

Interrupted aortic arch. A hybrid management: surgical-endovascular

Interrupción del arco aórtico. Un manejo híbrido: quirúrgico-endovascular

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ABSTRACT

Complex heart diseases represent a challenge for the cardiothoracic surgeon. Interruption of the aortic arch in adults is usually rare. The coexistence of arch interruption with other heart diseases that occur at this age is even rarer. The case presented here allowed surgical management by means of the interposition of a dacron graft, total correction of the common truncus arteriosus and placement of an endoprosthesis using endovascular therapy.

Keywords: aortic aneurysm, common arterial truncus, congenital heart disease, endovascular therapy, interruption of the aortic arch.

RESUMEN

Las cardiopatías complejas representan un reto para el cirujano cardiotorácico. La interrupción del arco aórtico en el adulto suele ser infrecuente. La coexistencia de interrupción del arco con otras cardiopatías que se presentan en esta edad es aún más raro. El caso aquí presentado permitió un manejo quirúrgico mediante la interposición de un injerto de dacrón, corrección total de tronco arterioso común y colocación de endoprótesis mediante la terapia endovascular.

Palabras clave: aneurisma aórtico, tronco arterioso común, cardiopatía congénita, terapia endovascular, interrupción del arco aórtico.

Interrupted aortic arch (IAA) is a rare heart disease, occurring approximately three times per million births.¹ It is the lack of continuity between the ascending and descending aorta. Rarely a condition that manifests in isolation. It is common to find AAI in association with other cardiac defects, for example: patent ductus arteriosus, ventricular septal defect mainly ventricular septal defect, left ventricular outflow tract obstruction, aorto-pulmonary window, aberrant innominate arteries.²

According to the classification of Celoria and Patton described in 1959, it is classified into three types: A, B and C.

Type A is a discontinuity of the distal arch to the left subclavian artery at the level of the aortic isthmus, type B is a discontinuity between the left common carotid artery and the left subclavian artery, and type C refers to a discontinuity that occurs between the innominate artery and the common carotid artery. Type A is the most commonly reported in adult cases at 79%, while type B is the most commonly detected in the neonate at 53%.³

Common truncus arteriosus, truncus arteriosus from its Latin name, is a rare cyanotic congenital heart disease.⁴ It has an estimated incidence of 1 in 10,000 births. It is described as a single arterial trunk arising from the heart through a

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single arterial valve. This artery is responsible for pulmonary, systemic and coronary circulation.⁵ Different classifications of the defect have been described. The first known classification was elaborated by Collett and Edwards in 1949, based on the disposition of the origin of the pulmonary arteries from the common trunk.⁶ Type I truncus arteriosus: there is a single pulmonary trunk connecting the right and left pulmonary arteries, this is the most common type. Type II: each main pulmonary artery has its own common origin in the trunk. Type III: each main pulmonary artery originates separately on the lateral side of the common trunk. Type IV: no pulmonary branch arises from the trunk, the pulmonary supply arises from collateral systemic arteries.⁷ As for the Van Praagh classification, specifies the presence or absence of ventricular septal defect. The classification involves in the nomenclature the letter A for presence and B for absence. Type I and type II are similar to that described by Collett and Edwards. Type III: describes a branch of the pulmonary artery that originates in the trunk and the rest of the lung irrigation through collaterals or branches arising from the aortic arch. IV: corresponds to a hypoplastic aortic arch, interrupted or with a patent ductus arteriosus.⁸

CASE DESCRIPTION

We present the case of a 37-year-old female patient whose personal pathologic history includes the detection of a murmur during infancy, and who required hospitalization during birth. Without further information provided by the patient, the complete cardiologic history is unknown. Her current condition began with dyspnea on medium exertion, chest tightness, palpitations, nausea, headache with intensity 9/10 of oppressive type, tinnitus and lipotimia for 15 consecutive days. She went to a local hospital in her city of origin where she was hospitalized due to the symptoms described above. During her hospitalization she developed syncope and diaphoresis.

