



# Epidemiology of Bronchiectasis

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*GEORGETOWN UNIVERSITY*

# Disclosures

- Principal Investigator/Grant support for clinical trials
  - Insmed (inhaled liposomal amikacin)
  - Bayer (inhaled ciprofloxacin)
  - Aradigm (inhaled liposomal ciprofloxacin)
  - Parion (inhaled mucolytic for PCD)
- Foundation support for Bronchiectasis Registry
  - COPD Foundation
- Consultant
  - Novartis
  - Raptor/Horizon
  - Xellia
  - Bayer
  - Electromed
- NO FDA Approved therapies

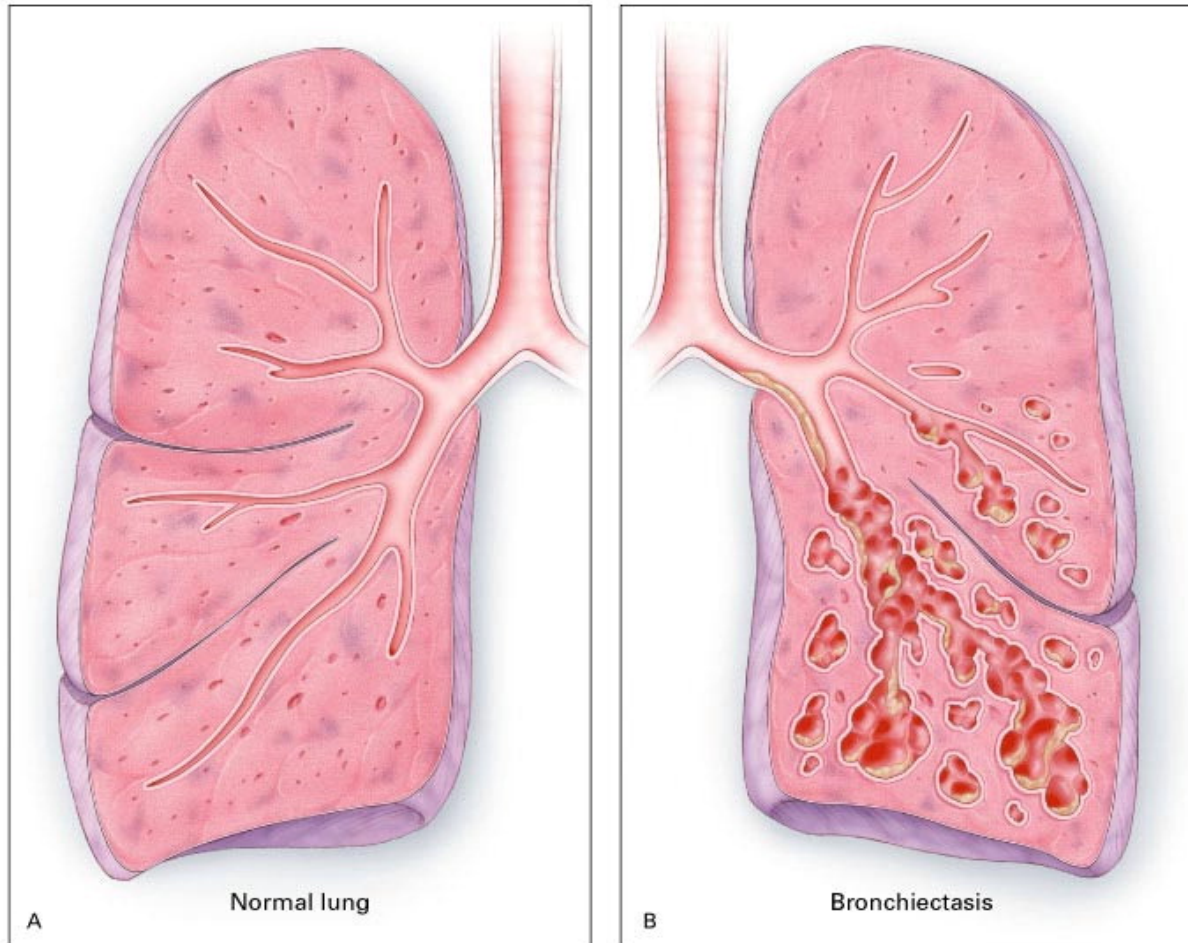
# Bronchiectasis “FAQ’s”

- **What is bronchiectasis?**
- **Why do I have it?**
- **Who else has it?**
- **Why is it increasing?**
- **Do I have COPD?**
- **What is the difference between bronchiectasis and NTM?**

# What is bronchiectasis?

- Abnormally dilated bronchi/bronchioles which leads to:
  - impairment of local host defenses
  - Chronic colonization with bacteria
  - Vicious cycle of airway inflammation and infection.
- Hence, an **anatomic** abnormality
- But, also a **disease state**
- **We diagnose it with high resolution CT scan**
- Symptoms
  - Chronic cough
  - Recurrent respiratory infections
- We are not including CF today.....
  - Multiple etiologies

