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Undiagnosed type B interrupted aortic arch without a patent ductus arteriosus identified during adolescence☆

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ABSTRACT

Type B interrupted aortic arch without a patent ductus arteriosus or ventricular septal defect is extremely rare. We present a case of this congenital abnormality with the initial presentation at 15 years of age. Our case demonstrates the efficacy of CT angiography in diagnosing this condition in addition to characterizing the anatomy of collateral blood flow. Furthermore, this study provides updated imaging information on interrupted aortic arch without a patent ductus arteriosus and will guide physicians in selecting the appropriate diagnostic imaging modality to evaluate this condition.

1. Introduction

Interruption of the aortic arch accounts for less than 1% of all cases of congenital heart disease and was first reported by Celoria and Patton in 1959 [1]. According to Celoria's revised classification, interrupted aortic arch is divided into three main types, dependent on the location of the interruption [2]. In type A, the interruption exists distal to the left subclavian artery. In type B, the interruption is between the left common carotid artery and the left subclavian artery. Finally, in type C, the interruption is between the right brachiocephalic artery and the left common carotid artery [3].

In infants, 53% of all interrupted aortic arch cases are type B, 43% are type A, and 4% are type C [4]. However, type A is the most common interrupted aortic arch in adults and accounts for 79% of cases, with types B and C accounting for 16% and 3% of cases respectively. Type B interrupted aortic arch is closely associated with 22q11.2 Deletion Syndrome, and may be caused by defective formation of the third and fourth arches during fetal development [5].

Early survival of infants with interrupted aortic arch is dependent on the patency of the ductus arteriosus and/or ventricular septal defect (VSD) [6]. Type B interrupted aortic arch without patent ductus arteriosus has not been documented since 1975 [7]. This case study presents an adolescent female patient with type B interrupted aortic arch without a patent ductus arteriosus. For the first time, detailed images from cardiac magnetic resonance (CMR), chest and neck computed tomography angiography (CTA), and 3-D reconstruction of such a case have been captured and are presented in this report.

2. Case Presentation

A 15-year-old female with DiGeorge's Syndrome presented to the pediatric cardiology clinic for evaluation of a heart murmur and shortness of breath. The patient reportedly had a history of bicuspid aortic valve and ventricular septal defect that never required medical therapy or surgical intervention. She occasionally experienced shortness of breath, and was suspected of having mild developmental delay. Physical examination was significant for a soft systolic ejection click associated with a 1 to 2/6 systolic ejection murmur. Physical examination was also notable for obesity and acanthosis nigricans on her neck. She denied chest pain, palpitations, dizziness, and syncope. Blood pressures were as follows: right arm 99/73, left arm 115/64, left leg 107/33, right leg 98/49.

The patient's EKG was unremarkable with no evidence of left ventricular hypertrophy. An echocardiogram was then performed, which

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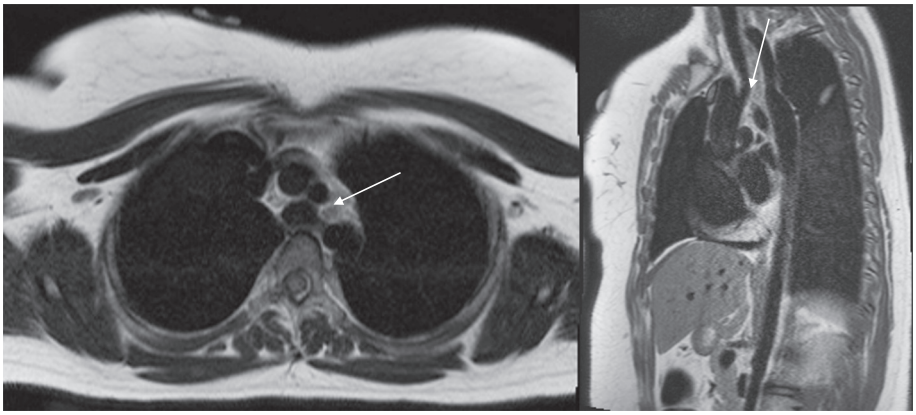

Fig. 1. Axial and candy cane DIR chest. Interruption of the aortic arch (white arrows) between the left common carotid and aberrant right subclavian arteries.

