Malignant tumors of the bile ducts remain a major management challenge. They occur most commonly in the proximal bile duct, at the hepatic duct confluence (hilar cholangiocarcinoma), or in the distal common bile duct near the papilla (distal cholangiocarcinoma) (Fig. 9–1). Resection offers the best chance of long-term survival, but many patients present with advanced disease that is not amenable to extirpation. The assessment and treatment of distal bile duct cancers is similar to that of cancers of the head of the pancreas. This chapter focuses on management issues specific to hilar cholangiocarcinoma.

**EPIDEMIOLOGY**

Cholangiocarcinoma is an uncommon disease. The annual incidence in the United States is 1 to 2 per 100,000, and it accounts for approximately 2 percent of all cancers.1 Men and women are affected in nearly equal proportions. Although occasionally seen in young patients, cholangiocarcinoma is primarily a disease of the elderly. The majority of patients are over 65 years of age, and the peak incidence occurs in the eighth decade of life.1 Untreated, hilar cholangiocarcinoma generally results in death within 12 months, from liver failure or from complications arising from biliary tract obstruction.

**ETIOLOGY**

Cholangiocarcinoma usually occurs sporadically, and most patients have no obvious risk factors. While the etiology of cholangiocarcinoma is unknown, there are several conditions that are associated with an increased risk.

**Primary Sclerosing Cholangitis**

Perhaps the most common associated condition in Western countries is primary sclerosing cholangitis (PSC), an autoimmune disease characterized by...
inflammation of the periductal tissues, ultimately resulting in multifocal strictures of the intrahepatic and extrahepatic bile ducts.\textsuperscript{2,3} The natural history of PSC is quite variable, and the true incidence of cholangiocarcinoma is unknown. The reported incidence in PSC ranges from 8 to 40 percent.\textsuperscript{2,3} Unlike most patients with sporadic cholangiocarcinoma of the extrahepatic biliary tree, patients with PSC are at risk for multifocal disease that is often not amenable to resection.

Congenital Biliary Cystic Disease

The increased risk of cholangiocarcinoma in patients with congenital biliary cystic disease (choledochal cysts, Caroli’s disease) is well described.\textsuperscript{4} Malignant degeneration is uncommon in choledochal cysts that have been completely excised early. However, the incidence of cancer increases substantially (approximately 15 to 20\%) in patients who are not treated until after the age of 20 years or in those previously treated by cyst drainage.\textsuperscript{5,6} The reason for the high incidence of cancer in patients with cystic diseases appears to be related to an abnormal choledochopancreatic duct junction that predisposes to reflux of pancreatic secretions into the biliary tree, bacterial contamination, and chronic inflammation.\textsuperscript{5-7} An increased incidence of cholangiocarcinoma has also been reported in patients subjected to transduodenal sphincteroplasty,\textsuperscript{8} and a similar mechanism may be responsible.

Hepatolithiasis and Biliary Parasites

Hepatolithiasis, also known as recurrent pyogenic cholangiohepatitis or Oriental cholangiohepatitis, is prevalent in Japan and parts of Southeast Asia. Obstruction of intrahepatic ducts leads to chronic recurrent episodes of cholangitis and stricture forma-

### Table 9–1. INCIDENCE OF HILAR CHOLANGIOCARCINOMA IN SELECTED SERIES

<table>
<thead>
<tr>
<th>Institution</th>
<th>Years*</th>
<th>Patients with Cholangiocarcinoma</th>
<th>Patients with Hilar Cholangiocarcinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>UCLA (1981)\textsuperscript{T}</td>
<td>24</td>
<td>96</td>
<td>47 (49%)</td>
</tr>
<tr>
<td>Mayo Clinic (1993)\textsuperscript{12}</td>
<td>9</td>
<td>171</td>
<td>79 (46%)</td>
</tr>
<tr>
<td>Johns Hopkins (1996)\textsuperscript{13}</td>
<td>23</td>
<td>294</td>
<td>197 (67%)</td>
</tr>
<tr>
<td>MSKCC (1998)\textsuperscript{14}</td>
<td>6</td>
<td>225</td>
<td>90 (40%)</td>
</tr>
</tbody>
</table>

\textsuperscript{T}MSKCC = Memorial Sloan-Kettering Cancer Center; UCLA = University of California, Los Angeles.
\textsuperscript{*}Time interval over which patients were accumulated.

