This Web cast has 2 parts:
• Part 1 – Keeping Lungs Healthy
• Part 2 – Treating Disease

Navigating the Hills and Valleys of CF Lung Disease: Treating Disease

August 16, 2007

| Adding *tomorrows* every day.

This Web cast is supported by an unrestricted educational grant from Genentech, Inc.
Navigating the Hills and Valleys of CF Lung Disease

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“The Problems” with CF Lungs

- Obstruction
- Infection
- Inflammation
- Progressive lung damage
“The Problems” with CF Lungs

Normal

CF
Signs & Symptoms

- Cough
- Increased mucus
- Shortness of breath
- Wheezing
- Decreased lung function
- Change in lung sounds/physical exam
- “Feel bad” or “run down”
Chronically Infected vs. Exacerbation

• They don’t “go away”/“always there”
• Maintenance therapies help keep the chronic bacteria “under control”
  – That’s why doing treatments is so important!
• Sometimes the bacteria “flares up”
• “Pulmonary Exacerbation”
Pulmonary Exacerbation

• A tough question
• Different CF care providers have different definitions
• Frustrating
• We developed a standard definition of pulmonary exacerbation
Pulmonary Exacerbation

• Lung function in our children with CF below national average!
• Reviewed medical literature
• Everyone received a “score” at each visit
• If “score” was above a certain number, exacerbation was diagnosed and treated
• Score and lung function tracked
Pulmonary Exacerbation Scoring Sheet

Type of Visit  □ Followup  □ Sick  □ Annual

Patient Initials ______  Patient Age _____  Physician _______  Date __________

Systemic Symptoms/Signs:

1. Fevers > 38°C (100.4°F) in the prior 2 weeks?
   No = 0  Yes = 1

2. Malaise or fatigue in the prior 2 weeks?
   No = 0  Yes = 1

3. Any increased or new school or work absenteeism in the prior 2 weeks?
   No = 0  Yes = 2

4. Anorexia or poor appetite in the prior 2 weeks?
   No = 0  Yes = 1

5. Wt. Loss (≥ 5%) or poor wt. gain compared to last clinic visit (or in the last 3mo.)
   No = 0  Yes = 2

SUM OF SYSTEMIC SYMPTOM SCORES: ______
Pulmonary Symptoms/Signs:

1. Increased cough (frequency, duration or intensity) for ≥1 week?
   None = 0  Mild = 1  Significant = 2

2. Major change in sputum (new onset, increased, change in consistency) or change in chest congestion for ≥1 week?
   None = 0  Mild = 1  Significant = 2

3. Increased DOE or SOB at rest?
   No = 0  Yes = 2

4. Change in chest exam (wheezes, crackles, rhonchi, decreased air entry) or Increased WOB or Respiratory Rate?
   No = 0  Yes = 2

SUM OF PULMONARY SYMPTOM SCORES: _____ (0-8)
Objective Measurements:
1. Decrease in FEV1 (compared to highest value of the prior six months 6 months)?
   - $< 10\% = 0 \quad > 10\% = 3 \quad > 15\% = 5$
2. New Chest Radiographic Abnormality?
   - None $= 0$
   - Increased air trapping, mucus or plugging or bronchiectasis $= 1$
   - New atelectasis or infiltrate $= 2$
   - Pneumothorax $= 5$
3. Hemoptysis?
   - None $= 0$
   - Streaked $= 3$
   - Increased or new onset $= 5$
4. Decreased SaO$_2$ from baseline (compared to the highest value of the prior 6 months)?
   - $< 4\% \text{ change} = 0$
   - $> 4\% \text{ decrease} = 2$
   - $> 10\% \text{ decrease} = 5$

**SUM OF OBJECTIVE MEASUREMENT SCORES:** _____ (0-5)

**TOTAL PULMONARY EXACERBATION SCORE:** _____ (0-17)
Akron Children’s Hospital CF Median FEV$_1$

- 6-13 Years
- 13-18 Years

<table>
<thead>
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<th>Year</th>
<th>6-13 Years</th>
<th>13-18 Years</th>
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<tbody>
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<td></td>
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<td>2006</td>
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Results

• The score is now “standardized” into our clinic visits
• Lung function continues to improve in our children with CF
• The score helps educate about exacerbations
Treating Pulmonary Exacerbations

- Severity of the symptoms
- Severity of CF - baseline CF lung health
- Recent and past throat or sputum cultures
- Past responses to different antibiotics
- Insurance/financial
- Following plan of care
- Support systems
- School/work situations
Treating Pulmonary Exacerbations

• Antibiotics
  – Oral
  – Inhaled
  – Intravenous (IV)

• Clearing the mucus (more or different airway clearance)

• Aggressive nutrition

• Monitoring the response
Choosing An Antibiotic

• Type of bacteria
• Age
• Previous antibiotics taken
• Insurance coverage
Oral Antibiotics

• May be used if symptoms are more mild and bacteria sensitive to medication
• Treatment a minimum of 2 weeks and often longer
• Follow up clinic visit to make sure you’re back to baseline
Inhaled Antibiotics

• Typically used along with oral or intravenous (IV) antibiotics
• Recommended to take after all other inhaled medicines and airway clearance
• Convenient & well tolerated
Intravenous (IV) Antibiotics

• Antibiotics are infused directly into the vein
  – Most effective way to treat a severe exacerbation
• More than one antibiotic is often used
• Length of treatment usually 2-3 weeks, but dependant on response to treatment
**Gram Stain**

*05/19/07* Greater than 25 polymorphonuclear leukocytes per low power field

Less than 10 epithelial cells per low power field

Moderate Gram negative rods

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**Sputum Culture**

*05/20/07* Normal oropharyngeal flora present.

