URSODEOXYCHOLIC ACID FOR CYSTIC FIBROSIS RELATED LIVER DISEASE, OUR EXPERIENCE

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Introduction: Abnormalities of LFT,s in CFALD have a low sensitivity and specificity and the presence of established cirrhosis will be diagnosed on imaging. Degree of benefit with UDCA is uncertain.

Aim: To Look at Mode of diagnosis for CFALD and Effectiveness & safety of UDCA in our unit.

Methods: Retrospective review on all patients currently receiving UDCA. Dose and duration of UDCA was recorded. Mode of diagnosis for CFALD was also noted. LFT and USS were analyzed with changes over time during treatment. Side-effects associated with UDCA were noted.

Results: 12 patients currently receiving UDCA were analyzed. M:F ratio was 7:5. Age ranged from 6.8 years to 18.7. Seven were homozygous for $\Delta F508$. USS and LFT,s in combination was most common mode of diagnosis in 67% cases, while USS or LFT,s alone was only used in 2 cases each. 85 LFT,s on 12 patients were analyzed. γ GT was raised in all 12/12 patients with improvement in 6/12(50%) during treatment. AST improved over time in 8/10 patients. ALT was normal in 3 cases while raised in 9 and improved in 4/9 cases over time during treatment. 46 USS results on 12 patients were analyzed. 33/46(77%) had inhomogenous texture in liver. Hepatomegaly was present in 8/12 and resolved in 4/8 during treatment. Splenomegaly was present in 10/12. No patient experienced side effects with UDCA.

Conclusions: USS in combination with LFT,s are most common mode of diagnosis for CFALD. In 50% patients improvement was noted in LFT,s over time during treatment with UDCA, it was safe and no significant intolerance or side effects were noted.