

AMSER Case of the Month: July 2019

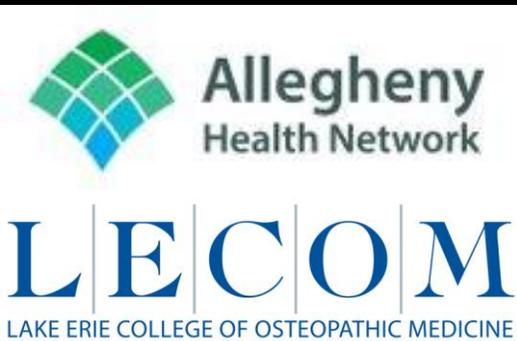
Febrile with Recurrent Pneumonia

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Patient Presentation

- HPI: 46 year old female with Kartagener's Syndrome and recent Pseudomonas infection s/p cefepime treatment, presented to ED with worsening shortness of breath. In the ED, the patient was febrile to 102 F and tachypnic. She was started on vancomycin, vefepime, doxycycline, and albuterol nebulizer and admitted to the ICU.
- PMH: Kartagener's Syndrome, Bronchiectasis, Sinusitis, Hyperlipidemia
- Surg Hx: Sinus surgery, appendectomy
- Med: Flonase, Advair, Singulair, Spiriva, Low-Ogestrel
- Allergies: Bactrim, Sulfa
- Family Hx: Father – CAD, Mother – Gallbladder disease
- Social Hx: No tobacco, alcohol, or drug use

What Imaging Should We Order?

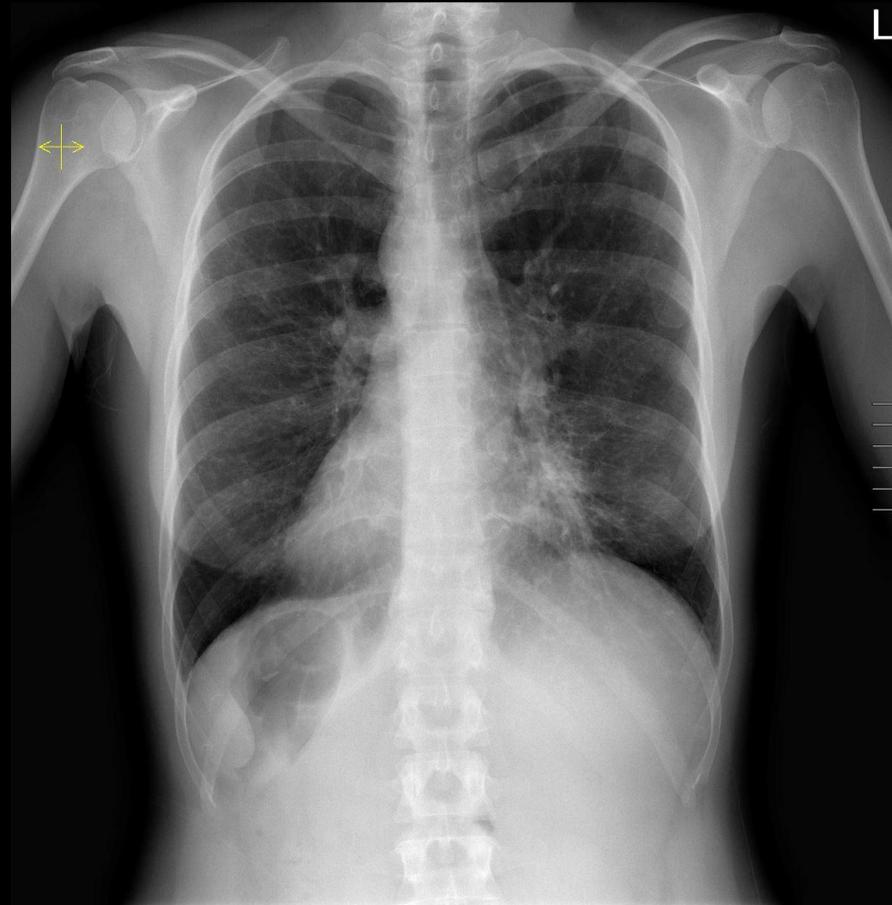
ACR Appropriateness Criteria

Variant 2:

