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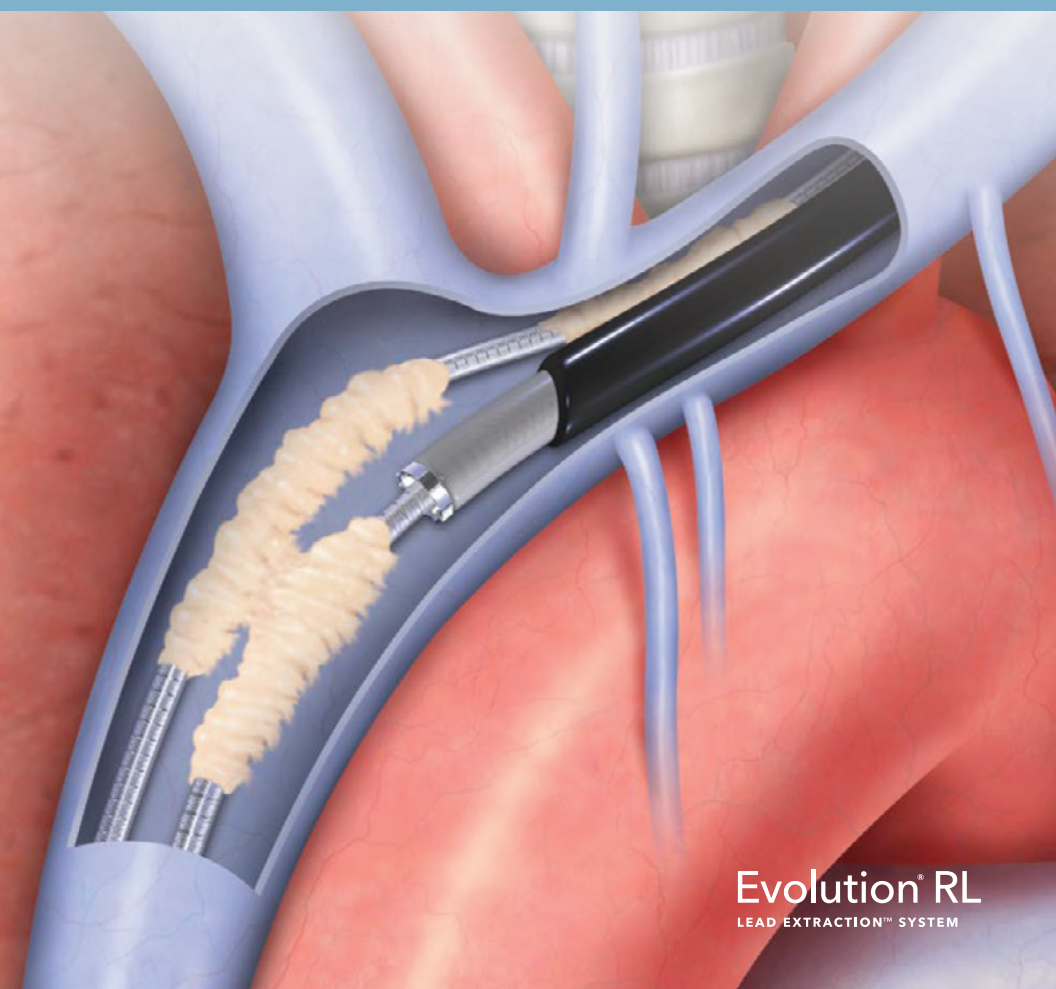


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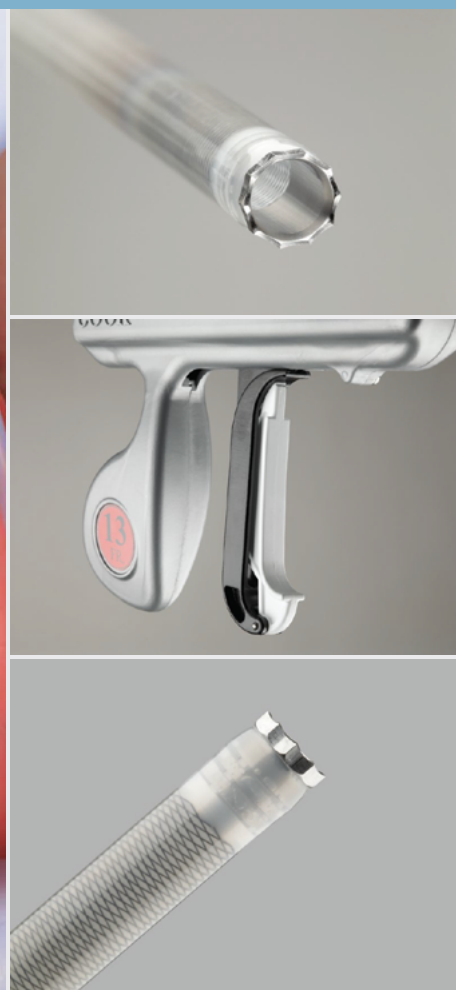
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# LEFT GONADAL VEIN TRANSPOSITION IN THE NUTCRACKER SYNDROME

## ABSTRACT

The cause of pelvic congestion symptoms is usually nutcracker syndrome. We present the clinical case of a 58-year-old female patient who consulted for lumbar and left flank pain, microhematuria, and dyspareunia of 10 years of evolution. Angiotomography showed compression of the left renal vein at the level of the aortomesenteric compass, associated with pelvic varices. It was resolved with surgical treatment by transposition of the left gonadal vein to the inferior vena cava and embolization of the distal bed with coils and foam. The patient had an uncomplicated postoperative period with complete resolution of symptoms.

**Keywords:** *nutcracker syndrome, pelvic congestion, venous transposition.*

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## INTRODUCTION

Nutcracker syndrome is caused by anatomical compression of the left renal vein, with venous hypertension and gonadal reflux.

Patients present clinically with hematuria, low back pain, left flank pain, and symptoms of pelvic venous congestion.<sup>1</sup>

It is essential to perform thorough imaging studies to identify anatomical abnormalities and hemodynamic alterations, thereby facilitating a definitive diagnosis. Conventional surgical treatment is a valuable tool for resolving this pathology. This article presents a clinical case treated using a hybrid approach, combining traditional surgery with embolization of the distal venous bed using coils and foam.

## CLINICAL CASE

A 58-year-old female patient with a history of arterial hypertension and hypothyroidism consulted for symptoms of lumbar and left flank pain, pelvic pain with dyspareunia, and long-standing microhematuria.

An angiotomography of the abdomen and pelvis showed signs of pelvic congestion with pelvic varices (*Figure 1*), compression of the left anterior variant renal vein (*Figure 2*), and an angle between the aorta and the superior mesenteric artery of less than 16 degrees (*Figure 3*).

We decided to perform conventional surgery with transposition of the left gonadal vein to the inferior vena cava, associated with embolization of the distal bed with coils and foam.

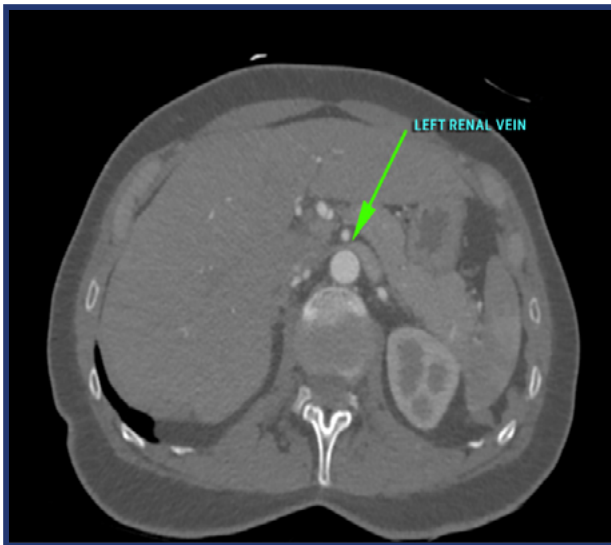
A supra-infra umbilical median laparotomy was performed, with mobilization and exposure of the left renal vein, the gonadal vein, and the inferior vena cava. Distally, phlebography was performed, which revealed numerous pelvic varicose dilatations. Selective cannulation with a microcatheter and embolization with 3% polidocanol foam, followed by coil placement, was performed.