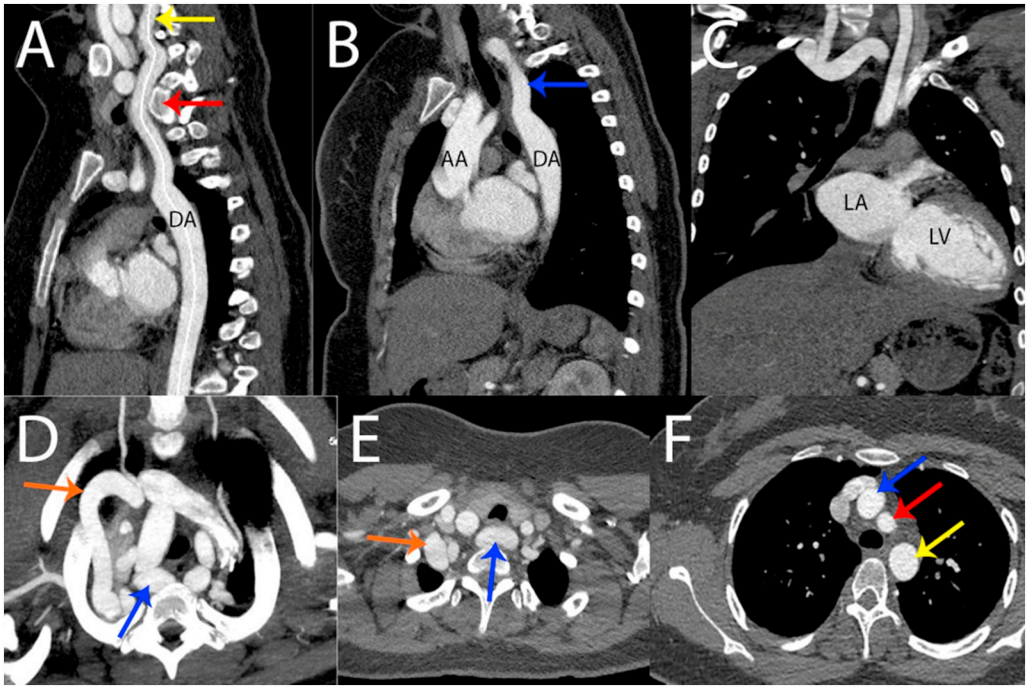

Fig. 2. Curviplanar (A) and sagittal chest CTA (B) shows IAA without a PDA and dilation of the right external carotid artery (A, yellow arrow) that provides blood flow to the right subclavian artery (A, red arrow) and descending aorta (DA). The dilated, tortuous subclavian artery passes behind the esophagus (B, blue arrow). (AA = ascending aorta). Coronal CTA chest (C) demonstrates a dilated left ventricle with ventricular non-compaction (LA = left atrium; LV = left ventricle). Axial MIP and thin slice chest CTA (C–F) shows the right common carotid (F, blue arrow) and left common carotid (F, red arrow) arteries originating from the ascending aorta (not shown) and the dilated, aberrant right subclavian (D–E, orange and blue arrows) supplying the descending aorta (F, yellow arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

demonstrated an abnormal aortic arch. Additional findings included dilated left ventricle (LV) with a thin interventricular septum, decreased global LV function, LV ejection fraction of 45–50%, reversal of the E/A ratio, partially fused bicuspid aortic valve, and abnormal flow in the descending aorta with forward flow in diastole. The subsequent differential included interrupted aortic arch, vascular ring, coarctation of the aorta, and ventricular noncompaction or spongiform cardiomyopathy.

Multiplanar, multisequence CMR imaging was performed without contrast. Computerized hemodynamic analysis demonstrated moderate dilation of the left ventricle with an ejection fraction of 42%. Right ventricular morphology and function were normal. CMR revealed a small interatrial diverticulum, LV non-compaction with a ratio of 2.5 and interventricular septal thinning with dyskinesia. MR imaging of the chest demonstrated a normal aortic root, 28 mm ascending aorta caliber, a collateral artery supplying the left-sided descending aorta coursing posterior to the esophagus (Fig. 1) and a small left pulmonary artery measuring 11 mm. Right and main pulmonary artery measurements were normal.

