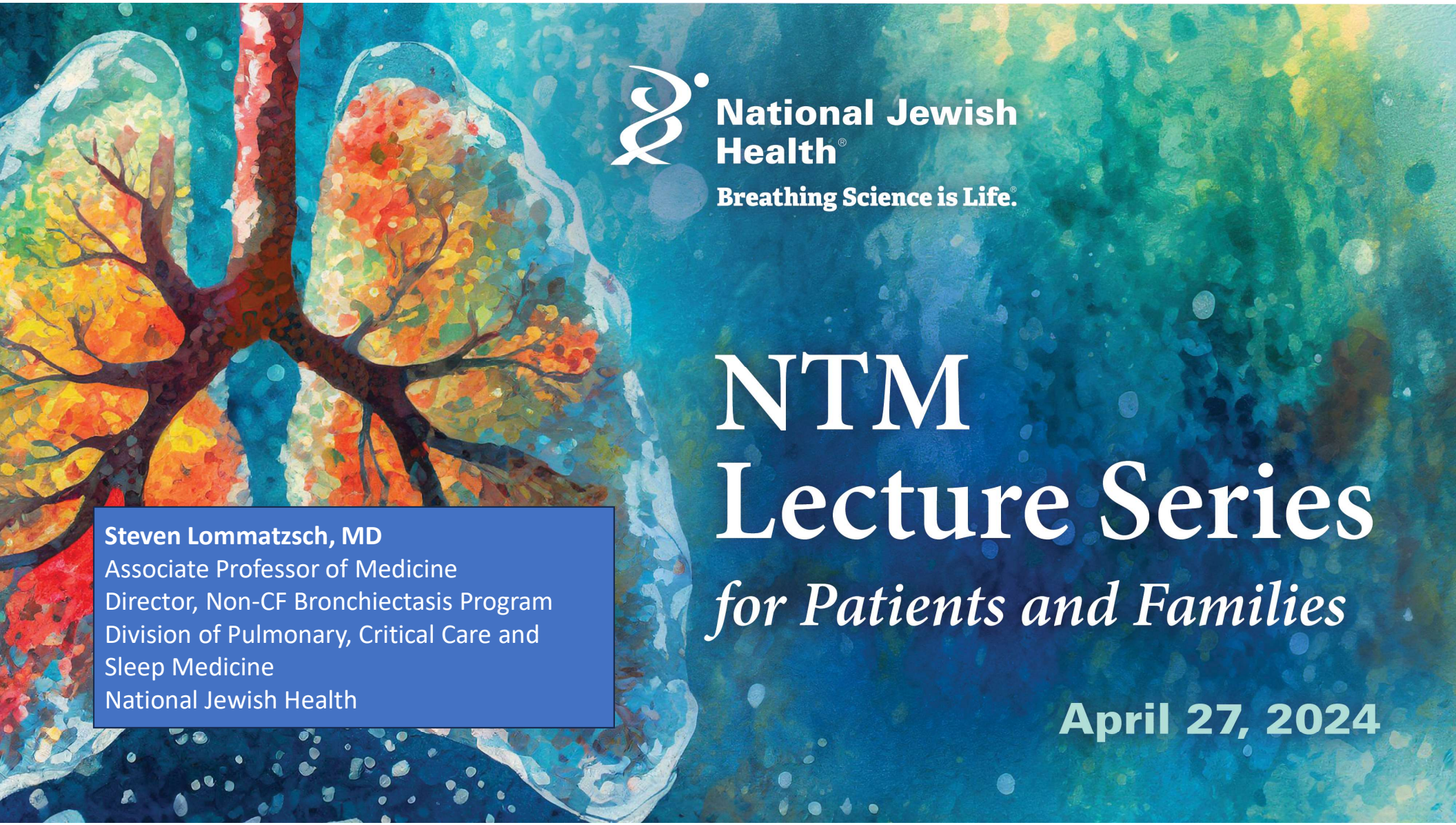
National Jewish Health®
Breathing Science is Life.®
NTM
Lecture Series
for Patients and Families
Steven Lommatzsch, MD
Associate Professor of Medicine
Director, Non-CF Bronchiectasis Program
Division of Pulmonary, Critical Care and Sleep Medicine
National Jewish Health
April 27, 2024