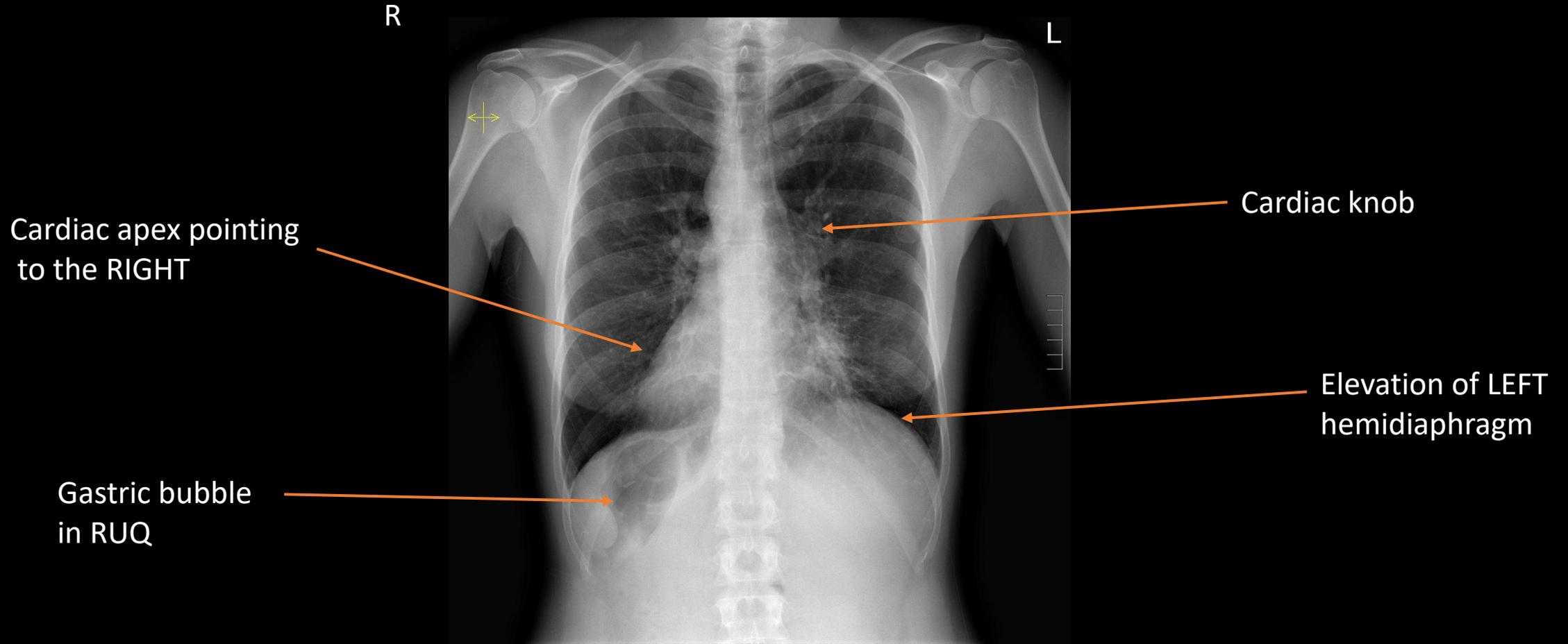
Acute respiratory illnesses in immunocompetent patients with positive physical examination, abnormal vital signs, organic brain disease, or other risk factors. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	⊕
US chest	May Be Appropriate	○
CT chest with IV contrast	Usually Not Appropriate	⊕ ⊕ ⊕
CT chest without and with IV contrast	Usually Not Appropriate	⊕ ⊕ ⊕
CT chest without IV contrast	Usually Not Appropriate	⊕ ⊕ ⊕
MRI chest without and with IV contrast	Usually Not Appropriate	○
MRI chest without IV contrast	Usually Not Appropriate	○

Findings: unlabeled



Findings: labeled



ACR Appropriateness Criteria

Variant 3:

Acute respiratory illness in immunocompetent patients with positive physical examination, abnormal vital signs, organic brain disease, or other risk factors and negative or equivocal initial chest radiograph. Next imaging study.

