

Cystic Fibrosis Research News

Title:

Supersonic Shear-Wave Elastography and APRI for the detection and staging of liver disease in paediatric cystic fibrosis

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What was your research question?

In assessing children with cystic fibrosis (CF), can the development of liver disease and progression to liver damage (scarring, referred to as fibrosis and cirrhosis) be detected using a non-invasive, ultrasound-based technique called Supersonic-Shear Wave Elastography

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(SSWE). SSWE measures liver stiffness, at the time of visit (point-of-care) relaying the measurements to the CF clinic.

Why is this important?

Children with CF may have thick sludgy bile that obstructs bile flow in the liver, up to 10% will develop CF-associated liver disease (CFLD) of sufficient severity, impairing good health and longevity. Current methods to detect CFLD are non-specific and assessment of advancing liver damage is difficult, especially in the early stages where only a liver biopsy is reliable to assess scar tissue. There is a need for a reliable, accessible, non-invasive test to detect and monitor the development of CFLD.

What did you do?

Between 2015-2018, this study enrolled 95 children with CF (55 with CFLD according to standard blood and USS tests, 41 without liver disease) and 29 healthy controls. SSWE was performed to obtain a liver stiffness measurement and to establish liver stiffness cut-off values for the presence of CFLD and the detection of advanced CFLD (cirrhosis and/or portal hypertension). The study also utilised a simple biochemical index derived from routine blood tests called aspartate aminotransferase-to-platelet ratio index (APRI). The APRI was combined with liver stiffness measurements in an attempt to improve the diagnostic accuracy for both detection of CFLD and presence of advanced CFLD.

What did you find?

SSWE was successful in all children. Liver stiffness was significantly higher in children with CFLD compared to children with CF without liver disease and compared to healthy controls. A cut-off value of 6.85kPa accurately discriminated the presence of liver disease in children with CF. A cut-off value of 9.05kPa accurately discriminated the presence of advanced CFLD (cirrhosis and/or portal hypertension) from children without advanced CFLD. When liver stiffness was combined with APRI the accuracy for detecting CFLD improved but not the discrimination of advanced versus mild CFLD.

What does this mean and reasons for caution?

SSWE used alone or in combination with APRI provides a reliable, non-invasive method to diagnose and assess the severity of liver disease in children with CF. SSWE can be performed adhoc or repeatedly, to identify and monitor liver disease development without the need for an invasive liver biopsy, which will improve patient experience and refine clinical care. SSWE-



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derived liver stiffness measurements for CFLD detection and disease progression should be interpreted by experienced clinicians in conjunction with other clinical findings.

What's next?

These observations require confirmation in larger CF patient groups to confirm liver stiffness cut-off values and better define their utility in the care of the individual patient. SSWE may become a useful test to study interventions directed at slowing the progression of CFLD in those where it is clinically significant.

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