

Sample FAQs

Q: What determines whether cholangiocarcinoma can be surgically removed?

This answer is excerpted from the e-book “100 Questions & Answers *About* Biliary Cancer” by Dr. Ghassan Abou-Alfa & Dr. Eileen M. O’Reilly which is available for free download through the Cholangiocarcinoma Foundation Website.

A: “Several factors determine whether cholangiocarcinoma can be removed. These include your general health condition, the condition of the cancer-free portion of your liver, and the distribution of the cancer and its relationship to vital structures in the liver including major blood vessels and bile ducts.

You must be in good health in order to tolerate general anesthesia and a major operation such as a liver resection. For instance, if a patient has a weak heart, an attempt at a liver resection may not be advisable as it may be too dangerous. Additionally, the noncancerous part of your liver must be healthy enough to tolerate a liver resection. The liver is a unique organ because it can regrow after part of it is removed. The liver is divided into right and left lobes and further divided into eight individual segments. Up to six of these segments (roughly 80%) of a healthy liver can typically be removed. Remarkably, the liver regrows within several weeks after removal, and normal liver function is restored within 4 weeks of a liver resection.

However, some patients with cholangiocarcinoma have cirrhosis of the liver (underlying liver malfunction) and may be able to have only a small percentage of the liver safely removed. Because patients with advanced cirrhosis often have **varices** (enlarged blood vessels at risk of major bleeding) or have blood that has trouble clotting, most patients with advanced cirrhosis cannot undergo any liver resection due to a high risk of bleeding to death during or shortly after the operation. These patients can also die from liver failure in the first few weeks after liver resection. While their liver may function well enough for everyday living, it may not be able to handle the stress of a liver resection.

In some patients, the portion of the liver that can remain following the operation is judged to be insufficient in size. Depending on the degree of cirrhosis and the amount expected to be removed, your surgeon may elect to grow a portion of your liver prior to the operation. This can be done using a procedure called **portal vein embolization**. During this procedure, an interventional radiologist injects a blocking material into the **portal vein** on the side of your tumor to decrease blood flow to that portion of your liver. This stimulates the side of your liver that is not affected to grow, reducing the percentage of your liver that is being removed. Having a large portion of your liver left behind following the operation will make it easier for you to recover.”

Q: What kind of scan is best to assess tumor response to treatment?

This answer is excerpted from the e-book “100 Questions & Answers *About Biliary Cancer*” by Dr. Ghassan Abou-Alfa & Dr. Eileen M. O’Reilly which is available for free download through the Cholangiocarcinoma Foundation Website.

A: “Since many patients who receive chemotherapy have metastatic cancer, a CT scan or an MRI is usually used to assess the status of the cancer. Before undergoing a CT scan, you will be given an oral contrast agent to drink immediately before the scan. An intravenous contrast agent will also be infused into your vein. The oral contrast agent will help the radiologist assess the intestinal tract, while the IV contrast agent will help visualize tumors in your liver, blood vessels, and other areas of the body. When you undergo a CT scan, you will lie down on a table and a donut-like part of the scanning machine will circle around you, taking pictures of the interior of your body. Depending on which parts of the body are scanned, the procedure may take anywhere from 10–30 minutes.

If you undergo an MRI, you will be given an intravenous contrast agent before lying on a table that is enclosed within the scanner. You may feel uncomfortable in such a tight place. An MRI can be noisy and typically take longer than a CT scan, often lasting around 45 minutes. Talk to your doctor and/or radiologist about which is the best test for you.

Occasionally, a PET scan may be recommended after a CT scan or MRI. A PET scan involves the injection of glucose into your vein and may help determine whether a finding on a CT scan or MRI is in fact cancerous or not. You can expect to have a repeat evaluation, preferably using the same type of scan, every 2 to 4 months. The radiologist can evaluate the cancer by measuring it and looking for changes compared to the previous scan. These measurements are compared with the original CT scan or an MRI that was taken before the start of therapy and any scans taken since.”

Q: What is hospice care? When is it time to start hospice?

This answer is excerpted from the e-book “100 Questions & Answers *About Biliary Cancer*” by Dr. Ghassan Abou-Alfa & Dr. Eileen M. O’Reilly which is available for free download through the Cholangiocarcinoma Foundation Website.

