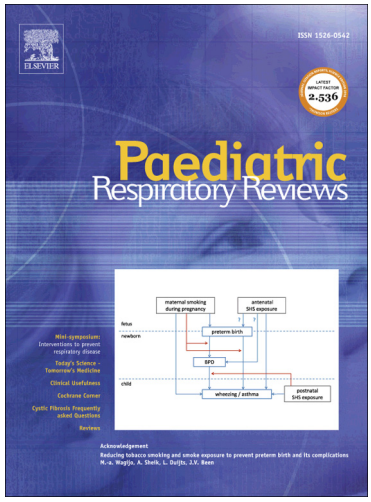

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Assessment of liver disease in cystic fibrosis

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Assessment of liver disease in cystic fibrosis

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Key words

Cystic fibrosis liver disease
Portal hypertension
Oesophageal varices
Liver transplantation

Abstract

Liver disease in cystic fibrosis has many causes, with biliary fibrosis due to abnormal CFTR protein predominating. Assessment requires aetiology to be defined. Biliary fibrosis may progress to cirrhosis and portal hypertension, which although initially asymptomatic, may cause varices and splenomegaly. Monitoring progression includes clinical and ultrasound assessment with endoscopic assessment of varices for those at risk. Extrapolated primarily from longitudinal assessment of viral hepatitis in adults, non-invasive elastography has a potential role. Evidence is lacking to support intervention strategies, but ursodeoxycholic acid and ligation of varices are widely applied. Indication and timing of liver transplantation are not clearly defined. Multidisciplinary approach is needed to tailor assessment and guide management.

Manuscript

Introduction

Liver disease may affect children and adults with cystic fibrosis (CF), although its significance, progression and impact on health may vary. Prevalence is reported to be between 10% and 30%, with onset of liver disease typically evident in the first decade, and prevalence of cirrhosis in children is suggested to be 5-10%. The Annual Report of UK Cystic Fibrosis Registry 2016 reports 1.3% prevalence of cirrhosis in those age < 16 and 3.7% age > 16. (1). However, epidemiological data are impacted on by the lack of consensus definition of liver disease, both relating to diagnostic criteria and also aetiology.

Aetiology