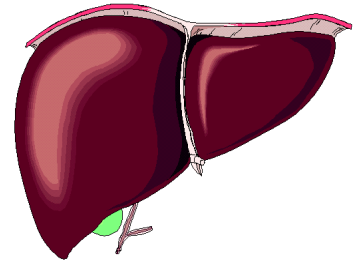


## EVALUATION & LISTING

### *Your Child's Liver Transplant Evaluation*

The University of Michigan is a national leader in liver transplantation, as well as the surgical and medical management of patients with liver disease. In the past 30 years, our physicians and staff have provided care to more than 2,200 adult and pediatric liver transplant recipients. At the University of Michigan Health System (UMHS), state-of-the-art equipment and facilities ensure patients receive the highest quality care. Electronic records are continually updated, meaning patient records are available to caregivers 24 hours a day, seven days a week. This system also allows for the tracking of all outside lab work and diagnostic studies for each patient.



***A Healthy Liver***

### *What is the Liver?*

The liver is an organ that is found in the upper right-hand side of the abdomen under the ribs. It is a dark reddish-brown organ that in an adult weighs about three pounds. It usually has a weight of around one to three percent of total body weight in both children and adults.

The liver has a right and left lobe. Blood enters the liver from the hepatic artery and the hepatic portal vein and leaves the liver from the hepatic vein. The blood from the artery carries most of the oxygen into the liver while the portal vein brings nutrients from the intestine.

The liver is estimated to have about 13 percent of the body's blood supply at any given moment, so the liver is a vascular organ (has a lot of blood vessels in it). The liver also has an extensive network of tubular structures called the biliary tree (bile ducts) that carries bile from the liver to the intestine.

Bile is a substance that carries wastes from the liver and is necessary to breakdown and to absorb fat from the intestine. We call the tubes that carry bile "bile ducts". Bile is made continuously in the liver and flows through the small bile ducts to larger bile ducts and empties into the small bowel. It is bile that helps digest the food in which we eat. Bile is the color of a copper penny or dark green. It is bile that turns our stool brown in color after digestion.

### *What Does the Liver Do?*

The liver has many functions, but it is primarily responsible for:

- Making and storing energy – glycogen and glucose
- Making proteins – some of these proteins help you not bleed from cuts
- Filtering out toxins that can make you sick
- Making bile to help digest foods and absorbing fat and fat-soluble vitamins

Other things that it does include: helps kill germs especially ones that come from the intestine, breaking down proteins, fats, and carbohydrates, and storing Vitamins A, E, D, and K.

### *Why Your Child May Need a Liver Transplant*

The liver can continue its normal jobs even when it has quite a bit of damage. However, when the liver becomes too badly damaged, it cannot grow enough new liver cells to heal itself. When this happens, normal liver tissue is replaced with scarred liver tissue, and the liver cannot continue to do the work it is supposed to do. When most of the normal liver tissue is replaced by scar tissue, the liver is said to be cirrhotic, and a liver transplant may be the only effective treatment. The indication for liver transplant at the University of Michigan Liver Transplant Program is severe, irreversible, acute or chronic liver failure for which there is no other effective medical or surgical therapy. Patients with rare liver-based metabolic defects or liver cancers also may be considered for liver transplant.

### *What Types of Liver Diseases May Lead to a Transplant?*

Some of the most common liver diseases that can lead to liver transplant include:

- Acute liver failure
- Alagille syndrome
- Alpha-1 antitrypsin deficiency
- Autoimmune hepatitis
- Biliary atresia
- Glycogen storage disease
- Hepatoblastoma
- Hepatocellular carcinoma



- Primary sclerosing cholangitis
- Progressive familial intrahepatic cholestasis (PFIC)
- Urea cycle disorders or other metabolic disorders
- Wilson's disease

There are effective medicines for some diseases, particularly if diagnosed early. However, in many cases we only have medicines that treat the complications of liver disease and not the underlying cause. In some individuals, the liver is not failing so all that is needed is treatment of the complications. In other cases, medicines do not solve the problem, and a transplant is required.