LOCATION

Although cholangiocarcinoma can arise anywhere within the biliary tree, hilar cholangiocarcinoma or tumors involving the biliary confluence are the most common (see Fig. 9–1). In most large series, the proportion of patients with tumors involving the hilus ranges from 40 to 60 percent (Table 9–1).\textsuperscript{12-14} Tumors of the distal bile duct are somewhat less common, accounting for 20 to 30 percent of all cholangiocarcinomas and 5 to 10 percent of all periampullary tumors.\textsuperscript{13,15,16} Approximately 10 percent of cholangiocarcinomas arise within the intrahepatic biliary tree.\textsuperscript{17-19} These tumors usually present as an intrahepatic mass, and although uncommon, jaundice may result from intrabiliary tumor extension (Fig. 9–2).

HISTOPATHOLOGY

The overwhelming majority of cholangiocarcinomas are adenocarcinomas, often well differentiated and mucin producing.\textsuperscript{1,20}

There are three well-described macroscopic subtypes of cholangiocarcinoma: sclerosing, nodular, and papillary.\textsuperscript{20} Sclerosing tumors account for the majority of cases. Sclerosing tumors are very firm and cause an annular thickening of the bile duct, often with diffuse infiltration and fibrosis of the periductal tissues (Fig. 9–3). Nodular tumors are characterized by a firm irregular nodule of tumor that projects into the lumen of the duct. Features of both types are often seen, hence the frequently used
descriptor “nodular-sclerosing.” The papillary variant accounts for approximately 10 percent of all cholangiocarcinomas and is more common in the distal bile duct. A polypoid mass that expands rather than contracts the duct is a characteristic feature (Fig. 9–4). Although papillary tumors may grow to significant size and appear surgically unapproachable (based on radiographic studies), they often arise from a well-defined stalk and histologically may be confined to the superficial layers of the duct wall (Fig. 9–5). Recognition of papillary cholangiocarcinomas is important since they are more often resectable and have a more favorable prognosis than the other types.

Longitudinal spread along the duct wall and periductal tissues is an important pathologic feature of cholangiocarcinomas. There may be substantial extension of tumor beneath an intact epithelial lining—as much as 2 cm proximally and 1 cm distally. The full tumor extent may thus be underestimated by radiographic studies and may not be fully appreciated by palpation at the time of resection. This feature underscores the importance of checking all resection margins with frozen-section histology at operation.

STAGING

According to the American Joint Commission on Cancer (AJCC), the staging of extrahepatic cholangiocarcinomas is based on the extent of the primary tumor (T stage), the extent of regional lymph node
involvement (N stage), and the presence of distant metastases (M stage) (Table 9–2). An alternative staging system proposed by Bismuth and Corlette classifies cholangiocarcinomas on the basis of their location with respect to the hilus and on the extent of ductal involvement.\textsuperscript{23} The AJCC staging is based largely on pathologic criteria and has little clinical applicability. The majority of patients in our practice have T3 (stage 4A) tumors based on invasion into the liver, but this alone says very little about their resectability. Moreover, this finding does not correlate with survival, provided that complete resection

