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Aetiology and outcome of acute liver failure in children in the United Arab Emirates

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Introduction: Pediatric Acute Liver Failure (ALF) is characterized by severely impaired liver function, with or without encephalopathy in children without previous liver disease. Geographic location affects aetiology, with Non-A-E hepatitis and drugs accounting for the majority of cases in the west.

Aim: We aim to review the aetiology, presentation and outcome of ALF in children in the UAE (Hepatic Encephalopathy).

Subjects & Methods: This is a retrospective, single centre study of children presenting with ALF from birth to 16 years over a 7-year period (September 2010-2017). We used the pediatric ALF Study Group criteria for defining ALF: (1) Absence of a previously known history of chronic liver disease, (2) biochemical evidence of acute liver injury, and (3) hepatic-based coagulopathy defined as PT≥15 s or INR≥1.5 not corrected by vitamin K in the presence of clinical HE or PT≥20 s or INR≥2 regardless of the presence or absence of clinical HE.

Results: 81 patients were identified (48 males and 33 females). Median age at presentation was 18 months (range 2 days-16 years). 12% presented in the first 4 weeks of life and 88% between 1-16 years of age. The aetiology was identified in 86% and included; 49% infection, 16% metabolic (The main cause of metabolic disease was *Wolcott Rallison syndrome*, seen in 46%), 15% acute circulatory failure, 14% indeterminate, 4% toxic and drugs, 1% infiltrative disease and 1% autoimmune hepatitis. Jaundice was seen in 42% at presentation (Median bilirubin 43 (range 2.2-600)) and didn't favor any aetiology. Encephalopathy was more significantly seen in the metabolic disease (77%, P=0.013). Renal failure was seen more significantly in acute circulatory failure (83%, P=0.008). INR was highest in toxic group (Median 6.5 (range 2.7-7.2)) and infiltrative disease (INR>10). Highest AST was in Indeterminate (Median 1059 (range 116-1435)) and Metabolic groups (Median 947 (range 38-9515)). Ammonia levels were highest in urea cycle defect (Median 455 (range 264-646)). Overall survival was 57% with improved survival in patients presenting after 1 month of age (68%). Only three patients were transplanted and they all survived. Metabolic, toxic and autoimmune disease had most favorable outcome with 60%, 100% and 100% survival, respectively.

Conclusion: ALF in the UAE has unique aetiology; with increased number of infections and reduced incidence of autoimmune hepatitis. The main metabolic disorder contributing to liver failure was *Wolcott Rallison syndrome* (a syndrome commonly seen in the Arab world and seen in children of consanguineous marriage). The presenting features and biochemical tests alluded to different aetiologies and can help target investigations. The mortality rate was high in our group; we feel this can be secondary to the different aetiology spectrum in our group in addition to lack of availability of liver transplantation in the UAE, and the need for children with ALF to travel for transplantation.

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