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Abstract Submission Form

TITLE	**Assessment of Fontan-associated liver disease in a single-centre Scottish cohort**
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ABSTRACT DETAILS:	
Background:	The Fontan operation is performed in individuals born with a single functional ventricle. The procedure separates venous return from the heart, and allows normal arterial oxygen saturations. It extends the life span of these patients, who are now surviving into adulthood. However, the Fontan circulation results in high hepatic venous pressure, chronic congestion and reduced portal blood flow. Fontan-associated liver disease (FALD) is an increasingly recognized complication, and can lead to cirrhosis, portal hypertension and hepatocellular carcinoma (HCC). Screening for liver disease is a critical part of long-term follow-up, although assessment of fibrosis in this cohort is not clear-cut.[1] Here we describe a model for out-patient hepatology review, and report baseline parameters of liver disease in new referrals.
Method:	All patients with a Fontan circulation referred to a hepatologist at the Royal Infirmary of Edinburgh from Dec 2017 to Apr 2019 were included. Demographic data, blood tests, imaging and echocardiogram results were recorded in a database at the time of clinic review.
Results:	18 patients were included. Mean age at review was 46, 50% were male. Average age at the time of the Fontan procedure was 9 years old. At clinic review, median duration of Fontan circulation was 19 years (range 8-45). Transient elastography (Fibroscan) results were available in 15 patients. Median liver stiffness was 14.9 KPa, range (7.9-34.3). Median MELDXI score was 7.1. Despite elevated liver stiffness results, ultrasound examination demonstrated normal liver appearance in 7/17 scans.
Conclusions:	Individuals with Fontan physiology are surviving longer, with universal development of hepatic fibrosis. Evaluation of the severity of liver disease is critical when those with a failing Fontan circulation are assessed for heart transplant, to determine whether concomitant liver transplant is indicated, and to exclude HCC. However, earlier identification of FALD allows appropriate screening for complications such as HCC, and timely discussions about long-term management including transplantation decision-making. We recommend assessment for FALD 10 years post-Fontan procedure, including clinical assessment, abdominal ultrasound, laboratory investigations, calculation of fibrosis scores and elastography. Following these individuals over time, and development of a UK registry, would help to improve our understanding of how best to assess fibrosis and predict severity of FALD.
References:	1. Gordon-Walker TT, Bove K, Veldtman G. Fontan-associated liver disease: a review. J Cardiol 2019; 74(3): 223-232