



Aorta-Right Atrial Fistula, Simulating IVC with Percutaneous Closure by CERA® Prosthesis

Fístula Aorta-Atrio Derecho, Simulando CIV, con Cierre Percutáneo por Prótesis CERA®

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SUMMARY

Aorta to right atrium fistula can be a congenital or acquired condition. Congenital fistulas are uncommon heart diseases and can be mistaken by other heart lesions like interventricular septal defect or atrial septal defect. They seem to be rare extra-cardiac vascular channels and their natural history, clinical presentation and treatment remain uncertain. Acquired fistula between aorta and right atrium is usually due to a post infectious rupture of sinus of Valsalva. The authors present a case of aorta-right atrium fistula that had been mistaken by perimembranous interventricular septal defect during echocardiography examination and confirmed by cardiac catheterization.

Descriptors: Fistula; Endocardial Cushion Defects; Heart Defects, Congenital; Echocardiography/diagnosis

RESUMEN

Fístulas entre la aorta y el atrio derecho, pueden ser de origen congénita o adquirida. Las fístulas aortocamerales congénitas son raras anomalías del corazón, que pueden ser confundidas con otros defectos como comunicaciones interventriculares o interatriales. Son raros canales vasculares extra cardíacos y su historia natural, presentación clínica y tratamiento adecuado todavía son oscuros. Las fístulas adquiridas, en general son resultantes de ruptura del seno de Valsalva pos-infecciosa. En este relato presentamos un caso de difícil diagnóstico ecocardiográfico, en el cual el diagnóstico de fístula aorta-atrio derecho fue confundido con el de comunicación interventricular perimembranosa y solamente confirmado después de estudio hemodinámico.

Descriptores: Fístula, Comunicación Atrioventricular, Cardiopatías Congénitas, Ecocardiografía/diagnóstico

Introduction

Aortocameral fistulas are rare anomalies of the heart. They can be congenital, in general, tunnel-shaped and winding, or they can be acquired, generally secondary to infections or surgical procedures. They can be confused with other septal defects, such as interatrial communication (IAC) and interventricular communication (IVC).

Echocardiography and cardiac catheterization are useful to provide diagnosis¹.

Case Report

A case of a 26-year old female patient is presented. The patient complained of dyspnea on exertion and little frequent palpitations. She was forwarded to our service

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to undergo transesophageal echocardiogram (TEE). She had been previously diagnosed with perimembranous IVC at transthoracic echocardiogram (TTE). TEE showed what we thought to be a subtricuspid IVC (partially occluded by septal leaflet tissue of the tricuspid valve) measuring approximately 0.3 cm with transeptal flow directed from the left ventricle (LV) to the right ventricle (RV). Maximum LV \Rightarrow RV systolic gradient estimated in 90 mmHg. (Figure 1).

Perimembranous IVC diagnosis was maintained despite opening visualization was quite difficult. The patient was forwarded to cardiac catheterization for the purposes of pre-operative assessment. During cardiac catheterization, continuous flow was noted from the sinus of right Valsalva to the right atrium, which led to the diagnosis of aortocameral fistula instead of IVC. The pro-

cedure chosen was percutaneous closure of the fistula with 6 asymmetric CERA® prosthesis. The procedure was successfully performed. (Figures 2 and 3).

Discussion

Aorta-right atrial fistula is found in an abnormal vascular connections group called aortocameral fistulas. These fistulas are rare extracardiac vascular channels. Their natural history, clinical presentation and adequate treatment are not clearly known yet². Extracardiac vascular communications may originate in any one of the three sinus of Valsalva, although they are rarely originated in a non-coronary sinus^{1,3}.

Commonly, aortocameral fistulas have a congenital origin. However, they may be described in association to prosthetic valve endocarditis after aortic root repair and percutaneous closure of septal defects⁴.

Most of the patients are asymptomatic. Nevertheless, symptoms like palpitations, mild dyspnea and recurrent respiratory tract infections may be experienced⁵. Despite the fact fistulas are identified at bidimensional echocardiogram, retrograde aortography combined with angiography are crucial to show the course and coronary ostiums¹.

Congenital aorta-right atrial fistulas can be classified into anterior or posterior, according to their origin and course in relation to the ascending aorta⁵. In our case, it was not possible to classify the fistula at the echocardiogram. Additionally, our case did not show exactly the characteristics of a congenital fistula (tunnel-shaped and winding). Instead, it resembled a communication between the sinus of Valsalva and the right atrial chamber. Thus, we cannot assure its origin, whether congenital or acquired.

