

# Double Choker: Double Aortic Arch with Bilateral Aortic Coarctation Associated with Heterotaxy-Asplenia Syndrome and Complex Atrioventricular Canal Defect



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## INTRODUCTION

Double aortic arch is a congenital anomaly that results from persistence of both aortic arches in utero and is the most common aortic arch anomaly resulting in a symptomatic vascular ring. The majority of cases of double aortic arch occur in isolation,<sup>1</sup> but intracardiac associations with septal defects, tetralogy of Fallot, and transposition of the great arteries have been reported.<sup>1-4</sup> We present the case of a patient with double aortic arch with bilateral aortic coarctation diagnosed by echocardiography with associated heterotaxy/asplenia syndrome, dextrocardia, right ventricular–dominant atrioventricular canal defect, and double-outlet right ventricle. To our knowledge, this is the fourth case in the world literature of double aortic arch with coarctation of both arches and the first case to be diagnosed using echocardiography with associated complex intracardiac disease.

## CASE PRESENTATION

A 21-year-old mother was referred for fetal echocardiography because of dextrocardia noted on obstetric ultrasound at 32 weeks' gestational age. Fetal echocardiography demonstrated heterotaxy syndrome with dextrocardia with cardiac segments {A,D,L} (situs ambiguous, D-looped ventricles, and L-malposed great arteries), right ventricular–dominant common atrioventricular canal defect, double-outlet right ventricle, and hypoplastic aortic valve and aortic arch. Our patient was born via cesarean section at 38 weeks' gestation, promptly intubated because of significant hypoxia, and started on prostaglandin immediately because of concern for aortic coarctation on fetal echocardiography. Postnatal transthoracic echocardiography verified the fetal findings of heterotaxy with dextrocardia and abdominal and atrial situs ambiguous (Figure 1). The inferior vena cava was

left of the spine, draining to the left-sided atrium separately from the hepatic veins. Bilateral superior venae cavae were demonstrated and drained into their respective atria, and all of the pulmonary veins drained into the left-sided atrium. Complex intracardiac anatomy consisted of a right ventricular–dominant common atrioventricular canal defect with D-looped ventricles (Figure 2, Video 1), double-outlet right ventricle (Videos 1-3), L-malposed aorta with a restrictive inlet-to-outlet subaortic ventricular septal defect (Figure 3, Videos 1-4), dynamic subvalvar and valvar aortic stenosis (Videos 2, 3, and 5), and a double aortic arch with severe coarctation of both right and left dorsal arches (Figures 4-6, Video 6). A large right-sided patent ductus arteriosus supplied the left-sided descending aorta, which joined the right-sided dorsal aortic arch segment (Figures 4 and 5, Video 6). The semilunar valves were in close proximity because of a hypoplastic conal septum, with the aortic valve arising leftward and slightly posterior to the pulmonary artery (Figure 3, Video 7). Cardiac magnetic resonance imaging (MRI) confirmed the echocardiographic findings, with the additional demonstration of asplenia, a right-sided stomach, and a transverse liver (Figures 6 and 7, Video 8).

In the setting of this complex cardiac anatomy, the patient developed significant pulmonary overcirculation and lactic acidosis with acute kidney injury. Bilateral pulmonary artery bands were placed, but the postoperative course was complicated by significant pleural effusions, hypotension, and lactic acidosis. The patient had a cardiac arrest requiring cannulation with venoarterial extracorporeal membrane oxygenation. Unfortunately, the combination of airway obstruction from the vascular ring, pulmonary overcirculation, and ventricular outflow obstruction in the context of a functional single ventricle was a lethal physiologic constellation in our patient.

## DISCUSSION

To our knowledge, only three prior cases of double aortic arch with bilateral coarctation have been reported<sup>5-7</sup>; these cases were diagnosed using invasive angiography and/or surgical examination. Ours is the first case describing the diagnosis of double aortic arch with bilateral aortic coarctation based solely on echocardiography as well as being associated with complex intracardiac anatomy. This case demonstrates the importance of imaging each patient with congenital heart disease using a systematic and critical assessment of each cardiac component, following the segmental approach.

Abnormalities of the aortic arch account for approximately 3% of congenital heart disease. Double aortic arch results when both the right and left embryonic fourth aortic arches persist as they join the aortic portion of the truncocoarctic sac to their respective dorsal aortae, both of which persist as well.<sup>1</sup> This anomaly accounts for approximately

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## VIDEO HIGHLIGHTS

**Video 1:** Subcostal coronal sweep from posterior to anterior demonstrating complex intracardiac anatomy.

**Video 2:** Apical view obtained from the right flank showing double-outlet right ventricle with a leftward aorta and a small, restrictive inlet-to-outlet ventricular septal defect. Subvalvar and valvar aortic stenosis.

