Radiographic studies (Figs 1–4) were obtained from a 60-year-old man with recurrent pneumonias for the past 15 years. He has chronic foul-smelling sputum, increasing shortness of breath, and increasing supplemental oxygen requirements. He reports a 50–pack-year history of tobacco use, but has not smoked for 12 years. In addition to bilateral crackles and rhonchi posteriorly, he exhibits digital clubbing. His FEV₁ is 1.52 L (48% of predicted); FVC, 2.4 L (61% of predicted); and FEV₁/FVC, 0.62. While he is receiving 5 L of oxygen by nasal cannula, his arterial blood gas analysis reveals a $P_{O_2}$ of 60 mm Hg, $P_{CO_2}$ of 41 mm Hg, and pH of 7.43. Sputum culture grows Pseudomonas species. Which one of the following statements concerning this case is not true?

A. A genetic linkage has been established in some cases.
B. The underlying pathologic finding is poorly formed or absent bronchial cartilage.
C. Nasal continuous positive airway pressure and airway stenting have been found to be effective.
D. Serum IgE levels are in the normal range.
E. Bronchial diverticula appear in some forms of this condition.

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Figure 2. Lateral chest radiograph showing tracheomegaly, hyperinflation, and cystic bronchiectasis with air fluid levels.

Figure 3. CT scan confirming tracheomegaly and bronchomegaly.

Figure 4. CT scan confirming tracheomegaly, bronchomegaly, and cystic bronchiectasis.
**Answer:** B. The underlying pathologic finding is poorly formed or absent bronchial cartilage.

This patient has tracheobronchomegaly or the Mounier-Kuhn syndrome, characterized by marked tracheal and bronchial dilatation sometimes associated with bizarre diverticula. Associated features include bronchiectasis and progressive obstructive lung disease with chronic purulent sputum production and recurrent pneumonias. Figures 1–4 demonstrate the tracheal and bronchial dilatation as well as cystic bronchiectasis. The transverse diameter of the trachea exceeds 29 mm; the transverse diameters of right and left mainstem bronchi exceed 26 mm and 23 mm, respectively. In normal subjects, the mean value for the trachea is 20 mm; for right and left mainstem bronchi, 14.5 each.

Histologically, patients with tracheobronchomegaly have severe atrophy of the longitudinal elastic fibers and thinning of the muscular layers of the major airways. This results in dilatation of both the membranous and cartilaginous portions of the large airways. The major physiologic problem is total collapse of the airways during expiration. Choice B is the only choice that is not true; it describes the pathologic findings of the Williams-Campbell syndrome, another cause of bronchiectasis and progressive obstructive lung disease.

The Mounier-Kuhn syndrome is found in patients with Ehlers-Danlos syndrome, Marfan’s syndrome, and cutis laxa. An autosomal recessive form has also been reported. The primary focus of treatment is the same as for any patient with bronchiectasis (postural or mechanical drainage, antibiotics, and treatment of obstructive airway disease). Pneumonic stenting with continuous positive airway pressure or mechanical stenting (Y-shaped stent) have been proven to be effective in some cases by improving lung function, facilitating bronchial drainage, and improving oxygenation.

IgE levels are normal in this disease but are elevated in two other bronchiectatic conditions: Job syndrome and proximal bronchiectasis associated with allergic bronchopulmonary mycosis.

**Suggested Readings**