Liver Dysfunction may be Associated with QOL in Adults with CF

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Background: More individuals with cystic fibrosis (CF) are living into their adulthood due to significant advances in medical care. The adult CF clinic at Indiana University cares for one of the largest adult populations with CF. Liver disease is common in children and adults with CF. However, the characteristics and consequences of CF liver disease (CFLD) in adults are not well understood. In this ongoing, IRB approved study, we systematically characterized the liver manifestations and their health related quality of life in adults with well characterized CF.
Methods: Individual patient demographics and clinical data were collected for study participants. Participants completed two quality of life study questionnaires: Chronic Liver Disease Questionnaire (CLDQ) and Revised Cystic Fibrosis Questionnaire (CFQ-R). 10mL of blood was collected for future research. Finally, participants underwent a bedside transient elastography via FibroScan®.
Results: There were 50 patients, with a mean age of 30.9 years, enrolled in the study. Pancreatic insufficiency was the most common co-morbidity, as it affected 90% of patients. The mean AST was 23.55 units/L, mean ALT was 25.20 units/L, and mean alkaline phosphatase level was 117.73 units/L. From the FibroScan®, the median liver stiffness measurement (LSM) was 4.65 kPa and the mean controlled attenuation parameter (CAP) was 219.52 dB/M. The mean CLDQ score was 5.343.
Conclusion: This ongoing study reveals that increased alkaline phosphatase and CAP scores are associated with a poor QOL. More data is needed to further understand the pathophysiology behind CFLD. Use of noninvasive imaging, noninvasive markers, and the CLDQ may aid in early identification of adult CFLD.