# Normal Lung and Airways and the Lung of a Patient with Bronchiectasis



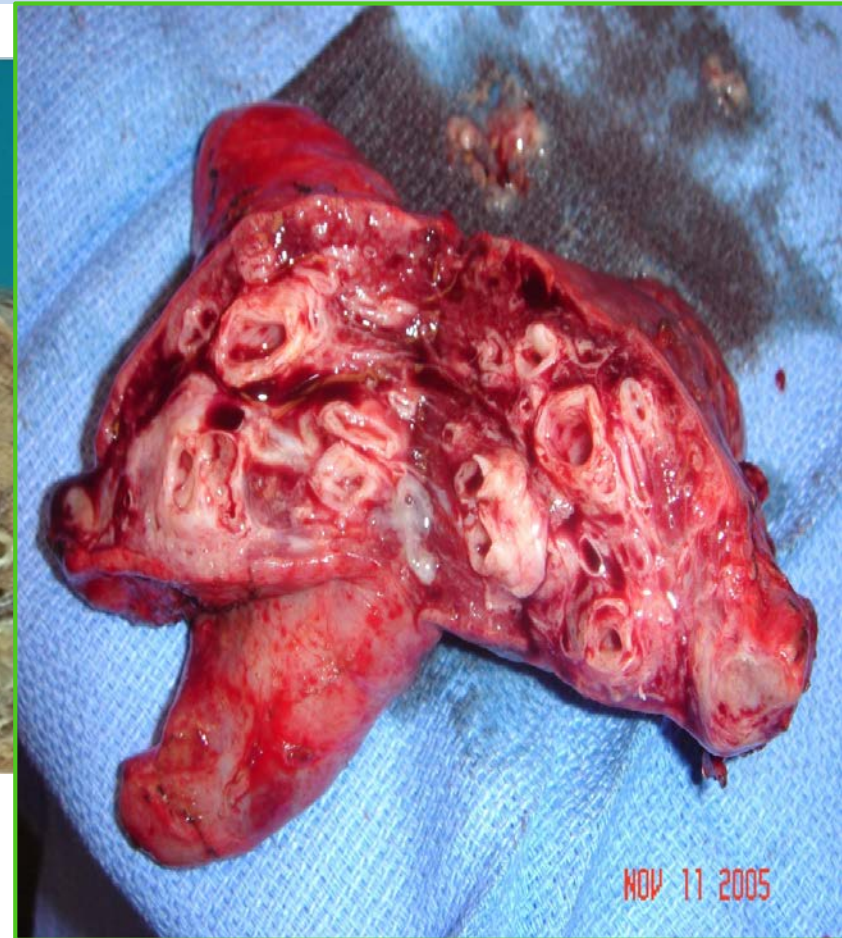
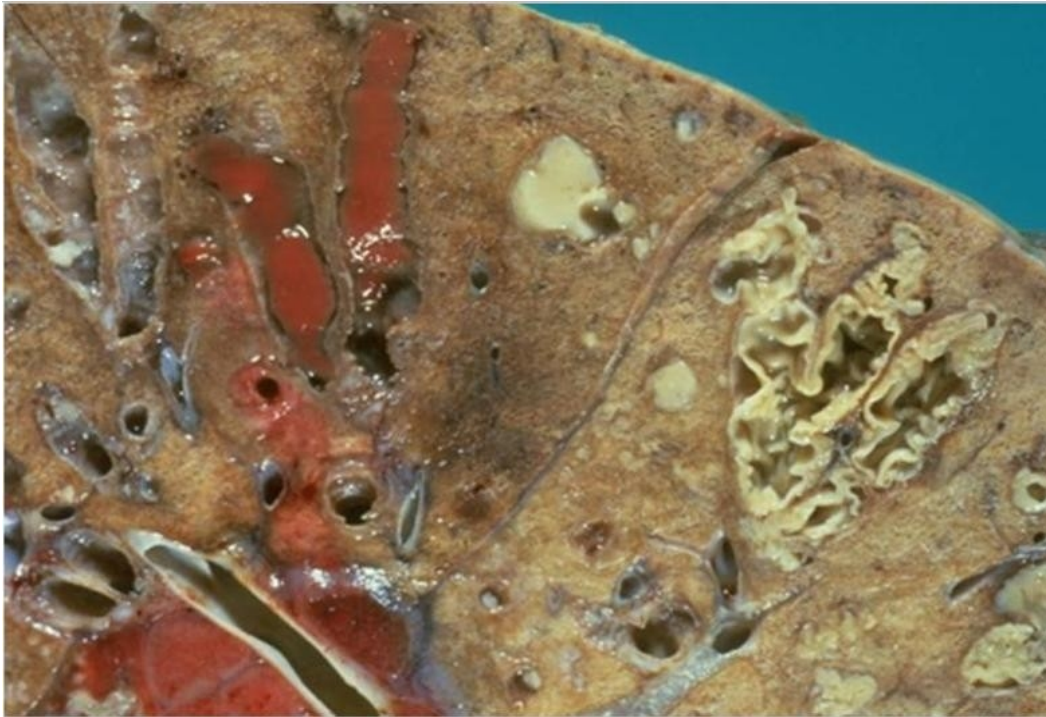
**Bronchiectasis**  
**Alan F. Barker, M.D.**  
**N Engl J Med 2002;**  
**346:1383-1393**