A CT angiogram of the chest and neck was performed to better delineate the vascular anatomy. 2D maximum intensity projection reformations and 3D post processing of the aorta and neck vessels were performed. This method identified a left aortic arch with an interrupted aortic arch between the left common carotid artery and the subclavian

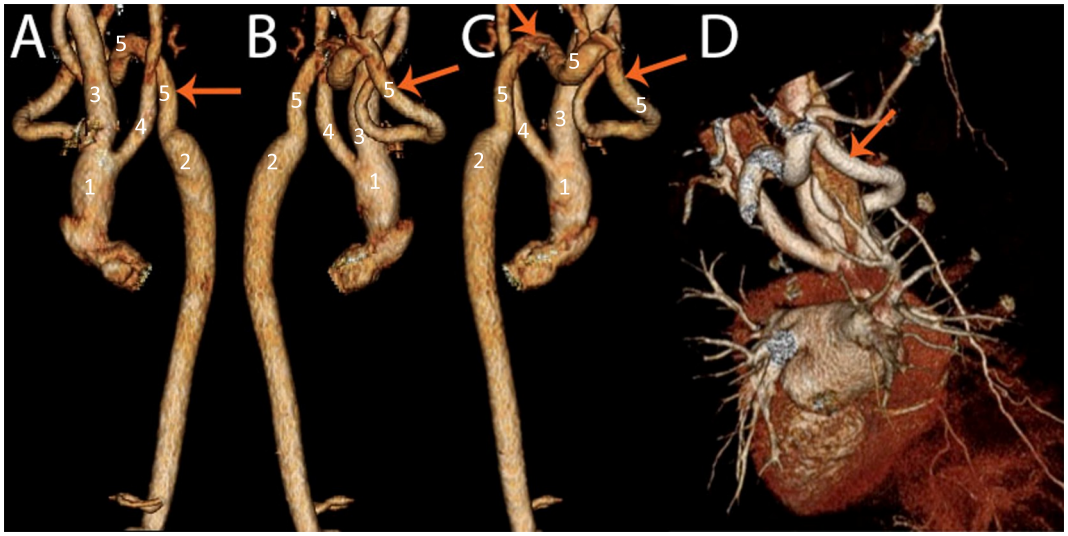

Fig. 3. Images depict 3D volume rendered CT angiography of type B IAA without a PDA (A-D). A through C are from anterior, right oblique, and posterior views, respectively. Note the dilated, tortuous, aberrant right subclavian artery supplying the descending aorta (orange arrows). (1 = ascending aorta; 2 = descending aorta; 3 = right common carotid artery; 4 = left common carotid artery; 5 = right subclavian artery). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

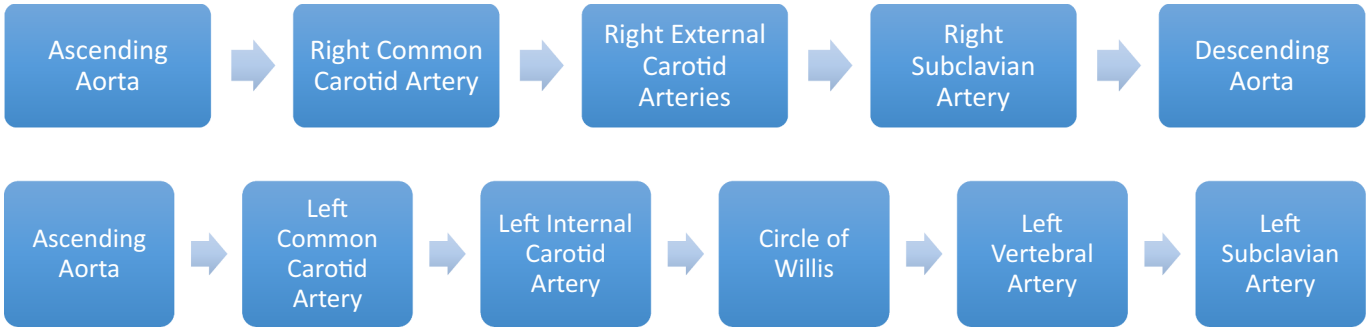

Fig. 4. Schematic depicting the complex course of blood flow between the ascending aorta and descending aorta (A) and left subclavian artery (B).

arteries. There was a tortuous aberrant right subclavian artery coursing posterior to the esophagus (Fig. 2).

There was no PDA or other direct communication of the aorta with the pulmonary arteries. The ascending aorta gave rise to independent right and left common carotid arteries. The proximal descending aorta gave rise to separate right and left subclavian arteries. The dilated right vertebral artery originated from the dilated right subclavian artery. The descending aorta was supplied via retrograde flow in the aberrant right subclavian artery (Figs. 3 and 4). The left subclavian artery was severely atretic at the ostium (Fig. 4). The left vertebral artery supplied the left subclavian artery via retrograde flow (Fig. 5). The left ventricle was dilated and had an abnormal configuration, with septal bowing toward the right ventricle. The left atrium was dilated with increased trabeculation. There was a small aneurysm in the interatrial septum with a base measuring 2.5 cm and a depth of 1.2 cm.

