

# Guidelines for bronchiectasis

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# European Respiratory Society guidelines for the management of adult bronchiectasis

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@ERSpublications

The publication of the first ERS guidelines for bronchiectasis <http://ow.ly/wQSO30dU0nE>

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BRITISH THORACIC SOCIETY  
GUIDELINE FOR BRONCHIECTASIS  
IN ADULTS

British Thoracic Society  
Bronchiectasis in Adults Guideline  
Development Group

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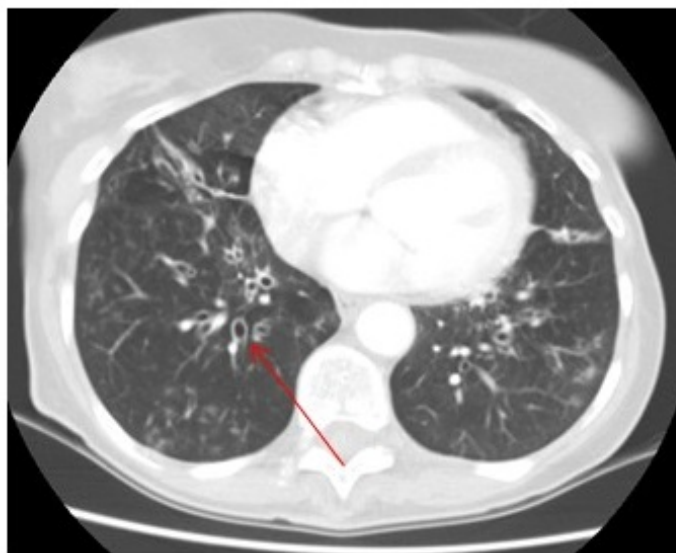
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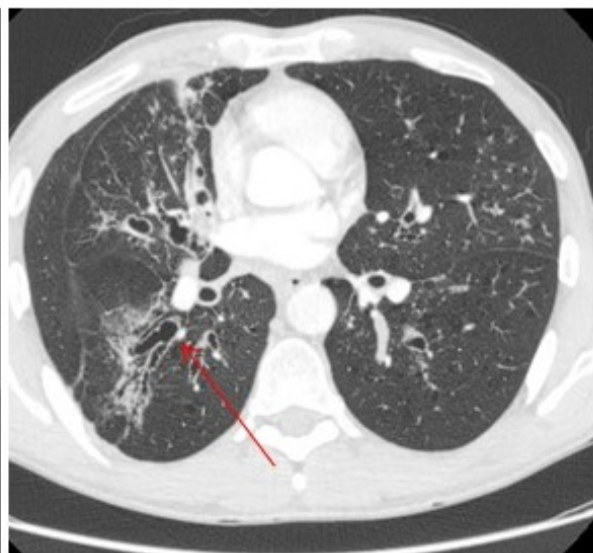
# What is bronchiectasis

A radiological finding of dilatation of the bronchi (usually on CT scan)

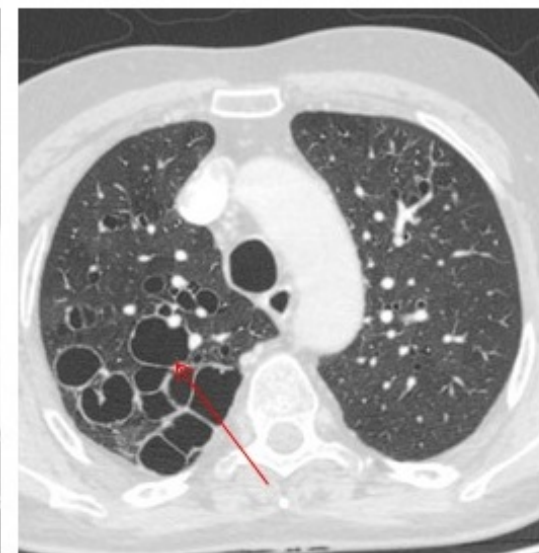
A disease associated with cough, sputum production and frequent chest infections