**Video 3:** Apical view obtained from the right flank with color Doppler showing double-outlet right ventricle with a leftward aorta and a small, restrictive inlet-to-outlet ventricular septal defect. Subvalvar and valvar aortic stenosis.

**Video 4:** Apical view with color-compare image of the restrictive ventricular septal defect in the inlet septum.

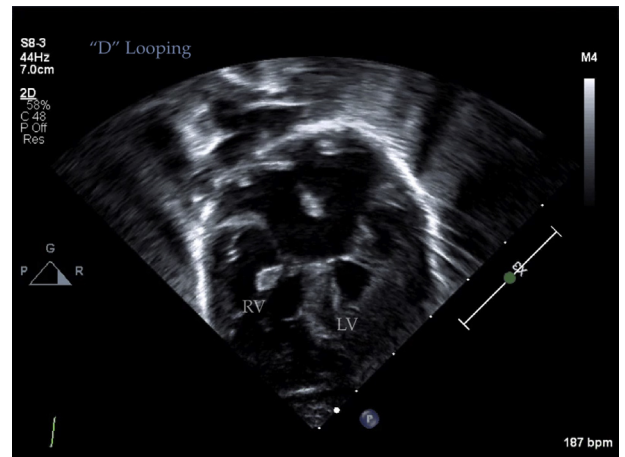
**Video 5:** Apical view of the outlets demonstrating funnel-like subaortic stenosis.

**Video 6:** Suprasternal notch color-compare sweep of the double aortic arch. Both hypoplastic arches are noted wrapping around the trachea.

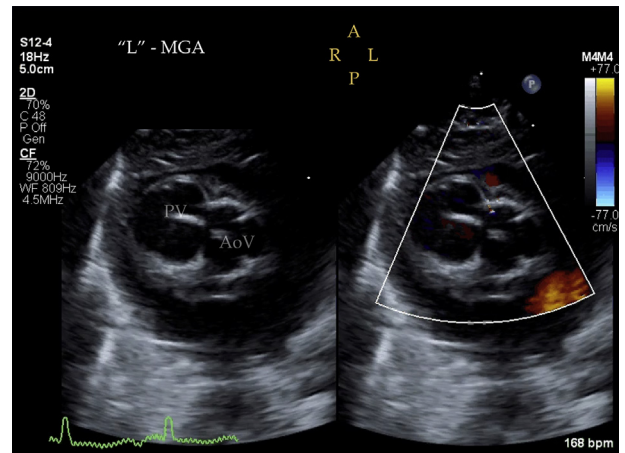
**Video 7:** Parasternal short-axis image of the semilunar valves demonstrating a leftward aorta. Notice that the semilunar valves are very close in proximity because of the hypoplastic conal septum.

**Video 8:** Segmented three-dimensional reconstruction demonstrating critical hypoplasia and long segment coarctation of both aortic arches (*red*) entering into the ductus (*green*). Hypoplastic left ventricle (*light red*), bilateral superior venae cavae entering into their respective atria. Left-sided inferior vena cava and accessory hepatic vein entering into the left atrium.

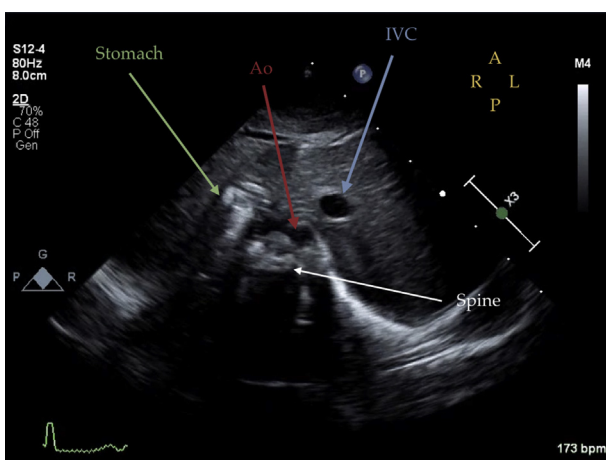
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**Figure 2** Apical four-chamber image from the right flank demonstrating D-looped ventricles. Hypoplastic and hypertensive left ventricle (LV) with tethering and severe stenosis of the left atrio-ventricular valve. RV, Right ventricle.



**Figure 3** Parasternal short-axis image of the semilunar valves demonstrating a leftward aorta. Notice that the semilunar valves are very close in proximity because of the hypoplastic conal septum. AoV, Aortic valve; MGA, malposition of the great arteries; PV, pulmonary valve.