Then, a transposition of the left gonadal vein to the inferior vena cava was performed (*Figure 4*). The patient experienced an uncomplicated postoperative period, characterized by complete remission of symptoms and the absence of microhematuria.

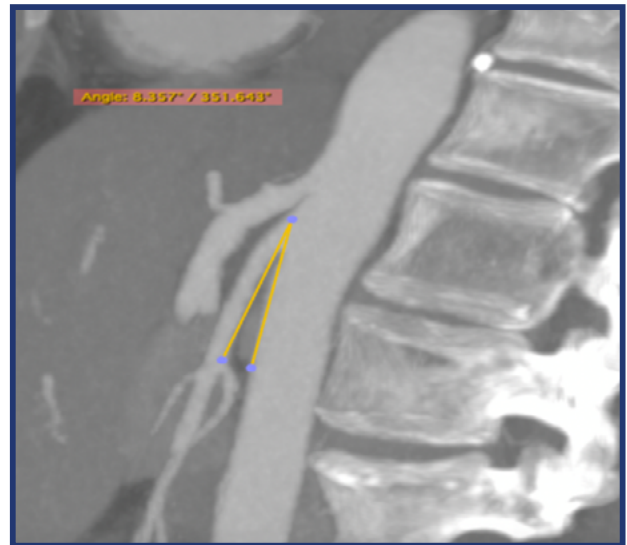


**FIGURE 1.** Angiotomography reveals the presence of pelvic varices.





**FIGURE 2.** Angiotomographic image showing compression of the left renal vein at the level of the aortic-mesenteric junction.



**FIGURE 3.** Angiotomographic image shows that the angle between the aorta and the superior mesenteric artery is less than 16 degrees.



**FIGURE 4.** Transposition of the left gonadal vein to the inferior vena cava.

## DISCUSSION

Symptoms of pelvic venous congestion (pelvic pain, dyspareunia, and dysmenorrhea) are sometimes attributed to the presence of gonadal reflux. In some cases, this is associated with the concomitant presence of compression of the left renal vein between the aorta and the superior mesenteric artery.

Although recognizing the symptomatology and subsequently diagnosing nutcracker syndrome are essential, patient selection and the strategy employed are fundamental to achieving satisfactory results.

Among the therapeutic possibilities is endovascular treatment with stent placement in the left renal vein, first described in 1996. Most of the available data is found in case reports. One of the most extensive published series is that of Chen et al., with 61 patients who underwent left renal vein stenting, with a median follow-up of 66 months. Complications observed were stent maldeployment requiring surgical intervention and stent migration to the right atrium, which required cardiac surgery. New studies report a stent migration rate of 6.7%; this may be of concern because the stents often migrate to the right-sided circulation.<sup>2</sup>

Another study by Wu et al. included 75 patients treated with renal vein stent angioplasty, of whom 6.6% had stent migration.<sup>3</sup>

Nowadays, the endovascular approach with stent angioplasty in the renal vein has become popular, but it is not without potential serious complications such as thrombosis or migration. The long-term patency and durability of the stent have not yet been elucidated. A crucial point to consider is that most patients affected are young women, making the durability of the proposed treatment of utmost importance.

Concerning conventional surgical treatment, the therapeutic options include transposition of the left renal vein to the inferior vena cava, bypass using the saphenous vein, and transposition of the left renal vein with a saphenous vein cuff. An additional surgical alternative is gonadal vein transposition. This technique is feasible, as in many cases, the gonadal vein is dilated, and this procedure can

alleviate the symptoms of pelvic congestion.<sup>4-5</sup> One of the advantages is that it can treat patients with posterior nutcracker syndrome, avoids transection of the renal vein, requires only one anastomosis, and avoids the need for removal of the saphenous vein. Due to the characteristics and applicability of gonadal vein transposition, it has become a viable surgical strategy for treating anterior and posterior nutcracker syndrome.<sup>6</sup>

## CONCLUSION

Nutcracker syndrome is a rare entity, and although multiple techniques have been developed for its treatment, the paucity of data and long-term follow-up do not allow for defining a single treatment strategy.

Some experts consider conventional surgery to be the primary treatment option. Although angioplasty with stenting has gained ground, the patency and long-term durability of this approach have yet to be clearly defined.

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## Declarations

The authors declare no conflict of interest.



# SURGICAL RESOLUTION OF MECHANICAL PROSTHETIC DYSFUNCTION IN A PATIENT WITH A PORCELAIN AORTA

## ABSTRACT

We present a clinical case with a satisfactory surgical outcome of a patient in the seventh decade of life with a diagnosis of dysfunction of a mechanical valve prosthesis in the aortic position and with a calcified aorta (porcelain aorta) from the root to the bifurcation of the iliac arteries. The decision for surgical replacement of the aortic valve was based on the presence of thrombosis and *pannus*, which, in addition, were the cause of severe aortic stenosis and secondary mitral insufficiency, also severe. This scenario is not accounted for in cardiovascular surgical risk scales, such as the EuroSCORE, the Society of Thoracic Surgeons (STS) risk scale, or the Parsonnet scale, among others. This adds a further risk for this procedure, as this fully calcified aortic morphology increases the predisposition for aortic dissection or rupture. For this reason, the repair is difficult because there is no adequate plane for the coaptation of the surgical suture. In this patient, age, the presence of comorbidities such as chronic renal failure, severe pulmonary hypertension, the need for reoperation, and tissues in poor condition are added as risk factors, despite which the postoperative result was satisfactory.

**Keywords:** *porcelain aorta, mechanical prosthetic dysfunction, high surgical risk, comorbidities, pulmonary hypertension, ventricular dysfunction.*

## Authors

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## INTRODUCTION

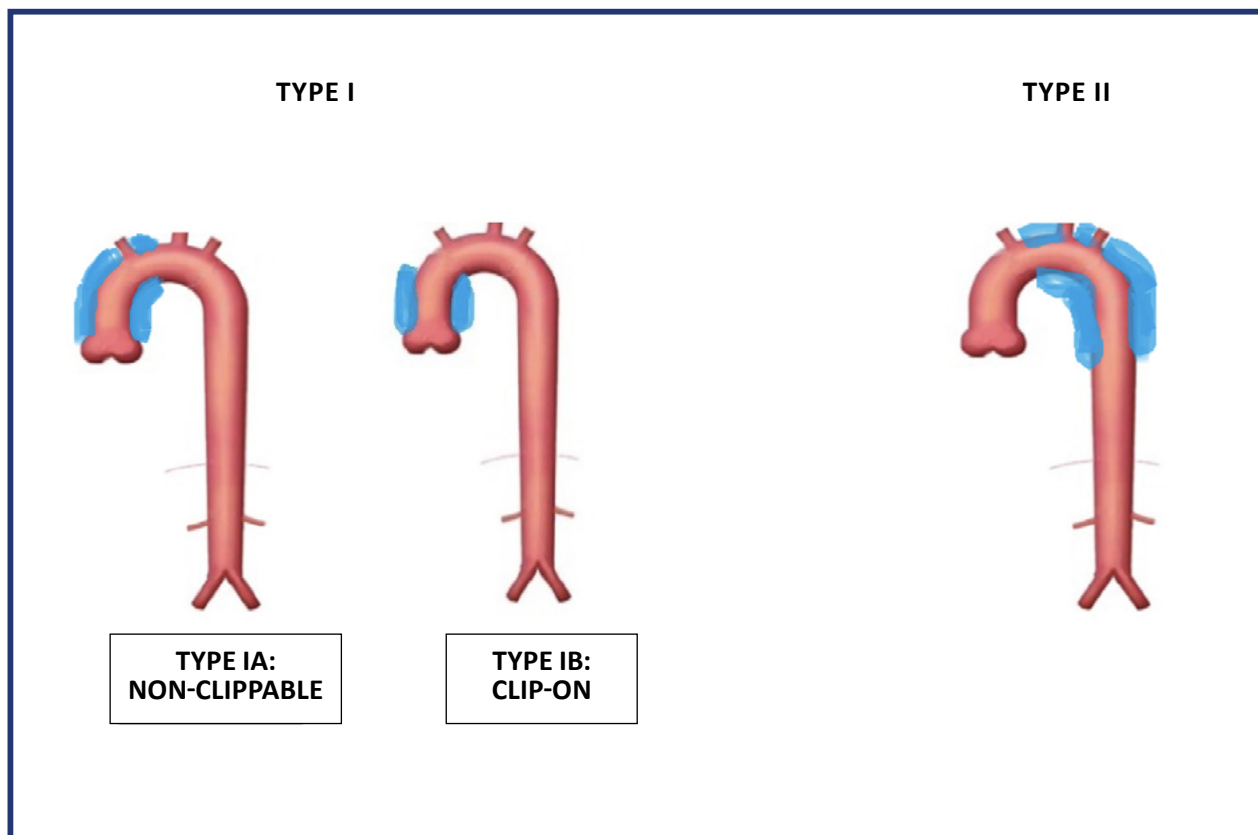
Severe aortic calcification, also called “porcelain aorta”, has a significant effect on patient prognosis. This is a generic entity characterized by the widespread and complete calcification of the thoracic ascending aorta, resulting from excessive calcium accumulation in the aortic wall, which may be related to atherosclerotic plaque. Aortic calcification may be located in the tunica intima, be eccentric and begin at the base of necrotic fibroadipose plaques (atherosclerotic type), or it may involve the tunica media (non-atherosclerotic). Van Mieghem<sup>1</sup> defines porcelain aorta as “significant circumferential calcification or severe atheromatous plaques of the entire ascending aorta extending into the arch such that aortic clamping cannot be performed”, which implies a significant risk during surgery.