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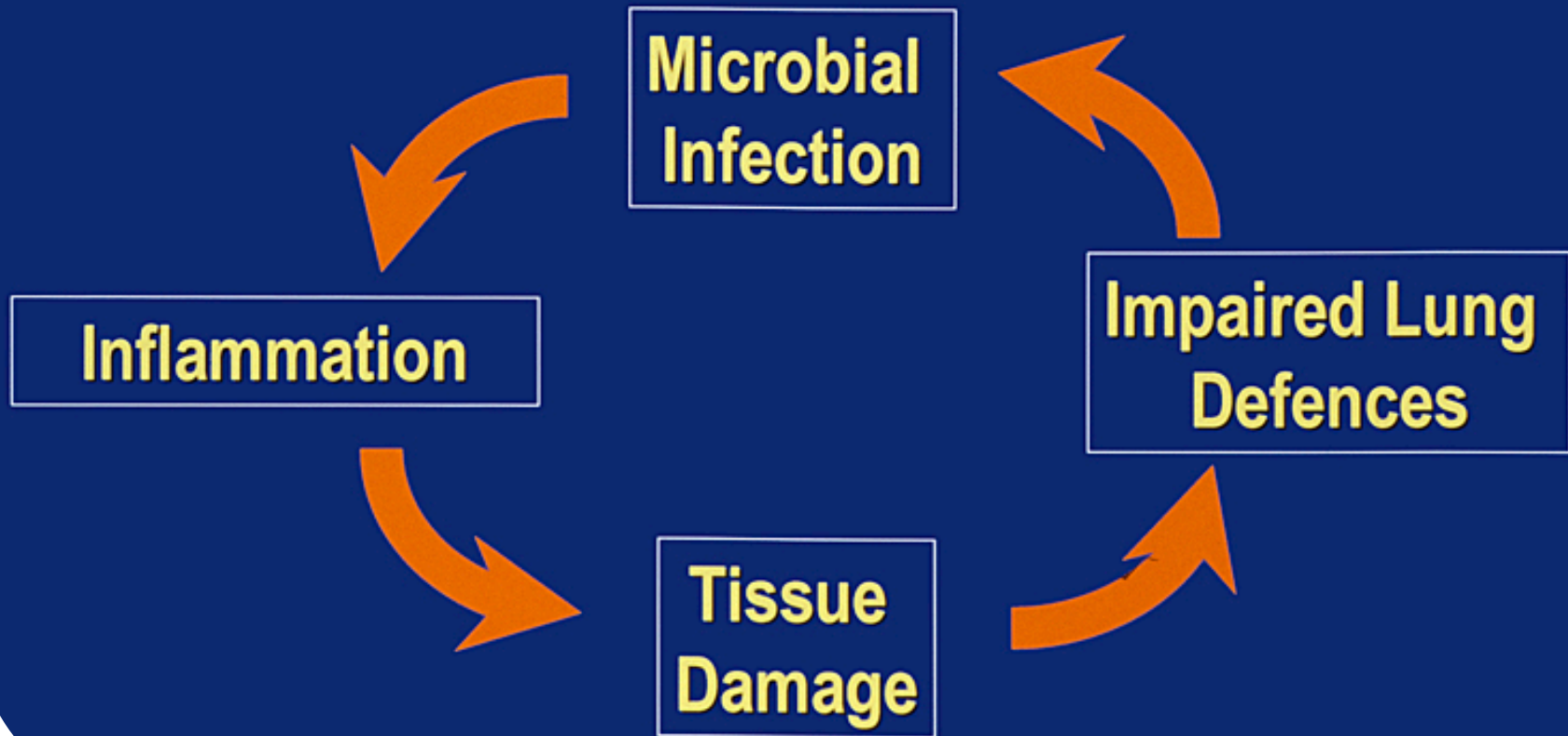
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# Bronchiectasis

