CASE REPORT
Vol. 9 No. 2 April-June 2024
doi: 10.35366/115157

Aortic coarctation versus aortic arch interruption: not all is what it seems

Coartación aórtica versus interrupción del arco aórtico: no todo es lo que parece

José J. Parra-Salazar,* Elizabeth Vera-Domínguez*

* Department of Surgery. Hospital General de Puebla "Dr. Eduardo Vázquez Navarro". Puebla, Mexico.

ABSTRACT

Interruption of the aortic arch is a rare congenital heart disease, representing 1% of the series. We present here the case of a young male patient with hypertension refractory to treatment, precordial pain, headache, decreased pulses in lower extremities. Initially diagnosed with aortic coarctation, surgical resolution was made. Aortic coarctectomy was planned. However, after surgical examination in the operating room, an interrupted aortic arch was found, and Dacron graft interposition was performed.

Keywords: aortic coarctation, congenital heart disease, interrupted aortic arch.

RESUMEN

La interrupción del arco aórtico es una cardiopatía congénita poco frecuente, representa el 1% de éstas. Se presenta el caso de un paciente masculino joven con hipertensión refractaria a tratamiento, dolor precordial, cefalea, disminución de los pulsos a nivel de extremidades inferiores. Inicialmente se diagnosticó como coartación aórtica, y se realizó cirugía. En un principio estaba planeada una coartectomía aórtica, sin embargo, tras la exploración quirúrgica, se evidenció un arco aórtico interrumpido, realizando una interposición de injerto de Dacrón.

Palabras clave: *coartación aórtica, cardiopatía congénita, interrupción del arco aórtico.*

INTRODUCTION

Interrupted aortic arch (IAA) is a rare heart disease, occurring approximately three times per million births.[1] Other sources suggest that it is found in 1% of congenital heart disease. We can define it as the lack of continuity between the ascending and descending aorta.

Its presentation is very common during the first weeks of life. Early intervention in the first weeks is mainly required to improve patient mortality.[2] It is rarely a condition that manifests in isolation. Most commonly, IAA is found in association with other cardiac defects such as patent ductus arteriosus, ventricular septal defect mainly ventricular septal defect, left ventricular outflow tract obstruction, aorto-pulmonary window, aberrant innominate arteries.[3] The main differential diagnosis in these patients is aortic coarctation, which is defined as an eccentric narrowing of an aortic focal segment.[4]

Anatomically, IAA can be classified into three types; type A refers to a discontinuity of the arch distal to the left subclavian artery at the level of the aortic isthmus, type B is a discontinuity between the left common carotid and the left subclavian artery, and type C refers to a discontinuity that occurs between the innominate artery and the common carotid

How to cite: Parra-Salazar JJ, Vera-Domínguez E. Aortic coarctation versus aortic arch interruption: not all is what it seems. Cir Card Mex. 2024; 9 (2): 52-56. https://dx.doi.org/10.35366/115157

Received: 07-22-2023. Accepted: 09-07- 2023.

Correspondence: Elizabeth Vera-Domínguez, MD. E-mail: elizabethvera988@gmail.com

artery.[5] In adulthood, type B is more common in 53% of cases, followed by type A in 43%, and type C in 4%.[6]

CLINICAL CASE

A 17-year-old male during childhood she presented with atypical precordial pain recurrent to physical activity, with subsequent cessation of symptoms at six years of age. At 17 years of age, began again with precordial symptomatology, with exacerbation and progression of precordial pain on two occasions, with intensity 10/10, lasting 15 minutes. In the last episode, a holocranial pulsatile headache with intensity 8/10 was present.

The patient was evaluated by Cardiology. A grade I/II systolic murmur was identified, with irradiation to the scapular region. On palpation of the extremities, absence of pelvic limb pulses was found. Computed tomography (CT) angiography with reconstruction was performed, showing para-intervertebral, internal mammary and bilateral axillary circulation, with intraluminal reinforcement upon administration of iodinated radiopaque, and evidence of coarctation of the proximal descending aorta *(Figure 1)*.

