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Case Report

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

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Keywords

- Case report
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Abstract

Introduction: Atrial septal defect (ASD) is the most common congenital heart disease, with three types: Ostium primum, secundum and sinus venosus. OS ASD is accessible to percutaneous closure in 90% of cases, which is indicated in case of right ventricle overload or paradoxical embolism and contraindicated in case of Eisenmenger physiology. 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 (less than 0.1/1000 births) making femoral access impossible and percutaneous closure unlikely. only a few cases of percutaneous closure; in this situation; are mentioned in the literature with few details concerning the technique and difficulties, here we describe the procedure as faithfully as possible with tips and tricks to solve all the difficulties encountered.

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

Therapeutic intervention: After being confronted to the categorical patient refusal of the surgery, we performed a literature review confirming that percutaneous ASD closure through the internal jugular vein is possible (07 cases described). 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 positioning of the guidewire for a better support. We finished by a simple manual compression before extubating the patient.

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

Conclusion: We described as faithfully as possible the sequence of events with all the difficulties encountered and their solutions. 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, and that gave us good stability to continue successfully the procedure. We probably 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.

Main take-away lesson: Percutaneous closure is the reference treatment for ostium secundum 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. General anesthesia is a reassuring attitude. 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; TEE: Transesophageal Echocardiography; TTE: Transthoracique Echocardiography

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] (Table 1). This procedure was first described in 1976 [1].

Table 1: Ooi yk ET AL study comparing percutaneous and surgical ASD closure [3].

Out Comes	Surgery	Percutaneous	*P*
Mortality	0	0	-
Age	4.5	5.6	<0.0001
Infection	Odds Ratio 3.73		<0.0001
Complications	Odds Ratio 6.66		<0.0001

According to ESC guidelines (Table 2) [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). After puncturing the femoral vein, a stiff guidewire is placed in the left superior 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 [6,7]. For this, prosthesis must be screwed on its rod, then brought back to its chamber and unbubbled before being slipped into its sheath [8,9] (Figure 1).

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

Association of ostium secundum ASD with an azygos continuation of the inferior vena cava, is very rare (less than 0.1/1000 births) [11] making femoral access impossible and percutaneous closure unlikely [12-14].

Patient information

De-identified patient specific information: We described a clinical case of 32-year-old female with desire for pregnancy, candidate for percutaneous closure of 20-21 mm OS ASD with right cavity dilatation, pulmonary systolic pressure at 40mmHg and good edges; discovered during an echocardiography made following a heart murmur finding (Figure 2).

Clinical findings

We started the procedure under local anesthesia, we punctured the right femoral vein with a 6Fr introducer, and then 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 ostium secundum

