The Heart Surgery Forum #2010-1100
14 (3), 2011 [Epub June 2011]
doi: 10.1532/HSF98.20101100

Online address: http://cardenjennings.metapress.com

Interrupted Aortic Arch or Extreme Coarctation? A Case Report and Review of the Literature

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ABSTRACT

The hallmark feature of aortic interruption that is useful in differentiating it from aortic coarctation is the "complete absence" of continuity between both parts of the interrupted segment. In this study, we reviewed the 28 patients diagnosed with isolated interrupted aortic arch (IAA) who reached adult age (> 20 years), aimed to review the validity of the Celoria-Patton classification in the literature, and reported the first microscopic pathology of the IAA in an adult.

INTRODUCTION

Interrupted aortic arch (IAA) was initially described in 1778 by Steidele and defined as the loss of continuity between the ascending and descending portions of the aorta in which the distal aortic circulation is supplied by a patent ductus arteriosus [Steidele 1778; Van Praagh 1971; Gokcebay 1972; Dische 1975; Reardon 1984]. Based on the absent segment of the aortic arch, Celoria and Patton described a classification system in 1959: type A, interruption distal to the left subclavian artery; type B, interruption between the left carotid and left subclavian arteries; and type C, interruption between the innominate and left carotid arteries [Celoria 1959]. IAA occurs in association with a wide variety of intracardiac malformations.

The disease is one of the rarest anomalies (3 instances per 1 million live births) among the congenital cardiac defects and one of the most rapidly deteriorating congenital heart diseases requiring emergent intervention; otherwise, death is inevitable within the first week of life. Without surgical intervention, 76% of the patients die in the newborn period, and 90% in the first year of life because of heart and renal failure or intracranial hemorrhage [Celoria 1959; Van Praagh 1971; Gockebay 1972].

On the other hand, a group of adult patients with IAA—concomitant with patent ductus arteriosus (PDA) and/or other congenital anomalies—who usually stay asymptomatic is described in the literature. All these patients were classified according to the Celoria-Patton classification. In the present study, we reported the first macroscopic pathology of an IAA in an adult and aim to review the validity of the Celoria-Patton classification within the literature.

CASE REPORT

A 44-year-old male patient presented at our center because of long-term exertional dyspnea, palpitations, and claudication of the lower extremities. General status of the patient was completely normal. On physical examination, peripheral pulses were palpable over the carotid arteries and in the upper limbs, but lower limb pulses were not. On admission, arterial blood pressure was 160/90 mmHg in both arms. Transthoracic echocardiography did not show any structural cardiac anomaly. Descending aorta could not be passed with a catheter during conventional angiography. In order to evaluate the aortic arch, thoracic multislice computed tomography (CT) angiography was performed, which revealed an interruption of the descending aorta beyond the left subclavian artery (Figure 1). The distal aortic part of the interruption was aneurysmal (diameter, 55 mm; length, 10 cm).

The right femoral artery and vein were cannulated following systemic heparinization. The left hemithorax was

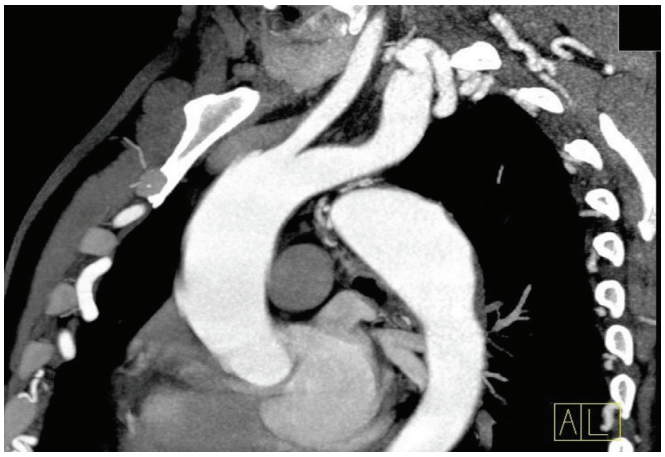

Figure 1. Computed tomography angiography revealing the interruption of the aorta.

Received July 25, 2010; received in revised form October 10, 2010; accepted October 19, 2010.