A transthoracic echocardiogram reported critical post-ductal aortic coarctation, bicuspid aortic valve with mild regurgitation, mild dilatation of the aortic root, left ventricular ejection fraction 60%, pulmonary arterial systolic pressure 17 mmHg, without data of pulmonary hypertension nor associated shunt. There was evidence of a left aortic arch with presence of coarctation at 39 mm from the emergence of the left subclavian artery, with narrowing up to 6 mm, with maximum gradient of 11 mmHg. A second control transthoracic echocardiogram was performed demonstrating situs solitus, levocardia, no atrial septal defects, no ventricular septal defects, left ventricular ejection fraction 83%, bicuspid aortic valve with mild dysplasia, left aortic arch with moderate hypoplasia and juxta-ductal aortic coarctation.

The patient was evaluated by cardiothoracic surgery and scheduled for surgical resolution of aortic coarctation by means of aortic coarctectomy.

Surgical resolution was approached by left posterolateral thoracotomy through the 3rd left intercostal space, without extracorporeal circulation. Dissection of the thoracic aorta and the isthmus up to the emergence of the brachiocephalic trunk was performed, as well as control of intercostal arteries in the thoracic aorta. Adequate control of the intercostal arteries was key to avoid bleeding due to the large number of collateral vessels developed. Apparent coarctation of the aortic arch distal to the left subclavian artery was observed.

Aortic arch and thoracic aorta clamping were performed. Before resolution of the aortic interruption, the ductus arteriosus was sectioned and sutured with 5-0 polypropylene. Subsequently, the hypoplastic segment was sectioned. An anastomosis is performed with a 16 mm Dacron graft with 4-0 polypropylene and a polytetrafluoroethylene (PTFE) felt patch is placed at the end of the aortic arch. The PTFE patch was placed at the junction between the end of the aortic arch and one end of the Dacron graft. This PTFE felt patch dressing or also called "Bigotera" will function as a reinforcement at the suture line of the anastomosis. The same technique was used to perform the anastomosis in the distal segment of the thoracic aorta *(Figure 2)*. Once both anastomoses were performed, hemostasis was verified. Surgical sealant (CoSeal) was placed in both graft anastomoses, pleural drains were inserted and surgical closure was carried out.