Overview of Bronchiectasis

- What is bronchiectasis
- How does one get bronchiectasis
- How is bronchiectasis treated
- Why is bronchiectasis relevant to Mycobacterial infection

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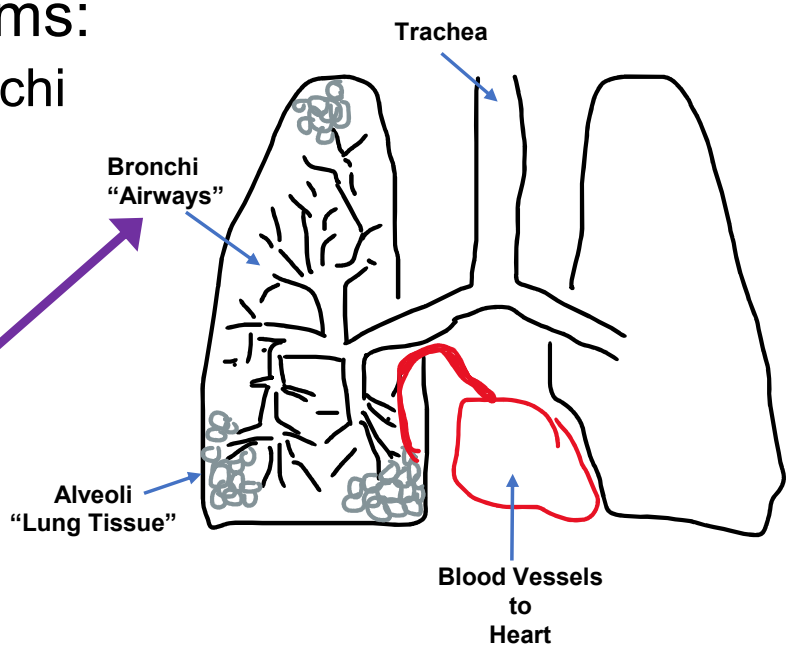
Normal Lung

- Composed of three main systems:
  - Airway System - Trachea & Bronchi
  - Tissue System - Alveoli
  - Vascular System - Blood vessels

Bronchiectasis is a problem with the BRONCHI = "Air tube problem"



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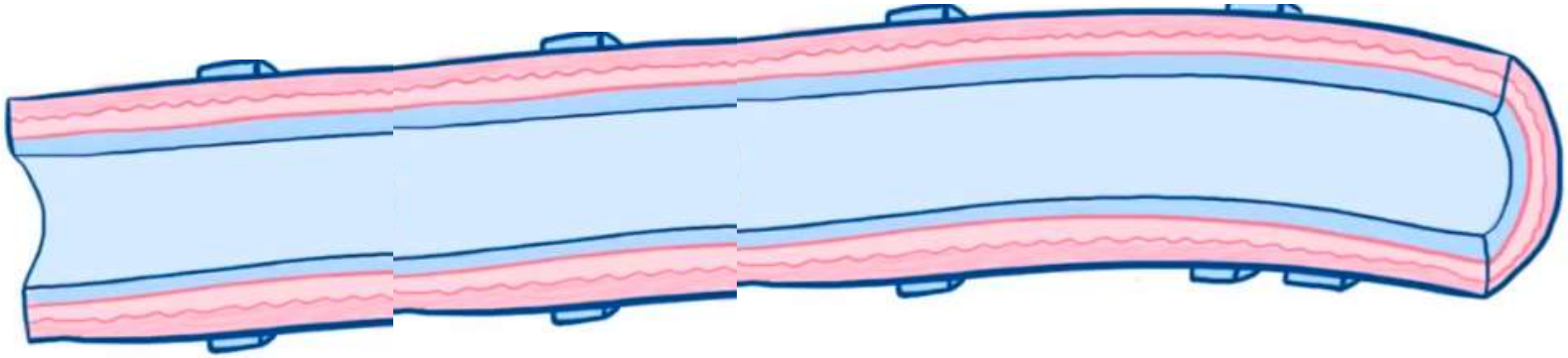
What is Bronchiectasis

From the Greek “bronkhos”
(windpipe or bronchial tubes) and “ektasis” (dilatation)[1]