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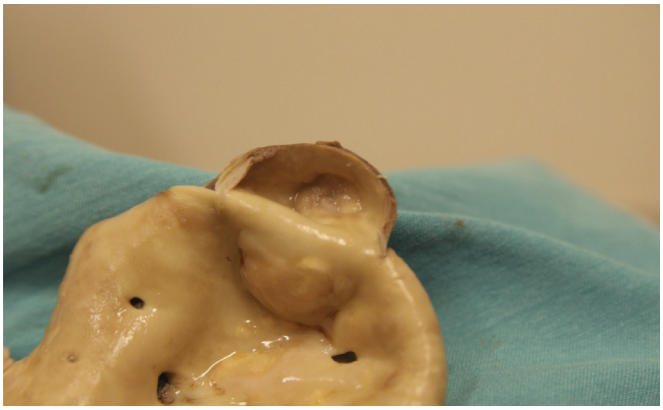

Figure 2. Macroscopic specimen of the interrupted aortic tissue.

accessed through a left posterolateral thoracotomy from the fourth intercostal space. The distal aortic arch and aneurysmal descending aorta were prepared and controlled with separate elastic tapes. There was no aortic neck below the left subclavian artery. The aneurysmatic portion of the descending aorta was heavily calcified. Special care was taken to avoid injury to the extensive collateral vessels. After the institution of cardiopulmonary bypass, the patient was cooled to nasopharyngeal temperature of 18°C. The aneurysmatic aorta including the interrupted segment was excised. There was no luminal and fibrous continuity between the proximal and distal segments. The ligamentum arteriosum was ligated and divided (Figure 2). A 24-mm single branched dacron graft (Polythese Polybranch; Perouse Medical, Ivry Le Temple, France) was interposed to the descending aorta with 4/0 polypropylene under total circulatuary arrest (TCA). After the proximal anastomosis was completed, TCA was terminated and circulation reinitiated via both the femoral artery and the side branch of the graft. Distal anastomosis was accomplished during the rewarming period. The perioperative period was uneventful, and the patient was discharged on the sixth postoperative day.

DISCUSSION

We conducted the current study using the PubMed/MEDLINE and Google Scholar databases, employing a search strategy that combined the words "interrupted aortic arch and adults." The world medical English literature contains only a few cases of isolated IAA in adults. In this study, we reviewed the 28 cases diagnosed with isolated IAA who reached adult age (>20 years) including our case (Table). In this series of patients, 71.5% were reported to be type A, 25% type B, and 3.5% type C aortic interruption. Five patients were treated conservatively, 1 patient underwent stent placement, and the remaining patients were treated surgically. The macroscopic pathological specimen was not stated in any report. Two patients had undergone anatomical graft interposition, and the others had extra anatomical bypass. In 2 patients, the type of surgical procedure was not specified.

If previous surgical intervention is not performed in a patient with IAA, this patient will very rarely reach adult age, depending on the adequacy of collaterals [Celoria 1959; Hokenek 2008]. There are some doubts about whether the anomaly is a real interrupted aortic arch or whether it is an aortic coarctation. The hallmark feature of aortic interruption that is useful to differentiate from aortic coarctation is the "complete absence" of continuity between both parts of the interrupted segment. The gold standard to distinguish these 2 pathologies is the direct vision during the operation and examination of the pathological specimen. The present report is unique in the literature that reveals the macroscopic specimen of the interrupted segment (Figure 2).

In the literature, there is lack of data that coarctation may progress to occlusion. A literature search reveals only 1 report with this kind of pathology, reported by Kaneda et al and diagnosed with just physical examination 23 years before, so the conclusion is not clear [Kaneda 2003].

Our search reveals the lack of universal nomenculature for IAA, especially for adults. According to the classification of Patton and Celoria, which only considers the location of the interruption on the aorta, our case can be classified as "Type A;" however, ductus arteriosus of our patient was not patent in contrast to the original definition of the authors, and this makes our definition questionable, as were the previous reports. Maybe our case can be classified as "isolated" IAA. Conversely to their clear cut-off points of the absent aortic arch and considering just the location of the aortic interruption without concomitant arch anomalies or intra-/extracardiac lesions such as ventriculosetal defect or PDA, many different variations exist. Moreover, with this classification, the matter of blood supply or function of the distal part of the interruption remains unclear.

The primary aim of the treatment in such patients is the correction of the interruption; however, additional intracardiac lesions should not be omitted, and the surgical strategy should be designed to take account of the entire cardiovascular pathology. Other surgical strategies, such as 2-stage repair, have been reported previously, but the accepted modern surgical concept is total correction consisting of both reconstruction of the aortic arch and repair of the intracardiac defects in a single-stage procedure. As mentioned previously, extra-anatomic bypass procedures are the most frequently preferred method of reconstruction in adult patients with IAA [Sai Krishna 2005]. The main reason for this surgical option is the risk of neurological events such as paraplegia. However, short aortic clamping times with a high descending aortic pressure may prevent this kind of complication.

