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EARLY PREDICTORS OF UNFAVORABLE OUTCOME IN NORTH AMERICAN INDIAN CHILDHOOD CIRRHOSIS

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Background: North American Indian Childhood Cirrhosis (NAIC) has only been described in the Cree-Ojibway First Nations of Northern Quebec. NAIC presents with transient neonatal jaundice and progresses to biliary cirrhosis often requiring liver transplantation (LT) in childhood. Only 30 patients have been described to date and risk factors associated with an earlier progression to LT have not yet been identified. Moreover, NAIC patients seem to experience more fractures than other cholestatic patients, but this has not been confirmed.
Aims: With this study, we aimed to identify predictors at 6 months from presentation that might suggest progression to end-stage liver disease as well as evaluate bone health in affected patients.
Methods: The records of all NAIC patients diagnosed between 2000-2020 were reviewed. Subjects were split into 2 groups based on whether they had undergone LT or not (No_LT) before age 18. Recorded complications included: hepatic encephalopathy (HE), variceal bleeding (VB), ascites, spontaneous bacterial peritonitis (SBP), bacteremia, and pulmonary shunts. Laboratory data (alanine aminotransferase, ALT; total bilirubin, TB) were collected at presentation and follow-up. Bone mineral density (BMD) of the lumbar spine (Z-scores) and number of fractures were compared between groups. NAIC patients were then compared to cohorts with other chronic cholestatic diseases such as biliary atresia (BA, n=24) and Alagille syndrome (AS, n=11).
Results: A total of 14 patients (M=9, F=5) were diagnosed with NAIC. Average age at presentation was 2.1 months (IQR 1-16.9 months), with 3 patients older than 18 months. Overall, 6 patients were transplanted (avg 8.6±1.7 years), one was listed for LT but died waiting, and 7 remained in a state of compensated cirrhosis. All complications were only observed in the LT group except for VB which also occurred in 2 patients of the No_LT group. Between presentation and 6 months, ALT and TB levels increased more in the LT vs No_LT group (p=ns). There was a greater variation of ALT/TB levels in the LT group (p=0.0047) even once the 3 patients with late referral were excluded (p=0.0381). No patient in the No_LT group had fractures, while 3 did in the LT group. BMD was lower in the LT group vs No_LT group (-

2.2±1.2 vs. -1.1±1.3, p=ns). NAIC patients had lower BMD (-1.7±1.3) than those with AS (0.7±0.9, p=0.003) or BA (-0.9±1.4, p=ns) and had a higher prevalence of fractures (21.4% vs. 12.5% for BA and 18.2% for AS patients).
Conclusions: In patients with NAIC, variation of ALT/TB levels at 6 months from presentation may be used as an early predictor of unfavorable outcome and progression towards end-stage liver disease. Patients who evolved to LT had more complications, higher prevalence of fractures and lower BMD values. Compared to children with BA or AS, NAIC patients had poorer bone health.

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