Normal Bronchial Airway



1. Chalmers JD. *Chest.* 2017;151:1204-1206.

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What is Bronchiectasis

From the Greek “bronkhos”
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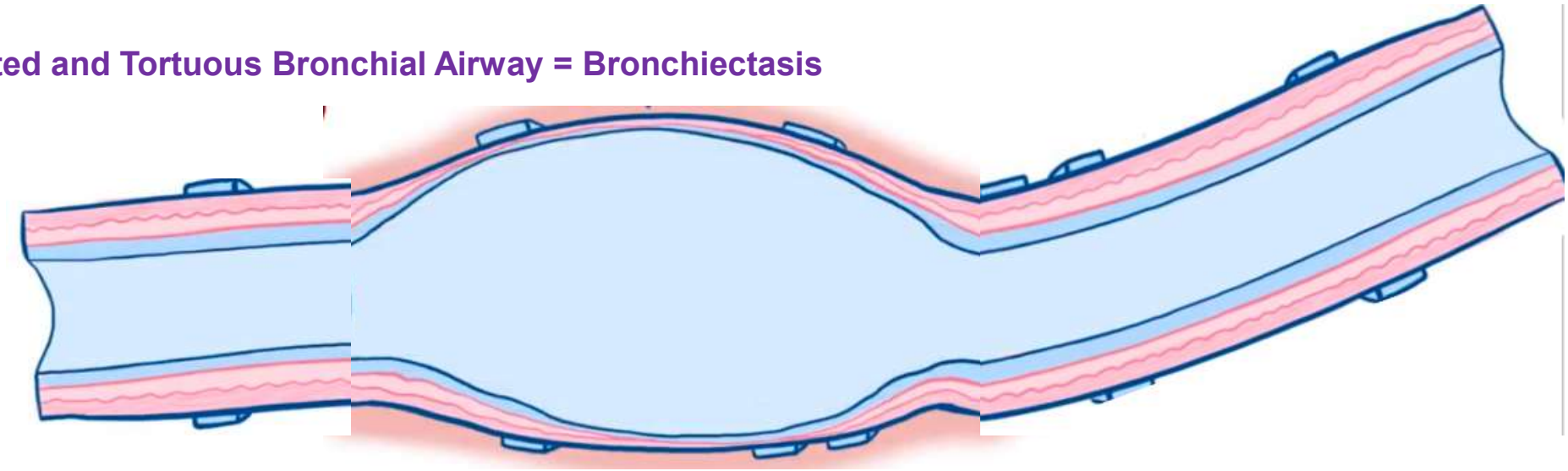
Dilated and Tortuous Bronchial Airway = Bronchiectasis



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What is Bronchiectasis

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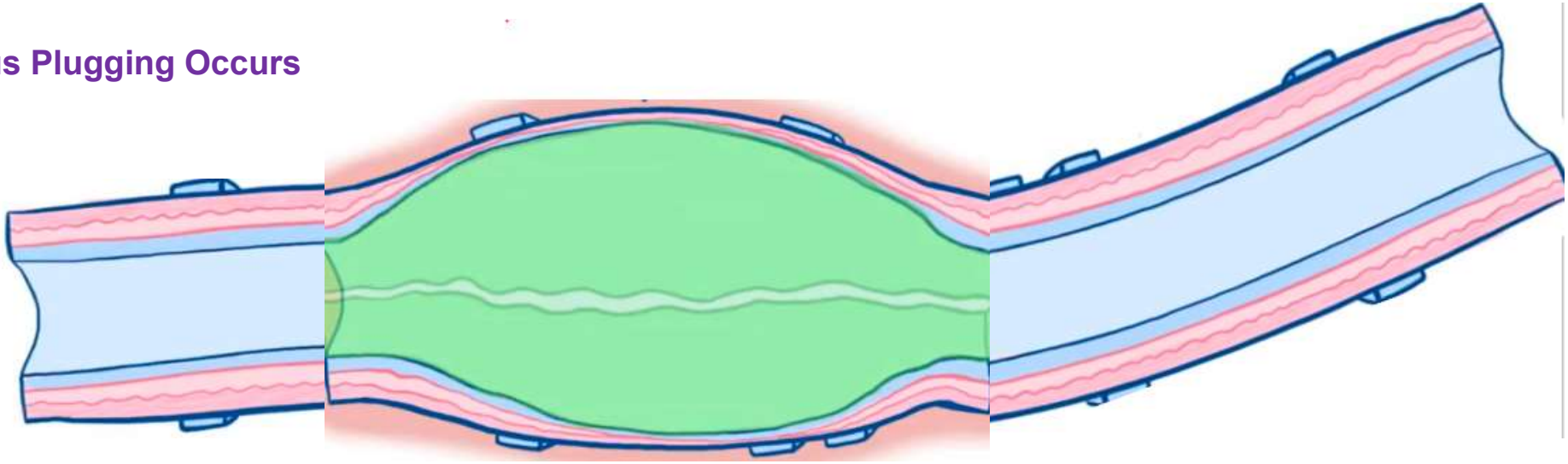
Mucus Plugging Occurs



