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Surgical Repair for Isolated Aortic Interruption in a Young Adult

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ABSTRACT

Interrupted aortic arch (IAA), a rare congenital malformation of the aortic arch, is defined as a loss of luminal continuity between the ascending and descending portions of the aorta. It is rarely diagnosed as an isolated anomaly in adulthood. Surgical repair is feasible through a sternotomy or thoracotomy incision. In this report, we describe the surgical repair of an isolated IAA in a 29-year-old patient by performing an ascending-to-descending aortic bypass via a sternotomy with cardiopulmonary bypass.

INTRODUCTION

Interrupted aortic arch (IAA) is a congenital malformation that occurs with an incidence of 3 per million live births [Collins-Nakai 1976]. It is defined as a loss of luminal continuity between the ascending and descending portions of the aorta. If there is no cardiac abnormality, collateral vessels between the proximal and distal aorta develop as the patient grows up, precluding early diagnosis in childhood. In adulthood, IAA entails a very poor prognosis without surgical treatment, owing to the progressive hypertension [Messner 2002; Sai Krishna 2005; Gordon 2011]. We review surgical strategies for such a rare pathology of the aortic arch.

CASE REPORT

A 29-year-old woman presented with progressively worsening headache. There was no previous diagnosis of a musculoskeletal disorder and no signs of infection, emotional stress, alcohol or tobacco use, changes in sleep patterns, medication use, or depression in the differential diagnosis of headache. On admission, the patient's blood pressure was 200/100 mm Hg in the right arm and 80/58 mm Hg in the left arm. The radial artery pulse was stronger in the right arm than in the left arm, and the pulses of the lower extremities were not palpable. Cardiac auscultation revealed a harsh systolic murmur over the right upper parasternal area. A transthoracic echocardiography evaluation revealed left ventricular diastolic dysfunction and mild concentric hypertrophy of the left ventricle. Subsequent imaging and angiography evaluations by computed tomography showed a type B interruption of the thoracic aorta between the left carotid artery and subclavian arteries (Figure 1). The examination also revealed dilatation of the brachiocephalic and right common carotid arteries.

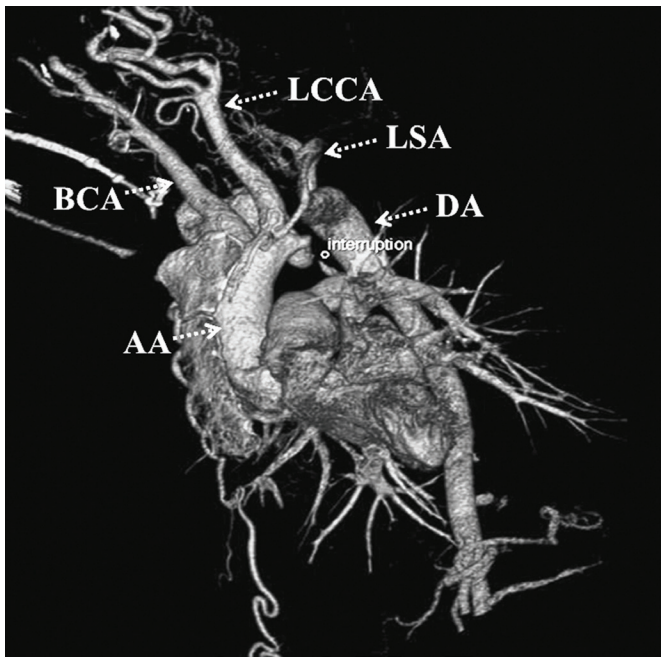

Figure 1. Preoperative 3-dimensional reconstructed computed tomography image shows complete interruption of the aortic arch (white dot) between the origins of the left common carotid and left subclavian arteries; also shown are the extensive collateral vessels. Note the presence of extensive and dilated collateral arteries that supply the descending aorta. LCCA indicates left common carotid artery; LSA, left subclavian artery; BCA, brachiocephalic artery; DA, descending aorta; AA, ascending aorta.

