

# Adolescent Tracheobronchomalacia

## Double Aortic Arches Revisited

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**T**he double aortic arch (DAA) is the most common vascular ring disorder recognized clinically.<sup>1,2</sup> Most cases are diagnosed in the neonatal population, with respiratory and gastroesophageal symptoms predominating. Several modalities have been used to identify this vascular anomaly, and diagnostic methods vary among institutions.<sup>1-4</sup> Direct endoscopic evaluation is a diagnostic tool available to clinicians, but imaging also has been shown to be an effective method for recognizing the DAA.<sup>4</sup>

A small fraction of patients born with this embryologic anomaly evade diagnosis into adolescence or adulthood. Similar to patients whose conditions are diagnosed during childhood, older patients who are symptomatic also require surgical correction of the aberrant vasculature. We recently encountered a patient with a DAA diagnosed at age 15 years after multiple years of treatment for pulmonary and gastroesophageal symptoms. We present the clinical and radiologic features of this patient's vascular anomaly and discuss the utility of multiple diagnostic approaches used to identify the DAA.

### REPORT OF A CASE

A 15-year-old girl sought medical attention for respiratory distress, including stridor and wheezing. The patient had a longstanding history of asthma exacerbations and refractory gastroesophageal reflux. Multiple medication regimens had been attempted for control of symptoms but met with little success. The patient had been evaluated several times with esophagoduodenoscopy and treated for observed gastritis.

Flexible laryngoscopy demonstrated symmetrically mobile vocal cords. Chest radiography demonstrated an opacity at the

level of the thoracic inlet, indenting the right contour of the trachea (**Figure 1**). Magnetic resonance imaging (MRI) demonstrated a DAA, with the right arch lying slightly higher than the left. Both arches measured approximately 14 mm in diameter (**Figure 2**).

The patient underwent surgical division of the DAA and ligamentum arteriosum. Direct laryngobrochoscopy was performed intraoperatively before cardiac repair. The arches had caused extrinsic tracheal and bronchial compression. No concomitant lesions were found. The patient did well postoperatively, and her medication intake was reduced.

### COMMENT

Various classification systems have been proposed in an attempt to describe aortic arch anomalies. Possibly the most simplified approach, suggested by Achiron et al,<sup>1</sup> is based on 3 variations of the aortic arch: right, left, and double. Although a right aortic arch is the most common anomaly (0.1% incidence reported in the adult population), DAA is the most common form of vascular ring disorder recognized clinically.<sup>1,2</sup> Double aortic arch results from the failure of the right dorsal aorta to regress embryologically, so that the resultant arch completely surrounds the trachea and esophagus.<sup>5,6</sup> Various levels of compression of each structure can

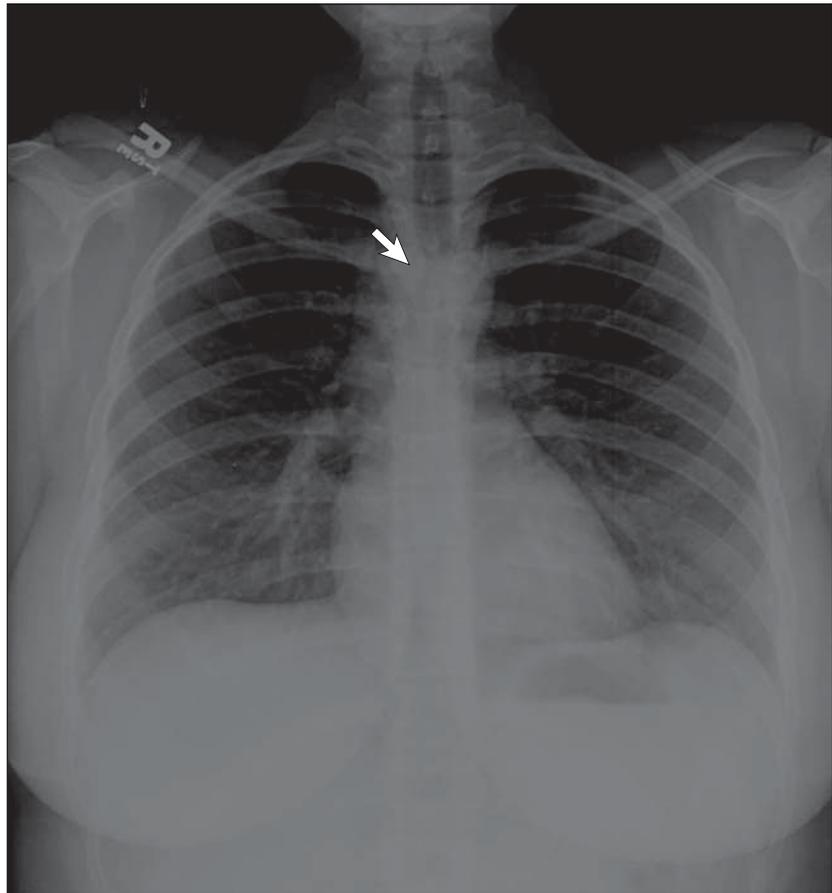
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be seen; not surprisingly, presenting symptoms are often respiratory or gastroesophageal in nature.<sup>2,3</sup>