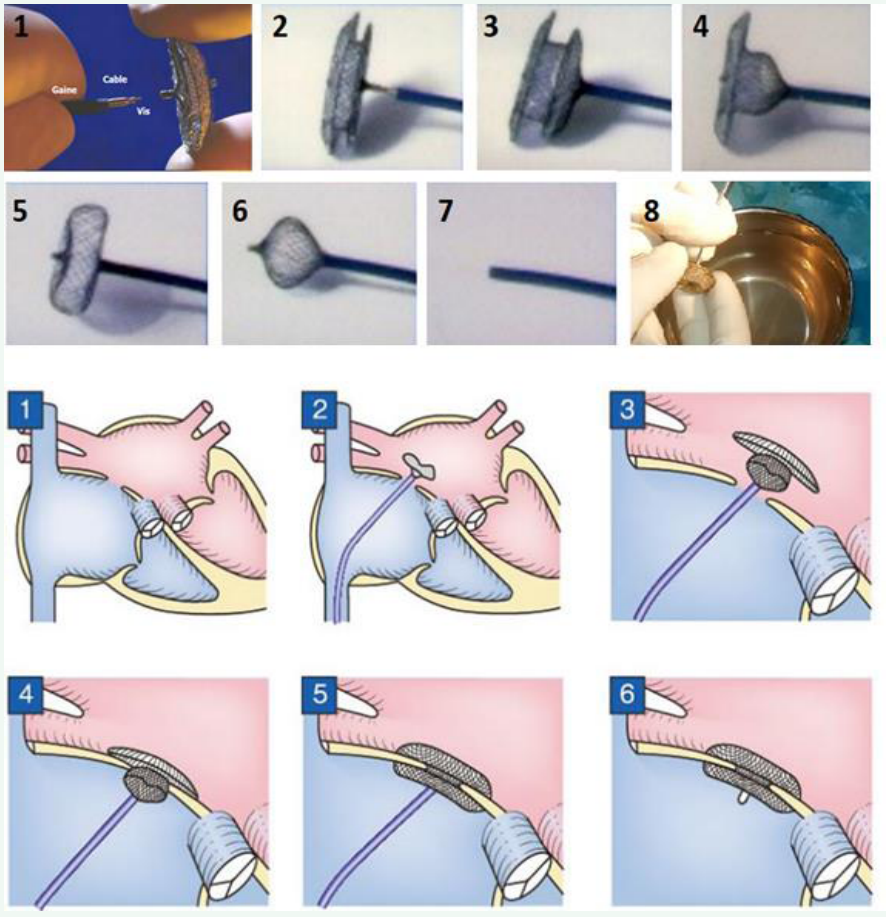

Figure 1 ASD device preparation and deployment.

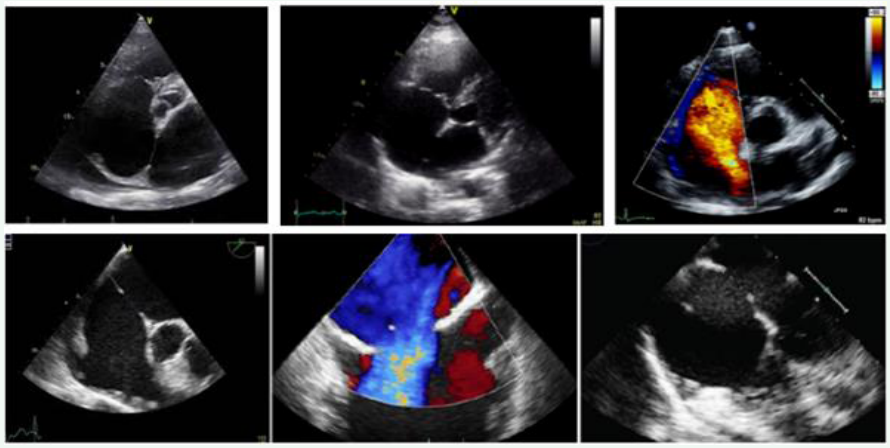

Figure 2 ASD TTE evaluation.

ASD associated with an azygos continuation of the inferior vena cava, making impossible the percutaneous closure procedure via the femoral approach. We interrupted the procedure and requested a CT angiography that confirmed the diagnosis. (Figure 3)

We discussed a surgical closure but we were confronted to the categorical refusal of the surgery by the patient. We performed a literature review confirming that access to percutaneous ASD closure through the internal jugular vein is possible (07 cases described) [12-14] .

We therefore performed an ultrasound of the jugular in the Trendlenburg position confirming its good caliber.

Therapeutic intervention

We returned to Cath Lab 1 month after the first attempt, the patient was put under general sedation, intubated and ventilated, in order to be able to guide the procedure by TTE and 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 very easily via the MPA and we positioned the 0.035'' stiff guide wire 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 at 23 mm (measurement confirmed 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.

Figure 3 Azygos continuation discovered in angiography and CT.

Table 2: ASD Closure ESC guidelines [5].		
Indication	**Class**	**Level**
Patient with significant shunt (RV volume overload) and PVR < 5 WU should undergo ASD closure regardless of symptoms	I	B
Device closure is the method of choice for secundum ASD closure when applicable	I	C
All ASDs regardless of size in patient with suspicion of paradoxical embolism (exclusion of other causes) should be considered for closure	IIa	C
Patients with PVR ≥ 5 WU but < 2/3 SVR or PAP < 2/3 systemic pressure and evidence of net L-R shunt (Qp:QS > 1.5) may be considered for closure	IIb	C
ASD closure must be avoided in patients with Eisenmenger physiology	III	C