Imaging findings were confirmed at surgery. The ligamentum arteriosum was ligated and divided. The interrupted arch was treated with an end-to-side 18 mm Dacron interposition graft after making longitudinal incisions in the distal ascending aorta and proximal descending aorta. Following creation of the prosthetic transverse arch, aortic pressures were equivalent above and below the diaphragm and intraoperative echocardiography showed a patent thoracic aorta. There were no operative or postoperative complications and the patient had equal pulses in all extremities. Repeat CT angiography of the chest was performed two days after surgery showing a patent graft (Fig. 6). The patient was doing well 6 months after surgery with no complaints.

3. Disscussion

In our study we provide a thorough imaging description of interrupted aortic arch with type B configuration without a patent ductus arteriosus presenting in mid-adolescence. To our knowledge, this is the

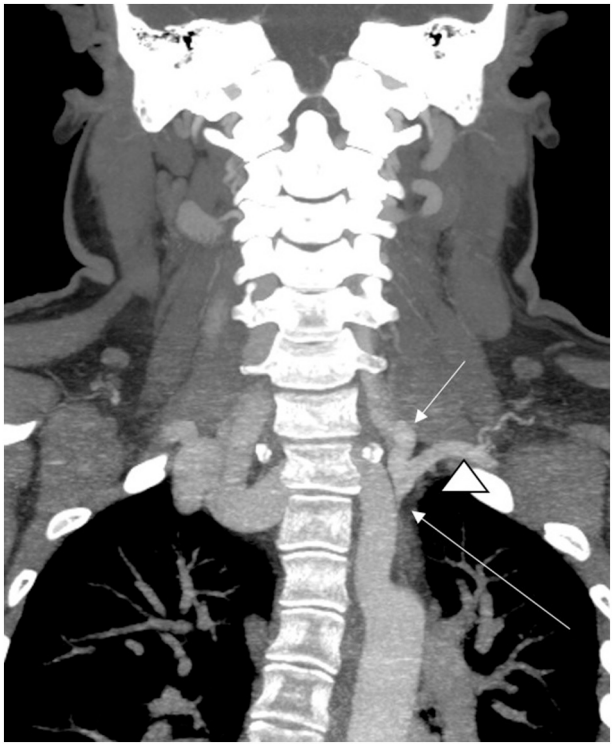

Fig. 5. Coronal maximal intensity projection image of the neck CT angiogram. Atretic ostium of the left subclavian artery (long arrow). Retrograde supply of the left subclavian artery (arrowhead) by the left vertebral artery (short arrow).

first time that cardiac MRI, chest CTA, and 3D reconstruction imaging of this condition has been published. Type B interrupted aortic arch without patent ductus arteriosus is extremely rare, with only one similar case reported since 1975 [7]. Cazavet et al. previously reported a 2.5-month-old girl with a similar unusual interrupted aortic arch [8]. One key difference was their patient had a small patent ductus arteriosus measuring 2 mm diameter and a 4 mm atrial septal defect. Also, their patient was diagnosed with the disorder as an infant. Finally, their case did not include cardiothoracic MR angiography. Previously reported cases in the last 40 years have only included type A interrupted aortic arch [4,6] or type B interrupted aortic arch [3,9]. Our case adds to the existing literature by describing the variety of collateral blood flow that can develop in type B interrupted aortic arch without a patent ductus arteriosus.

Most cases of interrupted aortic arch result in death in infancy if not diagnosed and treated promptly [7]. Generally, interrupted aortic arch presenting during infancy and early childhood is associated with patent ductus arteriosus and/or VSD. Case reports of those surviving beyond early childhood without surgery frequently lack patent ductus arteriosus and/or VSD, but instead have collateral circulation supplying the descending aorta.

Our case provides helpful information to direct a diagnostic approach for patients with interrupted aortic arch without patent ductus arteriosus. In our case it was difficult to clarify the vascular anatomy until CTA of the chest was performed. CTA also accurately demonstrated which vessels provided collateral blood flow. Chest MRA would probably provide similar results. The vertebral arteries, much like in previously documented case reports, play a major role in providing blood flow to the descending aorta in interrupted aortic arch without patent ductus arteriosus and must be clearly defined on imaging [3,4,6,7,9].

In summary, type B interrupted aortic arch without patent ductus arteriosus is extremely rare. Our case demonstrates the efficacy of chest CTA in diagnosing this condition in addition to characterizing the anatomy of collateral blood flow. Furthermore, this study provides updated imaging information on interrupted aortic arch without patent ductus arteriosus and will guide physicians in selecting the appropriate diagnostic imaging modality to evaluate this condition.

Author Declaration

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us.