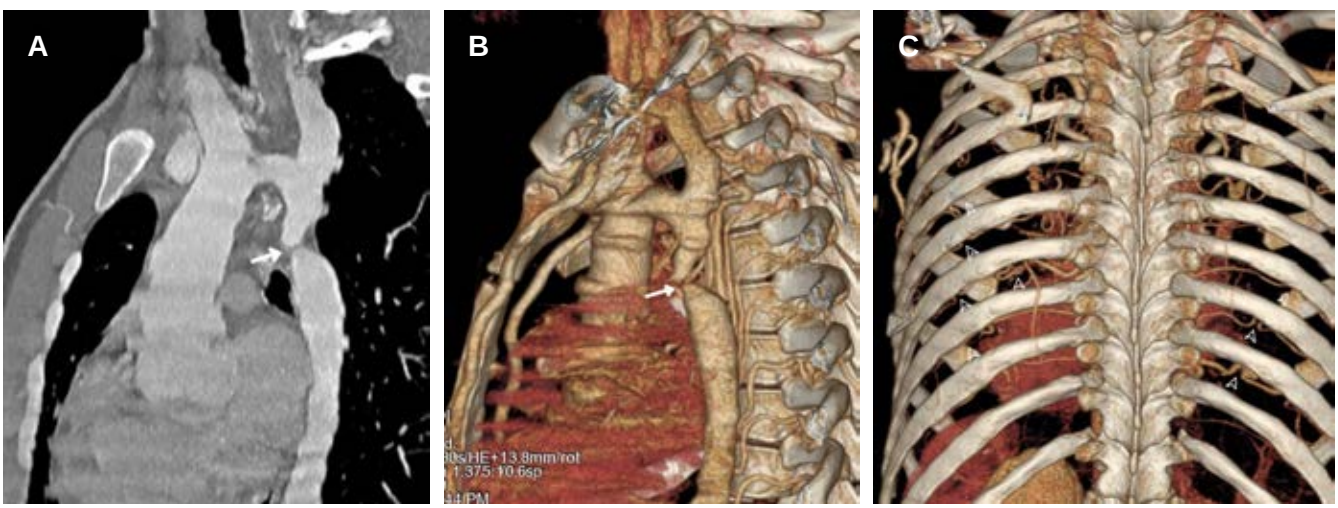

Figure 1: Preoperative angiotomography demonstrating defect distal to the left subclavian artery. A and B) White arrows indicating the defect. C) Arrow-heads on the costal ridge indicating increased flow in the posterior intercostal arteries, which generates erosion of the inferior costal border.

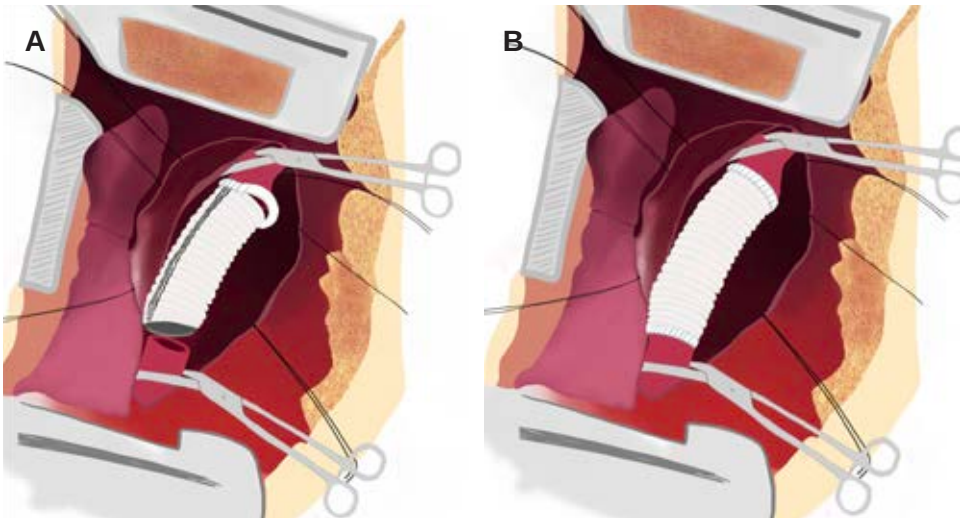

Figure 2:

Animated representation of Dacron graft interposition. A) Depicts proximal anastomosis of the graft to the aortic arch and placement of the polytetrafluoroethylene (PTFE) patch by continuous suture. B) Representation of the result of the interposition of the Dacron graft with the placement of PTFE patches for the reinforcement of both ends of the anastomosis.