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure9_3.png}
\caption{A. Endoscopic cholangiogram showing the typical irregular biliary stricture characteristic of sclerosing adenocarcinomas (arrow). These tumors cause a circumferential thickening of the duct wall and periductal fibrosis, which ultimately obliterates the lumen. (Reproduced with permission from Jarnagin WR, Saldinger PF, Blumgart LH. Cancer of the bile ducts: the hepatic ducts and common bile duct. In: Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. 3rd ed. Edinburgh [UK]: Churchill Livingstone; 2000.) \(B\), Coronal magnetic resonance cholangiopancreatography reconstruction showing an infiltrating tumor involving the hepatic duct confluence (arrow). The right and left intrahepatic ducts, which appear white, are dilated.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure9_4.png}
\caption{A. Transhepatic cholangiogram showing a markedly dilated bile duct from the confluence of the right and left hepatic ducts to the ampulla of Vater. Filling defects are apparent throughout the length of the duct (black arrows). A percutaneous biliary stent is in place (white arrow). \(B\). Cross-sectional computed tomography image of the same patient. The stent (white arrowhead) is seen within the duct, adjacent to a large tumor mass (white arrow). The underlying portal vein (black arrow) is free, and there is no evidence of lobar atrophy.}
\end{figure}
Figure 9–5. A, Excised bile duct from the patient discussed in Fig. 9–4. The duct has been opened longitudinally to reveal two separate polypoid masses (arrows). B, View looking down the incised bile duct, with one of the tumors seen arising from the mucosal surface. Most of the tumor mass extends into the bile duct lumen. C, Photomicrograph showing the malignant cells largely confined to the mucosa of the duct wall (arrow) (hematoxylin-eosin stain, low power view).
Table 9–2. CURRENT AMERICAN JOINT COMMISSION ON CANCER STAGING SYSTEM FOR CANCER OF EXTRAHEPATIC BILE DUCTS

<table>
<thead>
<tr>
<th>Stage</th>
<th>Tumor</th>
<th>Node</th>
<th>Metastasis</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Tis</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>1</td>
<td>T1</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>2</td>
<td>T2</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>3</td>
<td>T1 or 2</td>
<td>N1 or 2</td>
<td>M0</td>
</tr>
<tr>
<td>4A</td>
<td>T3</td>
<td>Any N</td>
<td>M0</td>
</tr>
<tr>
<td>4B</td>
<td>Any T</td>
<td>Any N</td>
<td>M1</td>
</tr>
</tbody>
</table>

Tis = carcinoma in situ; T1 = tumor invades subepithelial connective tissue or fibromuscular layer; T2 = tumor invades perifibromuscular connective tissue; T3 = tumor invades adjacent organs (liver, pancreas, duodenum, gallbladder, colon, stomach).

N0 = no regional lymph node metastases; N1 = metastasis to lymph nodes within the hepatoduodenal ligament (cystic duct, pericholedochal, and/or hilar lymph nodes); N2 = metastasis to peripancreatic, periduodenal, periporal, celiac, superior mesenteric, and/or posterior pancreaticoduodenal lymph nodes.

M0 = no distant metastasis; M1 = distant metastasis.

can be achieved. In the AJCC system, patients with involvement of N1- and N2-lymph nodes are staged similarly. This is inappropriate since patients with metastatic disease to N2-level lymph nodes (celiac, periduodenal, or retroperitoneal) are not candidates for resection and should be considered to have M1 disease. The Bismuth-Corlette system is more clinically relevant but is too simplified and, like the AJCC system, correlates poorly with resectability and survival. None of the current staging systems takes into account local factors such as vascular invasion or hepatic lobar atrophy, which are important determinants of resectability, which (in turn) is a major determinant of outcome. A more reliable and systematic method of selecting candidates for resection is clearly needed (see below).

DIAGNOSIS

Symptoms and Physical Examination

The early symptoms of hilar cholangiocarcinoma are nonspecific. Abdominal pain or discomfort, anorexia, weight loss, and pruritus are the most common symptoms and are seen only in one-third of patients. Most of these tumors come to attention because of jaundice or abnormal liver function tests. Jaundice is not present initially in cases of incomplete biliary obstruction (ie, right or left hepatic duct) or segmental ductal obstruction. Patients with papillary tumors of the hilus may have a history of fluctuating jaundice, which may be the result of loose tumor fragments intermittently obstructing the bile duct. In patients with cholangiocarcinoma and no previous biliary intervention, cholangitis is uncommon at initial presentation despite a 30 percent incidence of bacterial contamination (bacterbilia).