Cardiogenic shock and atrial fibrillation of medium ventricular response was detected, being managed by pharmacological cardioversion with amiodarone, later requiring norepinephrine and dobutamine to maintain perfusory mean arterial pressure. She was sent to the hospital unit where she was asymptomatic and hemodynamically stable after transfer.

A first transthoracic echocardiogram was performed with generalized dilatation in the four chambers, decreased contractile dynamics, vena cava with diameters of 21 mm, distensibility of 33%. Subsequently, upon admission to our hospital unit in charge of cardiology, a second transthoracic echocardiogram was performed. Cardiomyopathy in dilated phase of undetermined origin, left ventricular systolic dysfunction with left ventricular ejection fraction of 43%, with abnormal global and segmental contractility due to global hypokinesia. Severe dilatation of the right chambers. Moderate mitral and tricuspid insufficiency, mild aortic insufficiency and severe pulmonary insufficiency. Severe pulmonary arterial hypertension due to pulmonary artery systolic pressure of 100 mmHg. Diagnostic cardiac catheterization was performed by interventional cardiology with the results of severe pulmonary arterial hypertension unresponsive to oxygen challenge, and an aortogram with anatomy compatible with common arterial trunk type I was performed (*Figure 1*). Angiotomography with reconstruction of large vessels was performed, where the interruption distal to the subclavian artery and an aneurysmal dilatation were identified (*Figure 2*). It was decided to evaluate the patient by the cardiac surgery service.

For surgical resolution the sternotomy approach was performed. A 7 mm dacron graft anastomosis is performed at the right subclavian level. Arterial cannula of 19 mm is placed. Sternotomy is performed, finding multiple collateral vessels, subsequent opening of the pericardium, supra-aortic trunks are surrounded. Bicaval venous cannulation is performed. Heparinization and initiation of cardiopulmonary bypass. Temperature decreases to 28 °C, emptying (continuous retrograde

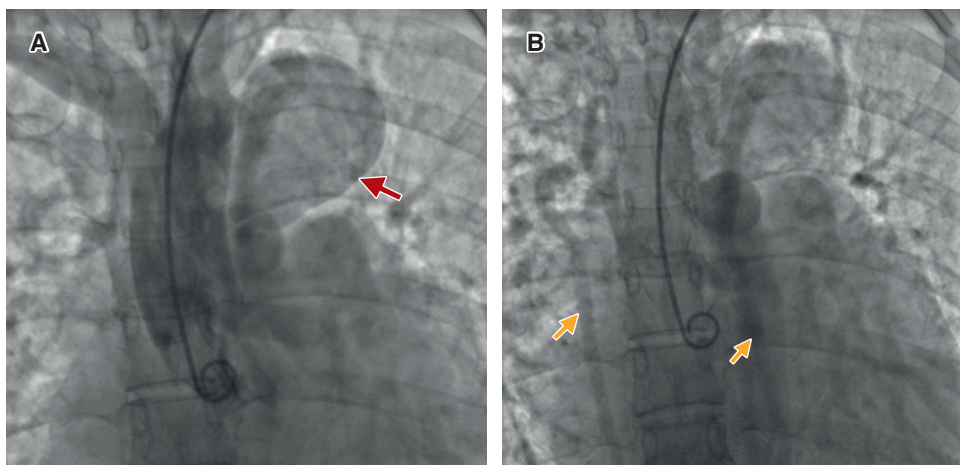


Figure 1:

A) Evidence of aneurysm in catheterization. **B)** Collateral vessels evidenced during catheterization.