A: “If your medical condition worsens, you may become debilitated and need a great deal of help and support. This can be provided through hospice care. **Hospice** is a whole-system care approach that addresses medical, physical, emotional, social, and spiritual needs for patients with advanced-stage disease. This care can be provided at home, especially if your family is able and ready to provide the physical care. A hospice registered nurse and possibly a hospice care physician will visit you on a regular basis and as frequently as is necessary. These regular assessments will ensure that you remain comfortable and that all of your needs are addressed. The hospice care team will consult regularly with your doctor. If necessary, you may be provided with a home health aide to assist you with your physical needs, such as bathing.

Although the hospice caring team will be present at your home for only a few hours a day, these resources are available 24 hours a day, 7 days a week in case an emergency arises.

You also may have hospice care provided at a hospice facility as an inpatient. This can be because of personal wishes, the lack of availability of family members, or if you require strenuous care that your family members cannot provide. You can still have quality time with your family and loved ones at an inpatient hospice facility, as most have extended visiting hours. Meanwhile, the staff will provide all of the care that you need.

You are entitled to hospice care after you and your doctor decide not to pursue any further active care, such as chemotherapy, and your doctor may recommend hospice care if your cancer is life threatening. Additional information is available at (www.hospice-info.net) and (www.hospicenet.org). Some hospice facilities provide palliative care or specific spiritual care. You can discuss with your doctor what options are available or visit the website of the National Hospice and Palliative Care Organization at (www.nhpco.org).”

Q: My mom’s afraid of chemotherapy. What should I do?

This FAQ is excerpted from the “Ask Dr. Giles” page on the Cholangiocarcinoma Foundation website. Dr. Geret Giles is a licensed psychologist who takes questions through the Foundation website from cholangiocarcinoma patients, family members and caregivers.

“My mother is scared about the pain and is on the fence to take the Chemo. We have talked with the Doctor about the drugs and the side effects and she feels as though she doesn’t want to prolong what is bound to happen anyway.

A: Choosing a treatment path is a very difficult and complicated task. It sounds like your mother has a specific path she wants to take and you are wondering if that is the best path for her quality of life and overall welfare. As you have mentioned, it is important to consider all sorts of variables when deciding upon the way to proceed. Here at cholangiocarcinoma.org you’ll find patients and their loved ones who have followed any one of a number of treatment options. Use the Patient Support options of the website to gather information and interact with others. Those who populate our website are the true treasure of this organization and an invaluable resource. Ultimately, however, it is important to take the time to clearly understand your mother’s wishes and then to align yourself with them. She will be ever grateful for that demonstration of love, support, and loyalty.”

Q: I don’t know even a single other person with cholangiocarcinoma, and wish I had someone to talk to about experiences with this rare cancer. How can I connect with others in a similar situation to mine?

One option is to seek a mentor through the Cholangiocarcinoma Foundation’s CholangioConnect program. CholangioConnect creates personalized connections that enable one-on-one support among cholangiocarcinoma fighters, survivors and caregivers.

This peer-to-peer connection platform helps anyone touched by cholangiocarcinoma, at any stage in their journey, find a mentor to ask questions and get support from someone who has personal experience with the disease. You can find out more about CholangioConnect at <https://cholangiocarcinoma.org/cholangioconnect/>

There are also social media resources available for cholangiocarcinoma patients and caregivers. For example, there is an active Cholangiocarcinoma Warriors Facebook site that you may consider joining. Major cancer hospitals and local cancer organizations often organize patient support groups. While cholangiocarcinoma may be too rare to have its own group, it may be possible to identify a liver cancer or gastrointestinal cancer support group in your area.

Q: Is it better to have a metal or a plastic stent?

A: Stent questions are best directed to doctors, because the decision about which type of stent will depend on the characteristics of the individual patient's case and anatomy (namely, where the bile duct blockage is located). There are many medical journal articles discussing plastic vs. metal stents for biliary strictures, and also on covered vs. uncovered metal stents, which can be found via a search engine. The general finding in many of the studies is that metal stents are preferred if the patient's life expectancy is longer than 3-4 months. This reflects that plastic stents need to be periodically replaced, while metal stents last much longer and are considered more permanent. An exception to this criterion occurs when the patient's treatment plans are still being worked out. In that case, the shorter-term plastic stents, because they can be removed, may be preferred as an interim treatment. But, to repeat the earlier point, the decision on stenting for an individual patient will consider the specifics of the case, so general findings may not apply.

