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A case never reported before: bilateral type A interruption in a double aortic arch without persistent ductus arteriosus

Un caso nunca antes reportado: interrupción bilateral de tipo A en un arco aórtico doble sin ductus arteriosus persistente

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Double aortic arch, interrupted aortic arch, persistent ductus arteriosus.

Palabras clave:
Doble arco aórtico, interrupción del arco aórtico, ductus arteriosus persistente.

ABSTRACT

We present the case of a four years seven-month-old patient referred to cardiologist for heart murmur. Echocardiography study revealed interruption of the aortic arch. Magnetic resonance imaging revealed type A bilateral interruption in double aortic arch, with obliterated ductus arteriosus, without septal defect, with collateral network made of two thick anastomotic arteries that join the subclavian arteries to the descending aorta. We haven't found this pathology reported in any medical literature. Asymptomatic patient received control as outpatient in Cardiology Department. Reexamined ten years later with multislice angiotomography, confirming the diagnosis.

RESUMEN

Paciente de cuatro años siete meses que es enviado a Cardiología, por encontrarse un soplo a la auscultación cardíaca. En el estudio ecocardiográfico se le diagnosticó interrupción del arco aórtico. Se estudia al paciente con Imágenes de resonancia magnética, encontrándose una interrupción bilateral Tipo A en un doble arco aórtico, con ductus arteriosus obliterado, sin defectos septales, con una red colateral formada por dos gruesas arterias anastomóticas que unen las arterias subclavias con la aorta descendente. El paciente asintomático es controlado en consulta externa de Cardiología. El paciente es reestudiado diez años más tarde con angiotomografía multicorte, confirmándose el diagnóstico.

INTRODUCTION

Interruption of the aortic arch is a rare congenital malformation, in which the luminal continuity between the proximal aorta and the descending aorta is lost.[1] The first description of this pathology was made by Steidele in 1778.[2] It is a rare pathology, representing less than 1.5% of all congenital heart diseases.[3]

Interruptions of the aortic arch are associated in 98% of cases with persistent ductus arteriosus, and in 90% of cases with interventricular communication,[4] less frequently with subaortic stenosis, transposition of large vessels, and with double outlet right ventricle.[5]

Celoria and Patton[6] classify interrupted aortic arches according to the site of interruption, in three types: type A, when the interruption is found distal from the origin of the left subclavian artery; type B, when it is located between the left carotid artery and the left subclavian artery; and type C, when it is located after the origin of the innominate artery. The most frequent type is B, then A.

Mollers and Edwards[5] refine this classification, creating subtypes according to the origin of the right subclavian artery.

- **Type I.** Interruption is located after the origin of left subclavian artery. Subtype a: right subclavian artery arises from innominate artery; and Subtype b: right subclavian artery arises from descending aorta, becoming an aberrant right subclavian artery.

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- **Type II.** Interruption after left carotid artery. Subtype a: right subclavian artery arises from innominate artery; Subtype b: right subclavian artery arises from descending aorta; and Subtype c: right subclavian artery arises from right pulmonary artery, becoming an isolated right subclavian artery.
- **Type III.** Interruption located after the origin to innominate artery.

When the interrupted aortic arch is accompanied by a persistent ductus arteriosus and interventricular communication, most of the arterial blood flows from the left ventricle to the right ventricle through the septal defect, and from the pulmonary artery to the descending aorta through the persistent ductus arteriosus. The birth of a child with this type of interruption constitutes a surgical emergency, because it is a ductal-dependent pathology. The physiological closure of the duct leads to heart failure, metabolic acidosis, and death. The therapeutic plan to save these children consists in treating the heart failure, infusing prostaglandin E1 to prevent the closure of the duct, and corrective surgery as soon as possible.[7,8]

Three percent of interruptions of the aortic arch[4] happen without persistent ductus arteriosus and without interventricular defect. The first case of this variant was reported by Pillsbury[9] in 1964, in a 16-year-old girl with type C aortic interruption. Afterwards, in 1967 Zetterqvist[10] reported the second case, and in 1970 Morgan[11] reported a third case. In a review of 105 cases of interruptions of the aortic arch, Moller[4] found only two cases with obliterated ductus arteriosus and without septal defect. Some authors have called this type of interruption «complete interrupted aortic arch»,[12,13] «adult-type interruption»,[12] «Solitary Interruption of the Aortic Arch»,[14] «isolated interrupted aortic arch».[15]

