



California Pacific
Medical Center

A Sutter Health Affiliate
With You. For Life.

Cholangiocarcinoma

LIVER DISEASE MANAGEMENT & TRANSPLANT PROGRAM

At California Pacific Medical Center we are committed to bringing new and advanced diagnostic tools, medical treatments and surgical options to the physicians and patients we serve. Through this disease and treatment profile, our physicians illustrate current and emergent treatment options we can provide for the specialized medical management of your patients.

Our promise to our patients is to deliver the highest quality expert care with kindness and compassion. We go beyond medicine to treat the whole person, not just the illness.

Medicine can transform a body. Going beyond medicine can transform a life.

For patient referrals:

1-888-637-2762

www.cpmc.org/liver

Beyond Medicine.

Overview

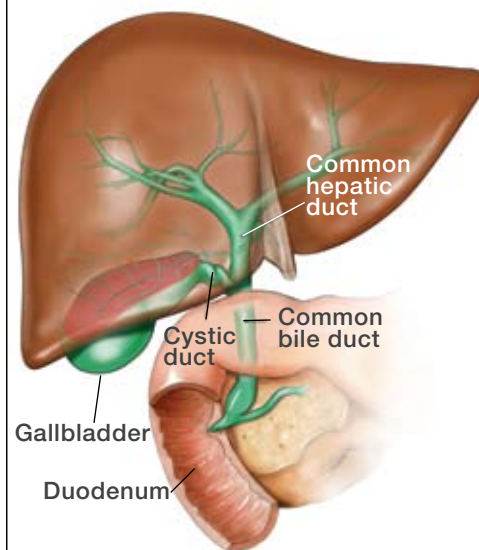
Cancer of the biliary system—including the gallbladder, bile ducts within the liver and bile ducts extending outside of the liver that drain into the intestine—is relatively uncommon, affecting about 9,000 individuals in the United States annually. Of these 9,000, about 2/3 have cancer limited to the gallbladder, while the remaining 1/3 have cancer of the intra- and/or extrahepatic biliary tree.

RISK FACTORS & PRESENTATION

Risk factors for biliary cancers include chronic inflammation from infection or autoimmune disease, particularly primary sclerosing cholangitis (PSC). Cholangiocarcinoma typically presents with signs of tumor obstruction of the bile duct. It may be localized (Klatskin's tumor) versus a diffuse process (cholangiocellular) within the biliary tree.

Early manifestations include pruritis (itchiness) and change in urine and stool color. Blood chemistry tests may reveal elevation of liver enzymes. Clinical jaundice occurs later and may be accompanied by abdominal pain, fever and weight loss. However, these clinical features are not specific to cholangiocarcinoma. A variety of other diseases and disorders ranging from benign (e.g. bile duct stone or inflammatory stricture) to malignant (e.g. pancreatic or liver tumors) may present similarly.

The Biliary Tree



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The gallbladder lies on the right of the liver. The biliary tree within the liver is referred to as "intra-hepatic" while "extrahepatic" refers to the biliary system outside the liver. A bile duct tumor(s) can occur in any of these locations.

Diagnosis

Distinguishing cholangiocarcinoma from mimicking conditions is imperative in guiding the appropriate treatment. A range of diagnostic tools is available to assess patients with suspected cholangiocarcinoma. These include noninvasive radiological imaging tests such as ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI) or "invasive" tests.

NON-INVASIVE

Ultrasound, CT and MRI

Ultrasound is often the initial study to rule out more common causes of jaundice. For individuals with suspected cholangiocarcinoma, evaluation begins with a computed tomography (CT) scan or magnetic resonance imaging (MRI). Specific CT imaging protocols and MRI sequences optimize visualization of the biliary system and liver. Computer workstations and software-enabled image reconstruction create 3D views of the biliary anatomy and identify the suspected region of the tumor from various angles. Additional information on the local tumor extent (including vascular involvement) and distant metastatic spread is seen on CT or MRI.