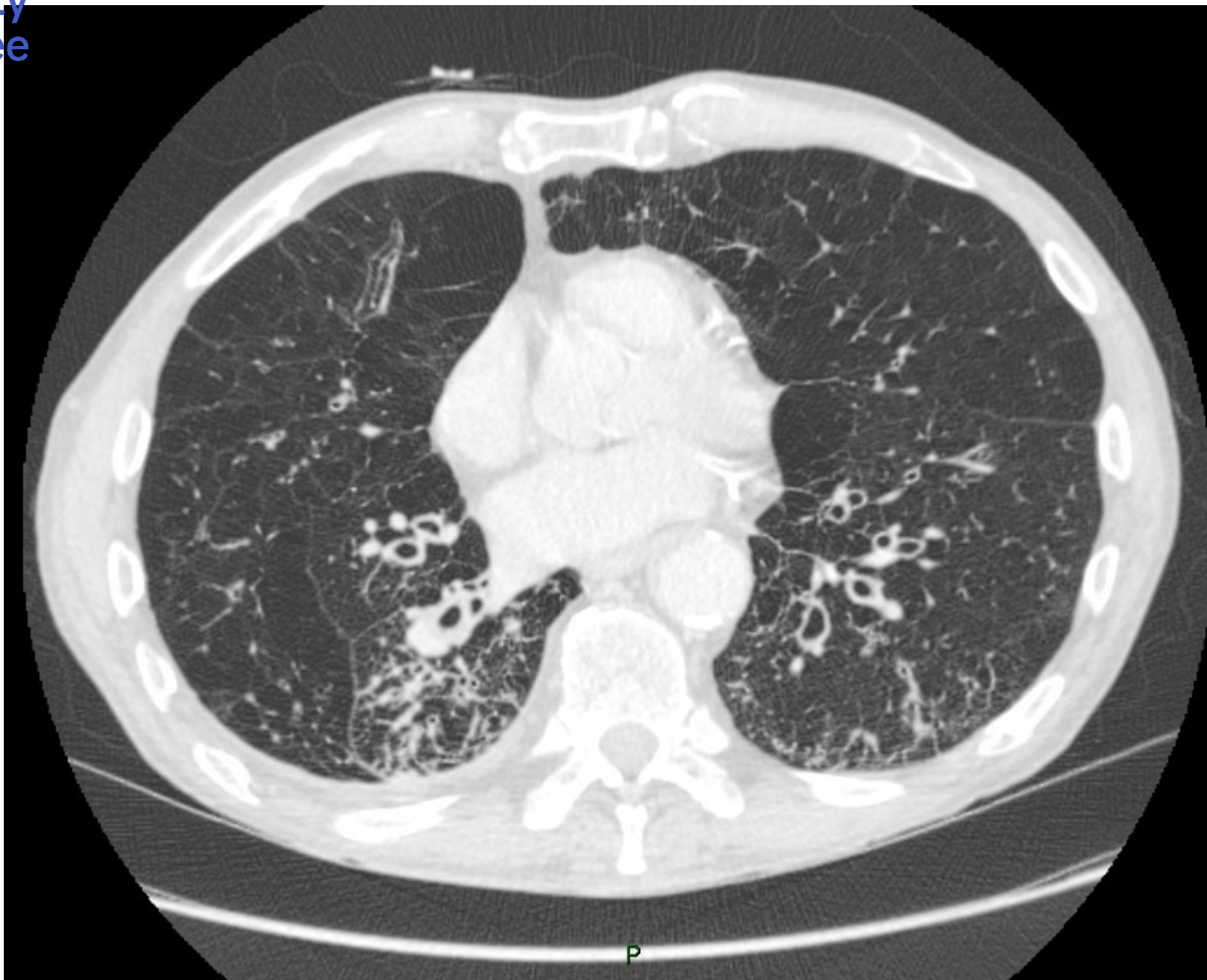
Cylindrical



Varicose



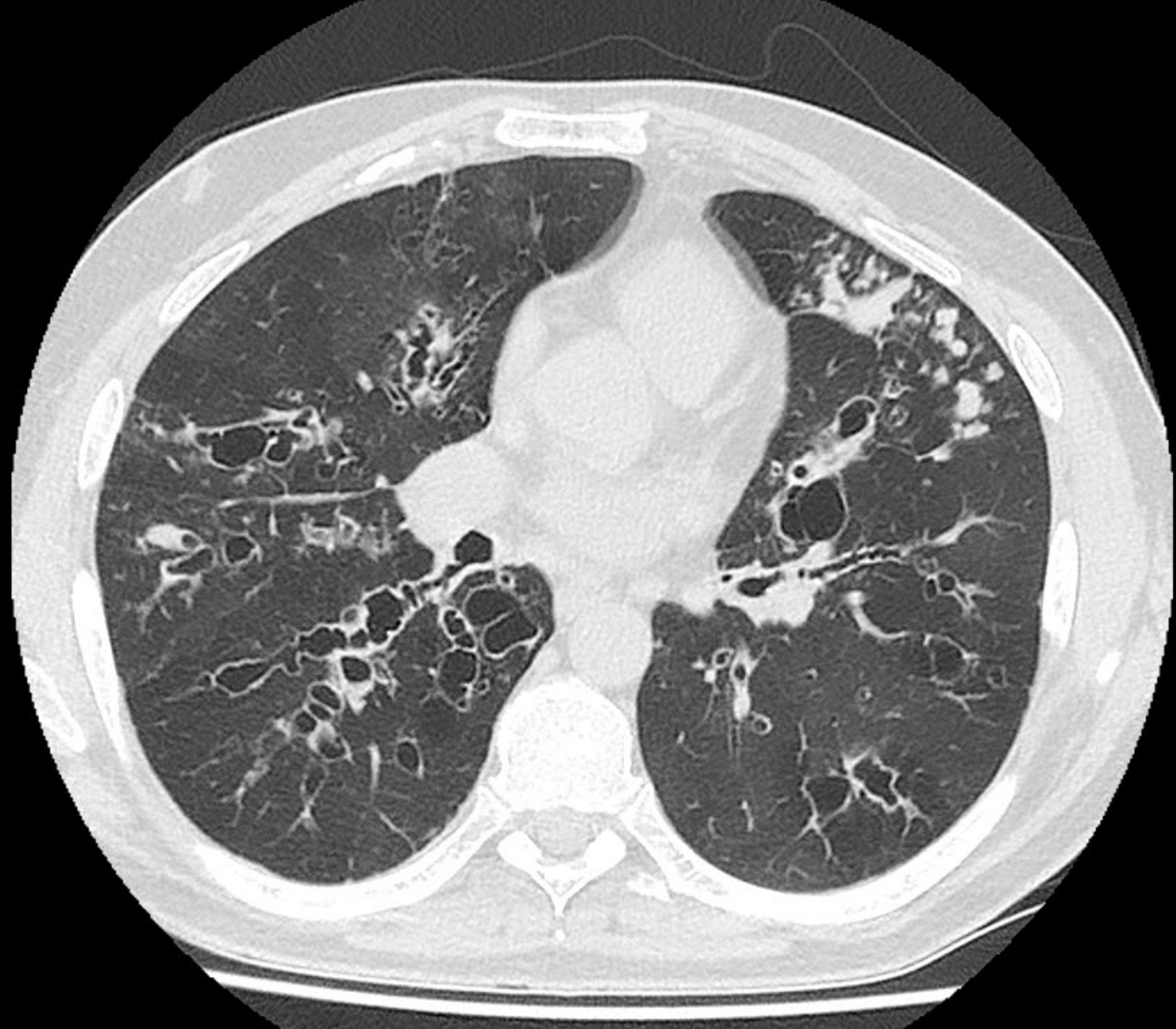
Cystic



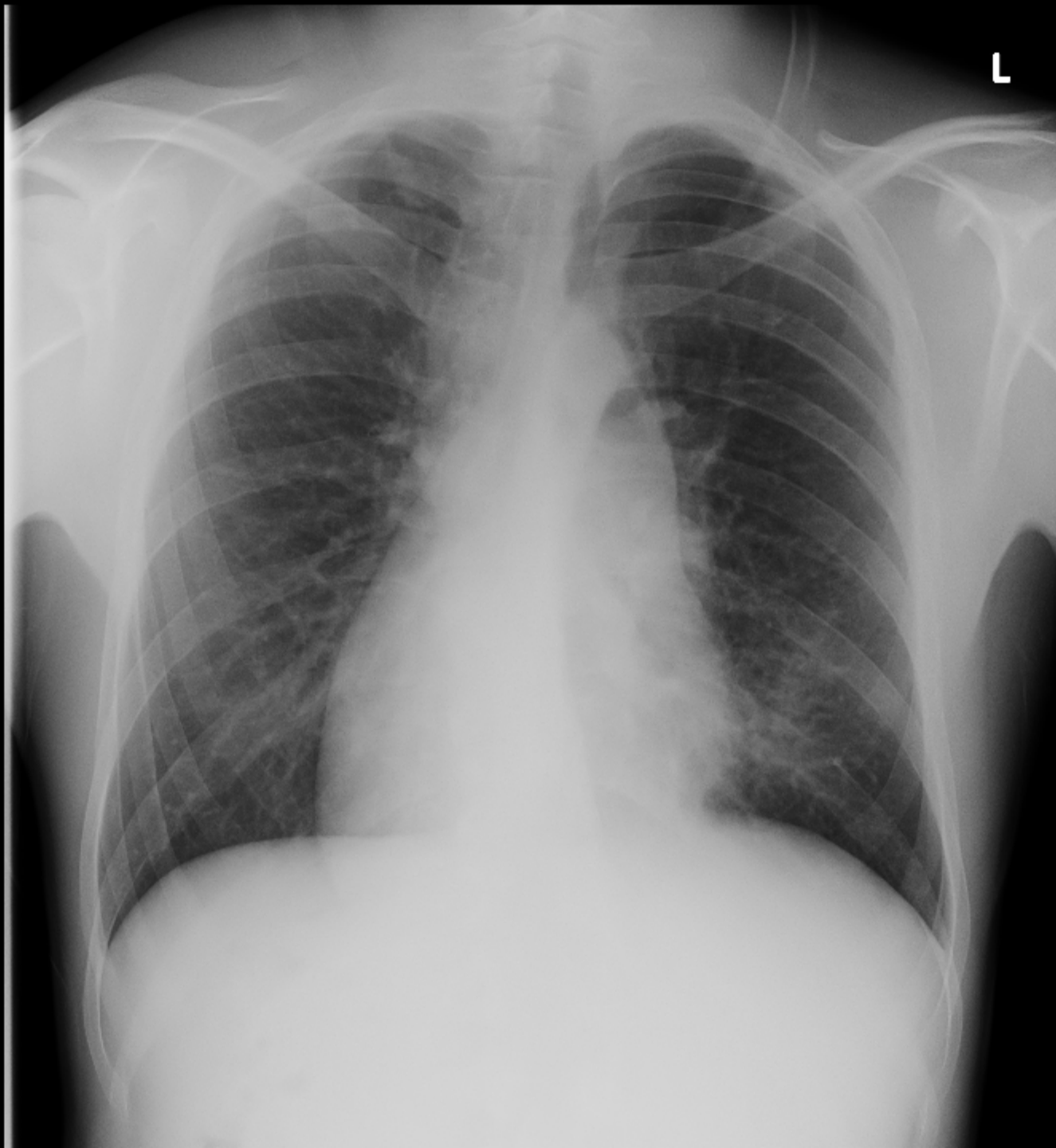


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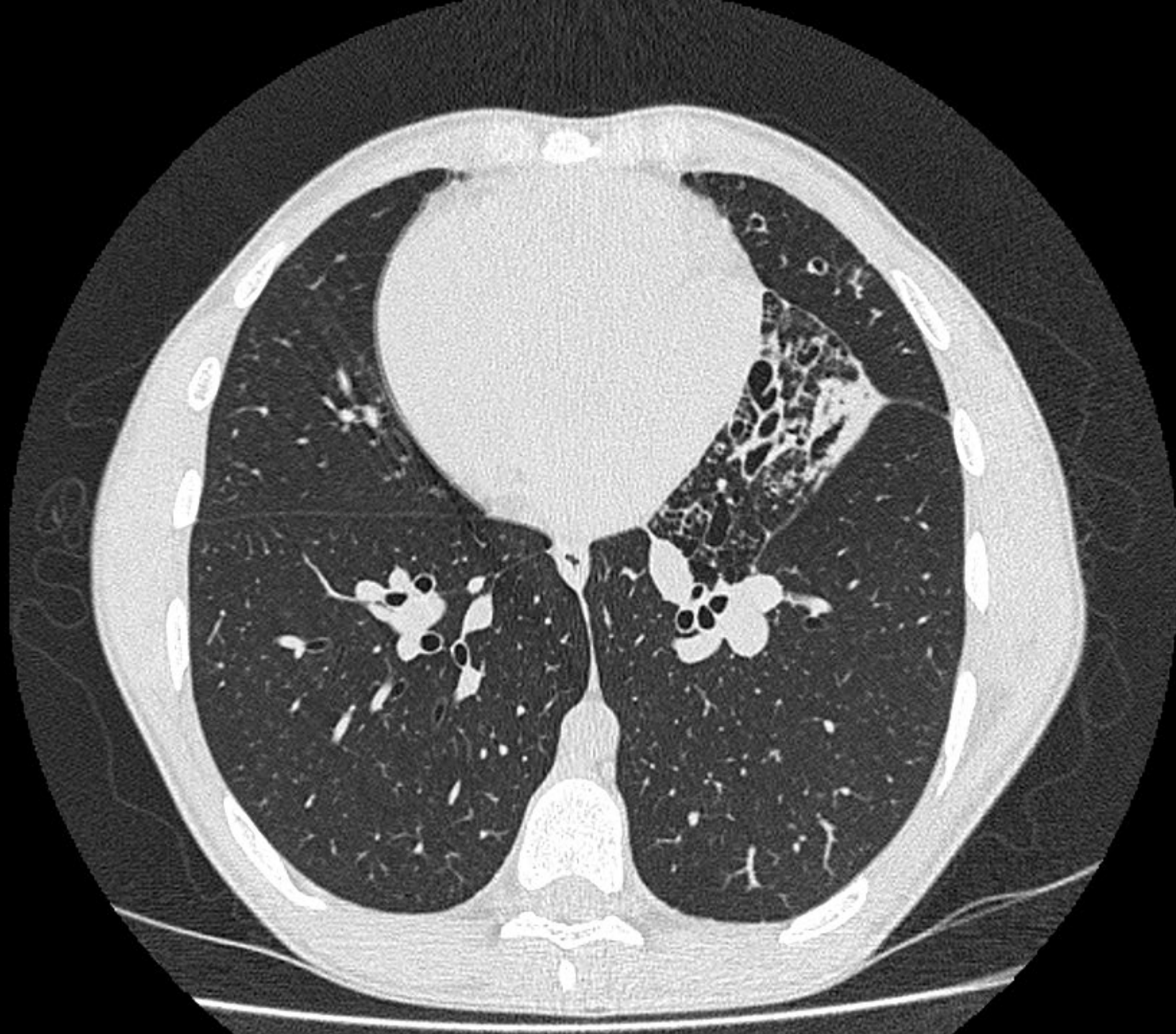
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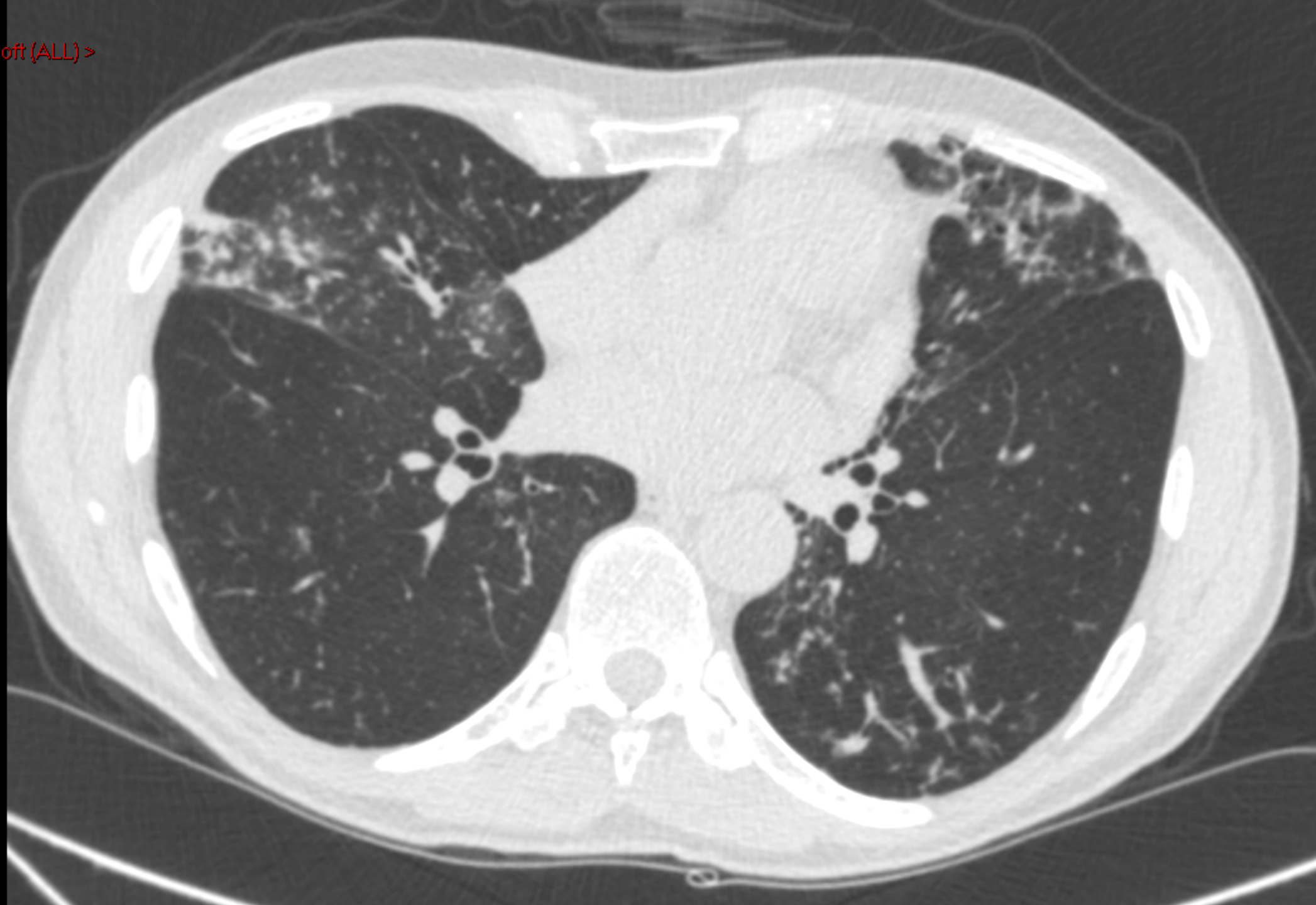


