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**Hilar cholangiocarcinoma**

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**Abstract**

Hilar cholangiocarcinoma is a rare tumor. It accounts for 2/3 of the tumors of the biliary tract. Untreated, prognosis is very poor. Surgery is the only therapy that offers the possibility of cure but is technically very complex. With recent improvements in the therapeutic strategies applied by multidisciplinary teams, survival rates in the different series currently range from 25% to 45%. A group of experts devoted to hilar cholangiocarcinoma (pathologists, gastroenterologists, radiologists, surgeons and oncologists) have reviewed and updated every open question in hilar cholangiocarcinoma.

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**Key words:** Cholangiocarcinoma; Hilar; Perihilar; Klatskin; Surgery; Cancer; Review

**Core tip:** Most remarked avances are: imaging methods have improved diagnostic sensitivity and specificity, especially for determining biliary and vascular involvement; there have been several proposals to improve the classic Bismuth-Corlette classification; pre- and post-operative care; technical aspects trying to obtain a R0 resection: widespread use of liver resection, resection of segment I and venous and arterial resection, refinement of post-operative histology and adjuvant therapies.

Ramia JM. Hilar cholangiocarcinoma

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**HILAR CHOLANGIOCARCINOMA**

Hilar cholangiocarcinoma (HC) is a rare tumor (0.74-1.05×100000 inhabitants), representing 3% of gastrointestinal cancers[1,2]. It accounts for 2/3 of the tumors of the biliary tract[1]. Although HC is frequently discussed at surgical scientific meetings and arouses great interest, it has far fewer citations than other HPB cancers and several questions regarding the condition remain unanswered in the medical literature.

Untreated, the prognosis of HC is very poor. Surgery is the only therapy that offers the possibility of cure but is technically very complex[1,3]. A few years ago, the number of patients operated and resected was very low, and 5-year survival was mediocre. With recent improvements in the therapeutic strategies applied by multidisciplinary teams comprising radiologists, gastroenterologists, surgeons and oncologists, survival rates in the different series currently range from 25% to 45%[3,4].

In the past decade, progress has been made in the following areas of the management and treatment of patients with HC: (1) Diagnosis: Imaging methods have improved diagnostic sensitivity and specificity, especially for determining biliary and vascular involvement. These methods include multi-detector computed tomography, Cholangio- magnetic resonance imaging, percutaneous transhepatic cholangiography, positron emission tomography/computed tomography, staging by laparoscopy; (2) Classification/staging: There have been several proposals to improve the classic Bismuth-Corlette classification, among them the Memorial Sloan-Kettering Center system, the 7th edition of TNM, and the most complete system so far devised by de Oliveira *et al*[2]. Correct preoperative staging is essential to identify the patients who should be operated; (3) Pre- and post-operative care: Several pre-operative measures have been used to minimize risks and improve results: uni- or bi-lobar pre-operative biliary drainage, the use of probiotics, autologous blood, replacement of drained bile, portal embolization, enteral nutrition, and so on[3]; (4) Technical aspects: The widespread use of liver resection (above 80% in the most recent series), resection of segment I, venous resection (performed in 6%-43% of patients), arterial resection, and the non-touch technique have improved survival for many reasons, but above all because they allow an increase in R0 resections. All these technical improvements have been achieved without significantly increasing morbidity and mortality[1-3]; mortality in HC surgery currently ranges between 0% and 11.9%[3]; (5) Liver transplantation: Once an absolute contraindication, liver transplantation now has a limited role and is appropriate in selected patients with HC; (6) Refinement of post-operative histology: This is essential for deciding which patients should receive adjuvant treatment; and (7) Adjuvant treatment: There have been advances in chemotherapy used, brachytherapy, photodynamic therapy, and so on.

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