

Clinical Pearl

Vascular ring “mimickring” asthma in a preschool girl with combined immunodeficiency: the benefits of lung function measurements at diagnosis and postoperative follow-up



Vascular rings are congenital anomalies of the aortic arch that result in compression of the tracheobronchial tree and/or esophagus, leading to respiratory and gastrointestinal symptoms. They can be classified as complete, when both the trachea and esophagus are fully encircled by a vascular anomaly, or incomplete, without full encirclement of both structures. Most children present in early infancy with nonspecific symptoms of dyspnea, cough, inspiratory or biphasic stridor, and sometimes an expiratory wheeze and feeding problems.¹ Only a few patients present much later.^{2,3}

Clinical Presentation

A 9-year-old girl was admitted to the pulmonology department for persistent symptoms of dyspnea, reduced exercise tolerance, and “noisy breathing” during the past 3.5 years. She was the third child of nonconsanguineous parents with 2 older healthy brothers. She was born small for gestation age, and her medical history revealed mild to moderate developmental delay during early childhood. She had a history of recurrent respiratory infections, wheezing episodes, and cough since early childhood. At the age of 5 years, she was evaluated in the pulmonology department and was diagnosed as having asthma. Despite treatment with fluticasone-salmeterol, she had recurrent wheezing episodes. At the age of 5.5 years, during an influenza B infection, she had acute necrotizing encephalopathy that presented with convulsions, vomiting, hepatic dysfunction, and coma. Treatment required endotracheal intubation and mechanical ventilation for 19 days. On discharge, she had neurologic sequelae that consisted of speech impairment, spasticity, left-sided hemiparesis, and intellectual disability. In addition, she had biphasic stridor that was appreciated as a postintubation stridor.

Supporting Data

At hospital admission, she was not in acute distress, was overweight (body mass index [a measure of weight in kilograms divided by height in meters squared], 27.55), and was dyspneic at rest with biphasic stridor. Her respiratory rate was 24/min, pulse was 80/min, oxygen saturation was 97%, and body temperature was 36.7°C. Wheezing, harder and longer expiratory phase, bilateral crepitant rales, and diffuse rhonchi were heard on auscultation of the chest. Other examination findings were normal (no

dysmorphism, no dysphagia, no signs of hypocalcemia). Body plethysmography revealed the following: forced vital capacity (FVC), 1.35 L (67%); forced expiratory volume in 1 second (FEV₁), 1.11 L (68%); FEV₁/FVC, 76; maximum expiratory flow at 50% of FVC, 1.29 L/s (51%); peak expiratory flow (PEF), 1.69 L/s (42%); total airway resistance, 0.7 kPas/L; (110%), expiratory airway resistance, 0.99 kPas/L (156%); total airway conductance, 1.43 (55%); residual volume, 0.75 L (105%); total lung capacity, 2.2 L (81%); post-bronchodilator study FEV₁, 0.93 L (decrease of 16%); and post-bronchodilator study FVC, 1.17 L (decrease of 13%) (Fig 1). The flow-volume curve was noted to have plateaus on both the inspiratory and expiratory limbs. Chest radiography revealed right aortic arch and tracheal stenosis. Barium contrast esophagography (barium swallow) revealed a posterior indentation of the esophagus. Computed tomography angiography of the chest revealed a vascular ring, which consisted of a right aortic arch with Kommerell diverticulum at the anomalous origin of the left subclavian artery left-sided ductus arteriosus (Fig 2). Bronchoscopy revealed tracheal dyskinesia. In addition, a small thymus was also visible on computed tomography. Immunologic workup revealed combined immunodeficiency (pan lymphopenia with hypogammaglobulinemia).

Because of immunodeficiency and conotruncal anomaly, Di George syndrome was suspected, but the results of fluorescence in situ hybridization analysis for microdeletions 22q11, 22q13, and 10p14 were negative. Surgical division of Kommerell diverticulum and left subclavian artery transfer was performed. Her breathing significantly improved (she was not dyspneic at rest, and her exercise tolerance improved) during the follow-up period of 6 months after the procedure. The PEF measurements revealed improvement from 1.69 L/s before the procedure to 3.27 L/s after the procedure, with persistent abnormality of the expiratory limb of the flow-volume curve. Immune status improved.