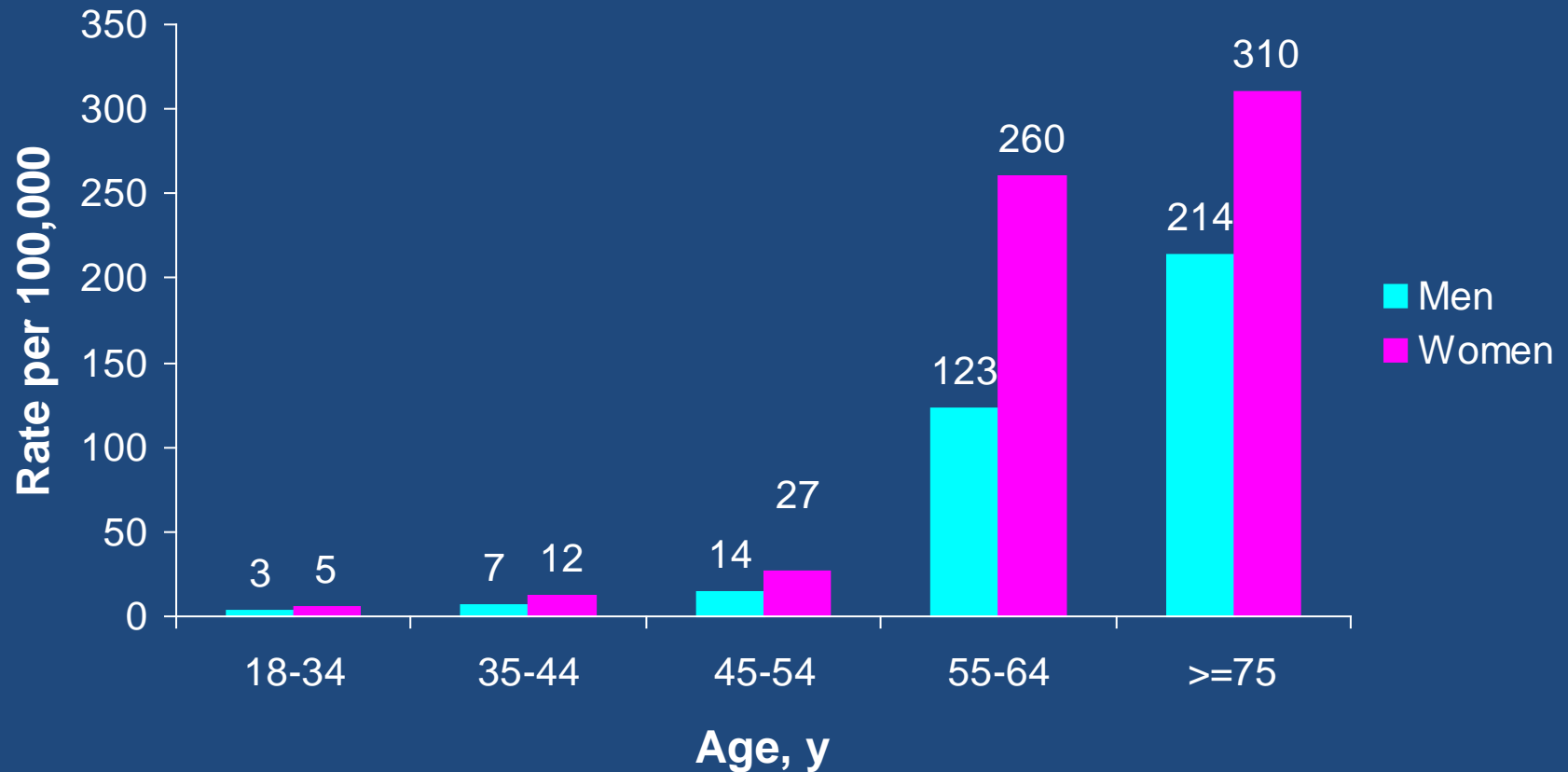


Courtesy of G. Huitt MD

# A VICIOUS CYCLE OF INFECTION AND INFLAMMATION



# Prevalence of Non-CF Bronchiectasis USA



Estimated US prevalence: 52 cases per 100,000

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



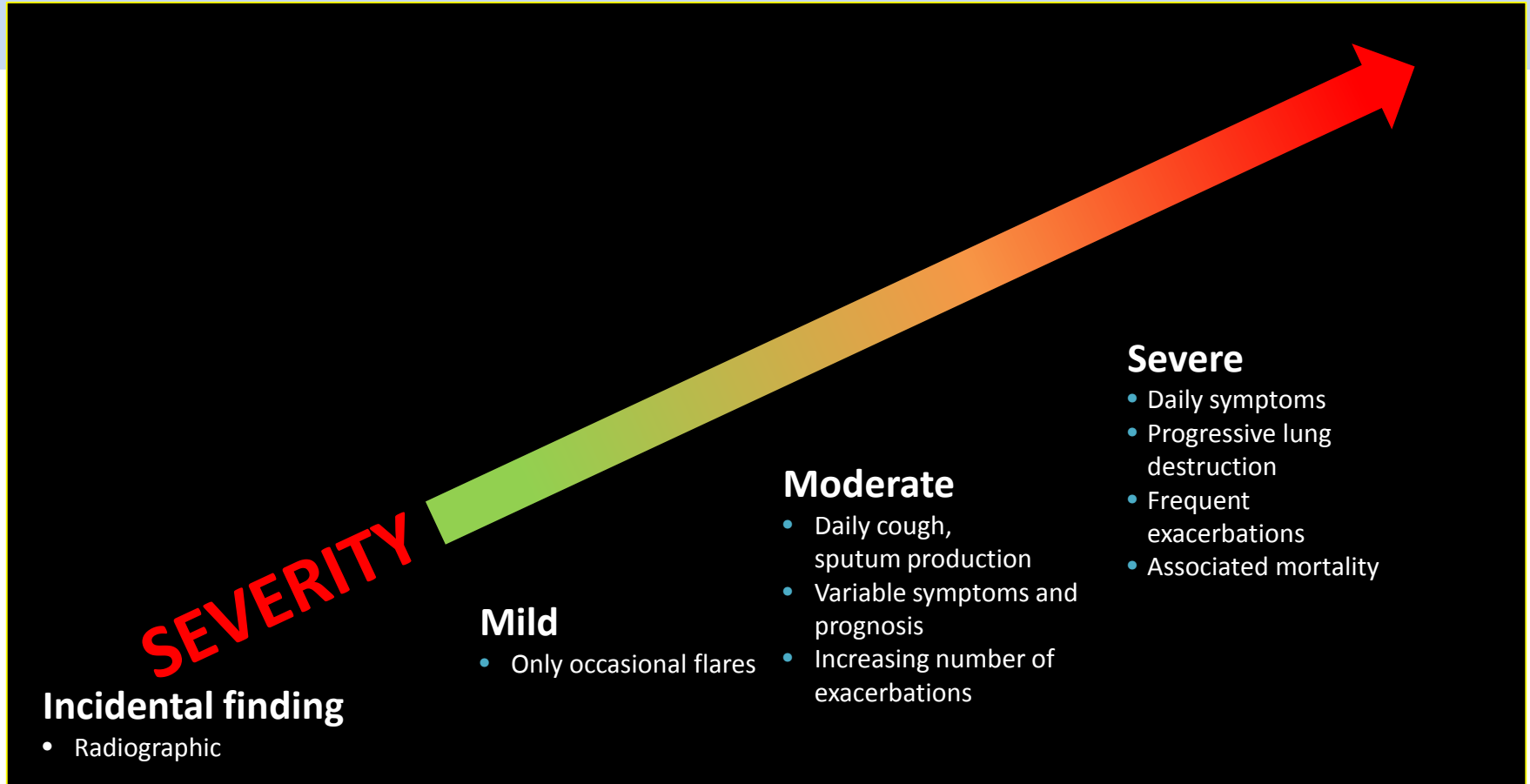
# Prevalence of NCFB Worldwide

- **Germany<sup>1</sup>**
  - 2013: 0.07%
  - Highest rate in men aged 75-84 years
- **United Kingdom<sup>2</sup>**
  - 2013: 0.56% in women, 0.49% in men
  - Increased prevalence observed between 2004 and 2013
- **Europe<sup>3</sup>**
  - Unknown prevalence
  - Data gathering underway: EMBARC European Bronchiectasis Registry
- **China<sup>4</sup>**
  - 2002-2004: 1.2% of individuals greater than 40 yrs
  - More men than women
  - “Not an orphan disease”
- **Republic of Korea<sup>5</sup>**
  - 2008: 9.1% in a computed tomography screened “healthy” population

