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Aorto-Cameral Fistula - Transcatheter Closure of Rare Complication: Case Series

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Abstract

Background: An aorto-cameral fistula (ACF) is a rare and complex cardiovascular condition involving an abnormal connection between the aorta and one of the heart chambers. It can be either congenital or acquired. While the majority of cases (73.5%) are treated with open surgery, percutaneous interventions (a less invasive, catheter-based procedure) are used in about 10% of patients. Three cases were reported where ACFs were closed using the percutaneous method. This less invasive approach is becoming more popular because it avoids many risks associated with surgery, making it a more desirable treatment option.

Cases: This report presents three unusual cases of aorto-cameral fistula (ACF) in two middle-aged women and one elderly male, all of whom experienced chest pain, palpitations, and progressive effort breathing. Two cases resulted from a ruptured sinus of Valsalva, and the third was idiopathic. In two instances, the abnormal connection was between the aortic sinus and the right atrium, while in the other, it was between the aortic sinus and the right ventricular outflow tract. Due to the lack of extensive clinical trials on ACF, treatment decisions are based on expert consensus. All three cases were successfully treated with percutaneous device closure.

Conclusion: These cases emphasize the importance of recognizing the rare presentation of aorto-cameral fistula (ACF) and demonstrate the effectiveness of aggressive, noninvasive treatment through percutaneous closure. This approach resulted in positive clinical outcomes, avoiding the need for more invasive surgical correction.

Keywords: ***Aorto-cameral fistula (ACF); Fistula; Infective endocarditis; Percutaneous device closure; Surgical correction; Right ventricular outflow tract***

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1. Introduction

Aorto -cameral fistula (ACF) is a rare but complex pathological condition characterized by the presence of blood flow between the aorta and any cardiac chamber [1]. The two main sub-groups of fistulas are congenital (coronary cameral fistulas and aorto-atrial fistula) and acquired aorto-cameral tunnels [2]. Congenital reasons accounting for around 12% of cases [3]. The prevalence of acquired causes is believed to be significantly higher and can result from a range of medical conditions, such as rupture of the sinus of Valsalva aneurysm, infective endocarditis, traumatic damage, aortic dissection, Behcet's disease, giant cell arteritis, Kawasaki disease or any vasculitis or, in rare cases, iatrogenic reasons (suture or perforation associated, such as after cardiac surgery or device closure [1,2]. The idiopathic atrial fistula of the aortic root (sinus of Valsalva), which was recently discovered by Campisi et al, is another extremely unusual defect [2]. The underlying disease needs to be understood thoroughly to enable planning the optimal therapeutic strategy. This report present a comprehensive overview of the clinical characteristics, subtypes of cases and timely aggressive management by percutaneous closure. Surgery is the choice of treatment, but isolated uncomplicated cases can be closed by percutaneous closure though it is very much challenging. Percutaneous closure can avoid many hazards of surgery so now a days it is becoming a more acceptable and demanding treatment option.

2. Case 1

A 35-years- old female, normotensive, non-diabetic, non-smoker, mother of 2 children presented with history of chest tightness along with pain & occasional breathlessness, palpitation for last 5 months. She also had a history of pedal edema 1 month back. She had no history of fever, recurrent repeated respiratory tract infection (RTI), Joint pain or swelling, any rash, weakness or any other complaints. No history of any cardiac surgery or any trauma. Her New York Heart Association class (NYHA) class was I, normotensive, acyanotic, no clubbing, no edema. No feature of vasculitis or Kawasaki was found. There was a systolic murmur over the upper right sternal border. All her blood reports were normal.

FIG. 1. **Chest Xray (CXR) showed cardiomegaly, right atrial enlargement but normal pulmonary vascularity.**

