Management of the Cystic Fibrosis Patient
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Where discoveries are delivered.

Learning Objectives
1. Describe the symptoms associated with Cystic Fibrosis (CF).
2. Describe how CF is diagnosed.
3. Describe the multi-system effects of CF.
4. Describe the typical management and treatment of CF.
5. Describe potential complications of CF.
6. Describe patient readiness for discharge home.

Etiology and Prevalence
• CF caused by recessive gene
  – Child must inherit two copies of defective gene—one from each parent
  – There is a 1 in 4 chance that the child of two carriers will have CF.
• 30,000 in United States
• 70,000 worldwide
• More than 10 million Americans have single copy of a CF mutation and are unaware!
Diagnosis of Cystic Fibrosis

• Diagnosis based on sweat test, genetic testing, and/or clinical presentation.
• Most commonly diagnosed in first year of life
  – Meconium ileus
  – FTT
  – Recurrent respiratory infections
• Adult diagnosis increasing (genetic testing advances)
• Some cases challenging to diagnose

Disease of Childhood?

![Figure 3: Number of CF Patients, 1986–2011](figure3)

Survival in Cystic Fibrosis

![Average Life Expectancy in Cystic Fibrosis](figure4)
Pathophysiology

- Defect in CFTR (Cystic Fibrosis Transmembrane Conductance Regulator)
- Affects sodium chloride ion transport
- Results in abnormal mucus
  - Excessive mucus production
  - Mucus thick and viscous
  - Abnormal mucus affects many systems

CF: A Multi-System Disease

Pulmonary System

Pulmonary Manifestations

- Bronchiectasis
- Bronchitis
- Bronchiolitis
- Pneumonia
- Ateletasis
- Hemoptyis
- Pneumothorax
- Reactive Airway Disease
- Cor Pulmonale
- Respiratory Failure
- Mucoid impaction of bronchi
- Allergic bronchopulmonary aspergillosis (ABPA)

The Vicious Cycle: Obstruction, Infection, and Inflammation
Respiratory Management

• Quarterly evaluation of pulmonary status at CFF accredited CF Center
• Long term maintenance therapies
• Consistent infection control procedures
• Rapid and aggressive treatment of pulmonary exacerbations
• Management of respiratory complications.

Maintenance Respiratory Therapies

• Airway clearance therapy
  – Chest physiotherapy (CPT)
  – Exercise
  – Flutter/acapella valve
  – IPV
  – “The Vest” by Hillrom or other airway clearance vest device.

Maintenance Respiratory Therapies, cont.

• Long term antibiotics/antifungals
  • Oral antibiotics
  • Inhaled antibiotics
    – Tobramycin 300 mg BID (Tobi)
      • FDA approved for admin. with Pari LC
      • Therapy targeted at gram negative bacteria
      • Decreased adherence due to nebulization time
      • May cause bronchospasm
      • 30 min neb time bid
    • New Tobi Podhaler
      • 7 min admin time
Maintenance Respiratory Therapies, cont.

- Inhaled antibiotics, cont.
  - **Aztreonam** 75 mg TID (Cayston)
    - FDA approved with Altera (e-flow) device ONLY
    - May see improved adherence due to decreased nebulization time of 3-5 minutes.
    - Note: DO NOT use Altera device for other medications without MD approval as doses require adjustment and may not be FDA approved with Altera.
    - Also targeting gram negative bacteria.
    - May also cause bronchospasm.
  - Compassionate use of non-FDA antibiotics may be seen in special circumstances.

- Mucolytics
  - Dornase alfa (Pulmozyme)
    - Usual dose 2.5 mg daily, may be increased to twice daily during exacerbation.
    - Requires separate nebulizer cup due to incompatibility with most other medications.
  - Acetylcysteine (Mucomyst)

- Bronchodilators
- Hypertonic saline (Hyper-Sal) 7%
- Other: albuterol, xopenex, atrovent, advair, spiriva
- Routine immunizations

Time Required for Daily Treatment Regimen

- Airway clearance therapy
  - Minimum 1 hour daily

- Inhaled medications
  - Tobramycin 30 min twice daily
  - Aztreonam 15 min three times daily
  - Dornase alfa 30 min daily
  - Hypertonic saline 30 min twice daily
  - Other inhaled medications
Time Required for Daily Treatment Regimen, cont.

