and compared to their histologic fibrosis stage. **Results:** We identified 137 pediatric patients (33% female; 45% Hispanic/Latinx, 11% Non-Hispanic Blacks, and 29% White) who fulfilled our criteria. NFS showed 0 patients meeting the upper cut-off, which would predict severe fibrosis. The lower cut-off predicted mild fibrosis with a sensitivity 98%, specificity 0%, positive predictive value 88%, and negative predictive value 0%; there were 0 true negatives. PNFS, with a cut-off of 26, predicted mild fibrosis with a sensitivity 74% and specificity 28%. Using a PNFS cut-off of 8, it showed sensitivity 52% and specificity 67%. **Conclusion:** The NFS and PNFS do not perform well enough to negate liver biopsy in pediatric patients. Despite the high sensitivity of NFS lower cut-off, the negative predictive value suggests a result above this cut-off lacks utility. The PNFS performed better than the NFS but not well enough to be used as an isolated screening tool. We continue to need non-invasive methods to reliably predict fibrosis in pediatric populations.

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1461

LONG TERM TUBE FEEDING IN YOUNG CHILDREN WITH ALAGILLE SYNDROME: DOES IT IMPACT HEIGHT?

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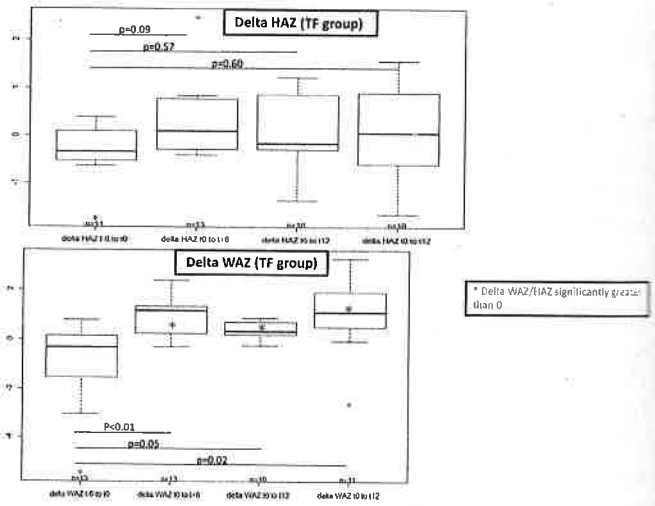
Background: The effect of nutritional intervention on growth in children with Alagille syndrome (ALGS) has not been studied. We aimed to describe nutritional management of ALGS children (≤5 years) in a large pediatric liver center and examine differences in growth before and after the initiation of tube feeding (TF). **Methods:** Retrospective study of children with clinically and/or genetically defined ALGS who received long term (≥1 year) TF. ALGS children without nutrition intervention (CG) were matched for age at time of TF initiation, sex and gestational age (GA). Z-scores of weight (WAZ) and height (HAZ) were grouped at: initiation of TF/control date (T_0); 1, 2, 3 and 6 months prior to TF (T_{-4}-T_{-1}); 1, 2, 3, 6 and 12 months (T_1-T_5) post TF. Statistical analyses were performed using R. **Results:** Nutritional interventions were initiated in 29/80 (36.2%) ALGS children: 23 TF, 2 TPN and 4 a combination of both. Of the 27 children that received TF, 13 (61.5% male) had sufficient data and received TF for ≥1 year; they were matched with 10 ALGS controls. Median (Q1;Q3) birth WAZ and GA were -1.2 (-1.6;-0.7) and 38 (37;39) weeks (TF vs CG: p-values ≥0.11). TF was initiated at a median age of 0.8 (0.6;0.9) years. Conjugated bilirubin, ALT, AST, GGT, and platelets at T_0 for TFG were not significantly different from the CG (all p-values ≥0.16). 3 children from each group were transplanted at a median age of 3.1 (2.8;4.3) years. Extrahepatic involvement including cardiac anomalies was comparable for both groups. Mean (SD) WAZ at T_0 were -3.8 (1.6) and -1.1 (1.5) for TFG and CG (p<0.001); HAZ at T_0 was significantly lower in TF than CG (-3.4 (-5.1;-2.7) vs -1.5 (-2.2;-1.0), p=0.01). Figure 1 shows that median delta WAZ, but not delta HAZ was significantly greater than 0 after initiation of TF. A generalized estimating equation including prematurity, age at initiation and group showed a significantly higher WAZ growth rate in the first six months and no difference for HAZ growth rate. **Conclusion:** One third of this ALGS cohort received tube feeding and/or TPN; no laboratory or phenotypic differences were identified between ALGS children who did or did not receive TF. Long term (≥1 year) TF leads to a sustained increase in WAZ, but no changes were seen in HAZ in the first year after TF initiation. This implies that weight is modifiable in ALGS in early childhood but that height may be determined by intrinsic genetic factors and/or cholestasis, as all grew well below WHO references.