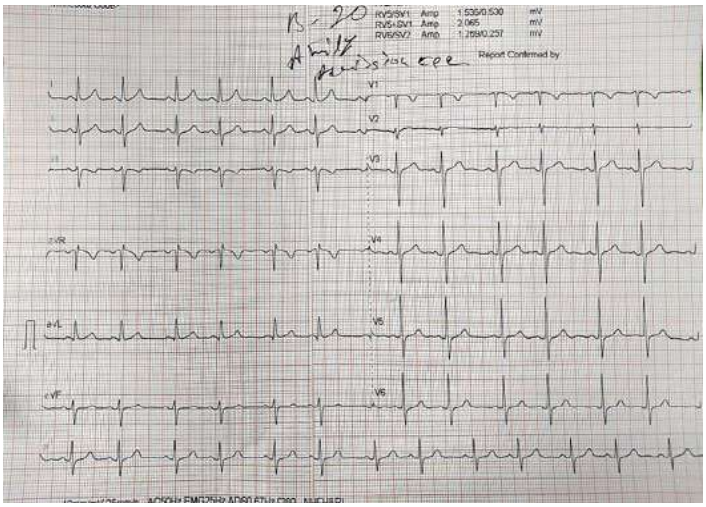

FIG. 2. **ECG showed sinus arrhythmia, left axis deviation.**

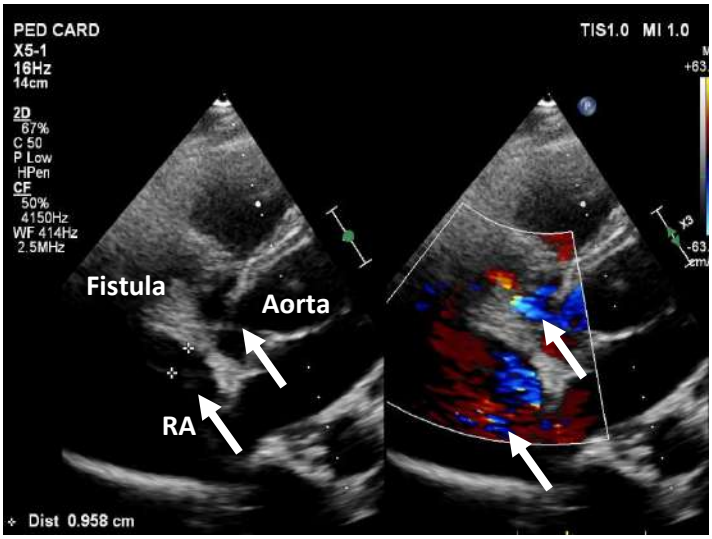

FIG. 3.

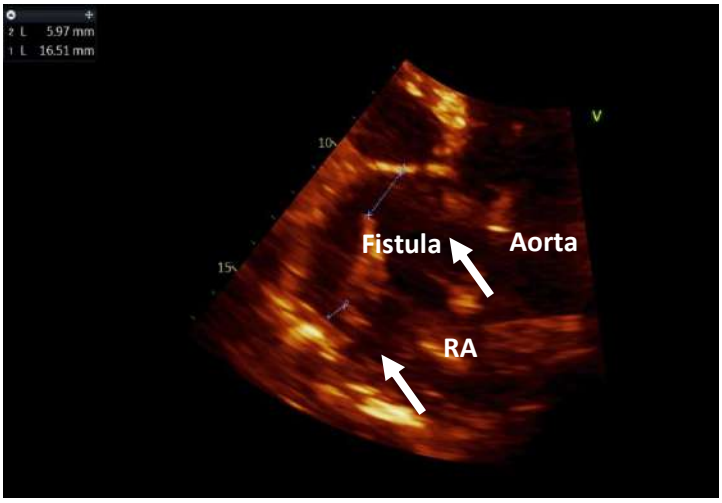

FIG. 4. **Transthoracic echo (3,4) showed large channel communicating dilated right coronary sinus to right atrium (RA).**

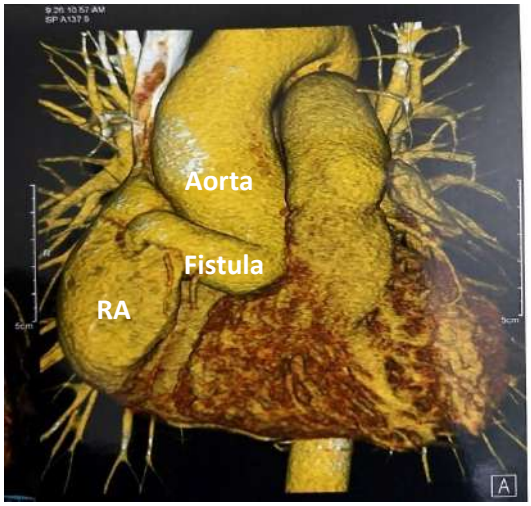

FIG. 5.

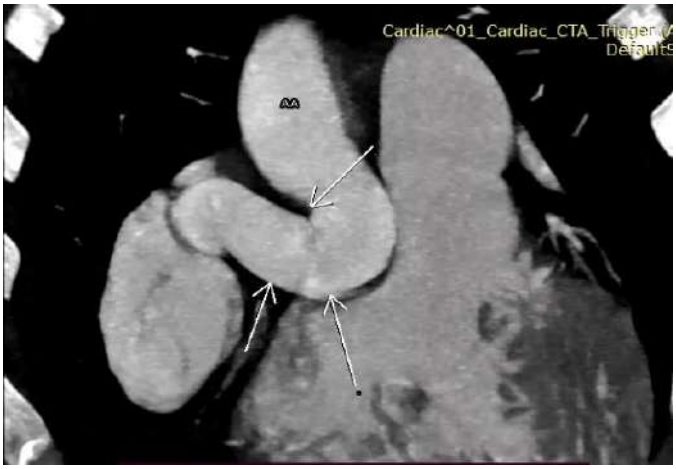

FIG 6. **CT angiogram showed (5,6) dilated ascending aorta and aortic root with large fistula communicating right coronary sinus with right atrium, which was tortuous with 180 degree turn with constriction at different site.**