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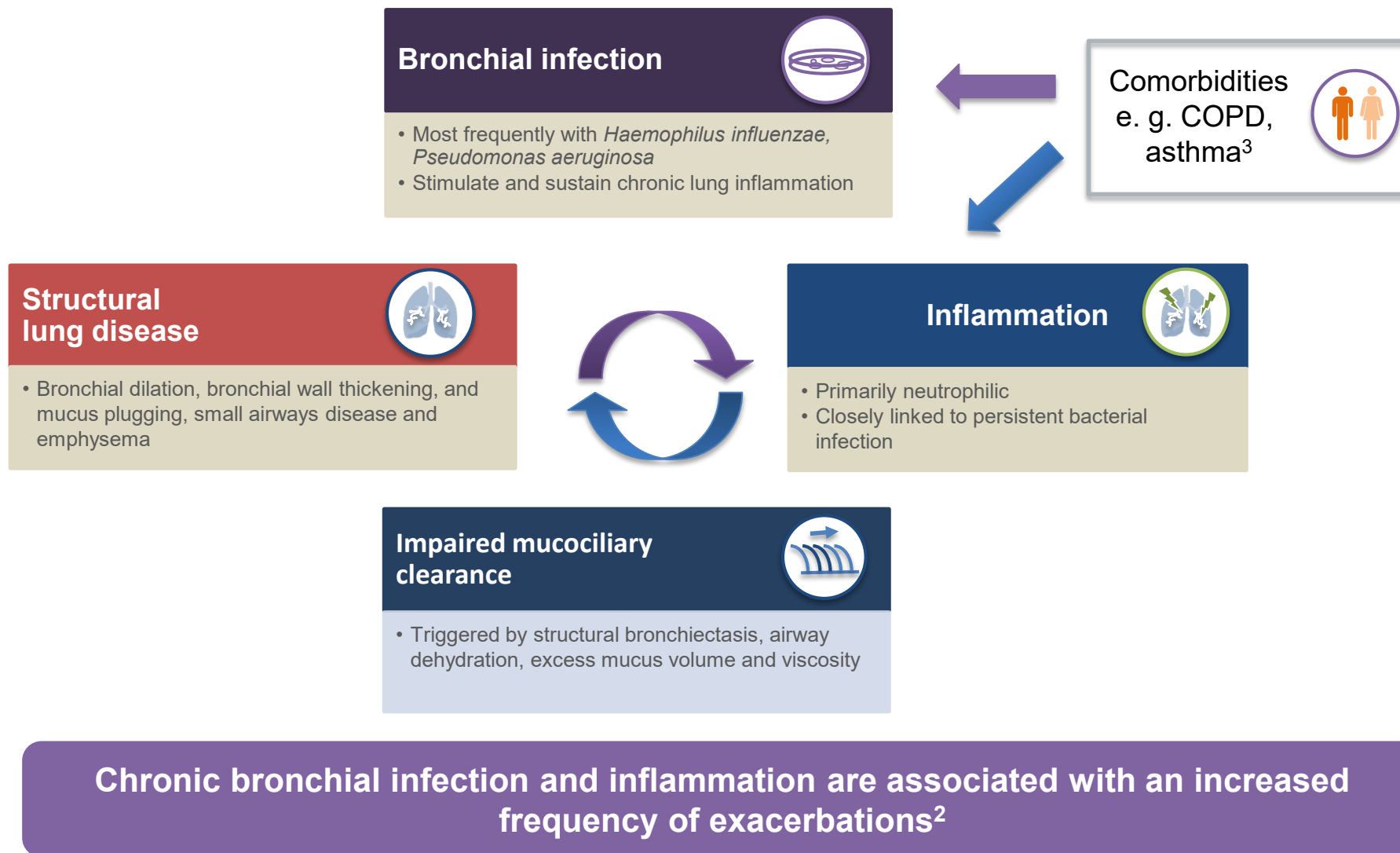




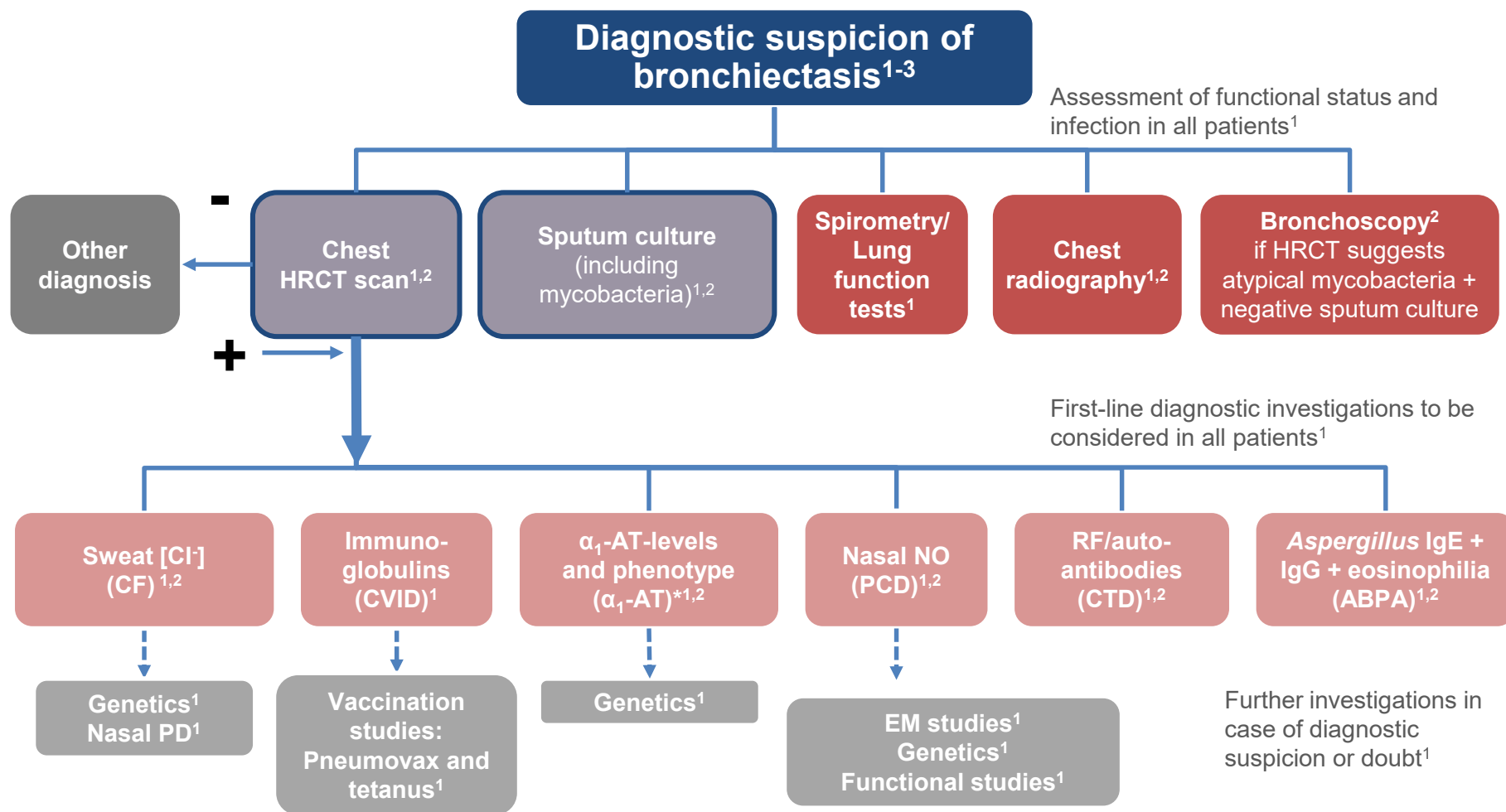




# Pathophysiology: Vicious Cycle hypothesis<sup>1,2</sup>



# Aetiological diagnosis of bronchiectasis



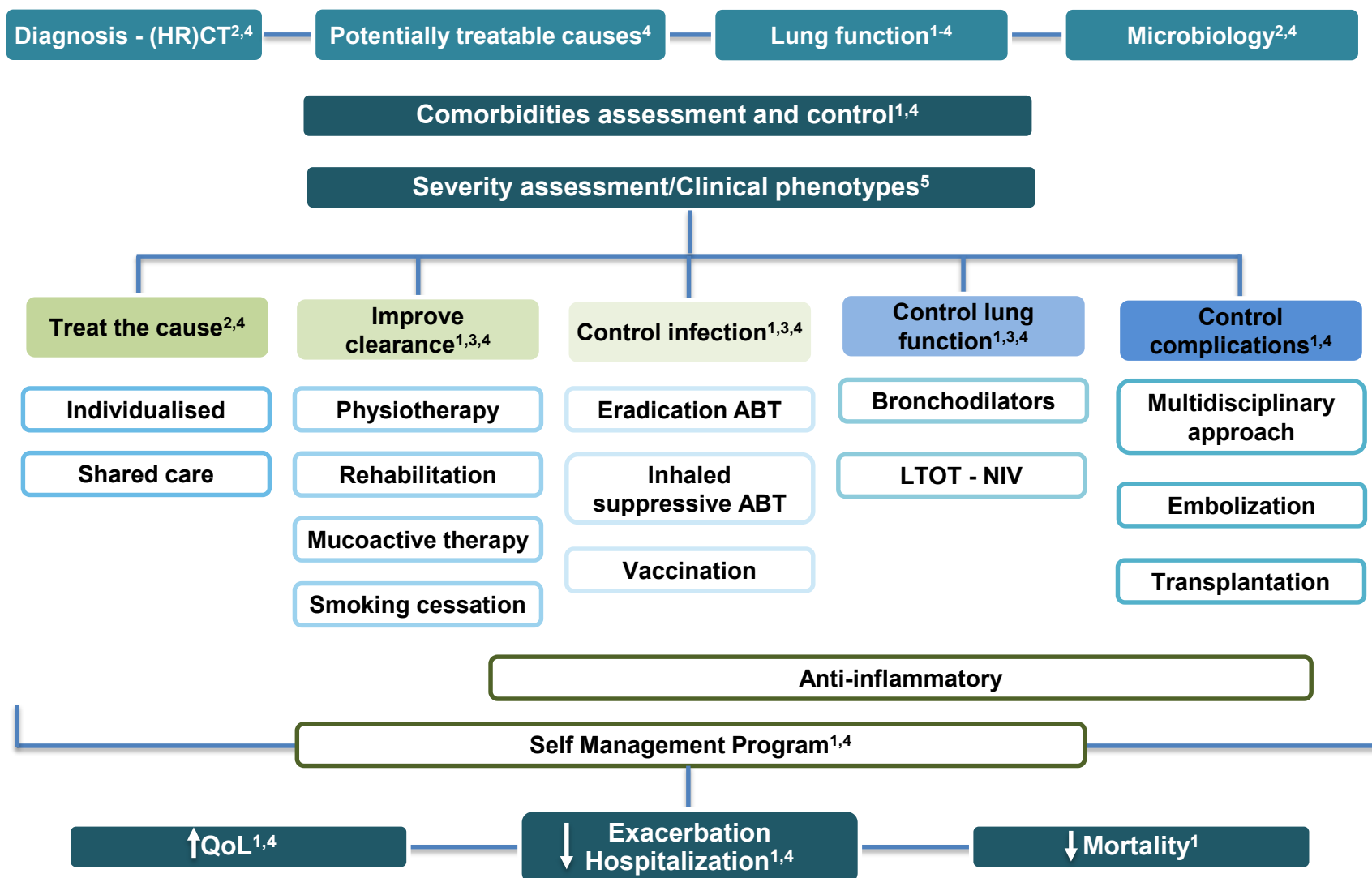
\*Routine screening not required unless the radiological investigations suggest basal emphysema.