Q: My mother was diagnosed at age 72. Is she too elderly to consider demanding treatments such as liver resection surgery or chemotherapy?

A: There have been a number of research studies on the experience of elderly patients with liver resection surgery, and the good news is it is generally safe, assuming the patient is healthy enough to undertake the procedure. Prior to major surgery for any patients, doctors will carefully examine their health status to ensure surgery can proceed safely. Here is a link to a recent study. <https://www.ncbi.nlm.nih.gov/pubmed/29135100>
In the case of chemotherapy, the question can be more complicated and also depends on the health status of the patient. Your mother's doctors will look at her overall health and functioning, for example, does she have conditions such as heart or kidney ailments or anemia. They will look at the medications she already takes, which might negatively interact with chemo. Many older patients are able to tolerate chemotherapy, but some are not.

Q: How do I choose a clinical trial? Are the results of on-going trials published so I can see which ones are having good results?

A: Here is an American Cancer Society link on choosing a clinical trial. <https://www.cancer.org/treatment/treatments-and-side-effects/clinical-trials/what-you-need-to-know/picking-a-clinical-trial.html>. The clinicaltrials.gov website has a similar page.

Information on the results of an on-going trial is often not publicly available. If a trial is Phase 2 or 3, then there may be published reports (e.g., from medical conferences or medical journal articles) on the results from earlier phases. Sometimes early results are described in company reports published for its investors. If you search the name of the drug, the name of the company and what is being treated (e.g., cholangiocarcinoma or solid tumors), you can sometimes find this sort of report.

Early numbers, however, may not be all that revealing. If the trial has just a few patients enrolled, or if it is early in the study time-line, then information would be extremely preliminary. As with any treatment, there is regrettably no sure way to know in advance if a specific patient will have a positive response.

Using clinicaltrials.gov and the resources posted on the Cholangiocarcinoma Foundation website, it is possible to compile a list of potential clinical trials. Announcements for clinical trials usually give a point of contact, someone you can call to ask about the trial and see if it is an appropriate option. Your doctors may also know about different trials and may be able to help you make contact. In addition to what you can find out about the trial itself, other factors are also important to consider, such as whether the trial is close to home, how logistically demanding is the trial (e.g., how frequently will the patient have to report to the trial site, what testing is required and how rigorous the treatment is), and how much financial cost/insurance coverage will there be.

Q: I was just diagnosed with cholangiocarcinoma, and the oncologist in my local clinic has told me he does not think my tumor can be removed surgically. Should I seek a second opinion?

A: There have been several studies suggesting that cholangiocarcinoma patients diagnosed in community medical facilities are far less likely to be sent for surgery than those treated by major cancer hospitals. This is a concerning finding, as surgery is currently the only curative treatment for cholangiocarcinoma.

This finding is due to the rareness of cholangiocarcinoma as well as the highly demanding nature of the types of surgery performed for this cancer (usually liver resection or whipple). Local medical facilities rarely see patients with this cancer and may not have much experience with the needed surgeries and treatments. For this reason, seeking a second opinion about your diagnosis and treatment options from a major cancer center experienced with cholangiocarcinoma is essential. The Cholangiocarcinoma Foundation website includes a list of major cancer hospitals with cholangiocarcinoma experience.

Q: I just got my blood test results, and my CA 19-9 number has gone up by five points. My oncologist did not seem concerned. Should I be worried?

A: CA 19-9 (carbohydrate antigen 19-9) refers to a tumor marker blood test that measures presence in your blood of a protein that is shed by cells. The protein can be excessively shed by tumor cells of several cancers including cholangiocarcinoma, so the test is used to help diagnose and monitor our cancer.

CA 19-9 is regularly measured for patients undergoing treatments and those being watched for recurrence. The number can fluctuate – doctors watch the trends over time more so than individual small changes. CA 19-9 can rise temporarily for non-cancer reasons such as inflammation in the bile ducts or pancreas. Doctors track a range of patient information, not just blood tests but also periodic scans and patient reports on symptoms and reactions to treatments. Looking at experiences reported on the Cholangiocarcinoma Foundation discussion board, patients have reported cancer recurrences without CA 19-9 rising, rising CA 19-9 with no recurrence, and of course, sometimes rising CA 19-9 as an early warning signal of progression or recurrence. Hence, CA 19-9 is an imperfect indicator of patient status, but nonetheless it tends to be the number patients watch like a hawk for clues on whether the cancer is stable or progressing.