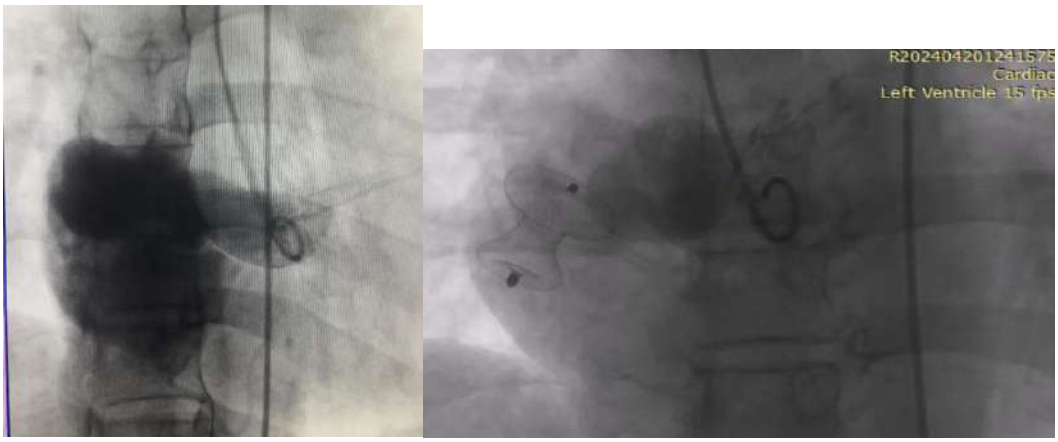

FIG. 7,8. **Cardiac catheterization showed the tortuous course of the fistula with safe deployment of device with no residual shunt.**

Cardiac catheterization revealed mild pulmonary arterial hypertension (40/10/18 mmHg) and a significant left-to-right shunt (Qp:Qs=1.8:1). The tortuous and constricted fistula tract was visualized, and the closure site was selected at the narrowest point. A 22/20-mm duct occluder was chosen, as it was sized 20%-40% larger than the tract.

Using a retrograde approach, right femoral venous and arterial access was obtained with 10-F sheaths, and the tract was crossed with a wire into the pulmonary artery. A delivery sheath was then advanced over the wire, and the occluder was deployed at the distal portion of the tract near the right atrium.

After deployment, angiography and echocardiography confirmed the secure placement of the device with no residual flow. Echocardiography later that evening and the following day also confirmed good device positioning and normal ventricular function. The patient was in stable physical condition and was discharged two days after the procedure. She returned for follow-up at one and three months, with continued good outcomes. Aspirin therapy was prescribed for six months.

3. Case 2

A 45-year-old woman presented with complaints of fatigue, breathlessness on exertion, and palpitations. Her symptoms had worsened over the last 5 months. The patient had an uncomplicated pregnancy at 24 years of age.

She had no history of fever, recurrent RTI, Joint pain or swelling, any rash, weakness or any other complains. No history of any cardiac surgery or any trauma. Continuous murmur found on the right sternal border.

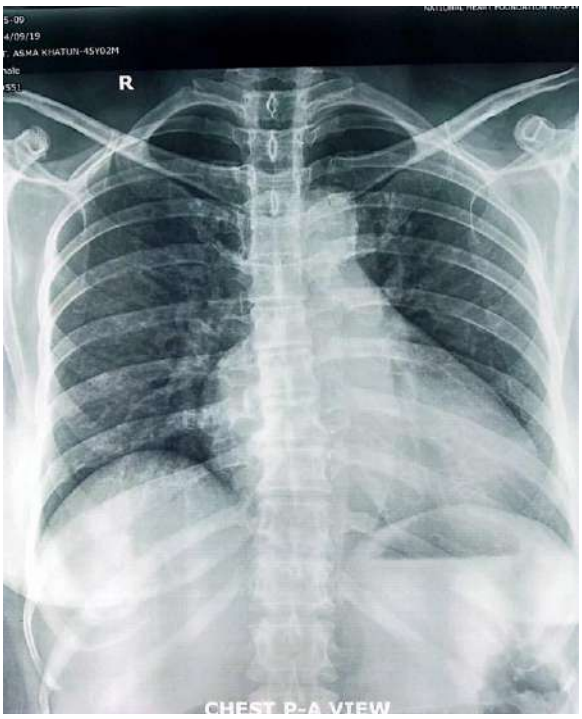

FIG. 9. **Chest radiograph showed dilated RA with normal congestion of the pulmonary vasculature.**

