Anatomic repair of interrupted aortic arch in adult

TO THE EDITOR. Interrupted aortic arch (IAA) is a rare congenital malformation frequently associated with other intracardiac defects, such as ventricular septal defect, patent ductus arteriosus and bicuspid aortic valve.[1]

When not surgically treated, 75% of patients die within the first month of life and 90% within the first years. The development of a collateral aortic circulation during gestation is the cause of perinatal survival and the diagnosis in adulthood.[1]

The worldwide literature reports less than 20 cases of interrupted aortic arch surgically treated in adults, testifying the rarity of this condition.[2]

In patients with IAA, symptoms usually occur in the neonatal period and clinical deterioration is often rapid.[1]

This letter reports the case of an asymptomatic patient who had IAA and was treated successfully with surgical repair.

A 21-year-old asymptomatic woman was referred to our hospital for evaluation of hypertension that was incidentally diagnosed two months before.

On physical examination, peripheral pulses were palpable over the carotid arteries and in the upper limbs. The blood pressure was 160/90 mmHg in the right arm and 170/95 mmHg in the left arm. However, there was a significant difference in blood pressure between upper and lower limbs. Bilateral femoral and popliteal pulses were extremely weak and cardiac auscultation revealed a regular tachycardia with grade 3 systolic murmur on the left second intercostal area.

The electrocardiogram showed left ventricular hypertrophy and regular rhythm at 105 bpm. Transthoracic echocardiography demonstrated concentric left ventricular hypertrophy with good left ventricular function and at suprasternal view aortic arch appeared to terminate or narrow after the left subclavian artery.

Cardiac catheterization was performed *via* the right brachial artery, which showed an interrupted arch distal to the left subclavian artery (Figure 1), and through transfemoral approach that showed the absence of the passage of catheter (Figure 2).

The patient underwent surgery through the fourth intercostal space with a left posterolateral thoracotomy. Isolation of the aorta was carefully performed to avoid excessive hemorrhage from the numerous collateral vessels that were isolated and cauterized or tied. Because of the development of collateral circulation, the clamp on the descending aorta did not produce significant decrease of the pressure distal to it and left-heart bypass was not used.

The aortic continuity was reconstructed by the interposition of a tubular prosthesis (Gelweave woven vascular prosthesis 18 mm) (Figure 3). The aortic cross-clamp time was 23.—minutes.

The postoperative course was regular, and no hemodynamic, neurologic, respiratory, or bleeding complications occurred. Postoperative mechanical ventilation time was three hours. Total blood loss during the first 24 hours was 300 mL. The postoperative echocardiogram showed a good surgical result and the absence of aortic obstruction. The hospital stay was four days.

IAA is a rare congenital malformation defined as a complete absence of flow between two portions of the aorta. Aortic coarctation instead is characterized by a narrowing of the aortic lumen with a flow of blood between the two ends. IAA has been classified by Celoria and Patton [1] according to the level of interruption which may be distal to the left subclavian artery (type A), between the left common carotid and left subclavian arteries (type B) and between the innominate and left common carotid arteries (type C). The most common type is B (53%), followed by A (43%) and C (4%).

Several methods can be used for the diagnosis of IAA. Two-dimensional echocardiography plays an important role in the delineation of IAA and for ruling out associated intracardiac anomalies. Cardiac catheterization is often necessary for definitive anatomical evaluation in adult patients with IAA. However, it may be difficult to perform in patients without a prior knowledge of vascular anatomy to ensure visualization of both proximal and distal segments. For this reason, some authors suggest other diagnostic techniques like MRI and CT.[2]

In our pediatric experience we usually recourse to cardiac catheterization only in some cases. In this specific case, however, we have applied the data collected from the literature for this unusual situation in an adult patient and we decided to practice cardiac catheterization.

The aortography demonstrated IAA, the site of interruption, and branching pattern of great arteries. However collateral vessels could not be defined completely since a large

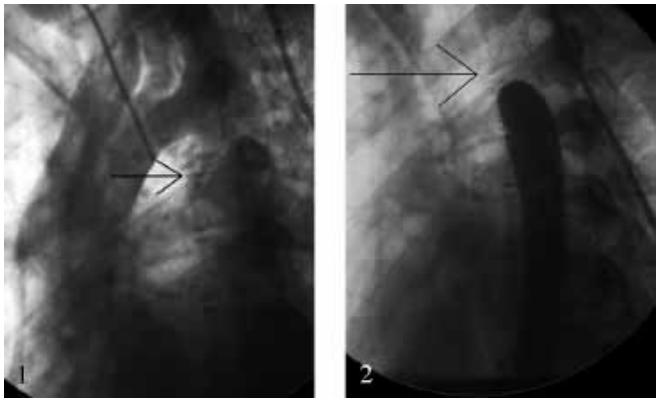

Figure 1.—Aortagraphy of interrupted aortic arch type A via the right brachial artery.
Figure 2.—Aortography through transfemoral approach.

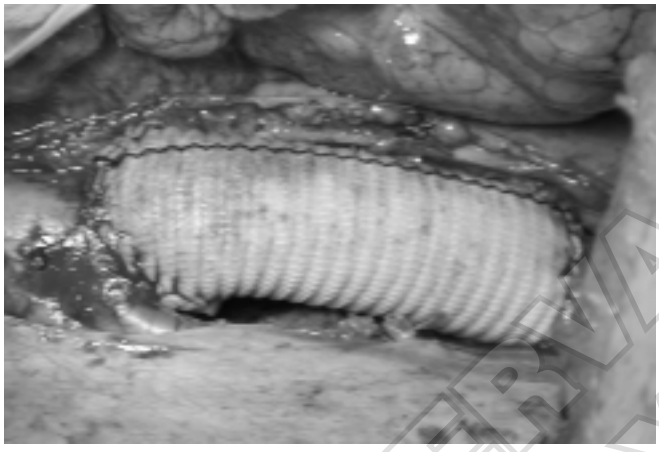

Figure 3.—Interposition of a tubular prosthesis.

volume of contrast material and prolonged invasive procedure time were required. Echocardiography showed no intracardiac anomalies, as well as other cases described in the literature,[1, 2] but only the presence of a bicuspid aortic valve.

In adults IAA is very rare. Some authors suggest that the extra-anatomic approach is the preferred intervention to correct an interrupted aortic arch.[2, 3] It consists of a ventral aortic repair through a midline sternotomy extended into an upper midline laparotomy and the anastomosis by a graft from the ascending aorta to the supraceliac abdominal aorta. This approach is used because of the risk of uncontrolled bleeding from the collateral aortic circulation with a thoracic surgical approach. Others suggest an anatomic approach through a thoracotomy.[4]

The risk of paraplegia after aortic coarctation repair in infants is estimated to be about 0.4%, but it increases to 3% in older children and adults.[5] Furthermore, there is no consensus regarding the optimal strategy for spinal cord protection in this type of surgery.

In our case we adopted the same technique used in neonatal and pediatric age. Having been able to isolate completely the section of aorta interrupted by multiple collateral arteries, was carried out a reconstruction of discontinuity. In adult the presence of collateral arteries developed reduces the risk of distal ipoperfusion induced by aortic clamping, but to prevent complications is well that the clamping is as short as possible.

We conclude that in this type of isolated IAA radical correction that we have practiced offers an excellent surgical approach for this rare condition in adults.

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