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1462

NATIVE LIVER SURVIVAL IN PATIENTS WITH FIC1 DEFICIENCY: IMPACT OF GENOTYPE, SERUM BILE ACID CONCENTRATIONS AND SURGICAL BILIARY DIVERSION.

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Background: Mutations in *ATP8B1* can lead to persistent cholestatic disease, also known as familial intrahepatic cholestasis protein type 1 (FIC1) deficiency or PFIC1. Apart from cholestasis and end-stage liver disease, patients may develop diarrhea, hearing loss, pancreatitis and allograft steatohepatitis. We analyzed factors associated with its prognosis, including the presence of predicted protein truncating mutations (PPTMs), serum bile acid concentrations (sBA) and surgical biliary diversion (SBD), using the largest genetically defined cohort of FIC1 disease to date. **Methods:** Our multicenter, retrospective cohort study included 130 patients with compound heterozygous or homozygous predicted pathological mutations in *ATP8B1*. Patients were categorized according to the number of PPTMs (defined as splice site, frameshift due to deletion or insertion, nonsense, duplication); FIC1-A (n=63, no PPTM), FIC1-B (n=30, one PPTM) or FIC1-C (n=37, two PPTMs). Cut-offs for sBA were based on ROC analyses. **Results:** Overall, serum bile acids concentration (sBA) at first presentation (below age 1y) was negatively associated with NLS during follow-up (%NLS at 10y: sBA<194 µmol/L, 57% vs. sBA≥194 µmol/L, 31%; P=0.03, left figure). The proportion of patients with sBA at presentation <194µmol/L was 45%, 60% and 68% in FIC1-A, FIC1-B and FIC1-C patients, respectively (P=0.09). Survival analysis showed that at age 18y, 44% of all patients were alive with native liver. Native liver survival (NLS) was comparable between the three groups (%NLS at 10y: FIC1-A, 67%; FIC1-B, 43%; and FIC1-C 58%, P=0.15). A lower fraction of FIC1-C patients underwent SBD (%SBD at 10y: FIC1-A, 66%; FIC1-B, 60%; and FIC1-C, 43%; P=0.02). Overall, undergoing SBD was associated with a decrease in sBA (pre-SBD 230µmol/L, post-SBD 74µmol/L; P=0.005). SBD tended to be associated with prolonged NLS (HR 0.53, 95%CI 0.28-1.01, P=0.05), as with a post-SBD sBA concentration <65µmol/L (P=0.05, right figure). The proportion of patients achieving a post-SBD sBA concentration <65µmol/L was comparable among FIC1-A, FIC1-B and FIC1-B patients (50; 40; and 63%, resp.; P=0.63), as was NLS at 10 years post-SBD (69; 45, 66%; resp. P=0.31). **Conclusion:** Patients with one or two predicted protein-truncating *ATP8B1* mutations do not differ in natural history from patients with other *ATP8B1* mutations. Irrespective of the type of *ATP8B1* mutations, serum bile acid concentration at initial presentation and surgical biliary diversion are associated with native liver survival in patients with FIC1 deficiency.