Clinical Outcome

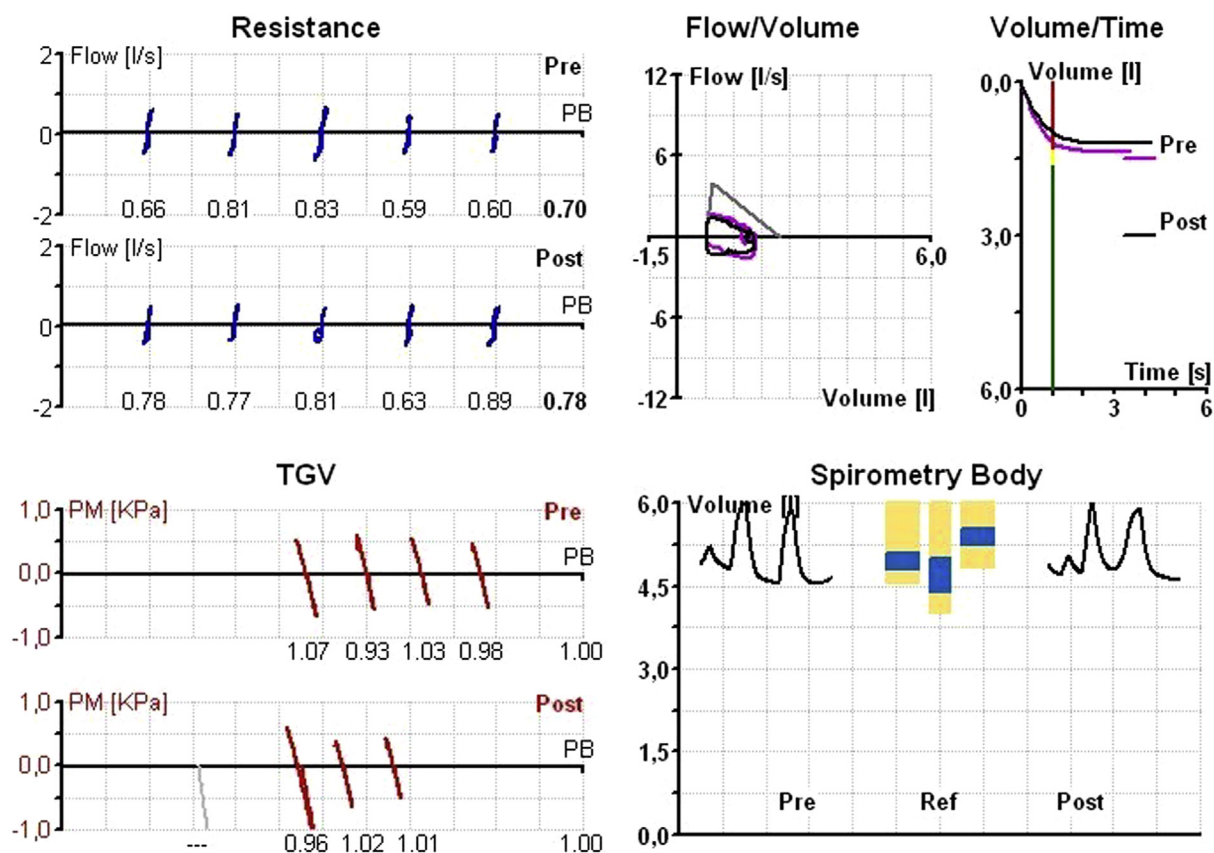
We describe a 9-year-old girl diagnosed as having a vascular ring. She was given a diagnosis of asthma but experienced ongoing respiratory symptoms despite asthma therapy. Abnormalities of the shape of the flow-volume curve prompted further investigations to evaluate for central airways obstruction, revealing the true diagnosis of vascular ring.

Stridor is a loud, harsh, high-pitched respiratory sound caused by abnormal air passage into the lungs. Inspiratory stridor suggests a laryngeal obstruction, whereas expiratory stridor implies tracheobronchial obstruction. Biphasic stridor from birth or early

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	Pred.	Pre	Pre/pred.	Post	Diff.
IVC-B	2,00	1,35	67%	1,17	-13%
IRV-B	-	0,93	-	0,49	-47%
ERV-B	-	0,25	-	0,42	63%
VT-B	-	0,28	-	0,27	-3%
FVCex	2,00	1,35	67%	1,17	-13%
FEV1	1,65	1,11	68%	0,93	-16%
FEV1/VCmax	85	76	90%	71	-6%
MEF25	1,31	0,79	60%	0,58	-27%
MEF50	2,55	1,29	51%	0,99	-24%
MEF75	3,62	1,51	42%	1,30	-14%
PEF	4,01	1,69	42%	1,42	-16%
RAWtot	0,63	0,70	110%	0,78	11%
RAW-ex	0,63	0,99	156%	1,13	15%
Gtot	2,60	1,43	55%	1,29	-10%
TLC-B	2,71	2,20	81%	1,75	-20%
TGV-B	1,33	1,00	75%	1,00	0%
RV-B	0,71	0,75	105%	0,58	-22%
RV/TLC-B	26	34	132%	33	-2%
TGV/TLC-B	-	45	-	57	25%

Figure 1. Body plethysmography revealing decreased forced expiratory volume in 1 second and peak expiratory flow measurements and abnormally shaped flow-volume curve.