- Oral medications
  - 30 min daily
- CFRD (CF related diabetes) management if applicable
- Total time required 3-5 hours daily when well
- With CFRD +1 hour=4-6 hours daily
- With IV therapy >8 hours daily

Patient adherence to treatment regimen

- Decreased adherence to CF treatment regimen common
- Adolescents/young adults high risk
  - National reported outcomes indicate accelerated decrease in lung function
- Inhaled aztreonam (Cayston)
  - Centers surprised with extent of non-adherence
- Non-judgmental approach

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Pulmonary Exacerbations
Pulmonary Exacerbations

- Progressive decline of lung function with acute worsening of respiratory symptoms, called "pulmonary exacerbations".
- General definition not yet developed

Clinical features of exacerbation:
- Increased cough
- Increased sputum production
- Shortness of breath
- Chest pain
- Loss of appetite
- Weight loss
- Lung function (PFT) decline

Flume, Patrick A. et al. Cystic Fibrosis Pulmonary Guidelines. Am J Respir Crit Care Med. 2009; 180: 802-808

Treatment of Exacerbations

- Loss of 5-10% FEV₁ may respond to oral antibiotics
- Loss of >10% usually requires IV therapy
- CFF data shows patients respond best with rapid and aggressive treatment of exacerbations.
- Routine treatments should be continued.

Hospitalization vs. Home IV

- Many factors affect decision
  - Degree of lung function decline
  - Family support
  - Living situation
    - Homeless
    - College dorms
    - Access to electricity, refrigeration, telephone
  - Insurance coverage
  - Work and school schedule
  - Family responsibilities
  - Adherence
  - Access to airway clearance therapy
  - Transportation

UC San Diego Health System
Treatment Starts with Sputum Culture

• Sputum obtained with each exacerbation
• Treatment based on identification of microorganisms.
• Most common:
  – Pseudomonas aeruginosa
  – Staphylococcus aureus (sensitive or MRSA)
  – Other gram negative organisms (Stenotrophomonas, Achromobacter)
  – Burkholderia cepacia (infection control implications)

Sputum Culture & Sensitivity

• How are cultures used?
• Limitations of microbiology data
• Petri dish vs. human immune system

Parenteral Antibiotics for treatment of CF Exacerbation

• Doses often higher due to altered pharmacokinetics caused by CF
• Two-drug combination therapy used to reduce risk of inducing resistance
• Clinical response often not seen for 4-7 days after treatment begins
• Treatment usually necessary for 2-4 weeks
Common IV Antibiotics

- Aminoglycosides
  - Tobramycin, gentamicin
  - Watch for s/s ototoxicity (tinnitus, vertigo, nausea, hearing changes)
- Beta-lactams
  - Ceftazidime, meropenem
  - Longer or continuous infusions increasingly common
  - Has been used via inhalation (off label)
- Anti-staph
  - MRSA vs sensitive staph

Airway Clearance

- Equally important to IV antibiotics
- Goal: 4 treatments/day
- Patients should be encouraged not to refuse treatments!
- RT is an excellent resource in treatment of CF patients
- Patients should be encouraged to get out of bed and walk!!

How to Determine Response to Treatment

- Return of FEV₁ towards baseline
  - For patients with low baseline FEV₁ (<1 liter) clinical exam more important
- Improvement in clinical exam
- Symptom improvement
  - Decreased cough
  - Decreased shortness of breath
  - Decreased sputum production
  - Increased activity/exercise tolerance
  - Decreased oxygen requirement
When is IV therapy discontinued?

- Return to baseline FEV₁
- Plateau in symptoms or pulmonary function
- Max duration 4 weeks
- Side effects
- Impaired kidney or liver function
- Patient request

What if patient does not respond?

- Change antibiotic regimen
- Evaluate other factors
  - Adherence
  - Airway clearance therapy
  - Reassess sputum cultures
  - Rest
- Cycle treatment
  - Month on/month off
  - Usually do not accept new baseline FEV₁ until several cycles of treatment.

