

## Late Diagnosis of Double Aortic Arch: Consequences on Long-Term Follow-Up

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**Summary.** Double aortic arch is the most common congenital anomaly of the aortic arch system, in which the trachea and esophagus are completely encircled by vascular segments of the aortic arch and its branches, often resulting in variable airway compression. One case of late diagnosis of this congenital malformation and long-term consequences of late surgical treatment with persistent tracheo-bronchomalacia and dynamic airway obstruction is reported. This report emphasizes the importance of an early diagnosis to minimise the progressive airways damage and subsequent respiratory symptoms, that need an accurate medical follow-up. **Pediatr Pulmonol.** 2014; 49:E75–E77. © 2013 Wiley Periodicals, Inc.

**Key words:** aortic arch; bronchomalacia; 4D CT scan; dyspnoea.

**Funding source:** none reported.

### CASE REPORT

DG, a 15-year-old boy, suffered from respiratory distress since the first week of life, associated to stridor. He was treated with anti asthmatic therapy due to the presence of recurrent episodes of “bronchial asthma”, recurrent cough along with a polysensitivity to aero-allergens. As a consequence of poor response to standard asthma treatments, a pH-impedanzometry test was performed and gastro-esophageal reflux (GER) was diagnosed and treated with PPI. After few years, despite conventional asthma treatment, DG still presented respiratory symptoms and further investigations were performed. Lung function test suggested significant intra-thoracic middle-airways compression with a forced vital capacity (FVC) of 70%, a forced expiratory volume in 1 sec (FEV1) of 67% and a FEV1/FVC ratio of 84%. Maximal expiratory and inspiratory flow-volume loop showed symmetric reduction of both maximal expiratory and inspiratory flows, a pattern that was suggestive of

a variable intra-thoracic obstruction. Finally a complete double aortic arch was diagnosed by CT scan after intravenous contrast medium injection at 7 years of age and surgically treated with dramatic clinical improvement and resolution of all symptoms.

Eight years after the operation, DG was completely symptoms-free with the exception of an increasing exercise induced dyspnoea not responding to bronchodilator treatment. A new assessment was planned including lung function test that showed a significant improvement of lung capacity (FVC of 147%, FEV1 of 142%, and a FEV1/FVC ratio of 83%) but still presenting a characteristic “box shape” during forced expiratory/inspiratory flow. An airway endoscopy and a dynamic chest CT scan with 4D dynamic reconstruction in free breathing were performed and revealed persistence of a footprint aortic arch on the lower third of the trachea without significant lumen stenosis. Interestingly, images acquired during end inspiration and forced expiratory phase confirmed the tracheal footprint at inspiratory phase (Image A, big

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Received 17 April 2013; Revised 2 June 2013; Accepted 2 June 2013.

DOI 10.1002/ppul.22852

Published online 16 July 2013 in Wiley Online Library  
(wileyonlinelibrary.com).

arrow) that did not significantly modified at forced expiratory phase (Image B, big arrow). Importantly though, at forced expiration, significant reduction in size of the right main bronchus (Image B, upper small arrow), and, at a lower degree, of the left main bronchus (Image B, lower small arrow) was revealed. These findings suggested a condition of persisting bronchomalacia and clearly explained the continuous exercise-induced symptoms.