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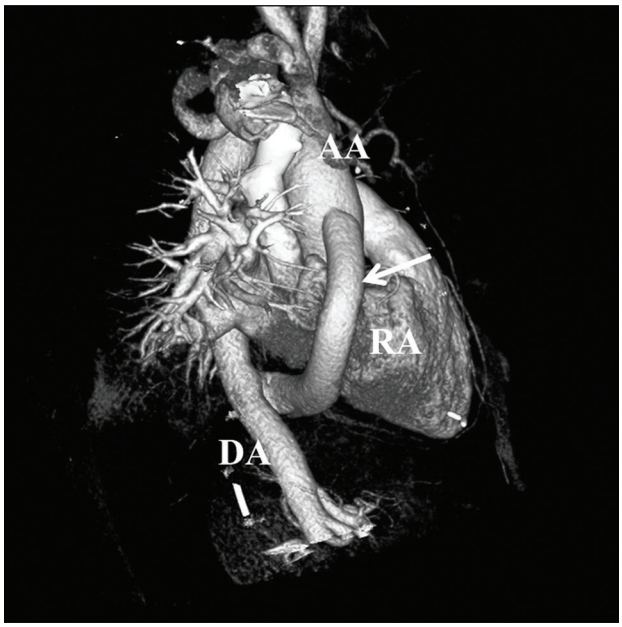

Figure 2. Postoperative 3-dimensional reconstructed computed tomography image shows extra-anatomic ascending-to-descending aortic bypass graft (white arrow) that encircles the right atrium. AA indicates ascending aorta; RA, right atrium; DA, descending aorta.

The descending thoracic aorta was supplied by extensive collateral vessels from the vertebrobasilar system, intercostal arteries, and the right internal thoracic artery, down to the posterior chest wall and the spine.

A single-stage operation was performed through a median sternotomy incision instead of a left thoracotomy incision. After systemic heparinization, the ascending aorta was cannulated before the origin of the brachiocephalic artery, and a 2-stage venous cannula was inserted into the right atrial appendage. Cardiopulmonary bypass was then initiated. Cardiac arrest was established with moderate hypothermia and antegrade blood cardioplegia through the aortic root. The apex of heart was positioned toward the left shoulder to expose the posterior pericardium. A vertical incision was made in the posterior pericardium, and the descending aorta was exposed to allow distal anastomosis of the graft. A side clamp was placed on the descending aorta, and a 14-mm Dacron graft (Hemashield; Maquet Medical Systems, Wayne, NJ, USA) was anastomosed end to side to the descending thoracic aorta with running 4-0 polypropylene suture. The graft was passed around the right atrium to reach the right aspect of the ascending aorta. After side-clamping of the ascending aorta, the proximal portion of the 14-mm graft was then anastomosed to the ascending aorta during cardiac arrest, in end-to-side fashion with running 4-0 polypropylene suture. There were no perioperative complications or postoperative bleeding. Cardiopulmonary bypass and aortic-clamping times were 120 minutes and 82 minutes, respectively. After the operation, the pressure difference between the 2 arms decreased from 120 mm Hg to 15 mm Hg. The patient was discharged from the hospital uneventfully on postoperative day 10.