Medical journal articles indicate that when CA 19-9 rises because a recurrence is afoot in a patient who has had surgery, in some cases the tumor marker can start moving upward as much as 9-10 months before anything is large enough to be seen on a scan. Doctors cannot treat what they cannot see, so it is generally wait and see until a recurrence reveals itself. Or maybe with time the conclusion will be this irritating number is moving around for some non-cancer reason. What is most important is to work cooperatively with your doctors as they monitor your status, so that any changes can be quickly caught and addressed.

Q: My father has cholangiocarcinoma, and we are thinking that he should have a biopsy so that he can be tested for genomic mutations. Is this always recommended?

A: Cancer research is evolving rapidly, and today there are many treatments in development that target specific genomic mutations. Up to about 40% of cholangiocarcinoma patients are estimated to have at least one treatable genomic defect. For this reason, it is generally recommended that cholangiocarcinoma patients have this testing.

There are some exceptions, where having a biopsy for testing would not be recommended.

1. The needed biopsy has a few downsides. Having a biopsy is not recommended if you are a surgical candidate because the doctors can take the sample tissue in conjunction with the surgery. A biopsy can disqualify patients from consideration for a liver transplant. The concern here is that there is a small risk of seeding cancer cells along the needle track. Patients in these cases should consult with their surgeons and other medical team members about the possible benefits of using tissue obtained through resection or transplant for genomic profiling.

2. Some patients might be considered too frail for this procedure. The doctor will look at the health status of the patient, as well as the location of the tumor or met, its size and accessibility, in considering the merits and potential for success (or not) of a potential biopsy in yielding a sufficient sample for the testing. If a patient is in very poor health, a doctor may feel that he or she is unlikely to qualify for clinical trials (the main option at the moment for

targeted treatments) and may question the wisdom of putting that patient through a biopsy procedure. If the patient is very frail, doctors may feel that the time it takes to receive the test results would be too slow if a patient needs urgent treatment.

Notwithstanding the above, if as a patient or caregiver, you wish to open the door toward finding and potentially benefiting from the emerging targeted treatments, it is important to have informed conversations with your medical team about the benefits you see from pursuing genomic/molecular testing, to see if objections to the testing can be overcome. Some of the concerns may be less relevant in the future as liquid biopsy techniques (through blood testing) evolve.

Q: How do I go about seeking a second opinion from a major cancer hospital? I am worried this will offend my local oncologist.

A: Many patients pursue second opinions. There are several ways to go about this. You absolutely should raise the idea with your current doctor and enlist his or her help. A second opinion is a common practice, particularly with a complex diagnosis, so your doctor should be supportive and helpful. Some reasons patients pursue second opinions include: to confirm the initial diagnosis, to be certain there are no options for surgery or transplant when the cancer is locally confined, and to investigate if there are other treatment options.

One approach is to ask around including with your current doctor for a referral to another cholangiocarcinoma expert, and approach that doctor's office directly. Or, on the websites for many of the major cancer hospital centers, you can find instructions for requesting a remote second opinion. For a remote second opinion, you will be given instructions on how to send all the medical records and scans, and the hospital will organize a review for you. Be sure it is a hospital with expertise with this rare cancer, not all hospitals have this. On their websites, hospitals offering this option will often post their charges and how long the process will take. Be sure to check your insurance coverage beforehand. For example, my understanding is that some insurers may not cover remote second opinions.

For many cholangiocarcinoma patients, the initial diagnosis is made via a hospital tumor board, which includes a group of doctors in different specialties, so multiple experts are involved reflecting the complexity of this rare cancer. If your diagnosis was not made via a multidisciplinary team (including specialties such as oncologist, radiologist, pathologist, liver specialist – hepatologist), then you may wish to ask your doctor about involving other specialties in looking at your case.

Q: I have been scheduled for surgery and my oncologist is saying it is up to me whether I have chemotherapy or radiation afterwards, that there is not a lot of evidence one way or another if it helps. How do I make this decision?

A: Until recently, the medical profession did not have strong statistical evidence that chemo or radiation given after surgery offered a benefit in delaying or avoiding recurrence of cholangiocarcinoma. There have been, however, a number of small scale studies suggesting benefits in terms of avoiding or delaying cancer recurrence from different post-surgery chemo or chemo/radiation treatments, particularly for patients with high risk features to their cancer.

