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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Undiagnosed Double Aortic Arch in an Adult With Repaired Tetralogy of Fallot

Anita Sadeghpour, MD,[a] Nakisa Khansari, MD,[a] Marziyeh Pakbaz, MD,[b] Kiara Rezaei Kalantari, MD,[c] Hamidreza Pouraliakbar, MD,[c] Maziar Gholampour Dehaki, MD[d]

ABSTRACT

Double aortic arch, the most common vascular ring, causes a complete ring surrounding the esophagus and trachea that leads to compressive symptoms. This report describes a young woman with a history of totally corrected tetralogy of Fallot who was a candidate for pulmonic valve replacement. A double aortic arch was detected incidentally by echocardiography and cardiac computed tomography. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2019;1:540–4) © 2019 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

The patient was a 22-year-old woman who had undergone total correction of tetralogy of Fallot with surgical closure of a patent ductus arteriosus 10 years earlier. She had been asymptomatic and under regular follow-up since then. At the last visit, however, she presented with new symptoms of exertional dyspnea and dysphagia. The physical examination was remarkable for a funnel chest and a diastolic murmur in the upper left sternal border. The patient's general physical appearance was that of an underweight woman who seemed younger than her chronological age.

PAST MEDICAL HISTORY

The patient's past history was remarkable for total surgical correction of tetralogy of Fallot.

INVESTIGATIONS

The electrocardiogram showed sinus rhythm, a right bundle branch block, and right-axis deviation. The

LEARNING OBJECTIVES

- Vascular rings are an uncommon type of congenital cardiovascular disease in which vascular structures completely or incompletely surround the trachea and esophagus.
- The most frequent type of complete vascular ring is DAA.
- DAA may cause symptoms resulting from its compressive effect on the esophagus and trachea.
- The combination of DAA and tetralogy of Fallot is extremely rare and may remain undetected until adulthood.
- Echocardiography can reveal abnormal arch anatomy and intracardiac anomalies.
- Cardiac computed tomography and magnetic resonance are valuable imaging methods for detecting DAA.
- Surgical intervention is the treatment of choice in symptomatic cases.

From the [a]Echocardiography Research Center, Rajaie Cardiovascular, Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran; [b]Department of Cardiovascular Disease, Hazrat-e Rasool General Hospital, Iran University of Medical Sciences, Tehran, Iran; [c]Department of Radiology, Rajaie Cardiovascular, Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran; and the [d]Department of Cardiac Surgery, Rajaie Cardiovascular, Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Informed consent was obtained for this case.

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chest radiograph showed a normal cardiothoracic ratio, prominent lung hila, and slight widening of the mediastinum. The transthoracic echocardiogram showed a normal-size left ventricle with moderate systolic dysfunction, severe right ventricular enlargement with moderate systolic dysfunction, normal mitral and aortic valves, moderate tricuspid regurgitation, severe free pulmonary regurgitation, no significant pulmonary hypertension, no residual ventricular septal defect, and no intracardiac or extracardiac shunt; however, it showed a double aortic arch (DAA) with a prominent right arch and a hypoplastic left arch (**Figure 1**, Videos 1 and 2). The DAA was confirmed by cardiac magnetic resonance (**Figure 2**) and computed tomography (CT) angiography (**Figure 3**, Video 3), which showed a complete ring surrounding the trachea and esophagus. The left subclavian and carotid arteries branched from the left aortic arch, and the right subclavian and carotid arteries branched directly from the right aortic arch, thus resulting in the classic "4-vessel sign." Other CT findings included stenosis of the origin of the left pulmonary branch artery, a right-sided descending aorta, and abnormal drainage of the right upper pulmonary vein to the superior vena cava. The anatomy and course of coronary arteries were normal.

MANAGEMENT

Because of her symptoms, the patient was scheduled for cardiac surgery and underwent successful pulmonary valve replacement, repair of the left pulmonary branch artery origin stenosis, repair of the abnormal drainage of the right upper pulmonary vein, and division of the nondominant left aortic arch. The patient experienced an uneventful postoperative course, and her symptoms of exertional dyspnea and dysphagia resolved.