$\alpha_1$ -AT, alpha-1 antitrypsin; ABPA, allergic bronchopulmonary aspergillosis; AT, Antitrypsin; CF, cystic fibrosis; Cl<sup>-</sup>, Chloride ion concentration; CTD, connective tissue disease; CVID, common variable immune deficiency; EM, electron microscopy; HRCT, high resolution chest tomography; Ig, Immunoglobulin; NO, Nitric oxide; PCD, primary ciliary dyskinesia; PD, potential difference; RF, rheumatoid factor.

1. Drain M, Elborn JS. Eur Respir Monograph 2011; 52:32-43; 2. British Thoracic Society Guidelines for Bronchiectasis in adults. 2018; (<https://www.brit-thoracic.org.uk/standards-of-care/guidelines/bts-guideline-for-bronchiectasis-in-adults-public-consultation/>); 3. Hill AT, et al. Eur Respir J 2017; 49.



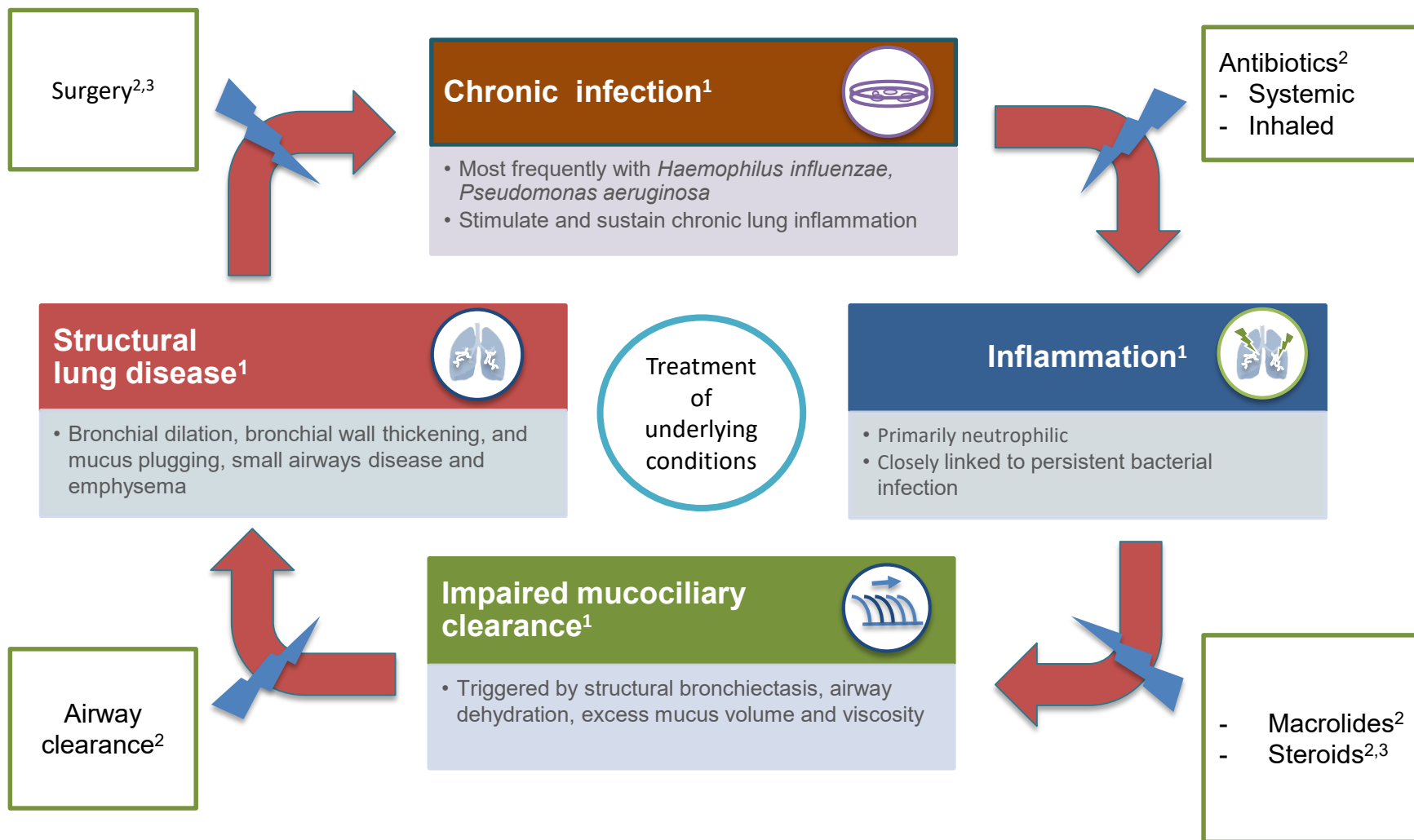
# A general overview on bronchiectasis management



ABT, antibiotic therapy; HRCT, high resolution computed tomography; LTOT, long-term oxygen therapy; NIV, non-invasive ventilation.

1. Martinez-Garcia MA, et al. Arch Bronconeumol 2018; 54:88-98; 2. Drain M, Elborn JS. Eur Respir Monograph 2011; 52:32-43; 3. Polverino E, et al. Eur Respir J 2017; 50:1700629; 4. British Thoracic Society Guidelines for Bronchiectasis in adults. 2018; (<https://www.brit-thoracic.org.uk/standards-of-care/guidelines/bts-guideline-for-bronchiectasis-in-adults-public-consultation/>); 5. Chalmers JD, et al. Am J Respir Crit Care Med 2014; 189:576-85.

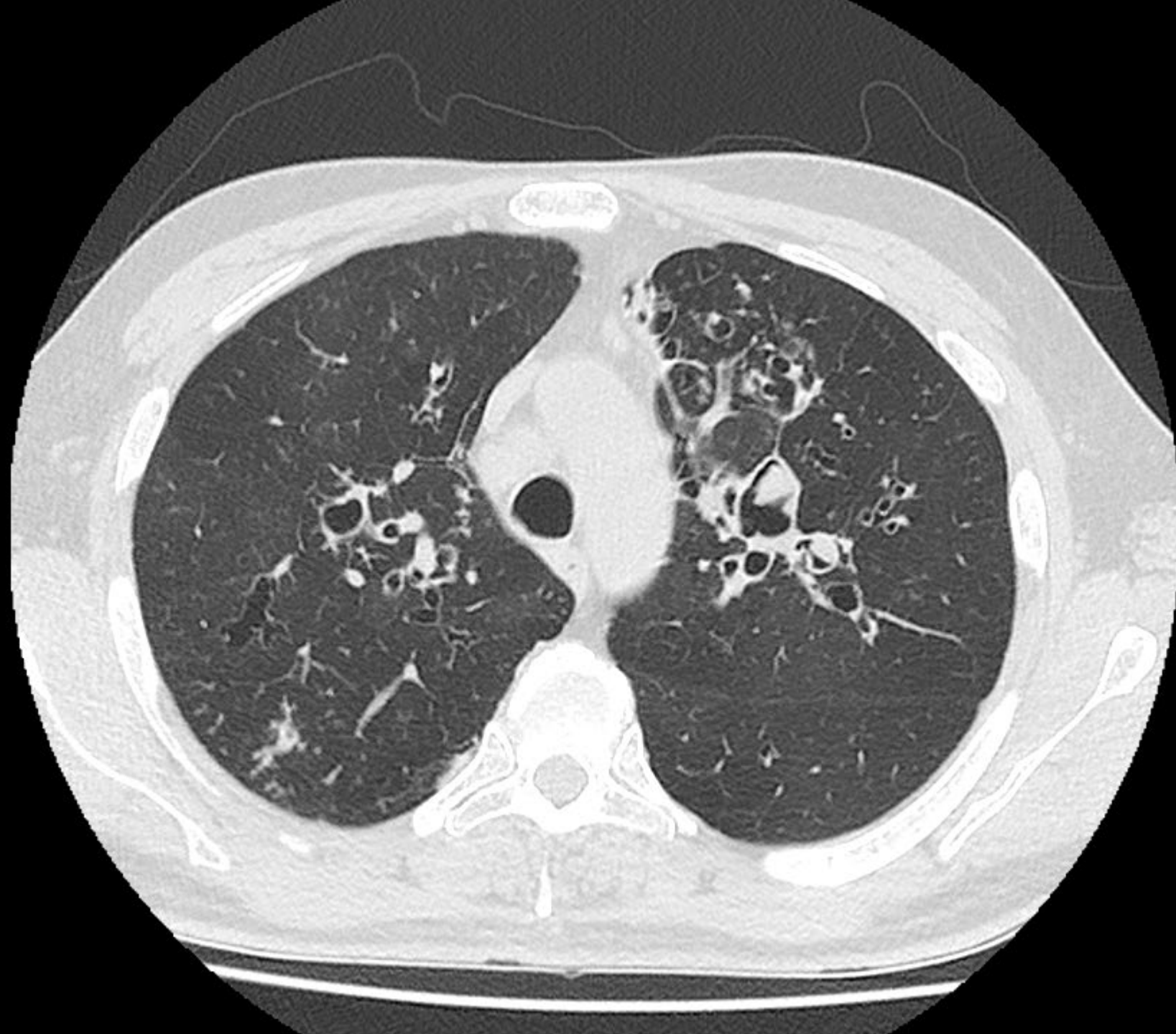
# Treatment of bronchiectasis





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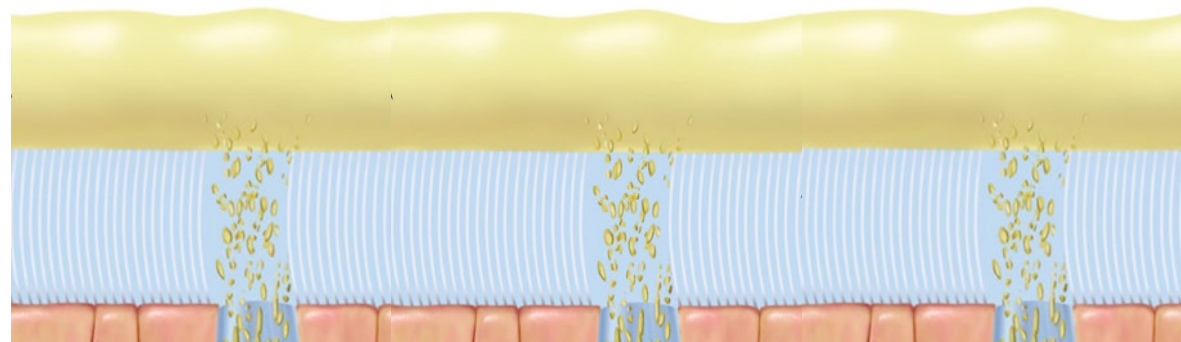
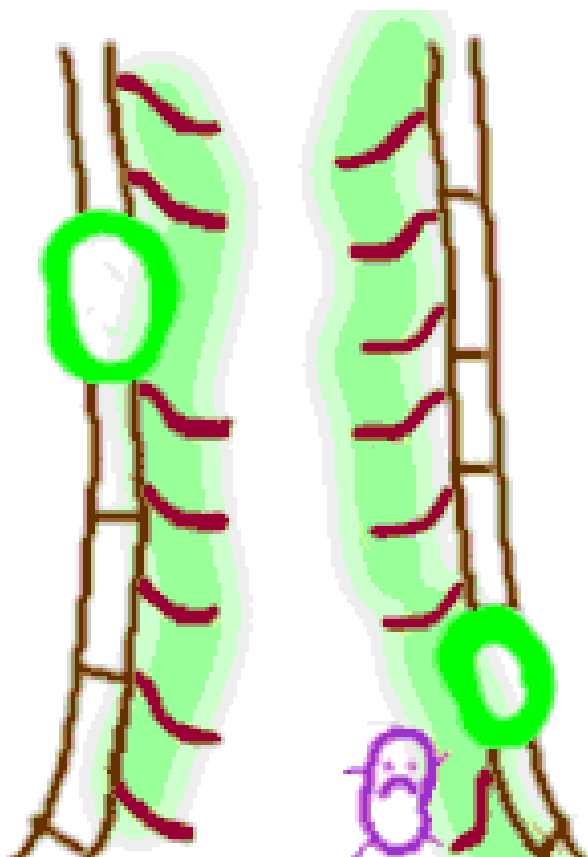
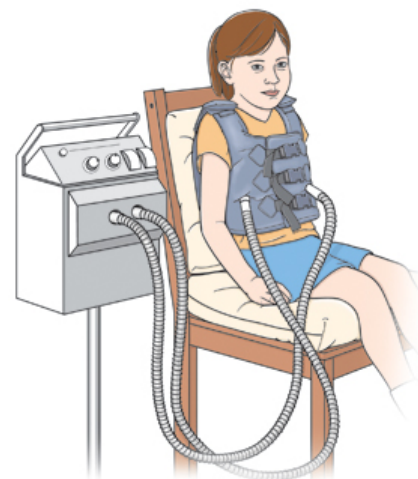
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# Airway clearance