Hence, the practice of many doctors in the U.S. has been to offer the option of post-surgery treatment to patients.

This past year, however, a new and large study undertaken in Britain showed an important survival benefit for patients treated with an oral chemo – capecitabine – after surgery. The study is referred to as “BILCAP” and its details can be consulted using an internet search. For patients with high risk features (e.g., when cancer is found in the surgical margins (R1) or lymph nodes), an adjuvant treatment also involving radiation may also be recommended.

Q: My doctor is recommending radiation treatment but I am worried that this will cause me to have another cancer down the road. Should I be concerned?

A: There is a small risk of developing a second cancer as a result of treatment with radiation and with some chemotherapies. Thankfully, this is a rare event. Over the more than ten years+ experience of the Cholangiocarcinoma Foundation discussion board, there has been only one patient report posted of a second cancer that seemed linked to a prior cholangiocarcinoma treatment. You can internet-search any treatment name plus “second cancer” and see some numbers on the risk – they will be tiny. Also, if a second cancer develops, usually this occurs some years after treatment, in these rare cases.

If you are concerned about such risks, this is a question to ask when discussing possible treatment. Doctors generally believe that the benefit of cancer treatments vastly outweighs such risks. Doctors are very aware of this concern – it comes up for example when considering the frequency of scans.

Q: My oncologist refuses to speculate on how long I might survive with different treatments for my cholangiocarcinoma. I have googled life expectancy information and the results are depressing. What can I expect?

A: Survival statistics are often misunderstood, and it is simply not possible for a doctor to assign expiration dates to patients. It is best not to focus on survival statistics. Often, they reflect past experience before newer treatments were available. They consider patients with many different characteristics and may not take into account your own situation.

Since numbers are out there, however, here is one way to think about it. Prognoses are usually given as medians, which mean that half will live longer, and some will live much longer than the number of months or years mentioned. Focus on reasons why you could be in the longer half – you are otherwise in good health, or you are getting good treatment, or you are determined and have a good family support system. This advice comes courtesy of an essay written by Harvard paleontologist Dr. Stephen Jay Gould. Dr. Gould was a Harvard paleontologist who was diagnosed with mesothelioma in 1982 and saw the median life expectancy was eight months. In his essay (link given below), Dr. Gould explains very eloquently the hope he saw in this number. He went on to live another 20 years.

Cholangiocarcinoma is a difficult cancer, but the medical science is rapidly evolving and new treatments are coming on line every year. The experiences of patients described in the medical literature and the Foundation’s discussion board shows a wide variation of experiences. Many

patients do outlive any guesses their doctors might make about their survival. There is always a reason for hope.

Here is Dr. Gould's essay:

<http://people.umass.edu/biep540w/pdf/Stephen%20Jay%20Gould.pdf>

Q: I was just diagnosed with cholangiocarcinoma. Should my children have genetic testing to determine if they are at risk of getting this cancer?

A: Cholangiocarcinoma is not considered a disease that is inherited. Below is a site that explains this. It does say that family members may have an increased risk. However, in a case where two family members share an illness, one question would be whether there is a genetic factor or something else that is shared, such as an environmental factor, for example, the CCA cases related to raw fish consumption in Asia.

<https://ghr.nlm.nih.gov/condition/cholangiocarcinoma#inheritance>

Q: I am puzzled why, given my diagnosis is intrahepatic cholangiocarcinoma and the tumor is in my liver, my doctor isn't recommending a liver transplant? Shouldn't this be an option?

A: The experience with liver transplant for intrahepatic CCA has not so far been positive. Recurrence rates tend to be high and overall survival has been disappointing, which is problematic given the scarcity of livers for transplant. For example, there have been studies of transplant outcomes for patients who had been diagnosed with hepatocellular carcinoma and then were discovered after liver transplant to have actually had intrahepatic cholangiocarcinoma. Sadly, a large proportion of these ICC transplant cases experienced recurrence.

In the future, doctors may develop methodologies for identifying subsets of ICC patients who may benefit from a liver transplant, but this is not the case today. This may involve certain patients at a very early stage who undergo a pre-transplant regime of chemo and/or radiation.

Here is an article reviewing the experience with liver transplant for all types of cholangiocarcinoma.

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4598610/>