ABBREVIATIONS AND ACRONYMS

CT = computed tomography

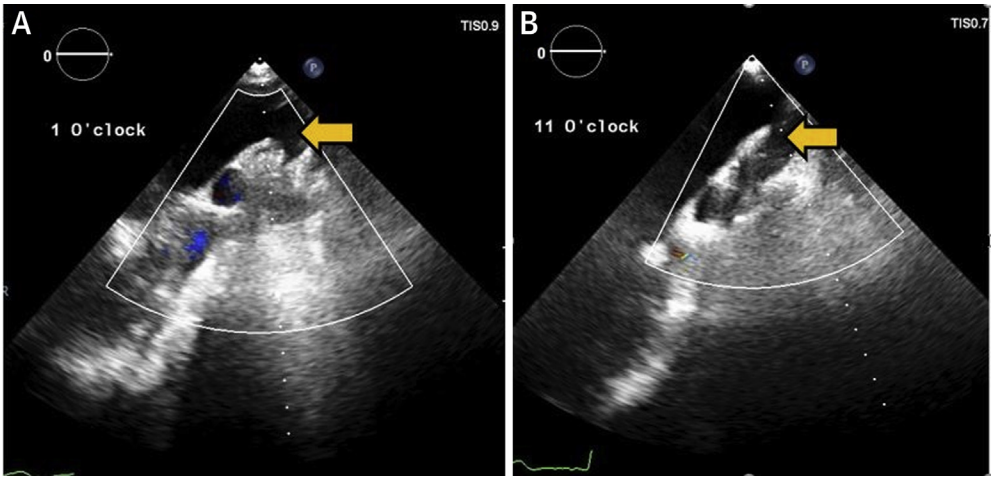
DAA = double aortic arch

DISCUSSION

Vascular rings are anomalies of the aortic arch system in which vascular structures (that are not necessarily patent) surround the trachea and esophagus and form a complete or incomplete ring around them (1). The most frequent type of complete vascular ring is the DAA, which surrounds the trachea and esophagus and may cause symptoms resulting from compression of these structures (2). In the case of severe compression, a DAA may be diagnosed early after birth in an infant with stridor, dyspnea, and feeding difficulties. Occasionally, when compression is minimal, a DAA maybe undetected until adulthood (3). Our patient was a young woman with a history of totally corrected tetralogy of Fallot who had an undiagnosed DAA that was detected incidentally by echocardiography and confirmed by cardiac CT.

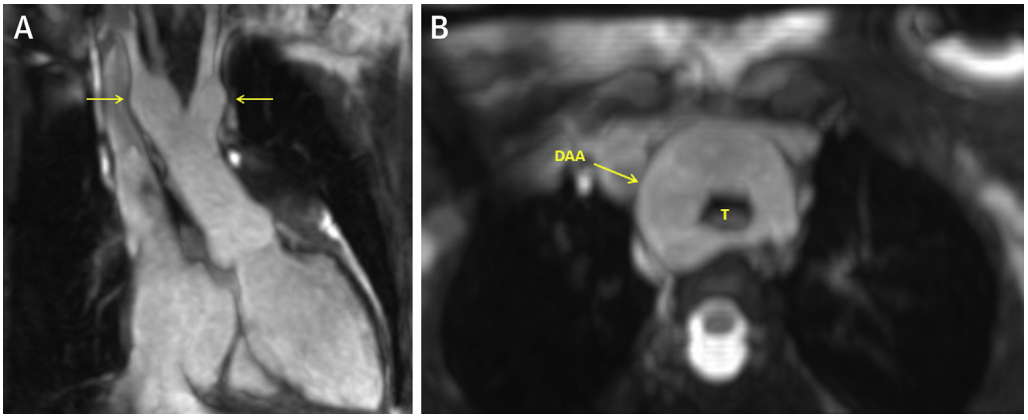
Indeed, in our patient dysphagia was not revealed to be a major symptom until we asked her about it. This shows the importance of precise history taking. Abnormal rearrangement of the 6 paired arch vessels during the fourth week of embryogenesis results in various forms of vascular rings (4,5). DAA is the result

FIGURE 1 Transthoracic Echocardiography of Double Aortic Arch



Suprasternal view of transthoracic echocardiography with the index marker of the probe pointing to **(A)** 1 o'clock revealing a hypoplastic left arch and to **(B)** 11 O' clock revealing the prominent right arch. The **arrows** show the arches in each view. See Videos 1 and 2.

