos continuation of the inferior vena cava, the right internal jugular vein remains a reasonable approach with particularities related to the superior 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.

Introduction

Atrial septal defect (ASD) is the most common congenital heart disease; its prevalence is around 56 to 100 cases per 100,000 births [1]; with three types: Ostium primum (20-35%), ostium secundum (50-70%) and "sinus venosus and coronary sinus" (10-15%) [2]. 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 described a case of 32-year-old female with pregnancy desire, candidate for percutaneous closure of 20-21 mm OS-ASD with right cavity dilatation, pulmonary systolic pressure at 40mmHg and good edges; discovered during TTE for a heart murmur (Figure 1).

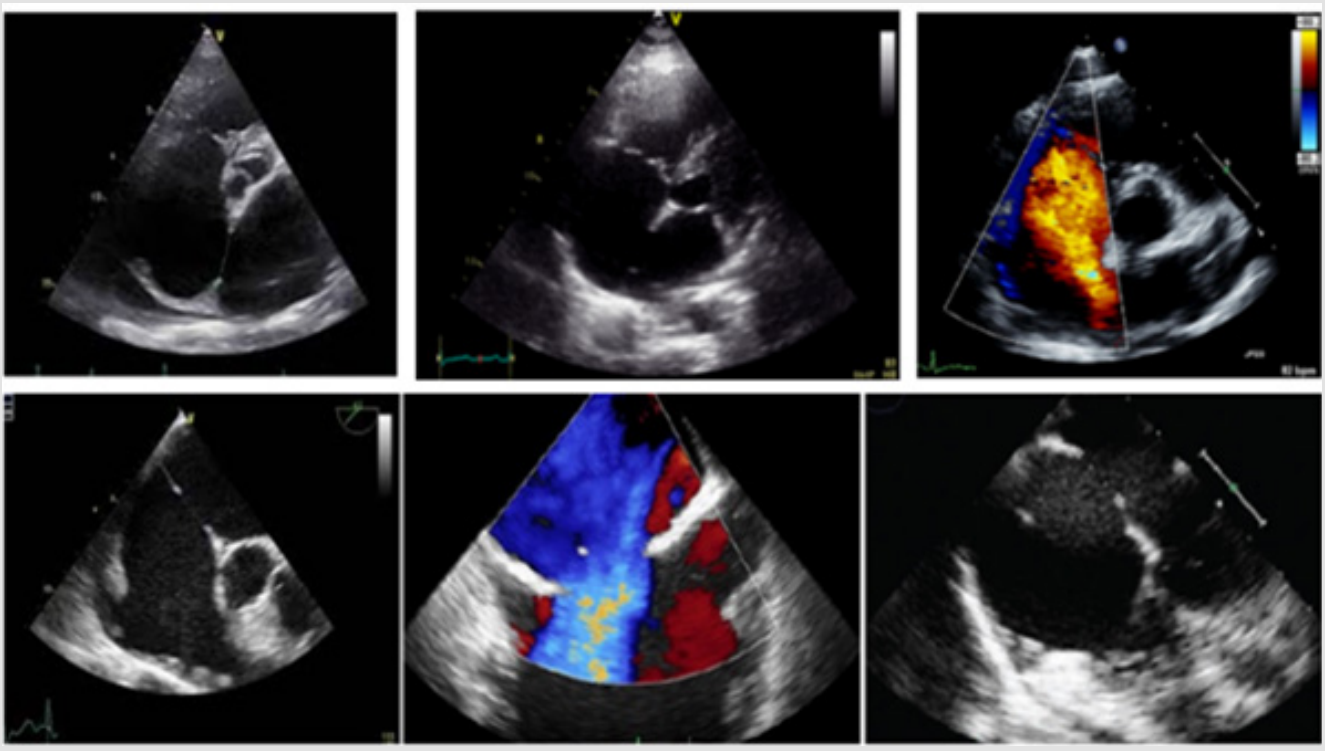

Figure 1: ASD TTE evaluation.

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, wich

was categorically refused by our patient. We therefore performed an ultrasound of the jugular vein in the Trendlenburg position confirming its good caliber.