The presence of porcelain aorta also influences the choice of procedure according to its location; however, to date there is no clear definition of how this term should be used when deciding the surgical technique, and it is often used as an exclusion criterion for conventional surgery in which aortic clamping and/or cannulation is required.

Amorim et al.<sup>2</sup> proposed a classification of porcelain aorta based on its location and the impact on the decision-making process in a heart team for specific therapeutic options, clarifying the types of procedures that carry a higher risk and are more suitable for the patient.

Amorim's classification recognizes two types of porcelain aorta. Type I involves the location of circumferential calcification in the ascending aorta. This is subdivided into type IA, with calcified aorta without the possibility of clamping (non-clampable), and type IB, with calcified aorta liable to undergo clamping (clampable). Type II deals with calcification of the descending aorta with or without inclusion of the aortic arch (*Figure 1*).

These cases can be satisfactorily resolved with excellent results through interventional procedures. However, such methods can only be performed in patients with a diseased native valve or dysfunctional biological valve prostheses. However, in patients with a dysfunctional mechanical prosthesis due to *pannus* with a high gradient, as in the case we present, the only option is surgical treatment despite the risks involved.



**FIGURE 1.** Amorim's classification of porcelain aorta, according to its location and the possibility of aortic clamping.

## CLINICAL CASE

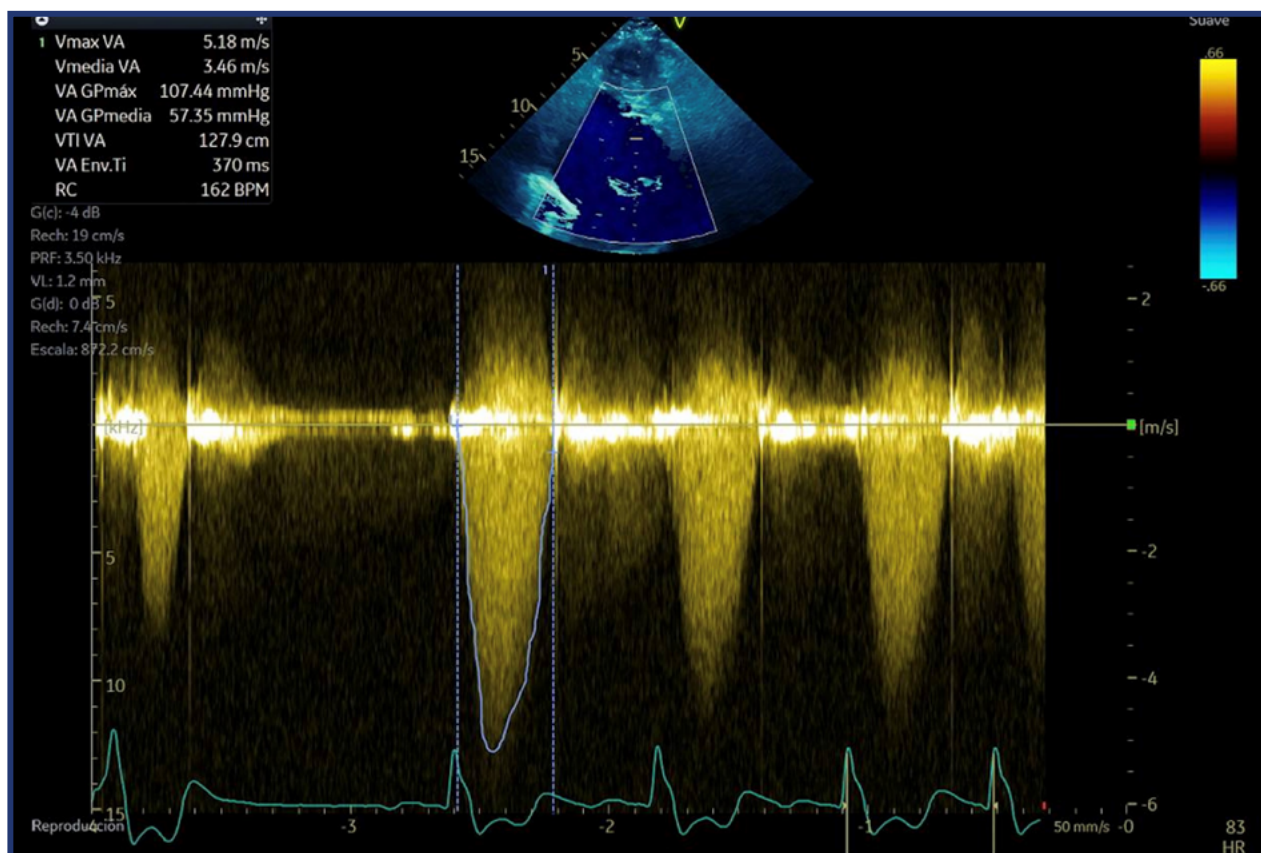
We present the case of a 67-year-old woman with a history of surgical reintervention 24 years ago for mechanical aortic valve replacement Carbomedics 21™ type in the aortic position due to valvular calcification. The patient presented systemic arterial hypertension of 10 years of evolution, type 2 diabetes mellitus diagnosed 8 years earlier, and hypothyroidism and chronic renal insufficiency stage 3b (both conditions of 5 years of evolution).

He consulted for dyspnea of one week's evolution. A transthoracic echocardiogram was performed, which showed the presence of a mechanical prosthesis in a dysfunctional bidisc aortic position with severe stenosis. The parameters studied showed a Vmax of 5.7 m/s, a maximum gradient of 107 mmHg, and a mean gradient of 57 mmHg. The aortic valve area was 0.27 cm<sup>2</sup>/m<sup>2</sup>. Aortic root with ectatic dilatation from the sinuses of Valsalva to the ascending aorta of 43 mm; mitral valve with undilated annulus (24 mm anteroposterior diameter and 30 mm bicommissural), thickened leaflets, predominantly the anterior and posterior leaflet with decreased mobility by tethering of its chordae tendineae, which

