



## Bronchiectasis

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## Bronchiectasis - Outline

- Definition & Radiology Review
- Epidemiology & Pathophysiology
- Etiologies
- Diagnosis & Work-up
- Treatment
- Case review

## Case

- 64 yo man with 5 years of chronic cough
- Dry cough with minimal mucous production
- Antibiotics at least 4 times/year
- Sinus disease with improvement s/p surgery
  
- Never smoker
- Grew up on a farm – livestock and chickens
- Works as an engineer
- PCP sent him for a CT scan

## And the CT Scan is Read As:

- BRONCHIECTASIS
  
- What IS this anyway?

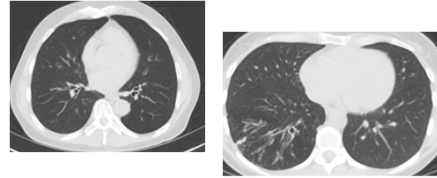
## Radiology

- What exactly do we see on imaging?
  - Bronchial diameter exceeding that of the adjacent pulmonary artery
  - Lack of normal tapering of terminal bronchioles in the lung periphery

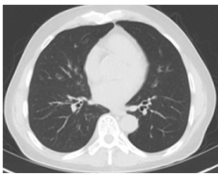
RadioGraphics 2015; 35:1011-1030

## Bronchiectasis

- Imaging: abnormal and permanent dilatation of the bronchi



Bronchiectasis



Normal

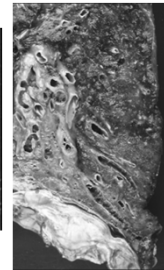
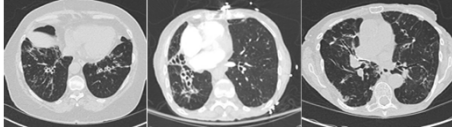


Image: Yale Rosen CC BY-SA 2.0  
[https://www.flickr.com/photos/pulmonary\\_pathology/3677946871](https://www.flickr.com/photos/pulmonary_pathology/3677946871)



## Bronchiectasis

- Clinical syndrome: cough, sputum production and recurrent bronchial infection
  - +
- radiological findings of dilated airways
- Occurs in multiple pathologic processes

Eur Respir J 2017; 50: 1700629

UpToDate Feb 2021. Clinical manifestations and diagnosis of bronchiectasis in adults

## Epidemiology

- Up to 500,000 US adults have bronchiectasis
- Prevalence increases with age, highest rates in adults >60 years old
- More common in women
- Extensive use of healthcare resources

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## Pathophysiology

- Two major factors
  1. Infectious insult
  2. Impaired drainage, airway obstruction or defect in host defense
- Airway neutrophils, cytokines and other immune responses cause abnormal dilatation and destruction of airways (bronchi and bronchiole walls)

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## Etiologies

### Acquired bronchial obstruction

- Foreign body aspiration
- Tumors
- Hilar adenopathy
- COPD
- Mucooid impaction
- Other

### Congenital anatomic defects

- Tracheobronchial
- Vascular
- Lymphatic

### Immunodeficiency states

- IgG deficiency
- IgA deficiency
- Leukocyte dysfunction
- Other rare humoral immunity immunodeficiencies

### Abnormal secretion clearance

- Ciliary defects
- Cystic fibrosis
- Young's syndrome

## Etiologies (continued)

### Infections

- Childhood infections
- Bacterial infections
- Viral infections
- Other infections

### Miscellaneous disorders

- Alpha-1-antitrypsin deficiency
- Recurrent aspiration pneumonia
- Rheumatic disease
- Inflammatory bowel disease
- Toxic fume & dust inhalation
- Chronic rejection after solid organ transplantation

## Etiologies

- Multiple etiologies can cause or contribute to pathophysiologic process
- Cystic Fibrosis (CF) Bronchiectasis
  - Recurrent and chronic airway infections
  - Most recognized cause
- Non-CF Bronchiectasis
  - All etiologies other than CF

## Non CF Bronchiectasis

- Airway Obstruction
- Defective host defense - common variable immunodeficiency
- Rheumatic disease
- Primary Ciliary Dyskinesia
- Infections
- Allergic Bronchopulmonary Aspergillosis (ABPA)
- Fibrosing Lung Diseases
- Aspiration
- Congenital

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## Airway Obstruction

- Foreign body aspiration
- Intraluminal obstruction lesion (carcinoid)
- Extra luminal compression (mass or lymph nodes)
  
- Focal bronchiectasis at sight of obstruction

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## Defective host defense

- Ciliary defects, prolonged immunosuppression, hypogammaglobulinemia (IPH), CVID
- Bronchial wall injury from repeated infections
- Recurrent sinus and respiratory infections
- ? IgG subclass deficiency

