

Hepatic sarcoid: ‘Satisfactory outcomes in orthotopic liver transplantation for hepatic sarcoidosis: the UK experience ’

Dr Ricky Sinharay ¹, Dr William JH Griffiths¹

¹ Department of Hepatology, Cambridge University Hospitals NHS Foundation Trust, Hills Road, Cambridge, CB2 0QQ UK

Introduction

- Sarcoidosis is an autoinflammatory granulomatous disease most commonly presenting with lung or intrathoracic involvement.
- Hepatic sarcoidosis is increasingly recognized and seen in 12-20% of cases. However, it remains a rare indication for liver transplantation.
- Hepatic sarcoidosis presents with sterile, well circumscribed, non-caseating epithelioid granulomas, cholestatic liver enzyme derangement, small and large duct biliary disease, and hepatosplenomegaly. Mass effect from granulomas and the development of liver cirrhosis leads to portal hypertension.
- In the present study, we present unpublished data from the UK Transplant registry on outcomes in hepatic sarcoidosis.

Methods & Materials

Patients listed for liver transplantation with a primary diagnosis of hepatic sarcoidosis were identified from the UK Transplant Registry between 2008 and 2021 (NHS Blood and Transplant Data). Data from this cohort were examined including demographics, graft and patient outcomes.

Results

- In the UK, 31 patients have been listed for liver transplantation due to hepatic sarcoidosis in the last decade.
- 22 patients received a liver transplant, 17 of whom are still alive today (Table 1).
- Three patients died whilst awaiting transplant and another six patients were removed from the list.
- The mean age at registration was 51.59 years (+/- 9.16 years).
- The mean UKELD score at transplant was 56.14 (+/- 4.06).

Age in years (Mean ± SD)	51.59 ± 9.16
UKELD score at registration (Mean ± SD)	56.14 ± 4.06
Total listed for hepatic sarcoidosis (n)	31
Total transplanted (n)	22
Patients died on waiting list (n)	3
Cause of death (n)	
Gastrointestinal haemorrhage	1
Multisystem failure	1
Other	1
Graft functioning following transplantation n (%)	18 (81.82%)
Probability of graft function from transplant (%)	
1 year	85.45 %
3 years	85.45 %
5 years	76.91%
Cause of graft failure (n)	
Recurrent disease	1
Biliary complications	1
Primary non function	1
Other	
Patient alive following transplantation n(%)	17 (77.27%)
Median patient survival from transplant (years)	6.56
Probability of patient survival from transplant (%):	
1 year	90.23%
3 years	90.23%
5 years	70.31%
Cause of death (n)	
Infection	1
Haemorrhage	1
Donor organ failure	1
Other	2
Confirmed cases sepsis n (%)	13 (59.10%)
Source of sepsis (n)	
Blood	1
Chest	2
Abdominal	3
Other	7

Table 1: NHS Blood and Transplant data between 2008 and 2021 showing patient and graft outcomes for patients receiving a liver transplant for hepatic sarcoidosis

Mortality data among patients with hepatic sarcoidosis

- The median patient survival from transplant was 6.56 years with a 1- and 3-year patient survival of 90.23% and 71% (Figure 1).
- Graft failure occurred in 4 of the transplanted cases.
- One case of graft failure was a result of recurrent hepatic sarcoidosis, whilst another was a result of biliary tract stenosis.
- Sepsis occurred in 13 cases (59.10%), however it was noted as the cause of death in only one case (4.55%)

