

Managing End Stage Pulmonary Disease

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Lung Functions

- Gas Exchange
- Filtering and Detoxification
- Metabolic/Synthetic Functions
 - Angiotensin conversion

Lung Anatomy

- Conducting Airways
 - Conduct air between larynx and alveoli
- Gas Exchange Surface
 - Blood-air interface
 - Two cell layers thick
 - 300 million alveoli
 - Surface area 450 to 1000 square feet

Normal Lung Function and Elasticity

- Lungs tend to collapse to a smaller size
- Chest wall tends to expand outward
- Lung expansion is maintained by a negative pressure in the pleural space
- Lungs distend easily with little pressure

Air Movement In Inspiration

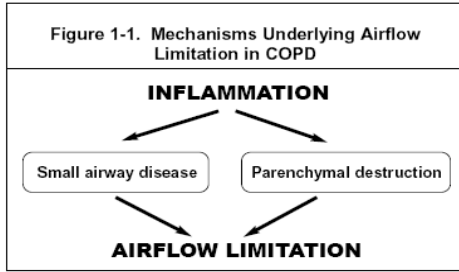
- The diaphragm contracts
 - pushes down on abdominal contents
 - lengthens the thoracic cavity
- Intercostal muscles contract
 - raise and lift ribs outward
 - expand the chest laterally
- Result is negative pressure within the chest
 - Air flows in easily to fill the "empty space"

Air Movement In Expiration

- Respiratory muscles relax
- Chest wall moves inward
- Elastic recoil pulls the lung inward
- Pressure within the chest increases
- Air flows outward passively

COPD

Figure 1-1. Mechanisms Underlying Airflow Limitation in COPD



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Classification of COPD

Figure 1-2. Classification of Severity of COPD

Stage	Characteristics
0: At Risk	<ul style="list-style-type: none"> normal spirometry chronic symptoms (cough, sputum production)
I: Mild COPD	<ul style="list-style-type: none"> FEV₁/FVC < 70% FEV₁ ≥ 80% predicted with or without chronic symptoms (cough, sputum production)
II: Moderate COPD	<ul style="list-style-type: none"> FEV₁/FVC < 70% 50% ≤ FEV₁ < 80% predicted with or without chronic symptoms (cough, sputum production)
III: Severe COPD	<ul style="list-style-type: none"> FEV₁/FVC < 70% 30% ≤ FEV₁ < 50% predicted with or without chronic symptoms (cough, sputum production)
IV: Very Severe COPD	<ul style="list-style-type: none"> FEV₁/FVC < 70% FEV₁ < 30% predicted or FEV₁ < 50% predicted plus chronic respiratory failure

Classification based on postbronchodilator FEV₁. FEV₁: forced expiratory volume in one second; FVC: forced vital capacity; respiratory failure: arterial partial pressure of oxygen (PaO₂) less than 8.0 kPa (60 mm Hg) with or without arterial partial pressure of CO₂ (PaCO₂) greater than 6.7 kPa (50 mm Hg) while breathing air at sea level.

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ASTHMA
Sensitizing agent



Asthmatic airway inflammation
CD4+ T-lymphocytes
Eosinophils

COPD
Noxious agent



COPD airway inflammation
CD8+ T-lymphocytes
Macrophages
Neutrophils

Completely reversible

Airflow limitation

Completely irreversible

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Air Movement In COPD

- Lungs are chronically over-inflated
 - Chest wall and diaphragm move less
 - Less negative pressure is generated in inspiration
 - Patients use accessory muscles or arms to lift and lengthen thoracic cavity
- Elastic recoil is reduced
 - Positive pressure in expiration is reduced
 - Expiration no longer passive but requires effort

Air Movement In COPD

- Airways are narrowed by inflammation
- Airways may collapse
 - Expiration is slower
 - Expiration takes more effort
- Cause of death:
 - infection
 - ventilatory failure
 - heart failure

Palliative Care For COPD Patients

- Positioning to allow lengthening of chest
 - head up
 - support for arms
- CPAP training
 - continuous positive airway pressure
- Cool air in motion
- Oxygen

Management of Severe COPD

Characteristics

- FEV₁/FVC < 70%
- FEV₁ < 30% predicted or presence of respiratory failure or right heart failure

Treatment

- Regular treatment with one or more bronchodilators
- Inhaled glucocorticosteroids if significant symptoms and lung function response or if repeated exacerbations
- Treatment of complications
- Rehabilitation ??
- Chronic oxygen therapy if respiratory failure