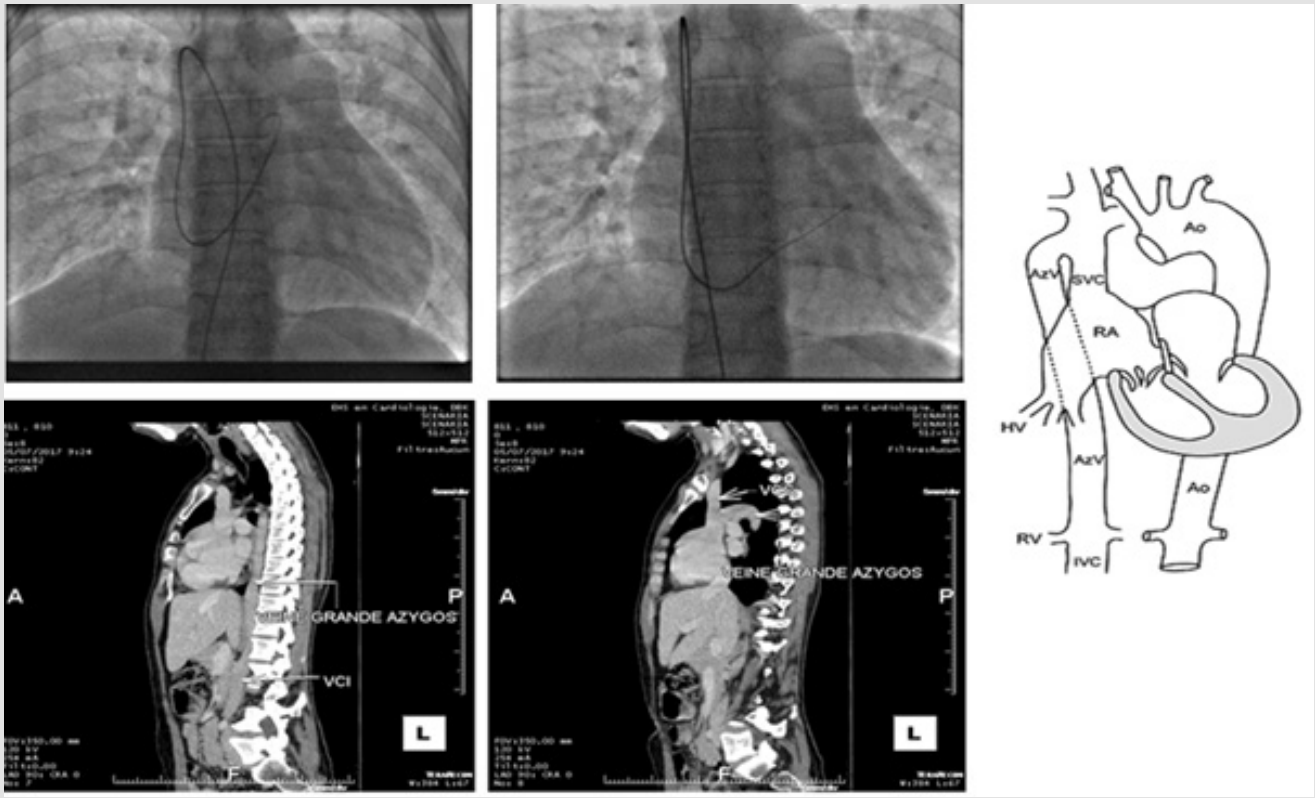

Figure 2: Azygos continuation discovered in angiography and CT.

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 was 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 + 2x7) 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 was 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 then 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). 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.

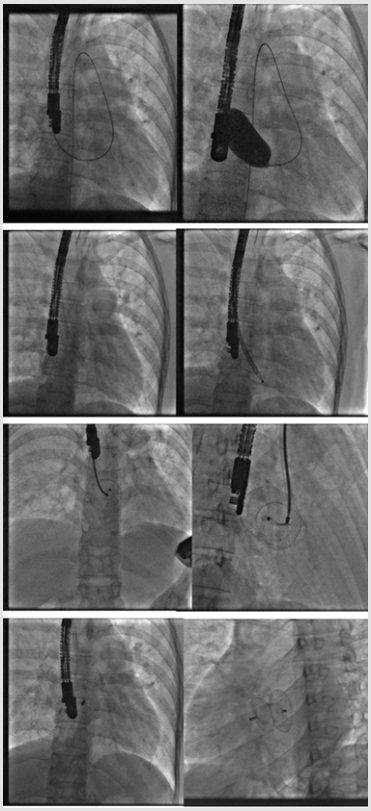

Figure 3: Procedure angiography.

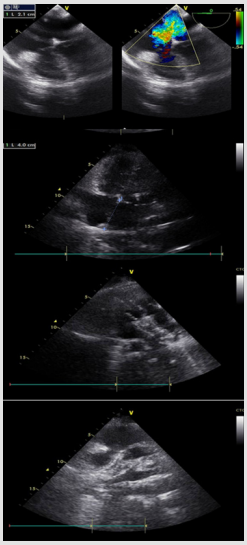

Figure 4: Procedure TTE.

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 discharged three days later under Aspirin for 6 months. Control 7 days, 1 and 3 months after, showed 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 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 pimary take-away lesson of this case: In case of ASD associated with an azygos continuation of the inferior vena cava, 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

- To know that percutaneous management of the association "ASD and azygos continuation" is possible
- 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 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 on reasonable request.

Competing Interests

The authors declare that they have no competing interests.

Funding

Not applicable

Authors' contributions

NZ was responsible for 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 in the writing of the manuscript.

AT participated in the realization of echocardiography and in 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.

Acknowledgements

We thank our paramedics who participated in the percutaneous procedure.

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ISSN: 2574-1241

DOI: 10.26717/BJSTR.2022.47.007488

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