FIGURE 2 Cardiac Magnetic Resonance of DAA

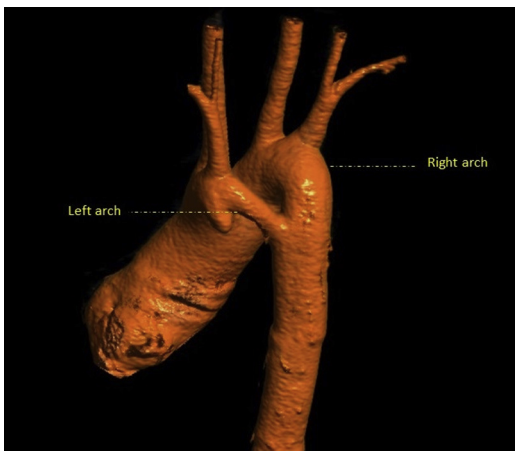


Cardiac magnetic resonance revealing the double aortic arch (DAA) in the **(A)** coronal and **(B)** axial views with a dominant right arch and a hypoplastic left arch **(arrows)** surrounding the trachea (T) and esophagus.

of the persistence of both fourth arches that join the descending thoracic aorta (**Figure 4**). The 2 arches surround and compress the trachea and esophagus (**Figure 5**). In most cases of DAA, 1 of the arches is dominant (the right arch is dominant in 75% of cases). In rare cases the arches are equal in size (6). Our patient had a DAA with a dominant right arch.

The DAA is usually an isolated lesion. However, it may be associated with other congenital anomalies in 22% of cases, including tetralogy of Fallot, transposition of the great vessels, and truncus arteriosus (2). Asymptomatic DAA with tetralogy of Fallot is extremely rare (7).

FIGURE 3 Cardiac Computed Tomography of Double Aortic Arch



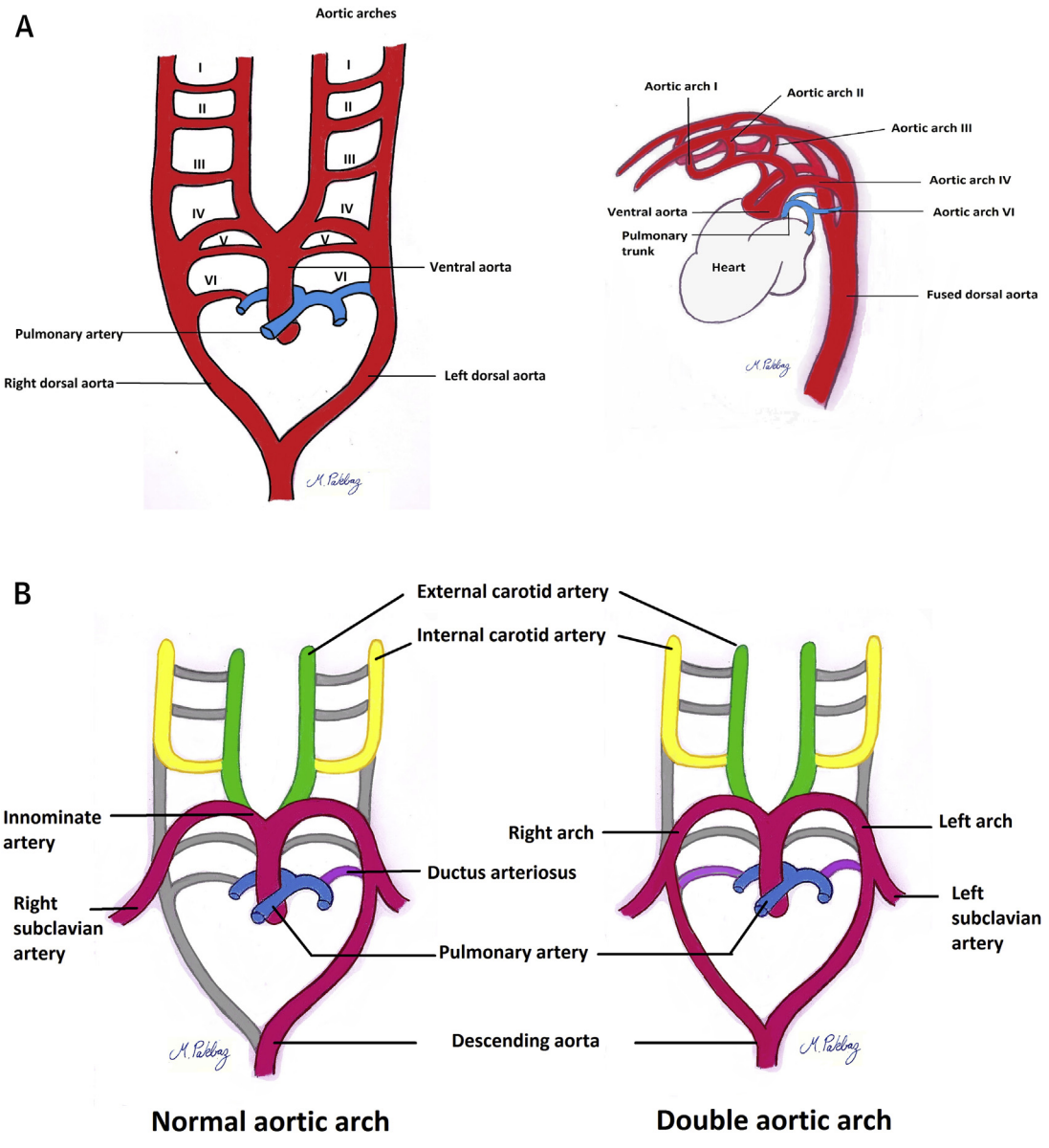
Cardiac computed tomography, volume-rendered reconstructed image showing the double aortic arch resulting in a vascular ring. See Video 3.