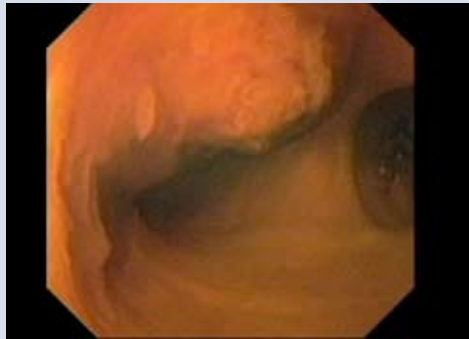
INVASIVE

Endoscopic Retrograde Cholangiopancreatography (ERCP)

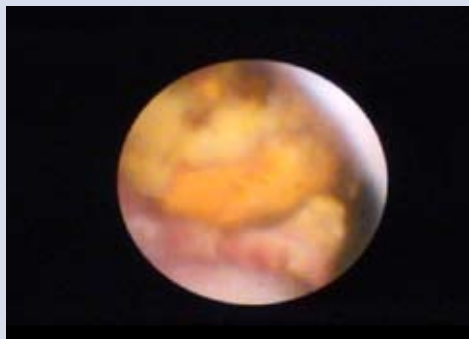
This diagnostic tool has been the mainstay in evaluating suspected cholangiocarcinoma. ERCP uses an endoscope to gain access to the bile duct from the duodenum. A small catheter inserted into the bile duct with contrast media subsequently injected provides radiographic images of the entire biliary system. ERCP also provides a means for passing tools into the bile duct to assist in diagnosis and/or treatment.

Cholangioscopy

Cholangioscopy involves insertion of a miniature endoscope (cholangioscope) into the bile duct to directly view the internal bile duct lining. The cholangioscope provides both visual assessment of strictures and a means to perform visual-guided biopsies. Recent advances in endoscopic imaging facilitate the differentiation of tumor from benign inflammatory tissue.



Bile duct tumor on direct video cholangioscopy.



Fiberoptic cholangioscopy using the SpyGlass® system reveals bile duct stone in a patient with suspected tumor.

Endoscopic Ultrasound (EUS)

EUS involves two complementary platforms: extraductal and intraductal. Extraductal ultrasound uses an ultrasound probe mounted at the tip of an endoscope to provide detailed, real-time

ultrasonic images of the extrahepatic biliary system without instrumenting the bile duct directly. Intraductal ultrasound involves the insertion of a catheter-based ultrasound probe into the bile duct. This probe, which can be advanced into the intrahepatic bile ducts, provides highly detailed images of the bile duct wall lining.

Percutaneous Transhepatic Cholangiography

Occasionally, to characterize the extent of a tumor into the liver, a radiopaque dye is injected into the biliary tree by placing a needle through the abdominal wall into the liver.