## DISCUSSION

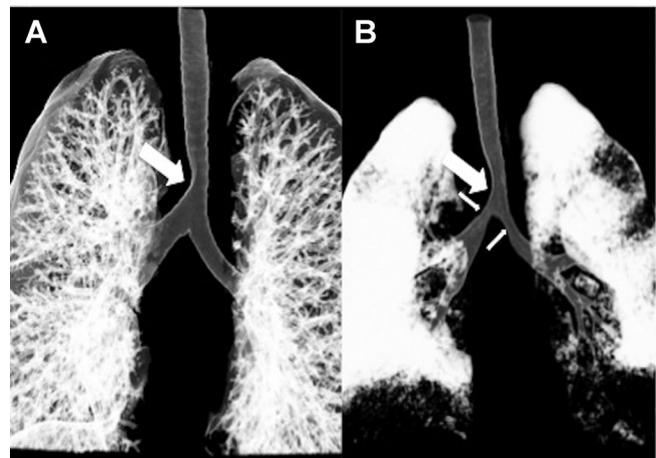
This report describes a late diagnosis of a double aortic arch and its consequences on a long-term follow-up. The literature shows a wide prevalence of this congenital vascular variation that encircles and compresses the trachea and/or the esophagus. Patients clinically display symptoms in the neonatal period or later in childhood, depending on the severity of the compression. According to the literature, respiratory symptoms appear to be more frequent than gastrointestinal symptoms of which choking with feeds is the most common.<sup>1</sup> Congenital stridor, a sign of obstruction located in the airway, is usually the clinical manifestation of laryngeal, tracheal, bronchial, or vascular abnormalities, and rarely the result of other conditions, such as infections or trauma.<sup>2</sup> Unfortunately incorrect or incomplete diagnosis is quite common particularly when congenital abnormality is associated with “asthma-like” respiratory symptoms in allergic patients. However, when asthma is unresponsive to treatment, other diagnosis should be considered, and more detailed examinations performed. GER as non-congenital comorbidity was detected in our patient and, together with a concomitant allergic sensitisation, it may have contributed to the delayed diagnosis.

Eight years after the surgical treatment DG presented an increasing exercise induced dyspnoea unresponsive to bronchodilator treatment. The lung function test was useful to confirm the clinical suspect of persisting airways-malacia. For a more specific definition, two different CT methods were adopted: end-expiratory imaging (at suspended end expiration) and dynamic expiratory imaging with reconstructions (in free breathing and during end inspiration and forced expiratory phase of respiration).<sup>3</sup> The recent advent of faster multi-detector row CT scanners, which can image the entire central airways in few seconds, has facilitated the ability to perform dynamic expiratory imaging.<sup>4</sup> It is known that positive pleural (intra-thoracic) pressure worsens the dynamic collapse of the airway in patients with trachea-bronchomalacia (TBM). Forced dynamic expiration produces a higher level of intra-thoracic/extra-tracheal pressure than does end-expiration or free breathing and can identify a greater degree of central airway collapse.<sup>5,6</sup> In our patient in fact, dynamic forced expiratory

acquisition identified airway lumen reduction at the level of the right main bronchus and the carina (Fig. 1A and B). Moreover, the same exam confirmed a persistent footprint on the lower third of the tracheal wall. Our experience suggests that dynamic forced expiratory imaging (compare to end-expiratory imaging or dynamic imaging during free breathing) in specific cases can be a useful tool to define in details TBM and clarify persisting respiratory symptoms.

Surgical treatment of double aortic arch eradicates symptoms in over 70% of patients although airflow limitation may persist. The persistence of symptoms postoperatively can be attributed to many reasons: residual tracheal compression, TBM, and tracheal stenosis, which are due to maldevelopment of trachea.<sup>7</sup> A delayed diagnosis with a late surgical treatment determines a bigger chance of residual TBM due to long-term tracheal compression. The consequent damage of the tracheal rings causes a localized area of tracheomalacia.<sup>1</sup> In fact, after an initial dramatic benefit the respiratory symptoms might rise again particularly related to the increasing physical activities during adolescence. Therefore an early diagnosis and surgical correction is crucial to obtain a better long-term outcome.

In conclusion, this report emphasizes the importance for pediatricians of a detailed knowledge of the most typical clinical presentations of such vascular variation, which is critical for an early diagnosis. Infants and young children with early stridor and persistent respiratory symptoms should be always investigated for the possibility of having vascular rings. Moreover, those



**Fig. 1.** A: 3D airway reconstruction acquired during end inspiration demonstrating, footprint aortic arch on the lower right wall of the trachea with slight reduction in size (big arrow). B: 3D airway reconstruction acquired during forced expiratory phase confirming footprint aortic arch on the right tracheal wall (big arrow) and significant reduction in size at forced expiration of the right main bronchus (upper small arrow) and, at a lower degree, of the left main bronchus (lower small arrows) suggesting bronchomalacia.

children with a late surgical treatment of such congenital abnormality need a close and longer medical follow-up. A dynamic forced expiratory imaging with reconstructions could be useful to exclude TBM in case of persistent respiratory symptoms and in rare cases may lead to reoperation.

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