Special Considerations

- Airway clearance therapy during IV antibiotic treatment
  - Patients with port may be unable to use “The Vest”.
  - May be able to “pad” area over port to enable patient to tolerate ACT.
  - Consider manual CPT or percussor.
  - ACT equally important to IV antibiotics.
Other Respiratory Complications
• Hemoptysis
• Pneumothorax
• ABPA
• Respiratory Failure

Hemoptysis
• Minor Hemoptysis
  – Common in patients as they get older
  – Often sign of pulmonary exacerbation
  – Treatment with Vit K, observation, antibiotics
• Major Hemoptysis
  – >240 ml (8 oz) blood in 24 hours or >100 ml/day for >3 days
  – Uncommon (<10% patients)
  – Requires emergent care, interventional radiology

Pneumothorax
• Patient usually presents with acute chest pain and shortness of breath
• Usually requires treatment with chest tube
• Small, asymptomatic pneumo may be managed with observation
• Management
  – Monitor sats, give oxygen (consider co2 retention), CT, local anesthesia, pain mgt, antibiotics, gentle CPT
ABPA

- Allergic Bronchopulmonary Aspergillosis (ABPA)
- Caused by an allergic reaction to aspergillus colonizing the sputum
- Treated with antifungals and/or steroids
- Diagnosed with sputum culture and aspergillus specific IgE values
- Patient often has severe asthma s/s in addition to cystic fibrosis
- Can have major impact on quality of life
- May be treated with xolair (anti IgE)

CF: A Multi-System Disease

Gastrointestinal System

- Malnutrition
- Steatorrhea
- Abdominal pain
- Distension
- Excessive flatus
- GERD
- Rectal prolapse
Nutritional Implications of CF

- 80-90% CF patients pancreatic insufficient
- Goal of enzyme replacement therapy is decrease malabsorption.
- Enzymes should NOT be reduced or discontinued to treat DIOS.
- Enzymes should be taken prior to meals/snacks.
- Partial or repeat dose if >45-60 min. last taken
- Pancreatic enzymes should be stored at room temp.

Nutritional implications cont.

- Energy requirements approximately 2x
- Protein requirements approximately 1.5-2X
- Energy needs STRONGLY INFLUENCED by severity of lung disease.
- Strong correlation between weight (BMI) and lung function (FEV1)
Pancreatic System

Abnormal glucose tolerance
CF Related Diabetes (CFRD)
Pancreatic malabsorption

CFRD
- Cystic Fibrosis Related Diabetes
- Characteristics of Type I and Type II
- Caused by increased mucus resulting in pancreatic scarring, and insulin resistance.
- Almost 50% of CF patients >30 years have CFRD.

CFRD subtypes
- CFRD with fasting hyperglycemia
  - Requires insulin
- CFRD without fasting hyperglycemia
  - Does not always require insulin
  - Often requires insulin during hospitalization
  - Almost always progresses to requiring insulin.
CFRD complications

- Ketoacidosis is rare.
- Macrovascular complications rare.
- Microvascular complications common.

CFRD Treatment

Oral hypoglycemics not used in CFRD due to inconsistent absorption.

Diet is NEVER restricted.

Patient should continue on high calorie, high fat, high protein diet and cover the carbohydrates with insulin.

Carb counting highly recommended.

Infection Control

- Almost all CF patients are on contact isolation
  - MRSA
  - Multi-drug resistant organisms (MDRO)
  - Pan-resistant pseudomonas or other organisms
  - Burkholderia cepacia
Burkholderia Cepacia

- Accelerated loss in lung function common
- Attempt to eradicate with initial colonization
- Requires confirmation at special lab
- Admit to different unit
- Staff members caring for a patient with cepacia should NOT care for other CF patients during same shift.
- Last appointment for clinic
- Prevented from attendance at all CFF functions
- Social isolation

What's New in CF?

- Potential new therapies in pipeline.
- CFTR potentiator or corrector therapies in clinical trials.
- New antibiotics.
- New methods of delivery for current medications.

Thank you

- Thank you from the entire Adult Cystic Fibrosis Team for taking such great care of our patients.
- We appreciate the high quality care and kindness that you show to our patients.
- Because of all of you……
There are many reasons to be HOPEFUL!!

Through quality care and continued research, persons with cystic fibrosis can rise above their challenges and achieve great things!!

UCSD Adult CF Team

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