

## Case Report

# Incidental Long Makuuchi Ligament and its Surgical Relevance in Pediatric Hepatic Resection

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## Abstract

The surgical relevance of Makuuchi ligament (Inferior Vena Cava ligament) has been less explored, especially in pediatric population. With the advent of major hepatectomies and liver transplantation indicated for various malignancies and metabolic conditions in children, knowledge about IVC ligament seems essential, in terms of its draining vessels, contained hepatic tissue and close relation with caudate lobe. Herein, we present a 2 years old female child diagnosed with hepatoblastoma (PRE-Treatment-EXTent of tumor II and POST-Treatment EXTent of tumor II) (PRETEXT II, and POSTTEXT II), with tumor involving right hepatic vein, who underwent right hepatectomy. Intraoperatively, after liver mobilization, we encountered a long IVC ligament, in the form of a broad membranous ligament bridging the left and right sides of caval groove encircling IVC completely. With the help of Cavitron Ultrasonic Aspirator (CUSA) and fine Ligaclips, the IVC ligament was meticulously ligated before tumor handling, preventing any major intraoperative bleeding, bile leak and possible tumor spread. Segments V-VIII was resected out to complete a formal anatomical right hepatectomy. The child was shifted to Intensive Care Unit (ICU) on ventilator for postoperative recovery and care. Postoperatively, we did not encounter any signs of hepatic failure and the subhepatic drain output was persistently minimal without any bleed or bile leak.

**Keywords:** IVC ligament; Makuuchi ligament; pediatric liver resection; Cavitron ultrasonic aspirator; Anatomic liver resection

## Introduction

With increasing advances in hepatic surgeries in pediatric population, including major hepatectomies and liver transplantation, the chances of manipulation of IVC ligament (otherwise known as Makuuchi ligament) has significantly increased [1]. Many cadaveric liver anatomies have been extensively studied to elucidate the surgical importance of IVC ligament mainly in hepatic resections. Makuuchi first described the importance of prior IVC ligament ligation in right hepatic lobectomies, in view of small vessels lurking in it [2]. Recently, the existence of small arteries, veins and lymphatics have been proven histologically in human cadaveric livers by Hirai et al. [3]. Some studies have reported the presence of hepatic tissue in this ligament, although not proven histologically [4].

These facts warrant careful manipulation of IVC ligament during hepatectomies especially in conditions of malignancies, keeping in mind the risks of intraoperative blood loss and tumor spread. The

handling becomes technically more difficult if extraordinarily longer Makuuchi ligaments are encountered, similar to the index case.

## Case Presentation

We received a two year's old well thrived female child, who was being evaluated outside for right flank swelling and occasional pain for 2 months. Her perinatal period was uneventful and she had no associated history of fever or bowel- bladder involvement. On examination, a large, firm, non-tender right hypochondriac mass of size 10 cm × 12 cm was palpated, reaching up to the epigastrium and moving with respiration. It was continuous with liver dullness on percussion.

Ultrasonography (USG) was suggestive of a well-defined hepatic mass measuring 10 cm × 10 cm × 9 cm, arising from segments V, VI, VII with central hypoechogenic area closely abutting right branch of portal vein. Contrast Enhanced Computed Tomography (CECT) chest, abdomen and pelvis depicted similar findings with exophytic component extending into the right subhepatic space. The lesion was found to cause significant mass effect, closely abutting with portal vein, IVC and bowel loops medially. It was supplied by branches of right hepatic artery and no evidence of vascular invasion found and labelled as hepatoblastoma PRETEXT II. The Makuuchi ligament was found in the form of a membranous tissue bridging the edges of caval groove with IVC completely embedded in it (Figure 1A). Liver enzymes were within normal range and tumor markers were slightly raised with Alpha-Fetoprotein (AFP)- 3249 ng/dl and beta human chorionic gonadotropin (Beta HCG)-0.24 mIU/ml. She received six cycles of neoadjuvant chemotherapy as per PLADO regimen under standard risk protocol.

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Post chemotherapy CECT showed a remarkable decrease in size of lesion (3 cm × 3 cm × 2.7 cm), involving segments V and VI with mild extension into right subhepatic space. It was closely abutting right branch of portal vein and IVC without any signs of invasions. There was no other evidence of distant metastasis (Figure 1B).