Treatment

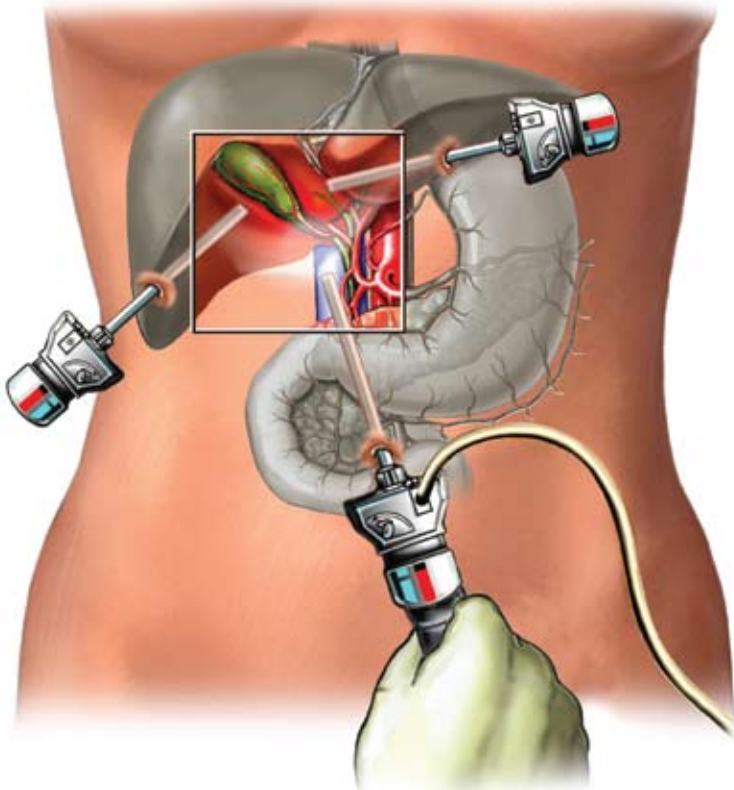
SURGICAL

For surgical interventions, cholangiocarcinomas are divided into intrahepatic and extrahepatic bile duct cancers. Intrahepatic tumors, when confined to a single lobe or segment of the liver, without evidence of metastasis, are resected with their surrounding liver lobe or segments, much as any primary liver tumor is addressed.



EUS reveals a mass within the extrahepatic bile duct.

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survival due to the frequency of recurrent tumor. The surgery itself has a perioperative mortality of 5-10% with major perioperative morbidity of up to 50%.

For tumors high in the biliary tree (Bismuth-Corlette Types IIIA and IIIB), the surgical resection involves a partial hepatic resection along with resection of the extra-hepatic biliary tree. The biliary system is then reconstructed as a hepaticojejunostomy to the remaining hepatic lobe.

For tumors low in the biliary tree (Bismuth-Corlette Type I lesions), a pancreatoduodenectomy is required in addition to the bile duct resection to effect a negative margin. Mid-duct tumors (Bismuth-Corlette Type II lesions) can be primarily resected without the need for either a hepatic resection or pancreatotomy.

In all cases, reconstruction after the resection involves bringing a fashioned limb of intestine up to the residual bile duct to permit biliary drainage into the intestines. This is often performed over a stent which remains in place for a period of time postoperatively. Postoperative radiation and/or chemotherapy may also be recommended, although studies to date have mixed results as to the benefits of adjuvant therapy with resected cholangiocarcinomas. Occasionally intraluminal brachytherapy is recommended with similar mixed results experienced and reported to date.

Laparoscopic evaluation of the liver and bile duct enables surgeons to determine the extent of disease prior to an open surgical procedure.

For extrahepatic bile duct cancers, if the lesion by preoperative evaluation appears resectable, the patient is prepared for the operating room and often undergoes a diagnostic laparoscopy immediately prior to the planned surgical resection to rule out unresectable tumor not evident by the preoperative evaluation.

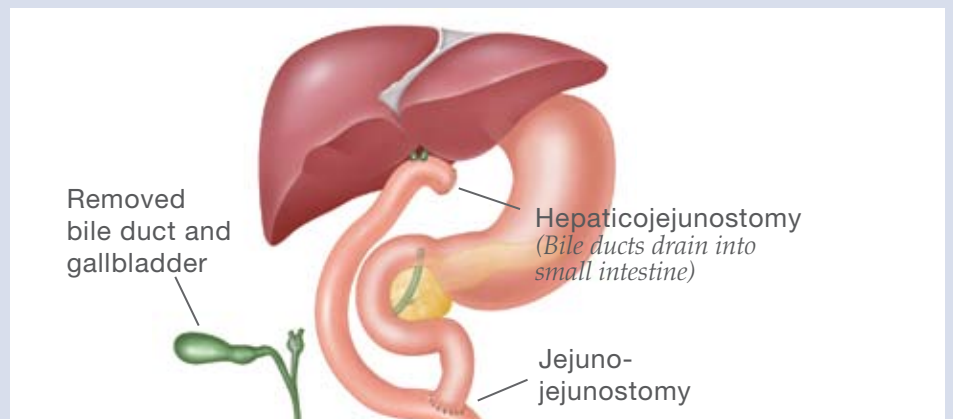
If this laparoscopic evaluation shows extensive disease which cannot be surgically removed, the procedure is stopped without making a large surgical incision. The individual is referred for endoscopic biliary stent decompression and concomitant treatment with radiation and/or chemotherapy.