Bronchodilators

- Less benefit in end stage disease
- Nebulizers vs Multi-Dose Inhalers +/- spacer
- B2 Agonists
- Anticholinergics
- Theophylline
- Patients may be psychologically dependent

Aerochambers



InspirEase

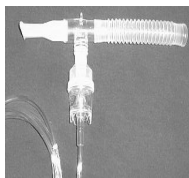


Cleaning and Care of InspirEase

o Cleaning and Care of InspirEase:

- Clean the InspirEase mouthpiece with warm water. (InspirEase is not dishwasher safe)
- Allow InspirEase to air-dry on a clean towel
- Do not clean the collapsible plastic bag. It should be replaced every 4 weeks or as needed
- The mouthpiece should be replaced every six months or when it is no longer working correctly
- Each starter kit comes with a mouthpiece and 3 collapsible bags. Collapsible bags can be bought separately, 3 in a box

What's With Nebulizers?



Traditional Nebulizer
Delivery System



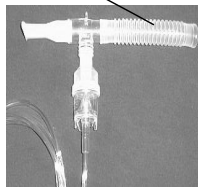
Jet Nebulizer
Delivery System

What's With Nebulizers?

- The traditional nebulizer delivery system can be used for most medications (i.e. albuterol & ipratropium)
 - These medications can also be mixed together & delivered via the nebulizer
- Pulmicort respules can only be administered using the jet nebulizer delivery system
 - Pulmicort respules should NOT be mixed with any other nebulized medications
 - However, other nebulized meds can be given via the jet nebs

Cleaning Nebulizers

- When cleaning the nebulizer, all parts can be cleaned with water and soap, except, the "tail"
- Washing the "tail" can potentially lead to bacterial growth
- Let all the parts air dry



B2 Agonists- Short acting

- Albuterol (Proventil, Ventolin)
 - Inhaler: 90mcg, 104mcg/act
 - Nebs: 0.63mg, 1.25mg/3ml
 - Oral SR: 4mg, 8mg tabs
- Levalbuterol (Xopenex)
 - Nebs: 0.63mg, 1.25mg/3ml
 - Every 6-8 hours
- Pirbuterol (Maxair)
 - Aerosol: 200mcg/inh
 - Every 4 to 6 hours
- Metoprolterenol (Alupent)
 - Aerosol: 0.65mg/act;
 - Nebs: 0.4%, 0.6%mg/3ml
 - Every 4 to 6 hours
- Adverse Effects:
 - Tachycardia, nervousness, tremor
 - Incr. serum glucose, ↓ serum K⁺

B2 Agonists – Long Acting

- Salmeterol (Serevent)
 - Aerosol discs: 50mcg/act
 - Every 12 hours
- Formoterol (Foradil)
 - Aerosol caps: 12mcg/act
 - Every 12 hours
- Adverse Effects:
 - Tremors, nervousness
 - Incr. serum glucose, ↓ serum K⁺

Anticholinergics

- Ipratropium
 - Inhaler: 18mcg/act
 - Aerosol: 0.02%, 0.03%, 0.06%
 - Every 6-8 hours
- Tiotropium (Spiriva)
 - Inhalation: 18mcg/capsule
 - Once a day dosing
- Adverse Effects:
 - Systemic absorption is low; Palpitations, nervousness

Combination Products

- Fluticasone & Salmeterol (Advair)
 - 100-50 mcg/dose
 - 250-50 mcg/dose
 - 500-50 mcg/dose
 - Every 12 hours
- Budesonide and Formoterol (Symbicort)
 - 80/4.5
 - 160/4.5
 - Turbo Inhaler 100 & 200
 - Every 12 hours
- Albuterol Ipratropium
 - Combivent Inh: 103-18mg/act
 - Duoneb Nebs: 2.5-0.5 mg/3ml

Corticosteroids

- Fluticasone (Flovent)
 - Inh: 44mcg, 110mcg, 220mcg/inh
 - Twice a day
- Budesonide (Pulmicort Turbohaler)
 - Aerosol Inh: 200mcg/inh
 - Nebs also available
 - Once or twice a day

Palliative Care For COPD Patients

- Corticosteroids - use generously
- Benzodiazepines – schedule, low non-sedating doses
- Low dose systemic opioids
- Secretions: “to dry or not to dry”
 - encourage hydration ?
 - avoid drying agents until dying

Treatment of Exacerbation

- Antibiotics in a patient that has no pneumonia on CXR
 - help resolve an exacerbation or decrease the risk of future deterioration
 - Beneficial if:
 - increased SOB, increased sputum volume and purulence
 - Antibiotics:
 - amoxicillin, smz/tmp, doxycycline
 - fluoroquinolone