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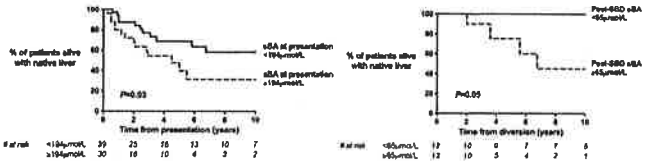

Figure. Proportion of patients with FIC1 deficiency that is alive with native liver, with a serum bile acid concentration at presentation <194μmol/L or ≥194μmol/L (left figure); and, with a post-surgical serum bile acid concentration <65μmol/L or ≥65μmol/L (right figure). Log-rank test.

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1463

NEUROPSYCHIATRIC COMORBIDITIES IN CHILDREN WITH NON-ALCOHOLIC FATTY LIVER DISEASE

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Background: There is a higher prevalence of depression in adults with non-alcoholic fatty liver disease (NAFLD) compared to adults without NAFLD. There are no reports on the prevalence of specific neuropsychiatric (NP) disorders in children with NAFLD. We hypothesized that among pediatric patients with NAFLD, there is a higher prevalence of NP disorders compared to a general pediatric population. **Methods:** This was a single-center cross-sectional retrospective chart review of children with NAFLD evaluated at Boston Children's Hospital (BCH) from 1995 - 2019. The prevalence rate of NP disorders was determined from patient, parent and clinician report at the first NAFLD evaluation visit. BCH NP prevalence rates were compared to those in the general pediatric population of the 2016 National Survey of Children's Health (NSCH) survey. Prevalence and relative risk (RR) was estimated from a general estimating equation with a log link, adjusted for age and sex and accounting for the complex survey sampling design to calculate the variance. Age is reported as median (interquartile range; IQR). **Results:** The BCH sample consisted of 370 children (30.5% female) with median age 13.0 (IQR10.8-15.8) years, while the NSCH included 50,212 children (48.9% female) with median age 8.1 (IQR 3.6-12.6) years. After adjustment for sex and age, 25.3% of BCH children with NAFLD were reported to have at least one NP disorder compared to 16.9% of NSCH children (RR=1.50; $P<0.0001$). There were statistically more children with depression (4.0% vs 2.2%; RR=1.85, $P<0.0001$), ASD (3.3% vs 2.0%; RR=1.66, $P=0.02$), and intellectual disability (4.1% vs 0.8%; RR=4.92, $P<0.0001$) in the BCH children. There were no significant differences in prevalence rates of developmental delay, ADD/ADHD, or anxiety between the groups. Restricting the data to 370 BCH and 6575 NSCH overweight and obese children (BMI ≥85th percentile), the BCH prevalence of at least one NP disorder was 38.4% compared to 28.2% for NSCH children (adjusted RR=1.36; $P<0.0001$), with the greatest disparity between populations due to intellectual disability (7.5% vs 2.2%; RR=3.39, $P<0.0001$). **Conclusion:** The prevalence of several neuropsychiatric disorders was higher among a cohort of children with NAFLD compared to that of a general pediatric population. Overweight and obese children with NAFLD were more likely to have at least one neuropsychiatric disorder compared to overweight and obese children without NAFLD. Further study to determine the significance of neuropsychiatric comorbidity in the prognosis and management of this population is warranted.

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1464

NORTH AMERICAN INDIAN CHILDHOOD CIRRHOSIS: IDENTIFYING FACTORS FOR DISEASE PROGRESSION

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Background: The Cree-Ojibway First Nations of Northern Quebec are the only group found to date to develop a disease historically named North American Indian Childhood Cirrhosis (NAIC). NAIC presents with neonatal jaundice and progresses to cirrhosis often requiring liver transplantation (LT) in childhood. Only 33 patients have been described so far with significant phenotype variability. Biochemical characteristics