Double aortic arch: a cause of stridor to remember

Vera Baptista,1 Inês Azevedo,2 Gisela Rio,3 Carla Moreira1

Department of Pediatrics, Hospital de Braga, Braga, Portugal
2Department of Pediatrics, Faculty of Medicine, Centro Hospitalar S João, Oporto, Portugal
3Department of Radiology, Hospital de Braga, Braga, Portugal

Correspondence to Dr Vera Baptista, verabaptista1@sapo.pt

DESCRIPTION
A previously healthy baby boy was admitted several times to the paediatric emergency department, from the age of 2 months, presenting respiratory distress, cough and wheezing. The episodes were managed as viral respiratory infections and inhaled bronchodilators, oral corticosteroids and chest physiotherapy were prescribed. At 6 months of age, during the third hospitalisation, a biphasic stridor was noted. The nasolaryngofibroscopy initially performed was unremarkable. At 7 months, a fibreoptic bronchoscopy showed a pulsatile tracheal compression, suggestive of a vascular ring. An MRI study showed a double aortic arch, confirming the clinical suspicion (figures 1 and 2).

Respiratory distress in children may or may not occur due to processes involving the respiratory tract.1 Many children are affected by recurrent lower respiratory tract infections and this is a frequent cause for urgent medical consultation. The challenge is to suspect and find the rare cases that might have an underlying disease.2 Stridor is a sign that suggests a significant obstruction of the large airways.

Vascular rings are rare congenital anomalies due to abnormal development of the aortic arch. In a double aortic arch, the ascending aorta bifurcates anteriorly to the trachea and oesophagus, completely encircling them. The arches then rejoin into a single descending posterior aorta. Vascular rings may cause a compression of the tracheobronchial tree and oesophagus, leading to respiratory infections, respiratory distress, stridor, wheezing, cough, dysphagia and feeding difficulties. The only definitive treatment is surgery.3

Learning points
▸ Many children are affected by recurrent lower respiratory tract infections; it is a challenge to find those cases that might have underlying lung or extrapulmonary disease.
▸ The clinician must have a high degree of suspicion in order to diagnose a vascular ring, as it is a relatively uncommon cause of respiratory distress.

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REFERENCES