Patients with interruptions of the aortic arch without persistent ductus arteriosus and without interventricular communication make up a special group of patients with an entirely different behavior than patients with interruptions of the aortic arch with persistent ductus arteriosus. Dische[14] finds that these patients have a longer survival rate, are diagnosed later in life, the clinical course is more benign, there are fewer complications, and the surgical treatment is more successful.[4,15]

In 1948, Edwards[16] described two types of double aortic arch: type 1, with two arches, both the right and the left arches being totally patent; and type 2, where the right arch is patent, but the left has some degree of atresia or interruption. Shuford in 1971[17] proposed a classification of the interruptions of the left arch in double aortic arches. Type A, located after the ductus arteriosus; type B, after the left subclavian artery; type C, between the common left carotid artery and the left subclavian artery; and type D, before the left common carotid artery. The type C interruption of the left arch creates an aortic arch very similar to the circumflex right aortic arch,[18,19] also called right aortic arch with retroesophageal component and aberrant left subclavian artery, for practical purposes the angiographic features of both anomalies are similar.[18] We have not found in the literature descriptions of bilateral interruptions in double aortic arches.

PRESENTATION OF THE CASE

We present the case of a four years seven months old male, first child, without significant prenatal or family medical history, normal height-weight and psychomotor development. Referred to cardiology by pediatrician who detected heart murmur.

Physical examination: weight 15.6 kg (50^{th} percentile), height 104 cm (50^{th} percentile). The fascies was normal, and no palate anomalies were found. Blood pressure: right arm: 90/60, left arm: 90/60, lower limbs 85/60. Saturation in four limbs: 91%. Cardiac auscultation: rhythmic and normophonetic sounds, holosystolic murmur of intensity 2+/6 in tricuspid foci, without irradiation.

Electrocardiogram: right ventricular hypertrophy. Chest X-ray: heart of normal shape and size.

Echocardiography: prominent left ventricle, mild failure of tricuspid valve. Probable interruption of aortic arch.

Magnetic resonance angiography (MRA): *Situs solitus* with levocardia, atrial-ventricular and ventricular-arterial concordance, integrity of interatrial and interventricular septum, normal drainage of systemic and pulmonary veins.

The 3D volumetric reconstruction images of the contrast-enhanced MRA *(Figure 1 A-C)* reveal a double aortic arch with interruption of the two arches after the origin of the subclavian arteries, presence of two thick anastomotic tortuous arteries arising from posterior aspect of the subclavian arteries and traveling back and down to join the descending aorta, which presents a diverticular formation in its upper end.

MRA diagnosis: bilateral type A interruption in a double aortic arch (according to Celoria's classification), with obliterated arterial duct, without interventricular communication.

Asymptomatic patient undergoes periodical controls in Cardiology Department.

After 10 years, patient is re-evaluated, by then he is 14 years, 7 months old, weight 57 kg (p57), height 150 cm (p5), blood pressure is 110/60 in upper limbs and 90/60 in lower limbs, saturation 91%.

Electrocardiography: right ventricular hypertrophy.

Multi-detector computed tomography angiography (MDCTA) is conducted. The 3D volumetric reconstruction images *(Figure 2 A and B)* show with more detail the findings of the MRA performed 10 years before. The two collateral vessels have a mean diameter of 10 mm and present a short extra-thoracic pathway located behind the third costal arches *(Figure 2C)*.

The current echocardiogram shows visceral *situs solitus*, prominent right chambers with tricuspid morphology similar to Ebstein's anomaly with mild insufficiency.

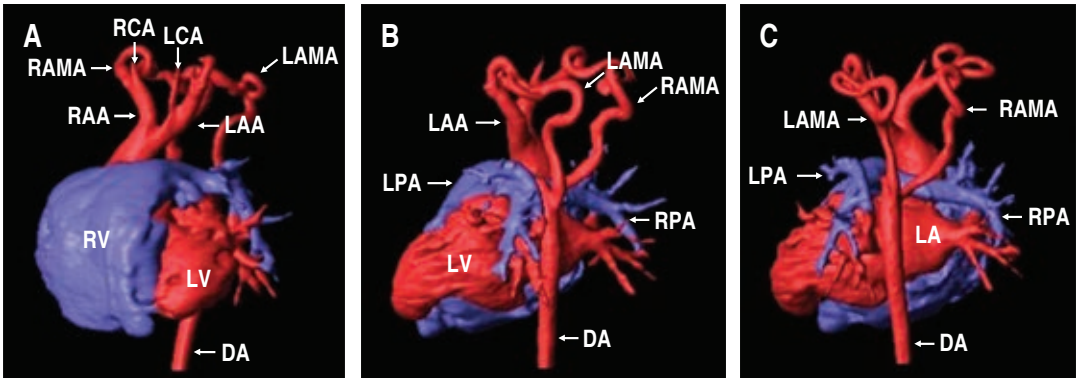
Patient is asymptomatic, has not undergone surgery.