1. Chalmers JD. *Chest.* 2017;151:1204-1206.

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Facts about Bronchiectasis

- Estimated that 350,000 to 500,000 adults in the US have the condition
- The condition is twice as common in women than men
- The disease increases in prevalence with increasing age
- The average number of times patients need to see their doctor in clinic to treat a respiratory illness is between 1 and 3 per year
- On average a patient with severe bronchiectasis is hospitalization once per year

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Diagnosis

- Symptoms may be common to many respiratory diseases
  - Cough, sputum production, shortness of breath, etc
  - Often takes exacerbation or acute event to come to appropriate medical attention
- Many diseases can cause bronchiectasis
- True diagnosis requires radiographic imaging with computed tomography (“CT scan”)

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Symptoms of Bronchiectasis

- Cough (98%)
- Chronic sputum production (78%)
- Dyspnea (62%)
- Fatigue (43%)
- Hemoptysis (27%)
- Wheezing (20%)

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Causes of Bronchiectasis

CONGENITAL	• Tracheobronchomegaly • Cartilage deficiency • Pulmonary sequestration • Yellow nail syndrome • Young's syndrome • Alpha-1 antitrypsin deficiency • Primary ciliary dyskinesia • Cystic fibrosis
IMMUNODEFICIENCY	• Hypogammaglobulinemia • CLL • Chemotherapy • Immunosuppression
POSTINFECTIOUS	• Bacteria • Mycobacterium • Aspergillus • Viruses
RHEUMATOLOGIC	• RA • SLE • Sjögren's syndrome • Relapsing polychondritis • IBD
ASPIRATION/ INHALATION	• Chlorine • Overdoses • Foreign bodies
Other	• ABPA

Abbreviations: ABPA, allergic bronchopulmonary aspergillosis; CLL, chronic lymphocytic lymphoma; IBD, inflammatory bowel disease; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus.