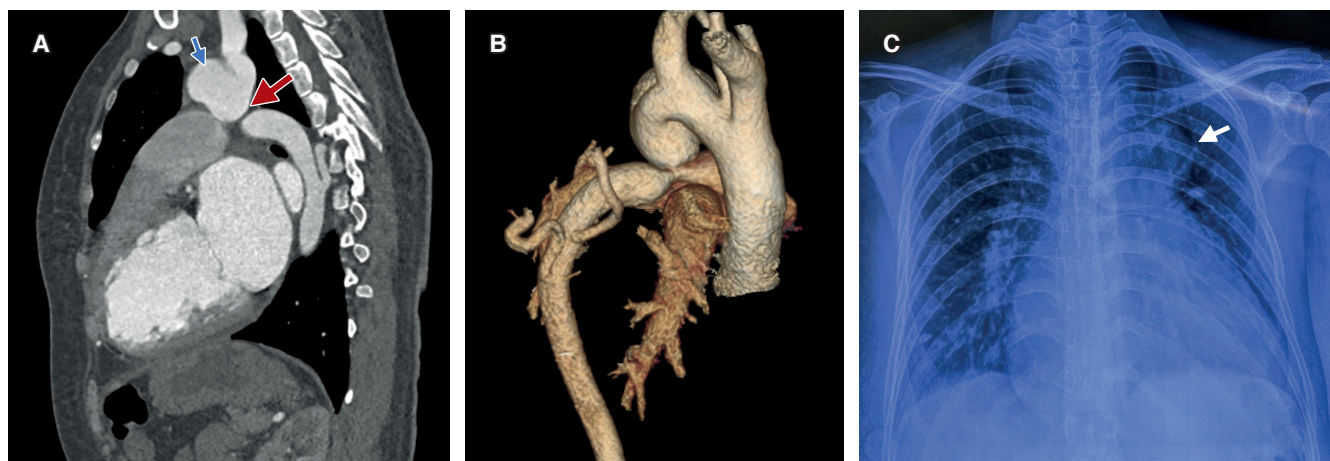


Figure 2: **A)** Sagittal CT scan showing aneurysm (blue arrow) and interruption of the aortic arch (red arrow). **B)** Reconstruction of great vessels. **C)** Preoperative chest X-ray showing a huge mass (white arrow).

perfusion through the brachiocephalic trunk). Aortic clamping and initiation of cardioplegia. Supra-aortic trunks and aortic isthmus are dissected up to their most distal portion. Aneurysm with fistula to the pulmonary artery trunk is identified. Aneurysm dissection of distal end of interruption of the aortic arch is performed, as well as pulmonary artery trunk identifying conduit. The aneurysm is resected, the pulmonary trunk is opened, the duct is sectioned and the descending aorta is identified. A 20 mm dacron graft is placed after placing the bigoteral in the distal end of the aortic mouth performing anastomosis with prolene 3-0 surgete, proximal anastomosis same technique (Figure 3). After clamping, bleeding occurred at the distal site of the anastomosis, so a 26 × 10 mm zenith endoprosthesis was positioned under fluoroscopy. Hemostasis was corroborated, epicardial cables, drains and sternal closure were placed. The findings were aneurysm in proximal end of 5 × 8 cm, interruption of the aortic arch type A and type I truncus arteriosus without ventricular septal defect. The pump time was 3 hours and 28 minutes and the aortic clamping time was 2 hours and 2 minutes. After surgery, the patient was admitted to the intensive care unit (Figure 4). He died fourteen days after surgery due to septic shock.

COMMENT

A complex congenital heart disease represents an operative challenge. The arteriosocomun trunk is a complex cardiopathy that represents a low percentage of congenital cardiopathies, less than 1%, other references determine that.⁹ Meanwhile, AAI represents a low incidence of 3 in 1'000,000 live births.¹⁰ We present here the case of a patient in the fourth decade of life, who presents a complex heart disease, involving interruption of the aortic arch, common truncus arteriosus and patent ductus arteriosus. In addition to the findings, an

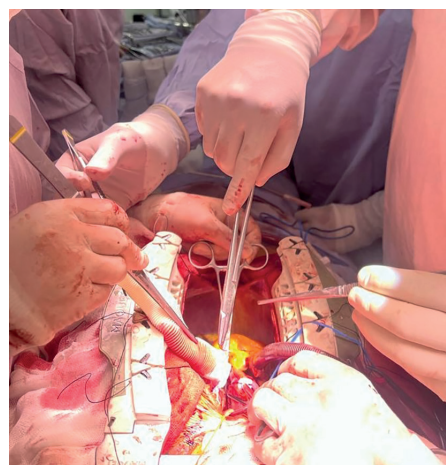


Figure 3: Dacron graft interposition.

aneurysm developed at the proximal level of the interruption. The occurrence of aneurysms in coexistence with congenital heart disease is extremely rare.