# 12 month study of 6% hypertonic saline



40 patients- 6% hypertonic saline vs 0.9% saline

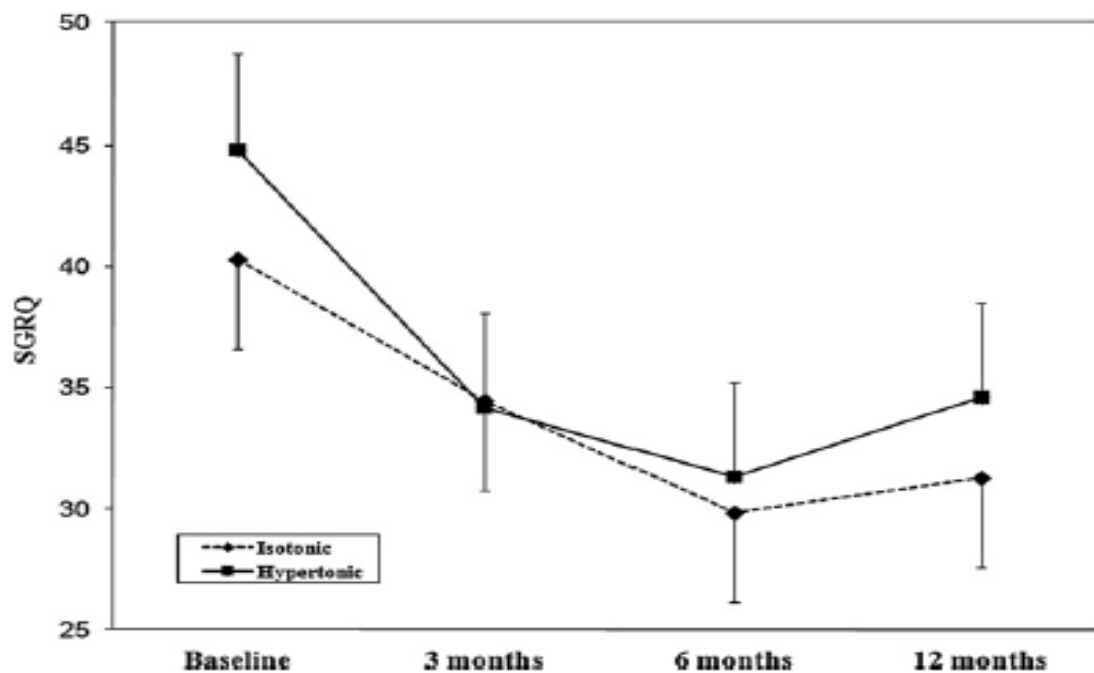


Figure 2 SGRQ Totals. No significant difference between groups at any time point.

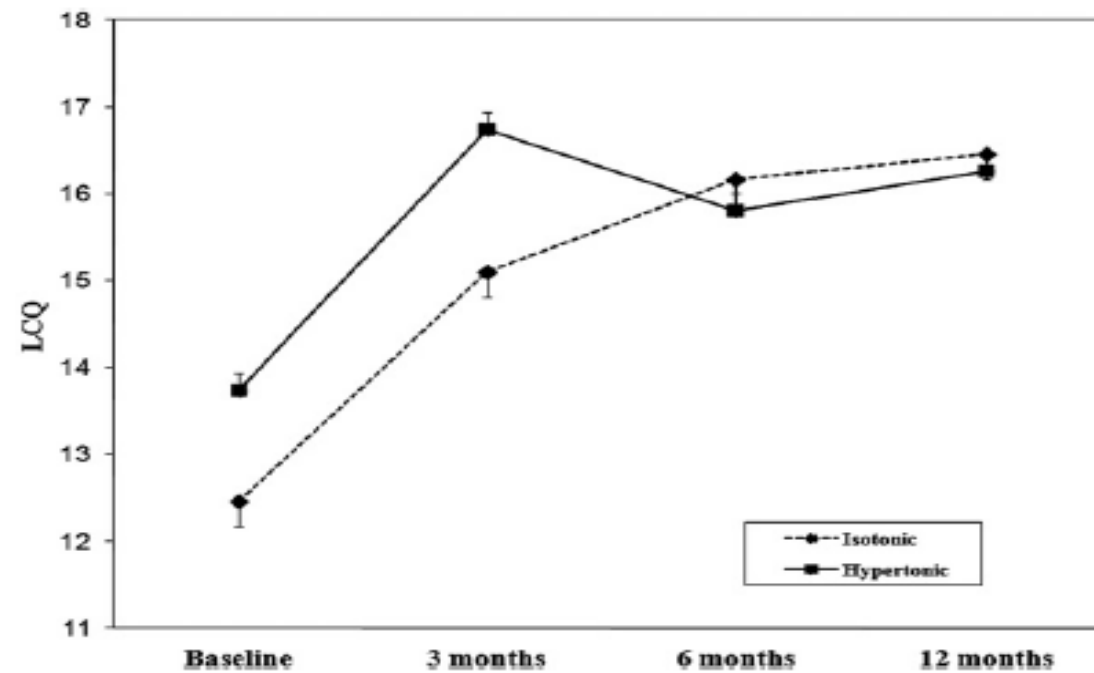
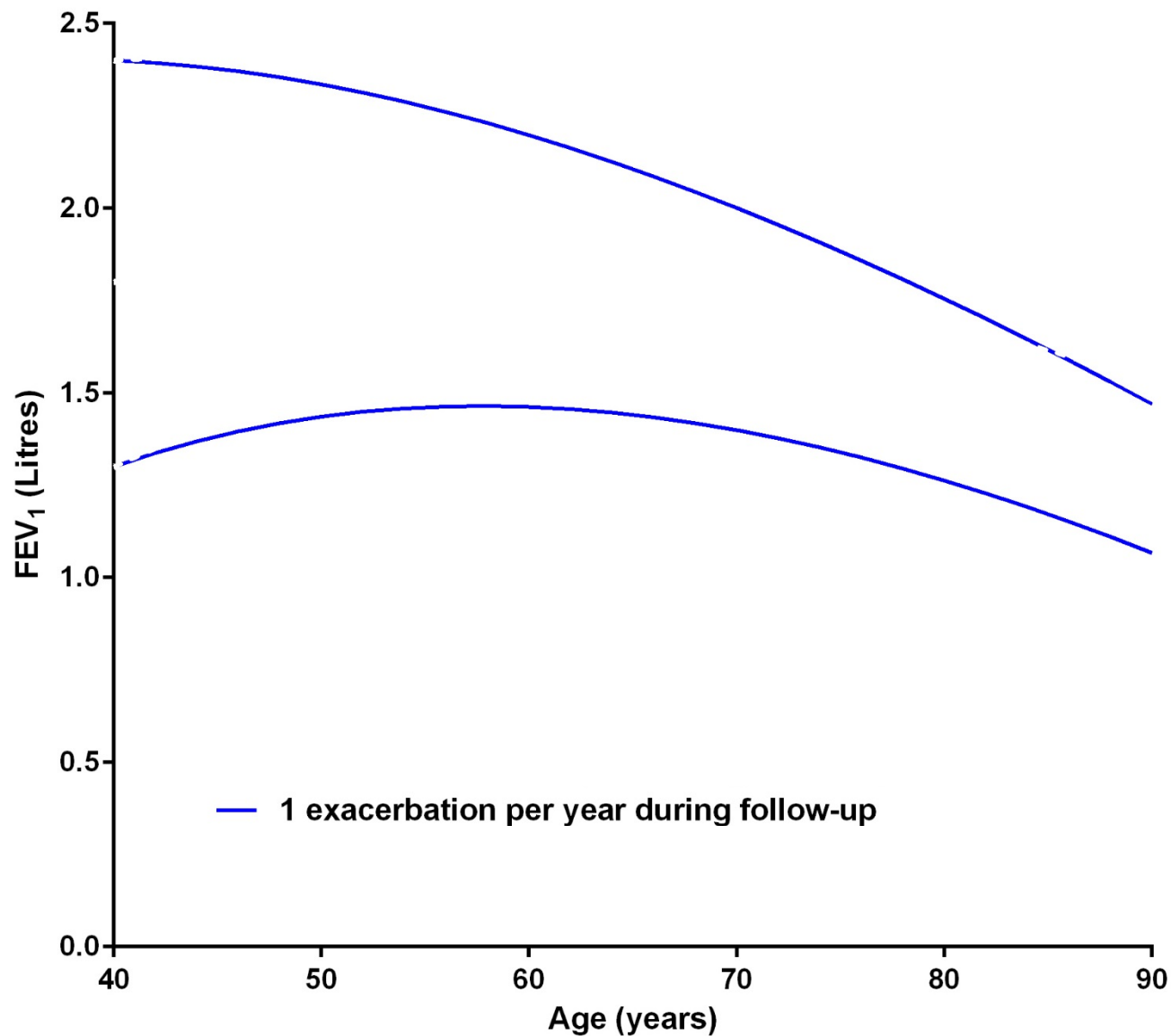


Figure 3 LCQ Totals. No significant difference between groups at any time point.