# Why is the prevalence increasing?

- More patients actually have it
- Better diagnostic tools
  - CT chest
- More recognition by clinicians
- But there is often a delay in diagnosis
  - Cough is a non specific symptom
  - Antibiotics are an “easy” solution
  - Sputum cultures are not commonly done in primary care practices in US

# Heterogeneous disease



1. Chalmers JD, et al. *Am J Respir Crit Care Med*. 2014;189:576-85.
2. Lonni S, et al. *Ann Am Thorac Soc*. 2015;12:1764-70

# US NCFB Registry Data

TOTAL ENROLLEES N=2000	
Gender	79% female
Age, median	64 years
Race/ethnicity	89% non-Hispanic white 7% Black 4% Asian
Mean BMI	23.2 kg/m <sup>2</sup>
Smoking	60% never 38% former 2% current
ENT co-morbidities	25%
Age at diagnosis, median	57 years



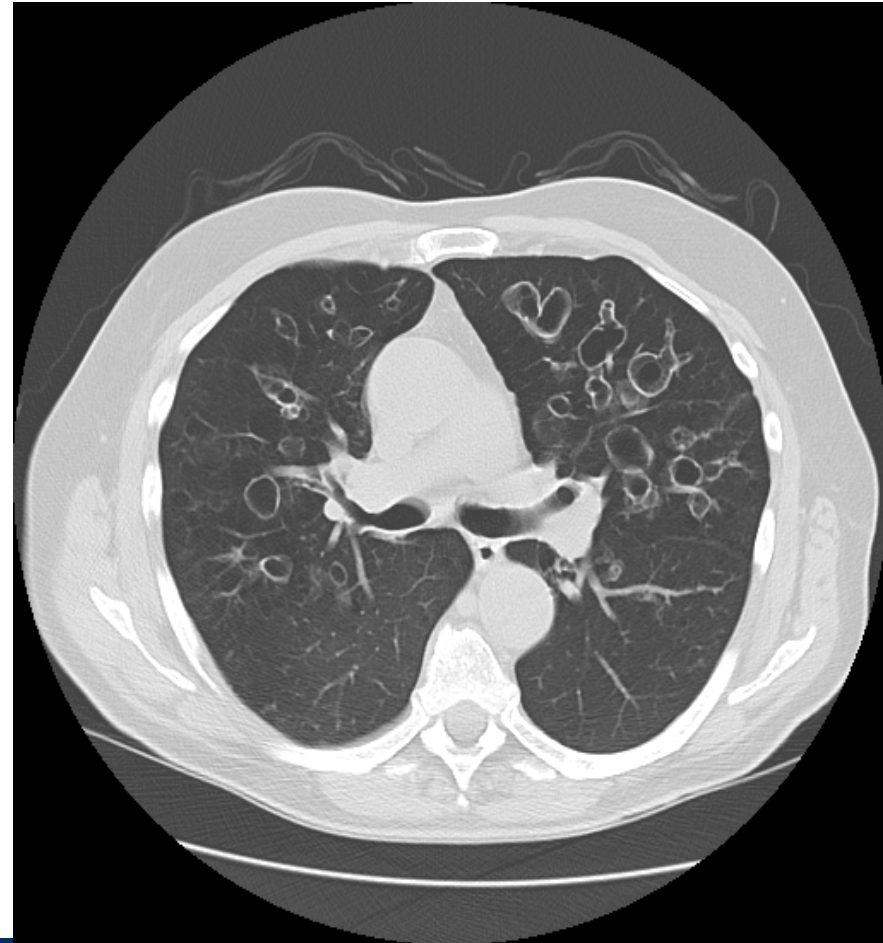
**BRONCHIECTASIS AND NTM**  
**RESEARCH REGISTRY**  
*A COPD Foundation Initiative*

