Bronchiectasis

Dilated, poorly clearing airways in a self-sustaining cycle · ERS 2025 guideline · often mislabeled COPD

Clearance is daily
antibiotics treat the flare

⚠ THE VICIOUS CYCLE, AND WHY IT DICTATES THE TREATMENT *three arms, because three links*

THE LOOP

Impaired mucociliary clearance → bacterial colonization → neutrophilic inflammation → airway wall destruction → worse clearance. Every therapy breaks one link, hence **three arms: clearance, infection, inflammation**. This is a **chronic disease with acute flares, not a run of unrelated pneumonias** -repeat antibiotic courses alone address one link intermittently while the others run continuously.

WHEN TO SUSPECT IT

Chronic daily purulent sputum (often dismissed as smoker's cough). **Recurrent pneumonia in the SAME lobe** -a structural problem until proven otherwise. **COPD that does not fit**: never-smoker, disproportionate sputum, frequent exacerbations, or repeatedly grows *Pseudomonas*. **Recurrent hemoptysis. RA or IBD** with respiratory symptoms -in RA the lung disease may precede the joints.

EXAM & BASELINE

Coarse inspiratory crackles are the commonest finding; **clubbing is uncommon, so its absence means nothing**. Spirometry is usually obstructive, part of why this gets misfiled as COPD; it neither makes nor excludes the diagnosis. **⚠ A NORMAL CHEST X-RAY DOES NOT EXCLUDE IT** -radiography is insensitive. Compatible history plus a clean film means get a CT, not reassurance.

HRCT DIAGNOSIS, AND READING THE DISTRIBUTION FOR THE CAUSE

Finding / pattern	Meaning
Diagnostic criteria	**Bronchoarterial ratio > 1** is the defining feature -airway wider than its companion artery, seen in cross-section as the **signet ring sign**. Supporting: **loss of normal distal tapering**, and **airways visible within ~1 cm of the pleura** where normal bronchi are too small to resolve
Upper lobe predominant	Suggests **cystic fibrosis** or **prior tuberculosis**
Central / perihilar + mucus plugging	Suggests **ABPA**, especially with asthma and eosinophilia
Middle lobe & lingula + tree-in-bud	Suggests **nontuberculous mycobacterial** infection
Focal, single lobe	**Exclude obstruction** (tumor, foreign body, compression) -bronchoscopy warranted. Or prior severe infection

FIND THE CAUSE: IT CHANGES TREATMENT IN A REAL MINORITY *the highest-yield thing available at the first visit*

Test	What it finds, and what changes
Serum immunoglobulins (IgG, IgA, IgM)	**Highest-yield treatable cause.** CVID and other antibody deficiencies present exactly this way, and **immunoglobulin replacement changes the disease course.** Add specific vaccine-antigen titers if borderline
Total IgE + *Aspergillus* IgE/IgG	**ABPA** -a steroid-treated disease that antibiotics will not touch
Sputum culture *including mycobacterial*	Sets the baseline flora that guides every future exacerbation. **Must be requested explicitly** -it is not part of a routine culture
CF testing (sweat chloride, CFTR)	**Test adults too.** Milder genotypes present in adulthood; diagnosis opens **modulator therapy** and a different care model. Prioritize with upper lobe disease, *S. aureus*/*Pseudomonas*, malabsorption or infertility
PCD · rheumatologic · alpha-1	**Primary ciliary dyskinesia** with lifelong symptoms, chronic sinus disease, otitis, infertility or **situs inversus** (Kartagener). **RA is well recognized and may follow the lung disease**; IBD associated. Alpha-1 deficiency carries family-screening implications

MANAGEMENT *ERS 2025: airway clearance training is now universal, not selective*

DAILY CLEARANCE = THE BACKBONE

Taught by a therapist, not mentioned in passing: active cycle of breathing, autogenic drainage, PEP/oscillatory devices, postural drainage. **The best technique is the one performed every day.** Add **nebulized hypertonic saline** -give a **bronchodilator first and watch the first dose**, since it can provoke bronchospasm. **Pulmonary rehab** for exercise limitation.

⚠ NEVER DORNASE ALFA HERE

The trap: **rhDNase is standard, beneficial therapy in CYSTIC FIBROSIS**. In **non-CF bronchiectasis a randomized trial found HARM** -more exacerbations and greater FEV_1 decline. The mechanisms differ, so **the CF evidence does not transfer.** Confirm which diagnosis you are treating before reaching for it.

EXACERBATION

Worsening cough, sputum volume/purulence, dyspnea or hemoptysis **sustained ~48 h+. Fever and infiltrates are often ABSENT**, so their absence excludes nothing. **Culture first, then treat empirically targeting the patient's OWN prior organisms**; cover *Pseudomonas* if previously grown. **~14 days, deliberately longer than pneumonia or COPD** -biofilm in abnormal airways relapses after short courses. **Intensify clearance**; steroids only for coexisting asthma/COPD.

PREVENTING THE NEXT ONE

Intervention	Detail and cautions
Long-term azithromycin (3× weekly)	For **≥ 3 exacerbations/year** despite optimized clearance, or chronic *Pseudomonas*. Benefit is **anti-inflammatory as much as antimicrobial**. **The threshold is a starting point, not a gate** -treat below it for severe flares, major quality-of-life impact, severe disease or immunodeficiency
⚠ Before any macrolide	**(1) EXCLUDE NTM** with mycobacterial cultures -monotherapy in unrecognized NTM **breeds macrolide resistance and wrecks the regimen you would later need. (2) Baseline ECG for QTc and liver function tests**; review interacting drugs; counsel on hearing changes
Pseudomonas	**The key prognostic marker** -more exacerbations, faster FEV_1 decline, worse quality of life, higher mortality. **Consider eradication on first isolation** rather than logging it as incidental. For chronic infection with frequent flares, **inhaled antibiotics** (tobramycin, colistin, aztreonam) give high airway levels with low systemic exposure; evidence is mixed and availability varies, so specialist-guided
⚠ Inhaled corticosteroids	**NOT routine therapy**, and they carry real **pneumonia and NTM risk** here. Use only for a genuine coexisting indication (asthma, eligible COPD)
Vaccinate · severity	Influenza, pneumococcal, COVID-19 -a prevented viral infection is a prevented bacterial flare. **BSI / FACED** scores stratify who needs aggressive prevention
Hemoptysis	From **hypertrophied bronchial arteries**. Massive: protect the airway, **bleeding side DOWN** to spare the good lung, **bronchial artery embolization** is definitive. **Hold clearance during active bleeding**

PEARLS & PITFALLS

- **Isolating NTM is not by itself an indication to treat** -diagnostic criteria must be met before committing to prolonged multidrug therapy.
- **Bronchiectasis and COPD genuinely coexist**, and the overlap group does worse than either alone -so finding one does not excuse you from looking for the other.

References: European Respiratory Society clinical practice guideline for the management of adult bronchiectasis (2025, replacing 2017) · British Thoracic Society guideline for bronchiectasis in adults · randomized trial evidence on recombinant human DNase in idiopathic bronchiectasis · randomized trials of long-term macrolide therapy for exacerbation prevention.