She underwent exploratory laparotomy with liberal right Makuuchi incision. Right and left triangular ligaments were divided to mobilize the liver. An exophytic mass of size 3 cm × 3 cm × 3 cm arising from segment VI of Right lobe of liver, densely adherent to right branch of portal vein and posterolateral aspect of intrahepatic IVC. Long Makuuchi ligament (vascular) of size 6 cm was noticed, with draining tributaries in to IVC (Figure 2A). Right anatomical hepatectomy performed along the Cantlie's line, exposing the surface drained by tributaries of middle hepatic vein with the help of Cavitron Ultrasonic Surgical Aspirator (CUSA) and clip ligators to achieve hemostasis (Figure 2B and C). The time of surgery was significantly prolonged due to incidental long Makuuchi ligament completely encircling the IVC with draining vessels. Chemotherapy related loss of planes and adhesions between Makuuchi ligament and IVC made it more tedious and need for meticulous dissection to avoid injury.

Post resection methylene blue injection into Gall Bladder (GB) after clamping distal common bile duct, revealed no evidence of bile leak from cut surface. Post operatively, the child was shifted on ventilator to pediatric ICU, where he was optimized and extubated after 48 hours. Weaned off gradually to room air. Feeds were started on day 4. The drain output gradually decreased and removed on day 10.

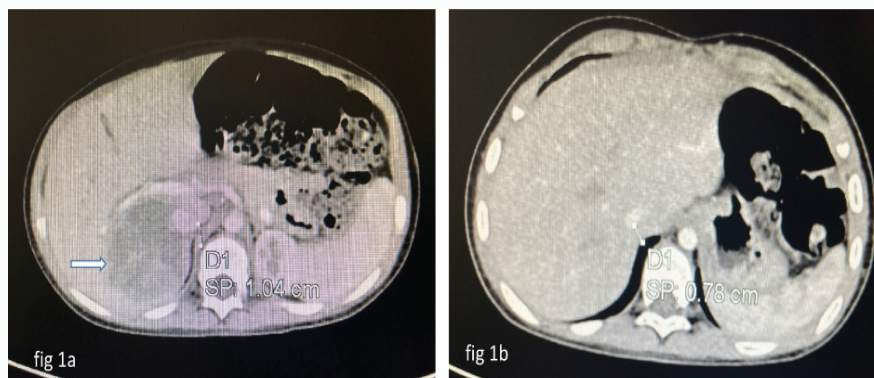
The histopathological examination of liver surface close to the Makuuchi ligament showed predominantly vascular channels devoid of viable tumor and predominantly post chemotherapy related changes, in the form of large areas of necrosis, hyalinisation, foamy histiocytic and inflammatory cell infiltration (Figure 3). The child is under regular follow up with normal radiology and doing well.

## Discussion

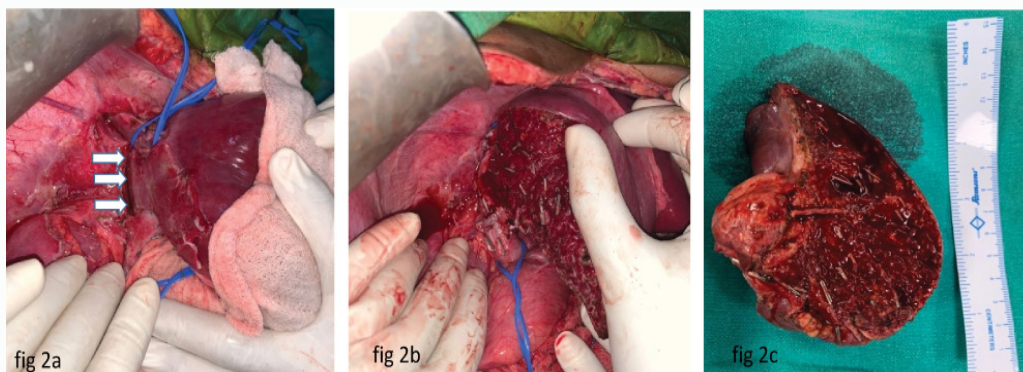
Although, the existence of Makuuchi ligament is not so widely accepted due to scarcity of data in the referential anatomical literature regarding its frequency and morphology, the surgical interest is in rising trend with ongoing development of liver surgeries.

The existence of IVC ligament dates back to the period of embryogenesis, where IVC is surrounded by hepatic parenchyma. At the end, it persists as an atrophied bridge of parenchyma which links the right and left hemiliver behind the IVC [2]. Morjane et al. [5] studied liver specimens from 43 human cadavers, where the IVC ligament persisted in 77% of cases. Further dissection of IVC ligament revealed the extrahepatic terminal part of Right Hepatic Vein (RHV) in all cases.