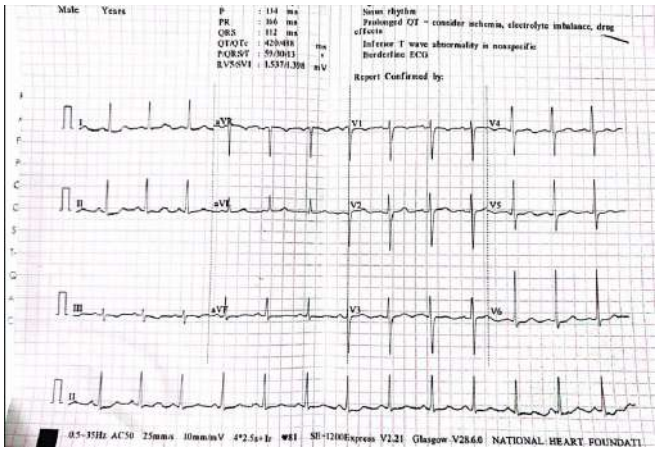

FIG 10. **ECG showed sinus rhythm with normal axis.**

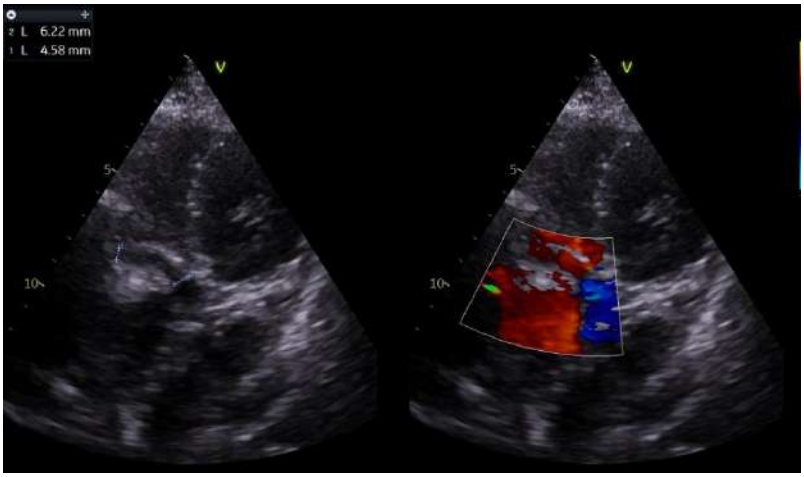

FIG. 11. **Transthoracic echo showed a short fistula formed by ruptured non coronary sinus to RA near right ventricular inflow part with dilated right atrium and left ventricle with good biventricular function.**

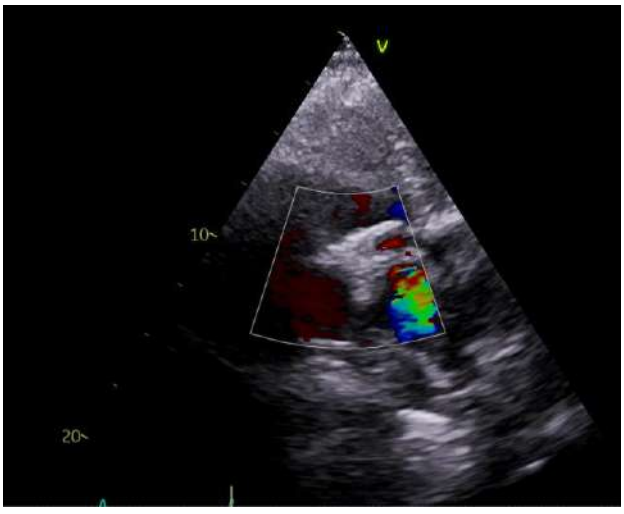

FIG. 12. **Device in situ with secured position with no residual shunt and well away from tricuspid valve with mild tricuspid regurgitation.**

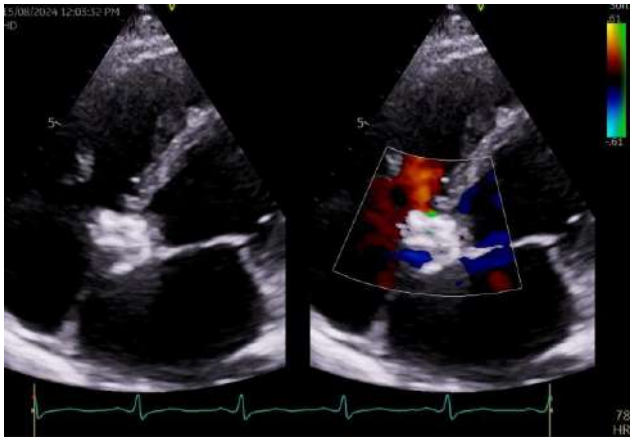

FIG. 13. **(12,13) Device in situ with secured position with no residual shunt with well away from aorta and tricuspid end.**

