Review Article

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# The Hilar Cholangiocarcinoma (Klatskin Tumor)

**Yigang Luo**

**Corresponding author**

Yigang Luo, Transplantation and Hepatobiliary Pancreatic Surgery, University of Saskatchewan, Ellis Hall, Rm161, 103 Hospital Drive, Saskatoon SK S7N 0W8, Canada.

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## INTRODUCTION

Hepatic hilar cholangiocarcinoma was initially reported by Altemeir in 1957 [1]. Klatskin [2] reported a series of 13 cases of hepatic hilar cholangiocarcinoma in 1965. About 60% of cholangiocarcinomas are composed of this tumor. Anatomically speaking, this tumor is located at a unique location, namely the hilar biliary bifurcation within a constrained tiny space. the liver (particularly the caudate lobe) and veins (portal vein, hepatic artery). Biology-wise, it often grows slowly and locally, with lymph node metastases, submucosal infiltration (up to 1.6 cm from the tumor's gross margin), and neurovascular infiltration; distant metastasis occurs less frequently. In terms of treatment, it is typically difficult to resect, particularly when attempting a R0 resection, and it does not react well to chemotherapy or radiation therapy. The high rate of local recurrence (>50%) causes therapy failure and unfavorable results.

### Staging/Classification

There have been several staging/classification systems over the years, such as AJCC (American Joint Committee of Cancer), Liver Cancer Study Group of Japan, MSKCC (Memorial Sloan Kettering Cancer Center), Bismuth/Corlette, and most recently European HPBA (European Hepato-Pancreato-Biliary Association). Dr. Henry Bisthmus (Figure 1) [4] is credited with the most well-known classification of the tumor.

according to the anatomic locations:

Tumors classified as

Type 1 affect only the hepatic bile duct;

Type 2 affect the bile duct bifurcation;

Type 3a affect both the right and left hepatic bile ducts;

Type 3b affects both the left and right hepatic bile ducts;

and so on.

e) Type 4: The tumor affects the hepatic bile ducts on both sides. Within clinical practice, this method is commonly utilized. It was predicated on the tumor's involvement with the biliary tree at the anatomic level. This does aid in the surgical plan's formulation. It does not, however, address the lymph node, metastasis, liver parenchyma, or vascular involvement status. Consequently, when liver resection and vascular resection/reconstruction are taken into account, it is less beneficial.

A classification system based on cancer growing pattern, mass formation, periductal infiltration, and intraductal growing was proposed by the Liver Cancer Study Group of Japan in 2000 (Figure 2) [5]. This was more about the biology behavior of cancer, with improved intraductal growth and mass formation prognoses. It is doubtful that detailed information regarding this classification would be available prior to surgery. Furthermore, the liver's lobar condition and vascular involvement were not described. As a result, this classification has little use in surgically evaluating resectability.

### Surgical management update

Five-year survival of hilar cholangiocarcinoma (Klatskin tumor) was less than 7.3% after initial local excision [9]. These were not good long-term results. An increasing number of studies in the 1990s revealed that R0 resection led to prolonged survival. It is discovered that there are many local recurrences and comparatively few distant metastases of Klatskin tumors. Consequently, it was encouraged to combine radical choledochectomy with liver lobectomy, particularly when the right hemi- or right extended hemihepatectomy was combined with caudate lobe resection [10,11]. The 5-year survival rate rose to between 30 and 40 percent. On the other hand, postoperative morbidity and mortality reached 59% and 11%, respectively. However, Chen XP et al. [12] released a study in the British Journal of Surgery in 2009.

With a 5-year survival rate of 34%, a minor limited hepatectomy may not always result in a worse prognosis for patients whose malignancies did not include vascular structures, since the postoperative morbidity and mortality from a major expanded resection were significantly reduced.

Abass S and Sandrassi C conducted a mega-analysis to review the topic of vascular repair and resection [13]. Out of 2457 instances, 669 patients underwent vascular resection; of them, 22–88% had a positive pathology result; 36–88% of patients had a R0 resection; the corresponding morbidity and mortality rates were 22–88% and 2–15%. Survival over five years was 20–56%. A multi-institutional review of 305 cases was described by Jong MC et al. in 2012 [14], demonstrating that portal vein resection should be performed when necessary to eradicate all illness. For certain patients with advanced hilar cholangiocarcinoma, a combination of liver resection,

extra-hepatice bile duct resection, and portal vein resection may provide long-term survival. For almost two decades, attempts have been made to treat cholangiocarcinoma by liver transplantation. However, the five-year survival rate was only 28% before to 2000. The outcomes with neo-adjuvant treatment were getting better with time. 2005 saw the Mayo Clinic's Rea SR et al. [15] implement a laborious and stringent preoperative selection and treatment strategy. In terms of 1, 3, and 5-year survival, the numbers were noticeably better: 92%, 82%, and 82%. Treatment options such as liver transplantation, however limited to a carefully selected patient population, more easily achieve R0 resection without causing problems with vascular involvement, intrahepatic bile duct, or lobar atrophy.

**New Clinical Classification Proposal**

Previous staging and classification systems only described surgical management related to cancer status and did not fully account for the therapeutic process. Three factors ultimately decide the outcome of a cancer patient's treatment.

a) The patient's overall health and tolerance to various therapeutic interventions;

b) The disease itself;

c) How the cancer responds to treatment.

Based on clinical treatment options according to the patient's state and cancer status, this new clinical classification has been developed. With ongoing advancements in therapeutic technologies, including as neoadjuvant therapy, chemotherapy, radiation therapy, and transplantation, more patients will receive radical treatment with better outcomes in the future. Here, a new clinical therapeutic classification is presented in order to account for all currently accessible therapeutic choices.

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