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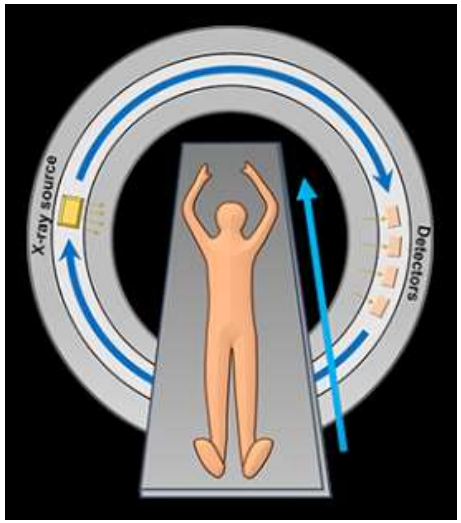
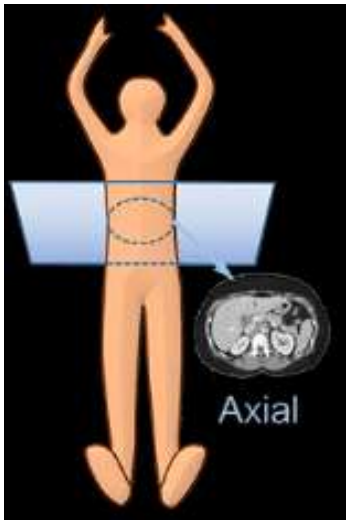
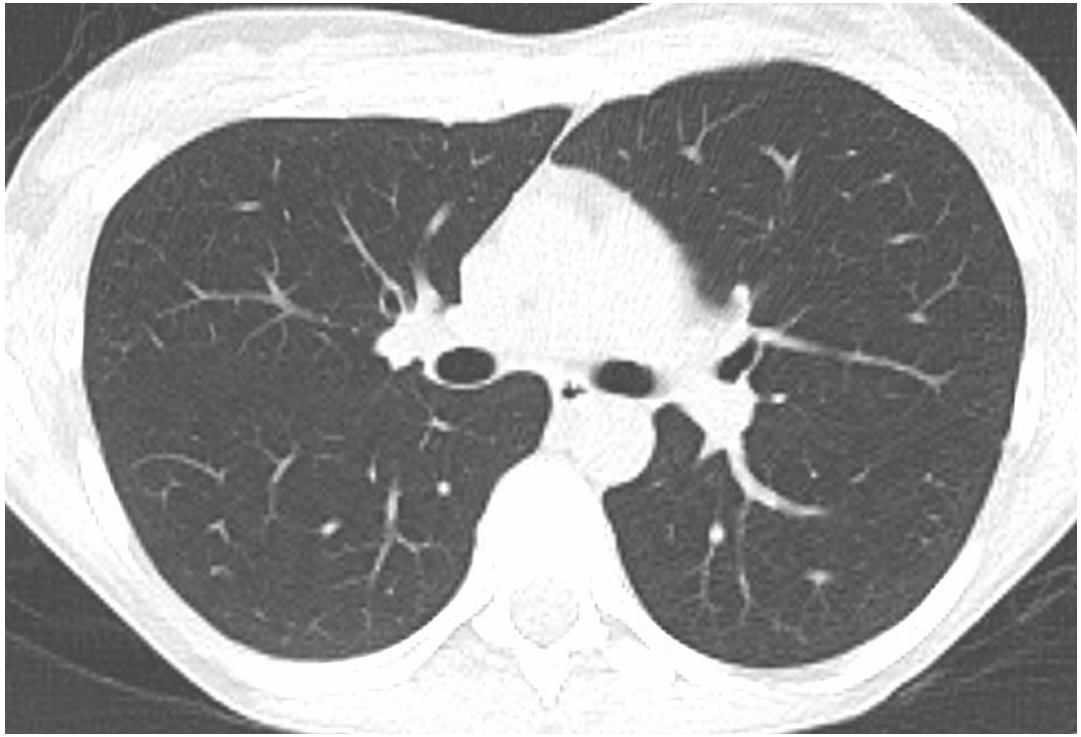
Imaging is Essential to Diagnosis

- Chest X-rays
- Chest CT scans







Example of normal chest CT scan

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CT Scan Makes the Diagnosis

Hill AT, et al. *Thorax.* 2019;74:1-69.

CT Features (≥1 of the following)

- Bronchoarterial ratio >1 (internal airway lumen/adjacent pulmonary artery) on CT scan
- Lack of airway tapering
- Airway visibility ≤1 cm of costal pleural surface or touching mediastinal pleura

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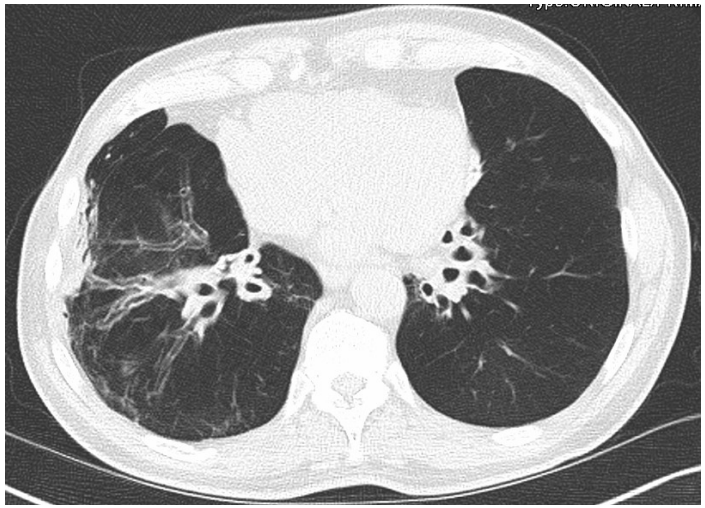
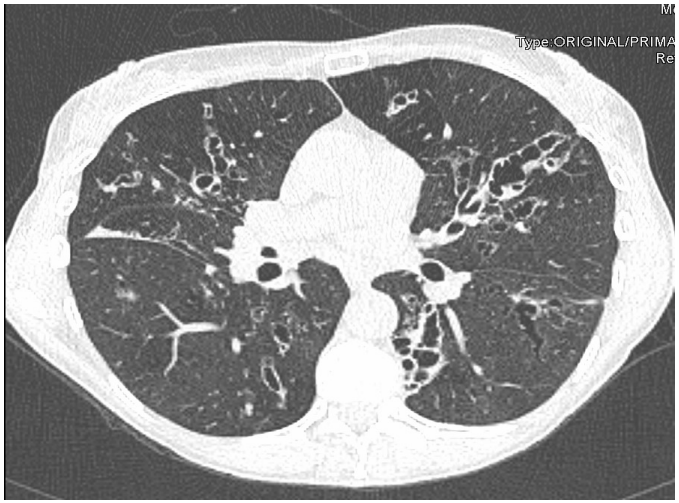
Types of Bronchiectasis seen on CT scan

