Congenital native interruption of aortic arch in an adult: Extra-anatomic approach by right-side thoracotomy

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We outline the case of an 18-year-old male patient with a congenital nonoperated interruption of the aortic arch. A right thoracotomy without cardiopulmonary bypass facilitated repair through an extra-anatomic bypass between the ascending and the supradiaphragmatic descending aorta. Results for the immediate and 2-year radiologic and clinical check-up were satisfactory. The most common complications in anatomic correction are stroke under selective cerebral perfusion, risk of paraplegia, and hemorrhage. We present a new technique for repair of interruption of the aortic arch in adults that avoids the need for extended dissection of the aorta and a partial occlusion clamp during anastomosis and allows for cerebral and medullar perfusion. (J Vasc Surg 2010;51:1525-7.)

The interruption of aortic arch (IAA) is a rare congenital malformation representing a neonatal surgical emergency. If left uncorrected, this defect is generally fatal within days. Its medical and surgical handling is currently well codified.[1] During the neonatal period, the complete correction must be done by sternotomy under extracorporeal circulation (ECC) and selective cerebral perfusion, with correction of the aorta by direct anastomosis or patch. Extremely rarely, these patients reach adulthood with well-developed arterial collaterals that allow an acceptable perfusion of the aorta after the interruption. These patients pose operating strategy problems, and surgical correction through sternotomy with ECC involves high risks of morbidity and mortality.[2] The most common complications are neurologic risk under a selective cerebral perfusion, risk of paraplegia by medullar ischemia, left recurrent nerve lesion, and hemorrhagic risk caused by injury of the collateral arteries.

We outline a case of an 18-year-old male patient with a congenital interruption of the aortic arch who benefited from a surgical approach through a right-side thoracotomy, without ECC. We believe this is the first description of this technique for IAA in adult.

CASE REPORT

The patient was diagnosed with a type A IAA (Table),[3] without ventricular septal defect or patent ductus arteriosus, at age 9 months during a routine medical visit. This patient, however, did not come back for consultation for 18 years because he presented no significant symptoms. The patient was referred to our department after loss of consciousness under exertion. Arterial pressure of the upper limbs while at rest was 140/100 mm Hg, the femoral pulse was barely perceptible, and bilateral intercostal pulses were noted. The blood pressure of the lower limbs was 120/80 mm Hg.

Magnetic resonance angiography (MRA) confirmed the diagnosis of type A IAA and showed a substantial system of collateral connections between the subclavian and intercostal arteries (Fig 1, *A*). Anatomic correction was ruled out because the extent of the collateral vessels rendered it too hazardous to attempt the extended dissection of the aorta required for its mobilization. The indication for an extra-anatomic bypass between the ascending aorta and the lower thoracic descending aorta, without ECC, was adopted.

Surgery was performed by means of a right posterior thoracotomy through the fifth intercostal gap. No rib resection was necessary. After resection of the left triangular diaphragmatic ligament, opening of the parietal pleura and anterior deviation of the esophagus, the supradiaphragmatic descending aorta was dissected over 25 mm (Fig 2).

A partial upper vertical pericardiotomy before the right phrenic nerve enabled us to approach the ascending aorta. An extra-anatomic bypass using a vascular prosthesis of 18-mm long piece of Hemashield (Boston Scientific, Natick, Mass) was inserted over the supradiaphragmatic descending aorta, clamped laterally; the tube was passed behind the esophagus in order to not compress this latter with a large bend at the thoracic right apex and the terminolateral anastomosis between the tube and the ascendant aorta by lateral clamping. The measure of the length of the prosthesis was made by passing it into the right costovertebral groove and then into the anterior mediastinum in front of the right pulmonary apex before the second anastomosis. The prosthesis was finally unclamped after carefully expelling bubbles and pasting both anastomoses with biological glue. The anastomoses were performed with continuous suture of 3-0 polypropylene. No collateral was affected.