We believe that in the present case aneurysmal dilatation at the distal portion of the interruption is not directly related to pathology of IAA, and such an aneurysm may be the result of hypertension related to an activated renin-angiotensin system, atherosclerotic disease, and massive collateral circulation.

Because our patient was symptomatic and had an enlarged descending aortic aneurysm, he underwent an anatomical surgical repair under deep hypothermia and TCA. To our knowledge, this case is one of the oldest adults with IAA to reach to the fifth decade without previous intervention and the first patient in the literature to undergo an anatomical repair via extracorporeal circulation and TCA.

Adult Patients with Surgical Repair or Medical Treatment of Interrupted Aortic Arch (IAA)*

	Reference	Patient Age, y	Sex	Type of IAA	Additional Disease/Procedure	Treatment
1	Kauff 1973	36	M	A	—	Subclavian bypass
2	Kauff 1973	49	M	B	—	Anatomical graft interposition
3	Milo 1982	65	M	B	—	Brachiocephalic bypass
4	Wong 1989	33	M	A	—	Medical
5	Burton 1995	24	M	A	Ascending aortic aneurysm + AI/root replacement	Ascening descending bypass
6	Canova 1995	72	F	A	AI/AVR	Ascending descending bypass
7	Ogino 1998	32	M	A	AI + sinus valsalva aneurysm	Subclavian descending bypass
8	Messner 2002	42	M	B	—	Subclavian descending bypass
9	Riess 2004	64	M	A	Coronary disease + AI; CABG + AVR	Ascending descending bypass
10	Sai Krishna 2005	64	M	A	Bicuspit aorta	Subclavian descending bypass
11	Sai Krishna 2005	40	M	B	Subarachanoid bleeding/ACA aneurysm repair	Ascending descending bypass
12	Sai Krishna 2005	46	F	A	Mid LAD disease	Ascending descending bypass
13	Sai Krishna 2005	34	M	B	—	Ascending descending bypass
14	Sai Krishna 2005	40	M	A	—	Ascending descending bypas
15	Sai Krishna 2005	23	M	A	—	Ascending descending bypass
16	Maier 2005	65	F	A	—	Medical
17	Sim 2007	43	F	C	—	Medical
18	Sari 2007	40	M	A	—	Surgery†
19	Gokce 2010	40	F	A	—	Surgery†
20	Casati 2008	35	M	A	MI/MVR	Subclavian descending bypas
21	Hokenek 2008	21	M	A	—	Anatomical graft interposition
22	Celkan 2008	35	M	A	—	Ascending descending bypass
23	Yildirim 2008	26	M	B	—	Medical
24	Kusa 2008	26	M	B	—	Stent
25	Alam 2009	72	M	A	—	Medical
26	Sakellaridis 2010	62	M	A	—	Subclavian descending bypass
27	Mirat 2010	60	M	A	—	Ascending descending bypass
28	Teskin 2010 (present case)	46	M	A	Descending aortic aneurysm	Anatomical graft interposition

*AI indicates aortic insufficiency; AVR, aortic valve replacement; CABG, coronary artery bypass graft; ACA, anterior cerebral artery; LAD, left anterior descending coronary artery; MI, myocardial infarction; MVR, mitral valve replacement.

†Type of surgery is not clarified.

We performed an anatomical graft interposition for the descending aorta under extracorporeal circulation; due to the interruption, the aorta was closed to the left subclavian artery. Proximal anastomosis was successful under TCA for maximal surgical exposure by eliminating substantial collateral circulation. Both TCA and aortic cross clamp times were short in our patient (22 minutes and 24 minutes, respectively), and we did not observe any kind of neurological, renal, or respiratory complications. The protective effects of hypothermia are largely presumed to be secondary to decreased tissue metabolism and a generalized reduction in energy-requiring processes in the cell [Lillehei 1969; Mault 1993; Kouchoukos 2001; Kouchoukos 2004].

As a conclusion, IAA in adulthood is one of the rarest anomalies of cardiovascular disorders and must be treated with either anatomic or extra-anatomic reconstruction as soon as it is diagnosed. The current classification system for this group of patients just considers the location of the interrupted aortic arch without taking into account other anatomic patterns and associated cardiac anomalies. We believe that such a classification does not meet the real anatomical or clinical situation.

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