1. US NCFB registry data, per A.E O'Donnell, Georgetown University, Washington, D.C.
2. Aksamit T et al. CHEST 2017;151: 982-992

# Why do I have bronchiectasis?

- No underlying disease
  - Idiopathic bronchiectasis
- “Post infectious”
- Immunologic deficiencies/abnormal “host”
- Rheumatologic abnormalities
- Congenital abnormalities
  - Alpha one anti-trypsin deficiency
  - Primary ciliary dyskinesia
  - Cystic fibrosis
- Aspiration

# Focal vs Diffuse Bronchiectasis



# Focal vs diffuse bronchiectasis

## Focal

- Anatomic abnormalities
  - Post infectious scarring
  - Airway obstruction
    - Tumor
    - Foreign body
- Aspiration
  - Neurologic disorders
  - Prior head and neck cancer

## Diffuse

- Pulmonary only disease
  - Prior infection
  - Prior inhalation injury/aspiration
  - Asthma/COPD
- Sino-pulmonary disease
  - Congenital etiologies
  - Immunodeficiency
- Other systemic diseases
- Idiopathic

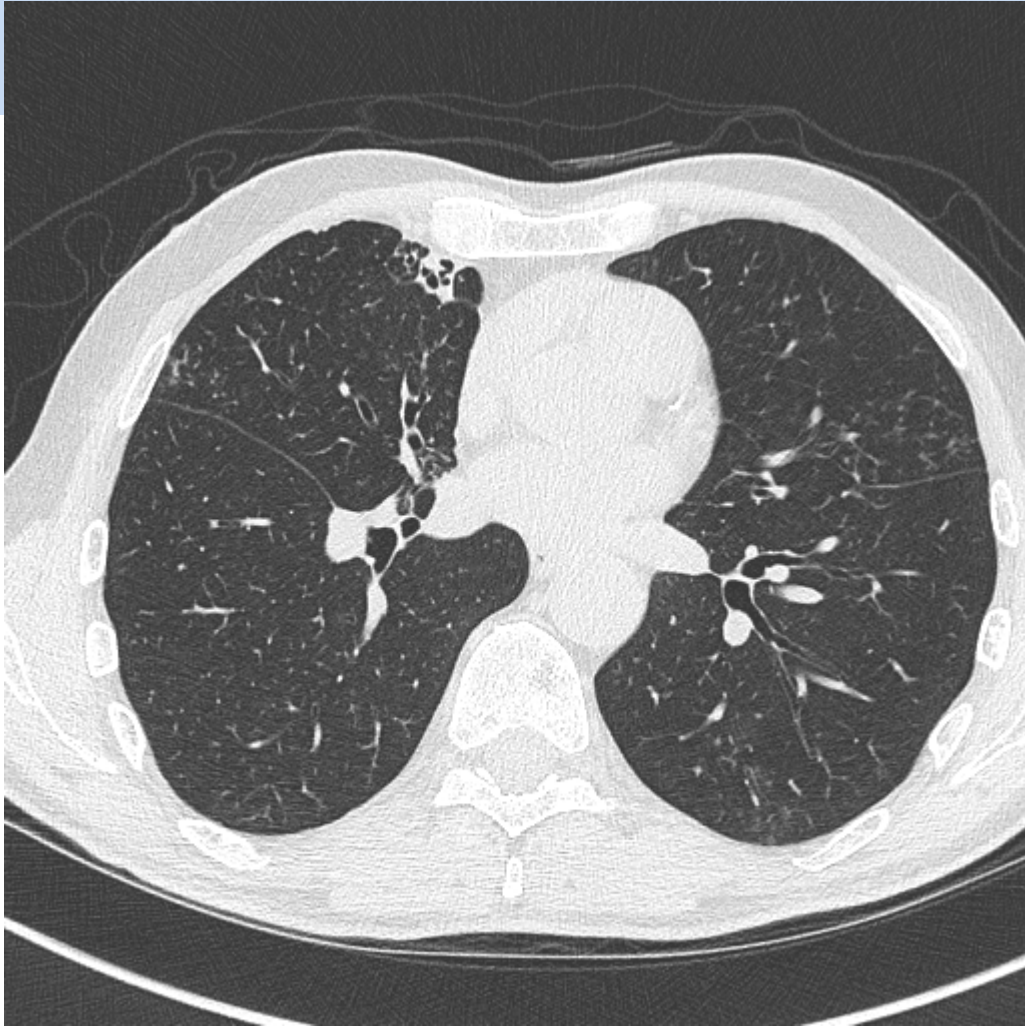
# Evaluation for focal and diffuse bronchiectasis

