Cystic Fibrosis

What is Cystic Fibrosis (CF)?

– CF is a genetic disease passed from parents to child; two defective genes are needed (one from each parent) in order for the disease to be present.

– People with CF have thick, sticky mucus which clogs some organs in the body, mainly lungs, pancreas and liver.

– Mucus build up in the lungs allows bacteria to get stuck in the airways, causing infections and inflammation, which can lead to lung damage over time.

– Mucus build up in the pancreas and liver prevents the intestine from absorbing certain nutrients from food.

How is Cystic Fibrosis Diagnosed?

– Oregon and Washington test for CF on newborn screen.

– A sweat test detects high chloride on the skin which determines the diagnosis for CF.

– A blood test identifies which two genes are defective on the CF gene.

How is Cystic Fibrosis Treated?

– Because there is no cure for CF at this time, goals of treatment are to reduce symptoms and slow the progression of your disease.

Treatments to help the lungs include:

– Medicines to thin the mucus in the lungs (Pulmozyme™, sodium chloride 7%).

– Inhaled antibiotics fight bacteria that cause infections (TOBI™, Cayston™).

– Chest physiotherapy (CPT) – therapy to break up and loosen mucus in lungs.

What are signs and symptoms of Cystic Fibrosis?

- Salty tasting skin
- Not growing or gaining weight normally, difficulty maintaining weight
- Lung infections
- Persistent coughing sometimes with mucus
- Greasy, foul smelling bowel movements that may float in the toilet bowl
- Belly pain, diarrhea or constipation
- Nasal polyps – small growths in the nose

Treatments to help the **digestive tract** include:

- Enzymes (Creon®, Zenpep®) taken at every meal and with snacks to help break down food in the intestines.
- Specific vitamins that are not absorbed.

New Treatments

- Ask your doctor if you are eligible for new medications.
- For information about new treatments, visit the CF Foundation website: **www.cff.org/treatments/pipeline** **[this hyperlink doesn't work]**

Points to remember

- Make sure your vaccinations are up to date.
- Maintain a smoke-free environment, including your home and car.
- Schedule regular appointments with your CF care provider.

For additional information

- Cystic Fibrosis Foundation: 1-800-FIGHT-CF, **www.cff.org**
- Boomer Esiason Foundation: 1-516-746-0077, **www.esiason.org**

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