Bronchiectasis and Airway Clearance
Underdiagnosed and Undertreated

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Disclosures

- I receive speaking/consulting fees from Insmed
- I give unbranded disease state talks on Mycobacterium Avium Complex pulmonary disease
Objectives

• Understand the definition and pathophysiology of bronchiectasis
• How to diagnose bronchiectasis
• Basic management of bronchiectasis
• Review the vital role of airway clearance in the management of bronchiectasis
• When does vest therapy come into play
Bronchiectasis

• Greek: bronkhia (airway) and ektasis (widening)
• Features similar to COPD/asthma
  – Inflamed and easily collapsible airways
  – Obstruction of airflow
  – Frequent exacerbations
• Irreversible dilation and destruction of bronchial walls

Barker AF. NEJM. 2002
Bronchiectasis

Normal bronchus

Bronchiectasis

Normal cell wall
Normal mucus

Damaged cell wall
Increased mucus
Bronchiectasis

- Nonspecific symptoms
- Often misdiagnosed and treated as asthma or COPD
- Often not considered in the differential diagnosis of chronic cough
- Significant utilization of health care system:
  - Hospitalizations
  - Clinic visits
  - Imaging and laboratory tests
  - Antibiotics
Epidemiology

- First described by Rene Laennec in 1819
- Prevalence varies from country to country
- Estimated 350,000 – 500,000 in the US
- More Common in women and older age

https://www.regencyhistory.net/2016/03/regency-medicine-invention-of.html
Epidemiology

- Considered untreatable prior to the development of antibiotics in the 1950s
- Common in Western world around WWI
  - 40% mortality before age 40
- Successful pertussis and measles vaccination reduced mortality in developed countries
  - “orphan disease” and research shifted away

Altenburg et al. Neth J Med. 2015
Types of Bronchiectasis

- Cylindrical bronchiectasis
- Varicose bronchiectasis
- Cystic bronchiectasis

Symptoms

- Cough
  - Dry or productive
- Shortness of breath
- Recurrent pneumonia
- Frequent asthma or COPD exacerbations
- Hemoptysis
- GERD or recurrent sinus infections?
Causes

• Primary immunodeficiency (CVID)
• Post-infection (TB, measles, pertussis)
• Non-tuberculous mycobacteria
• Impaired muco-ciliary clearance (CF, PCD)
• Toxic inhalation (GERD, chronic aspiration)
• Airway obstruction (tumor, foreign body)
• Obstructive airway disease (COPD, asthma, A1AT)
Causes

• Inflammatory bowel disease
• Autoimmune disease (RA, Sjogren’s, sarcoidosis)
• Interstitial lung disease (traction bronchiectasis)
• Secondary immunodeficiency (HIV, hematologic malignancy)
NTM

https://radiopaedia.org/cases/pulmonary-mycobacterium-avium-complex-infection-1
ABPA
Infectious and Inflammatory Cycle

• Inflammatory insult
  – Disordered muco-ciliary clearance
    • Airflow obstruction
      – Chronic or recurrent infections
        » Bronchial wall damage

• Inflammatory insult
  – ...
    • ...
      – ...
Bronchiectasis Cycle

1. Inflammation
2. Bacterial colonization
3. Mucus accumulation
4. Infection
5. Permanent airway dilation, loss of cilia
6. Neutrophil-derived proteases
7. Airway remodeling

Initial insult
Bronchiectasis and COPD

- Moderate-to-severe COPD
  - New bacterial infection
  - Increased bronchial inflammation
    - Antibiotic response
    - Immune response
      - Bacterial eradication?
        - Yes
        - No

- Cole's vicious circle
  - Persistent bronchial inflammation
    - Bacterial and inflammatory proteolytic molecules
    - Impaired mucociliary clearance
    - Chronic bronchial infection
      - Bronchial wall destruction and distortion
**Work-up**

- Once diagnosis made by HRCT:
  - Spirometry
  - Sputum cultures including AFB and Fungal
  - Immunoglobulins (IgG, IgE, IgA, IgM)
  - A1AT
  - PCD suspicion: Nasal nitric oxide (eNO) and ciliary biopsy with electron microscopy
  - Autoimmune serologies (ANA, ANCA, ENA, RF, anti-CCP)
  - HIV
Figure 3. Radiographic signs of bronchiectasis. A = Bronchus terminating in a cyst; B = lack of bronchial tapering as it travels to the periphery of the lung; C = signet ring sign (bronchus is larger than the accompanying vessel); D = mucus plug (mucus completely filling the airway lumen).
Treatment