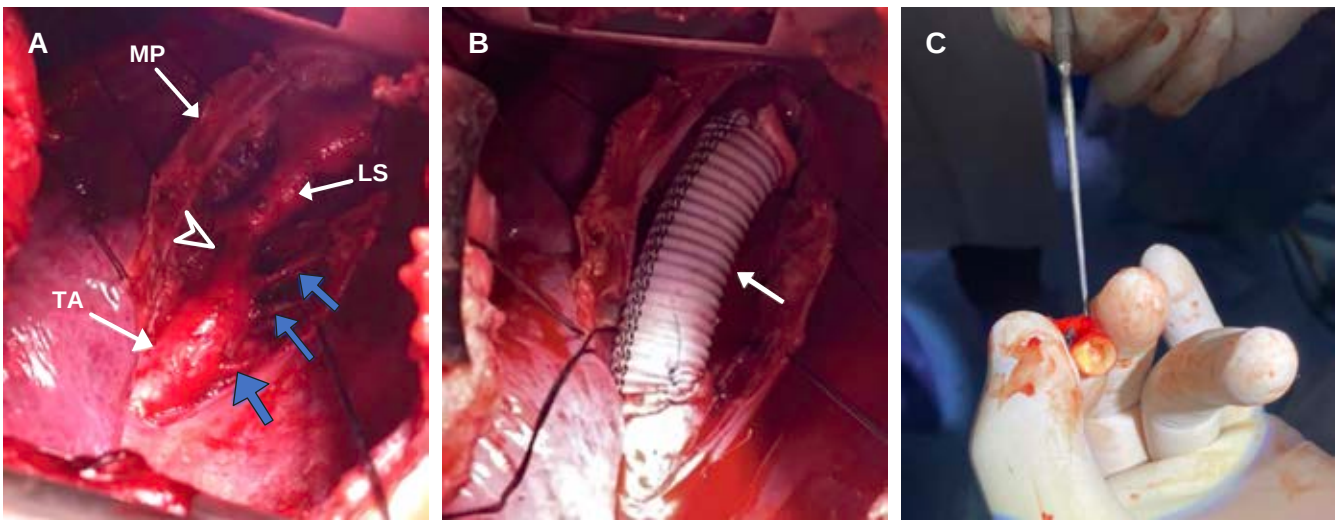

Figure 3: A) Image showing the defect (arrow head) prior to defect repair showing left subclavian artery (LS), vertebral arteries (blue arrows). B) Image showing defect repair with 16 mm Dacron graft (white arrow). C) Exploration of the hypoplastic segment with the nerve hook. The lumen of the defect segment was not perforated.
MP = mediastinal pleura. LS = left subclavian artery. TA = thoracic aorta.

The aortic cross-clamp time was 54 minutes. The patient was evaluated in the operating room by anesthesiology with transesophageal echocardiography and a residual gradient of 5 mmHg was identified.

Once the procedure was completed, the hypoplastic segment was explored. When attempting to explore the segment with the nerve hook, the segment did not pass through the section of what was believed to be the coarctation. At that point, an aortic arch interruption was diagnosed in the operating room, as opposed to an aortic coarctation as preoperatively diagnosed *(Figure 3)*.

A transthoracic echocardiogram during the first day in the intensive care unit reported no data of pulmonary hypertension, no shunts observed, preserved systolic function with ejection fraction of 56%. At the level of the thoracic aorta, a prosthetic tube with laminar flow, without gradient and without leakage was observed. Postoperative CT angiography was made reporting vascular structures at the thoracic level and its collaterals within the normal range, prosthetic tube at the level of the origin of the descending aorta intact and without contrast leakage *(Figure 4)*.

Postoperative course was uneventful. Length of stay in the intensive care unit was three days. In-hospital stay was seven days. Follow-up was performed two weeks after hospital discharge at the outpatient clinic. Patient evolution has been favorable without requiring subsequent rehospitalization.

COMMENT

Interrupted aortic arch is a rare congenital heart disease in which there is an interruption of the lumen and anatomical continuity between the ascending and descending arteries of the aorta.[3] The most common type found in adulthood is type A, in 79%. Such as the one found in the case presented here, type A, distal to the left subclavian artery. Due to this disposition of the defect, there is a difference in oxygen saturation where the upper extremities have greater oxygenation than the lower extremities.[5] IAA is considered incompatible with life when the ductus arteriosus closes and the defect is not corrected. Most cases of IAA are associated with patent ductus arteriosus in 98% of cases.[4] The survival of cases where the interruption persists into adulthood is due to the development of an extensive collateral network, which is vital for maintaining distal flow. Patency of the ductus arteriosus is another key point that allows survival in adult patients.[7]

The presence of IAA, patent ductus arteriosus and ventricular septal defect has been termed the triad of interrupted aortic arch.[3] In the case we presented herein, the ductus arteriosus was present, but with absence of ventricular septal defect.