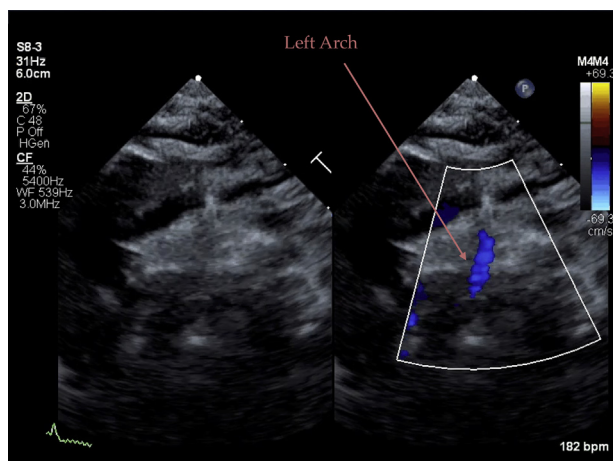


**Figure 1** Subcostal image demonstrating abdominal situs ambiguous. A right-sided stomach, midline liver, and left-sided inferior vena cava (IVC) are seen. Ao, Aorta.

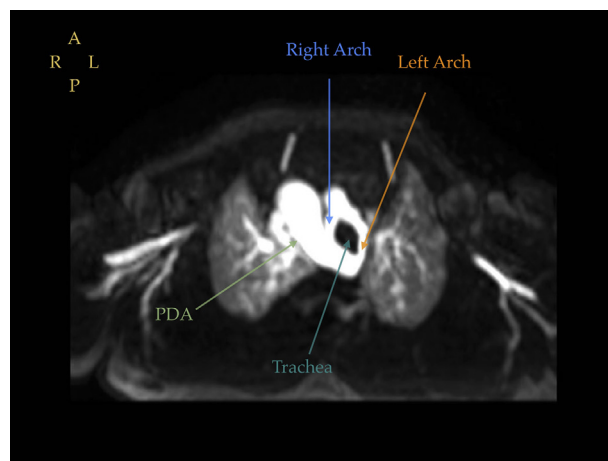
30% to 45% of vascular rings and is the most common symptomatic vascular ring on the basis of surgical series; the true incidence of asymptomatic vascular rings is difficult to ascertain.<sup>8</sup> The ascending aorta

bifurcates into a right-sided and a left-sided aortic arch. The right arch gives rise to the right common carotid and right subclavian arteries, while the left arch gives rise to the left common carotid and left subclavian arteries. Usually, double aortic arch presents with one arch larger than the other, including selective hypoplasia or atresia of the other arch. Most commonly, the right aortic arch is larger and dominant and the left aortic arch is hypoplastic or atretic. Although bilateral ductus arteriosus can be present, the ductus arteriosus (or ligamentum arteriosum) is typically left sided and needs to be considered during surgical repair.

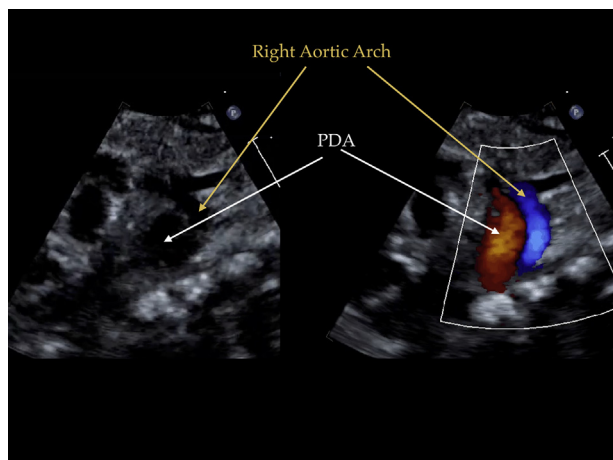
Echocardiographic assessment of double aortic arch is performed in the suprasternal notch and high parasternal views. In the case of a double aortic arch, a transverse sweep from inferior to superior can demonstrate the ascending aorta in cross-section, giving rise to both a right aortic arch and left aortic arch encircling the echogenic and bright trachea (Video 6). The sweep can also demonstrate the equidistant origins of the common carotid and then subclavian



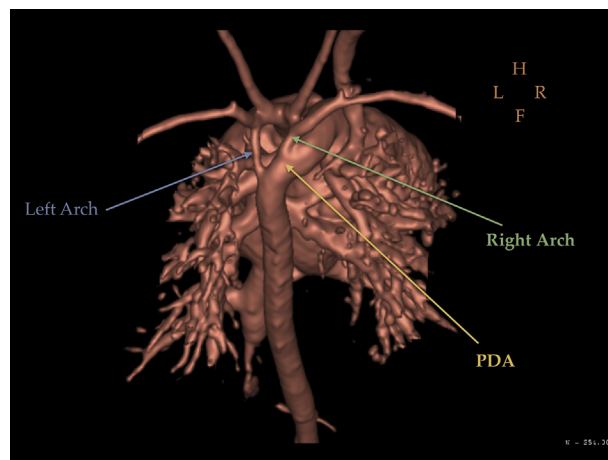
**Figure 4** Suprasternal notch long-axis view of the severely hypoplastic left arch.



