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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-jejunosotomy 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

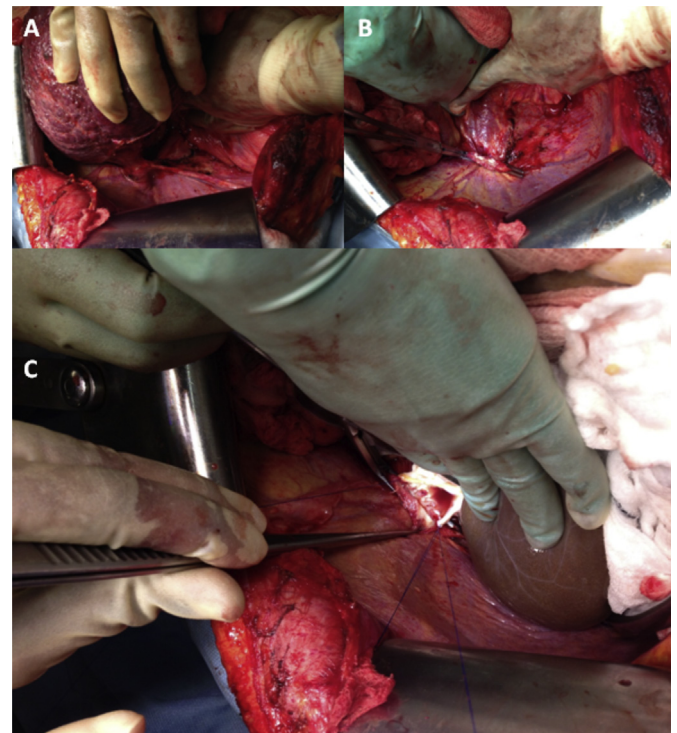


Fig. 1. Intraoperative imaging of absence of IVC. 1A) The native liver is lifted up to demonstrate hepatic veins directly draining to the heart; 1B) The conjoined hepatic veins over a vascular clamp after hepatectomy; 1C) Implantation of the liver graft with top end of the graft cava directly anastomosed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

February 2011, for a bi-segmental liver resection (S5–S8) for multiple hepatocellular carcinoma (HCC) related to chronic hepatitis B virus (HBV) infection. At the staging Computer tomography (CT) scan the absence of retrohepatic IVC was noted and documented. The IVC was replaced by a dilated azygos continuation of the IVC above the level of the renal veins up to the top of the liver (Fig. 2). During the surgery, the absence of retrohepatic IVC was confirmed and dilated retroperitoneal, paravertebral and perigastric varices were noted. The liver was cautiously posteriorly dissected from the diaphragm, due to the extended adhesions from the previous surgery, and the hepatic veins, draining directly into the atrium, were clamped before the liver was removed (time of hepatectomy 3 h and 12 min). A graft from a donor after brain death (DBD) was implanted by suturing the donor superior IVC into the recipient common orifice of the hepatic veins with continuous 4/0 prolene (CIT 728 min, WIT 30 min). In the presence of anomalous hepatic arterial inflow to the graft, aortic conduit was used with appropriate reconstruction of accessory vessels. The operation time was 6 h and 20 min. Patient had an uneventful post-operative course and was discharged after 9 days. The patient is alive and well at 4 months of follow-up.

3. Discussion

Perhaps the absence of IVC in adult LT recipient makes the hepatectomy phase more simple and straightforward, provided this anomaly is identified prior to the operation. Surprised identification may pose challenges to the unprepared transplant surgeon, and in some occasions there may be technical challenges of creating the hepatic venous outflow depending of the graft choice, for example live donor transplantation. Moreover, early identification may help graft preparation. More importantly, this obviates the need for more technically challenging hepatectomy, temporary porto-caval shunt and caval occlusion for the implantation phase. For transplant centres still performing venous by-pass techniques, the need for such complex procedure that is not without any complications may not even arise in the absence of IVC.

The embryological origin of IVC is complex: normally IVC is composed of four segments including hepatic, suprarenal, renal and infra-renal; it is formed between the 6th – 8th weeks of gestation, coinciding with the development of the liver, spleen, heart and lungs congenital anomalies of IVC are numerous and classified in 14 variations [7]. The most common vascular anomalies of IVC include the left or the double IVC, the entire/partial absence of IVC (or azygos continuation of the IVC), circumaortic or retro-aortic left renal vein and the absence of the infrarenal IVC [8]. Those congenital malformations are rare and often difficult to define with complex nomenclature (Table 1), but perhaps the anatomical

squeal of major anomalies remain the same, and characterized by an alternative passage venous return from the lower part of the body diverted to the chest (and eventually to the heart) via the azygos system of veins. In some cases the co-dominance of both right and left (hemi) azygos systems that is termed as double IVC, whereas in the left dominant azygos return may be termed as left side IVC with dilated left azygos system is seen to the left side of aorta.

