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Noninvasive Follow-up of Liver Involvement of Cystic Fibrosis Patients

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Objectives: Cystic fibrosis is the most common inherited disease in white population, with an incidence of 1 in 3000 newborns. Progress in our understanding of the disease and the impact of this on management has been rapid over the past 20 years. Hepatobiliary complications commonly occur in cystic fibrosis with increasing prevalence due to longer life expectancies and widespread screening efforts. Shear-wave elastography (SWE) is a novel noninvasive method that involves application of local mechanical compression on soft tissue using focused ultrasonography and acquiring strain images that show tissue response. We aimed to compare abdominal ultrasonography and SWE and also clinical and laboratory findings of children with cystic fibrosis prospectively. The measurements were compared with clinical data, biochemistry parameters and ultrasound findings.

Methods: A total of 14 consecutive patients with documented CF were prospectively studied. In 13 case, elastography measurement was performed at the same time with abdominal ultrasonography evaluation of the liver. One patient had clinical and ultrasonographic evaluation, but not elastography.

Results: Measurements were performed in 14 CF children (3 boys, 11 girls). The mean age of the patients was 5.2 (0.7-16). The mean length of the patients was 105.1 cm (66-143) and the mean weight was 18.3 kg (6.4-35.5). Abdominal ultrasonography revealed hepatosteatosis and hepatomegaly in 2 patients and only hepatomegaly in 1 patient. Liver function tests of patients were as follows; the mean AST level was 41 U/L (21-87) and the mean ALT level was 29.1 U/L (10-67). The INR values of only two patients were slightly higher. Eleven patients had pancreatic insufficiency and patients were receiving pancreatic enzyme replacement therapy. When the sputum cultures and colonization status of the patients were evaluated, 3 patients had Pseudomonas aeruginosa colonization and 1 patient had Burkholderia cepacia colonization and 1 patient had Pseudomonas aeruginosa and Burkholderia cepacia colonization. One patient has hepatomegaly and normal liver function tests and the patient's Kpa value was 6,36; two patients with elevated liver function tests and hepatosteatosis had Kpa values of 5.15 and 19.3; 1 patient with elevated liver function tests had a Kpa of 6.38. Abdominal ultrasonography and liver function tests of other patients were normal. The mean Kpa values of the patients were 8.68 (4.1-20.2).

Conclusion: Gastrointestinal complications should be closely monitored in cystic fibrosis. Although there is no definite ranges for children in SWE, it is important to monitor and evaluate with clinical and other laboratory and imaging methods.

Keywords: Cystic fibrosis, liver, elastography