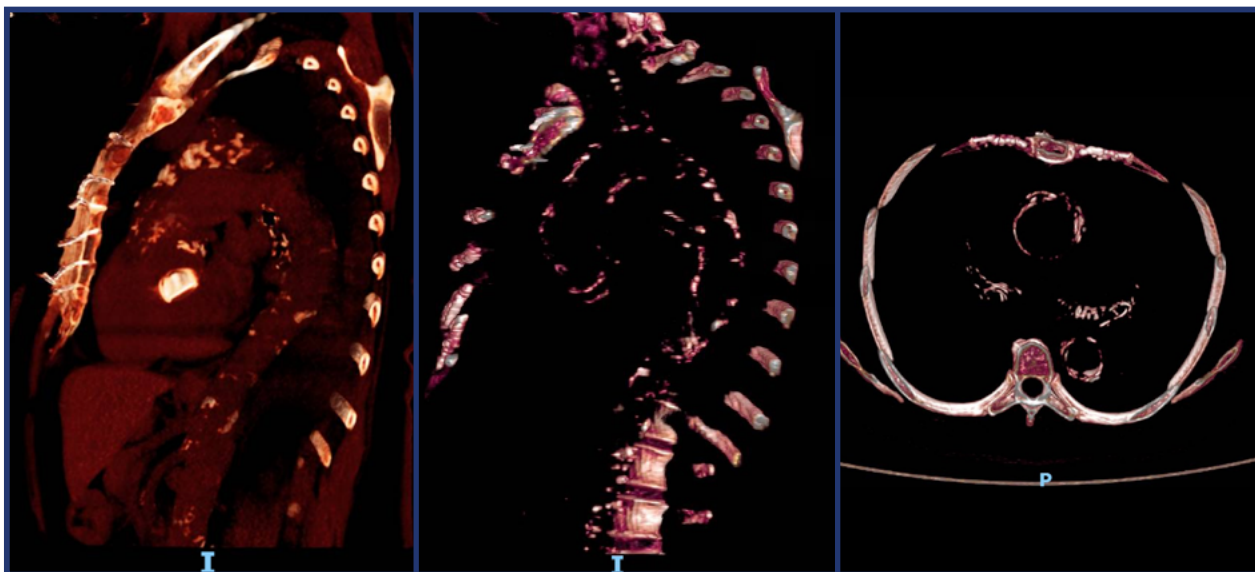
conditions a severe mitral insufficiency with 9 mm vena contracta. The subvalvular apparatus showed no alterations. The left ventricular ejection fraction was 65%. Right ventricular dilatation was observed, with preserved global systolic function. Pulmonary artery systolic pressure was 85 mmHg.

The study protocol included a transesophageal echocardiogram, which reported the presence of a mechanical prosthesis in the aortic position with adequate posterior disc mobility and restriction of anterior disc mobility by mixed mechanism: *pannus* in the anterior annulus and a rounded, hypoechoic, vibratile image with dimensions of 8 × 6 mm at hour 6 compatible with a thrombus, mitral valve with thickened leaflets and severe insufficiency, and tricuspid valve also with severe insufficiency (Figure 2).

Cardiac catheterization was performed, which revealed no angiographic lesions but did show the presence of calcium plaques in the aortic root. Angiotomography was requested, reporting a porcelain aortic artery at the root, which continued into the descending aorta and extended to the bifurcation of the iliac arteries (Figure 3).



**FIGURE 2.** Mean transvalvular aortic gradient of 57 mmHg and a maximum velocity of 5.18 m/s are observed, suggesting severe aortic prosthetic dysfunction.



**FIGURE 3.** Angiotomographic image shows calcification of the aorta artery from its root to the bifurcation of the iliac arteries.

A medical-surgical session was conducted during which the patient's history and the diagnostic studies performed were reviewed. A EuroSCORE score of 78.5 was calculated, representing a very high risk. Nevertheless, a surgical resolution for valve replacement versus aortic valve toilette (prosthesis cleaning) plus mitral valve replacement was considered as the first option, so urgent programming was decided.

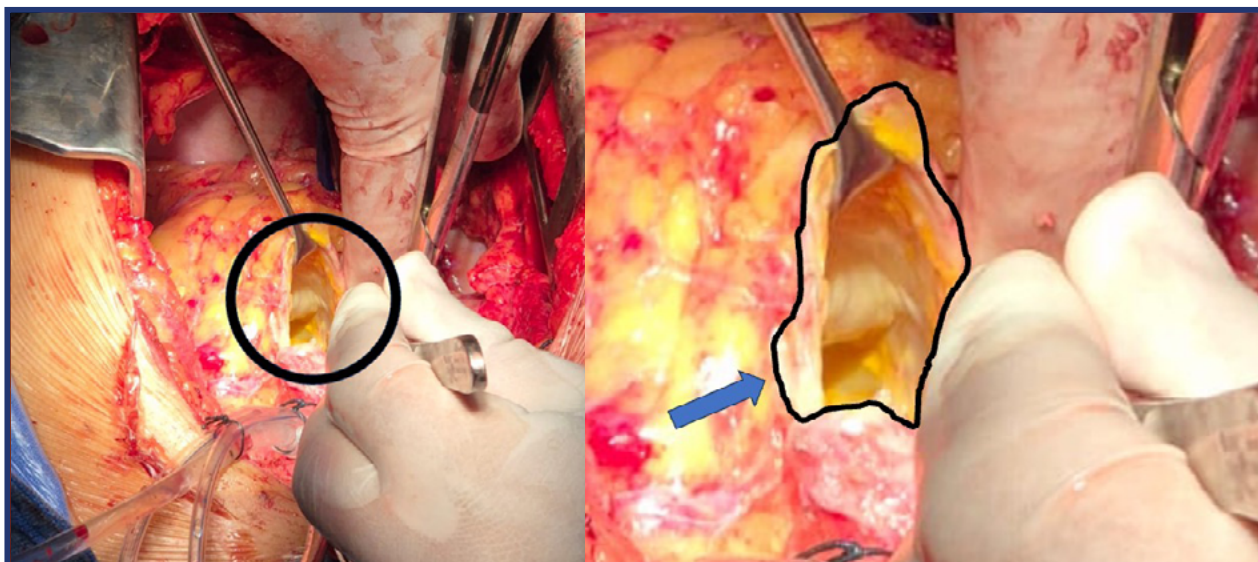
The patient presented with sudden hemodynamic deterioration and dyspnea, so she was transferred to the post-surgical intensive care unit, where she received advanced treatment for heart failure and renal failure with hemodialysis and adjustment by nephrology. She presented improvement in cardiac and renal functions, so it was decided to perform surgery. A median sternotomy was performed and, simultaneously, dissection and cannulation of the femoral artery were performed to avoid manipulation of the porcelain aorta. This surgical approach allowed, in the event of requiring aortic root replacement, the resection of most of the ascending aorta and the removal of adhesions remaining from the previous surgery. Central venous cannulation was performed in the right atrium to achieve better venous drainage instead of femoral vein drainage. Once the middle mediastinum was exposed, a calcified plaque was observed along the entire length of the ascending aorta (Figure 4). Cardiopulmonary bypass (CPB) was initiated, and a ventricular drainage cannula (Vent™) was placed in the right superior pulmonary vein, which brought the patient's temperature to 28 °C as part of the surgical plan, which included subsequent ventricular fibrillation. High aortic

clamping was performed near the arch, where a minor amount of calcified plaque was noted, with aortotomy at the site of previous raffia. Anterograde cardioplegia (Custodiol™) was infused directly into the left coronary *ostium* and then into the right. Once cardiac arrest was reached, a 10 × 8 mm thrombus was observed on the aortic side of the valve prosthesis and *pannus* around the valve circumference on the ventricular side (Figure 5). A *toilette* with total resection was performed. Once the *toilette* was completed, persistent prosthetic dysfunction was detected due to restricted mobility in the hemi-discs, with inadequate opening and closing of both leaflets; therefore, prosthetic replacement was decided. The mitral valve had thickened leaflets and calcification in P2 and P3. It was agreed to resect and replace the mitral valve. A Mitris 27 Edwards Lifesciences™ prosthesis was placed in the mitral position, and an Inspiris 21 Edwards Lifesciences™ prosthesis in the aortic position. Aorto-orrhaphy with the Carrel technique was performed without incident. Cardiopulmonary bypass time was 112 minutes, and aortic clamping time was 115 minutes. Cardiopulmonary bypass was successfully destented with supportive infusion of vasoactive amines (noradrenaline) and inotropic amines (dobutamine).

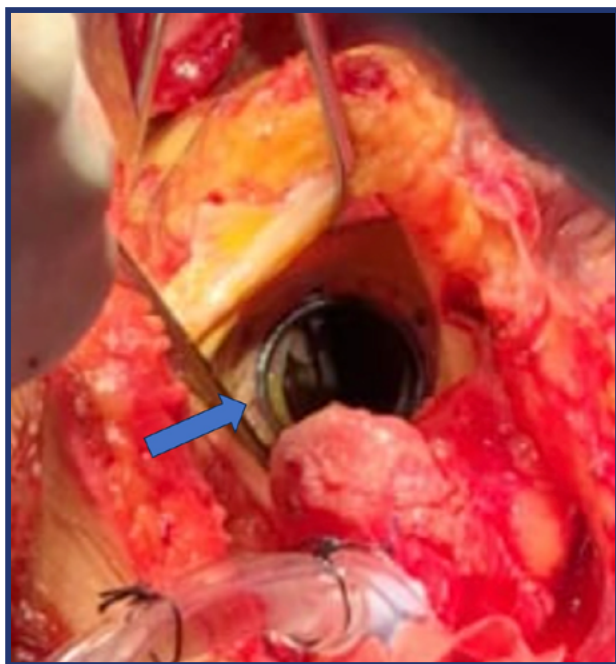
## DISCUSSION

The patient presented here had a porcelain aorta in its entirety, with severe calcification of the ascending aorta. This condition represents a surgical challenge and is associated with high morbidity during surgical aortic valve replacement to correct aortic stenosis.