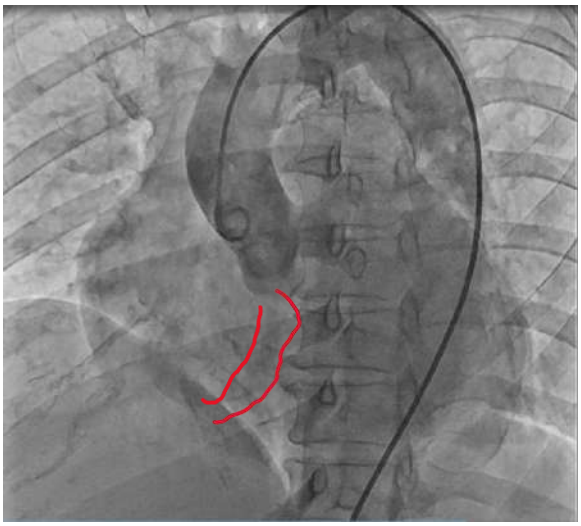

FIG. 14. **Ruptured non coronary sinus to RA near right ventricular inflow.**

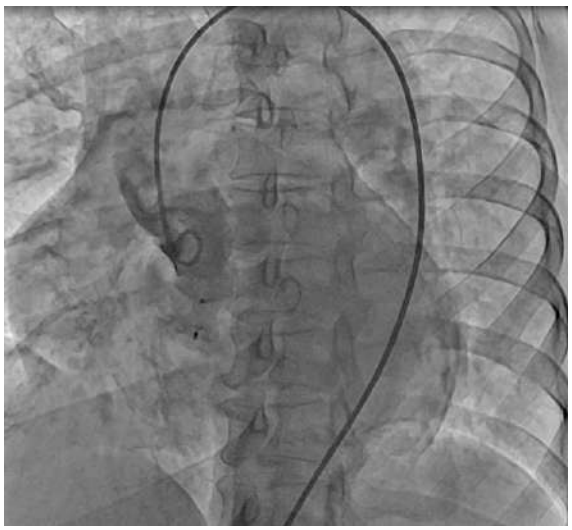

FIG. 15. **Device in safe position with no residual flow with no aortic regurgitation.**

In cardiac Cath we found a significant shunt (1.5:1) so we planned to close. Angiography confirmed the diagnosis. Coronary angiogram showed normal finding. Arteriovenous loop was formed. Amplatzer 16/14 mm duct occlude (ADO I) (St.Jude Medical, approach with 9 size Amplatzer delivery sheath and deployed with the guidance of check angiography and echocardiography after confirming secured position.

Before release confirmed coronary supply, aortic and tricuspid regurgitation or any distortion of surrounding structure. Patient was discharged after two days with the advice aspirin for six months. Recently this patient came to her first follow up after one month and found asymptomatic with no residual shunt with good health status.

4. Case 3

62-year-old man presented with progressive onset of breathlessness, palpitation, and deterioration in exercise capacity. He had no history of fever, trauma or any previous surgery. On examination, he was functionally NYHA III, pulse-70/min, blood pressure-160/60 mmhg, with continuous murmur IV/VI in left parasternal area.

Chest X-ray showed cardiomegaly with increased pulmonary blood flow. Transthoracic & trans oesophageal echocardiography was performed and showed non coronary sinus ruptured into proximal right ventricular outflow tract (RVOT) (maximum & minimum diameter and the length of the aneurysm were taken). The ventricular septum showed no defect, and no aortic regurgitation was present.