The physical findings are often nonspecific. Jaundice will usually be obvious. The gallbladder is usually decompressed and nonpalpable with hilar obstruction; thus, a palpable gallbladder suggests a more distal obstruction or an alternative diagnosis. Patients with long-standing biliary obstruction and/or portal vein involvement may have findings consistent with portal hypertension.

Alternative Diagnoses

The vast majority of patients with proximal biliary obstruction and jaundice have cholangiocarcinoma. However, alternative diagnoses are possible and can be expected in 10 to 15 percent of patients. Gallbladder carcinoma, Mirizzi syndrome, and idiopathic benign focal stenosis (“malignant masquerade”) are the most common conditions that may be mistaken for cholangiocarcinoma. Over a 28-month period at Memorial Sloan-Kettering Cancer Center, we evaluated 109 patients with obstruction involving the biliary confluence consistent with hilar cholangiocarcinoma. In 90 patients, this diagnosis was confirmed while 19 patients were found to have other conditions (9 with gallbladder cancer, 6 with malignant masquerade, 3 with neuroendocrine tumor of the bile duct, and 1 with Mirizzi syndrome).

Distinguishing gallbladder carcinoma from hilar cholangiocarcinoma can be difficult. A thickened and irregular gallbladder with infiltration into segment IV, selective involvement of the right portal pedicle, and obstruction of the common hepatic duct with occlusion of the cystic duct on endoscopic cholangiography are all suggestive of gallbladder carcinoma. Mirizzi syndrome is a benign condition resulting from a large gallstone impacted in the neck of the gallbladder (Fig. 9–6). Benign focal strictures (malignant masquerade) can occur at the
hepatic duct confluence but are uncommon (Fig. 9–7). Relying on the results of percutaneous needle biopsy or biliary brush cytology is dangerous since they are often misleading and because one may miss the opportunity to resect an early cancer. It is often impossible to establish the diagnosis unequivocally before operation, and this is indeed unnecessary in nearly all cases since lesions at the biliary confluence (including benign strictures) are best assessed and treated at operation. We firmly believe that in the absence of clear contraindications (see below), exploration is indicated in all patients with suspicious hilar lesions.

**TREATMENT**

**Preoperative Evaluation**

Evaluation of patients with hilar cholangiocarcinoma is principally an assessment of resectability,
since resection is the only effective therapy. First and foremost, the patient’s general health and fitness for major surgery (possibly including partial hepatectomy) must be examined. The presence of significant comorbid conditions, chronic liver disease, and/or portal hypertension generally precludes resection. Patients with potentially resectable tumors may develop biliary tract sepsis, often after intubation of the biliary tree. These patients require resuscitation and treatment of the infection before surgery is undertaken.

The preoperative evaluation must address four critical determinants of resectability: (1) tumor extent within the biliary tree, (2) vascular invasion, (3) hepatic lobar atrophy, and (4) metastatic disease. Tumors that extend into the second-order biliary radicles bilaterally are not amenable to resection with clear margins, but this is often difficult to determine before operation. Likewise, the presence of metastatic disease (to distant sites or N2-level lymph nodes) is a contraindication to resection. Lobar atrophy is an often overlooked but important factor in assessing the resectability of these tumors (see below). Long-standing biliary obstruction may cause moderate atrophy whereas concomitant portal venous compromise induces rapid and severe atrophy of the involved segments. On cross-sectional imaging, atrophy is characterized by a small and often hypoperfused lobe, with crowding of the dilated intrahepatic ducts (Fig. 9–8).