# Lung function decline



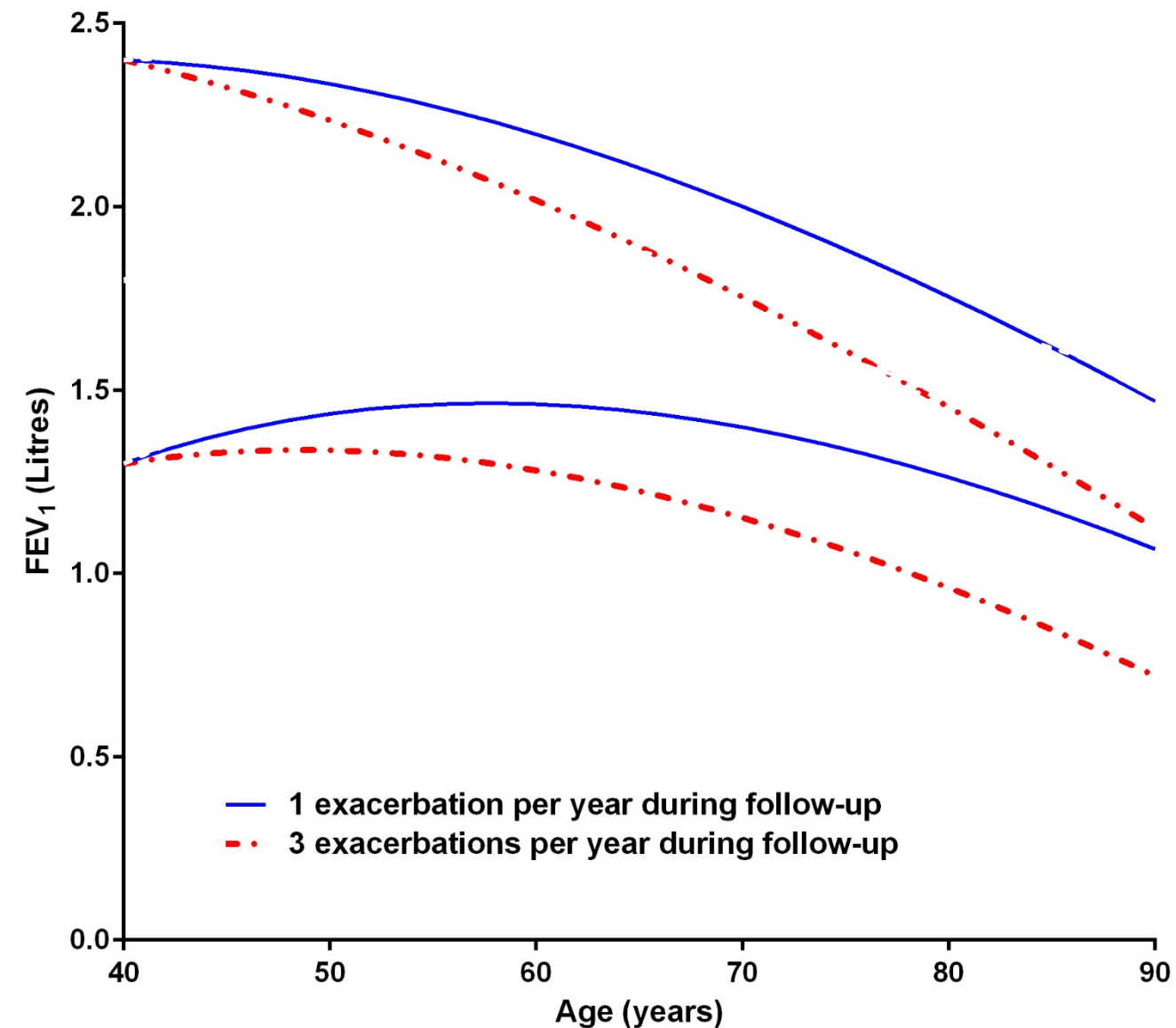
Multivariate linear mixed model over 4 years follow-up

Exacerbations independently accounted for 11ml lung function decline per event

Other risk factors included

- Baseline FEV1
- Smoking
- Symptoms
- Airways disease

# Lung function decline



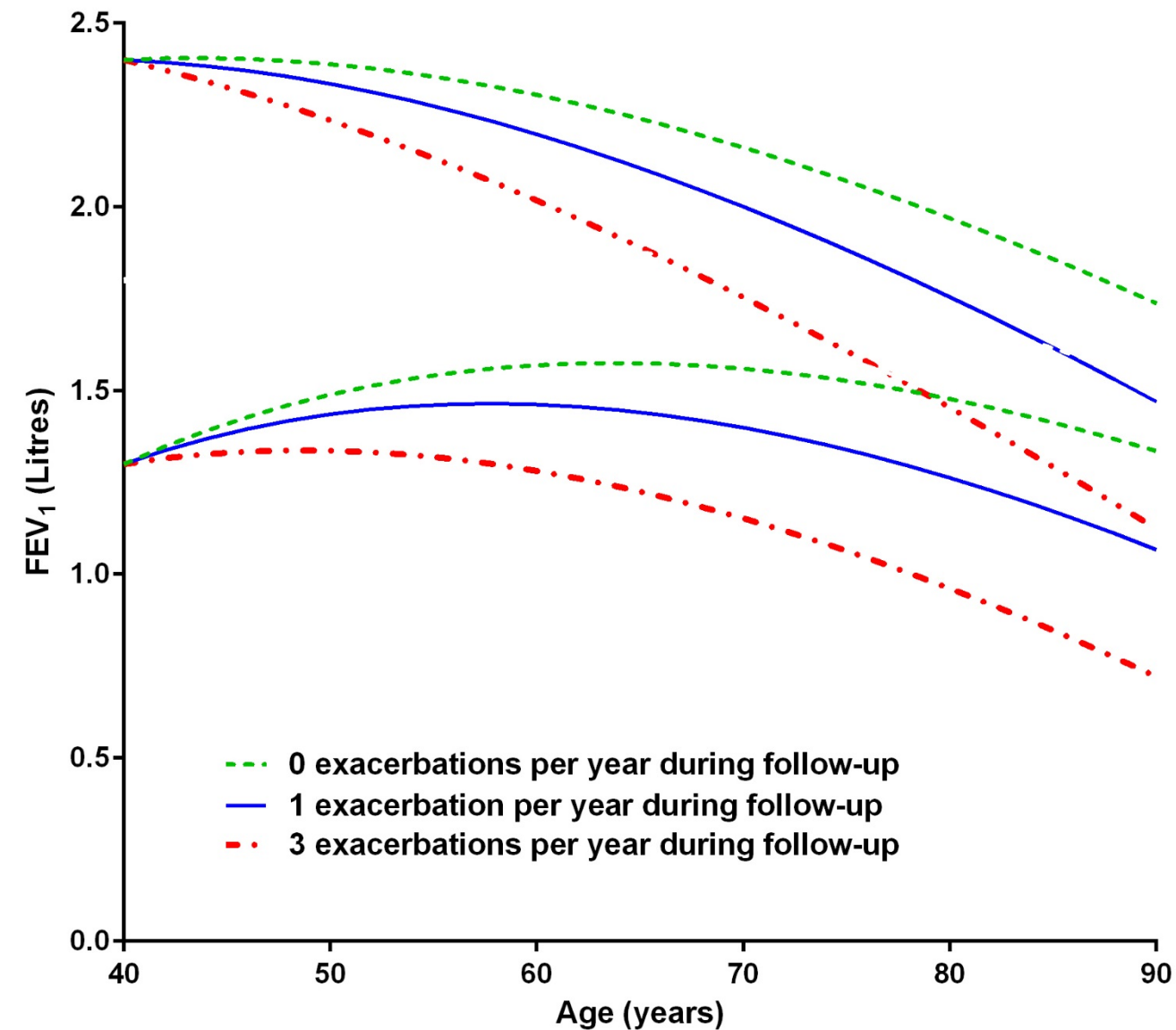
Multivariate linear mixed model  
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Exacerbations independently  
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decline per event

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# Lung function decline



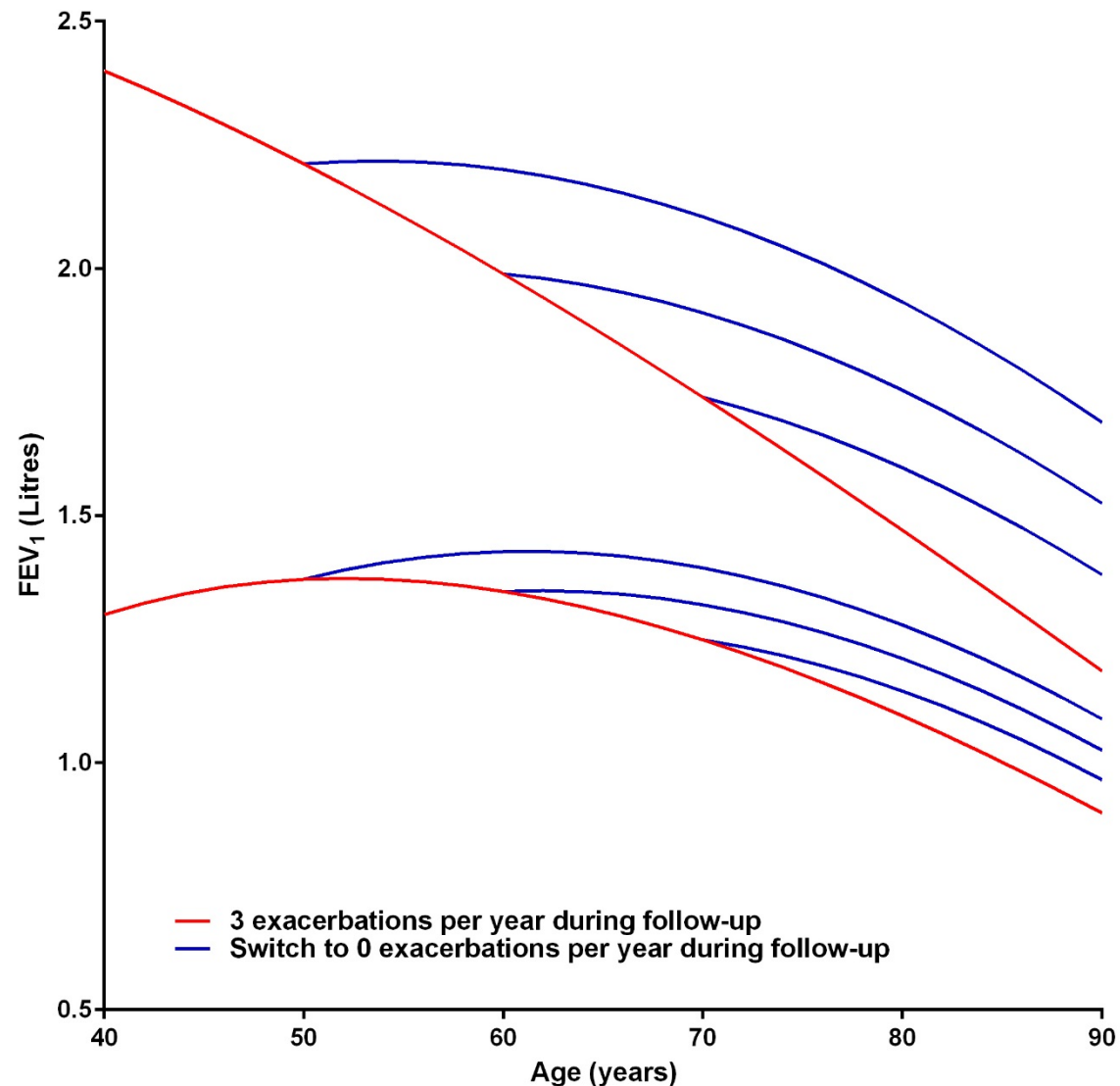
Multivariate linear mixed model  
over 4 years follow-up

Exacerbations independently  
accounted for 12ml lung function  
decline per event