DISCUSSION

We present the case of a patient that, at age 4, was diagnosed with type A bilateral interruption of double aortic arch, without persistent ductus arteriosus and without septal defect.

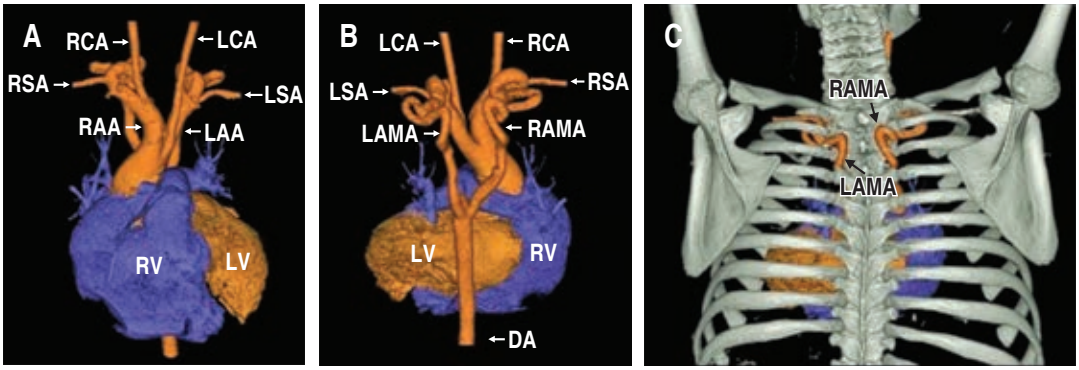
There are two classifications of interruptions of the aortic arch according to the site: Celoria and Patton[6] classify the interruptions in the single aortic arches and Shuford[17] classifies the interruptions of the left aortic arch in double aortic arches. Our patient has a double aortic arch but with bilateral interruption. We have arbitrarily chosen Celoria's classification to call it «type A bilateral interruption», which means that the interruption is found in both arches, after the origin of the subclavian arteries. We have found no reports of similar cases in the medical literature.

Depending on the presence of persistent ductus arteriosus, the interruptions of the aortic arches can also be classified in interruptions with persistent ductus arteriosus and interruptions without persistent ductus



DA = Descending aorta, LA = Left atrium, LAA = Left aortic arch, LAMA = Left anastomotic mediastinal artery, LPA = Left pulmonary artery, LV = Left ventricle, RAA = Right aortic arch, RAMA = Right anastomotic mediastinal artery, RCA = Right carotid artery, RPA = Right pulmonary artery, RV = Right ventricle.

Figure 1: Magnetic resonance angiography when patient was four years, seven months old. 3D volumetric reconstruction images. **A)** Anterior view; **B)** Left lateral view; **C)** Posterior view.

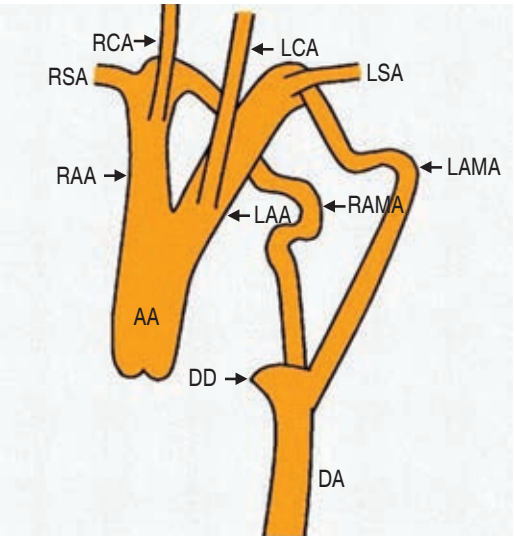


LAA = Left aortic arch, LAMA = Left anastomotic mediastinal artery, LCA = Left carotid artery, LSA = Left subclavian artery, RAA = Right aortic arch, RAMA = Right anastomotic mediastinal artery, RCA = Right carotid artery, RSA = Right subclavian artery.