Procedure	Appropriateness Category	Relative Radiation Level
CT chest without IV contrast	Usually Appropriate	⊗ ⊗ ⊗
CT chest with IV contrast	May Be Appropriate (Disagreement)	⊗ ⊗ ⊗
US chest	May Be Appropriate	○
CT chest without and with IV contrast	Usually Not Appropriate	⊗ ⊗ ⊗
MRI chest without and with IV contrast	Usually Not Appropriate	○
MRI chest without IV contrast	Usually Not Appropriate	○

Findings: unlabeled



Findings: labeled

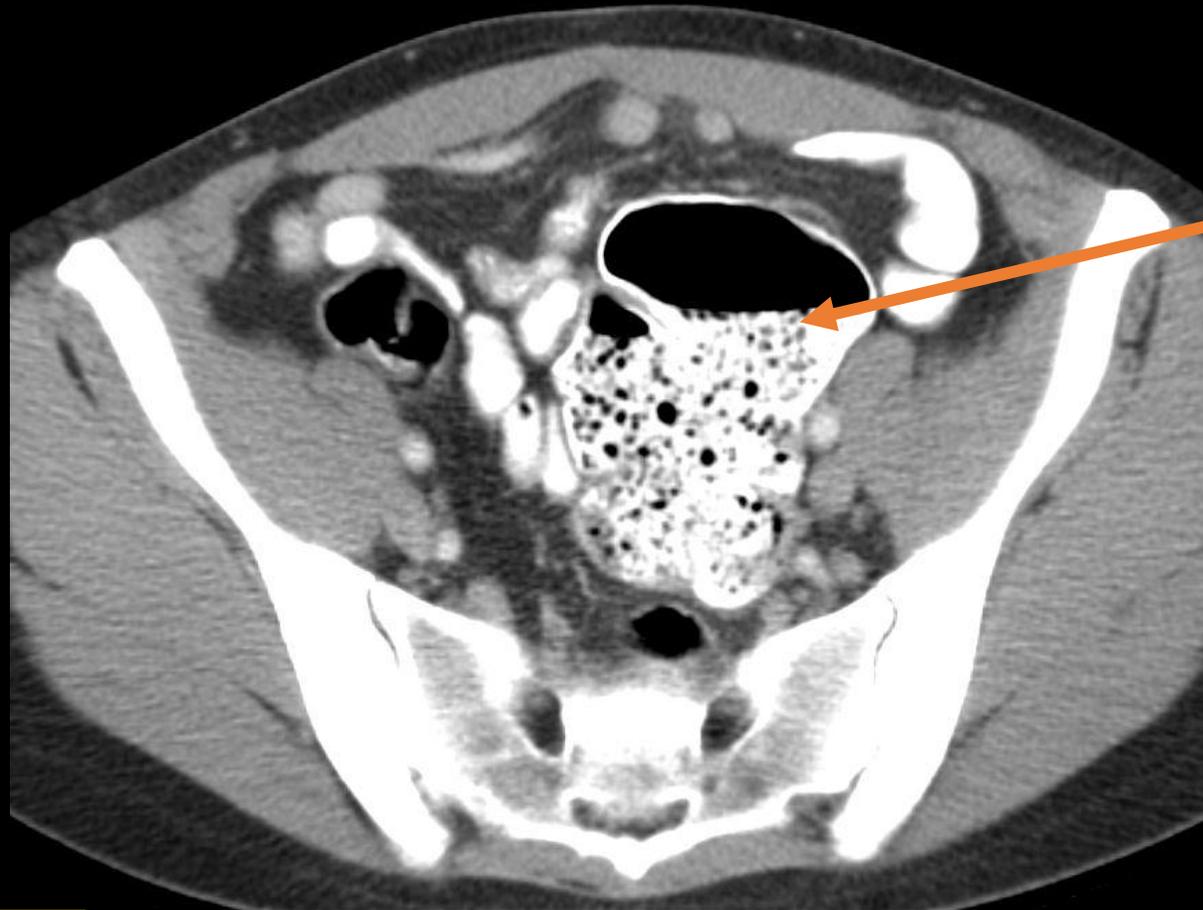


CT of the abdomen show the stomach and spleen are in the RIGHT upper abdomen and liver is in the LEFT.

