

NTM Lecture Series for Patients



September 21, 2019
NATIONAL JEWISH HEALTH

Bronchiectasis and NTM Overview

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Disclosures

Insmmed: advisory board member and speaker

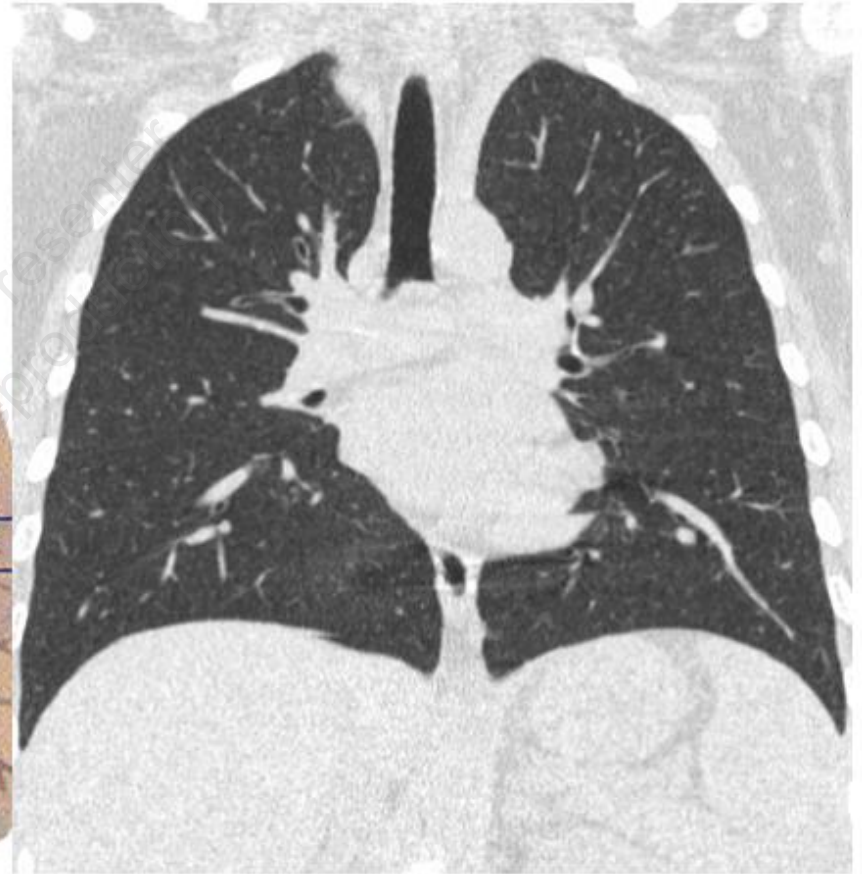
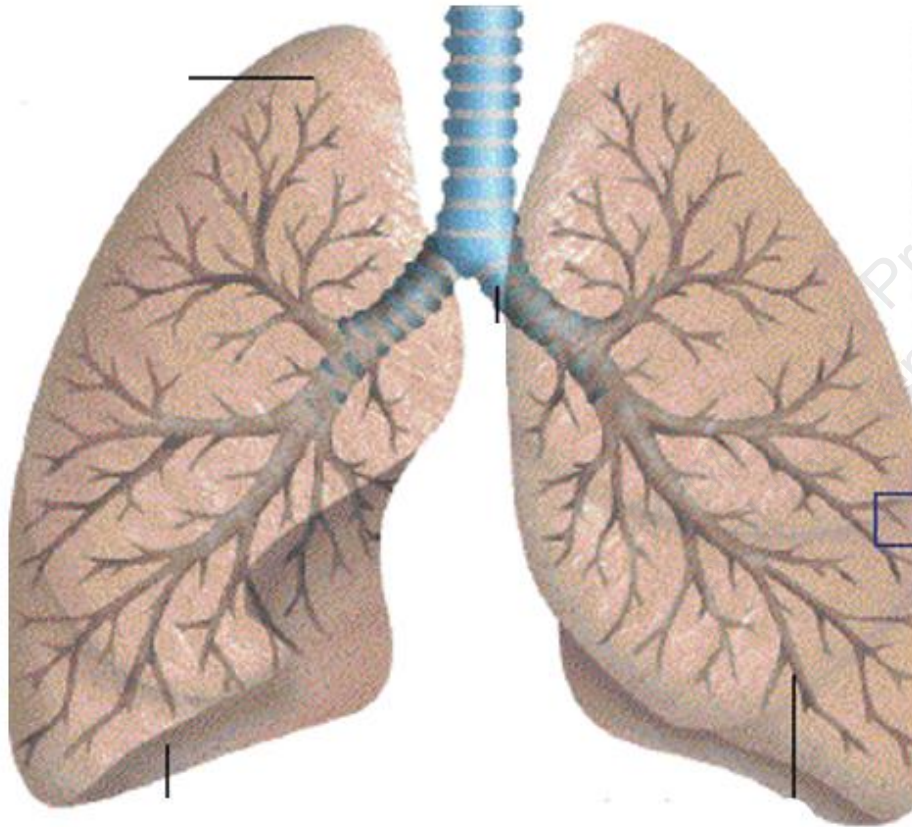
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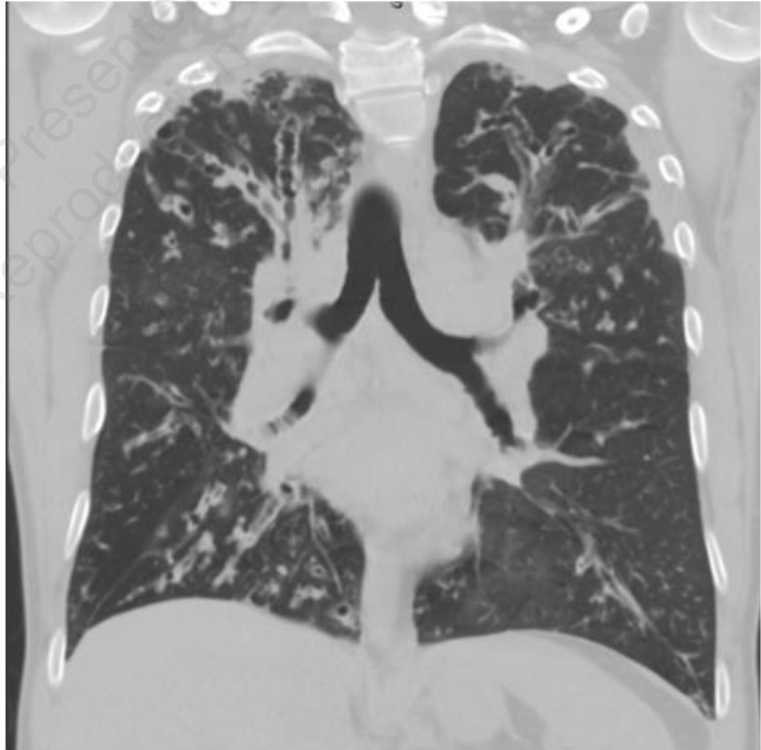
**Dr. Rene Laennec
1819**