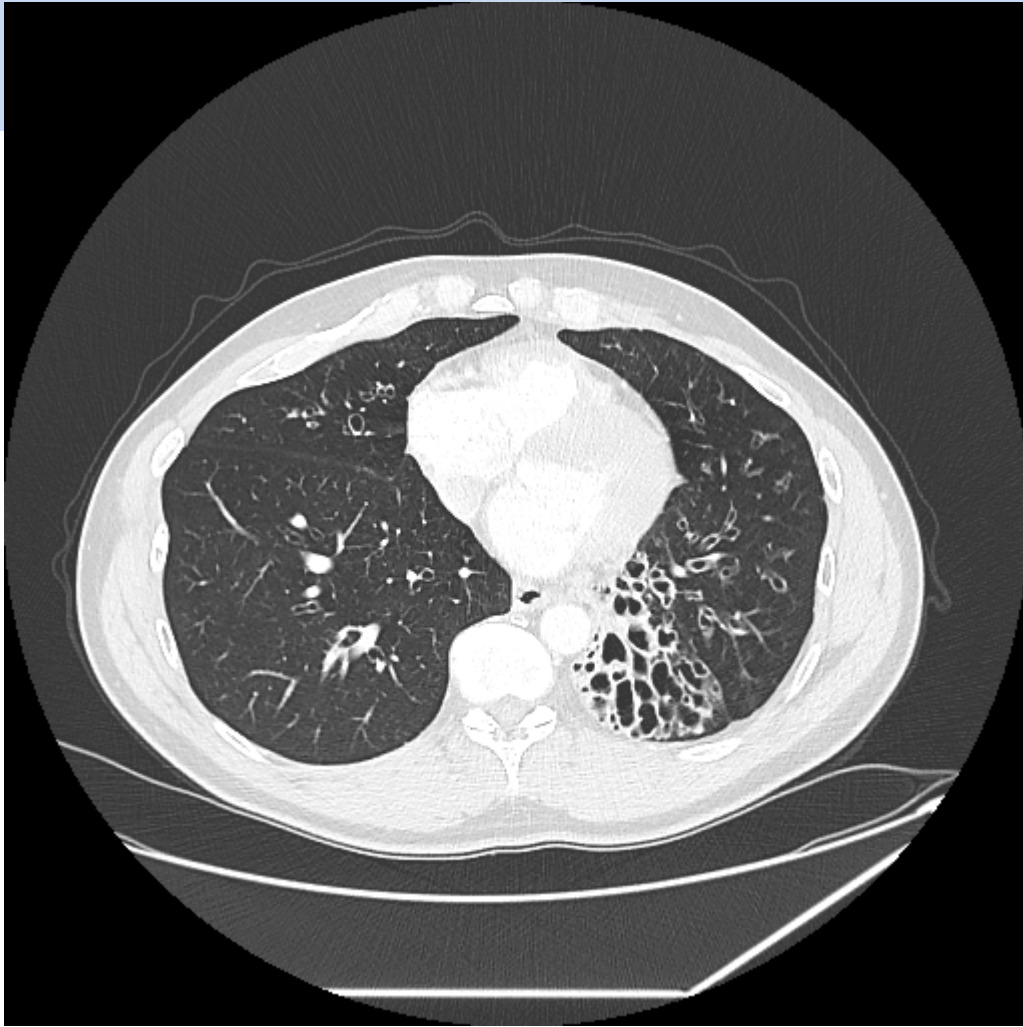
- Overactive or underperforming immune system
  - Immunologic evaluation
    - IgG, IgA, IgA, Ig E
    - HIV testing
- Evaluation for other immunologic disorders
  - Allergic bronchopulmonary aspergillosis
  - Rheumatoid arthritis
  - Sjogren's syndrome
  - Inflammatory bowel disease
- Structural work up
  - Bronchoscopy
- Evaluation for reflux/aspiration

