

prednisolone was thought to be unacceptable for those patients: Four of the 22 patients (18%) had either experienced significant weight gain with prednisolone or were already overweight, and another four of 22 (18%) had worsening diabetic control or prednisolone-induced diabetes. Five of 22 (23%) had other unspecified side effects of prednisolone, and one patient had budesonide used due to failure of other induction medications. Twelve of 22 patients (55%) receiving budesonide achieved a normal alanine aminotransferase level within 12 weeks of commencement. The median wean time from budesonide was 7 months (range, 2–20 months). Four of 22 patients (18%) were unable to cease budesonide completely due to AIH flares on lower doses.

Conclusion: Budesonide has been shown to provide an alternative option as induction and maintenance therapy for patients with AIH who are intolerant of prednisolone. Further studies are needed to compare the role of budesonide versus prednisolone as first-line or second-line therapy in induction of remission in AIH and the long-term outcomes.

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Hepatic predominant presentation of Kawasaki disease in an adolescent girl: Case report

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Case Report: A previously well 16-year-old girl from north Queensland presented to a regional hospital with a 3-day history of high fevers (39.8°C) accompanied by myalgia, anorexia, and diarrhea, with a single bloody bowel motion on the day of admission. On a clinical examination, she was febrile and appeared generally unwell. An erythematous maculopapular rash extended over her stomach and arms. There were no other focal signs of infection. Admission blood test results showed a normal hemoglobin level of 136 g/L (reference interval [RI], 115–160 g/L) and normal white cell count of $6.1 \times 10^9/L$ (RI, $4-11 \times 10^9/L$) but significantly elevated inflammatory markers, with a C-reactive protein (CRP) level of 244 mg/L (RI, < 5 mg/L) and erythrocyte sedimentation rate (ESR) of 45 mm/h (RI, < 12 mm/h). Her liver function testing (LFT) results were the most remarkable, with an alanine aminotransferase (ALT) level of 239 U/L (RI, < 45 U/L), aspartate aminotransferase (AST) level of 163 U/L (RI, < 35 U/L), alkaline phosphatase (ALP) level of 174 U/L (RI, 30–110 U/L), gamma-glutamyltransferase (GGT) level of 171 U/L (RI, < 55 U/L), total bilirubin level of 80 µmol/L (RI, < 20 µmol/L), and conjugated bilirubin level of 58 µmol/L (RI, < 4 µmol/L). Regarding liver synthetic function, she had an elevated INR of 1.4 (RI, 0.9–1.2) and a low albumin level of 18 g/L (RI, 29–42 g/L). Hemolysis was excluded, with a haptoglobin level of 2.7 g/L (RI, 0.36–1.95 g/L) and normal blood film. Initial abdominal ultrasound showed a mildly hyperechoic hepatic texture with normal biliary tree. The patient commenced empiric treatment with intravenous cefotaxime 1 g 6-hourly and doxycycline 100 mg 12-hourly. Despite antibiotics, she continued to have recurrent febrile episodes. Extensive infectious disease screening was performed with blood cultures and serology for hepatitis A/B/C/E, cytomegalovirus, Epstein-Barr virus, Q fever, *Mycoplasma pneumoniae*, leptospirosis, brucellosis, streptolysin, Rickettsia, HIV, tuberculosis, Legionella, and dengue fever. Autoimmune studies, including antinuclear antibodies, extractable nuclear antigen, tissue transglutaminase, anti-LKM1, and ANCA, were negative. Over the next 5 days, the patient developed a diffuse swelling of her hands and feet, with an associated progression of her rash that resulted in desquamation of her digits. Clinically, her tongue took on a strawberry-red appearance, prominent cervical lymphadenopathy developed, her sclera became injected, and she became visibly jaundiced. A repeat ultrasound scan of her liver showed a potential narrowing of the hepatic artery without other hepatic or biliary changes. By day 3 of her presentation, serum bilirubin level reached its peak of 163 µmol/L (< 20), with LFT derangement largely improving throughout her admission. She was given a provisional diagnosis of Kawasaki disease (KD) and commenced treatment with intravenous immunoglobulin (IVIg)