**FIGURE 4.** Intraoperative photograph showing calcification of the entire circumference of the aorta.



**FIGURE 5.** Presence of *pannus* on the ventricular side of the valve prosthesis, leading to severe stenosis.

Calcification of the aortic walls limits the options for aortic clamping and cannulation and increases the risk of embolism when the aorta is manipulated. For this reason, identification of risk factors associated with complications is essential.

A porcelain aorta is not an absolute contraindication for aortic valve replacement and/or coronary bypass grafts. Still, it requires a special strategy and individualized approach to minimize the risk of embolic complications and technical problems during aortotomy opening and/or closure.

As described by Carrel et al,<sup>3</sup> the overall incidence of significant calcification of the ascending aorta is 5-8%

of patients scheduled for aortic valve replacement and/or coronary artery bypass graft placement. In our center, the incidence is 5% of cases. However, given the increased longevity of patients undergoing surgery and the diagnostic studies available today, the incidence may increase.

Porcelain aorta is generally considered a contraindication for conventional surgery because each aortic manipulation may involve a risk of calcific embolization, resulting in neurological complications.<sup>4</sup> For this reason, catheter-based procedures such as percutaneous aortic valve implantation (TAVI) are widely recommended, with good results.<sup>5</sup>

In this patient with mechanical prosthesis dysfunction in the aortic position, TAVI was not feasible. However, there are multiple strategies available to minimize the risk of complications during a surgical procedure in porcelain aorta; among them, individualize each case based on medical history (such as previous surgery). It is also essential to design various surgical plans with different therapeutic options before starting surgery, such as starting with femoral cannulation in reoperations and avoiding excessive manipulation of the aortic root, optimal cardiac emptying using ventricular drainage, moderate hypothermia to cause ventricular fibrillation, and infusing direct cardioplegia in the coronary *ostia* and avoiding cardioplegia cannula in the aortic root. If the patient has a history of previous surgery, it is recommended to perform the aortotomy, if possible, at the previous surgical site.

Kramer et al.,<sup>6</sup> compared the results of surgical versus transcatheter treatment. Analyses before and after the comparison demonstrated similar results, with slightly better outcomes, particularly in terms



of 5-year survival after surgery; this highlights the usefulness of surgical replacement with appropriate patient selection. Patients with smaller calcifications and pinchable aortas can often tolerate surgical replacement, which may be necessary in the context of concomitant disease. Regarding postoperative complications, improving tools for patient selection is critical to provide optimal treatment. Quantitative assessment of calcifications helps aid patient selection.

In the population of patients studied with porcelain aorta, there was a high prevalence of risk factors typical of atherosclerotic disease, history of peripheral artery disease, and coronary revascularization. Consequently, it is likely that, in this cohort of patients with porcelain aorta, the surgical risk was higher, given that porcelain aorta is not considered in risk scales such as EuroSCORE or STS.<sup>2</sup> This is important to consider from a purely surgical point of view, as risk is often underestimated when calculated on these scales. However, when these patients are evaluated with cardiohemodynamic studies, the risk is higher.

The case presented here is interesting because with comorbidities, heart failure, and chronic renal failure, the mortality rate is 75% when evaluated with the EuroSCORE; when the porcelain aorta is added, this rate increases to almost 100%. If we add to this the fact that the patient had a dysfunctional mechanical prosthesis, valve replacement can only be performed by a surgical approach, with an ominous prognosis. However, the postoperative outcome was satisfactory.

## CONCLUSION

Patients with porcelain aorta should be meticulously evaluated when any procedure is considered, regardless of the operative treatment approach. In

patients requiring concomitant procedures (as in this case, surgical replacement), it can be performed with a similar incidence of in-hospital complications and 5-year survival to those of the transcatheter procedure. The absence of a “clampable zone” and the increased total volume of calcifications are associated with increased morbidity and mortality.

## Declarations

The authors declare no conflict of interest.

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# VENTRICULAR MYXOMA: AN UNUSUAL PERIOPERATIVE FINDING

## ABSTRACT

Myxomas are benign cardiac tumors predominantly located in the left atrium and are more common in women aged 30 to 60 years. This article presents the case of a 36-year-old woman with a complex medical history who was diagnosed with an incidental finding of an intraventricular myxoma. Imaging studies, including echocardiogram and MRI, revealed cardiac masses. During surgery, lesions were found in the right atrium and a mass in the left ventricle. Pathology confirmed the diagnosis of cardiac myxoma, suggesting a possible association with Carney syndrome due to the patient's history. This case highlights the rarity and complexity of myxomas, underscoring their significance in clinical practice and the necessity for a multidisciplinary approach to facilitate early diagnosis and treatment.

**Keywords:** cardiac masses, myxoma, Carney syndrome.

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## INTRODUCTION

Myxomas are benign intracardiac tumors of primary origin; 75% are located in the left atrium (mitral annulus or edge of the fossa ovalis), followed by the right atrium (20%), and simultaneously atrioventricular (5%). Most are asymptomatic, although patients may have obstructive, embolic, and constitutional symptoms.<sup>1,2</sup> Its presentation is more frequent in women from the fourth to the sixth decades of life.<sup>3</sup> About 10% correspond to Carney complex or “myxoma syndrome”, generally in young female patients with unusual skin freckles; they are usually ventricular, multiple, and recurrent, associated with non-cardiac myxomatous tumors and endocrine tumors.<sup>4</sup> However, the exact etiology remains unclear. The following is a clinical case of an intraventricular mass that underwent cardiac surgery.

## CLINICAL CASE

The patient is a 36-year-old woman with a medical history of hypothyroidism, bilateral hypoacusis, hypoalbuminemia, bilateral superficial venous insufficiency, and chronic pigmentation of the lower limbs under study; surgical history of drainage of a collection in the left buttock and cesarean section; history in first-degree relative (brother) with elephantiasis and bilateral hypoacusis. The patient was admitted to the health institution for presenting with an abscessed cellulitis in the left buttock. With no other findings on physical examination on admission, she underwent drainage surgery. In the postoperative period, she presented grade I edema in the lower limbs, so studies were requested; among them, a transthoracic echocardiogram showed a mass dependent on the interventricular septum, with preserved right and left ventricular function, type II diastolic dysfunction, left atrium with mild dilatation, mitral sclerosis with traces of insufficiency, and mild tricuspid insufficiency, with systolic pressure of the pulmonary artery calculated at 33 mmHg.

Cardiac magnetic resonance imaging was then requested, which identified a mass attached to the interventricular septum, measuring 14 × 14 mm, with a thickening similar to that detected in the right atrium. The presumptive diagnoses were lymphoma, myxoma, or papillary fibroelastoma.

Elective surgery was indicated for resection of the intracardiac mass and pathological study of the specimen.

The pre-anesthetic assessment showed that the patient was ASA 3, with no predictors of difficult airway, high surgical risk, with EuroSCORE II, and an in-hospital mortality risk of 0.62%.

In the echocardiographic evaluation, a round intraventricular left mass was visualized, and another smaller one was in the right ventricle.

Under noninvasive vital signs and invasive blood pressure monitoring, conventional anesthetic induction was performed with lidocaine (1 mg/kg), propofol (2 mg/kg), fentanyl (4 mcg/kg), and rocuronium (1.2 mg/kg). Direct laryngoscopy (Cormack-Lehane I) and orotracheal intubation were performed. Anesthetic maintenance was performed with a balanced technique (sevoflurane and remifentanyl). Volume-controlled pulmonary ventilation was indicated, with a tidal volume of 6 mL/kg and an FiO<sub>2</sub> of 0.45. The surgical procedure was started with the patient on extracorporeal circulation.

Intraoperative transesophageal echocardiography showed preserved left ventricular function, without valvulopathies, the presence of multiple masses, one of 3 cm<sup>2</sup> pedicled and adhered to the interventricular septum, another of smaller size (0.4 cm) in the right ventricle adhered to the septum, infiltration and a possible mass in both inferior and superior vena cava towards the cresta terminalis posterior to the base of the pedicle in the interventricular septum (Figure 1).

