Many of you have noticed already that you are unable to obtain acetylcysteine, also known as Mucomyst, from your pharmacy. There is a nationwide shortage. A release date was initially thought to be mid May but this has changed and acetylcysteine is still not readily available. Please make your provider aware if you are unable to obtain any so that a temporary solution can be arranged.

Mucomyst/Acetylcysteine

Vitamins and Cystic Fibrosis

Malabsorption of fat-soluble vitamins can occur in patients who have cystic fibrosis, especially in those patients who are pancreatic insufficient. These patients can experience a blockage in the ducts that lead from the pancreas to the small intestine. If these ducts are blocked, then the pancreatic enzymes needed to digest the protein and fat from food cannot be delivered, which can result in malabsorption. The vitamins A, D, E, and K are vitamins that need fat in order to be absorbed; if fats from the diet are not digested or stored properly, these vitamins will not be absorbed well. Fat-soluble vitamins play important roles in one’s overall health:

- Vitamin A – important for vision, gene expression, reproduction, embryonic development, growth, and immune function
- Vitamin D – important for bone health, muscle function, innate immunity, cardiovascular disease, and diabetes
- Vitamin E – antioxidant that helps prevent cell membrane damage
- Vitamin K – cofactor required for the activity of several key proteins in the coagulation (blood clotting) pathways

Products that have been specially formulated to contain fat-soluble vitamins include ADEKs, ABDEKs, and AquADEKs. ADEK vitamins are available as chewable tablets, ABDEKs are available as chewable tablets, softgels, and drops, and AquADEKs are available as softgels and drops. ABDEKs and AquADEKs contain greater amounts of vitamin A, D, and K as compared to ADEKs. These products also contain water-soluble vitamins, such as vitamin B, folic acid, and vitamin C (among others). The labels will give more specific information, and vitamin needs are evaluated by your physician to help decide what may be lacking and how to treat that deficiency.

It is important to remember to take these vitamins with pancreatic enzymes. The enzymes will help the vitamins be absorbed more easily.

<table>
<thead>
<tr>
<th>Content</th>
<th>ADEK chew tab</th>
<th>ABDEK drops</th>
<th>ABDEK chew tab</th>
<th>ABDEK softgel</th>
<th>AquADEK drops</th>
<th>AquADEK softgel</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Fat-Soluble Vitamins</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vitamin A</td>
<td>9,000 IU</td>
<td>4,627 IU/1 mL</td>
<td>16,000 IU</td>
<td>16,000 IU</td>
<td>5,751 IU/1 mL</td>
<td>18,167 IU</td>
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<tr>
<td>Vitamin E</td>
<td>150 IU</td>
<td>50 IU/1 mL</td>
<td>200 IU</td>
<td>200 IU</td>
<td>50 IU/1 mL</td>
<td>150 IU</td>
</tr>
<tr>
<td>Vitamin D</td>
<td>400 IU</td>
<td>500 IU/1 mL</td>
<td>1,000 IU</td>
<td>1,000 IU</td>
<td>400 IU</td>
<td>800 IU</td>
</tr>
<tr>
<td>Vitamin K</td>
<td>150 mcg</td>
<td>400 mcg/1 mL</td>
<td>800 mcg</td>
<td>800 mcg</td>
<td>400 mcg/1 mL</td>
<td>700 mcg</td>
</tr>
<tr>
<td><strong>Water-Soluble Vitamins &amp; Zinc</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thiamin (B1)</td>
<td>1.2 mg</td>
<td>0.5 mg/1 mL</td>
<td>1.5 mg</td>
<td>1.5 mg</td>
<td>0.5 mg/1 mL</td>
<td>1.5 mg</td>
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<tr>
<td>Riboflavin (B2)</td>
<td>1.3 mg</td>
<td>0.6 mg/1 mL</td>
<td>1.7 mg</td>
<td>1.7 mg</td>
<td>0.6 mg/1 mL</td>
<td>1.7 mg</td>
</tr>
<tr>
<td>Niacin</td>
<td>10 mg</td>
<td>6 mg/1 mL</td>
<td>10 mg</td>
<td>20 mg</td>
<td>6 mg/1 mL</td>
<td>20 mg</td>
</tr>
<tr>
<td>Pyridoxine (B6)</td>
<td>1.5 mg</td>
<td>0.6 mg/1 mL</td>
<td>1.9 mg</td>
<td>1.9 mg</td>
<td>0.6 mg/1 mL</td>
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<tr>
<td>B12</td>
<td>12 mcg</td>
<td>4 mcg/1 mL</td>
<td>6 mcg</td>
<td>6 mcg</td>
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<tr>
<td>Biotin</td>
<td>50 mcg</td>
<td>15 mcg/1 mL</td>
<td>100 mcg</td>
<td>100 mcg</td>
<td>15 mcg/1 mL</td>
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<tr>
<td>Folic acid</td>
<td>200 mcg</td>
<td>---</td>
<td>200 mcg</td>
<td>200 mcg</td>
<td>---</td>
<td>200 mcg</td>
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<tr>
<td>Ascorbic Acid (Vitamin C)</td>
<td>60 mg</td>
<td>45 mg/1 mL</td>
<td>100 mg</td>
<td>100 mg</td>
<td>45 mg/1 mL</td>
<td>75 mg</td>
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<tr>
<td>Pathothenic Acid</td>
<td>10 mg</td>
<td>3 mg/1 mL</td>
<td>12 mg</td>
<td>12 mg</td>
<td>3 mg/1 mL</td>
<td>12 mg</td>
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<tr>
<td>Zinc</td>
<td>7.5 mg</td>
<td>5 mg/1 mL</td>
<td>15 mg</td>
<td>15 mg</td>
<td>5 mg/1 mL</td>
<td>10 mg</td>
</tr>
</tbody>
</table>