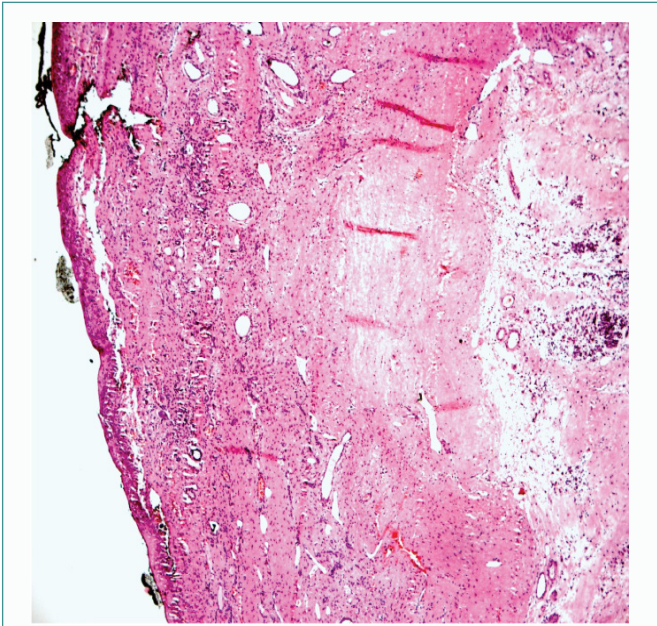
As indicated from many researches, the Makuuchi ligament seems to be a broad band like connective tissue which bridges both edges of caval groove thus embedding IVC [3]. Hirai et al. [3] stressed upon the fact that employment of energy sources in the form of endo linear staplers, laparo- ultrasonic coagulating shears are beneficial to decrease the risk of bile leak and bleeding [4]. They demonstrated their results based on histology of 16 specimens from human



**Figure 1 :** (1a) Contrast enhanced computed tomography (Pre chemotherapy)- broad membranous tissue (Makuuchi ligament) bridging the edges of caval groove with IVC completely embedded in it. Hepatic mass measuring 10 cm × 10 cm × 9 cm, arising from segments V, VI, VII with central hypoechoic area closely abutting right branch of portal vein (arrow). (1b) Post chemotherapy pictures showed drastic resolution of lesion closely abutting IVC.



**Figure 2 :** Operative pictures (2a) Suprahepatic and infrahepatic IVC control- Makuuchi ligament completely embedding IVC (white arrow). (2b) Right anatomical hepatectomy performed along the Cantlie's line, exposing the surface drained by tributaries of middle hepatic vein. (2c) Post hepatectomy specimen.



**Figure 3:** Histopathological examination showed a liver surface of attached Makuuchi ligament (inked) with presence of multiple vascular channels with a lesion comprising post chemotherapy related changes (H & E, 100X).

cadavers. Due to presence of larger number of vessels and lymphatics in the caudal portion, IVC ligaments were preferably be separated in the craniocaudal direction. Thus, concluding that extrahepatic major hepatic veins should be dissected after hemostatic ligation of IVC ligament.

This surgical plane of dissection between the Makuuchi ligament and the IVC becomes obliterated after chemotherapy induced fibrosis as was in our case. Utmost care and patience is required to avoid inadvertent injury to IVC during this part of surgery. In another cadaveric liver study, Morjane et al. [5] showed that IVC ligament being a terminal part of RHV, right hemiliver resection with elective vascular control was anatomically possible in 85% of cases and difficult to perform in 15% of the cases.

Kogure et al. [6] revealed that IVC ligament is not a simple connective tissue, rather a degenerated band of hepatocytes comprised of portal triads with portal venules, hepatic artery branches and biliary radicals etc [6]. Similar findings were contributed by Sarmiento et al. [4] suggesting the existence of hepatic tissue with well-formed portal triads in Makuuchi ligament.

Champetier et al. [7] identified aberrant biliary radicals IVC ligament which were in continuity with other- intrahepatic biliary ducts with irregular islands of hepatocytes distributed in between.

## Conclusion

The IVC ligament was inconstant but frequent anatomic structure, situated above the right suprarenal vein covering the inflow of right hepatic vein into the IVC. It is the persisting terminal extrahepatic part of RHV which may enable elective vascular control during right hemiliver surgery.

The consistent presence of portal triad components and occasionally ectopic hepatocytes suggest significant relevance for hepatic surgeons. These components may harbor carcinogenic potential which may necessitate checking for presence of tumor in IVC ligament, as well as in the IVC in the preoperative and intraoperative analysis, which again demands telltale awareness among surgeons dealing with liver pathologies in children.

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