Cystic Fibrosis Research News

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What was your research question?

Is there a relationship between abnormal baseline research ultrasound findings and demographic and clinical features?

Why is this important?

Currently there are no accepted markers that can identify children with CF who may be at risk for advanced liver disease (with or without portal hypertension) later in life. By identifying particular factors that are associated with an abnormal pattern on consensus research ultrasound, we can potentially identify specific early risk factors for an abnormal liver ultrasound.

What did you do?

We performed a research ultrasound of the liver on 719 children (ages 3 to 12) with CF with pancreatic insufficiency and without known cirrhosis. The pattern of the ultrasound was determined by the consensus of 3 radiologists and defined as either normal or abnormal. We then compared many clinical and historic variables from the CF Foundation Patient Registry between those with normal ultrasound patterns and those with abnormal patterns to see if we could determine any relationships.

What did you find?

We found that 82% of children had a normal pattern. However, 18% had an abnormal pattern and were older compared to those with a normal pattern. Early infection (<2 years) with Pseudomonas was associated with a normal ultrasound. CF related diabetes and ursodeoxycholic acid use was associated with a higher risk of an abnormal ultrasound.

What does this mean and reasons for caution?

Our findings demonstrate that the rate of abnormal ultrasound patterns is higher than previously reported and there is an association between CF-related diabetes and glucose intolerance and US identified liver disease later in life. It is interesting that early Pseudomonas infection appeared to be protective for US identified liver disease later in childhood. We still need to be cautious as the

ultrasound methods used are descriptive and may not directly correlate to the severity of liver disease in CF.

What's next?

This is an ongoing study to see if outpatient ultrasound can identify risk for advanced liver disease. In addition, we are looking at early infection, antibiotics, blood tests and other biomarkers in determining risk. The study now includes transient elastography and magnetic resonance elastography.