2 g/kg over 2 days, with low-dose aspirin 150 mg per day, then transferred to a tertiary facility. At the tertiary center, IVIg was continued and cardiac investigations with computed tomography coronary angiography and transthoracic echocardiography were performed, with unremarkable results. The patient was subsequently discharged home. One week after discharge, the patient reported ongoing low-grade fevers (37.6°C) and progressively became unwell with malaise, lymphadenopathy, and a recurrence of conjunctival injection. She received a further 1 g/kg of IVIg over 2 days. Results of blood tests at this time showed an ESR of 80 mm/h (1–20), CRP of 29 mg/L (0–6), total bilirubin level of 25 µmol/L (2–20), normal liver function, and normal white cell count but elevated eosinophils ($1.53 \times 10^9 [0.04-0.40]$). On a follow up 1 month later, she had made a full recovery. Additionally, her liver function, blood counts, immunoglobulins, and CRP have returned to normal ranges.

Discussion: This case report documents the clinical experience of an adolescent patient presenting with symptoms consistent with the clinical criteria for KD. This patient's presentation is interesting, given the predominant hepatic involvement, compared with the more common cardiac involvement. A literature review found that adult and pediatric KD cases were not phenotypically alike, with older patients more likely to have elevated transaminase levels, though less likely to develop coronary artery involvement.¹ This is in keeping with the clinical presentation in this patient, and given a reduced propensity for cardiac involvement, it may imply a better prognosis. However, poor outcomes in older patients are in part suggested by diagnostic delay, with consequently delayed management resulting in coronary artery complications.² Hepatic involvement is a well-documented complication of KD, with 65% of patients with adult-onset KD developing hepatitis. Despite the association with treatment failure, patients are typically asymptomatic, with resolution of liver dysfunction linked to overall disease activity. The pathogenesis of LFT derangement in patients with KD is incompletely understood but is thought to be multifactorial. In a subset of cases, patients present with obstructive jaundice and hydrops of the gall bladder. This has been theorized to be due to compressive lymphadenopathy of the porta hepatis. In less common cases, patients with KD have been found to present with obstructive jaundice but without sonographic evidence of hydrops or mechanical obstruction. To our knowledge, in addition to this case, there have been five previous reported cases of obstructive jaundice without evidence of hydrops of the gall bladder. In these cases, patients were investigated with radiographically normal hepatobiliary anatomy. In all cases, LFT results returned to normal ranges after administration of IVIg.

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Cirrhosis Care (CirCare) study: Working towards better patient outcomes

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Background and Aim: Optimizing patient care for patients with advanced cirrhosis is challenging, as many are receiving multiple medications, have dietary restrictions, and may require repeated hospital admissions each

year. There is considerable interest in the use of patient-reported outcome measures, such as health-related quality of life (HRQoL), to inform patient care and to improve timely identification and control of symptoms, with the ultimate aim of enhancing physical and psychological well-being. The CirCare study, initiated in 2017, aimed to identify potentially modifiable patient and health service factors associated with patient outcomes (survival, hospital admissions, and patient-reported outcome measures) in a large cohort of patients with cirrhosis.

Methods: Consecutive patients attending hepatology/gastroenterology clinics or admitted to the Royal Brisbane and Women's, Prince Charles, Mater, Logan, or Princess Alexandra hospitals with a diagnosis of cirrhosis were eligible. The primary data include patient interview data collected face-to-face at recruitment from June 2017 to December 2018; clinical data from patients' medical records; and follow-up data from hospital medical records, emergency room presentations, cancer registry, death registration, and Medicare, via data linkage. Here, we present study enrolment data.