The pathogenesis of the absence of IVC remains controversial. The entire absence of IVC might be the result of numerous abnormalities during the fusion and regression of the embryonal veins (such as vitelline, supracardinal, subcardinal, and posterior cardinal veins) or caused by a peri-natal thrombotic event, which causes degeneration and disappearance of the previously existent IVC [9]. Therefore, the venous outflow from tributaries of IVC develops a massive venous collateral circulation. The infrahepatic absence or interruption of IVC is caused by the absence of fusion of the right subcardinal vein to the liver and it is characterized by the presence of IVC from the common iliac vein up to the renal veins, from which point it continues posteriorly to the diaphragmatic crura as dilated azygos vein. Similar anomalies are not uncommon to encounter in organ donation, especially in the paediatric population (Fig. 3) [10,11]. The prominent azygos vein joins the superior vena cava at the normal location in the right paratracheal area. The hepatic veins drain in a short suprahepatic segment of the IVC (1–2 cm) and joins the right atrium immediately after the diaphragm [12–14].

LT in recipients with absence of infrahepatic IVC have been reported mainly in children, which incidence is up to 27.5% of paediatric recipients who required LT for biliary atresia. The earliest cases of biliary atresia associated with unsuspected congenital absence of infrahepatic IVC, preduodenal portal vein and anomalous hepatic artery had all fatal outcome due to poor inflow in two cases and venous outflow obstruction in one case; therefore it was emphasized for the first time the great importance of pre-operative recognition of vascular anomalies and technical adaptations for the successful of the surgery [15]. Later, several authors reported that left lateral segment graft could be successfully implanted for the syndromic biliary atresia with absence of IVC by the interposition of a neo-cava created with a cadaveric iliac vein graft interposition anastomotized by a triangular orifice with the left lateral segment [16–20]. More recently, a successful case of living donor LT using a right-lobe liver without middle hepatic vein (MHV) was reported in an infant with biliary atresia and interrupted retrohepatic IVC. In this case, the donor liver graft had three major hepatic veins draining segments V–VII and VIII, which were confluent in order to create a large triple orifice by using a venous graft; this new single orifice was then end-to-end anastomosed to the recipient's suprahepatic IVC [21].

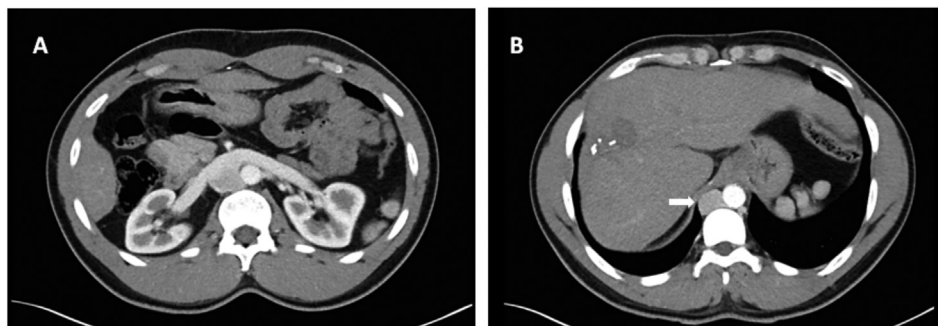


Fig. 2. Cross sectional imaging findings of absent inferior vena cava. 2A) IVC in a deep position just to the right of the aorta at the level of both right and left renal veins, 2B) lack of retro(intra)-hepatic course of the IVC and its course deep to the diaphragm.

Table 1
Characteristics (IVC most common anomalies adaptation of listed anomalies by Bass et al.³).