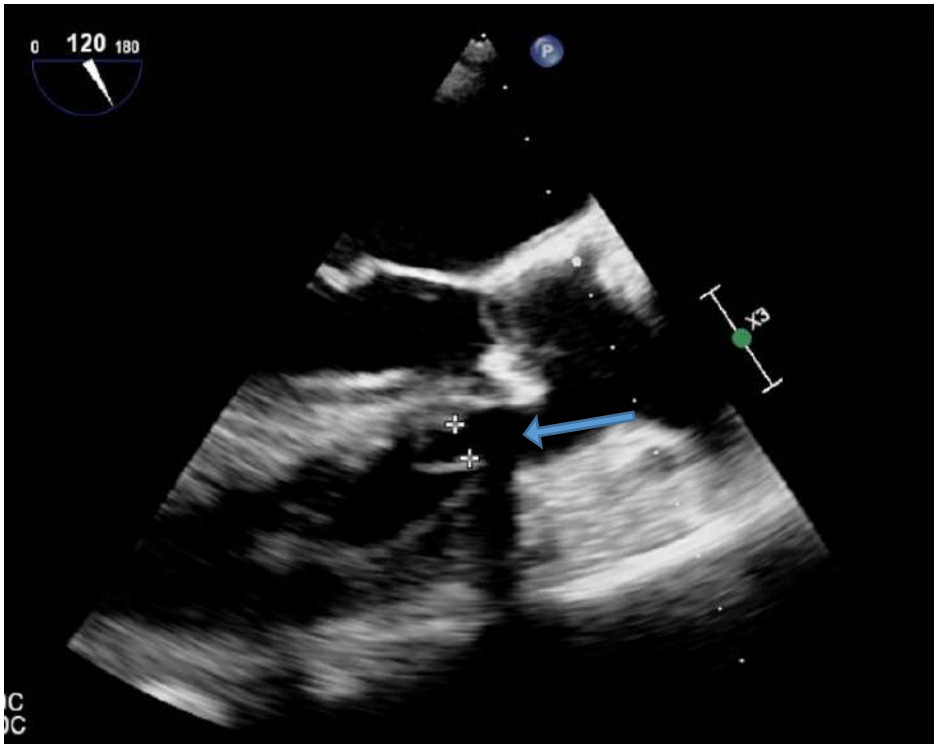

FIG. 16. **TEE showed ruptured non coronary sinus into right ventricular outflow tract.**

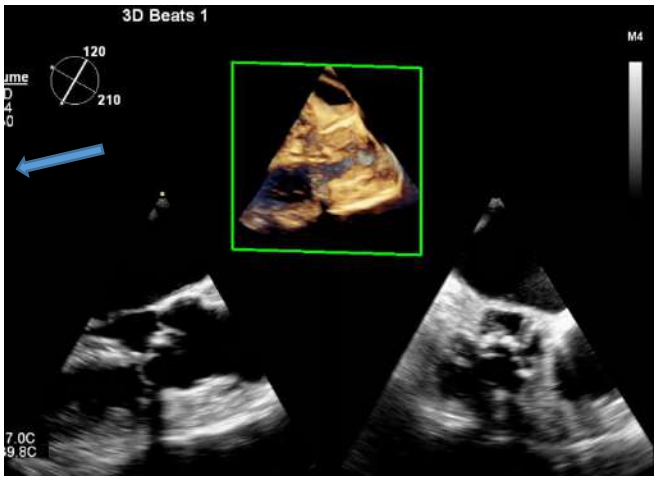

FIG. 17.

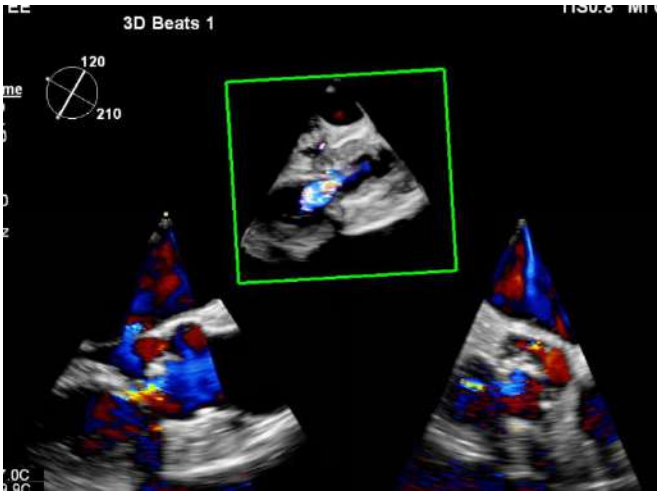

FIG. 18. **TEE 3D (17,18) showed ruptured non coronary sinus from different plane into right ventricular outflow tract.**