COPD

- Pain
 - Generally due to chest muscle fatigue

- NSAIDs or acetaminophen often beneficial

Restrictive Lung Disease

- Lungs are stiff
 - reduction in lung tissue elasticity reduces the lung recoil (and therefore the lung volume) that causes the amount of air-intake/inhaled within the lungs
- Inspiration requires work, expiration is passive
- Work of breathing increases
- Volume of each breath is decreased
- May compensate with increased rate
- Cause of death:
 - ventilatory failure
 - infection

Palliative Care In Restrictive Disease

- Positioning – based on patient preference
- Oxygen – may desaturate with activity
- Steroids - may help early, less benefit late
- Bronchodilators have little benefit
- Benzodiazepines for dyspnea
- Opioids for dyspnea
- Cough management
 - usually suppress if non-productive

Dyspnea

- Dyspnea is the sensation of difficult breathing
 - Mechanism is not entirely clear
- Occurs in up to 70-80% of hospice patients

Dyspnea

- **May represent abnormal relationships between neuromuscular effort and air movement in the lung**
 - Respiratory muscle fatigue may contribute
- **Patient perception of the meaning of the underlying**
 - Cause may lead to increasing distress
- **May occur without lung or cardiac disease**

Palliative Care Of Dyspnea

- **Identify potentially reversible causes**
 - bronchospasm, CHF, infection
 - PE, effusions, anemia
- **Treat reversible causes if appropriate**
- **Evaluate for use of oxygen or a fan**
- **Consider bronchodilators**
 - especially if bilateral wheezing
- **Consider corticosteroids if**
 - COPD, PF, asthma or tumor are present

Treatment Of Potentially Reversible Causes Of Dyspnea

Cause of Dyspnea	Physical Signs	Treatment Options	Route
Bronchospasms	Wheezing Decreased air movement Nonproductive cough	Bronchodilators – adrenergic and anticholinergic Corticosteroids	Inhaled Nebulized IV, PO, SC, PR
Congestive Heart Failure	Inspiratory rates Edema Elevated JVP Orthopnea	Diuretics Cardiac medications Morphine Oxygen	PO, IV, Neb PO PO, SL, SC, IV, PR Inhaled
Bacterial Pneumonia	Productive cough with changing sputum Localized rales or consolidation, fever, chills	Antibiotics, Expectorants, Cautious hydration Morphine for pain/dyspnea Oxygen	PO PO PO, IV, SC PO, SL, IV, SC, PR Inhaled

USE OF OXYGEN

- For hypoxemic patients oxygen may improve cognition or decrease confusion.
- For normoxic patients who feel short of breath, oxygen is seldom beneficial.
 - Treat for anxiety
 - Try a fan first if oxygen saturation are normal
- For hypoxemic patients who are not alert,
 - Some feel offers little benefit except postponing death

Benzodiazepines for Dyspnea

- Schedule ATC
- Anxiety is usually a component of feeling short of breath
- Start low and titrate up
 - Lorazepam (Ativan) 0.25 to 0.5mg every 6-8 hours
 - Diazepam (Valium) 1 to 2.5mg every 8 -12 hours
- Goal is to relieve anxiety and calm patient should not sedate at appropriate doses

Opioids For Dyspnea

- Use Morphine unless contraindicated
 - Oxycodone or hydromorphone can be used if patient has renal failure
- Start with Morphine 2.5mg PO every 1 hour PRN if opioid naïve
 - Titrate up as needed
- Schedule every 4 hours if needed frequently
 - Use breakthrough doses as needed
- Seldom need more than 5-10mg every 4 hours
- Increase benzodiazepine before going above 10-20mg of morphine every 4 hours

Symptomatic Treatment Of Dyspnea

Treatment	Indication	Dose	Frequency	Route
Oxygen	Hypoxemia	2-5 liters/min	PRN or continuously	Inhaled
Fan or cool air toward face	Dyspnea with or without hypoxemia	As tolerated	PRN or continuously	Environmental
Morphine* (oral)	Dyspnea	If opioid naïve, 2.5 mg, titrate up to 10 mg as needed If on opioids for pain, 1/6 to 1/10 of daily dose	4 to 6 hrs and/or PRN q 1-2 hrs PRN q 1-2 hrs	PO, SL, PR