The patient's immediate postoperative progress was satisfactory. He was extubated 6 hours after surgery and left the intensive

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Table. Interrupted aortic arch classified by Celoria and Patton[3]

Classification	*Interruption of the aortic arch between:*
 Type A	the left subclavian artery and the descending aorta at the isthmus
 Type B	the left common carotid and left subclavian arteries
 Type C	the innominate and left common carotid arteries

care department after 24 hours. The postoperative MRA showed good patency of the anastomoses. The result of the 2-year radiographic and clinical check-up was satisfactory (Fig 1, *B*).

DISCUSSION

Surgical correction of IAA in adults is problematic. The substantial collateral bypasses that have developed render any extended dissection of the isthmic aorta extremely difficult and any attempt at mobilizing the aorta highly risky.

Anatomic correction of stenosis of the arch in children has the virtue of facilitating normal growth of the aorta, and prosthetic fabrics can cause serious vascular distortion during growth of the aorta.[4] Anatomic repair also prevents complications in the long run that might affect left ventricle function as well as the development of false aneurysms at the distal anastomosis. Indeed, the elastic properties of the aorta wall prevent peaks of pressure at each systole.

In adults, however, techniques of extra-anatomic bypass have produced good medium-term results[5] and represent a good alternative to conventional surgery. The advantages include being relatively simple procedures that have a low risk of morbidity and death. This case was unique because the patient was an adult with a complete IAA who had not undergone any previous aortic surgery. Most reports are of patients who had aortic surgery in childhood and have been reoperated on with this technique for residual lesions.[6] Also, this case is important because the vascular surgeon may see such patients who are not under the care of any cardiac surgical service, unlike most patients, and can treat them, especially if the technique does not need ECC.

Without any associated intracardiac lesion, the right-side thoracotomy has several advantages, including good exposure of the ascending aorta and the supradiaphragmatic descending aorta, no handling of the heart, avoiding the recourse to ECC, low partial clamping of the descending aorta prevents medullary ischemia, and finally, thoracotomy prevents pericardiac symphysis and allows simple subsequent heart surgery. In fact, because it involves an