According to the cases reported in a review by Gordon et al.[1] where IAA persisted into adulthood, the age range for surgical correction was 18 to 72 years. Of 38 cases, only five were in the age range of 18 to 21 years, such as our case. Predominant sex in the revision was male. Our case presented here was a male.

The diagnosis is mainly suspected on clinical presentation of symptoms. The most common finding is hypertension refractory to treatment. Another finding on physical examination is the discrepancy in blood pressure in the extremities. On measurement, the pressure in the lower extremities is usually more diminished. Other symptoms include claudication, evidence of aortic insufficiency and congestive heart failure.

The studies of cabinet that support the diagnosis are the radiography of thorax of first instance. This study usually demonstrates notches at the level of the inferior border of the costal arches. This is known as Roesler's sign, due to tortuosity in the intercostal arteries, due to increased flow. A transthoracic echocardiogram with Doppler is performed to identify the discontinuity in the flow. Magnetic resonance angiography helps to demonstrate heart malformations.[3] Especially in our case presented here, a diagnostic echocardiogram was performed without the presence of an interruption. Coarctation is evidenced presenting a gradient, so initially the diagnosis of aortic coarctation is given by cardiology.

Although the initial proposal was to treat the patient by interventionist methods, after diagnostic doubt, surgical approach was considered. In a series of cases analyzed by Gordon et al.,[3] 54% of the adult patients with IAA underwent surgical treatment by lateral thoracotomy sternotomy for its correction. Eight percent received interventional management by stents and 10% refused surgical management and received only medical treatment. Pharmacological therapy was applied to reduce pulmonary hypertension.

A review by Krishna et al.[8] analyzed the experience in one center with a population of 12 patients. Ten extra-anatomic bypasses were performed in a single stage. Of this group, five patients underwent ventral aortic repair via midline sternotomy. This approach was preferred due to excessive bleeding with the left posterolateral thoracotomy approach. This technique avoids the extensive net of collateral vessels in the chest wall. In two patients, a descending thoracic to subclavian aortic bypass graft was performed. These patients were boarded

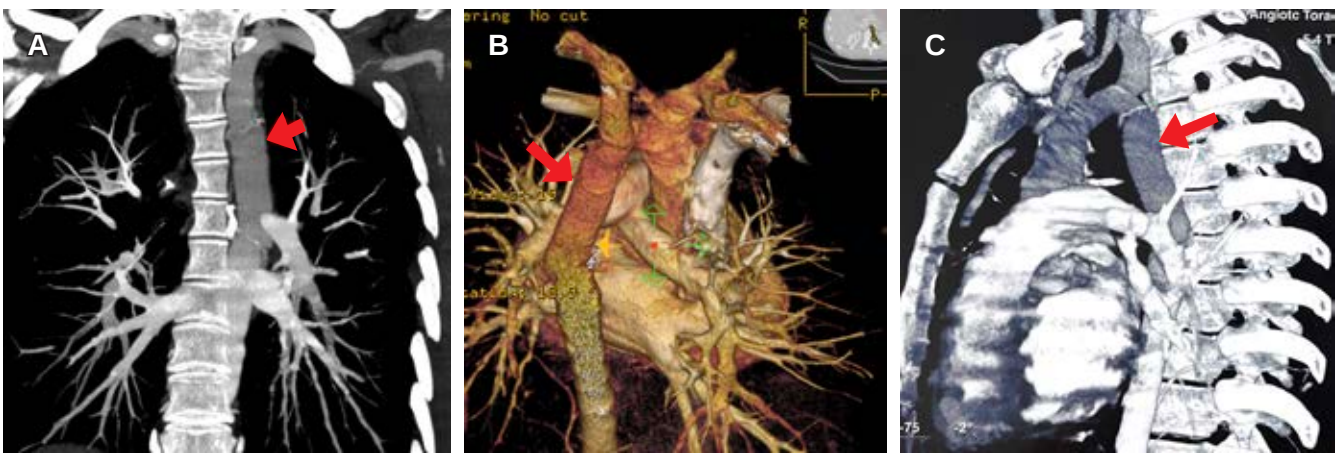

Figure 4: Angiotomography images with reconstruction of great vessels showing a graft at the level of the descending aorta. A) coronal plane. B) Reconstruction of great vessels posterior view. C) Sagittal plane. Red arrows indicate graft location.

previously approached by posterolateral thoracotomy. All repairs were performed without extracorporeal circulation.