Radiographic Studies

High-quality radiographic studies are critical in selecting patients for resection. In the past, computed tomography (CT), percutaneous transhepatic cholangiography (PTC), and angiography were considered the standard investigations. Currently, we rely almost exclusively on magnetic resonance cholangiopancreatography (MRCP), and duplex ultrasonography (US). These noninvasive studies provide the same information regarding the extent of disease as the aforementioned studies but with less risk.

Magnetic resonance cholangiopancreatography has emerged as a powerful investigative tool, and we prefer it to the more invasive endoscopic retrograde cholangiography (ERCP) or PTC. Several studies have demonstrated the utility of MRCP in evaluating patients with biliary obstruction. It may identify the tumor and level of obstruction and may also reveal obstructed and isolated ducts not seen by PTC. It also provides information regarding the patency of hilar vascular structures, the presence of nodal or distant metastases, and lobar

Figure 9–8. Left, Cross-sectional computed tomography image of a patient with cholangiocarcinoma involving the left hepatic duct. The left lobe, clearly demarcated from the right, is shrunken and hypoperfused (arrows). The bile ducts, which appear black, are dilated and crowded. Right, T1-weighted gadolinium-enhanced cross-sectional magnetic resonance image of a different patient with hilar cholangiocarcinoma involving the left hepatic duct. The left lobe is small, and the bile ducts (which appear black) are dilated and crowded. There is evidence of hypoperfusion of the left lobe, which appears darker. There is also evidence of hypoperfusion of the right posterior sector, which also appears darker and is clearly demarcated from the well-perfused right anterior sector (arrowheads) but does not as yet show evidence of atrophy. These radiographic features suggest that the tumor involves the left hepatic duct and the left portal vein and is beginning to involve the right portal pedicle. (Reproduced with permission from Jarnagin WR, Saldinger PF, Blumgart LH. Cancer of the bile ducts: the hepatic ducts and common bile duct. In: Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. 3rd ed. Edinburgh [UK]: Churchill Livingstone; 2000.)
atrophy (Figs. 9–9 and 9–10). Furthermore, unlike direct cholangiography, MRCP does not require biliary intubation. Potential infectious complications, which may increase operative morbidity, are thus avoided.

Duplex US is a valuable study in patients with hilar obstruction; in experienced hands, US rivals all other modalities for assessing extent of disease and vascular involvement36–38 (Fig. 9–11). In our practice, MRCP and duplex US have become the preferred investigations.

**Patient Selection**

Distinguishing resectable from irresectable tumors demands careful consideration of all available data. Bilateral tumor extension to second-order intrahepatic biliary radicles and encasement or occlusion of the main portal vein clearly preclude resection. On the other hand, involvement of the right or left branch of the portal vein does not necessarily preclude resection. In this situation, complete resection will require ipsilateral partial hepatectomy, but this would not be possible if the contralateral lobe is atrophic or if the tumor extends to the contralateral second-order biliary radicles. The detailed criteria of irresectability are outlined in Table 9–3.

Despite improvements in radiologic techniques, only 20 to 50 percent of patients submitted to exploration have tumors that can be resected.13,14,39 We have therefore proposed a preoperative staging system aimed at improving patient selection.14 Based on the radiographic interpretation of local tumor
extent (T stage), this system analyzes resectability and considers the following factors: extent of biliary ductal involvement, portal venous involvement, and lobar atrophy (Table 9–4). Analyzing our patient data by using the proposed system, we found that resectability and the need for partial hepatectomy both increased progressively with increasing T stage while 5-year survival was inversely related (Table 9–5). By contrast, there was no correlation between these variables and AJCC stage. The proposed stag-
Hilar Cholangiocarcinoma

The treatment goals in patients with hilar cholangiocarcinoma are resection with negative histologic margins and the restoration of enterobiliary continu-