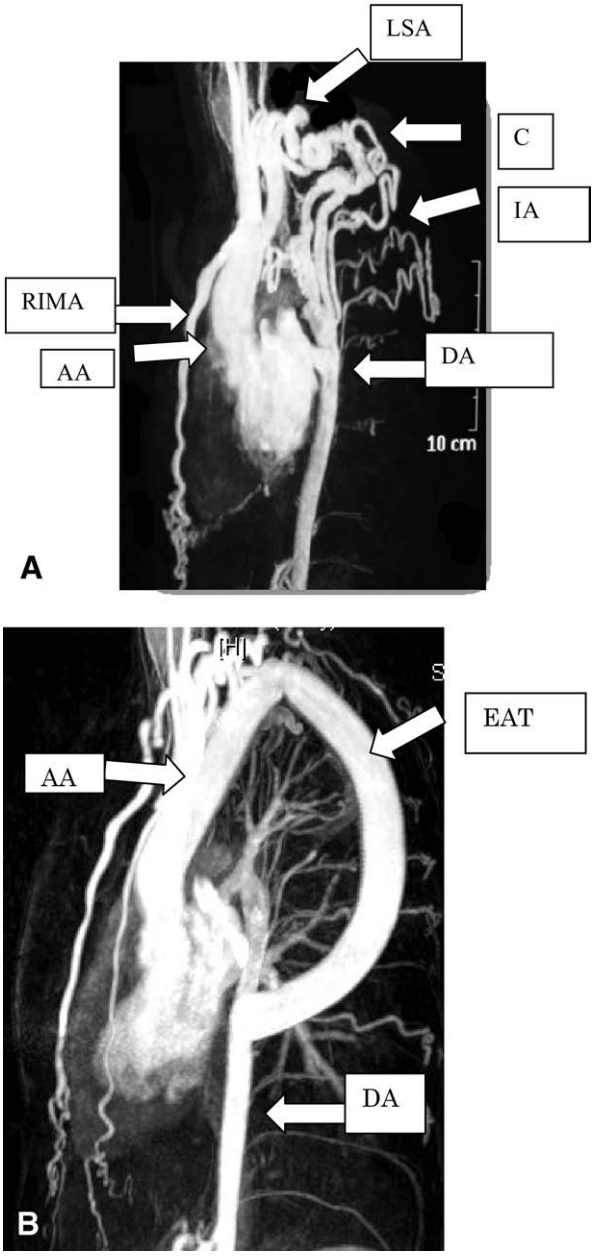

Fig 1. A, Preoperative magnetic resonance angiography (MRA) profile shows the type A aortic arch interruption and the collateral bypass originating from the left subclavian artery *(LSA)*. **B,** Postoperative MRA profile shows the permeability of the extra-anatomic tube *(EAT)*. *AA,* Ascending aorta; *C,* collaterals; *DA,* descending aorta; *I,* intercostals; *RIMA,* right internal mammary artery.

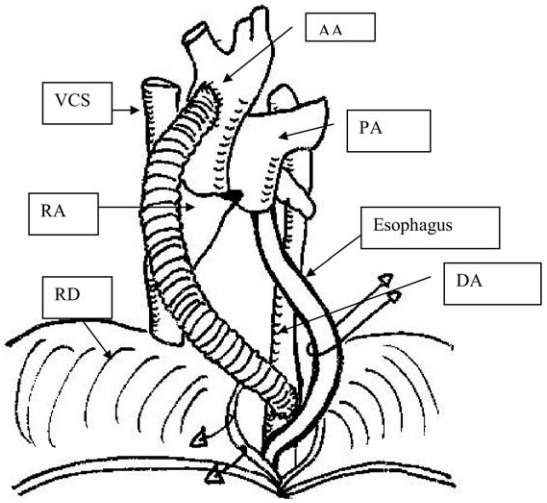

Fig 2. Scheme after the extra-anatomic tube (EAT) bypass shows that the thoracic descending aorta *(DA)* is behind the esophagus, which has been mobilized to the anterior side, and both anastomoses have been done after partial clamping of the aorta. *AA,* Ascending aorta; *PA,* pulmonary artery; *RA,* right atrium; *RD,* right diaphragm; *VCS,* vena cava superior.

aortic left arch like in any patient with normal anatomy, it will be logical to perform a left thoracotomy; nevertheless, the existence of multiple collaterals and the difficulty to access the ascending aorta makes the right approach more judicious.

The unique difficulty is accessing the descending aorta; that is why we must free the esophagus and enclose it in order to recline it; therefore, we have an excellent approach of the descending aorta, which must only be liberated by 3 cm. Lateral clamping for each anastomosis maintains an arterial flow and avoids the problems of cerebral ischemia and medullar ischemia. For these reasons, a preoperative MRI was sufficient to analyze the collaterals and decide about the side of the bridging; arteriography was not necessary. No cerebral monitoring was necessary.

On the other hand, the collaterals provide downstream vascularization, which has been constituted gradually. With sternotomy, it is more difficult to liberate the descending aorta and leads to the need to push the heart backwards, which may be destabilized. Only concomitant heart lesions justify sternotomy, making it possible to perform the extra-anatomic bypass and cardiac repair in a single operation.[7]

This technique could also be of considerable utility in correcting aortic recoarctations and avoiding iterative left-sided thoracotomy, which increases the exposure to neurologic lesions,[8] such as of the left recurrent nerve, and the risk of medullar ischemia. In aortic recoarctations, the use of balloon angioplasty or stents, or both, as an alternative to open surgery has had favorable immediate and midterm results.[9]

CONCLUSIONS

The right thoracotomy presented in this technique for repair of IAA in an adult avoids the need for extended dissection of the aorta and partial occlusion clamp during anastomosis. It allows cerebral and medullar perfusion and thereby incurs a lower operating risk.

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