Most patients had normal biochemical liver profiles, with mean values for bilirubin 18.2 ± 10.6 µmol/L, ALT 26.5 ± 10.6 IU/L, albumin 47.5 ± 5.3 g/L and platelets 169 ± 55 x10^9, despite hepatic imaging of parenchymal abnormalities. Two patients were found to have hepatocellular carcinomas (HCC), BCLC stage B and C at diagnosis. There was a total of 4 deaths over a median follow-up of 2.8 years, of which 2 deaths were liver-related.

Conclusions Elevated hepatic stiffness and ultrasound appearance in keeping with severe fibrosis are common in adult Fontan patients, despite normal liver enzymes and synthetic function. FNH is the most common abnormality on imaging in high risk patients. The incidence of HCC justifies surveillance in this group of patients, however the optimal surveillance protocol remains to be established.

Results 19 patients were identified (12 male: 7 female); mean age at death was 61. 11/19 patients had alcoholic liver disease (ALD), 5 had ALD/Hepatitis C. 12 patients died on the ward, 3 in ICU, 2 at home and 2 at a nursing home. Child-Pugh Scores (CPS) ranged from B-6 to C-13 and average MELD scores was 23 (range 8–38). None were eligible for transplantation: 12 due to active alcohol use, 3 due to co-morbidities, 1 malignancy, 2 unknown. Median number of admissions in the year preceding death was 2 (range 0–5). Predominant symptoms prior to death were respiratory distress, confusion and pain. Average interval between admission and death was 24 days. 13/19 patients were referred for inpatient palliative care input. Although all patients had a DNACPR notice in place, the average DNACPR-to-death interval was just 20 days (range 0–139 days). EO decisions were made ‘early’ (DNACPR-to-death time >21 days) in patients (n=6) who had gradual disease evolution and/or a long period of contact with the service. In those with ‘short’ DNACPR-to-death times (n=13), 9 had been hospitalised 2 or more times in the year prior to death (this being a known marker of poor prognosis), and 2 had been admitted 4 or 5 times. 5/19 patients had a documented Amber Care Bundle referral (prognostic uncertainty tool). 14 patients had a documented ceiling of care discussion; of these 12 were for ward-based care only and 2 were for Level 3 escalation.

Conclusions Patients with end stage liver disease continue to be engaged in EO and treatment escalation discussions relatively late, despite clear indicators of poor prognosis (including recurrent admissions and non-transplantable status) within the previous year. Those well known to specialist teams who deteriorate gradually have a greater chance of expressing their preferences. Increased awareness of poor prognostic features is required in the secondary care setting.