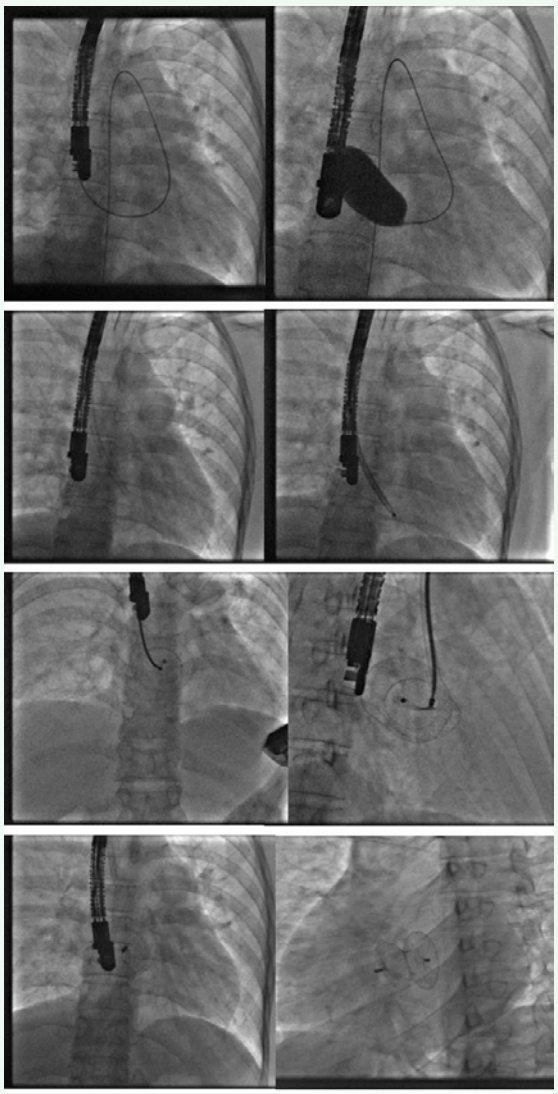

Figure 4 Procedure angiography.

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 transport, 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 4: Device positioning, deployment and release under echocardiography).

After confirming the correct positioning of the prosthesis and the absence of conflict with the atrioventricular valves (especially the mitral valve), the pulmonary veins and the superior vena cava, we released successfully the Occluder by anti-clockwise rotation of its rod (Figure 5: Device positioning, deployment and release under echocardiography).

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 chose to perform simple prolonged manual compression before waking up and extubating the patient.

The patient 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 led to right upper limb anesthesia and paresis and which regressed after 3 days. The patient discharged three days later under Aspirin for 6 months.

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

DISCUSSION

We have described as faithfully as possible the sequence of events in the patient because the cases described in the literature have not covered all the difficulties encountered during this unusual procedure.

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

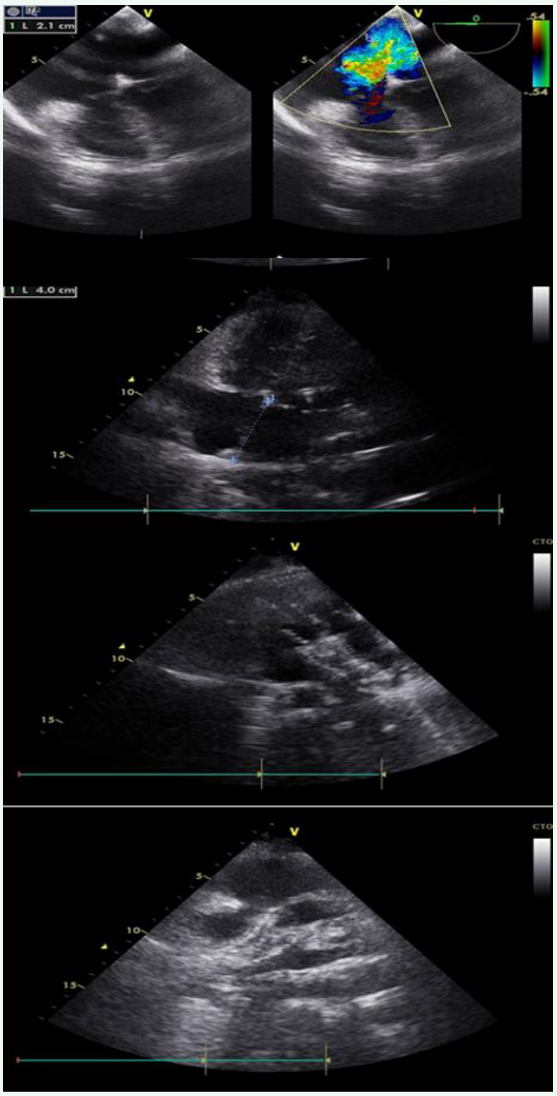

Figure 5 Procedure TTE.