Results: A total of 1065 patients with cirrhosis were identified during the recruitment period. Of 746 invited, 581 patients were interviewed, with 165 refusals (78% response). Patients not invited included 112 who attended the clinic at a time when the research nurse was busy interviewing another patient and did not have time to approach the patient, 130 who did not attend the clinic, and 77 who were not eligible. Age and sex of patients who were missed or refused were not significantly different from patients recruited. Of the 581 recruited patients, 70% were male, 5% were Aboriginal and/or Torres Strait Islander, 44% had less than senior school education level, 23% were currently employed, 28% lived alone, 35% lived in the most disadvantaged areas (quintiles 4–5), 14% had English as not their first language, and 15% were inpatients at recruitment. Most patients (82%) reported moderate–high need for assistance with at least one out of the 50 items and reported unmet needs due to symptoms (70.4%), psychosocial issues (57.9%), or day-to-day activities (48.3%), and related to information about liver disease (45.5%). Our cohort had significantly lower HRQoL scores as measured by the generic Short Form-36 (SF-36). SF-36 scores were lower for all domains (except role emotional) compared with the healthy population (normative Australian data);¹ and inpatients had lower scores than outpatients for all domains ($P < 0.05$). The Chronic Liver Disease Questionnaire (disease-specific HRQoL instrument) scores for our cohort were comparable to those reported for patients with cirrhosis;² inpatients had lower (worse) scores ($P < 0.05$) than outpatients (Fig. 1).

Conclusion: Lower HRQoL is seen in this cohort of patients with cirrhosis, particularly among those admitted with decompensated cirrhosis. As clinical and follow-up data become available, we plan to assess the patients' support needs and their association with HRQoL, hospitalizations, and survival. We believe that the findings of this study will help inform the design of targeted strategies to improve outcomes for patients with cirrhosis.

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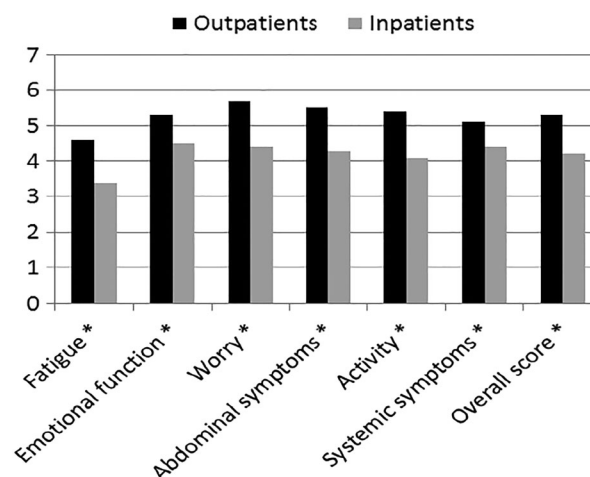


Figure 1 Chronic Liver Disease Questionnaire domain means.

* $P < 0.05$ for inpatient versus outpatient.

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Cirrhosis and compliance with European Association for the Study of the Liver hepatocellular carcinoma guidelines 2018: Profiling of a busy Brisbane tertiary public hospital

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Background and Aim: Hepatocellular carcinoma (HCC) is now the fifth most common cancer in the world, occurring most often in the population with cirrhosis. Recently published guidelines from the European Association for the Study of the Liver (EASL) for the management of HCC suggest recommendations for surveillance, leading to reduced lifetime health care spending and improved years of quality of life.¹ The new EASL guidelines were used to benchmark our public service against international recommendations. The secondary aims were to measure compliance with surveillance intervals for reported cirrhosis complications (varices surveillance and bone mineral density loss) and to examine incidence of hepatic encephalopathy, use of hepatic venous wedge pressure measurements, and use of transjugular intrahepatic portosystemic shunt (TIPS) procedures.

Methods: All patients with cirrhosis (compensated and decompensated) ($n = 204$) and patients with hepatitis B virus ($n = 9$) who met requirements for HCC surveillance criteria between January 2017 and December 2018 were audited for compliance with twice-yearly abdominal ultrasound scans, alpha-fetoprotein (AFP) measurement, surveillance endoscopy for varices, and dual-energy X-ray absorptiometry scans.

Results: Between January 2017 and December 2018, a total of 213 patients met the criteria for HCC surveillance. Of the 213 patients with cirrhosis, fatty liver disease was the most common etiology (80% compensated and 20% decompensated) (Table 1). Ten patients with HCC were identified (0.05%). Two patients died, and three patients received interventional radiological intervention (transarterial chemoembolization) without evidence of recurrence. Of the 112 of 213 patients who had complex hepatic encephalopathy testing (Stroop/pencil/flicker), 46% were found to have minimal hepatic encephalopathy. Compliance with screening recommendations is shown in Table 2.

Conclusion: Compliance with ultrasound scanning fared better than AFP