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## Rheumatic & systemic disease

- RA and Sjogrens syndrome can be complicated by bronchiectasis
- Inflammatory bowel disease (UC > Crohn's)
- Bronchiectasis can occur prior to rheumatic symptoms/diagnosis
- Mechanism not known
- RA + bronchiectasis (and COPD) has higher mortality than other bronchiectasis associations

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## Primary Ciliary Dyskinesia

- Immotile-cilia syndrome with defect in airway cilia
- Autosomal recessive with 30+ variants
- Recurrent infections – upper and lower respiratory tracts
- Bronchiectasis middle lobe and lingula
  
- Nasal nitric oxide analysis (low level is consistent with PCD)
- Extended genetic testing

UpToDate Feb 2021. Overview of Primary Ciliary Dyskinesia  
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## Infections

- Multiple infections associated with bronchiectasis
  - Bacterial
  - Atypical bacteria (mycoplasma, chlamydia and legionella species)
  - Viral
  - Mycobacterial (TB and NTM)
  - Nocardia
  
- Childhood and recurrent infections

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## Mycobacterial Infections

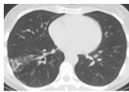
- Sequela of virulent infections
  - Direct tissue injury
  - Obstruction from enlarged lymph nodes
  
- Bronchiectasis is both a risk and consequence

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## Infections – NTM: chicken or egg?



- Underlying bronchiectasis may have NTM infection or colonization
- NTM infection (MAC and M abscessus) can cause bronchiectasis
- Fibronodular bronchiectasis caused by MAC usually in women >60 in RML and lingula.
  - Peribronchial inflammation and thickening that leads to bronchiectasis



UpToDate Oct 2020. Overview of NTM infections

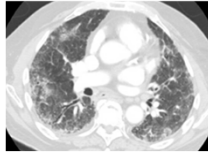
## Allergic Bronchopulmonary Aspergillosis (ABPA)

- Complex hypersensitivity reaction in response to colonization of the airways with *Aspergillus fumigatus*
- Occurs in patients asthma or cystic fibrosis (CF)
- CT with peripheral and central airway bronchiectasis
  
- Blood eosinophilia
- Elevated plasma IgE
- Precipitating specific antibodies Aspergillus

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## Fibrosing Lung Diseases

- Sarcoidosis – upper/central airways
- Idiopathic Pulmonary Fibrosis (IPF) – lower airways
- Sequela of acute respiratory distress syndrome (ARDS) – middle lobe and lingula



RadioGraphics 2015; 35:1011–1030

## Other contributors

- Vitamin D Deficiency
  - Observational study of 402 patients with bronchiectasis
  - 50% deficient and 43% insufficient
  - Deficient patients with more pseudomonas colonization, more exacerbations and worse symptoms
- Cigarette smoking → COPD
  - Causal role not clear
  - Repeated infections/exacerbations can accelerate disease

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## Other contributors

- Chronic aspiration
  - Airway destruction from acidic GI contents
  - Lower lobe airways
- Alpha-1 Antitrypsin (A1AT) deficiency – most associated with premature panlobular emphysema
  - Abnormal elastase
  - Lower lobe predominance

RadioGraphics 2015; 35:1011–1030

## Congenital syndromes

- Williams-Campbell Syndrome: rare cartilage deficiency of the mid-order bronchi.
- Swyer-James Syndrome: post infectious bronchiolitis obliterans
  - smaller lucent lung usually accompanied by diffuse bronchiectasis

RadioGraphics 2015; 35:1011–1030

## Work-up

- Imaging → CT Chest (preferably thin sections)
  - CXR insensitive
- Bronchoscopy
- CBC with differential, Immunoglobulins, autoimmune, sweat chloride, CFTR gene mutation, nasal nitric oxide analysis, PCD gene testing, A1AT, RAST (aspergillus testing)
- Cultures – AFB, fungal
- PFTs (often obstructive impairment)

RadioGraphics 2015; 35:1011–1030

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease

## Bronchiectasis - Exacerbations

- Deterioration of 3 or more symptoms for  $\geq$  48 hour
  - Cough
  - Sputum volume and/or consistency
  - Sputum purulence
  - Breathlessness and/or exercise intolerance
  - Fatigue and/or malaise
  - Hemoptysis
- Mucous tends to more tenacious and concentrated as compared to healthy controls and other conditions