Figure 2: Contrasted multidetector computerized tomography. 3D Volumetric Reconstruction Images. **A)** Anterior view; **B)** Posterior view; **C)** Posterior view of 3D volumetric reconstruction of heart, vessels and thoracic cage. Observe in **C** the extrathoracic pathway of RAMA and LAMA behind third costal arches.

arteriosus. Interruptions accompanied by persistent ductus arteriosus correspond to 97% of the cases they are diagnosed early during childhood, constitute a surgical emergency, because they are ductal-dependent, and when the ductus arteriosus closes the patients die. While, interruptions of the aortic arch that occur in not accompanied by persistent ductus arteriosus occur in less than 3%,[4] are diagnosed later, often in adults, are more benign and have fewer complications. The case we report belongs to the second group, the patient has obliterated ductus arteriosus, no septal defect, was diagnosed at age 4, is practically asymptomatic, at present he is 14 years old and in good health. He has the characteristics of what Dische[14] calls «solitary interrupted aortic arch».

It has been confirmed that when children are born with an interruption of the aortic arch with persistent ductus arteriosus, they lack a collateral network between the ascending and descending aortas; that is why when the ductus arteriosus closes a catastrophe ensues. Children whose ductus arteriosus is obliterated in utero have time to develop collateral vessels. The same happens with children born with extreme aortic coarctations. Children born with type B and C interruptions of aortic arches[10-13,20] with obliterated ductus arteriosus develop intracranial collateral networks between the arteries arising from the ascending aorta and those arising from the descending aorta. In children born with type A interruptions of the aortic arch with obliterated ductus arteriosus, where the interruption is located after the origin of the left subclavian artery, as in the case we report, all the supra-aortic arteries arise from the ascending aorta, none from the descending aorta, which means that the intracranial anastomotic networks cannot be created, the only possibility is the creation of thoracic and thoraco-abdominal collateral networks. Among the thoracic collateral networks, one of the most important is the one described by Kirks in extreme aortic coarctations[21] as anastomotic mediastinal arteries, formed by the anastamosis of the upper intercostal artery, a branch of the subclavian artery, and the third intercostal arteries, branches of the descending aorta, this is the type of anastomotic arteries our patient has *(Figure 3)*. In the images of the volumetric reconstruction of the MRA *(Figure 1 A-C)*, as well as in the MDCA *(Figure 2 A and B)*, we see two thick and tortuous anastomotic arteries, which connect the subclavian arteries to the descending aorta, with a short extra-thoracic pathway behind the third costal arches *(Figure 2C)*. These mediastinal anastomotic arteries have a mean diameter of 10 mm, capable of carrying an adequate blood flow to the descending aorta and the lower limbs. Blackford,[22] in a case of extreme aortic



AA = Ascending aorta, DA = Descending aorta, DD = Ductus diverticulum, LAA = Left aortic arch, LAMA = Left anastomotic mediastinal artery, LCA = Left carotid artery, LSA = Left subclavian artery, RAA = Right aortic arch, RAMA = Right anastomotic mediastinal artery, RCA = Right carotid artery, RSA = Right subclavian artery.

Figure 3: Schematic drawing of double aortic arch with bilateral interruption after origin of subclavian arteries, presence of two thick mediastinal anastomotic arteries that join subclavian arteries to descending aorta.

coarctation, describes an anastomotic artery similar to the one in our case, between the costocervical trunk and the descending aorta, which had a diameter of 14 mm.

In the upper end of the descending aorta *(Figure 3)* we see a saccular dilatation ending in a cul-de-sac and joining the pulmonary artery: this is a ductal diverticulum,[23] the remnants of the ductus arteriosus obliterated during fetal life.

Cases of interruption of the aortic arch, published in the medical literature, have been diagnosed traditionally using echocardiography and angiography with catheterization,[5,9,11,12,14,24,25] others with magnetic resonance imaging[15,26,27] and some with CTA.[10,20,28] In our hospital we prefer to study cardiopathies and vascular rings in children using magnetic resonance and in adults using multidetector computerized tomography. Our patient was studied with magnetic resonance when he was four years, seven months old, and with multidetector computerized tomography, ten years later. We have not found in the medical literature another case studied with the two imaging modalities.

CONCLUSION

This is an exceptional case of a bilateral interruption in a double aortic arch, with obliterated arterial ductus, causing the intrauterine development of two thick anastomotic arteries, which allowed the asymptomatic growth of the child. We haven't found any similar case reported in medical literature.

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