References:
Vitamins [online]. Leeds Regional Adult and Paediatric Cystic Fibrosis Units, St James’s University Hospital, Leeds, UK. Available from http://www.cysticfibrosismedicine.com
Airway Clearance Therapy: The Vest

The Vest is a widely used therapy for airway clearance. This assists in the removal of retained secretions. There are guidelines for settings that include pressure, frequency and length of time. Each vest company has a set of guidelines to follow based on research available.

Many large Cystic Fibrosis Centers have adopted their own guidelines. The University of Minnesota has adopted vest settings that many of you have expressed interest in.

Minnesota Vest Therapy Settings
Settings 8, 9 and 10. Each at a pressure of 10 for 5 minutes each.
Settings 18, 19, and 20 Each at a pressure of 6 for 5 minutes each.
Deflate the vest after each setting and cough 3 times.

The St. Alexius Heart and Lung Cystic Fibrosis Center will usually follow the settings recommended by the Vest company but will support individuals who utilize the Minnesota Vest Therapy settings. Please inform your provider of any changes you make in your settings and of the results in your airway clearance.

Recipe Corner

Grape Slush
2 Grape Juice Bars
2 Tablespoons Corn Syrup
½ Cup Grape Juice or 7 Up
1 Tablespoon Corn Oil
Blend in Blender
493 Calories per serving

Fortified Milk
1 Quart whole milk
1 Cup instant non-fat dry milk
Pour liquid milk into deep bowl. Add dry milk and beat slowly with mixer until dry milk is dissolved. Usually less than 5 minutes. Refrigerate. The flavor improves after several hours.
1 Cup contains: 209 Calories, 14 Grams Protein, 8 Grams Fat, 461 mg Calcium

Great Strides Walk

The Great Strides: Taking Steps to Cure Cystic Fibrosis walk that had previously been planned for June 18th, has been postponed until September 10, 2011 due to flooding issues. This is the largest national fundraiser event held by the Cystic Fibrosis Foundation. The money raised is used to support vital Cystic Fibrosis programs. We are fortunate to have growing numbers of walkers each year and are looking forward to raising the bar for more walkers. Please come out and join us for a fun filled morning.

The Drug Development Pipeline

The Cystic Fibrosis Foundation continues to show its progress in medication development by tracking it on the Drug Development Pipeline. This pipeline is available to view on the CFF website www.cff.org. The progress of a medication is watched as it travels through preclinical trials and 3 different phases of clinical trials. Clinical trials allow actual patients to become involved in the research. Medications that successfully pass the third phase of clinical trials are then submitted to the Federal Drug Administration (FDA) for approval.

We have been watching a genetic modulator called VX-770 which has recently completed phase 3 trials. Here is a statement put out by the Cystic Fibrosis Foundation:

“VX-770: The program is sponsored by Vertex Pharmaceuticals, Inc. and partially funded by CFFT. VX-770 is a new compound, called a “potentiator,” designed to act upon a non-functional but properly localized CFTR protein to help to open the chloride channel in CF cells. Phase 3 trials have been completed in pediatric and adult people with CF who have one copy of the G551D mutation in their CF gene. There were no safety issues in these trials and the treatment groups met the primary endpoint for improved lung function and secondary endpoints of reduced pulmonary exacerbations, increase in weight, and increase in quality of life measures. Extended open label trials are ongoing. A New Drug Application is planned to be submitted to the FDA in 2011.”

We invite you to check out the progress of many other medications on the Drug Development Pipeline at www.cff.org.

Important Dates

Upcoming Clinic Dates: August 16th, September 20th and October 18th.
Luncheon Dates: October – Topic to be announced.

Your CF Team Members:
Dr. Carla Zacher  Dr. James Hughes
Sarah Mork, RN  Deb Fueller FNPc
Lynn Feist, LRD  Kaitlyn Towery, LSW
Stephanie Friedt RRT.RPFT
Deb McPherson PharmD