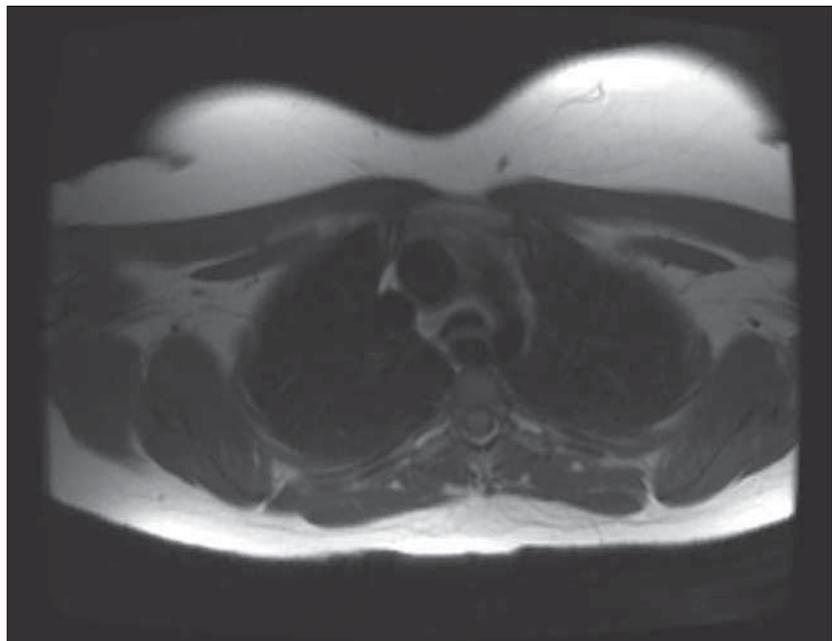
Due to the compressive features of DAA, most patients present in early infancy.<sup>1</sup> In a study by Alsenaidi et al,<sup>2</sup> all patients with DAA (n=81) were symptomatic before age 3 years, and 91% of patients had respiratory symptoms. Gastroesophageal symptoms were also common, and 40% of patients had issues such as choking and vomiting.

Multiple modalities have been used to diagnose DAA. Plain chest radiography, computed tomography, MRI, laryngobronchoscopy, tracheobronchography, and prenatal Doppler ultrasonography have all been used.<sup>1-4</sup> In most institutions, patients with respiratory symptoms are evaluated with an initial chest radiograph. Although many false-negative results are obtained, it has been suggested that specific radiographic findings can be highly indicative of DAA. Pickhardt et al<sup>4</sup> retrospectively evaluated the preoperative chest radiographs of 41 children with surgically proven vascular rings. Specific findings from this study included increased retrotracheal opacity (79%), tracheal bowing (92%), and tracheal narrowing (77%). Lateral radiographs were found to be slightly more reliable than frontal radiographs in the diagnosis of vascular rings. However, the variability between the 2 methods was not statistically significant. Pickhardt et al state that, in their study, no symptomatic patients with vascular rings had normal chest radiographs; therefore, they consider chest radiography to be a useful screening tool for diagnosing vascular rings in symptomatic patients.