Liver transplant is offered to children with a diagnosis that is improved by the procedure. However, it is not a good treatment for some diseases or conditions, such as certain types of infections and cancers that will recur after a transplant. There can be other reasons why a child might not be a candidate for a liver transplant. Because a liver transplant is not perfect (still around a 10 percent death rate), there must be enough benefit from the transplant to justify the potential risk of the surgery. Liver transplant is not a cure that lessens the need for medical care and medicines. Instead, it should be viewed as trading one chronic disease for another. It does offer long-term survival and a good quality of life for the majority of children who have this operation. However, there remains a life-long obligation to take care of the liver which includes clinic visits, blood tests, and daily medications for the rest of a child's life.

## The Referral Process

The referral process can start in many ways for your child. Your local doctor or gastroenterologist may contact the transplant office with your child's clinical history and concerns about his/her liver function. The doctor may feel that it would be best if the transplant team evaluates your child for possible treatment options including liver transplant. There may be situations where your child is in a local hospital and it is felt that they should be transferred directly to the University of Michigan for an urgent evaluation and assessment of treatment options, including liver transplant.

If your child's evaluation will be done in the outpatient clinic, the transplant nurse coordinator will contact you to discuss your child's health history, review the evaluation process with you, and obtain general information. The transplant nurse coordinator will request that medical records be obtained from the referring physician before the transplant evaluation. The records will need to include the following:

- Medical history
- Surgical notes
- X-ray (imaging) reports (i.e., CT scans and ultrasounds of the liver, including copies of the scans on CD-ROM)
- Current and past laboratory results
- Liver biopsy results and slides for review with the pathologist
- Insurance coverage for the evaluation and possible transplant

Once an appointment has been arranged, your referring doctor will be notified of the date and time of your child's evaluation. A letter will be sent to your home that outlines the evaluation process and includes an appointment slip with the date and time of the clinic visit and directions to the Pediatric Transplant Clinic.

*The Purpose of the Liver Transplant Evaluation is to:*

- Introduce you to the members of the University of Michigan Pediatric Liver Transplant Team and confirm the diagnosis of the liver disease including a review of previous liver biopsies and radiological studies. In most cases, the referring doctor already has made the diagnosis. However, all the information will need to be reviewed to make sure that the Pediatric Liver Transplant Team understands each child's medical condition as completely as possible and is in a position to make the best possible recommendation for each child.
- Determine how severe the liver disease is in order to select the best options for management, which may include liver transplant. If liver transplant is an option, this evaluation will help the Pediatric Liver Transplant Team determine the urgency of the need for transplant.
- Inform you and your family about the liver disease, the transplant operation, risks and benefits of both a liver transplant and the medications used following transplant, and other possible ways to treat the liver disease.

The patient evaluation requires a full morning visit and includes several tests to gather information the team needs to make a decision about liver transplant. It may be necessary for your child to undergo testing in the afternoon or on additional days, as determined after the evaluation.

## Tests That May Be Required as Part of the Evaluation

Among the tests that may be required are the following:

- Blood tests, including viral studies
- Imaging studies – x-rays, ultrasounds, or CT scan/MRI
- A liver biopsy

### Blood Tests

Your child will need to have blood drawn. Blood tests determine the severity of your child's liver disease and the presence or absence of infections such as Hepatitis B or C or HIV (AIDS virus). These tests may include:

- **Liver Enzymes** – Elevated levels of liver enzymes can alert physicians to liver damage or injury, since a damaged or injured liver leaks enzymes into the blood stream.
- **Bilirubin** – Bilirubin is produced by the liver and is excreted in the bile into the intestine. High levels of bilirubin often indicate a block in the flow of bile or a defect in the processing of bile by the liver. There are two types of bilirubin – conjugated (direct) and unconjugated (indirect). The conjugated form is the one that is associated with liver disease. However, once we establish that there is liver disease, we usually focus on the total bilirubin.
- **Albumin, Total Protein and Globulin** (protein tests) – Below-normal levels of proteins made by the liver are found in many chronic liver disorders.
- **Clotting Studies**, such as international normalized ratio (INR) and prothrombin time (PT) – These are tests that measure the time it takes for blood to clot and are often used before a liver transplant. Blood clotting requires vitamin K and proteins made by the liver. Liver cell damage and bile obstruction can interfere with proper blood clotting.