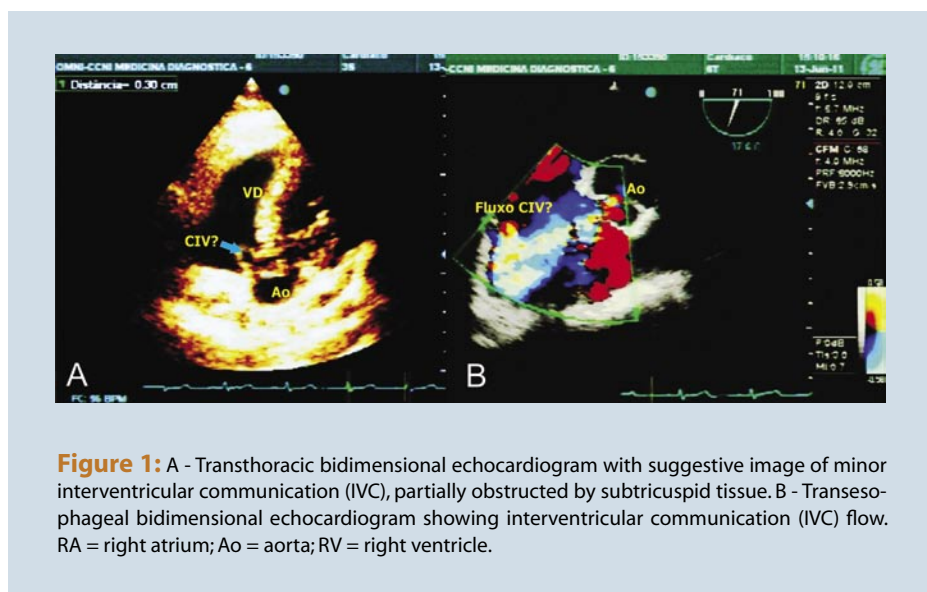


Figure 1: A - Transthoracic bidimensional echocardiogram with suggestive image of minor interventricular communication (IVC), partially obstructed by subtricuspid tissue. B - Transesophageal bidimensional echocardiogram showing interventricular communication (IVC) flow. RA = right atrium; Ao = aorta; RV = right ventricle.

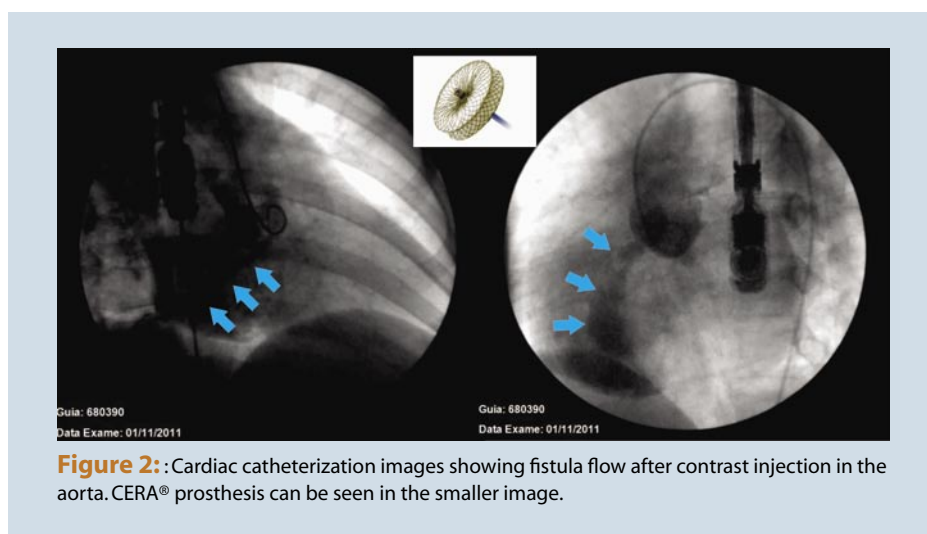


Figure 2: Cardiac catheterization images showing fistula flow after contrast injection in the aorta. CERA® prosthesis can be seen in the smaller image.