Other risk factors included

- Baseline FEV<sub>1</sub>
- Smoking
- Symptoms
- Airways disease

# Could treatment prevent lung function decline



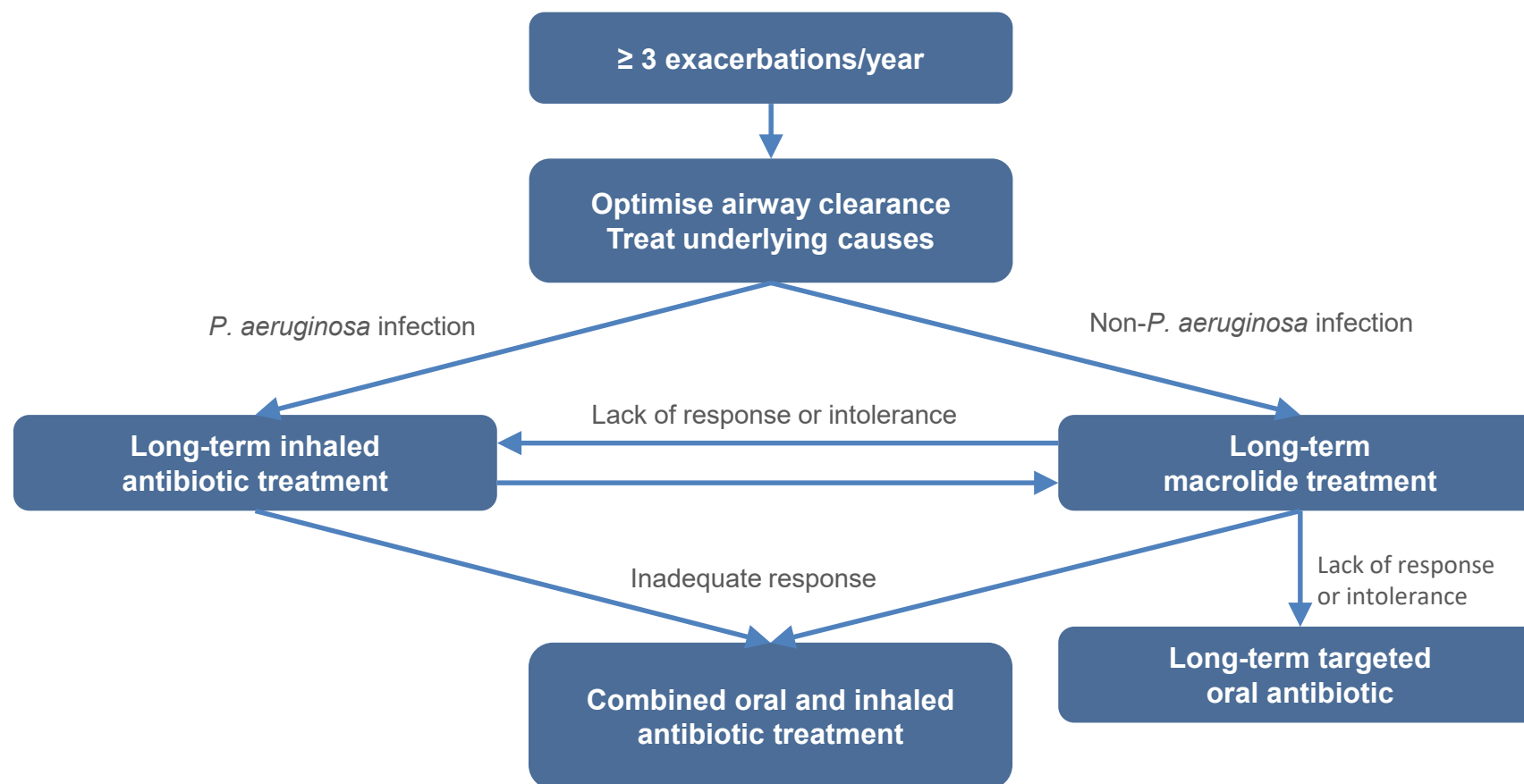
Hypothetical model of introducing a therapy at age 50, 60 and 70 which prevents exacerbations

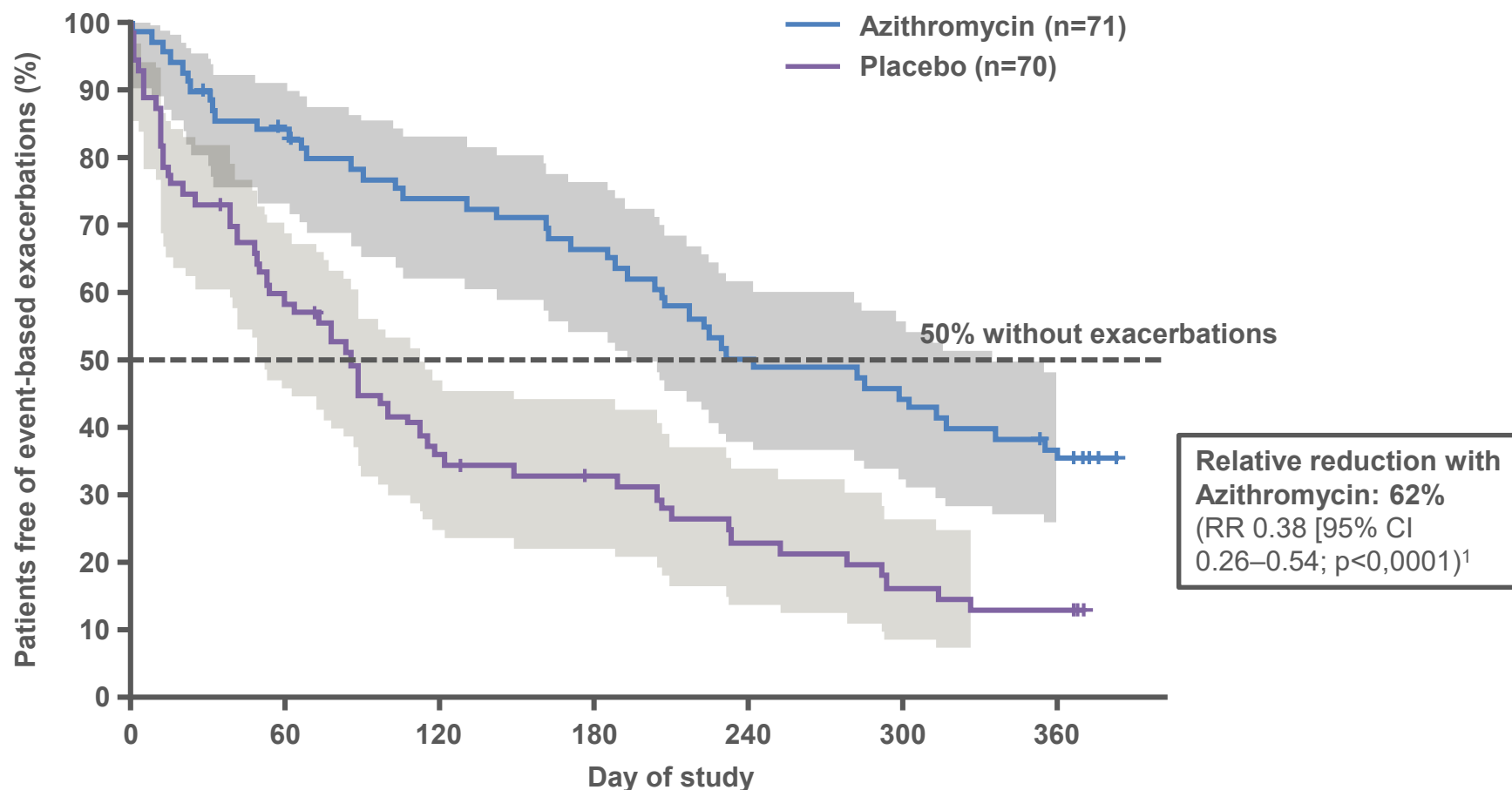
Data shows a marked effect of exacerbation reduction which is greatest in patients with preserved lung function



# ERS recommendations for long-term antibiotic treatment of bronchiectasis

- Acute exacerbations of bronchiectasis should be treated with 14 days of antibiotics in normal cases
- Long-term antibiotic treatment ( $\geq 3$  months) should be offered to the patient in case of  $\geq 3$  exacerbations per year:

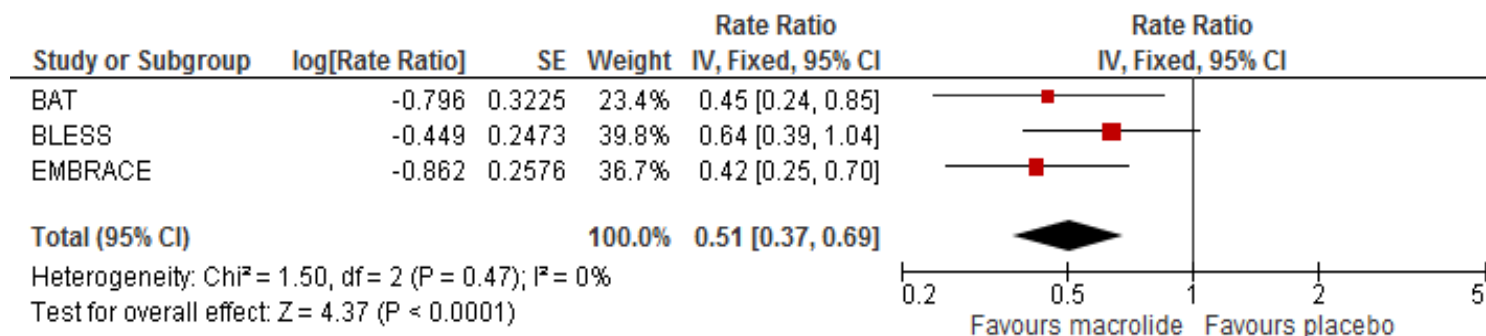




- 1 of 3 patients with bronchiectasis receive long-term macrolide treatment<sup>2</sup>
- Attention: Risk of developing a macrolide resistance  
→ Always culture (at least 3 times) for NTM before starting macrolide therapy



# Individual patient data meta-analysis: macrolides



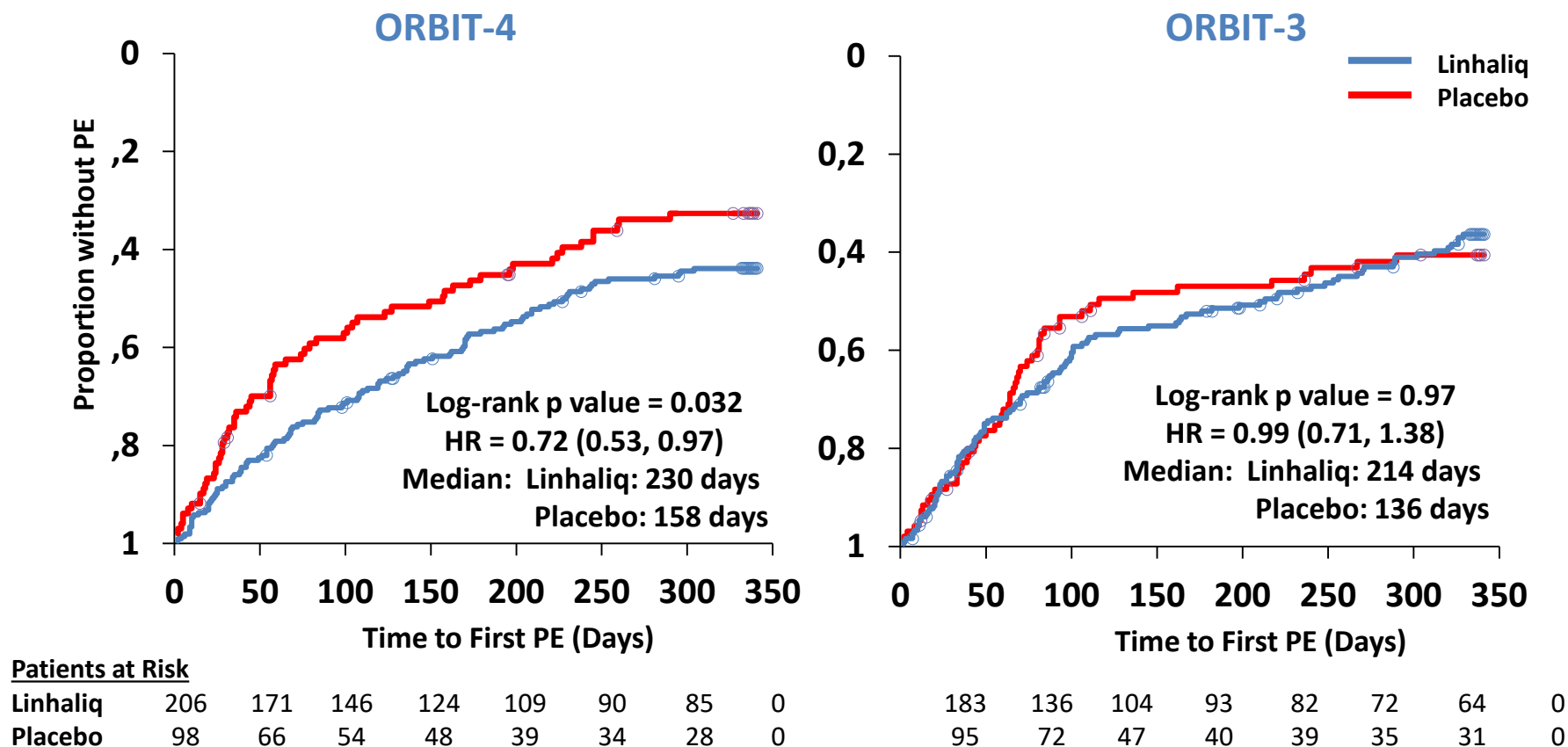
Macrolides overall reduce exacerbations by **50%**

