

Letter

A 16-YEAR-OLD BOY WITH DELAYED GROWTH AND MODERATE PERSISTENT ASTHMA

To the Editor:

A 16-year-old boy, hitherto healthy except for severe anorexia, was referred for unresponsive bronchial asthma. At the age of 12 years, he was diagnosed as having partial growth hormone deficiency and received replacement therapy. At the age of 13 years, he started psychotherapy for anorexia. Exercise-induced asthma developed in his 15th spring with grass pollen allergy. Spirometry performed 2 months before admission revealed the following: forced vital capacity (FVC), 93%; forced expiratory volume in 1 second (FEV_1), 46%; FEV_1/FVC ratio, 45%; and forced expiratory flow between 25% and 75%, 27% of expected normal value.¹ These findings suggested severe asthma, partially responsive to bronchodilator (postbronchodilation FEV_1 increased by 17% of baseline¹). The results of thoracic x-ray radiography were unremarkable. Despite increasing doses of fluticasone propionate, 800 $\mu\text{g}/\text{d}$ or less, and montelukast, 10 $\mu\text{g}/\text{d}$, the boy developed daily cough, dyspnea, and relapsing episodes of fever and bronchitis. Because of this failure of asthma control, the patient (by then aged 16.7 years) was referred to our institution in December 2005. Weight and height were under the third percentile for his age. Initial examination revealed delayed puberty, stridor at normal tidal breathing, and expiratory wheeze and bilateral crackles at auscultation. The boy claimed not to be fit enough for sports. Skin prick testing confirmed hypersensitivity to grass, mites, and cat dander. Cough was present at rest and became paroxysmal after running. Basal spirometry confirmed severe dysventilation (Figure 1a), and the airway resistance loop obtained by body plethysmography was open (Figure 1c). Spirometry indicated severe dysventilation (FVC, 90%; FEV_1 , 38%; Tiffenau Index, 35%; and maximum expired flow rate at 50% [$MEF_{50\%}$], 17% of expected normal value). After salbutamol, 400 μg , the FVC increased to 97% and the FEV_1 to 40%, whereas the Tiffenau Index and $MEF_{50\%}$ remained unchanged. The midinspiratory flow at 50% ($MIF_{50\%}$) was 0.79 L/s, the ratio of forced inspiratory volume in 1 second to forced inspiratory vital capacity was 16%, and the $MEF_{50\%}/MIF_{50\%}$ ratio was 0.77. Normal lung volumes (total lung capacity, 109%; respiratory volume, 124%; ratio of total lung capacity to respiratory volume, 26%) with airway resistance of 325% of expected value and a reduction of specific airway conductance of 22% were obtained at body plethysmography (Figure 1c). These findings prompted morphologic evaluations. A supracarinal high-resolution computed tomographic scan showed severe compression of the tracheal lumen under

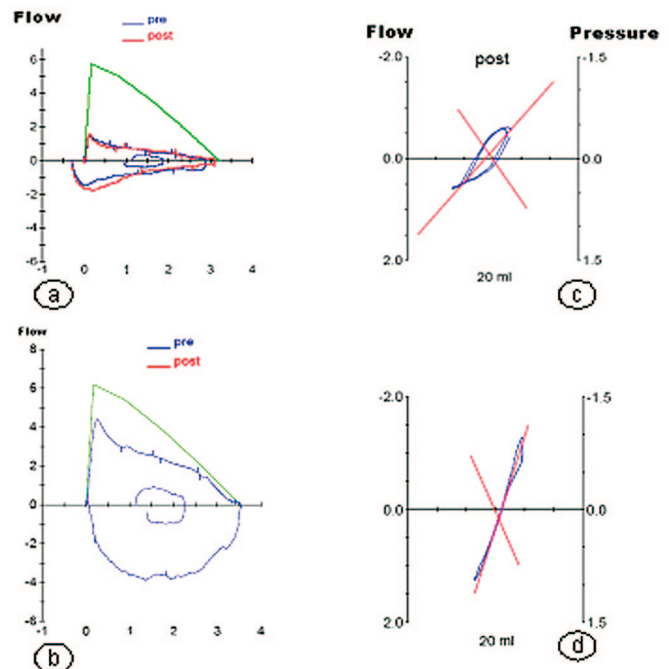


Figure 1. Pulmonary function tests: spirometry before (blue) and after (red) salbutamol, 400 μg , before (a) and 6 months after (b) surgery. Airway resistance and tidal breathing flow/pressure loop before (c) and after (d) surgery.

the pressure of the esophagus. The dilation of the thoracic esophagus, burdened with abundant food material, reached 4 cm in diameter. Dislocation of the trachea and extensive compression of the main bronchi were seen. Achalasia determining megaesophagus with tracheal compression was confirmed at barium esophagography. After laparoscopic Heller myotomy, dyspnea resolved immediately and cough disappeared in a week.

Spirometry was diagnostic in this case. A flattening of the expiratory part of the curve suggested a variable intrathoracic obstruction (eg, bronchial tumor or mediastinal disease). A flattened inspiratory part of the loop indicated variable extrathoracic obstruction (vocal cord dysfunction or paralysis). The fact that both of these conditions were present suggested fixed intrathoracic or extrathoracic obstruction, for which the most common causes are tracheal stenosis, tracheal tumor, and foreign body (Figure 1a). The same kind of curve flattening had already been observed with the tidal breathing flow/volume loop. At plethysmography, airway resistance findings further supported the presence of a severe obstruction. The resistance loops are remarkably open, indicating the presence of trapped air (Figure 1b). These findings are more suggestive of a proximal obstructive disease than of an

Authors have nothing to disclose.

asthma profile. After surgery, asthmatic symptoms disappeared and controller therapy could be discontinued in 6 months. Anorexia disappearance was clearly due to recovery from dysphagia. Spirometry at 1 and 6 months after surgery showed a progressive increase of both expiratory and inspiratory parameters: FEV₁, 67% and 88% of predicted; and MEF_{50%}, 53% and 63% of predicted (Figure 1b). The shape of the airways resistance loop normalized after a month with no further evidence of trapped air. Residual volume reverted to 93% of predicted after 3 months with a decrease in airway resistance (146% of predicted) and an increase in specific airway conductance (62%) (Figure 1d). Within a year the boy's height and weight normalized for his age. A computed tomographic scan at 9 months showed the esophagus with a clearly reduced lumen (maximum diameter, 2 cm), tapering in shape, and a minimal tracheal lumen of 1.2 cm.

Congenital achalasia due to a neuromuscular defect is rare (0.5 to 1 per 100,000 population per year), with a prevalence of approximately 8 per 100,000 population.² When not associated with more complex congenital malformations,³ the condition runs the risk of delayed diagnosis and/or misdiagnosis.⁴ In our case, routine chest x-ray examination did not reveal any pathologic findings, and pulmonary function test results were interpreted as indicative of asthma. This case extends the differential diagnosis of asthma to tracheal obstruction in adolescents, whereas larger airways obstructive conditions are recognized in the asthma workup of infants and children.⁵ We suggest that undiagnosed congenital conditions such as megaesophagus should also be ruled out and that spirometry is instrumental in revealing an unexpected diagnosis as in this hyposomic boy referred for asthma.

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