**-inventor of the
stethoscope**

**-first to describe
bronchiectasis**



Bronchiectasis



Cylindrical

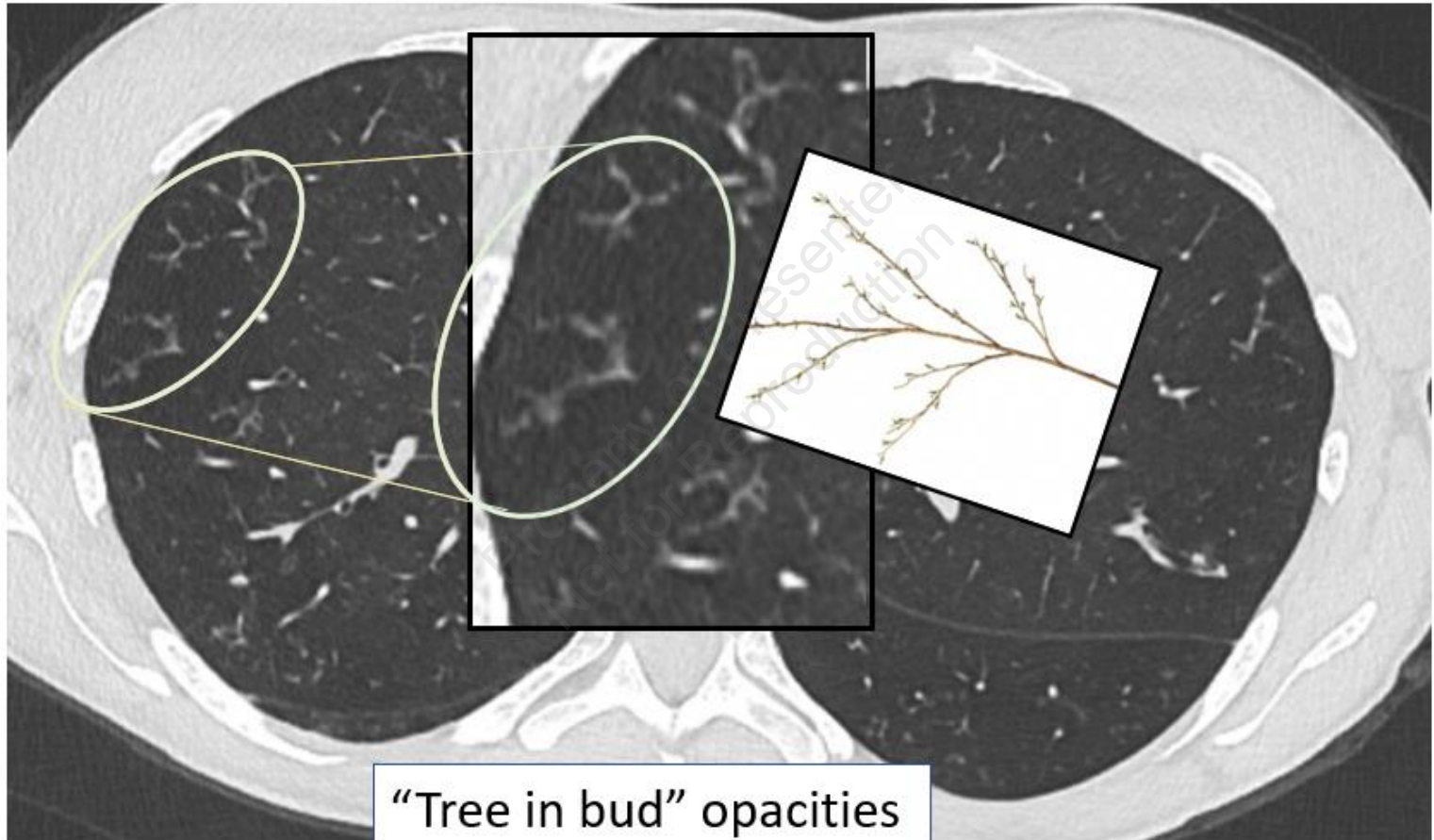


Varicose



Cystic



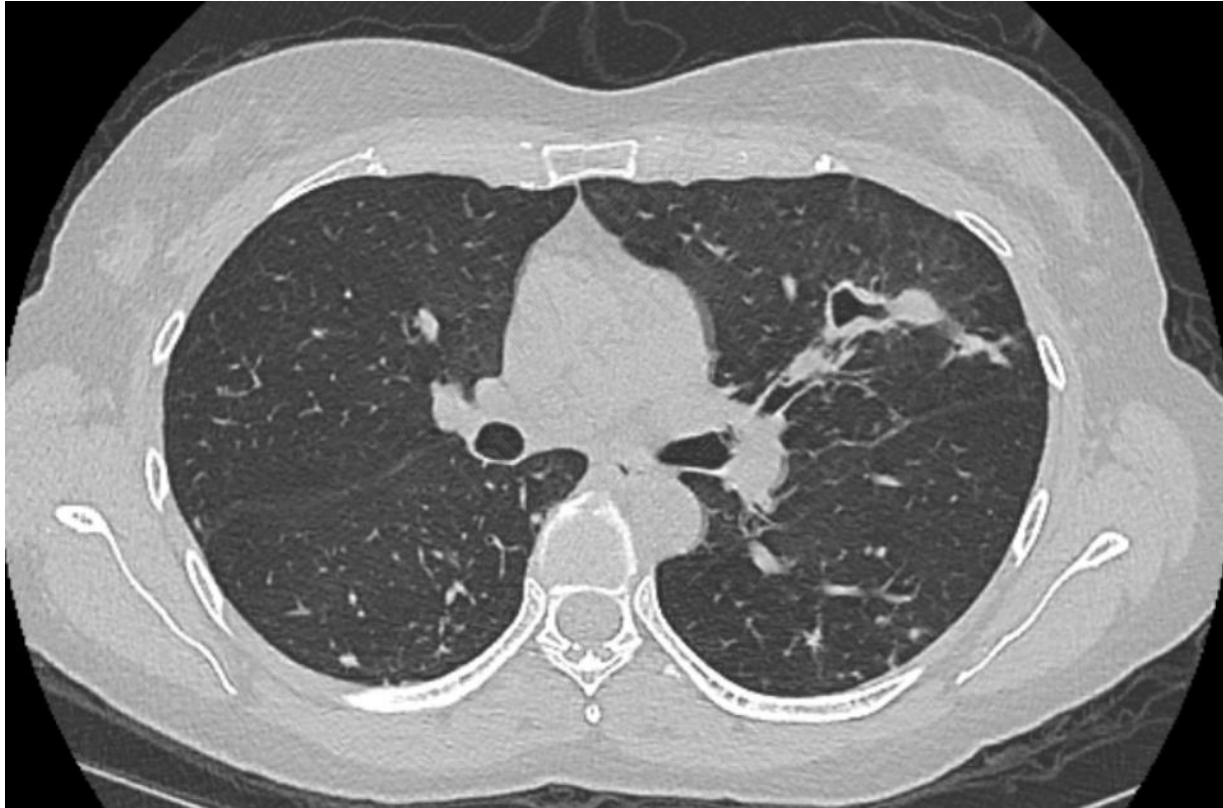


“Tree in bud” opacities

