



Review

Adult liver transplantation in the congenital absence of inferior vena cava



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HIGHLIGHTS

- Congenital absence of the infrahepatic inferior vena cava (IVC) amongst adult liver transplant recipients is rare.
- Radiological identification of vascular anomalies by routine pre-transplant imaging (CT/MRI) during transplant assessment work up is essential to plan appropriate strategies.
- Preservation of adequate length of native hepatic veins as they join the right atrium allows satisfactory outflow reconstruction with classic piggyback technique.

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ABSTRACT

Whereas congenital absence of inferior vena cava observed in paediatric population more often than not, as an isolated or syndromic variety, this is seldom encountered in adult liver transplant recipients. There appear few sporadic reports in the literature on experience of such anomaly in adults. Given the rarity of situation, surprising encounters of such anomalies may pose challenge to the unprepared transplant surgeon and unfavourable outcomes may even have resulted in under-reportage of this condition. In this brief report we document our recent experience with two such cases and this is supplemented with extensive reference to the literature on classification of such anomalies with the endeavour to document implications of such in the adult liver transplant setting.

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1. Introduction

Congenital absence of the entire infrahepatic inferior vena cava (IVC) is a rare type of venous malformation, which prevalence is about 0.07–8.7% in the general population [1]. These anomalies are frequently associated with cardiovascular malformations, biliary atresia, asplenia and polysplenia syndrome and variety of clinical presentations exist [2]. In paediatric population when it is usually associated with biliary atresia, the condition is complicated up to 7–13% by polysplenia syndrome. Many children with associated significant cardiovascular anomalies do not survive till adulthood. In those surviving till adulthood these anomalies more likely to be of isolated variety; hence they become incidental findings in the majority, meanwhile a proportion of them present with distal limb venous insufficiency or related to deep venous thrombosis [3]. As the cross sectional imaging is routine nowadays some other asymptomatic patients are diagnosed with this malformation once imaged for different reasons.

Liver transplantation (LT) in paediatric recipients in the absence of infra-hepatic IVC has been vastly reported; biliary atresia represents the most common indication for paediatric transplantation. Only a few cases of LT in adult with absence of IVC have been reported and all these reports originate from the recent past [4–6]. In those settings, radiological investigations before the surgery permit to recognise vascular anomalies as absence of IVC, which is extremely useful to plan the suitable surgical techniques for each recipient. Barring the LT alone, these anomalies have implications on widely practiced cadaveric organ donation as well, hence the transplant community would benefit from extensive literature review that accompanied these two cases of adult recipients with absent infra-hepatic IVC who underwent LT during the last 12 months in our institution.

2. Case reports

2.1. Case 1

A fast track liver graft was offered from a donation after cardiac death (female of 50years, donor warm ischemia time 25 min). At the time of offer 02:30hrs of cold ischaemic time (CIT) had been elapsed, however considering the offer it was deemed to be acceptable. With the logistical pressures of minimising the CIT, a suitable local recipient was mobilised whilst the liver graft was routed to the transplanting centre. The recipient was a 59 years-old man with diagnosis of chronic liver failure due to primary sclerosing cholangitis (PSC) with model for end-stage liver disease (MELD) score of 19. Prior to listing for LT, following imaging were performed: ultrasound scan (US), which visualized a chronic hepatic parenchyma distortion and patent hepatic and portal veins; magnetic resonance cholangiopancreatography (MRCP), which reported multiple dilatation and strictures of intrahepatic ducts and atrophic left kidney, but no reference to the absence of infrahepatic IVC. Whilst the patient was being inducted, the images were reviewed and the absence of IVC was detected at this stage. Discussion with the anaesthetic team was made at this point, specially the beneficial aspect of this anomaly with regards to the transplant operation without the need of performing a porto-caval shunt or side clamping of cava that would ensue reduced venous return during the implantation phase.

The image findings were confirmed at the laparotomy, and the three hepatic veins directly drained to the right heart (Fig. 1A). Standard dissection of hilum followed by clamping the hepatic veins before completing the explant by cutting in to the hepatic veins. The explant phase of the operation was thus lasted only 50 min, an added advantage in the setting of a liver graft from a donor after cardiac death (DCD) already in transit. After the hepatectomy the three hepatic vein orifices were conjoined to form one single outflow tract, which was anastomosed end-to-end fashion with the suprahepatic IVC of the graft with 4/0 prolene continuous (Fig. 1B and C). Rest of the anastomoses were fashioned as standard. The liver was well reperfused without bleeding from the outflow reconstruction. An end-to-end arterial anastomosis and hepatico-jejunostomy were performed. During the intraoperative period the patient remained hemodynamically stable. The total operative time was 4 h and with the implantation time of 34 min. Despite accepting an offer with a liver graft already in ice, the CIT elapsed was 394 min. After surgery the patient spent 3 days in intensive care unit uneventfully and was discharged after 11 days with an excellent recovery. At 1 year of follow-up, the recipient is alive and well with normal liver function tests.

2.2. Case 2

A 32 years old man referred for LT for hepatocellular carcinoma (HCC) recurrence, having previously undergone liver resection in

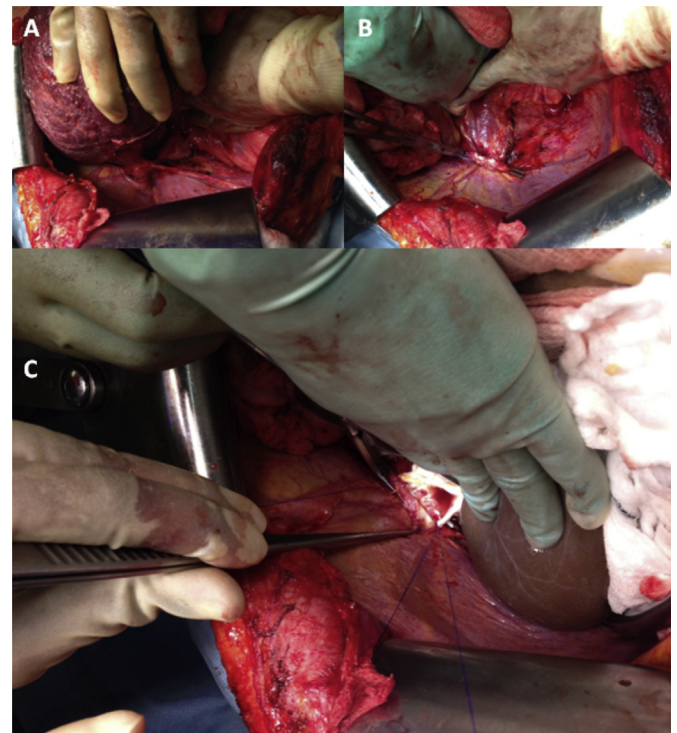


Fig. 1. Intraoperative imaging of absence of IVC. 1A) The native liver is lifted up to demonstrate hepatic veins directly draining to the heart; 1B) The conjoined hepatic veins over a vascular clamp after hepatectomy; 1C) Implantation of the liver graft with top end of the graft cava directly anastomosed to the three hepatic veins.

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