

Cystic Fibrosis Research News

Title:

**Prevalence of Elevated Liver Enzymes in Children with Cystic Fibrosis
Diagnosed by Newborn Screen**

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What was your research question?

How frequently are routine annual blood tests used to monitor the liver (ALT, AST and GGTP) abnormal in children and young adults who were diagnosed with cystic fibrosis (CF) by newborn screening.

Why is this important?

The liver can be affected in CF and about 5-10% of people with CF can develop advanced liver scarring called cirrhosis. This seems to occur by the age of 15 years. Liver blood tests are recommended annually. How frequently and to what degree these tests are abnormal in stable children and young adults with CF can help care providers decide how they should respond to abnormalities. We also wanted to know if abnormal liver blood tests might help early identifications of patients who may be at risk of developing cirrhosis. Since Colorado was one of the first states to institute newborn screening for CF we had over 20 years of data to inform this study.

What did you do?

We compiled up to 20 years of annual liver blood test results of children and young adults who had been diagnosed with CF through newborn screen or the presence of meconium ileus, who are followed by the Colorado CF Center.

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We determined the frequency of any abnormal value for AST, ALT or GGT as well as values that were 1.5, 2 or 3 times the upper limit of normal for the individual test. We investigated demographic and clinical factors that may be associated with abnormal liver blood test results. We then determined if early abnormalities in AST, ALT or GGT (before 5 years of age) was associated with a subsequent diagnosis of advanced liver disease.

What did you find?

Most patients with CF had at least one abnormal AST, ALT or GGT in the first 20 years of life. By 20 years of age about 60% had an abnormal AST, 95% an abnormal ALT and 50% an abnormal GGT. No one had a value greater than 3 times the upper limit of normal. The incidence of AST, ALT or GGT greater than 2 times the upper limit of normal was about 85%, 70% and 85% consecutively, by 20 years of age. Persistently abnormal liver blood tests were less common for ALT and GGT. Females were more likely to have persistently abnormal AST or ALT and individuals of Hispanic ethnicity were more likely to have persistently abnormal GGT. Patients with an elevated GGT before 5 years of age had a higher risk of cirrhosis.

What does this mean and reasons for caution?

Liver blood tests are an important tracking tool for annual CF visits. Early identification of patients who are at risk for developing cirrhosis may be aided by these liver blood tests. However, we only looked at blood tests at annual visits. Liver blood tests can be high due to many causes in addition to CF, such as infections, medications, as well as under or over nutrition. Thus liver blood tests are just one tool that is used to determine the overall health of the liver in individuals with CF.

What's next?

Future studies will look into other tests of the liver (specialized blood tests, imaging and genetic tests) to determine if there are more comprehensive tools, identifying individuals who have advanced liver disease and those at risk for advanced liver disease to help us study potential new therapies.

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