Anomaly	Prevalence	Embryological origin	Clinical significance
Left IVC (Typically the left IVC joins the left renal vein, which crosses anterior to aorta in the normal manner)	0.2–0.5%	Regression of right supracardinal vein, persistence of the left supracardinal vein.	- Misdiagnosis as left sided para aortic lymphadenopathy.
Duplicated IVC	0.2–3%	Persistence of <i>both</i> supracardinal veins.	- Misdiagnosis of lymphadenopathy. - Recurrent pulmonary embolism following IVC filter placement.
Azygos continuation of IVC/Absence of hepatic IVC (The retrohepatic IVC is often not completely absent but drains via a short course directly into the right atrium. The azygos enters the SVC in the right paratracheal space. The gonads drain into ipsilateral renal veins)	0.6%	Theorized to be failure to form the right subcardinal-hepatic anastomosis; resulting in atrophy of the right subcardinal vein. Blood is then shunted through the retrocrural azygos vein (partially derived from thoracic segment of the right supracardinal vein).	- Misdiagnosis of right paratracheal mass/lymphadenopathy. - Associated with congenital heart disease, asplenia, polysplenia. - Increasingly recognised in asymptomatic patients since the advent of cross-sectional imaging.
Circumaortic Left Renal Vein (Two left renal veins present. The superior renal vein passes anterior to aorta and inferior renal vein passes posterior to aorta)	8.7%	Persistence of dorsal limb of left renal vein and dorsal arch of the renal collar (the intersupracardinal anastomosis).	- Misdiagnosis of retroperitoneal adenopathy.
Retroaortic Left Renal Vein	2.1%	Persistence of the dorsal arch of the renal collar (the ventral arch regresses).	Preoperative recognition.
Double IVC with Retroaortic Right Renal Vein and Hemiazygos continuation of the IVC	–	Persistence of left lumbar and thoracic supracardinal vein and left suprasubcardinal anastomosis, failure of right sub-cardinal-hepatic anastomosis. The right renal vein crosses into the left IVC posterior to the aorta and continues as the hemiazygos. The hemiazygos may cross at T8/T9 into azygos. Alternatively it may drain into the coronary vein via a persistent left SVC or into an accessory hemiazygos continuation to the left brachiocephalic vein.	- Alternate drainage pathway of hemiazygos into the coronary vein or left brachiocephalic vein may be mistaken for mediastinal mass and aortic dissection. - 1 case report of death from inadvertent ligation of hemiazygos to azygos continuation of left IVC during thoracic surgery. - 1 case report of Budd-Chiari syndrome. In this case the bulk of hepatic venous drainage was via the right renal vein to hemiazygos continuation of left IVC.
Double IVC with Retroaortic Left Renal Vein and Azygos continuation of IVC	–	Persistence of left supracardinal vein and dorsal arch of renal collar (regression of ventral limb). Additionally, the subcardinal-hepatic anastomosis fails.	Preoperative planning.
Circumcaval Ureter (Retrocaval Ureter) (The proximal ureter courses posterior to the IVC, then emerges to the right of the aorta)	–	Right supracardinal system fails to develop but right posterior cardinal vein persists.	Partial right ureteric obstruction and recurrent UTI Mostly on the right.
Absent infrarenal IVC with preservation of suprarenal portion (External/internal iliac veins join and form large ascending lumbar veins draining to azygos and hemiazygos)	–	Failure of all three paired venous systems or perinatal IVC thrombosis.	Mistaken for paraspinal mass.

Abbreviations: IVC, inferior vena cava; SVC, superior vena cava; UTI, urinary tract infection.

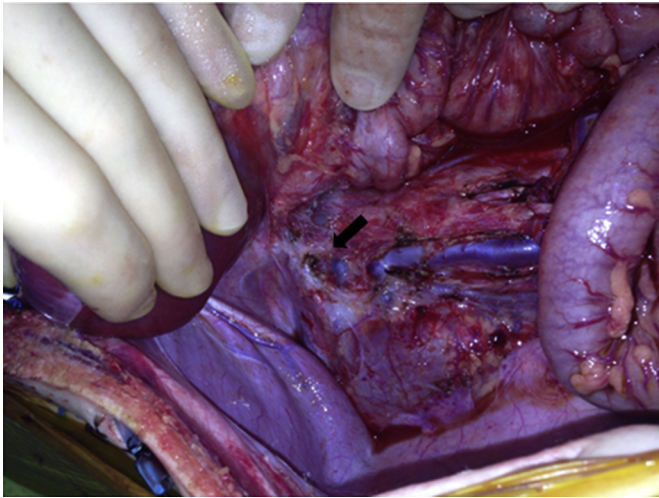


Fig. 3. Intraoperative imaging of congenital absence of IVC and azygous continuation. The azygous course is under the psoas muscle distal to the renal vein (two right renal veins in this case) insertion (arrow). The liver is held up which drains to the right heart through hepatic veins.

To the best of our knowledge, only three cases of adult LT with absence of IVC have been reported (Table 2) [4–6]. The first case described was a successful orthotopic LT where the graft was implanted by end-to-end suprahepatic IVC anastomosis similar to the technique adopted in our case [4]. Hatipoglu et al. [5] described a living donor LT performed in a recipient with fibrosis and atrophy of the retrohepatic IVC, in which the donor right lobe was implanted with a right hepatic vein end-to-side anastomosis to the recipient suprahepatic veins. Recently, one case of orthotopic LT in a patient with complex IVC anomaly (duplication of the infrarenal IVC, azygos continuation of the IVC, agenesis of the hepatic portion of the IVC and presence of several anomalous veins communicating the common iliac vein and the IVC of one side with the contralateral side) was also reported [6].