**Figure 6** Cardiac magnetic resonance axial image of both aortic arches encircling the trachea, as well as the right-sided ductus. *PDA*, Patent ductus arteriosus.



**Figure 5** Suprasternal notch long-axis image of the severely hypoplastic right-sided aortic arch entering into the right-sided ductus. *PDA*, Patent ductus arteriosus.



**Figure 7** Cardiac magnetic resonance three-dimensional reconstruction demonstrating critical hypoplasia and long segment coarctation of both aortic arches entering into the ductus.

arteries arising from their respective ipsilateral aortic arches. A long-axis view of each aortic arch should be demonstrated to define its caliber from the ascending to transverse to descending aorta and evaluate for arch hypoplasia or coarctation. In double aortic arch, the larger dominant aortic arch needs to be identified. The nondominant arch and ligamentum arteriosum are typically ligated and divided to release the vascular ring. In our case, the long-axis view of both the right- and left-sided arches demonstrated aortic arch coarctation (Figure 4 and 5).

Although echocardiography can define aortic arch anatomy and evaluate for intracardiac anomalies, the inherent limitations of ultrasound in the evaluation of a vascular ring must be acknowledged. Visualization of posterior vascular structures can be difficult, for example, in cases in which there may be a patent posterior limb. It is important to note that in the unique case of a double aortic arch with an atretic left aortic arch distal to the left subclavian artery, the echocardiographic findings may appear similar to a right aortic arch with mirror-image branching. In these cases, supplemental imaging with cardiac computed tomography or MRI can be helpful to demonstrate the posterior limb of the atretic left-sided arch that completes

the vascular ring. In addition, echocardiography cannot define the degree of tracheal or esophageal compression to prognosticate the clinical manifestations of a vascular ring. Therefore, supplemental imaging with either cardiac computed tomography or MRI should be obtained to confirm the diagnosis as well as aid in surgical planning. In our case, advanced imaging by cardiac MRI confirmed the unique finding of double aortic arch with bilateral coarctation (Figures 6 and 7, Video 8).

Symptoms in patients with double aortic arch are more common than with other types of vascular rings. Tracheal narrowing and compression can cause stridor, dyspnea, and/or frequent respiratory infections. Esophageal compression can cause swallowing and feeding difficulties, with dysphagia or vomiting. Some patients may therefore be detected by chest radiography or bronchoscopy (obtained to evaluate respiratory symptoms) or barium swallow studies (obtained to evaluate feeding difficulties). In these cases, echocardiography should be considered as the next step in diagnosis, as an efficient, noninvasive modality to confirm a vascular ring due to double aortic arch as well as other variations of a vascular ring or sling.

Surgical repair involves division of the smaller arch, which is usually the left arch, as well as ligation and division of the ductus arteriosus (or ligamentum arteriosum). Typically, this is done through a thoracotomy on the side of the nondominant arch.<sup>9</sup> The anatomy of both aortic arches and ductal anatomy therefore must be critically assessed and understood before surgical repair to avoid catastrophic results. Short- and long-term outcomes of isolated double aortic arch repair are excellent, with high rates of survival and low rates of reintervention.<sup>2</sup>

Only a small percentage of patients with double aortic arch (5%–18%) have associated intracardiac lesions; when present, tetralogy of Fallot is the most common intracardiac conotruncal anomaly.<sup>4,5,8,9</sup> We report a rare case of a double aortic arch with bilateral aortic coarctation in combination with extremely complex cardiac anatomy. The combination of airway obstruction from the vascular ring, pulmonary overcirculation, and ventricular outflow obstruction in the context of a functional single ventricle was unfortunately a lethal physiologic constellation in our patient.

## CONCLUSION

Although double aortic arch typically presents with a dominant right arch and hypoplastic left arch, bilateral aortic coarctation can be present, and therefore each arch should be critically assessed using echocardiography. Advanced multimodality cardiac imaging should be adjunctive to confirm the echocardiographic findings and to delineate anatomic relationships with the trachea. This arrangement should be considered as a differential diagnosis when dealing with patients diagnosed with double aortic arch and in patients diagnosed with complex intracardiac disease.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2020.02.001>.

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