

BRONCHIECTASIS

ETIOLOGY

Airway abnormality

- Pneumonia
- Airway obstruction by tumor or foreign body

Congenital disorder

- Cystic fibrosis
- Primary ciliary dyskinesia
- Alpha-1 antitrypsin deficiency
- Mounier–Kuhn*, Williams–Campbell, and Ehlers–Danlos syndromes

Immunodeficiency

- Common variable immunodeficiency
- Acquired immunodeficiency
- HIV infection
- Hematologic cancer

Autoimmune disorder

- RA, Sjögren's syndrome, Systemic Sclerosis
- Inflammatory bowel disease

Aspiration syndrome

- Vocal cord disease or dysfunction
- Esophageal disease or dysmotility

Allergic bronchopulmonary aspergillosis

WORKUP

Prior imaging, bronchoscopy-BAL

Sweat Cl testing, CFTR mutations

Genetic testing

α-1 antitrypsin levels and phenotyping

CT findings

Ig levels, HIV tests, Ab levels before and after vaccination

ANA, anti-CCP, RF

GI workup

MBS, esophageal motility testing

Aspergillus-specific IgE, aspergillus skin-prick testing, IgE level

* Mounier–Kuhn syndrome primarily etiology is congenital but can be acquired in the context of chronic airway injury connective tissue diseases.

TREATMENT

- Treatment of underlying condition
- Improve clearance and drainage of airways secretions
 - Maintenance of adequate systemic hydration
 - Nebulization with hypertonic solutions (saline 7% or NAC 10 or 20%) twice daily or NAC 600 mg P.O q12h
 - Use of OPEP (AirPhysio, Acapella or Aerobika) to be used frequently at least q12h x2 min
 - If frequent flares persist, consider HFCWO-VEST therapy and or continuous high-frequency chest wall oscillation device (CHFO)
 - CHFO- Volara (Baxter) device provides:
 - Continuous positive expiratory pressure (CPEP) for continuous lung expansion

- Continuous high frequency oscillation (CHFO) to promote mobilization of secretions
 - The system can also provide aerosolized medications approved for nebulization and supplemental oxygen
- Treatment of airways obstruction and inflammation:
 - Brensocatib is a dipeptidyl peptidase 1 (DPP1) inhibitor that targets neutrophil-mediated diseases.
 - Reduce the annual rate of exacerbations, prolong the time to the first exacerbation and increase the chances of patients remaining exacerbation-free.
 - If documented airway obstruction: LABA with or without inhaled steroids and albuterol q4h as needed
- Treatment and prevention of Infection:
 - Vaccines: Influenza yearly, RSV following CDC recommendations, and Pneumococcal (one dose PCV21 - CAPVAXIVE if not vaccinated or if unknown)
 - Antibiotics as needed for flareups
 - Antibiotics by Nebulization for patients with more than 3 exacerbations yearly despite systemic antibiotics
 - Tobramycin 300 mg plus albuterol 2.5 mg every 12 hr by nebulization
 - Consider other options if frequent exacerbations
 - Macrolides: If more than 2 exacerbations yearly, absence of non-TB mycobacteria and absence of cardiovascular disease along with normal QT interval: Azithromycin p.o. 500 mg MWF
 - Rotational Antibiotics: if more than 3 exacerbations yearly and not a candidate for macrolides.
 - Use one antibiotic at each time for 10 days followed by 20 days off. Start with one antibiotic and then switch to a different one the next month. After 3 months, then the cycles.

1. Ciprofloxacin or Levofloxacin 500 or 750 mg 1 tab by mouth every 12 hr x 10 days.
2. Trimethoprim/Sulfamethoxazole DS 1 tab by mouth every 12 hr times 10 days.
3. Doxycycline 100 mg 1 tab by mouth every 12 hr times 10 days.