Computed tomography and MRI are also valuable modalities by which aortic anomalies can be diagnosed. They are efficient and noninvasive. Although distal airway malacia has been shown to be more difficult to diagnose with dynamic computed tomography than with imaging studies such as bronchography,<sup>3</sup> the false-negative rate for vascular rings is nearly nonexistent, and unsuspected vascular rings are often detected.<sup>1,2</sup> One disadvantage of these imaging techniques is that in patients who require respiratory sup-



**Figure 1.** Chest radiography demonstrates an opacity at the level of the thoracic inlet, indenting the right contour of the trachea (arrow).



**Figure 2.** Axial magnetic resonance imaging reveals a double aortic arch with tracheal compression.

port, endotracheal intubation can increase the degree of airway patency by acting as a stent, resulting in an

underestimation of tracheal compression. A second disadvantage of MRI alone is the time required to

complete it and the possibility that sedation will be required.<sup>7</sup>

Direct bronchoscopy has been used to determine the presence of vascular rings and is an effective mechanism by which tracheobronchomalacia secondary to compression can be diagnosed.<sup>8</sup> However, bronchoscopy carries with it the limitations that are associated with general anesthesia and may be difficult in the neonatal population with borderline lung reserve and small airway diameter, as suggested by Mok et al.<sup>3</sup> In addition, the degree of airway compression and the diagnosis of tracheobronchomalacia require spontaneous respiration. This allows expiratory airway collapse to be visualized by bronchoscopy without the influence of positive airway pressure. Other limitations such as splinting of the lesion with the bronchoscope have been described as contributing to increased false-negative results in this population.<sup>3</sup> In addition, when a vascular anomaly is recognized by bronchoscopy, further diagnostic imaging is often required preoperatively to evaluate the aberrant vasculature in more detail.

In a series of patients with vascular rings (n=64), Shah et al<sup>8</sup> reported preoperative use of echocardiography (96%), chest radiography (93%), barium swallow (75%), MRI (60%), and computed tomography (59%). Direct laryngobronchoscopy was performed in only 20% of patients. In our patient, chest radiography and a confirmatory MRI were the diagnostic tools used. Several other authors have indicated that routine evaluation of patients with vascular rings does not include direct laryngobronchoscopy.<sup>9,10</sup>

Barium swallow, echocardiography, and chest radiography may be sufficient for initial evaluation of a patient with a suspected vascular ring. Although direct laryngobronchoscopy may not be necessary for the diagnosis of a DAA, we believe that it should be performed during the treatment of a patient with a vascular ring. Preoperatively, it can show the presence of a concomitant airway lesion and demonstrate the degree of vocal cord mobility. In patients with congenital cardiac anomalies, there is an increased incidence of synchronous airway le-

sions.<sup>10</sup> In addition, airway endoscopy can serve as a preoperative baseline for the patient. Imaging modalities such as chest radiography are effective in the symptomatic population, but patients who are asymptomatic or those who have lesser degrees of airway compression will benefit from direct visualization via bronchoscopy. This information can contribute to decisions about which patients may ultimately require surgical intervention.<sup>4</sup> The degree of secondary tracheobronchomalacia can be assessed. Furthermore, there is a risk of iatrogenic vocal cord paresis during vascular ring repair. Finally, bronchoscopy has been used intraoperatively, during ring repair, to visualize tracheal release.<sup>7</sup>

Due to the embryologic variability of DAA, a minority of these patients reach late adolescence or adulthood without a formal diagnosis. Although most patients with a DAA have a history of mild symptoms, some patients are entirely asymptomatic and require no treatment. Kajikawa et al<sup>11</sup> described an asymptomatic 64-year-old man with a DAA diagnosed after chest radiography demonstrated a widened mediastinum. Although the patient received conservative treatment, symptomatic patients require surgical correction of the vascular anomaly. Retrospectively, the patient described in this report had a long history of symptoms that, although nonspecific, are classically associated with the presence of a DAA. The prevalence of adolescent and adult patients with this vascular anomaly remains low. However, it is a diagnosis that should be considered in a patient with refractory respiratory and gastroesophageal symptoms.

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