• Different for each diagnosis
  » Immunoglobulin replacement
  » Antibiotics for NTM
  » Steroids and anti-fungal for ABPA
  » Immunosuppression for autoimmune disease
  » Recurrent aspiration – medical and surgical management
  » CPAP for tracheobronchomegaly
  » Inhaled steroids for asthma phenotype
Reflux Pillow

1. MedCline is used in your own bed.
2. You sleep comfortably in the perfect position.
3. Harmful acid stays in your stomach, where it belongs.
Treatment

• Optimizing treatment of the underlying cause
• Antibiotics targeting culture results for acute exacerbations/infections
• Chronic macrolide and/or inhaled antibiotics for prophylaxis
  • Daily azithromycin if 2 > exacerbations per year
• Airway clearance crucial
  » Therapeutic and prophylactic
Follow-up/Prevention

• 2-4 visits per year
• Spirometry 2x per year
• Sputum culture at least 2x per year
• Vaccinations
• Review of airway clearance and exercises
  • Pulmonary rehab
Clear Airways

- Less mucus
- Improved ventilation
- Reduced obstruction
- Less bacterial load
- Less infection
- Less exacerbations

Respiratory Therapists

• The most important caregivers in the successful management and treatment of bronchiectasis patients

• Airway clearance education, follow-up on techniques, adding new maneuvers, pulmonary rehab coach, induced sputum, etc.
Airway Clearance

- Most important treatment to break the cycle
- Time intensive
- Compliance can be poor
  - Can take weeks before starting to see results
- Not enough patient education
  - Poor understanding of why it is needed
Airway Clearance

- Exercise
  - 150 min moderate intensity exercise per week (treadmill, stationary bike, yoga, core strength)
  - Pulmonary rehab
- Bronchodilators
- Hypertonic saline
- Positive expiratory pressure devices (acapella)
- Vest
- Breathing maneuvers / postural drainage and percussion
Airway Clearance

“The Triple”

1. Nebulized albuterol BID
2. Hypertonic saline BID
3. Acapella BID or more (10-20 breaths followed by 2-3 huff coughs)
Huff Cough

- Forced expiration technique, alternative to deep coughing
  - Deeper breaths and using diaphragm + abdominal muscles to make a series of rapid exhalations to help move mucus to larger airways

https://quizlet.com/488727702/unit-ii-2-c-supporting-oxygenation-3-questions-flash-cards/
Postural Drainage
IMPACT

• Individual management of patient airway clearance therapy
  – Toolset developed to help assess, evaluate, and educate patients on airway clearance options
  – Structured discussion in the office
  – Improved compliance by understanding barriers
  – Stresses long term benefit
AIRWAY CLEARANCE ASSESSMENT

There are no right or wrong answers to this survey. Please answer as truthfully as possible so we can work together to find the best airway clearance treatment(s) for you and your lifestyle.

1. Which airway clearance methods are you currently using? Please circle the device(s) you are using:
   - ExerTec (please specify which type)
   - HulCoughing
   - Positive Expiratory Pressure (PEP) (e.g., PEP Valve, PEP Mask)
   - Oscillating Positive Expiratory Pressure (OPEP) (e.g., AsapRela, AirPulse, Long Tube)
   - Chest Physical Therapy (CPT)/Postural Drainage
   - High-Frequency Chest Wall Oscillation (Heimovest® The Vest®, InCourage®, SmartVest®, Monovest®)
   - Active Cycle of Breathing Techniques (ACBT)/Autogenic Drainage
   - Bronchodilators (e.g., Levibuterol, Albuterol, I Nebulizers)
   - Hypertonic Suer Solution Concentration:
   - Other:

2. To what extent do you agree or disagree with each of the following statements? Check the box that applies to each statement. Not at All True √ A Bit True √ Occasionally True √ Somewhat True √ Completely True √
   - I understand what airway clearance is.
   - I am not able to explain the benefits of airway clearance.
   - I believe airway clearance is an important part of my care routine and makes me healthier.
   - I am not aware of all of the airway clearance options that are available to me.
   - I am willing to work closely with my care team to find the airway clearance routine that is best for me.
   - My airway clearance skills need improvement.
   - I consistently do my airway clearance routine each day.
   - I am unhappy/disappointed with my current airway clearance routine.
   - I continue doing my airway clearance routine when I am traveling.
   - I am not comfortable doing airway clearance in front of friends.
   - I am able to set aside time each day to perform airway clearance.
   - My airway clearance routine gets in the way of doing things I want to accomplish each day.
   - I am able to find time each day to perform airway clearance.