Nodular bronchiectasis

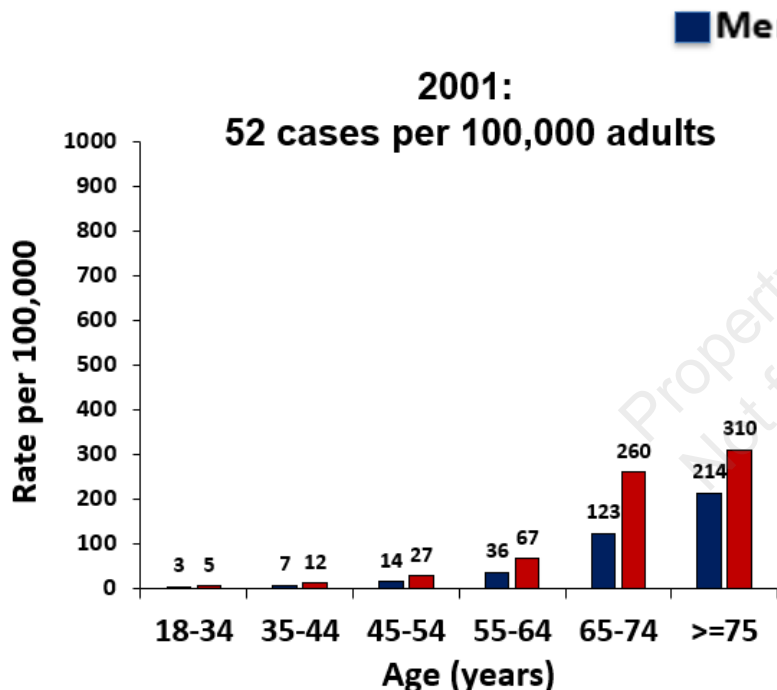


Cavitary bronchiectasis

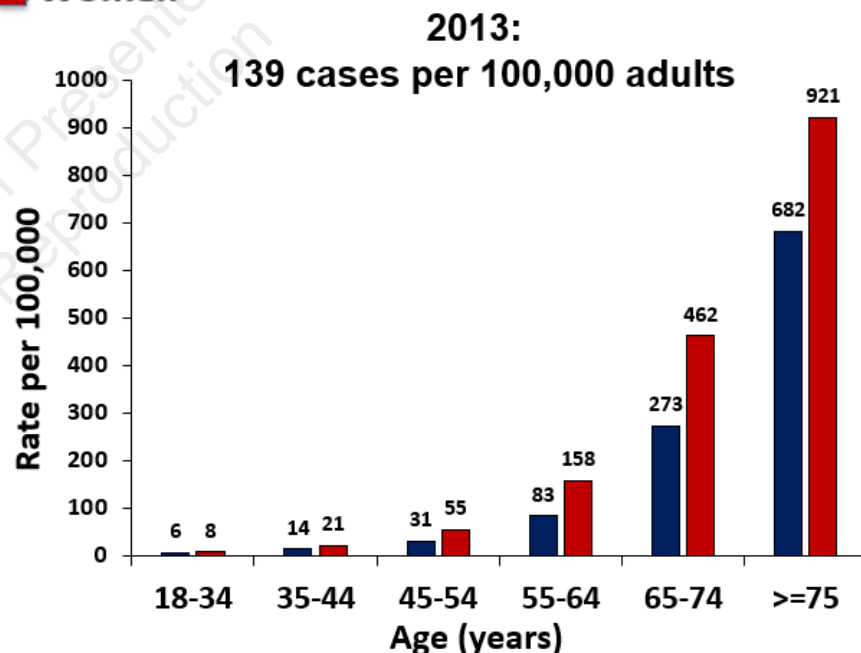


Prevalence of Bronchiectasis in US

Retrospective Analysis of Health-care Claims for Bronchiectasis