Tumor Stage Biliary Involvement Ipsilateral Lobar Atrophy Ipsilateral Portal Vein Involvement Main Portal Vein Involvement
T1 Hilus and/or right or left hepatic duct No No No
T2 Hilus and/or right or left hepatic duct Yes No No
T3 Hilus and/or right or left hepatic duct Yes/No Yes No
T4 Secondary biliary radicles bilaterally Yes/No Yes/No Yes

T1 tumors are confined to the hilus with or without involvement of the right or left hepatic duct and are not associated with atrophy or portal vein involvement. T2 tumors are associated with ipsilateral lobar atrophy but not portal vein branch involvement. T3 tumors are characterized by ipsilateral portal venous branch involvement and may or may not be associated with lobar atrophy. T4 tumors are defined as irresectable, based on tumor extension to secondary biliary radicles bilaterally or encasement/occlusion of the main portal vein.

Adapted from Burke E, et al. Hilar cholangiocarcinoma. Patterns of spread, the importance of hepatic resection for curative operation, and a presurgical clinical staging system. Ann Surg 1998;228:387 by permission of the publisher Lippincott Williams and Wilkins.
Hepatic transplantation is associated with a prohibitively high incidence of recurrence, which has led many centers to abandon its routine use in this disease. All patients should be evaluated for possible resection before any intervention. Ill-advised attempts to place biliary stents, especially when not indicated, may result in complications that delay operation, and stent-associated infection and inflammation may hinder further radiographic and operative assessment. If resection is clearly not feasible, then transtumoral drainage or intrahepatic enterobiliary bypass offers satisfactory palliation (see below).

At operation, the objective should be the complete resection of the tumor, with negative histologic margins. Partial hepatectomy is usually required to achieve this goal. Several recent series show a strong correlation between negative histologic margins and the percentage of patients subjected to partial hepatectomy (Table 9–6). Additionally, tumors that extend into the left hepatic duct usually involve the principal caudate lobe ducts and require en bloc caudate lobectomy (see Fig. 9–10).

Operative Technique

The operation begins with a thorough exploration of the abdomen. Contrary to popular belief, metastatic disease is common in patients with hilar cholangiocarcinoma. Performing staging laparoscopy before proceeding to open exploration may help identify some patients with unresectable disease.

The details of resection are illustrated in Figures 9–12 to 9–16. The duodenum and liver are mobilized, and the gallbladder is taken down. The bridge of liver tissue that often connects segments III and IV should be divided to expose the left hepatic duct at the base of the umbilical fissure. Lowering the hilar plate will expose the biliary confluence. Palpation of the duct in this area will give the surgeon a sense of the tumor extent. The distal bile duct is transected and elevated upwards, allowing inspection of the anterior wall of the portal vein for evidence of tumor invasion; a portion of the distal duct should be sent for frozen-section histology. Once it has been determined that resection is feasible and will leave behind a tumor-free liver remnant with intact blood supply and biliary drainage, the surgeon should then control and divide the inflow blood supply to the lobe that is to be removed. The contralateral hepatic duct above the tumor should be divided likewise, and a portion should be sent for frozen-section histology. The hepatic vein draining that portion of the liver to be excised should be exposed and divided extrahepatically. The parenchymal transection should now be pursued. Finally, enterobiliary reconstruction is completed, using a 70-cm Roux-en-Y loop of jejunum.

Results

It must be stressed that with the approach outlined above, long-term survival can be achieved with an acceptable operative mortality. Patients resected with negative histologic margins survive significantly longer than those with microscopically positive margins (Table 9–7). Over the past 20 years, there has been a steady increase in the use of hepatic resection in patients with hilar cholangiocarcinoma, resulting in an increase in the proportion of R0 resections (ie, resections with neg-