Saccular/cystic

Cylindrical/tubular



Varicose



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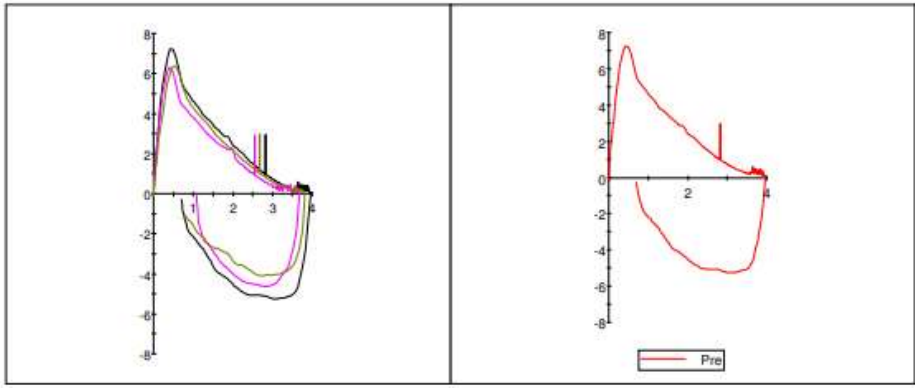
Other Testing

- "Breathing Test" – Spirometry
- Sputum cultures
  - Sometimes bronchoscopy
- Laboratory bloodwork
  - CBC and diff
  - RF/CCP, SSA, SSB, ANA, ANCA
  - IgE (initial per BTS)
  - Aspergillus precipitins (initial per BTS)
  - IgG, IgA, and IgM (initial per BTS)
  - Consider HIV testing
  - A1AT level/genotype
  - Antibody titers to pneumococcal vaccination (consider work-up per BTS)
  - Testing for PCD and CF (CF is first line per BTS if under 40yo)

---- SPIROMETRY ----	Pred	LLN	Actual	%Pred
FVC (L)	3.57	2.85	3.96	110
FEV1 (L)	3.08	2.47	2.84	92
FEV1/FVC (%)	86.46	74.73	71.75	82
FEF Max (L/sec)	6.68	5.06	7.24	108
FEF 25-75% (L/sec)	3.60	2.36	1.98	55
FIF Max (L/sec)	4.24		5.24	123
FEF50%/FIF50% (%)	90-100		50	
Expiratory Time (sec)			6.80	
Back Extrap Vol (L)			0.08	
FIVC (L)			3.25	



Treatment and Management Gaps

- There are currently no guidelines for the management of bronchiectasis in the United States
  - British Thoracic Society guideline, 2019 (updated from 2010)[1]
  - Thoracic Society of Australia and New Zealand position statement, 2023 (updated from 2015)[2]
  - European Respiratory Society guidelines, 2017[3]
- There are no therapies that are currently FDA-approved for the airway condition of bronchiectasis
- Much of the treatment of NCFBE has been influenced by cystic fibrosis research and management recommendations

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Treatment Starts With Identifying Cause

Condition / Disease	Treatment
ABPA	Oral steroids +/- oral antifungal
Alpha-1 antitrypsin deficiency	Alpha-1 protein replacement
Aspiration/GERD	Treat GERD and speech therapy
Cystic fibrosis	CFTR modulator therapy
Immunodeficiency (CVID)	IVIg replacement therapy
Infection (TB, NTM, etc)	Antibiotics
Rheumatologic/Autoimmune/ Inflammatory Diseases (RA, Sjogren's, IBD, etc)	Immunosuppression

Abbreviations: ABPA, allergic bronchopulmonary aspergillosis; CFTR, cystic fibrosis transmembrane conductance regulator; CVID, common variable immunodeficiency; GERD, gastroesophageal reflux disease; IBD, inflammatory bowel disease; IVIg, intravenous immunoglobulin; NTM, nontuberculous mycobacteria ; RA, rheumatoid arthritis; TB, tuberculosis.

