Kartagener's Syndrome: *recurring lungs infections and COPD exacerbations*

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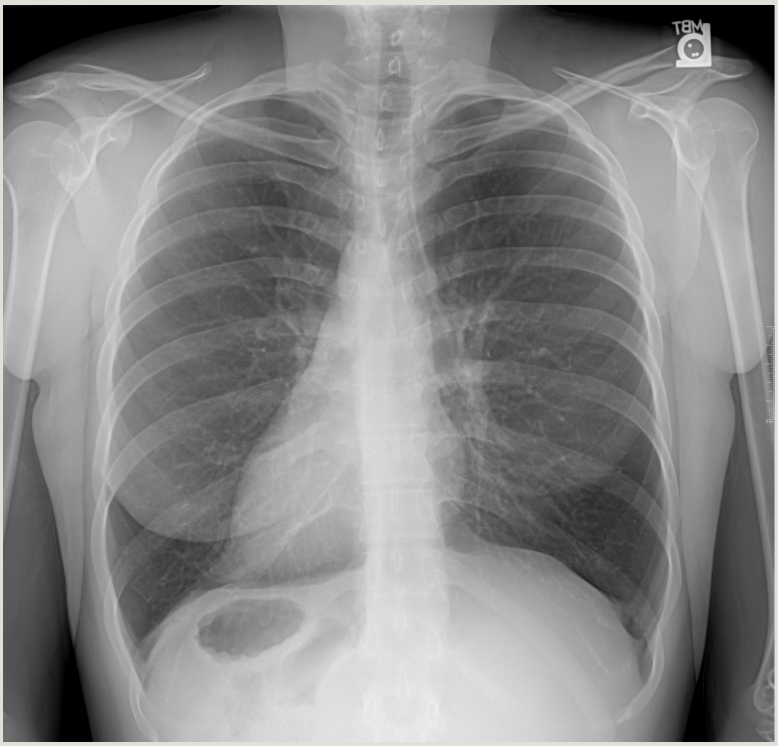

Figure 1 CXR demonstrates situs inversus (right sided heart, aorta and stomach bubble) and a dilated bronchial tree, mainly in upper and lower lobe on the left side.

Case
29-year-old female with Kartagener's Syndrome presented to clinic with chronic cough, sputum production and dyspnea. The diagnosis was made at age 2 by sinus biopsy. The patient had problems with hypoxemia as an infant. There were recurring infections of lungs, ears and nose since age 2. A rotation of Levaquin, Bactrim and Augmentin was tried for the treatment of bronchiectasis, but it failed. The patient is currently treated with inhaled Tobi, albuterol and a vibrating vest. Despite this treatment 3-4 admissions to the hospital for exacerbations of COPD occur yearly.

Pulmonary Function Tests
FVC - 65.7% of predicted
FEV1 - 28.8% of predicted
FEV1/FVC - 0.474

Primary ciliary dyskinesia is a genetic disorder with manifestations present from early in life. Approximately one half of the patients with primary ciliary dyskinesia have situs inversus, and these patients are diagnosed with Kartagener's syndrome (KS). This syndrome consists of a classic triad of sinusitis, situs inversus, and bronchiectasis. KS is inherited with an autosomal recessive pattern; its incidence is about 1 in 30,000 live births.

The diagnostic criteria recommended for this syndrome are a history of chronic bronchial infection and rhinitis from early childhood, combined with one or more of following features: (a) situs inversus or dextrocardia in the patient or a sibling, (b) living but immotile spermatozoa, (c) tracheobronchial clearance, which is absent or nearly absent. Treatment includes antibiotics to treat upper and lower airway infections. Obstructive lung disease/bronchiectasis should be treated with inhaled bronchodilators, mucolytics, and chest physiotherapy.

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