Weycker, et al. *Clin Pulm Med.* 2005;12:205



Weycker, et al. *Chron Respir Dis.* 2017; 14: 277



BRONCHIECTASIS AND NTM RESEARCH REGISTRY

A COPD Foundation Initiative

NTM Lecture Series for Patients

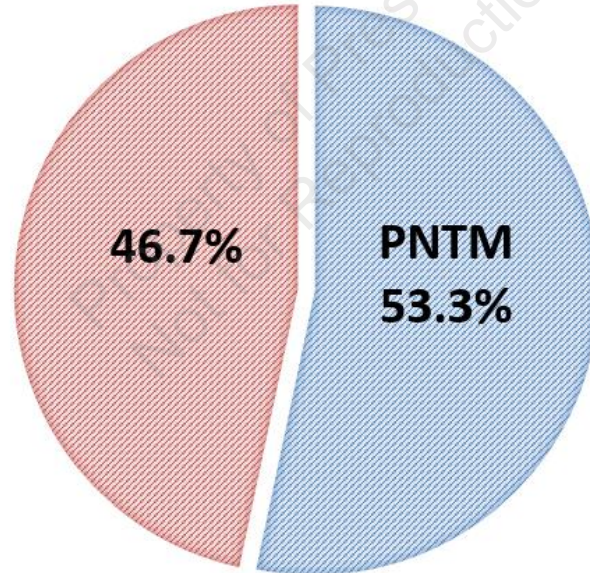
<https://www.bronchiectasisandntminitiative.org>



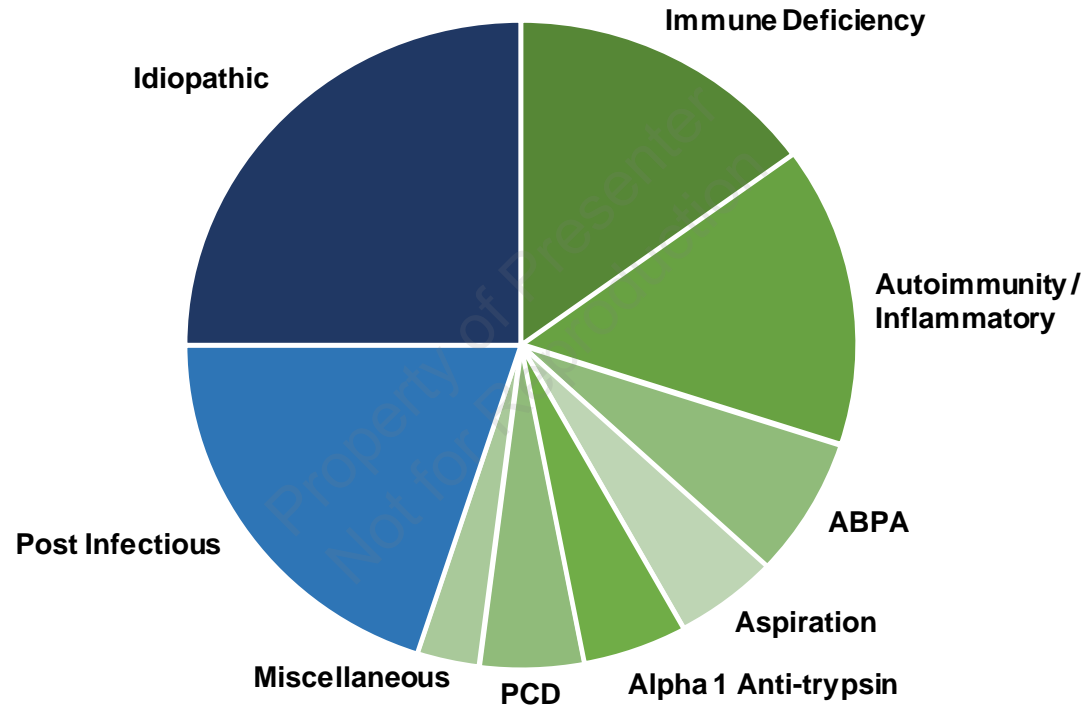


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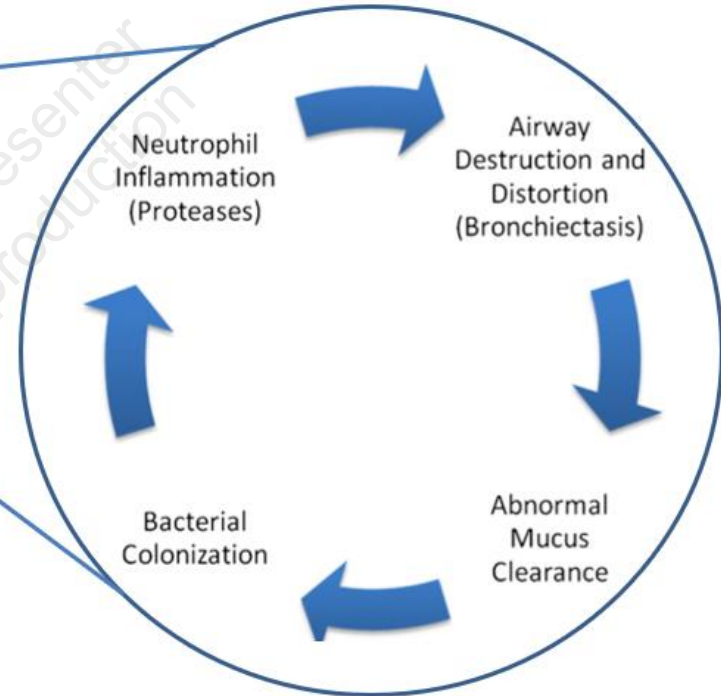