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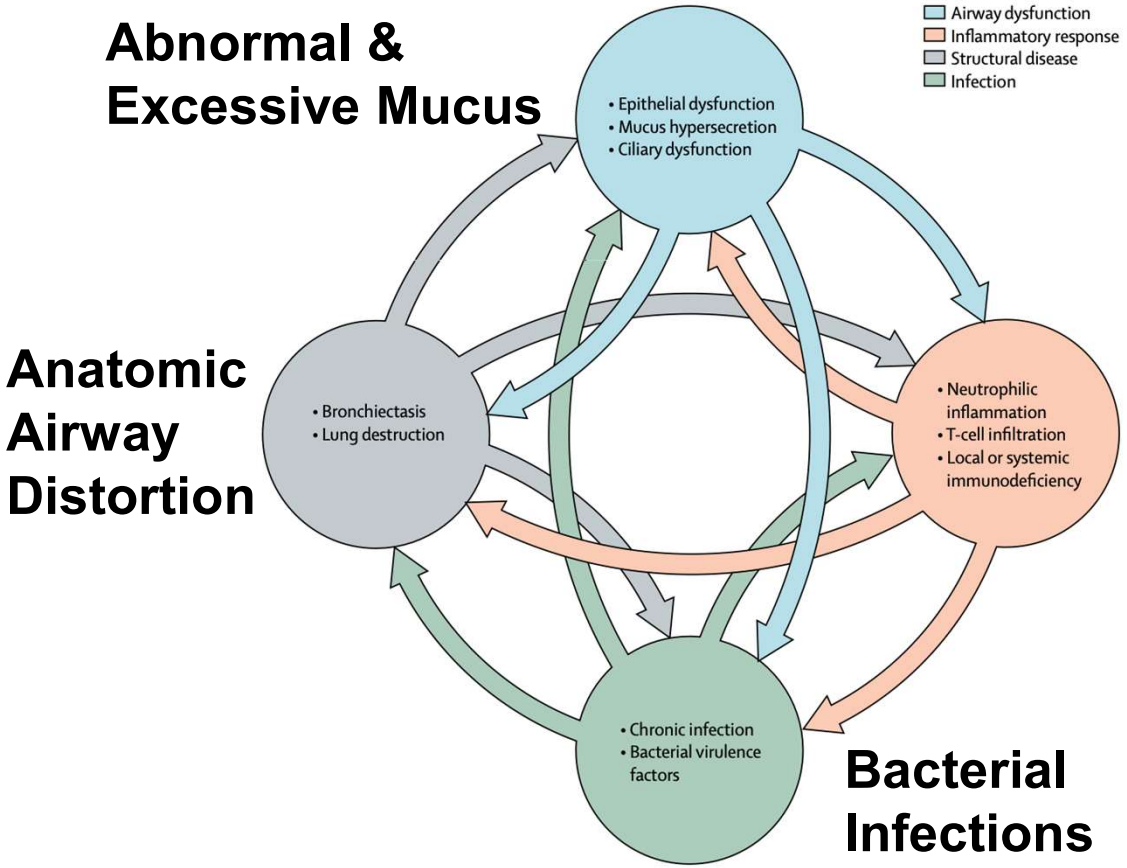
Targeted Treatment—The Vicious Vortex



Abnormal & Excessive Mucus

Anatomic Airway Distortion

Neutrophilic Inflammation

Bacterial Infections

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The 3 Cornerstones of Management

1. Airway clearance
2. Airway clearance
3. Airway clearance

Amazingly this cornerstone is often **forgotten** and **overlooked**!