We opted for general anesthesia and intubation in order to reduce stress in the Cath lab (stress of the patient and the healthcare team) and to facilitate the guidance of the procedure by TEE. We almost no longer perform TEE for ASD closure procedures; TTE alone is enough (except for children and in complex anatomies).

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 probably 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).

CONCLUSION

Percutaneous closure is the reference treatment for ostium secundum ASD; the current prostheses are well adapted [2]. The immediate and medium-term results are better than surgery with a lower complications rate [3].

The pimary take-away lesson of this case: In case of ostium secundum ASD associated with an azygos continuation of the inferior vena cava, the femoral approach is impossible, 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.

PATIENT PERSPECTIVE

The patient was very satisfied with the result despite the complication at the puncture site, she says she is delighted and reassured to have a pregnancy.

INFORMED CONSENT

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

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.

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REFERENCES

1- Van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011; 58: 2241-7.

2- Naqvi N, McCarthy KP, Ho SY. Anatomy of the atrial septum and interatrial communications. J Thorac Dis. 2018; 10: S2837-S2847.

3- Ooi YK, Kelleman M, Ehrlich A, Glanville M, Porter A, Kim D, et al. Transcatheter Versus Surgical Closure of Atrial Septal Defects in Children: A Value Comparison. JACC Cardiovasc Interv. 2016; 9: 79-86.

4- Martin SS, Shapiro EP, Mukherjee M. Atrial septal defects - clinical manifestations, echo assessment, and intervention. Clin Med Insights Cardiol. 2014; 8: 93-8.

5- Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al. ESC Scientific Document Group. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021; 42: 563-645.

6- Gabriele Egidy Assenza, Luca Spinardi, Elisabetta Mariucci, Anna Balducci, Luca Ragni, Cristina Ciuca, et al. Transcatheter Closure

of PFO and ASD: Multimodality Imaging for Patient Selection and Perioperative Guidance. JCDD. 2021; 8: 78.

7- Chan KC, Godman MJ, Walsh K, Wilson N, Redington A, Gibbs JL. Transcatheter closure of atrial septal defect and interatrial communications with a new self expanding nitinol double disc device (Amplatzer septal occluder): multicentre UK experience. Heart. 1999; 82: 300-6.

8- Harper RW, Mottram PM, McGaw DJ. Closure of secundum atrial septal defects with the Amplatzer septal occluder device: techniques and problems. Catheter Cardiovasc Interv. 2002; 57: 508–524.

9- ASD_Closing-procedure.

10- Yang MC, Wu JR. Recent review of transcatheter closure of atrial septal defect. Kaohsiung J Med Sci. 2018; 34: 363-369.

11- Boffa GM, Chioin R, Stritoni P, Daliento L, Congedo E, Sperandeo V, et al. Azygos or hemiazygos continuation of the inferior vena cava. An angiographic findings of 10 cases. G Ital Cardiol. 1980; 10: 1063-8.

12- Seshagiri RD, Patnaik AN, Srinivas B. Percutaneous closure of atrial septal defect via transjugular approach with Blockaid device in a patient with interrupted inferior vena cava. Cardiovasc Interv Ther. 2013; 28: 63-5.

13- Oliveira EC, Moura MAG, Almeida JA, Ribeiro ALP, Nascimento BR. Percutaneous closure of ostium secundum atrial septal defect using left internal jugular vein access in a child with situs inversus and absence of inferior caval vein. Cardiol Young. 2019; 29: 1310-1312.

14- Ozdemir E, Emren S V, Eren N K, Nazli C, Tokac M. Transjugular closure of ASD in a patient with interrupted inferior vena cava. Int J Cardiovasc Acad. 2018; 4: 15-18.