PNTM = Pulmonary Non-tuberculous mycobacteria



Etiology of Bronchiectasis

Pathophysiology of Bronchiectasis

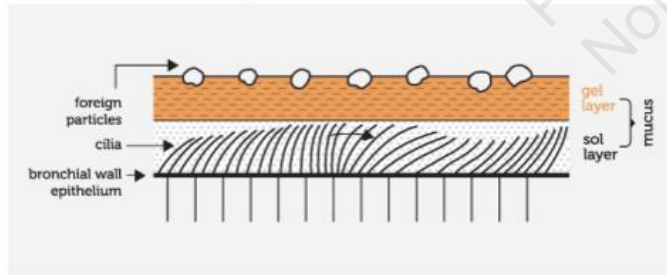


McShane, et al. *Am J Respir Crit Care Med* 2013; 188:647
Cole, *Eur J Respir Dis Suppl* 1986; 147:6

Sputum is not the same as mucus

Mucus

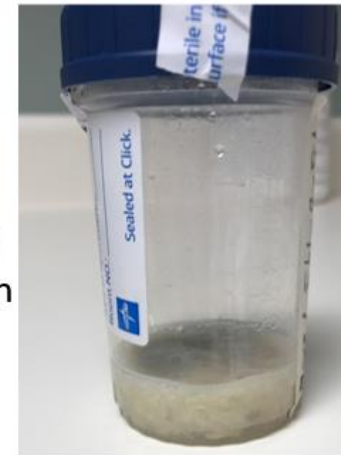
- Mucin Glycoproteins
- Antimicrobial and anti-inflammatory properties
- Cleared by cilia



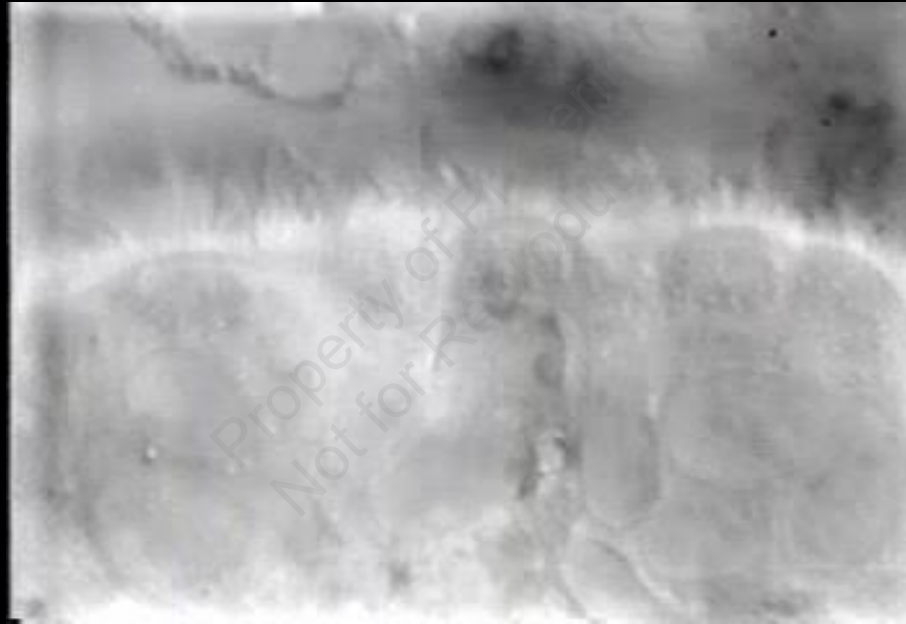
Bronchiectasis Toolbox: www.bronchiectasis.com.au

Sputum

- Large polymers that include:
 - DNA
 - Filamentous actin
 - Proteoglycans
 - Bacteria
 - Inflammatory cells
 - Impairs cilia motion



