primary studies - published, non RCT

Implications of carrier identification in newborn screening for cystic fibrosis.

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Study design (if review, criteria of inclusion for studies)

Prospective multicenter case-controlled cohort study.

Participants

Children with pancreatic insufficient cystic fibrosis (CF) aged 3-12 years without known cirrhosis underwent screening US.

Interventions

This study examines whether heterogeneous (HTG) pattern on liver ultrasound (US) identifies children at risk for advanced cystic fibrosis liver disease (aCFLD). Participants with HTG were matched (by age, Pseudomonas infection status and center) 1:2 with participants with normal (NL) US pattern. Clinical status and laboratory data were obtained annually and US bi-annually for 6 years.

Outcome measures

Primary endpoint was development of nodular (NOD) US pattern consistent with aCFLD.

Main results

722 participants underwent screening US, with 65 HTG and 592 NL. Final cohort included 55 HTG and 116 NL with â‰¥ 1 follow-up US. ALT, AST, GGTP, FIB-4, GPR and APRI were higher, and platelets were lower in HTG compared to NL. HTG had a 9.5-fold increased incidence (95% confidence interval [CI]:3.4, 26.7, p

Authors' conclusions

Research US finding of HTG identifies children with CF with a 30-50% risk for aCFLD. A score based on US pattern, age and GPR may refine the identification of individuals at high risk for aCFLD.

See also

Arch Dis Child Fetal Neonatal Ed. 2003 Nov;88(6):F467-71.

Keywords

ultrasound; diagnostic procedures; non pharmacological intervention - diagn;