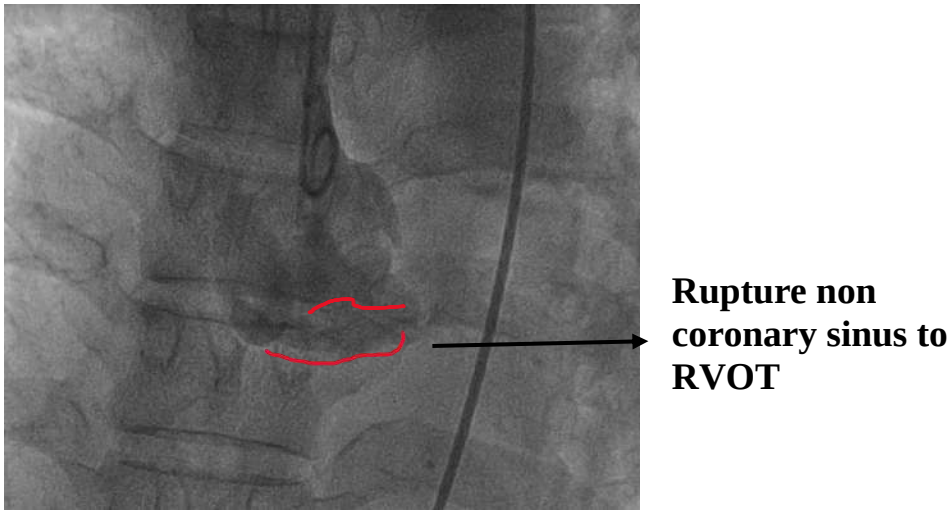

FIG. 19. **Root aortogram showing aneurysmal ruptured non coronary sinus to Right ventricular outflow tract (RVOT).**

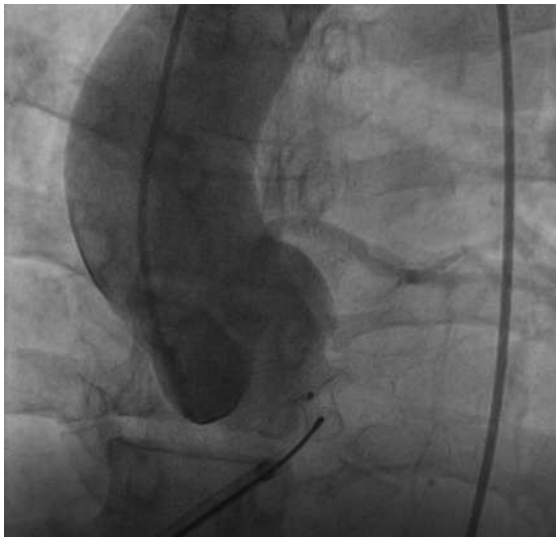

FIG. 20. **Deployment of ADO I (12X10mm) device after confirming its secured and stabilized position.**

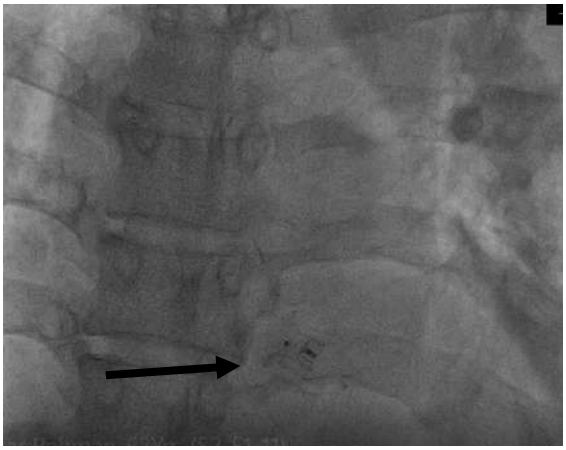

FIG. 21. **Deployed device with stabilized position.**

Cardiac catheterization was performed and did not reveal any coronary artery disease and showed noncoronary sinus ruptured into right ventricular outflow tract; the mean pulmonary artery pressure was 40/15 mmHg and the Qp/Qs was 1.8:1. The RSOV was crossed with a 7F Judkin Right catheter (Cordis Corporation) and a 0.035” × 260 cm straight tipped Terumo wire (Terumo Corp, Japan) from the aortic side. The wire was manipulated into pulmonary artery (PA) and snared through the right femoral vein with a 10 mm Goose Neck Snare (Microvena, MN, USA), to form an arteriovenous loop. The delivery sheath was passed from the venous end and pushed over the wire across the ruptured sinus of Valsalva (RSOV). The Amplatzer 12/10 mm duct occlude (ADO I) device was loaded into the sheath. The aortic retention disc was opened into the ascending aorta and the entire system was pulled back till it anchored at the aortic end of the RSOV. At this point, the other end of the device was delivered by stabilizing the loading cable and pulling back the sheath. The entire maneuver was performed under fluoroscopic and transesophageal echo guidance. A check angiogram was done to confirm the position of the device. Once it was found to be optimum & quantify no aortic regurgitation (AR) & coronary artery was well away then the device was released. He was given Aspirin (5 mg/kg/day) and clopidogrel for six months following the procedure. He came for follow up after one. three. six, one year later found with good state.

