

An Uncommon Cause of Wheeze

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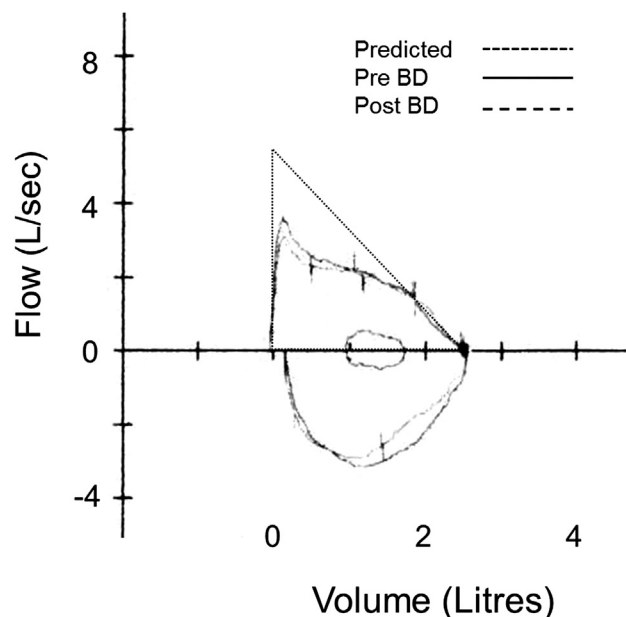


FIGURE 1. Flow volume loop, demonstrating flattening of the expiratory limb.

An 11-year-old girl was referred for the assessment of a mild wheeze on exertion. She had an intermittent cough and wheezing episodes associated with both exercise and upper respiratory tract infections. In addition, there was a description of episodic inspiratory stridor particularly when the patient had an upper respiratory tract infection. At birth, there was a history of “extremely vigorous” suctioning, which was thought to have led to laryngotracheomalacia as a cause of her symptoms. However,

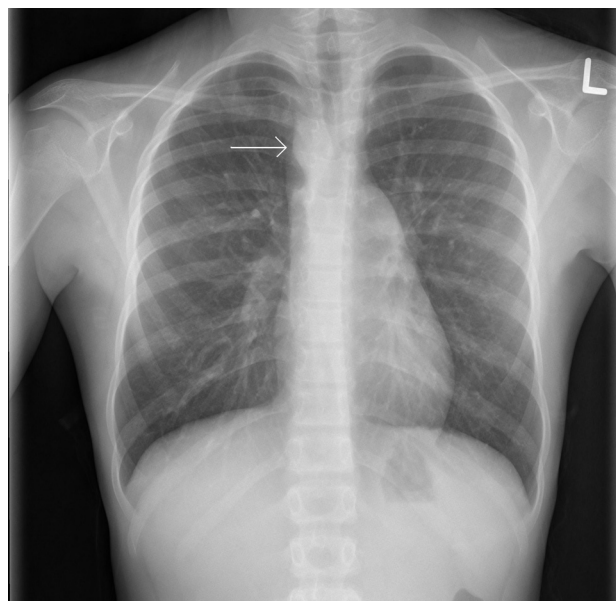


FIGURE 2. Chest radiograph, revealing a right-sided aortic arch (arrow) and the absence of a left aortic knob.



FIGURE 3. Coronal reformatted image of the computed tomography of the chest, revealing a right-sided aortic arch (Ao) with a vestigial left aortic arch (A).