<table>
<thead>
<tr>
<th>Author</th>
<th>Period of Study (yr)</th>
<th>Resected (N)</th>
<th>Partial Hepatectomy (%)</th>
<th>Negative Margins (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cameron (1990)</td>
<td>15</td>
<td>39</td>
<td>20</td>
<td>15</td>
</tr>
<tr>
<td>Hadjis (1990)</td>
<td>8</td>
<td>27</td>
<td>60</td>
<td>56</td>
</tr>
<tr>
<td>Nimura (1990)</td>
<td>10</td>
<td>55</td>
<td>98</td>
<td>83</td>
</tr>
<tr>
<td>Klempnauer (1997)</td>
<td>25</td>
<td>147</td>
<td>79</td>
<td>79</td>
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<td>Burke (1998)</td>
<td>6</td>
<td>30</td>
<td>73</td>
<td>83</td>
</tr>
</tbody>
</table>

Adapted from Burke E, et al. Hilar cholangiocarcinoma. Patterns of spread, the importance of hepatic resection for curative operation, and a presurgical clinical staging system. Ann Surg 1998;228:390 by permission of the publisher Lippincott Williams and Wilkins.
Figure 9–12. Initial operative approach. The round ligament has been divided and is held upwards, exposing the undersurface of the liver. The hilar plate at the base of segment IV has been lowered, exposing the tumor at the biliary confluence. The bridge of liver tissue attaching segments III and IV has been divided; this provides greater exposure of the left hepatic duct. The gallbladder has been taken down, and the distal bile duct has been divided. (Reproduced with permission from Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. Edinburgh [UK]: Churchill Livingstone; 1997 [CD-ROM].)

Figure 9–13. Upward traction on the divided bile duct and periductal lymphatic tissues exposes the underlying portal vein. Developing the plane between the tumor and the portal vein is the only way to assess vascular invasion. Provided that the vein is not involved, a plane between the tumor and the portal vein can easily be developed. Once it has been determined that resection is feasible, the right hepatic artery is divided and retracted upwards with the tumor. The left hepatic artery is retracted laterally, away from the tumor. (Reproduced with permission from Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. Edinburgh [UK]: Churchill Livingstone; 1997 [CD-ROM].)

Figure 9–14. The left hepatic duct is divided well beyond the tumor, and a portion is sent for frozen-section analysis. Further dissection exposes the portal vein confluence, and the right branch is divided. (Reproduced with permission from Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. Edinburgh [UK]: Churchill Livingstone; 1997 [CD-ROM].)

Figure 9–15. The hepatic parenchyma is divided after extrahepatic control of the right hepatic vein has been achieved. We crush the parenchyma with a Kelley clamp and control intrahepatic vessels and ducts with clips and ties. The central venous pressure is closely monitored and kept under 5 mm Hg until the liver has been divided. (Reproduced with permission from Blumgart LH, Fong Y, editors. Surgery of the liver and biliary tract. Edinburgh [UK]: Churchill Livingstone; 1997 [CD-ROM].)
Palliation

The majority of patients with cholangiocarcinoma have irresectable disease at presentation or at laparotomy. Palliative biliary decompression may be accomplished surgically or by percutaneous transmural biliary stenting. Adequate drainage of hilar tumors is more difficult to achieve endoscopically, and this approach is associated with a higher failure rate and greater complications. Most patients found to have irresectable tumors at presentation are submitted for percutaneous drainage whereas those found to have irresectable disease at laparotomy may be suitable for intrahepatic enterobiliary bypass. External biliary drainage catheters can often provide relief of jaundice or itching. When left in place for long periods of time, however, these catheters can be a source of cholangitis and may require frequent changing. In addition, tumor growth along the tube tract can occur, which can sometimes result in a painful mass in the subcutaneous tissues (Fig. 9–17). Self-expandable biliary endoprostheses, such as Wallstents, are permanent internal devices that avoid many of the problems and complications associated with external drainage catheters. These stents, while an improvement, are prone to occlusion secondary to tumor growth.