Group	Macrolide	Placebo	NNT
1-2	0.32	1.0	1.5
3	0.77	1.35	1.7
4+	1.14	2.11	1.0

Largest benefit seen in patients with *Pseudomonas aeruginosa* where the benefit is 64% (RR 0.36 (0.20-0.67)).



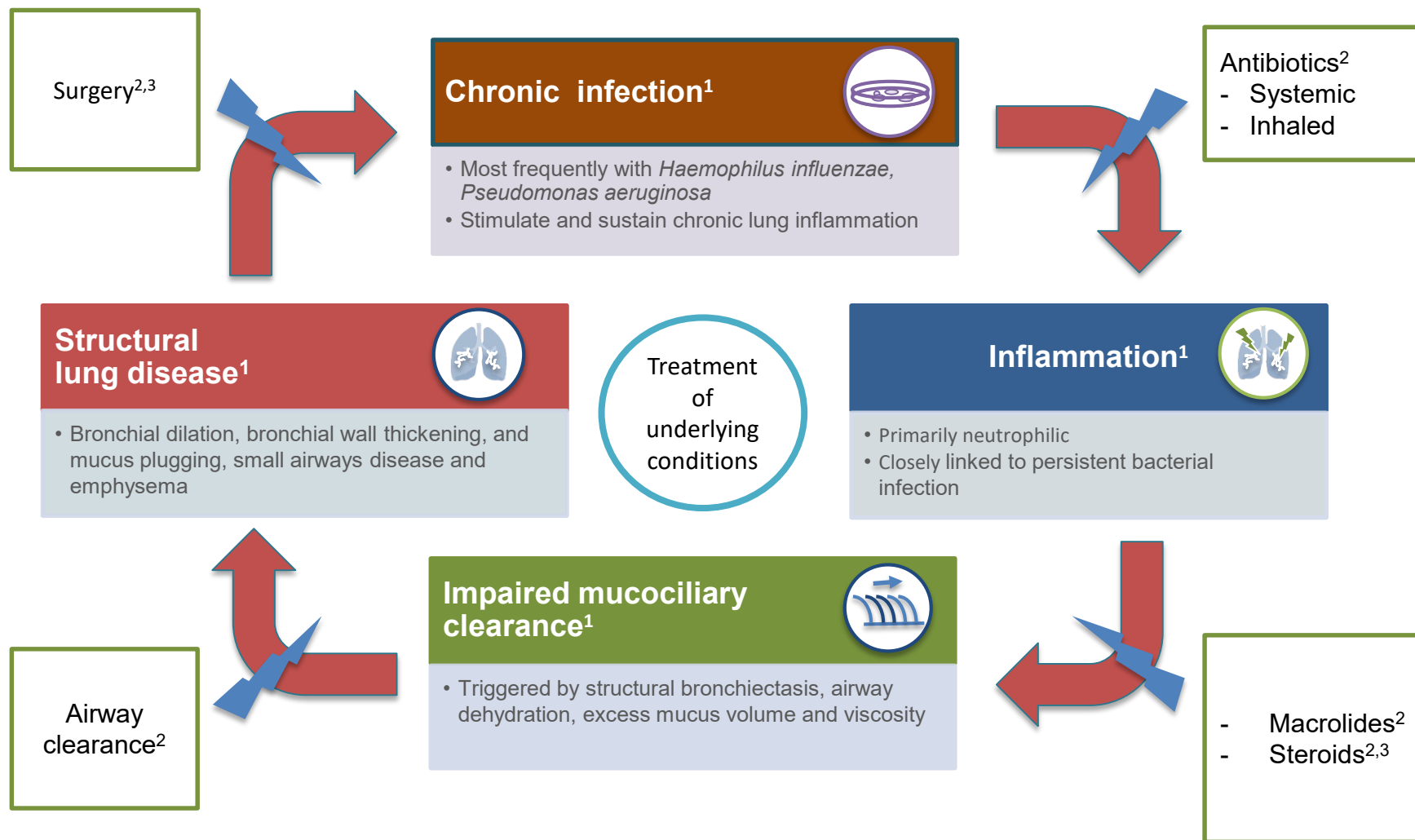
# Inconsistent results with inhaled antibiotics



Summarized recommendation	Strength of recommendation	Quality of evidence
Perform a minimum bundle of tests including differential blood count, serum immunoglobulins, and testing for ABPA in newly diagnosed patients	Conditional	Very low
Treat acute exacerbations of bronchiectasis with 14 days of antibiotics	Condition	Very low
Patients with a new isolation of <i>Pseudomonas aeruginosa</i> should be offered eradication antibiotic treatment	Conditional	Very low
Do not offer eradication antibiotic treatment to patients following new isolation of pathogens other than <i>P. aeruginosa</i> .	Conditional	Very low
Do not offer inhaled corticosteroids for the treatment of bronchiectasis	Conditional	Low
Do not offer statins for the treatment of bronchiectasis	Strong	Low
Offer long term antibiotic treatment for patients with three or more exacerbations per year*	Conditional	Moderate
Offer mucoactive treatment for aptients who have difficulty expectorating sputum and poor quality of life where standard airway clearance techniques have failed to control symptoms	Conditional	Low
Do not offer recombinant DNase for the treatment of bronchiectasis	Strong	Moderate
Do not routinely offer long acting bronchodilators for patients with bronchiectasis	Conditional	Very low
Offer long acting bronchodilators for patients with significant breathlessness on an individual basis	Conditional	Very low
Do not offer surgical treatments with the exception of patients with localised disease and high exacerbation frequency despite optimal medical care	Conditional	Very low
Patients with chronic productive cough or difficulty to expectorate should be taught airway clearance techniques	Conditional	Low
Patients with impaired exercise capacity should participate in pulmonary rehabilitation and take regular exercise	Strong	High



# Treatment of bronchiectasis



## “Treatable” (Therapeutic)

### Chronic airway infection

- Antibiotic therapy
  - Inhaled
  - Targeted
  - Macrolides

### Pathogen acquisition



- *Pseudomonas* eradication therapy

### Immunodeficiency

- Ig replacement
- Prophylactic antibiotics

### NTM



- Antibiotic therapy

### ABPA

- Corticosteroids
- +/- antifungals

### Airflow obstruction & Functional impairment

- Pulmonary rehabilitation
- Bronchodilators



### Sputum production

- Airway clearance
- Mucoactive drugs

### Asthma & eosinophilia



- Inhaled corticosteroids

### Low BMI

- Nutrition

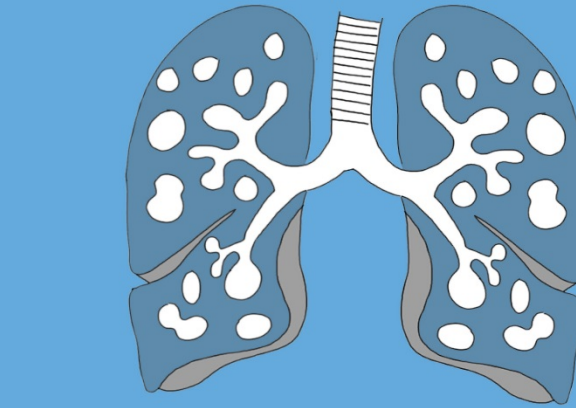
### GORD

- PPI
- +/- prokinetics



### Other comorbidities

- Treat appropriately



# BRONCHIECTASIS “TRAITS”

## “Other factors”

- Ethnic differences
- Environmental exposures
- Climatic variation
- Lifestyle factors

## “Targetable” (Endophenotypic)

### Microbial (bacterial) dysbiosis

- Probiotics



### Mycobiome (fungal) dysbiosis

- Anti-fungal

### Neutrophil dysfunction

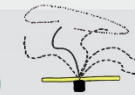
- NE inhibitors



### Protease mediated lung damage

- Protease inhibitors

### Ciliary dysfunction (Primary or secondary)



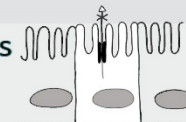
- Airway clearance
- CFTR potentiator therapy

### Systemic inflammation & vascular dysfunction

- Anti-inflammatory therapy

### CFTR dysfunction

- CFTR potentiators
- CFTR correctors



### Innate immune deficiency

- TLR-based therapeutics
- Antibiotic prophylaxis

# Acknowledgements

## Executive group

Stefano Aliberti  
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## iABC co-ordinator

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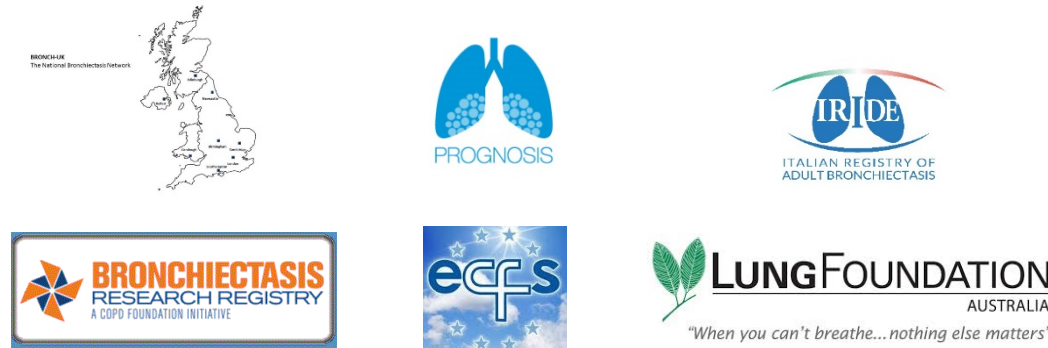
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Antoni Torres  
Montserrat Vendrell  
Tobias Welte  
Robert Wilson

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[www.bronchiectasis.eu](http://www.bronchiectasis.eu)

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