

Type B Interrupted Aorta in an Adult Patient

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ABSTRACT

Introduction: Interrupted aortic arch is a rare congenital malformation characterized by a complete loss of luminal continuity between the ascending and descending aorta. It is often diagnosed during the neonatal period.

Case presentation: We presented a 51-year-old male patient with interrupted aortic arch type B who was treated successfully with posterolateral thoracotomy without using cardiopulmonary bypass.

Conclusion: The prognosis for interrupted aortic arch depends on the associated congenital anomalies, but the outcome is usually very poor unless there is surgical treatment. Survival into adulthood depends on the development of collateral circulation.

CASE PRESENTATION

A 51-year-old male patient attended to our hospital, complaining of early fatigue with minimal effort. He also had a history of hypertension, which had been treated β -blocker. On his physical examination, the peripheral pulses were normally palpable in the upper extremities, but they were reduced in the lower limbs. Electrocardiography showed sinus rhythm with mild left ventricular hypertrophy. Chest radiography showed an increased cardiothoracic index. Laboratory results were within normal ranges. The patient underwent preoperative evaluation by 2-dimensional echocardiography, which showed normal ejection fraction (EF%60) with minimally mitral and aortic insufficiency. Aortic interruption has shown 0.5 cm before the subclavian artery, which has no continuity to the descending aorta.

An angiography was performed via the femoral approach for coronary artery imaging. Since 0.38-inch guide wire could not be inserted through the descending aorta region distal to the left carotid artery, we made aortography and diagnosed interrupted aorta. His coronary arteries were normal. Also cardiac MRI showed type B IAA (Figure).

SURGICAL TECHNIQUE

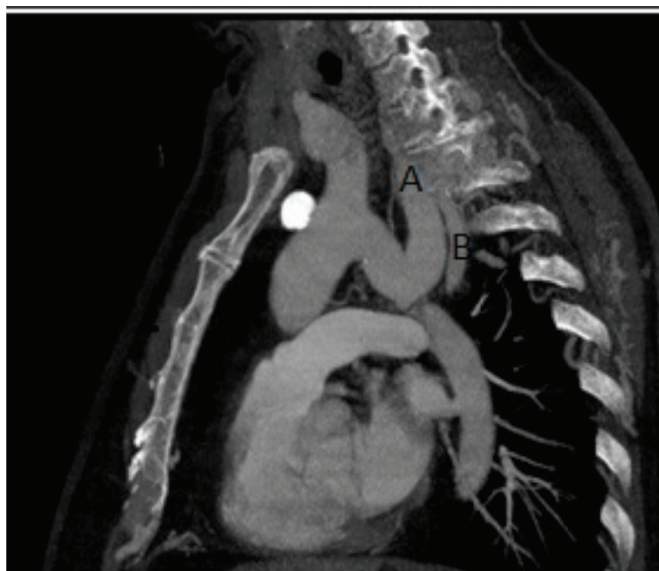
Elective surgery was planned for the patient, and a single-stage operation was performed via a left posterolateral thoracotomy. The left carotid artery, arcus aorta, and descending aorta were exposed. We put a side clamp on the proximal part of the interrupted segment of the arcus. A bypass was performed between the distal part of the left carotid artery and descending aorta using an 18-mm Dacron tubular graft (Vascutek, Gelweave, Terumo Cardiovascular Systems, Inchinnan, Renfrewshire, Scotland). The proximal and distal anastomoses were done with 4-0 polypropylene sutures using an end-to-side technique. The repair was performed without cardiopulmonary bypass. The patient was monitored for hemodynamic parameters in the intensive care unit for the first 24 hours following surgery; the period was uneventful. The patient had no renal, neurologic, or gastrointestinal complications. The hypertension was controlled with oral administration of 50 mg of Metoprolol and 5 mg of Ramipril. The patient was discharged from the hospital without any complications on postoperative day 5.

DISCUSSION

Interruption of the aortic arch (IAA) is an extremely rare congenital malformation that occurs in 3/1,000,000 live births and accounts for 1% of all congenital heart disease [Messner 2002]. It was described for the first time in 1778 [Steidele 1778] and Celoria and Patton defined the first classification system of IAA [Celoria 1959], which is still popular today. According to this classification system, the interruption is identified as Type A when the site of the aortic arch discontinuity is distal to the left subclavian artery; Type B when the interruption site is between the left subclavian artery and the left carotid artery (in our case); and Type C when this segment is between the left carotid artery and the innominate artery. Type B is the

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Patient's cardiac MRI showing type B IAA

most common (53%), followed by Type A (43%), and Type C (4%). Several methods can be used for the diagnosis of IAA. Although it has some limitations, echocardiography is the procedure of choice for the initial diagnosis of IAA in almost all cases [Akdemir 2004]. Cardiac catheterization is the most widely used technique for the definitive diagnosis; however, it may be difficult to perform this in patients without a prior knowledge of their vascular anatomy to ensure visualization of both the proximal and distal segments [Yildirim 2008]. Also, MR angiography or CT angiography can be used for confirmation. It presents as severe congestive heart failure in the neonatal period, and 90% of affected neonates die at a median age of 4 days. In the few cases reported in adults, the presentation varies from asymptomatic status to differential blood pressure recordings in the extremities and systemic arterial hypertension with its attendant complications. Survival into adulthood is dependent upon the development of substantial collateral circulation [Collins-Nakai 1976]. These collateral vessels are subject to atrophy, atherosclerosis, and even spontaneous rupture, resulting in secondary complications [Prasad 1988]. Of the patients with interruption [Messner 2002; Prasad 1988; Kauff 1973; Milo 1982; Todoric 1985; Burton 1995; Ogino 1998], 10 had undergone successful surgical repair, and most procedures were done in a single stage by means of an extra-anatomic approach. Extra-anatomic approach performed via sternotomy with cardiopulmonary bypass [Messner 2002]. Type A IAA can be repaired with the same technique of aortic coarctation because of its anatomical localization [Braunlin 1983]. In infants after the resection of

the narrowed segment, end-to-end anastomosis is performed. However, in adults with Type A IAA, it is difficult to perform end-to-end anastomosis. In most cases, graft interposition is the first choice of surgical treatment. But in literature for Type B patients, this method isn't described. It has been done with sternotomy and cardiopulmonary bypass (CPB) successfully for many cases [Erkanli 2012].

According to our experience, the surgical repair of Type B IAA in adults is feasible and safe through a posterolateral thoracotomy incision without using CPB for anatomically suitable patients.

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