Table 9–7. INFLUENCE OF HISTOLOGIC MARGINS ON SURVIVAL AFTER RESECTION OF HILAR CHOLANGIOCARCINOMA

<table>
<thead>
<tr>
<th>Study</th>
<th>R0 Resections</th>
<th>R1 Resections</th>
<th>R0 Survival (mo)</th>
<th>R1 Survival (mo)</th>
<th>Operative Mortality (%)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hadjis 1990</td>
<td>12</td>
<td>15</td>
<td>43</td>
<td>25</td>
<td>7</td>
<td>.04</td>
</tr>
<tr>
<td>Pichlmayr 1996*</td>
<td>91</td>
<td>27</td>
<td>26</td>
<td>13</td>
<td>10</td>
<td>.01</td>
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<tr>
<td>Burke 1998</td>
<td>25</td>
<td>5</td>
<td>&gt; 60</td>
<td>22</td>
<td>7</td>
<td>.01</td>
</tr>
</tbody>
</table>

R0 = resection with negative histologic margin; R1 = resection with positive histologic margin; p = probability.


Adapted from Burke E, et al. Hilar cholangiocarcinoma. Patterns of spread, the importance of hepatic resection for curative operation, and a presurgical clinical staging system. Ann Surg 1998;228:391 by permission of the publisher Lippincott Williams and Wilkins.

Table 9–8. EXPERIENCE WITH 269 HILAR CHOLANGIOCARCINOMA PATIENTS OVER 20 YEARS

<table>
<thead>
<tr>
<th></th>
<th></th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients evaluated</td>
<td>131</td>
<td>48</td>
<td>90</td>
</tr>
<tr>
<td>Patients resected</td>
<td>27</td>
<td>21</td>
<td>30</td>
</tr>
<tr>
<td>Negative margin</td>
<td>15 (56%)</td>
<td>14 (67%)</td>
<td>25 (83%)</td>
</tr>
<tr>
<td>Hepatectomies performed</td>
<td>16 (60%)</td>
<td>9 (43%)</td>
<td>22 (73%)</td>
</tr>
<tr>
<td>Operative mortality (%)</td>
<td>7.4</td>
<td>5</td>
<td>6</td>
</tr>
<tr>
<td>Median survival (mo)</td>
<td>25</td>
<td>36</td>
<td>40</td>
</tr>
<tr>
<td>5-yr survival (%)</td>
<td>22</td>
<td>NR</td>
<td>56</td>
</tr>
</tbody>
</table>

NR = not reported.
Chemotherapy has not been shown to improve survival in patients with unresectable disease, and its use in this setting is difficult to justify outside the context of a clinical trial.

Palliative Radiation Therapy

Patients with irresectable locally advanced tumors may be candidates for palliative radiation therapy. A combination of external beam radiation and intraluminal iridium-192 delivered percutaneously is typically used. Several authors have demonstrated the feasibility of this approach although improved survival compared with that achieved with biliary decompression alone has not been documented in a controlled study.47,57–59

Photodynamic Therapy

Ortner and colleagues reported their limited experience with photodynamic therapy in patients with irresectable hilar cholangiocarcinoma.60 This approach has been used successfully as a palliative measure for tumors of the esophagus, colon, stomach, bronchus, bladder, and brain. Although promising as a means of providing biliary decompression without stents or perhaps as a way of treating those with microscopically involved bile duct margins, photodynamic therapy clearly needs further scrutiny before it is widely adopted.

Figure 9–17. Computed tomography scan showing tumor growth along a percutaneous tube tract (arrowhead), with a large subcutaneous tumor mass.

Figure 9–18. A, Photograph of a Wallstent removed at operation in a patient with hilar cholangiocarcinoma. Notice the ingrowth of tissue into the stent wall. B, Computed tomography scan of a patient with recurrent cancer after resection. A Wallstent had been previously placed to relieve a biliary obstruction (black arrowhead); it subsequently became occluded and caused biliary obstruction proximal to the stent. The left hepatic duct (white arrowhead) is markedly dilated as a result of the occluded stent within its lumen.
REFERENCES