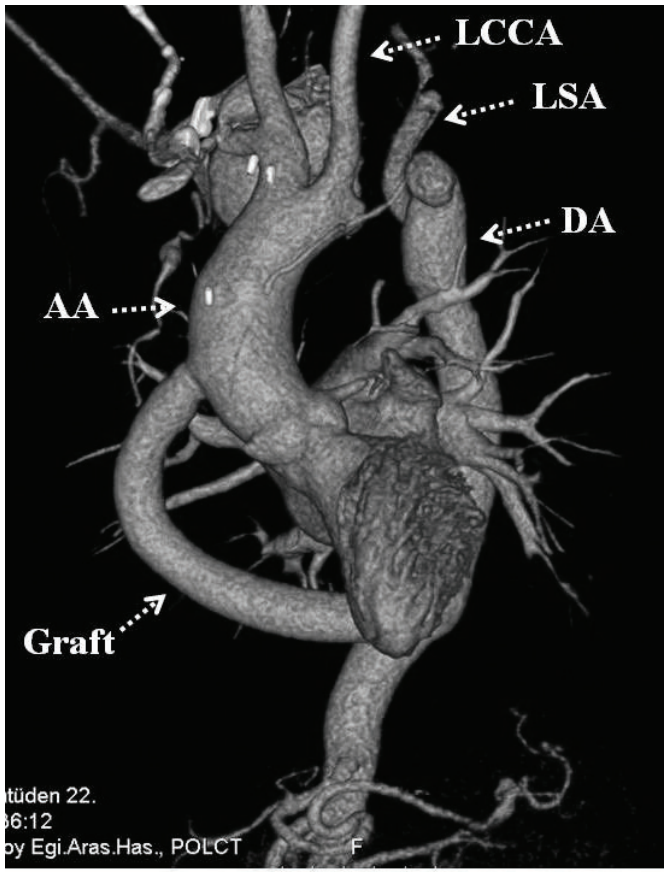

Figure 3. Postoperative 3-dimensional reconstructed computed tomography image shows the interruption of the aortic arch and ascending-to-descending aortic bypass graft. LCCA indicates left common carotid artery; LSA, left subclavian artery; DA, descending aorta; AA, ascending aorta.

At the 6-month follow-up after the surgery, a 3-dimensional computed tomography angiography examination revealed a patent graft and good distal flow with palpable peripheral pulses (Figures 2 and 3).

DISCUSSION

IAA is a rare congenital malformation defined as a complete absence of antegrade blood flow between 2 portions of the aorta [Collins-Nakai 1976; Messner 2002; Sai Krishna 2005; Gordon 2011]. In 1959, Celoria and Patton proposed a classification system that is still used [Celoria 1959]. This classification system divides this clinical entity into 3 types, depending on the site of the discontinuity. In type A, the interrupted-aorta discontinuity occurs distal to the left subclavian artery. A type B aortic interruption (the most common discontinuity) occurs between the left carotid and subclavian arteries. The rarest form is type C. In this type, the discontinuity occurs between the brachiocephalic and left carotid arteries.

IAA generally presents in childhood. IAA is extremely rare in adult patients. The small number of cases that have been reported involved an isolated congenital malformation

or occurred in combination with other defects in the same patient [Messner 2002; Sai Krishna 2005; Gordon 2011]. Survival into adulthood depends on the development of a substantial collateral circulation. These collateral vessels are subject to atrophy, atherosclerosis, and even spontaneous rupture, thereby producing secondary complications. In adult patients, the clinical presentation varies from an asymptomatic status to different blood pressures in the extremities and systemic arterial hypertension with its associated complications.

In the treatment of isolated IAA, operations can be performed through a sternotomy or thoracotomy incision [Gordon 2011]. A thoracotomy incision allows adequate exposure of the left subclavian artery as an inflow for a distal aortic bypass in a type A interruption. In a type B or type C interruption, however, surgical repair can be performed via a sternotomy incision with an extra-anatomic posterior pericardial approach [Sai Krishna 2005] or via an anatomic bypass with an end-to-end anastomosis [Daebritz 1999; Korkmaz 2011]. In the posterior pericardial ascending-to-descending aortic bypass procedure, the posterior pericardium is opened, and the thoracic aorta is dissected and looped. The distal anastomosis is performed first. The graft is then carried anterior to the inferior vena cava along the lateral border of the right atrium. The graft is anastomosed proximally onto the right lateral aspect of the ascending aorta. Cardiopulmonary bypass and cardioplegic arrest are used to retract the heart cephalad during exploration of the retropericardial portion of the descending aorta. This approach gives an excellent exposure of the thoracic descending aorta during the distal anastomosis of the bypass graft. It may also allow repair of coexisting cardiac and/or ascending aorta pathologies. Nevertheless, a sternotomy approach can be beneficial to avoid extensive surgical dissection that might cause perioperative hemorrhage from dilated collateral arteries; injury to the phrenic, recurrent, or vagus nerves; or injury to the left lung.

Although most procedures are feasible by means of an extra-anatomic posterior pericardial approach, anatomic ascending-to-descending aortic bypass via a thoracotomy can be an alternative strategy, without cardiopulmonary bypass [Daebritz 1999; McKellar 2007]. Prolonged cardiopulmonary bypass and aortic clamping can be avoided to reduce such neurologic events as spinal ischemia or cerebrovascular accidents. Side-clamping of the descending aorta allows continuation of blood flow to the intercostal arteries; however, injury to collateral blood vessels, lung parenchyma, recurrent laryngeal or phrenic nerves, the esophagus, lymphatic channels, and the spinal cord may develop.

In conclusion, ascending-to-descending bypass via a sternotomy incision and the use of cardiopulmonary bypass is feasible, and this approach can be the preferred choice of treatment because of its technical ease and likelihood of fewer complications.

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