Findings: unlabeled



Findings: labeled



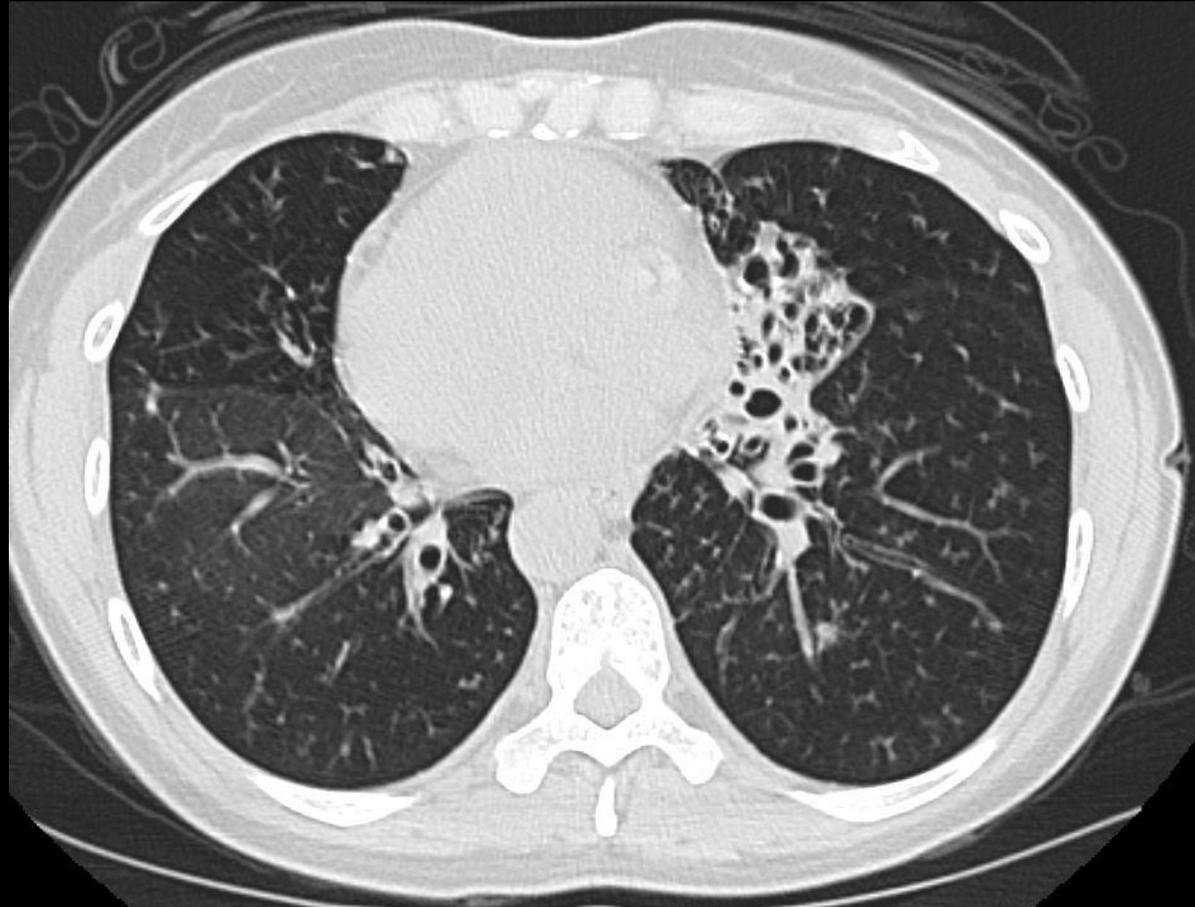
Cecum

R

L

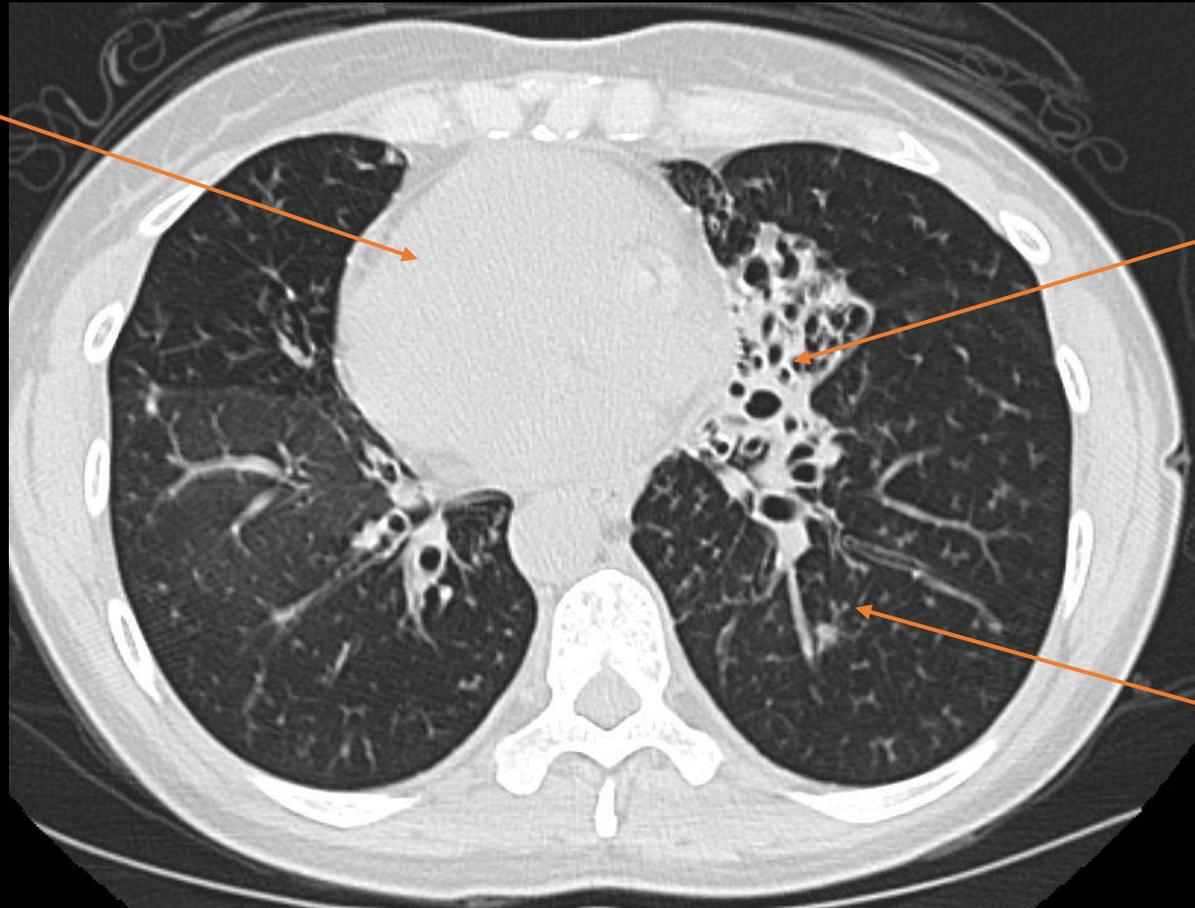
CT of the pelvis shows the cecum is in the LEFT lower quadrant

Findings: unlabeled



Findings: labeled

Heart



Bronchiectasis/dilated airways with peribronchial wall thickening

Scattered areas of "tree-in-bud" nodular opacities

R

L

Diagnosis

Kartagener's Syndrome with Situs Inversus and
Recurrent Bronchiolitis/Bronchopneumonia

Case Discussion

- Initial Presentation

- Recurrent infections of upper and lower respiratory tract in childhood
- Common symptoms included: chronic cough, nasal congestion, mucopurulent sputum, recurrent sinusitis, and recurrent otitis media

- Imaging modalities

- Plain CXR reveals situs inversus with dextrocardia, right-sided stomach bubble, and right aortic arch
- CT scan of the chest reveals dextrocardia and bilateral bronchiectasis.
- CT scan of abdomen reveals left-sided liver, right-sided stomach, and right-sided spleen.

Case Discussion

- Kartagener Syndrome is an autosomal recessive genetic disorder presented as primary ciliary dyskinesia with complete reversal of circulatory system and viscera (situs inversus totalis).
- Evidences of situs inversus totalis are seen with liver and cecum on the left, and spleen and stomach on the right of the body.
- Extensive bronchiectasis noted with bronchial dilation and peribronchial wall thickening, worst in the anatomic left middle lobe. Scattered bilateral “tree-in-bud” nodular opacities throughout the lung. These constellation of findings are consistent with bronchiolitis or bronchopneumonia.

Case Discussion

- Diagnosis of Kartagener Syndrome included triad of: situs inversus, chronic sinusitis, and bronchiectasis.
- The patient had repeat bacterial infections which caused an acute exacerbation of her bronchiectasis.
- Commonly, patients will have recurrent aspiration/pneumonia due to inability of cilia to clear airway secretion
- Recommendation of long term preventive antibiotic therapy such as macrolide.
- Vaccination against influenza and pneumococcus.

References:

- American College of Radiology. ACR Appropriateness Criteria for acute Respiratory Illness in Immunocompetent Patients. Available at <https://acsearch.acr.org/docs/69446/Narrative/>. Accessed May 16 2019.
- UptoDate. Clinical manifestations and diagnosis of bronchiectasis in adults. Accessed May 17 2019.
- UptoDate. Primary ciliary dyskinesia (immotile-cilia syndrome). Accessed May 16 2019