In the literature have been recently described two cases that have some similarity to the case presented here. One of the cases reported in the literature reported a male patient in the sixth decade of life, with a diagnosis of type A thoracic arch interruption and post-interruption thoracic aortic aneurysm. In this patient no ductal finding was described. The clinical presentation in this patient was characteristic of dyspnea and wheezing, as well as a history of arterial hypertension. Surgical resolution was by sternotomy, with interposition of a termino-terminal dacron graft.¹¹

Another case found in the literature corresponds to a woman in her sixth decade of life with a diagnosis of

interrupted aortic arch with post-interruption aneurysm and bicuspid aortic valve. As in the previous case, there was a history of hypertension previously. The clinic involved decreased exercise tolerance, as well as claudication in the lower extremities. In this case, the patient did not receive any medical-pharmacological or medical-surgical care.¹² A case found which does not correspond to a rupture with aneurysm, is the one published by Ture, et al,¹³ where the 5-month-old patient, who presented the diagnosis of common truncus arteriosus in coexistence with ruptured aortic arch type B and crossed pulmonary artery. The resolution was surgical, the aortic interruption was repaired, the Lecompte maneuver was performed, where the anterior pulmonary artery was brought to the aorta. It represents a rare case. In the presentation of our case, in comparison with the cases found in the literature, we can find that our patient did not present with the classic clinic in patients with interruption in adulthood. Patients who prevail into adulthood with this malformation usually present with hypertension, claudication, cardiac insufficiency.⁷ Open repair of the interruption allows the interposition of a graft in the interruption, in the specific case of our case involving an aneurysm proximal to the interruption, allowing correction of this defect.

In our case, an endoprosthesis was placed after graft interposition. Initially, endovascular management had been planned. Due to the conditions and the complexity of the cardiopathy, it was decided to use sternotomy, with hybrid management of the endoprosthesis placement. The type of endovascular prosthesis used in this case was the Cook Medical Zenith Alpha Thoracic® (Cook Medical, Bloomington, Indiana). This device is a modular and tubular endovascular grafting system. It allows its adaptability to tortuous anatomies and tortuous access vessels. It can be used as an auxiliary component to increase the length by overlapping or extending

the coverage of a graft and is adaptable to the curvature of the aortic arch.¹⁴ This last proposed use is the one attributed to it during our surgical procedure. As it is a device that allows overlapping, it allows bleeding to be controlled and provides support to the distal portion of the graft. The preparation of cardiothoracic surgeons in training to achieve mastery of the new techniques is fundamental. The management of endovascular techniques is fundamental in preparation. Not only does it involve preparation and knowledge of devices, but skill acquisition is critical. Often, this acquisition of these skills is implemented in descending order of hierarchy. A great example of the preparation of cardiothoracic surgeons is the Canadian system, where it has allowed flexibility to the contemporary surgical environment.¹⁵ A fundamental part of the program involves familiarization with endovascular surgery, as well as rotations that allow for the development of skills in transcatheter aortic valve replacement.

CONCLUSIONS

Few cases have been described of over interruption of the aortic arch and the development of aneurysms. The case presented here, allowed surgical management by interposition of a dacron graft, total correction of common truncus arteriosus and stenting by endovascular therapy. This allows a hybrid management of the pathology. As innovations develop, the cardiac surgeon has more similarities with vascular surgeons. This advancement is a constant challenge to the cardiac surgeon's skill development.

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Figure 4: Postoperative radiography in intensive care.

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