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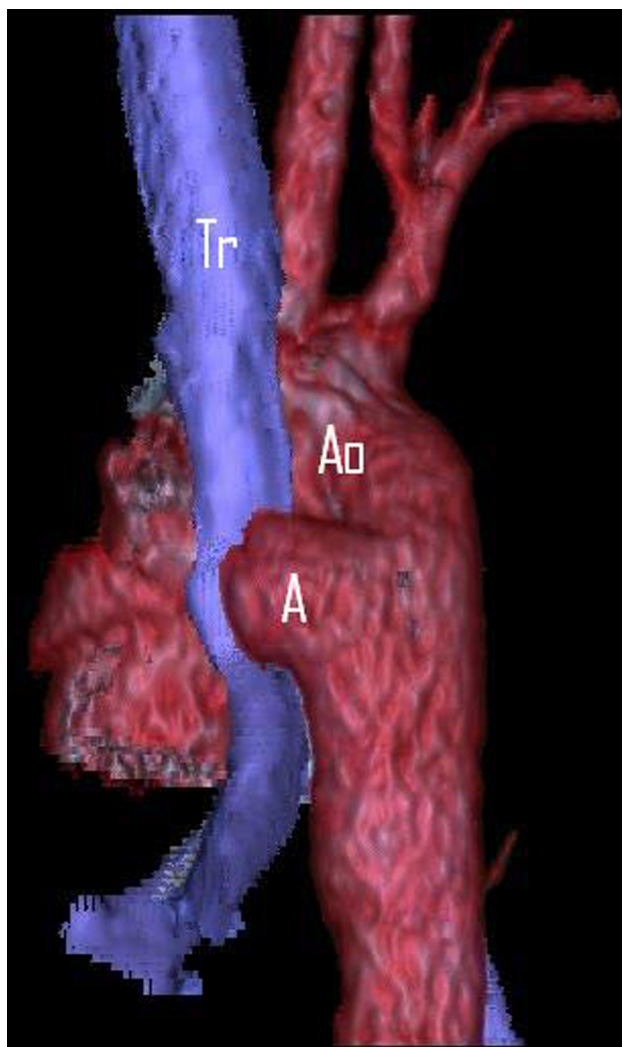


FIGURE 4. Left anterior oblique view of a 3-dimensional volume-rendered image with trachea (Tr), aorta (Ao), and vestigial left arch (A).

given her episodic wheezing and prolonged cough with upper respiratory tract infections, there also was a concern for asthma. Thus, she was initially treated with salbutamol on an as-needed basis followed by a trial of inhaled corticosteroids. The inhaled corticosteroids were of no clinical benefit, and the salbutamol did not completely resolve the patient's symptoms.

Spirometry showed flattening of the expiratory limb (Figure 1), which suggests a variable intrathoracic obstruction. A subsequent chest radiograph revealed a right-sided aortic arch (Figure 2), and thus a computed tomography of the chest was recommended to further characterize the great vessels. The scan revealed a right-sided aortic arch with a vestigial left-sided aortic arch (Figures 3,4) and secondary tracheal compression with leftward shift of the trachea (Figure 5). The finding of tracheal compression secondary to a right-sided aortic arch was the most likely explanation for this patient's symptoms. She subsequently



FIGURE 5. Axial contrast-enhanced computed tomography image of the thorax, showing the ascending (*) and descending (**) portions of the right-sided aortic arch. The vestigial portion of the left arch (A) leads to trachea compression with a left shift.

was referred to a pediatric cardiologist who, after a thorough assessment, decided that corrective vascular surgery for this patient was not warranted given the risks involved. The patient continued to have mild, intermittent wheezing on exertion and during exercise but over the course of the following year improved significantly even in the absence of pharmacotherapy or surgical intervention.

Under normal vascular development, the aortic arch passes in front of and to the left of the trachea.¹ However, in this patient, the arch passes to the right and behind the trachea. As the aorta crosses anterior to the vertebral bodies and posterior to the trachea, it may (and in this case did) lead to tracheal compression.² Although it occurs in approximately 0.1% of the population, this congenital malformation is one of the most common major malformations of the great vessels,^{3,4} and is due to the aortic arch developing from the right (instead of the left) fourth branchial arch.¹ Patients with this malformation usually are asymptomatic but do rarely develop inspiratory stridor, expiratory wheezing, and dyspnea.⁵ Consultation to cardiology or vascular surgery is warranted if the patient is symptomatic; however, surgical intervention is not considered prudent unless significant symptoms occur. This case was documented to highlight the point that other etiologies for wheezing besides asthma must be considered when symptoms are not convincing for asthma or when patients do not follow the normal response to therapy.

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