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

We further confirm that any aspect of the work covered in this manuscript that has involved either experimental animals or human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

We understand that the Corresponding Author is the sole contact for the Editorial process (including Editorial Manager and direct communications with the office). He is responsible for communicating with the other authors about progress, submissions of revisions and final approval of proofs. We confirm that we have provided a current, correct email address which is accessible by the Corresponding Author and which has been configured to accept email from Jeremy.burt.md@flhosp.org.

Declarations of Interest

Authors declare there are no conflicts.

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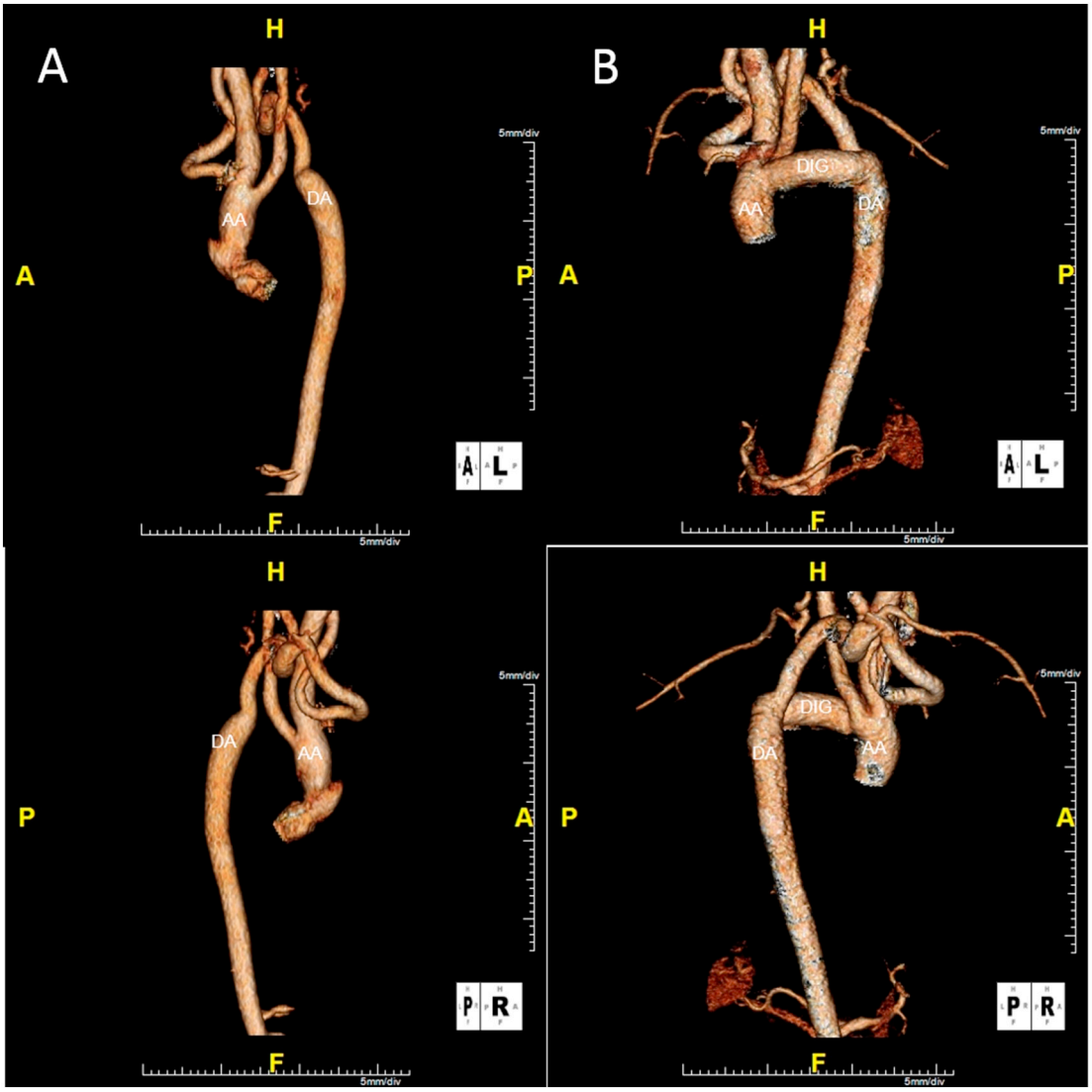

Fig. 6. Images depict 3D volume rendered CT angiography of type B IAA without a PDA before (A) and after (B) surgical repair. DIG = Dacron interposition graft; AA = ascending aorta; DA = descending aorta.

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