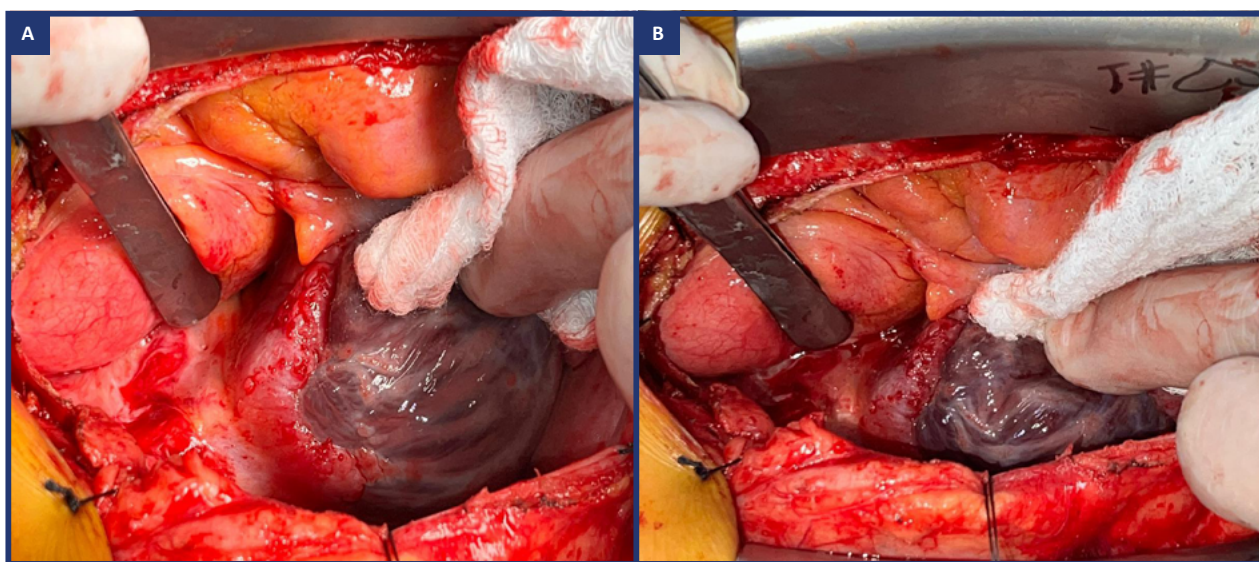
Surgery was performed by median sternotomy with findings of thickened cardiac tissue, vessels with thickened adventitia, two lesions with irregular borders of 5 × 6 × 3 cm in the right atrium, at the superior cavo-atrial junction and extending towards the appendage, and another lesion on the lateral aspect of 3 × 3 cm; these lesions were on the epicardium but infiltrated the adjacent tissues. No lesions were observed inside the right atrium; in the



**FIGURE 1.** Preoperative transthoracic echocardiogram: 3 cm<sup>2</sup> pedunculated mass attached to the interventricular septum.

left ventricle, a mass of 3 × 2 cm was observed adhered to the interventricular septum under the mitral valve, of yellow color and gelatinous consistency. Samples were sent for pathological study (*Figures 2A and 2B*). The lesions in the right atrium were not completely resectable because they infiltrated the entire wall. At the end of the surgical procedure, a bilateral spinal erector block was performed at the T5 level guided by ultrasound with simple bupivacaine 0.5%; the patient was extubated 10 minutes after anesthetic closure, under deep inspiration. The patient was transferred to the intensive care unit with an adequate ventilatory pattern, no

pain, and stable vital signs. No intraoperative or postoperative complications were observed, and the patient was discharged from the hospital on the fifth postoperative day. Microscopic analysis showed lesions compatible with cardiac myxoma. Further immunohistochemical evaluation showed a B-cell phenotype (positive for CD20, PAX5, CD79A) of atypical blast lymphoid cells, along with partial positivity for BCL6, IRF4, BCL2, CD30, and C-MYC (<40% of cells). It was negative for CD10, CD5, CD15, ALKc and HHV8, and a very high Ki67 proliferation index (>80%). In situ hybridization for Epstein-Barr virus was positive in a diffuse pattern.



**FIGURES 2A AND 2B.** Irregular border lesions measuring 5 × 6 × 3 cm at the superior cavo-atrial junction.

## DISCUSSION

Intracardiac tumor lesions are infrequent in healthy adults. They can be of primary or secondary origin. The incidence of primary intracardiac tumors is low (between 1.38 and 30 per 100,000 persons per year); 80% are benign, and 20% are malignant. Myxomas account for 50% of benign tumors, followed by lipomatous tumors (21%) and papillary fibroelastomas (16%).

The clinical presentation is variable, ranging from asymptomatic, incidental findings, signs of heart failure and shock, to sudden cardiac death. Influencing factors include the size, mobility, and location of the lesion within the cardiac cavity.<sup>5</sup>

Morphologically, myxomas are usually round, polypoid in appearance, lobulated, and have a smooth surface. Histologically, they contain primitive proliferating cells. On immunohistochemistry,

they are positive for vimentin, calretinin, S100, nonspecific enolase, factor VIII, CD31, and CD34.<sup>6</sup>

Approximately 10% of myxomas are associated with Carney syndrome, an autosomal dominant genetic disorder characterized by mutations in the *PRKAR1A* gene. These tumors tend to present at a younger age, with the involvement of multiple cardiac chambers and have a higher recurrence rate of up to 20%. In addition, thyroid involvement, benign or malignant, has been reported to occur in 25.7% of patients with Carney syndrome and cardiac myxoma.<sup>7-9</sup>

Based on the findings in the literature and the characteristics of the patient presented here (a young woman with thyroid and cutaneous involvement, who was found incidentally to have a ventricular myxoma and participation of the right ventricle, right atrium, and cavo-atrial junction), it can be

concluded that this is an atrial myxoma associated with Carney's complex.

The review and description of this clinical case enable us to update the clinical and paraclinical presentation of this disease, which is a rare and low-incidence situation of interest to the scientific community.

#### Declarations

The authors declare no conflict of interest.

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# RESOLUTION OF MITRAL ANNULAR DISJUNCTION THROUGH MINIMALLY INVASIVE SURGERY

## ABSTRACT

We present the case of a 60-year-old patient with a history of hypertension, diagnosed with mitral insufficiency in the context of functional class 3 dyspnea.

Mitral annular dysjunction is a structural anomaly characterized by the separation of the myocardium of the left ventricle and the mitral annulus that supports the posterior leaflet during systole. A strong link between mitral annulus disjunction and arrhythmogenic valve prolapse is recognized in this entity.

**Keywords:** mitral annular dysjunction, mitral valve, mini-invasive surgery.

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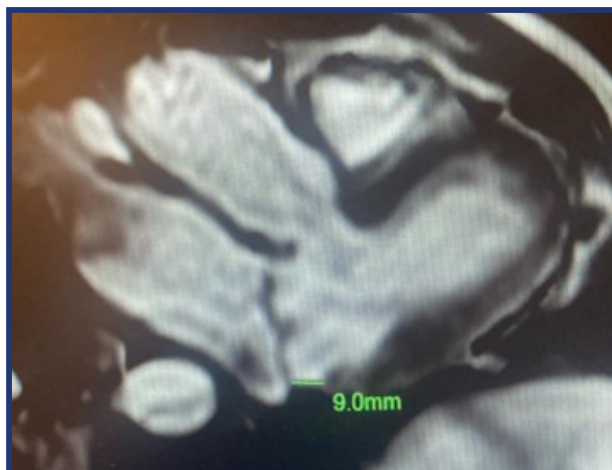
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## INTRODUCTION

Mitral annular disjunction is a structural anomaly characterized by the separation of the left ventricular myocardium and the mitral annulus supporting the posterior leaflet during systole. A clear association between mitral annulus disjunction and arrhythmogenic valve prolapse is recognized in this entity<sup>1-4</sup>. The first description of this pathology was in the 1980s by Hutchins et al.<sup>5</sup> after performing autopsies on deceased patients with mitral valve prolapse. This entity gained importance in 2005, when Eriksson et al.<sup>6</sup> reported that understanding this pathology was crucial for achieving better long-term results<sup>7</sup>.