### *Your Child's Blood Type*

When receiving a transfusion, the blood received must be a compatible type with your child's type of blood, or an allergic reaction will occur. The same allergic reaction will occur if the blood contained within a donor organ enters your child's body during a transplant. Allergic reactions can be avoided by matching the blood types of your child and the donor. We need to document your child's blood type twice in order to list for a liver transplant.

### *Viral Studies*

These tests determine if your child has antibodies (evidence that they had a previous infection or vaccination) to viruses, such as cytomegalovirus (CMV), Epstein-Barr virus (EBV), Herpes virus, and varicella (chickenpox). Cytomegalovirus may become active when you are exposed to immunosuppressive medications and may increase the chance of infection after transplant.

### **MRI, CT and Ultrasound**

Your child may get an MRI, CT scan, or an ultrasound as part of the evaluation.

The **MRI (magnetic resonance imaging)** is a painless test that uses radio waves to take pictures of the liver. The MRI helps the transplant team examine the liver's size, shape, position, and blood supply. Because your child needs to remain still for this study, it may be necessary for your child to be sedated by a doctor or nurse specialized in sedating children.

A **CT (computed tomography)** scan uses X-rays to take pictures of your child's body. It can be used to take pictures of many different parts of the body. The machine takes pictures of very small sections of the body part being scanned. The child will lie on a table that moves through the middle of a "donut-hole" in the CT machine. Nothing will hurt or touch your child during the scan. Because your child needs to remain still for this study, it may be necessary for your child to be sedated by a doctor or nurse specialized in sedating children.

An **ultrasound** takes pictures of the liver using sound waves but is less detailed than an MRI or CT scan. It, also, has a feature called Doppler (like what police use in their radar or weathermen use) that allows us to look at blood flow in the liver blood vessels.

### **Liver Biopsy**

Your child also may need to have a liver biopsy. The liver biopsy is an important procedure that may be used during the evaluation of your child for liver transplant and during your child's post-transplant care.

The liver biopsy helps doctors determine the severity of liver disease by looking at the cells of the liver. It also can be used to help the Pediatric Liver Transplant Team determine the reason for an increase in liver enzymes.

A liver biopsy is generally performed in the C. S. Mott Children's Hospital Operating Room Suite. The child receives medications to help decrease the pain and also to help the child not remember the procedure. We generally perform an ultrasound to find a safe spot to do the liver biopsy. Once the child is appropriately sedated, the gastroenterologist (liver doctor) will

clean the area so that the risk for infection is low. Then, the gastroenterologist will stick a small needle into the right side of the abdomen in the area where the liver is located. When the needle is withdrawn, there will be a tiny piece of the liver (no larger than the size of a pen refill) inside the needle. This will be sent to the laboratory for us to examine under a microscope. This procedure usually takes less than five minutes once a child is sedated. With any procedure, there are associated risks. Your gastroenterologist will review these risks with you before performing the biopsy. Following the liver biopsy, your child will need to stay for a minimum of four hours. If you live far away from the hospital, or your child is under two years of age, your child will need to stay at the hospital overnight for observation.

#### You Should Know

No patient will be put on the liver transplant list until all of the necessary testing has been completed and reviewed by the Pediatric Transplant Team and confirmed that a liver transplant is the best option for your child.

### Inpatient Transplant Evaluation

There are times when a child is admitted to the hospital and his/her entire evaluation is completed while as an inpatient. Your child will complete the same standard testing that would normally be done in the outpatient setting. This is typically done when a child is acutely ill and would need a transplant within the next few days to weeks after evaluation.