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease

## Guidelines - 2017

### European Respiratory Society guidelines for the management of adult bronchiectasis

Eva Polverino<sup>1</sup>, Pieter C. Goeminne<sup>2,3</sup>, Melissa J. McDonnell<sup>4,5,6</sup>, Stefano Aliberti<sup>7</sup>, Sara E. Marshall<sup>8</sup>, Michael R. Loebinger<sup>9</sup>, Marlene Murriss<sup>10</sup>, Rafael Cantón<sup>11</sup>, Antoni Torres<sup>12</sup>, Katerina Dimakou<sup>13</sup>, Anthony De Soya<sup>14,15</sup>, Adam T. Hill<sup>16</sup>, Charles S. Haworth<sup>17</sup>, Montserrat Vendrell<sup>18</sup>, Felix C. Ringshausen<sup>19</sup>, Dragan Subotic<sup>20</sup>, Robert Wilson<sup>1</sup>, Jordi Vilaró<sup>21</sup>, Bjorn Stallberg<sup>22</sup>, Tobias Wette<sup>19</sup>, Gernot Rohde<sup>23</sup>, Francesco Biasi<sup>2</sup>, Stuart Elborn<sup>24</sup>, Marta Almagro<sup>25</sup>, Alan Timothy<sup>25</sup>, Thomas Ruddy<sup>25</sup>, Thomy Tonia<sup>26</sup>, David Rigau<sup>27</sup> and James D. Chalmers<sup>28</sup>

## Treatment - Exacerbations

- Antibiotics – choice of agent based on cultures
- 14 days of treatment
- Long term antibiotics (> 3 months) in adults with 3 or exacerbations/year
  - Inhaled antibiotics with chronic *P. aeruginosa* infection
  - Macrolide therapy
- Eradication therapy with new isolation of *P. aeruginosa*
  - Combination of oral, IV and/or inhaled therapies

Eur Respir J 2017; 50: 1700629



## Treatments - inhalers

- Inhaled corticosteroids and long-acting bronchodilators – not recommended for routine use
- Continued in patients with co-morbidities of asthma and/or COPD
- Trial of short or long acting bronchodilators in certain patients (significant breathlessness)

Eur Respir J 2017; 50: 1700629

## Treatment – airway clearance

- Mucous clearance therapies
  - Directed cough
  - Exercise
  - Forced expiration
  - Chest physical therapy – postural drainage, hand or mechanical chest clapping
  - Oscillation vest
  - Vibratory Positive Expiratory Pressure (PEP)



VibroPEP by Comtek  
MediGlobe.com/medtek.com

UpToDate Oct 2020. Bronchiectasis in adults: Treatment of acute exacerbations and advanced disease



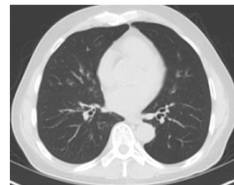
Acapella Vibratory PEP  
DME Supplies

## In practice....

- Steroids – most often inhaled
- Inhaled and oral antibiotics
- Clearance aides (flutter/acapella and chest vest)
- Nebulized hypertonic saline

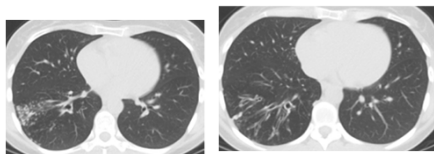
## Case 1

- Recurrent cough and sinus drainage
- Improves with oral steroids
- IgG, IgM and IgA low end of normal



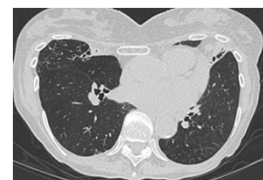
## Case 2

- 37 yo old with 4 years of chronic cough
- BAL with MAC
- Chronic sinus disease



## Case 3

- 87 yo old with mild, intermittent, chronic cough
- Minimal antibiotic, bronchodilator or steroid use



## Summary – Bronchiectasis

- Irreversible regional or diffuse bronchial dilatation
- Repeated pattern of airway infection, inflammation, and injury
- Multiple causes, including congenital diseases, infection, obstructing lesions, immunodeficiency, aspiration
- History + lab work + imaging findings can help with diagnosis
- Treatment consists of antibiotics, steroids (inhaled, oral), bronchodilators, mucous clearance assistance as well as treatment of underlying conditions

Eur Respir J 2017; 50: 1700629

## References

- UpToDate
  - Clinical manifestations and diagnosis of bronchiectasis in adults
  - Clinical manifestations and diagnosis of allergic bronchopulmonary aspergillosis
  - Primary Ciliary Dyskinesia (immotile-cilia syndrome)
  - Overview of nontuberculous mycobacterial infections
- Polverino E, Goeminne PC, McDonnell MJ, et al. European Respiratory Society guidelines for the management of adult bronchiectasis. *Eur Respir J* 2017; 50: 1700629 [https://doi.org/10.1183/13993003.00629-2017]
- Milliron, B et al. Bronchiectasis Mechanisms and Imaging Clues of Associated Common and Uncommon Diseases *RadioGraphics* 2015; 35:1011–1030