## CLINICAL CASE

We present the case of a 60-year-old patient with a history of hypertension, diagnosis of mitral regurgitation, and functional class 3 dyspnea. In the preoperative studies, the echocardiogram revealed valve prolapse, a 9 mm mitral annulus disjunction, and severe mitral regurgitation, with no evidence of arrhythmic events. The EF was 65%, there was eccentric hypertrophy, non-dilated right chambers, mild tricuspid and pulmonary regurgitation (*Figure 1*). Magnetic resonance imaging showed a myxomatous mitral valve and 9 mm annulus disjunction, without motility alterations, with severe mitral insufficiency and mild aortic insufficiency. A cine coronary angiography was also performed, in which no significant lesions were observed.

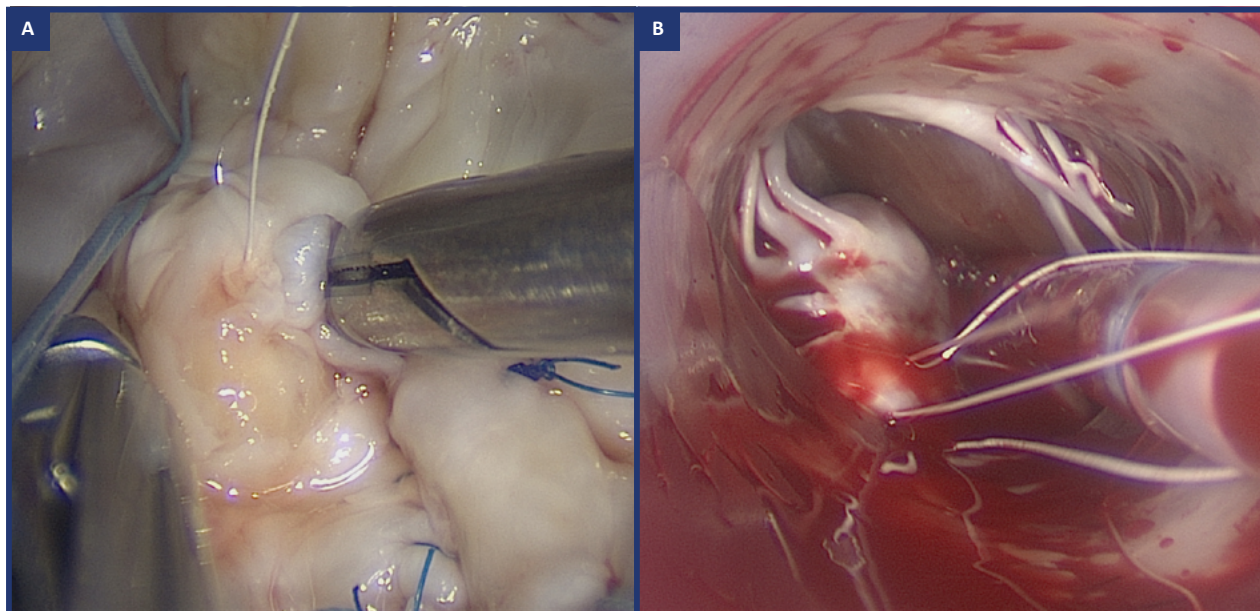


**FIGURE 1.** Echocardiogram showing atrial displacement of the mitral annulus plane.

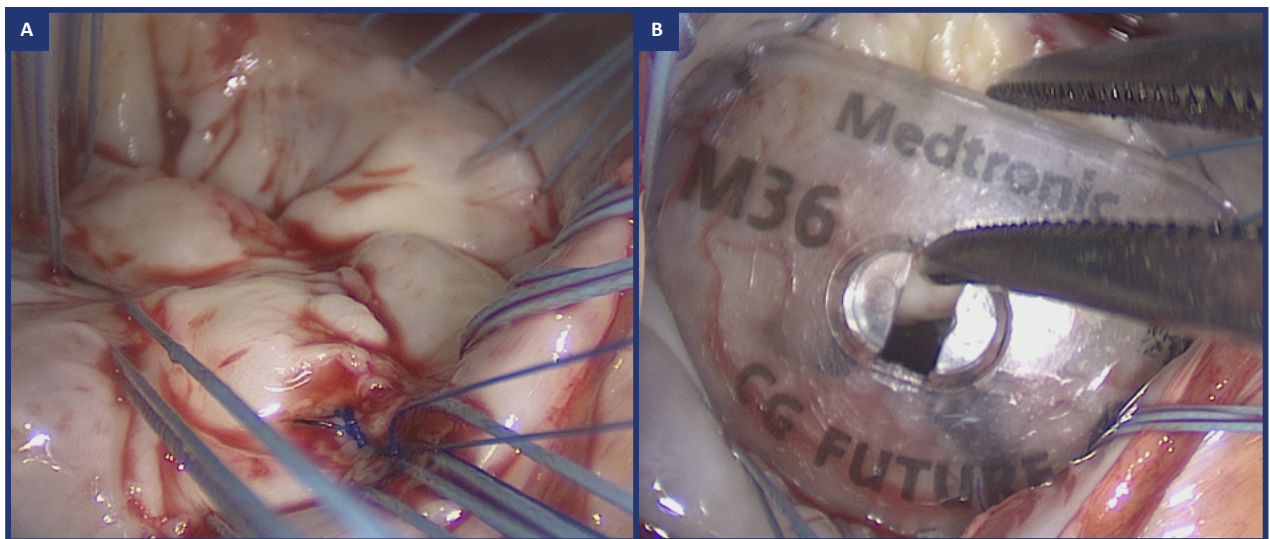
## TREATMENT

A minimally invasive video-assisted mitral valvuloplasty by right thoracoscopy was performed. The procedure consisted of quadrangular resection of P2 (*Figure 2A*), sliding of the posterior leaflet with incorporation of three pairs of neochords (*Figure 2B*), and fixation of the annulus to the ventricle with U-stitches with pledgets (*Figure 3A*) and a number 36 flexible ring (*Figure 3B*). The intraoperative control echocardiogram showed residual mild mitral regurgitation.

The patient underwent the postoperative period without complications; he remained for hours in the cardiovascular recovery room. He was discharged on the fourth day after surgery, without complications.



**FIGURE 2.** A: Quadrangular resection of P2. B: Neocord.



**FIGURE 3. A:** Annuloplasty points. **B:** Mitral annulus measurement with measuring device 36.

## DISCUSSION

The case presented here highlights the importance of early diagnosis and surgical intervention in patients with mitral annulus disjunction, as adequate repair can restore valve functionality, prevent deterioration of cardiac function, and reduce the occurrence of ventricular arrhythmias associated with this condition.

Minimally invasive surgery is presented as a practical option for the management of severe mitral regurgitation associated with annular disjunction, with good postoperative results and rapid recovery.

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## Declarations

The authors declare no conflict of interest.

# FIRST CASE OF BRANCHED THORACIC ENDOGRAFT TO THE SUBCLAVIAN ARTERY IN COLOMBIA

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## ABSTRACT

This case report describes an 83-year-old female patient with a history of recovered heart failure and multiple comorbidities, presenting a high risk of sudden cardiac death. She was referred to after experiencing a cardiogenic syncope, which led to the placement of a bicameral pacemaker. Additionally, a fusiform aneurysm was detected in the descending aorta, near the left subclavian artery. Treatment was performed via endovascular intervention using a second-generation device with a branched stent in the left subclavian artery, yielding favorable good outcomes.

**Keywords:** *descending aortic aneurysm, subclavian artery, endovascular intervention.*



## INTRODUCTION

Endovascular surgery has become the first-line treatment for aortic diseases across all its sections, including both the thoracic and abdominal regions, since the early cases<sup>1,2</sup>. Aortic arch lesions have historically represented a significant challenge<sup>3</sup>, and continue to pose new challenges. Hybrid surgery, which combines a less aggressive endovascular treatment of the aortic arch and bypasses of the supra-aortic trunks, partially or totally, is currently a viable alternative. In this context, branched stent techniques offer new therapeutic possibilities.