Figure 1

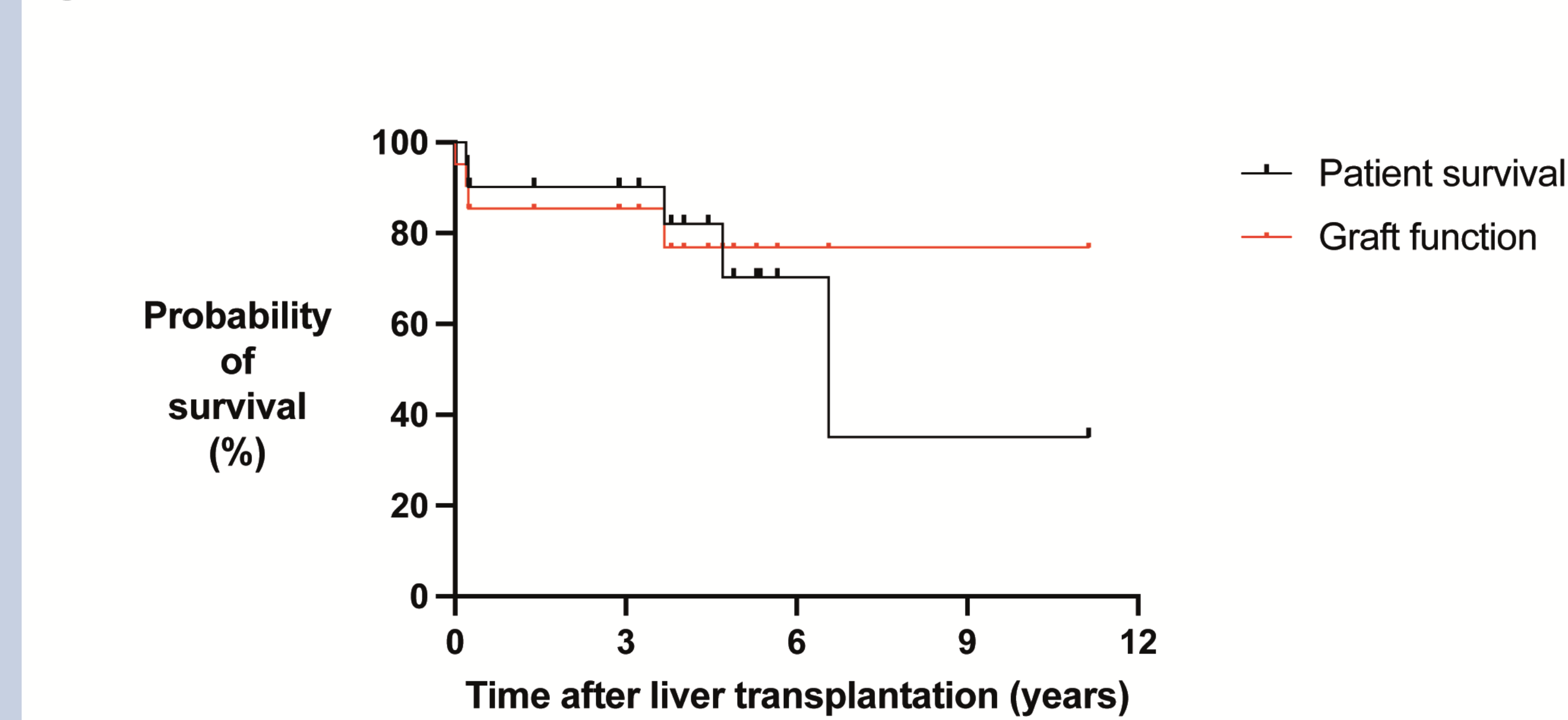


Figure 1:Kaplan-Meier Curve showing the probability of patient and graft survival following liver transplantation for hepatic sarcoidosis in the UK between January 2008 and April 2021 (n=22)

Conclusion

- Hepatic sarcoid is a rare indication for liver transplant.
- NHSBT data between 2008 and 2021 shows that patient survival for those receiving a liver transplant in the UK for hepatic sarcoidosis was satisfactory in the short to medium term.
- US data between 1987 and 2007 suggested 1- and 5- year patient survival of 78% and 61% respectively which was worse than for other cholestatic liver disease but notably in an older era than the UK cohort.
- Although not common, recurrent sarcoidosis in the donor liver occurs and may respond to increased immunosuppression.
- Limitations of the study include its small dataset, the retrospective study design and the reliance of registry data on centers submitting up to date information. The registry does not contain data on immunosuppression used, disease recurrence or details on the presence of extra-hepatic sarcoidosis.
- There is a marked paucity of data on the use of immunosuppression and hepatic sarcoidosis in the transplant setting, in order to demonstrate improvement in liver biochemistry, histology, graft and patient outcomes. A UK-wide collaboration across transplant centers would be a welcome step to improve data collection and inform transplant listing decisions.