In cases herein hepatectomy phase was straightforward due to the preoperative identification of the absence of IVC, which permitted to the surgeons to pre-plan the surgical options. This included the planned implantation technique with “classical” piggyback technique as opposed to more commonly used “modified” piggyback technique. Ensuring the top end of vena cava is left open is one important surgical step in the backbench preparation of the liver and extension grafts may become necessary if this end is very short. Another technical modification is the obviated need for temporary porto-caval shunt. In cases where IVC is absent, porto-caval shut is usually not required however, if a shunt become an absolute necessity, a jump graft could be fashioned between the PV and the Azygous continuation as the vein dips deep in to the psoas muscle just above the renal veins. Therefore, the key features to look for are the IVC or Azygous continuation running behind the liver, or the absence of intrahepatic course of such veins.

The two cases presented in this report confirmed that the absence of infrahepatic IVC can be totally asymptomatic in adult recipients and may be presented as surprising finding during the surgery unless attention is paid for all cross sectional images. Previous radiological investigations as CT and MRCP are essential to identify such vascular anomalies. Once the absence of infrahepatic IVC is recognized, it requires only a slight modification of the implantation technique but it doesn't have any implication for the transplant. The preservation of adequate length of suprahepatic hepatic veins as they join the right atrium allows satisfactory outflow anastomosis.

Table 2
Adult LT recipients with congenital absence of IVC reported in literature.

Year	Author	Number of cases	Age of Patient (years)	Primary disease	IVC anomaly	Other anomalies	Surgical procedure and IVC anastomosis	Outcome
2012	Zinsler M.J. and Hanto D.W.	1	50	HCC/HCV chronic disease	Interrupted IVC with azygous continuation	–	OLT: end-to-end suprahepatic IVC anastomosis	Discharged on 9 POD, uncomplicated
2012	S.Hatipoglu et al.	1	21	Congenital hepatic fibrosis	Retro-hepatic IVC fibrotic and thrombosed	Pronounced dilatation of venous plexus (azygous, hemiazygos paravertebral, cutaneous, iliac)	LDLT: Donor right hepatic vein end-to-side anastomosis to the suprahepatic IVC	Alive at 5 months
2014	Julio C. U. et al.	1	57	HCC/HCV chronic disease	Absence of retro-hepatic IVC, azygos continuation of the IVC, duplication of the infrarenal IVC	Anomalous veins communicating the common iliac vein and the IVC of one side with the contralateral side	OLT: The right, middle, and left hepatic veins were sectioned and joined in a single, wide cuff, using venoplasty. This single orifice was anastomosed to the suprahepatic IVC of the new liver	Discharged on 6 POD, uncomplicated
2014	Angelico R. et al.	2	59	Primary Sclerosing Cholangitis	Retro-hepatic IVC absence	Atrophic left kidney	OLT: Recipient three hepatic vein orifices conjoined to form one single outflow tract for anastomosis with donor IVC	Discharged on 10 POD, uncomplicated
			35	HCC/HBV-related chironis	Retro-hepatic IVC absence	–	OLT: Donor suprahepatic IVC anastomosed with common orifice of the recipient hepatic veins draining into the right atrium	Alive at 13 months
								Discharged on 7 POD, uncomplicated
								Alive at 4 months

Abbreviations: IVC, inferior vena cava; HCC, hepatocellular carcinoma; HCV, Hepatitis C virus; HBV, Hepatitis B virus; OLT, orthotopic liver transplant; LDLT, Living donor liver transplantation; POD, post-operative day.

Ethical approval

Not applicable.

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None.

Author contribution

Angelico R designed the research and wrote the paper.

Stonelake S collected the data, literature review and drafting of manuscript.

Perera DS collected the data, literature review and drafting the manuscript.

Mirza DF edited and approved the final manuscript.

Russell S important intellectual content, anaesthetised for cases.

Muiesan P performed operation (2), edited and approved the final manuscript.

Perera MTPR performed operation (1), conceptualised the paper, wrote and approved the final manuscript.

Conflict of interest

None.

Guarantor

Perera MTPR.

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Not applicable.

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Abbreviations

IVC: Inferior vena cava

LT: Liver transplant

CIT: Cold ischemia time

PSC: Primary sclerosis cholangitis

MELD: Model for end-stage liver disease

US: Ultrasound scan

MRCP: Magnetic resonance cholangiopancreatography

CT: Computerised tomography

DCD: Donation after circulatory death

DBD: Donation after brain death

HCC: Hepatocellular carcinoma

HBV: Hepatitis B virus

MHV: Middle hepatic vein