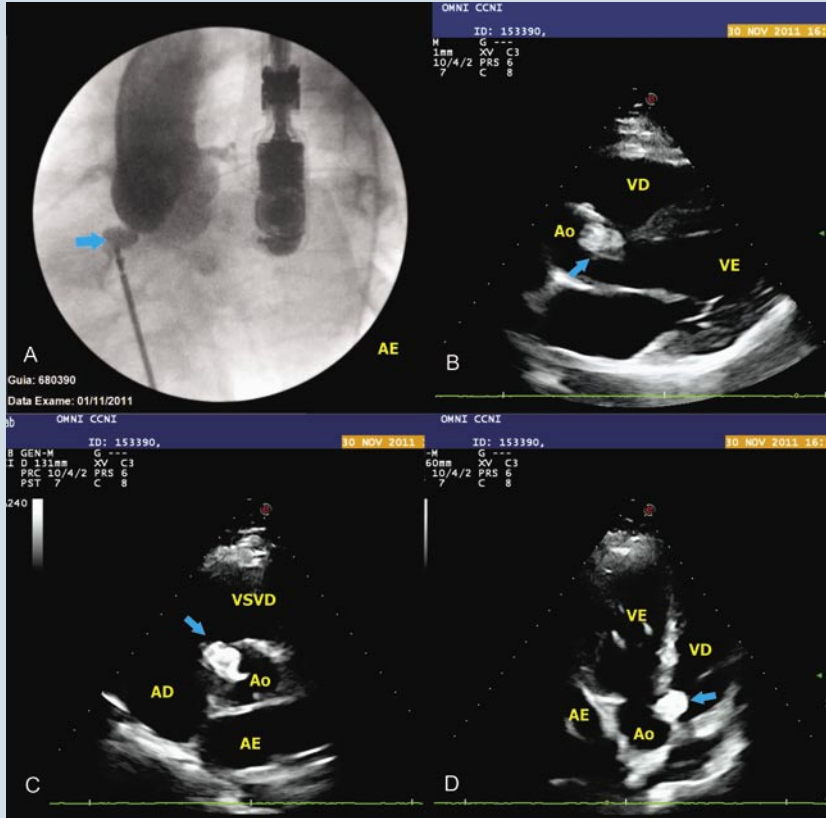


Figure 3: A - Cardiac catheterization images after prosthesis implant (arrow). B - Longitudinal parasternal view at bidimensional echocardiogram showing CERA® prosthesis (arrow). C - Transversal parasternal view at bidimensional echocardiogram showing CERA® prosthesis (arrow). D - Apical 5-chamber view at bidimensional echocardiogram showing CERA® prosthesis (arrow). LA = left atrium; Ao = aorta; RV = right ventricle; LV = left ventricle; DORV = double outlet of the right ventricle.

Fistula closure is mandatory in case of symptoms. However, closure is recommended in asymptomatic patients as it is a low-risk procedure and due to the risk of ventricle overload, bacterial endocarditis, pulmonary vascular disease, aneurysm formation and even spontaneous rupture due to continuous permeability⁶.

In perimembranous IVCs, many times it is possible to see at the echocardiogram a small aneurysm in the membranous septal region, which corresponds to the involvement of the tricuspid subvalve tissue in an attempt to spontaneously close the defect⁷. A very similar aspect was noted in this case report, but in our case it was a slight dilation of the sinus of Valsalva. Additionally, mosaic flow in the region with high gradient confirmed the wrong diagnosis provided by echocardiography of minor perimembranous IVC. It was in fact continuous flow of the fistula, with difficult alignment with Doppler.

There are many treatment options which vary according to the type of fistula, gauge, winding, calcification, course and relation of coronary ostiums to the aortic opening of the fistula⁵.

In case of tunnel-shaped fistula, embolization or even surgical closure are good options. In our case, due to the anatomical aspect, the hemodynamic team selected percutaneous closure of the fistula through the same technique, usually used for the occlusion of interventricular communications. Some details were assessed more strictly, such as the distance of the right coronary artery ostium to the sinus of Valsalva (not to obstruct the coronary artery) and the opening of the leaflet before device release (not to cause aortic valve dysfunction).

The literature shows few cases of percutaneous occlusion of extracardiac fistulas.

Ribeiro et al.⁸ reported a case of a 40-year old patient with ruptured aneurysm of the right sinus of Valsalva to AD, unde-

termined etiology, which evolved to congestive cardiac failure and LV dilation. Aortic root angiography showed opening measuring around 3 mm in diameter. ADO II prosthesis was implanted easily. No residual flow after the procedure, and aortic valve leaflet movement was not compromised.

Chandra et al.² reported a case of a 12-year old girl with history of dyspnea on exertion and palpitations. Angiography showed a great fistula largely originated in the right sinus of Valsalva, anterior to the origin of the right coronary artery, with narrow end in the posterior wall of the right atrium. Coronary angiography did not show any changes. Fistula was closed by the placement of a device in the aortic end. Whether the fistula should have been closed on both ends and what would be the risk of thromboembolic in the systemic circulation with only on distal portion closed is still not clear².



The continuous use of ADO II prosthesis in a larger number of patients with congenital or structural cardiopathies is crucial to determine the actual role of this new device in lesion percutaneous closure⁸.

CERA occluder is a self-expandable device made of nitinol truncated cone, coated with ceramics. This provides greater flexibility and allows the over dimensioning of the device in relation to the diameter of the channel without causing damage to surrounding structures. Implant protocol and follow-up are the same used for Amplatzer[®] Duct Occluder I (ADO I) device. Due to easiness of implantation, greater flexibility and fistula location, the hemodynamic team chose to implant this device⁹.

Conclusion

Percutaneous closure of aorta-right atrial fistula may be the best treatment option for selected cases. The procedure is safe and effective. It must be performed by an experienced team. It has been reported by other authors. It is a good option for surgical therapeutics, and it is well established in literature¹⁰.

In difficult and doubtful echocardiography diagnoses of perimembranous IVC, complementation with hemodynamic studies may contribute to the correct diagnosis, in addition to detecting other less frequent anatomical changes.

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