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Components of Treatment

1. Mucus Management
2. Inflammation Attenuation
3. Infection Control

- Decreases progression of airway distortion and scarring
- Maintains better lung function

- Helps control patient symptoms
- Prevents illness / hospitalization
- Decreases likelihood of needing oxygen therapy

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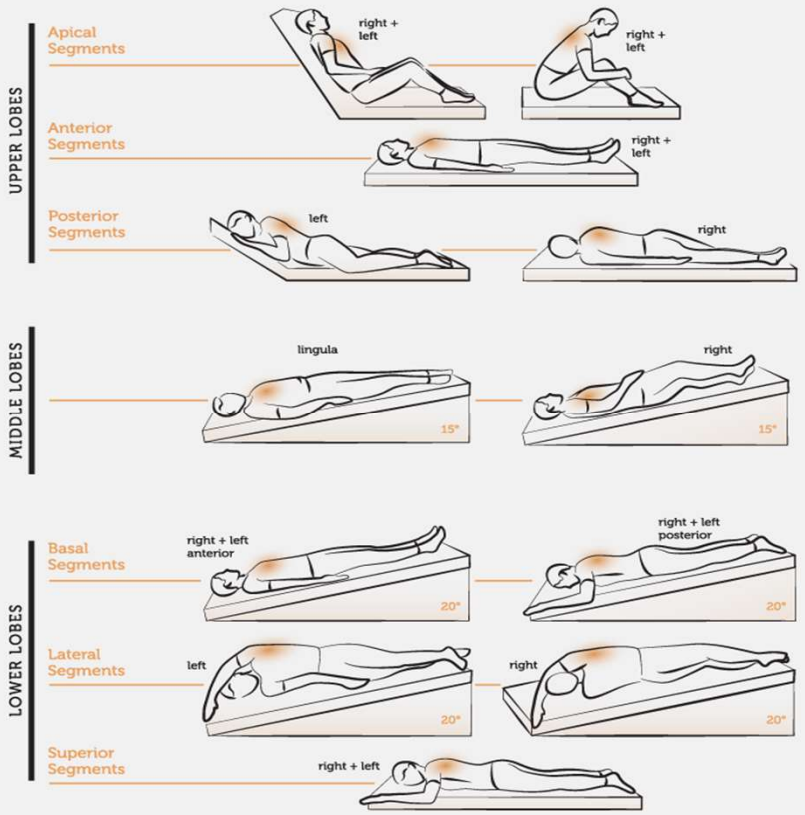
Airway Clearance - Mechanical

- Manual Chest Physiotherapy
- Active Cycle Breathing, Autogenic drainage, Huff Coughing
- Postural Drainage
- Positive expiratory pressure devices
- Oscillating devices, High-frequency chest wall oscillation, Flutter, Acapella devices
- Inspiratory muscle training
- Aerobic training/exercise



Airway Clearance - Pharmacologic

- Hypertonic saline (0.9%, 3%, 7%, 10%)
  - HR-QOL, 6MWT improvement, decrease healthcare utilization
- N-acetylcysteine (NAC) or “Mucomyst” nebulization
- Bronchodilator therapy – SABA before saline / airway clearance

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I've heard it all….

- "I can cough it up, so I don't need to do my airway clearance."
- "I do not get anything up when I use it, so I stopped."
- "I use it when I start to get sick."

- If bronchiectasis is a disease of distorted airways getting plugged with mucus and trapping bacteria in that mucus, then the treatment starts with getting that mucus out to clear the lung of bacteria/infection.

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Inflammation Attenuation

- Azithromycin daily therapy
  - Decrease exacerbations
  - Reduces Sputum Production
  - Improve lung function
  - Improve Quality of Life

"Macrolide" antibiotics are:

- Azithromycin

- Clarithromycin

- Erythromycin

It is important to **exclude** NTM infection with sputum cultures prior to starting therapy to avoid breeding resistance!

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Control Infections

- Treat Exacerbations
- Chronic suppressive inhaled antibiotic treatment
- NTM – MYCOBACTERIAL THERAPY

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Monitoring and Follow-up

- Regular visits with symptom assessments
- Spirometry – clinic based / home spirometry
- Sputum cultures
- Imaging / CT imaging (radiographic progression)
- Re-education and goals discussions

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Future Treatments

- Treatments that decrease neutrophil activity
- Biologic agents target inflammation
- Nebulized immunoglobulin therapy
- Inhaled ascorbic acid and glutathione
- CFTR potentiator therapy
- Novel antimicrobial development

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Thank you

- Any questions?

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