3. Which of the following might get in the way of doing your current airway clearance routine or adding a new airway clearance option? Check all items that apply to your situation.
   - High out-of-pocket cost
   - It’s difficult to set up
   - It’s too complicated
   - It takes too much time
   - It disrupts my daily life
   - I can’t travel with it
   - Prefer to exercise

4. Use this space to add any thoughts not addressed in the sections above.
# Your Action Plan

A copy of this Action Plan should be provided to the patient at the conclusion of the clinic visit for reference at home. Retain a copy in the patient's chart for review during the next visit.

## Year Care Team

**Recommended Airway Clearance Routine:**

- Exercise (Please specify which type):
- Huff Coughing
- Positive Expiratory Pressure (PEP) (e.g., PEP Valve, PEP Mask)
- Oscillating Positive Expiratory Pressure (OPEP) (e.g., Acapella, Aerobika, Lung Fluter)
- Chest Physical Therapy (CPT) / Postural Draining
- High-Frequency Chest Wall Oscillation (HFCWO) (e.g., Vest, InCourage, SmartVest, Monarch)
- Active Cycle of Breathing Techniques (ACBT) / Autogenic Drainage
- Bronchodilators (Albuterol, Levalbuterol, and Ipratropium)
- Hypertonic Saline Concentration: _____ %
- Other:

**How Many Times Each Week?**

**How Long Each Time?**

## Conducting Airway Clearance

- Chest Physical Therapy
- Using the Acapella
- Using the Aerobika
- Postural Draining
- Vest Therapy (mobile)
- Vest Therapy (stationary)
- Huff Coughing

## Peer Support Videos

- Active Cycle of Breathing
- Hypertonic Saline
- Fitting airway clearance into a busy day
- How airway clearance improves overall well-being
- Doing airway clearance while "on the go"
- Doing airway clearance while on vacation
- Why Airway Clearance is Needed
- Bronchiectasis: Role of Airway Clearance Techniques
- Overview of Airway Clearance Techniques

## Conducting Airway Clearance

- Doing manual chest physiotherapy (from CF Foundation)
- Using the Acapella (from Smith Medical)
- Using the Aerobika (from International Biophysics Corporation)
- How to Use the InCourage Vest (from Respironics)
- Performing Huff Cough (from University of Michigan)
- Active Cycle of Breathing (from CF Foundation)

## Next Clinic Visit

**Date:**

**Time:**

I have reviewed this Action Plan with my care team and I agree to follow through with each of the recommended items listed above.

**Healthcare Provider Signature:**

**Your Signature:**

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Video Library

Use these helpful, short videos to get educated about various airway clearance options.
Links to videos and websites are owned and operated by third parties, which are under their exclusive control, provided for your information and convenience at your own risk and are not an endorsement of the linked content or such third parties.

Conducting Airway Clearance

- Exercise
- Huff Coughing
- Bronchodilators
- Hypertonic Saline
- Using a Nebulizer

About Airway Clearance

- How Airway Clearance Improves Overall Well-being
- Review of Airway Clearance Therapy
- Tecnicas para la limpieza de las vías respiratorias
- What is Bronchiectasis?
- Que es la Bronquiectasia
- Bronchiectasis: The Role of Airway Clearance Therapy
High Frequency Chest Wall Oscillation (HFCWO)
• Series of oscillating motors creating different pressure waveforms to provide disruption in the lungs and mobilize secretions

• Criteria in confirmed bronchiectasis patients:
  – Daily productive cough for at least 6 months
  – Two or more exacerbations/infections requiring abx per year
  – Failure of other modes of airway clearance

• No single vest is better than the other, each has their own physical features that may appear to certain patients (portability, weight, etc.)
Afflovest

- FREES PATIENTS.
- MOBILIZES SECRETIONS.
- ADVANCES THERAPY.
High Frequency Chest Wall Oscillation (HFCWO)

- Three modes of oscillation treatment
  - Percussion
  - Vibration
  - Drainage

- Three levels of intensity
  - 5 Hz
  - 13 Hz
  - 20 Hz
Summary

• Bronchiectasis is underdiagnosed and thus often incorrectly treated
• When diagnosed, bronchiectasis is often under treated
• In addition to treating the underlying cause, an aggressive airway clearance regimen is crucial to improving patients quality of life in both the short and long run
Summary

• The RT plays a vital role in the successful management of bronchiectasis patients
• Airway clearance regimens are individualized to each patient and understanding the basis behind each method is important for adherence
• Patient education is key for buy-in and optimal compliance
I'm happy to sidestep any questions you may have.
Thank You

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Every life deserves world class care.