



The Childhood Liver Disease Research and Education Network strives to provide information and support to individuals and families affected by liver disease through its many research programs.

## **CYSTIC FIBROSIS LIVER DISEASE**

### **What is Cystic Fibrosis Liver Disease?**

Cystic Fibrosis (CF) is a condition affecting the lungs, pancreas, liver and other organs that is caused by changes (mutations) in a gene called “*CFTR*” (Cystic Fibrosis conductance Transmembrane Regulator). The primary problems in most patients with CF are decreased functioning of the pancreas and chronic lung disease.. Mild liver involvement with abnormal liver blood tests or fat in the liver is common. In some patients, the liver problems caused by CF are much more important than the lung disease. Advanced liver disease in individuals with CF is called CF liver disease (CFLD). Severe scarring of the liver, called cirrhosis, can occur in CFLD.

### **What are the symptoms of the Liver Disease in Cystic Fibrosis?**

Many CF patients with liver involvement do not have any symptoms. In patients with advanced CFLD, patients may have a large liver and spleen. Problems with weight gain can occur. As the scarring in the liver becomes more severe, the pressure in the vein leading to the liver (the portal vein) rises and complications of portal hypertension can occur. These can include a large spleen, fluid accumulation in the abdomen (ascites) and bleeding from dilated veins (varices) in the esophagus or stomach. Rarely jaundice and bleeding problems can occur.

### **What is the Lung Disease in Cystic Fibrosis?**

In cystic fibrosis, the salt balance in the body is disturbed. Because there is too little salt and water on the outside of the cells, the thin layer of mucus that helps keep the lungs free of germs becomes very thick and difficult to move. And because it is so hard to cough out, this mucus clogs the airways and lead to infections that damage lungs.

### **What Problems Can Cystic Fibrosis Cause in Other Organs?**

**Pancreas:** In about 90% of patients with CF the tubes that drain the pancreas are blocked and the release of the natural enzymes that aid in the digestion of fat and protein is prevented; this is called "exocrine pancreatic insufficiency". Affected patients need to take pancreatic enzymes with food to help with digestion. A few patients can develop inflammation of the pancreas (pancreatitis).



**Diabetes:** CF related Diabetes can occur in individuals with CF after their teenage years.

**Meconium ileus:** About 10% of patients with CF are born with thick secretions blocking the lower part of the small intestine called meconium ileus. Later in life, thick secretions can cause a bowel blockage called DIOS (distal ileal obstruction syndrome) or "Meconium Ileus Equivalent".

Constipation is common in CF.

### **How do you get Cystic Fibrosis Liver Disease?**

It is not known why some people with CF develop CFLD and why others do not. It is believed that the role of the CFTR protein is to add liquid to bile and that in CF the bile becomes thick and leads to plugging of the bile ducts resulting in inflammation and scarring.

### **How is Cystic Fibrosis Liver Disease found and diagnosed?**

CF liver disease is found during physical exam by a doctor and by blood tests. Sometimes ultrasound, CT scans, liver biopsy and other tests may add information.

### **What is the Treatment for Cystic Fibrosis Liver Disease?**

There is no known cure for CFLD, but there are ways to prevent or reduce health problems related the liver disease of CF. Intestinal bleeding can occur in severe CFLD, and medicines and procedures can be used to treat this. Patients with CFLD should be immunized against hepatitis A and B to prevent further injury to the liver. They should avoid alcohol use.

### **What is the Outlook for someone with CF Liver Disease?**

Most patients with CFLD can lead long and productive lives. However, those with severe lung problems or progressive liver disease may have a shortened life span.

### **Where Can I Get Other Information About Cystic Fibrosis Liver Disease?**

The **Cystic Fibrosis Foundation** is dedicated to education and support for patients and families with Cystic Fibrosis. The Cystic Fibrosis Foundation supports research and education in CF.

There is a detailed website with information: <http://cff.org>

The Cystic Fibrosis Foundation can also be contacted by mail, phone and email.



THE CHILDHOOD LIVER DISEASE RESEARCH AND EDUCATION NETWORK

Atlanta • Baltimore • Chicago • Cincinnati • Denver • Houston • Indianapolis • Los Angeles • New York • Philadelphia • Pittsburgh • St. Louis • San Francisco • Seattle • Toronto

Cystic Fibrosis Foundation

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Other information:

<http://www.nlm.nih.gov/medlineplus/cysticfibrosis.html>