Isolate 01 Pseudomonas aeruginosa (m)

Many

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<table>
<thead>
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<th>Isolate</th>
<th>ISO# 01</th>
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<tr>
<td>Antibiotic</td>
<td>KB</td>
</tr>
<tr>
<td>Aztreonam</td>
<td>S</td>
</tr>
<tr>
<td>Ceftazidime</td>
<td>S</td>
</tr>
<tr>
<td>Ciprofloxacin</td>
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<tr>
<td>Gentamicin</td>
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<td>Meropenem</td>
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<td>Piperacillin</td>
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<tr>
<td>Ticarcillin</td>
<td>S</td>
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<tr>
<td>Tobramycin</td>
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</tbody>
</table>

*S=Sensitive I=Intermediate R=Resistant  MIC results are reported in ug/ml*
Common Antibiotics

- Oral - Cipro, Bactrim, Doxycycline, Keflex
- Inhaled - TOBI®, Cayston® (2010)
- IV - Tobramycin, Ceftazadime, Meropenem, Aztreonam
Time on Antibiotics

- Goal: back to baseline
- Duration is guided by response, e.g., symptoms and monitoring
- Important to finish full antibiotic course - even if you are feeling better to avoid organism resistance!!!!
When to Stop Treatment

• How you/your child feels (appetite, activity, breathing, cough, mucus, etc.)
• Side effects of medications
• Laboratory tests (PFT, X-ray, etc.)
• Physical exam (lung sounds, vital signs, etc.)
Home Therapy

• **Advantages**
  – Convenience
  – Cost

• **Disadvantages**
  – Time consuming
  – Less monitoring of response
  – Other conditions (CFRD, asthma)
  – IV access
  – Telephone access
Hospitalization

• **Advantages**
  – Closer monitoring
  – Rest for home caregivers
  – Other conditions cared for (CFRD, asthma)
  – 24 hour support
  – May get better faster

• **Disadvantages**
  – Regimented
  – Cost
  – Isolation
  – Missing activities
  – Different caregivers
  – Risk of new infection
Not Feeling Better

- Don’t wait - be timely
- Ask questions
- Communicate changes
- Follow up clinic visit(s) is extremely important
- People with CF and families are the center of the CF team
Pulmonary Exacerbation
Preventing an Exacerbation

• Follow your plan of care….

BUT

• Sometimes you just get sick & not all exacerbations can be prevented
• When you’re feeling ill, call your CF care center
Order of Inhaled Medications

• Bronchodilators
• Get the mucus out – Pulmozyme®; hypertonic saline
• Airway clearance
• Inhaled antibiotics - TOBI®, Cayston® (2010)
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Navigating the Hills and Valleys of CF Lung Disease

- Peak of Health
- Feeling a little tired
- Loss of appetite
- Coughing more
- Pulmonary Exacerbation
- Hills and Valleys of CF
Therapies for Cystic Fibrosis

Thanks to Cystic Fibrosis Foundation-supported research, people with cystic fibrosis (CF) have more therapy options than ever before. There are drugs to help clear the thick CF mucus from airways, reduce inflammation and aerosolized antibiotics for CF. The Foundation is dedicated to investing in promising research that will lead to more new treatments and a cure for cystic fibrosis.

- **Clearing the Airways**
  - Inhaled Medications
  - Antibiotics
  - Other Drugs
- **Implanted Devices**
- **Nutrition & Eating Right**
- **Alternative Therapies**

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**Clearing the Airways**

People with cystic fibrosis do airway clearance techniques (ACT) to loosen and get rid of the mucus from the lungs. Clearing mucus helps to reduce the severity of lung infections and improve lung function.

Some airway clearance techniques require help from family members, friends or therapists. Adults with cystic fibrosis can do many airway clearance techniques themselves.

One technique is called “postural drainage and percussion.” People with cystic fibrosis sit, stand or lie in a position that will help free up mucus as their chest and back are pounded or clapped. Sometimes a patient will use a mechanical “vest,” or blow into a device that shakes the mucus loose, to help clear their airways.
Staying Healthy

Make a Plan to Stay Healthy

Most people with cystic fibrosis are able to manage their disease—with routine treatment and visits to an accredited care center.

Knowing your CF is important to stay healthy. Staff at CF Foundation-accredited care centers partner with people with CF to develop individual treatment plans. These plans typically include high-calorie, high-fat diets, therapies to loosen the clogged mucus from their airways, and mucus-thinning drugs and antibiotics when needed.

By following a treatment plan developed with their CF care center team, many people with CF can slow down the progression of their disease. A healthier body is better able to deal with bacteria and chronic lung infection.

Avoid the Spread of Germs

Cystic fibrosis puts the airways at risk for lung infections. There are, however, effective ways to lessen the risk. One way is to limit contact with known germ sources.

Although germs are everywhere and cannot be avoided, one of the best ways to keep from catching or spreading germs is through effective hand-washing, whether with soap and water or alcohol-based hand gels.

Everyone with CF should avoid unnecessary contact with people who have a cold or any other contagious illness, and should cough and sneeze into a tissue.

Learn more about methicillin-resistant Staphylococcus aureus (MRSA). It's all about good hand hygiene!
• Originally broadcast in 2007
• Now in 2 parts
  – Keeping Lungs Healthy – Part 1; and
• Please contact your CF Care Center or the CF Foundation to get your questions answered
  – CF Foundation - info@cff.org or 1-800 FIGHT CF
Thank You

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