## CASE REPORT

This is the case of an 83-year-old female patient referred for cardiogenic syncope. Her medical history includes chronic hypertension, type II diabetes mellitus, non-oliguric chronic kidney disease, permanent atrial fibrillation, heart failure with reduced ventricular function, myocardial

revascularization surgery, and percutaneous coronary intervention<sup>4</sup>. She was referred from another facility due to syncope and a thoracic aortic aneurysm; she was admitted after experiencing cardiogenic syncope, which required the implantation of a bicameral pacemaker at the site of origin due to the risk of sudden cardiac death. A fusiform aneurysm was found in the descending aorta, with angiographic imaging (*Figure 1*) showing aneurysmal dilation of the thoracic aorta in the proximal segment near the left subclavian artery, measuring 54 mm, and a distal aneurysm, proximal to the superior mesenteric artery, measuring 45 mm (*Figures 2 and 3*). Treatment was decided upon involving endovascular repair of the thoracic aorta using a branched stent, followed by a review by the electrophysiology team due to a 200 ml hematoma at the pacemaker insertion site. This led to the decision to explant the device and perform a new contralateral implantation.



**FIGURE 1.** Descending aortic aneurysm near the left subclavian artery.



**FIGURE 2.** Descending aortic aneurysm near the superior mesenteric artery.



**FIGURE 3.** Angiotomographic image showing distal and proximal involvement.

## RESULTS

Emergency surgery was performed, it was decided not to perform a cervical debranching<sup>5</sup>, and to perform endovascular repair treatment of the thoracic aneurysm with a branched stent, given the critical conditions of the patient, her longevity and to minimize the high risks of morbidity and mortality; Castor™ stent with subclavian branch, made of braided polyester and

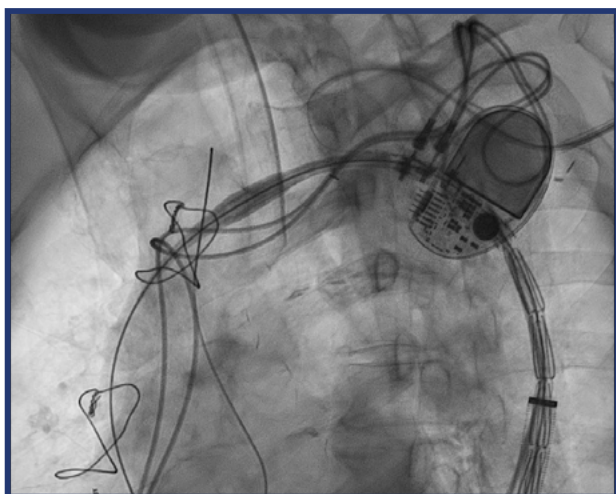
nitinol, placed in zone 2, was used. The bicameral pacemaker was repositioned, and the hematoma was drained, with a sample taken for culture; the result was negative. The pacemaker generator was placed in the retropectoral space (reimplantation), and the wound was closed. The right common femoral artery was then dissected and repaired, with placement of a 6F introducer. The left humeral artery was punctured



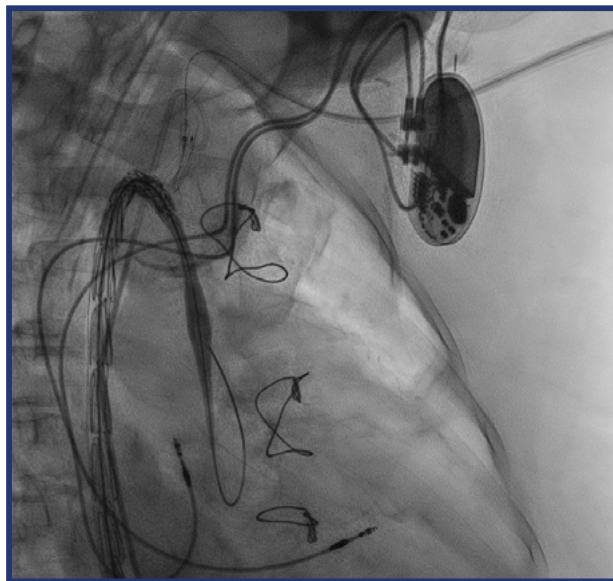
with an 8F introducer, and the left femoral artery with a 6F introducer. In the thoracic aortogram, an aneurysm was observed in sector 4 of the thoracic aorta, with extension towards the splanchnic plane and severe atheromatosis in the iliac arteries and abdominal aorta (*Figures 4 and 5*). The subclavian branch was modeled with a Medtronic™ 9 × 20 balloon (*Figure 6*). Postoperative control showed adequate exclusion of the aneurysm in sector 4, without endoleaks, without displacements, and with patency in the subclavian artery (*Figure 7*). In the abdominal aortogram, adequate patency of the splanchnic vessels was observed, without dissections (*Figure 3*). A right femoral arteriography was performed with extensive endarterectomy and closure of the arteriotomy with Prolene 6/0™. Hemostasis of the access sites was achieved using fibrillar SurgiSeal™, and the access wounds were subsequently closed. During the procedure, the patient presented elevated blood pressure, which required infusion of nitroglycerin at 0.5 mcg/kg/min. Subsequently, she was transferred to the ICU for intensive management and hemodynamic monitoring. The patient made a satisfactory recovery, without neurological symptoms, and was discharged four days after surgery.

## DISCUSSION

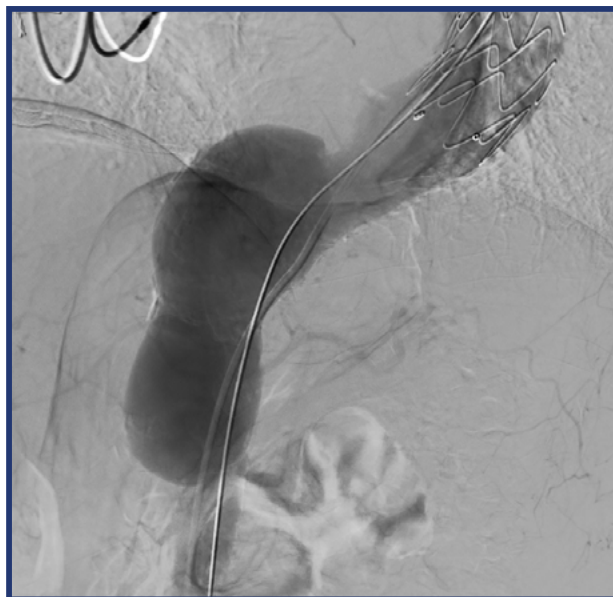
Endovascular treatment with state-of-the-art devices, such as branched stents, combined with extra-anatomic cervical revascularization of supraoptic vessels, is a valuable therapeutic option, especially for elderly patients with multiple comorbidities<sup>6</sup>. This strategy reduces the high risks of stroke, renal failure, paraplegia<sup>7,8</sup>, and death<sup>4</sup>, improving survival and offering a definitive and less invasive solution for complex patients. However, potential long-term complications should also be considered, such as stent collapse with risk of sudden death, the need for rescue surgeries or poor sealing at the anchor site<sup>9,10</sup>.



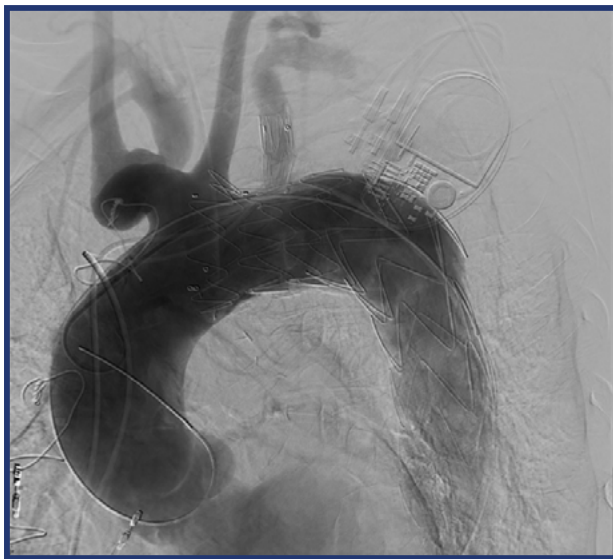
**FIGURE 4.** Device advancement in the descending aorta.



**FIGURE 5.** Lateral view.



**FIGURE 6.** Deployment of the endoprosthesis.



**FIGURE 7.** Total exclusion of the aneurysm.

## CONCLUSION

The latest generation of thoracic endografts, especially those with branched devices, has proven to be a valuable tool in the treatment of aneurysmal diseases. Their applicability, both in the short and long term, represents an effective therapeutic option, particularly in the population with high morbidity and mortality.

## Declarations

The authors declare no conflict of interest.

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