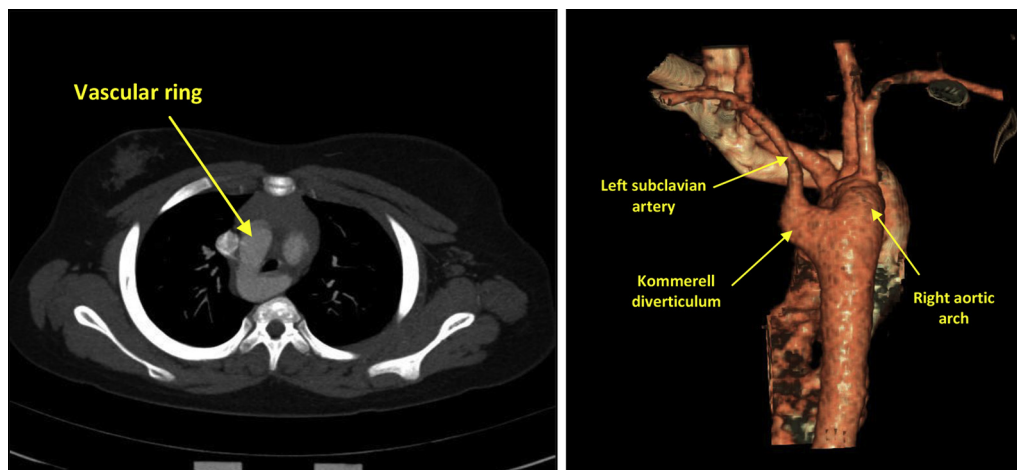


Figure 2. Computed tomography angiogram of the chest showing a Vascular ring, consisting of a right aortic arch with Kommerell diverticulum at the anomalous origin of the left subclavian artery left-sided ductus arteriosus.

infancy is caused by fixed proximal airway obstruction, which can be intrathoracic (tracheobronchial tree) or extrathoracic (nose, pharynx, larynx, trachea). Patients with biphasic stridor are often initially misdiagnosed as having asthma because of noisy respirations, although stridor is never a sign of asthma but rather of proximal airway compromise.³ Stridor differs from wheeze in that it has a different pitch and harsher sound than the more musical pitch of a wheeze.⁴ Although our patient had respiratory symptoms from early childhood, biphasic stridor was not noticed until 5.5 years, when it was misdiagnosed as a postintubation stridor.

The diagnosis of asthma in children requires a careful review of a child's current and past medical history, family history, and a physical examination, results of lung tests (spirometry with bronchodilator test and challenge testing), and finally a therapeutic trial of medications.⁵ It is of the utmost importance to properly interpret results of spirometry in children with chronic respiratory symptoms, especially the shape of flow-volume curve. Characteristic changes of the shape of the flow-volume curve should prompt physicians to consider a diagnosis of central airway obstruction from vascular rings or other causes.⁶ Typically, with a fixed intrathoracic tracheal obstruction, such as complete vascular ring, there is a plateau of both inspiratory and expiratory flows.⁷ With variable intrathoracic obstruction, as might be seen with incomplete vascular rings⁶ or tracheomalacia, a plateau is present on expiratory flow but lacks on inspiration.⁷

Assessment of the FEV₁ and PEF together with flow-volume curve tracings may be helpful in screening for central airway obstruction. In patients with central airway obstruction, PEF is significantly decreased, compared with FEV₁, and a value of FEV₁/PEF greater than 8.8 mL/L/min has been suggested as an indicator of this condition in children.⁸ This case highlights the value of lung

function measurements and the importance of careful flow-volume curve examination in making the right diagnosis of vascular ring in children.

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References

- [1] Bakker DA, Berger RM, Witsenburg M, et al. Vascular rings: a rare cause of common respiratory symptoms. *Acta Paediatr.* 1999;88:947–952.
- [2] Turner A, Gavel G, Coutts J. Vascular rings—presentation, investigation and outcome. *Eur J Pediatr.* 2005;164:266–270.
- [3] Sturm J, Knutsen AP. Vascular ring presenting as asthma in an 8-year-old. *Ann Allergy Asthma Immunol.* 2012;108:291–293.
- [4] Spencer S, Yeoh BH, Van Asperen PP, Fitzgerald DA. Biphasic stridor in infancy. *Med J Aust.* 2004;180:347–349.
- [5] Global Initiative for Asthma, National Heart, Lung and Blood Institute. *Global Strategy for Asthma Management and Prevention* 2006. Bethesda, MD: Global Initiative for Asthma, National Heart, Lung and Blood Institute, <http://www.ginasthma.org/Guidelineitem.asp>. Accessed December 29, 2007.
- [6] Parker JM, Cary-Freitas B, Berg BW. Symptomatic vascular rings in adulthood: an uncommon mimic of asthma. *J Asthma.* 2000;37:275–280.
- [7] Pellegrino R, Viegi G, Brusasco V, et al. Interpretative strategies for lung function tests. *Eur Respir J.* 2005;26:948–968.
- [8] Linna O, Hyrynkangas K, Lanning P, Nieminen P. Central airways stenosis in school-aged children: differential diagnosis from asthma. *Acta Paediatr.* 2002; 91:399–402.