The onset of symptoms is directly related to the severity of anatomic tracheoesophageal compression by the surrounding vascular ring. If the compression is severe, symptoms such as dyspnea, recurrent pneumonia, and feeding difficulties may be observed at birth. However, when compression is minimal, a DAA maybe undetected until adulthood, as occurred in our patient (3).

Many diagnostic modalities may help in detecting this anomaly. The chest radiograph may be otherwise normal or may show mediastinal widening, bilateral aortic knobs, and/or indentation of the trachea (7). Echocardiography may reveal the abnormal anatomy of the aortic arch and may also aid in detecting intracardiac anomalies. Currently, CT and cardiac magnetic resonance are the most accurate diagnostic modalities for evaluation of vascular rings and permit characterization of the location and severity of airway and esophageal obstruction (7). In patients presenting with dysphagia, a barium esophagogram may show left and right notches in the esophagus (2).

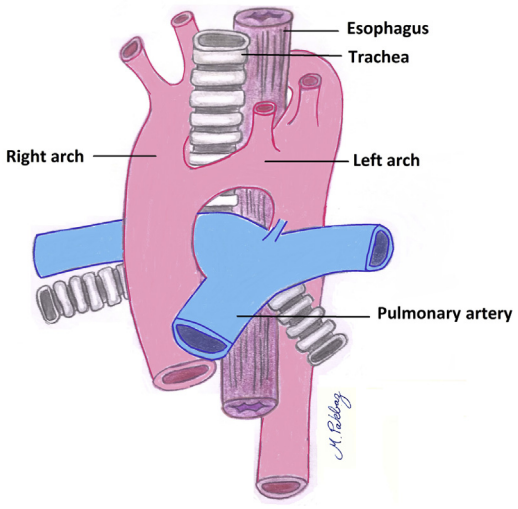
Surgical resection of the minor aortic arch is the only curative choice for symptomatic patients to relieve the compressive features. This arch can be approached through a median sternotomy or left posterolateral thoracotomy (8). The success rate of surgery is high in infants with DAA (9). There are some reports of successful surgical treatment performed in adult patients with DAA (10).

FIGURE 4 Aortic Arch Embryology



(A) The aortic arch develops from 6 symmetrical paired vessels and the paired dorsal aortas. During embryogenesis, there is an aortic arch and a ductus arteriosus on each side at first. **(B) (Left)** further development of the arch system. The **gray segments** normally regress. The **colored segments** finally contribute in the arch structure and branches. Each **arch** gives rise to the ipsilateral branches of carotid and subclavian arteries. Normally, an interruption occurs in the right arch between the right subclavian and descending aorta, and the right ductus arteriosus and dorsal aorta regress **(left).** Failure of this interruption and regression results in the formation of a double aortic arch **(right).**

FIGURE 5 Anatomic Orientation of Double Aortic Arch Related to the Esophagus and Trachea



FOLLOW-UP

Our patient became symptom free following surgery, and she is doing well on her regular follow-up visits.

CONCLUSIONS

We reported a case of repaired tetralogy of Fallot in a patient who was a candidate for redo surgery for severe pulmonary regurgitation and who had a DAA that was discovered incidentally by transthoracic echocardiography and confirmed by CT angiography.

ADDRESS FOR CORRESPONDENCE: Dr. Marziyeh Pakbaz, Department of Cardiovascular Disease, Hazrat-e Rasool General Hospital, Iran University of Medical Sciences, Sattar-khan Avenue, Niyayesh Street, Tehran 1445613131, Iran. E-mail: marzi.pakbaz@gmail.com.

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KEY WORDS congenital heart disease, double aortic arch, tetralogy of Fallot, vascular ring

APPENDIX For supplemental videos, please see the online version of this paper.