Symptomatic Treatment Of Dyspnea

Treatment	Indication	Dose	Frequency	Route
Morphine* (parenteral)	Dyspnea	Parenteral dose is 1/3 of oral or rectal dose. If opioid naïve, 1-2 mg, titrate up as needed If on opioids for pain, 1/6 to 1/10 of daily dose	4 to 6 hours and/or PRN q 1-2 hrs PRN q 1-2 hrs	SC, IV
Lorazepam**	Anxiety	0.25 to 1 mg Titrate up to 2 mg if needed	4 to 8 hrs Or PRN q 4 hrs	PO, IV, PR

Symptomatic Treatment Of Dyspnea

Treatment	Indication	Dose	Frequency	Route
Corticosteroids	Bronchospasm COPD	Prednisone 20 to 60 mg Dexamethasone 4 to 12 mg	Daily Once daily or divided doses Avoid bedtime	PO PO, SC, IV, PR
Bronchodilators Adrenergic Anticholinergic	Bronchospasm Airway obstruction COPD	Metered dose inhaler or nebulizer	2 to 12 hrs and/or PRN q 4 hrs	Inhaled aerosol

Risk of Treating Dyspnea

- Easy to overdose if treating physical signs rather than symptoms
- Treat patient reports of discomfort
 - not respiratory rate or effort
- Goal is to relieve subjective symptoms of distress
- Not necessary to decrease respiratory rate for comfort

Respiratory Secretions

- **Sources:**
 - Sinuses or upper respiratory infection
 - Saliva
 - Infection or inflammation in lung
 - Increased vascular permeability
 - Normal pulmonary fluid (mucus)

Secretion Management Issues

- Removing secretions is the first choice
- Thin secretions are easiest to remove
- Pulmonary mucus is usually a good thing
- Cough may be exhausting or ineffective
- Some secretions can be prevented, some can't
- Dried secretions can cause problems
 - difficult to bring up
 - may block airways
 - infection

Respiratory Secretions

- **Can the patient cough?**
 - If yes, thin secretions
 - If no, decrease secretion production
- **Is coughing exhausting the patient or causing pain?**
 - If yes, suppress cough and dry secretions
- **Are thick secretions a risk for the patient?**
 - Ex – Trach, ALS, chronic bronchitis
 - If yes, use drying agents cautiously
- **How long is the patient likely to live?**
 - If more than 48 hours, try to avoid drying agents

Treatment of Secretions

- **To decrease secretion viscosity (thin secretions):**
 - Systemic Hydration
 - Guaifenesin 200-400 q 4 hrs or 600-1200mg q 12 hrs
 - Saline Nebs
 - Mucomyst Nebs
- **To decrease mucus and saliva secretion:**
 - **Atropine** 2-4 drops 1% ophthalmic solution PO or SL every 2-4 hours PRN
 - **Scopolamine** 1-3 patches (onset of action 4-12 hours)
 - **Hyoscyamine** 0.125-0.25 mg po q 4 hrs
 - **Glycopyrrolate** 0.1-0.2mg SQ, IV q8-12h or every 4 hours PRN
 - **Diphenhydramine** 25-50 mg PO, IV, IM every 6 hours PRN

Treatment of Cough

- **If productive – usually due to infection or bronchitis**
 - Thin secretions
 - Suppress with opioids if needed for sleep/fatigue
- **If non-productive**
 - Evaluate for bronchospasm or reflux
 - Suppress if not reversible
 - **Hydrocodone**
 - 5 mg P.O. every 4-6 hrs PRN
 - **Morphine**
 - 2-5 mg P.O. every 4 hrs PRN
 - **Lidocaine** (1-2% solution for injection)
 - 3ml nebulized every 4 to 6 hrs

Suggested Reading

- Oxford Textbook of Palliative Medicine, 2nd ed. p 583-616.
- A Physician's Guide to Pain and Symptom Management in Cancer Patients by Janet L. Abraham MD. Johns Hopkins Univ. Press. p 297-305
- Respiratory Physiology - The Essentials by John B. West MD. The Williams and Wilkins Co.
- Respiratory distress in the terminally ill patient by Sam Ahmedzai. Respiratory Disease in Practice. 1988; Oct/Nov: 20-29.
- Advanced Lung Disease - Palliation and Terminal Care by John Hansen-Flaschen MD. Clinics in Chest Medicine 18(3): 645-655.

CASE

55 year old male with end stage COPD
 2 L/m oxygen
 Advair 1 unit BID
 Combivent 2-4 puffs q4hprn
 Theophylline 300mg BID
 Tenacious yellow sputum:
 Becoming fatigued from effort to get sputum up.
 Very short of breath
 Unable to sleep or eat
 What would you suggest?