Normal Cilia – Real Time



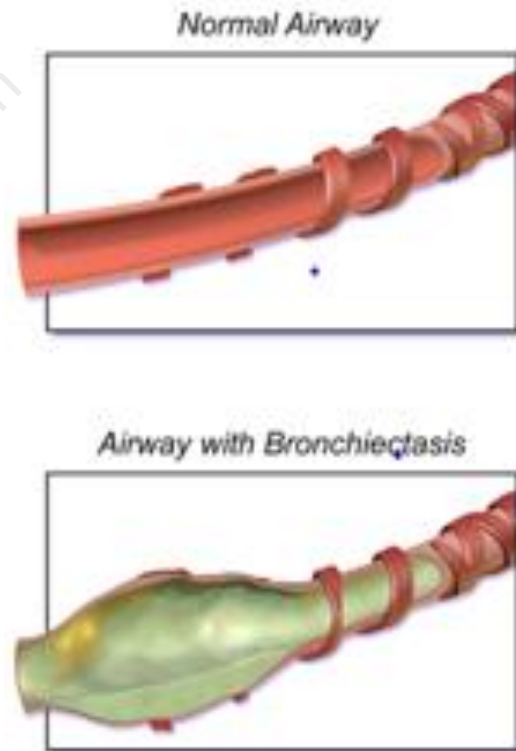
J. Carson

Airway clearance



Coughing is not effective in bronchiectasis

- Bronchial wall instability and “floppy” airways close prematurely
 - ***Expiratory flow is reduced, thereby limiting the effectiveness of the cough***



Airway Clearance Techniques

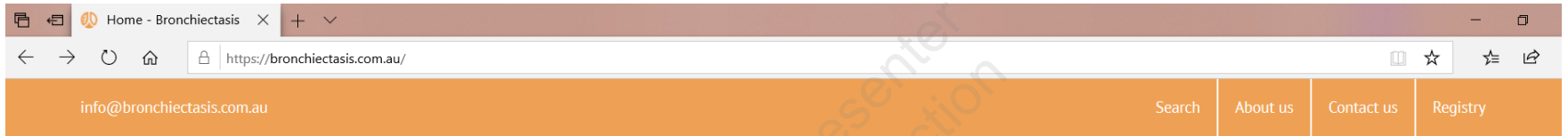
- ✓ Allow air to move behind obstruction and ventilate distal regions
- ✓ Modulate expiratory airflow in a way to propel secretions proximally up the airways

Modes of Airway Clearance

to be CUSTOMIZED to patient preference and success

<u>Breathing Techniques</u>	<u>Devices</u>	<u>Nebulized Solutions</u>	<u>Assistance</u>
Active Cycle of Breathing	Positive Expiratory Pressure (PEP) Mask	Hypertonic saline (7%, 3%)	Percussion
Autogenic Drainage	Positive Expiratory Pressure (PEP) with Oscillation	Albuterol	
Huff	High Frequency Chest Wall Oscillation (HFCWO) "Vest"	<u>Acetylcysteine</u>	
Postural Positioning			

www.bronchiectasis.com.au



[Bronchiectasis](#) [Assessment](#) [Physiotherapy](#) [Indigenous](#) [Paediatrics](#) [Resources](#)

- Principles of airway clearance
- Techniques
- Exercise

The Bronchiectasis Toolbox is a multidisciplinary resource for the management of people with bronchiectasis. The content, based on national and international guidelines, is designed to provide guidance for health professionals who are providing care to children or adults with bronchiectasis.

Endorsed by the Thoracic Society of Australia and New Zealand





- Airway Clearance in the Normal Lung
- Hydration and humidification
- Management Plan
- Choosing a technique
- Case Study

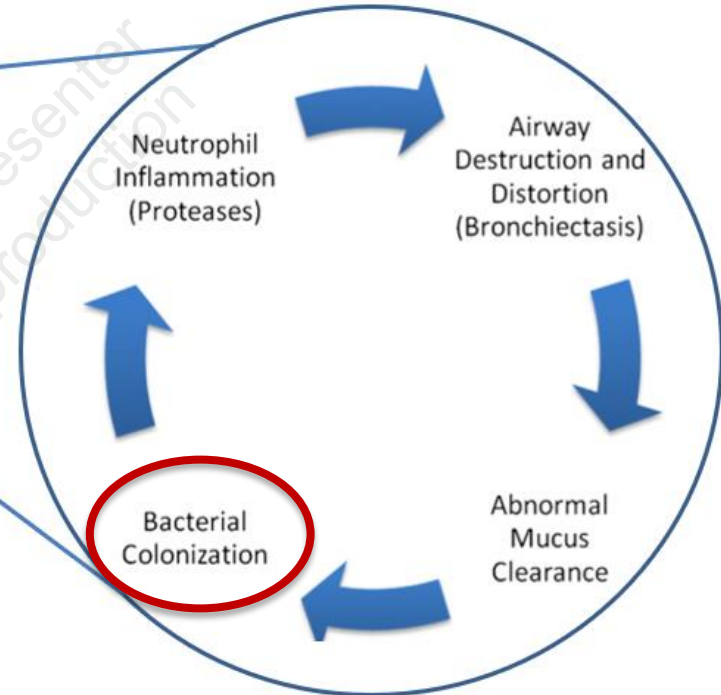


- Videos of airway clearance techniques
- The active cycle of breathing technique
- Forced Expiration Technique
- Positive Expiratory Pressure Therapy
- Oscillating Positive Expiratory Pressure Therapy
- Autogenic Drainage
- Gravity Assisted Drainage
- Manual Techniques
- Inhalation Therapy via a Nebuliser
- Afflovest
- Expiration with an open glottis in the lateral posture