Undoubtedly, advances in surgical techniques have allowed lower risk of morbidity and mortality, with favorable results. For this reason, surgical repair is the treatment of choice in adults with IAA.[9] The aim of surgical treatment in IAA is to repair the defect and repair some other lesion in a single stage. Extra-anatomic bypass, end-to-end anastomosis, graft interposition or percutaneous stent placement have been proposed as surgical resolutions.[10] End-terminal anastomosis is suggested in newborns, infants, older children and adults, while graft interposition is an excellent option in adult populations.[3]

In the case presented here, we decided to perform a section of the portion where the defect was located and subsequently a Dacron graft interposition was performed. The patient had no bleeding during the postoperative period. No additional interventions were required. At two months of follow-up the patient is stable. Systemic arterial hypertension has subsided. No additional hospitalization was required.

REFERENCES

1. Gordon EA, Person T, Kavarana M, Ikonomidis JS. Interrupted aortic arch in the adult. J Card Surg. 2011;26(4):405-409. doi: 10.1111/j.1540-8191.2011.01273.x.
2. Carlomagno G, Altiero M, Ferrara F, et al. Late diagnosis of interrupted aortic arch with massive collateral circulation in a former competitive athlete with early-onset hypertension. Circ Cardiovasc Imaging. 2021;14(2):e010818. doi: 10.1161/CIRCIMAGING.120.010818.
3. Dong SW, Di DD, Cheng GX. Isolated interrupted aortic arch in an adult: A case report. World J Clin Cases. 2021;9(4):992-998. doi: 10.12998/wjcc.v9.i4.992.
4. Franconeri A, Ballati F, Pin M, Carone L, Danesino GM, Valentini A. Interrupted aortic arch: a case report. Indian J Radiol Imaging. 2020;30(1):81-83. doi: 10.4103/ijri.IJRI_273_19.
5. Nasr VG, DiNardo JA. Interrupted aortic arch. In: The pediatric cardiac anesthesia handbook. Hoboken, NJ: Wiley Blackwell; 2017. pp. 187-190.
6. Patel DM, Maldjian PD, Lovoulos C. Interrupted aortic arch with post-interruption aneurysm and bicuspid aortic valve in an adult: a case report and literature review. Radiol Case Rep. 2015;10(3):5-8. doi: 10.1016/j.radcr.2015.06.001.
7. Silva J, Guiomar N, Silva MP, Caeiro D, Gama V. Interrupted aortic arch in an adult. Eur J Case Rep Intern Med. 2017;4(9):000692. doi: 10.12890/2017_000692.
8. Sai Krishna C, Bhan A, Sharma S, Kiran U, Venugopal P. Interruption of aortic arch in adults: surgical experience with extra-anatomic bypass. Tex Heart Inst J. 2005;32(2):147-150.
9. Firouzi A, Sadeghipour P, Pouraliakbar H, et al. Paradigm shift in the management of isolated interrupted aortic arch in adulthood. Curr Probl Cardiol. 2021;46(3):100717. doi: 10.1016/j.cpcardiol.2020.100717.
10. Patel DM, Maldjian PD, Lovoulos C. Interrupted aortic arch with post-interruption aneurysm and bicuspid aortic valve in an adult: a case report and literature review. Radiol Case Rep. 2015;10(3):5-8. doi: 10.1016/j.radcr.2015.06.001.

Funding: none.

Disclosure: the authors have no conflict of interest to disclose.