TABLE 1. **Summary of three cases with aorto-cameral fistula (ACF).**

Age (year)	Sex	Symptom duration(month)	Communicating chamber	Cause	Device size	Follow up	Complication
35	F	5	Right coronary sinus to right atrium	Idiopathic	22/20	4 month	No
45	F	5	Non coronary sinus to right atrium	Ruptured Sinus of valsalva	16/14	1 month	No
62	M	11	Non coronary sinus to right ventricular outflow tract	Ruptured Sinus of valsalva	12/10	5 year	No

5. Discussion

Aorto-cameral fistulas (ACF) are classified into congenital (aorto-atrial fistulas), which are less common (12%) than those of acquired causes [2,3]. The first case was an aorto-atrial fistula in which the aortic root was dilated, and the cause was idiopathic, which is very rare [2]. Other two cases were due to ruptured sinus of Valsalva. Among the acquired cause ruptured sinus and infective endocarditis is most common [2]. Valsalva aneurysms may involve all 3 sinuses, more frequently the right (75%-90%) and noncoronary (10%-25%), and rarely the left coronary sinus are involved [4]. Both cases of ruptured sinus of Valsalva involved the noncoronary sinus. This anomaly can remain unrecognized for many years [5,6]. Last case was diagnosed at the age of 62 year; he had also gone unrecognized for a long time. Patients of all ages have been reported to be affected by aortic atrial fistula, with cases ranging from as young as 5 days to 85 years old [2].

When the cause of the ACF is due to a ruptured sinus of Valsalva aneurysm, the right atrium, along with the right ventricle, are the most common receiving cardiac chambers. This is because the right coronary sinus, which straddles the tricuspid valve, and therefore is in anatomic proximity to both of these chambers. When the ruptured sinus of Valsalva aneurysm specifically involves the non-coronary sinus, the adjacent right atrium is the most common receiving chamber [4]. This reported case series, aorto-atrial fistula was identified between the right coronary sinus and the right atrium. Additionally, two other cases involved ruptured sinus of Valsalva, with the non-coronary sinus rupturing into the right atrium and the right ventricular outflow tract.

Depending on the type of fistula, small shunts may initially show no symptoms at all, with a constant murmur on physical examination serving as the only indicator of a diagnosis. However, as spontaneous resolution is uncommon, these smaller fistulas will eventually cause progressive cardiac dysfunction. Significant and potentially fatal outcomes include severe dyspnea, congestive heart failure, and even mortality can arise from larger fistulas and the shunts that occur from them [1].

While transthoracic echocardiography is a useful tool for identifying this condition in patients, the diagnosis becomes clearer with transesophageal echocardiography, which has a clarity rate of 97%. For a conclusive diagnosis and assessment of coronary vascular architecture, coronary angiography and aortography are particularly significant [7].

In a systematic review, ACF was corrected via an open surgical approach in 73.5% of all cases. In 10.3% the fistula was closed via a percutaneous intervention, whilst in 4.4% of cases a conservative medical approach was advocated (e.g., diuretics and blood transfusions), due to the high surgical risk. When percutaneous closure of the fistula tract was employed, closure with an Amplatzer device was the treatment of choice (71.4%), followed by coil embolization (14.3%), covered stents (7.15%) and finally balloon closures (7.15%) [3,2]. Treatment plans are determined by the treating physicians' consensus and expert opinion as no clinical studies have been conducted on these patients.

Traditionally, surgical closure has been the mainstay of treatment for ACF, with an operative mortality rate of <5% and excellent long-term outcomes [8,9]. Nevertheless, these patients remain at risk of prolonged hospital stays and postoperative complications such as chest pain and septicemia, making percutaneous device closure an attractive alternative [8]. ACF (isolated RSOV) have been successfully closed percutaneously using transcatheter devices [10].

Success rates up to 90% have been reported in catheter-based closures [11]. In order to determine the best device type and size, as well as the precise moment of deployment to achieve total occlusion of the fistula without obstructing flow in normal coronaries, temporary occlusion of the fistula with a balloon catheter and simultaneous selective coronary injection is advised [12].

The first reported case, temporarily occlusion of the tract with a balloon was done due to its significant tortuosity. During balloon occlusion, both coronaries were assessed and determined the size of the distal tract where it opens into the right atrium.

Complications, although rare, include cardiac perforation, fistula formation, thrombosis, and device embolization into the systemic or pulmonary circulation [11]. Post-deployment follow-up includes the assessment of coronary blood flow, aortic valve function, and the presence of thromboembolism. After the deployment of a device, a short course of anticoagulants or antiplatelet drugs is recommended to prevent thromboembolism until the endothelialization of the device occurs [11]. Regarding follow up, Frequent exercise and relaxation is needed. To rule out late sequelae, echocardiogram is required to look for potential regional wall motion abnormalities and ECGs are required to rule out probable myocardial ischaemia [12].

6. Conclusion

Aorto-cameral fistula (ACF) is a rare and complex condition that necessitates corrective treatment, even in asymptomatic cases, as the disease can progressively worsen if left untreated. Treatment options include both surgery and the transcatheter approach, with the latter gaining popularity due to its favorable outcomes and less invasive nature. However, caution is essential because complications can occur during or after the procedure, making it crucial to have surgical backup available in case of emergencies.

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