RESECTABLE LESIONS

If the initial laparoscopic exploration appears favorable or there is low suspicion of extensive disease, the open surgical procedure ensues

with planned removal of the tumor. The primary surgical objectives are to obtain margins free of residual tumor and removal of associated lymph nodes which may also be involved with the tumor. Surgical resection offers the only opportunity for cure which, unfortunately, remains low with approximately 25-30% five-year

Roux-en-Y Hepaticojejunostomy Procedure performed for cholangiocarcinoma and biliary injuries.



UNRESECTABLE LESIONS

For lesions found at surgical laparotomy to be unresectable, the objective changes from removing the entire lesion with negative margins to instead decompressing the objective changes from removing the entire lesion with negative margins to instead decompressing the obstructed biliary tree. Depending on the type of tumor and its position in the biliary system, the biliary bypass procedure employed would vary. After biliary decompression, the patient, once recovered from surgery, is referred for consideration for radiation and/or chemotherapy. Unfortunately, with nonresectable lesions, limited life expectancy with median survivals of two to 10 months is anticipated.

TRANSPLANTATION

Liver transplantation is generally contraindicated in patients with cholangiocarcinomas. However, a very small minority of carefully screened patients found to have surgically unresectable disease may be considered for liver transplantation under experimental protocols with a favorable long-term survival benefit.

Stent Therapy

Endoscopic therapy consists of dilation of the obstructive biliary segment (with rigid or balloon dilation catheters) and metallic biliary stenting. The goal of treatment is to permanently widen the bile duct to a diameter that allows unimpeded bile flow. Endoscopic therapy may require several interval procedural sessions.

For unresectable lesions in which endoscopic stenting is not possible, a percutaneous stent placed through the abdominal wall can be an acceptable alternative to effect biliary drainage.

Medical Treatment

For unresectable tumors and often as adjunctive therapy for resected tumors, physicians may employ chemotherapy, radiotherapy and/or brachytherapy to palliate symptoms and prolong survival as well as increase the opportunity for surgical cures.

As only a small percentage of patients are cured with surgery alone, our focus at California Pacific Medical Center is to integrate all available treatments. We also periodically offer experimental therapies.

Why Choose Us

Hepatobiliary and pancreas diseases—disorders of the liver, bile ducts, gallbladder and pancreas—form a complex set of medical problems whose treatment often requires equally challenging surgical procedures. At California Pacific Medical Center, we have been leaders in hepatobiliary and pancreas surgery since the founding of our liver transplant program in 1988. Our doctors are closely involved in clinical research and surgical innovation. Annually, our physicians provide care to some 4,000 hepatobiliary and pancreas patients, both in San Francisco and at our network of outreach sites in California and Nevada.

For patients requiring hospitalization, we have a dedicated critical care liver unit, hospitalists who specialize in hepatobiliary disease, physician assistants, on-call anesthesia staff and a specialized O.R. nursing team. At California Pacific, our focus is on providing experienced, personalized care for all patients.

CANCER NAVIGATION SERVICE

Our Cancer Care Navigation Service provides individuals and families assistance with appointment scheduling, patient education and support service referral. Call 1-866-975-COPE (2673) or email patientnavigation@sutterhealth.org.

GENETIC RISK ASSESSMENT

The Cancer Genetic Risk Assessment Program at California Pacific offers individuals with a personal or family history of cholangiocarcinoma the opportunity to learn more about the genetic nature of their disease and whether they may be predisposed to other cancers which they could monitor. Individuals meet with our genetic counselor during which an evaluation of one's medical and family history is performed, as well as a detailed risk assessment and genetic education. If appropriate, genetic testing may be offered and facilitated by the genetic counselor. A genetic risk assessment may assist in medical management decisions such as aggressive cancer screening and preventive measures. For more information, call the Cancer Genetic Risk Assessment Program at 415-600-5961 or visit www.cpmc.org/services/cancer-genetesting/

For more information

Hepatobiliary Program

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San Francisco, Calif. 94115
www.cpmc.org/liver

Hepatologists

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Fax. 415-600-1020

Hepatobiliary Surgeons

Tel. 415-600-1010
Fax. 415-600-1012

Complex Gastroenterology

Tel. 415-600-1151
Fax. 415-600-1416

Oncologists

Tel. 415-923-3012
Fax. 415-928-4840

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