- Why prescribe exercise in bronchiectasis
- Exercise prescription

Pathophysiology of Bronchiectasis



McShane, et al. *Am J Respir Crit Care Med* 2013; 188:647
Cole, *Eur J Respir Dis Suppl* 1986; 147:6

Nebulized Antibiotics for *Pseudomonas*

- Tobramycin
 - Colisitin
 - Gentamicin*
 - Aztreonam
 - Ciprofloxacin
- ✓ Reduce bacterial load

* The only inhaled antibiotic shown to reduce exacerbations in study

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 - ✓ Variable improvement in Quality of Life
 - Probably due to inadequate quality of life tools!

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 - Probably because study subjects have been too heterogenous

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 - ✓ Have not *consistently* reduced exacerbations
 - ✓ **NOT FDA approved!!**

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Arikayce – FDA approved for REFRACTORY pulmonary MAC disease



Diagnosis of pulmonary NTM

Symptoms:

Cough

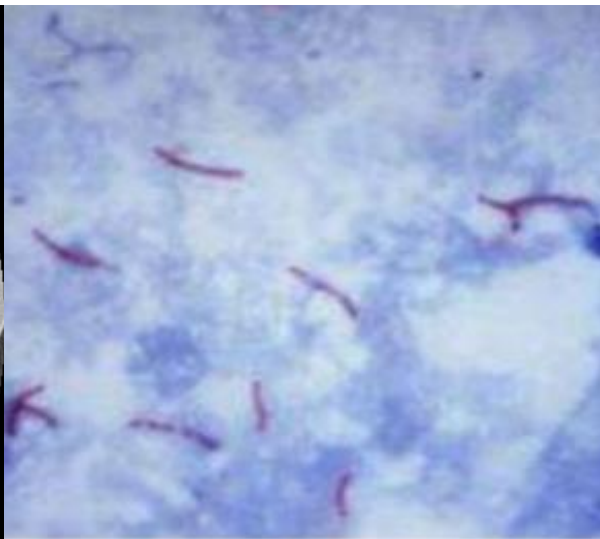
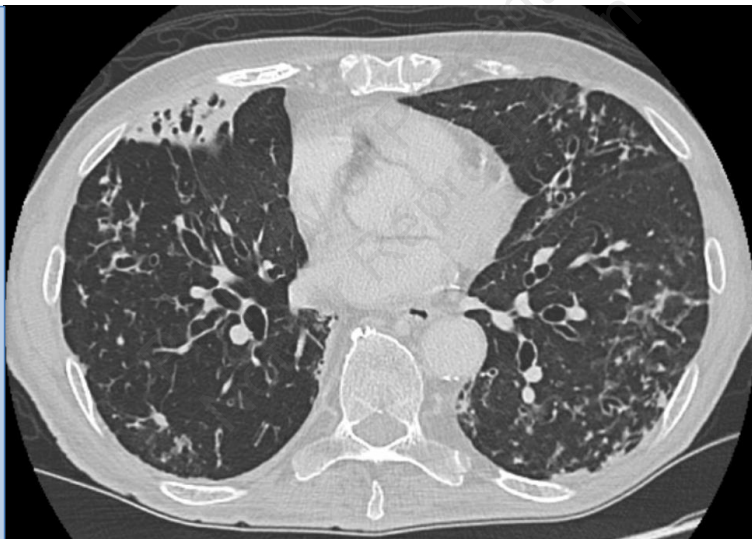
Sputum production

Fatigue

Weight loss

Coughing up blood

Chest discomfort



Diagnosis of pulmonary NTM

Colonies of Mycobacteria on Ogawa's medium



M. avium



M. kansasii



M. goodii

Whole-exome sequencing for NTM

1. PNTM patients *and* unaffected family members vs. controls:
 - More changes in CF, cilia, & connective tissue genes
2. What about “unaffected” family members?
 - Many had bronchiectasis and/or overlapping physical features
3. More *immune gene* changes only in PNTM affected

Whole exome data support:

- “Susceptible persons” model of PNTM disease
- The more “mild” changes in relevant gene categories, the greater risk of developing bronchiectasis and PNTM

Thank you



Mias using the nebulizer (credit: Denver Zoo)