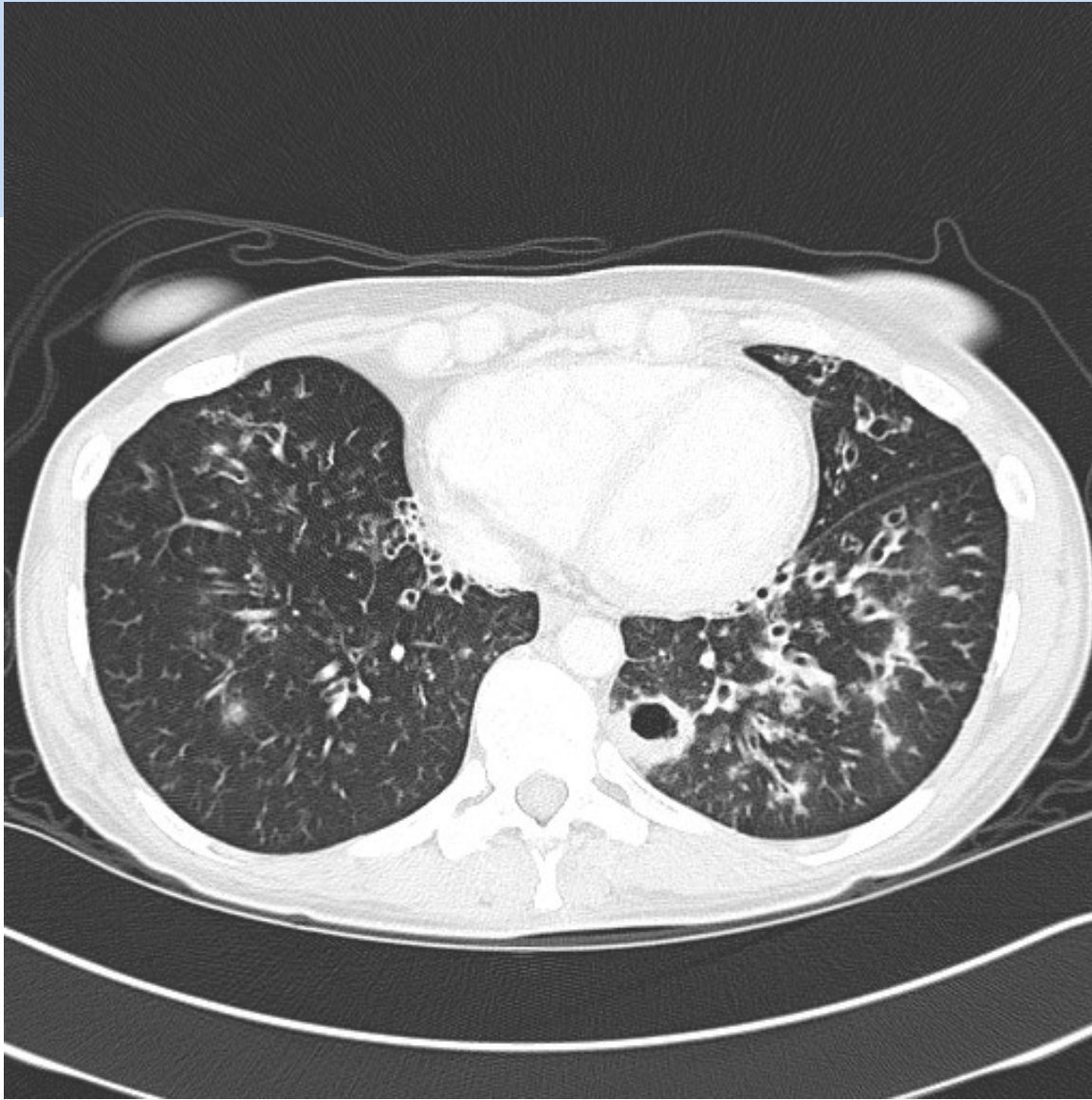


# Evaluation of focal or diffuse bronchiectasis

- Does the patient need a genetic work up?
  - AAT deficiency
  - Evaluation for cystic fibrosis
    - Sweat test
    - Genetic evaluation
    - Co-morbidities
    - Specific microbiologic findings
      - Staphylococcus aureus
      - B. Cepacia
  - Evaluation for ciliary dysfunction
    - History
      - Neonatal respiratory distress
      - Ear and sinus problems
      - Infertility
    - Genetic testing







# Do I have COPD?

- COPD and bronchiectasis are not the same
- COPD
  - Smoker's disease
  - Occasionally COPD patients develop bronchiectasis
  - COPD patients with significant cough and sputum production should be evaluated for bronchiectasis
- Many bronchiectasis patients are misdiagnosed
  - COPD
  - Bronchitis
  - Asthma
  - pneumonia

# Bronchiectasis and NTM

- Bronchiectasis is the anatomic abnormality
  - NTM is one type of infection that occurs in BE
    - Other bacteria may also be present
    - Pseudomonas/ other gram negatives and gram positives
  - Fibrocavitary vs nodular bronchiectasis
    - Fibrocavitary: NTM is a consequence of BE
    - Fibronodular: NTM may be the cause
  - Fibrocavitary: men and women
  - Fibronodular: female predominant

# Fibronodular vs fibrocavitary



# What makes me cough and produce sputum?

- Routine microbiology
  - Pseudomonas
  - Other gram negatives
  - Staphylococcus/streptococcus
  - nocardia
- Non tuberculous mycobacteria
- Fungi
- Country/region specific microbiology



# Is there a blood test for my disease?

- Is there a test that can predict exacerbations?
  - No
- What about a test to assess bacterial load?
  - Not perfect
- A prognosticator biomarker?

EVIDENCE is lacking

# What is my prognosis?

- Is my disease going to progress?
  - Perhaps
  - We have some predictors regarding outcomes
- How do I live better with my disease?
  - We will get to that
- Can I be cured of bronchiectasis?
  - Probably not, though surgery can be curative for some

# What you need to ask your physician

- Confirm bronchiectasis
- Discuss an evaluation for causes
  - Treatable etiologies
  - Inherited diseases
- Confirm and monitor sputum cultures
  - Routine bacteria
  - NTM
- Targeted multimodality treatment
- New treatments/clinical trials

# What I have learned from my patients

- Delay in diagnosis
- Lack of good explanations regarding the disease
  - Physicians need to do better
  - Team approach needed for the disease
- Psychosocial aspects of the disease
  - Cough/sputum
  - Fear of exacerbations/progression
- Burden of treatments

# What I have learned from my patients



# Bronchiectasis

- It is the underlying disease for many patients
- Worldwide incidence/prevalence increasing
- There are multiple causes
  - But we often don't identify a cause
  - “chicken and egg” question with NTM infection
- Evaluation should be tailored to the patient
- Microbiology results important

# Resources

- NTM IR website
  - <https://www.ntminfo.org>
- Bronchiectasis tool box
  - <http://bronchiectasis.com.au>
- Bronchiectasis News Today
  - <https://bronchiectasisnewstoday.com>
- US Bronchiectasis Registry
  - <https://www.copdfoundation.org/Research/Bronchiectasis-Research-Registry/Learn-More.aspx>

3<sup>rd</sup>

# WORLD BRONCHIECTASIS CONFERENCE

*Patient Session*

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**JULY 14, 2018**

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