When a child has liver problems,

the Johns Hopkins Pediatric Liver Center can provide the care and support that the child—and the family—needs. In some instances, a transplant may be necessary. The team at the Pediatric Liver Center works closely with the Johns Hopkins Comprehensive Transplant Center to manage the transplant process and long-term care of these children.

As the only option for pediatric liver transplants in the state of Maryland, Johns Hopkins provides crucial services and support to our patients and their families. We have been performing pediatric liver transplants for over 25 years and have transplanted livers into all age groups, from infants as small as 5 pounds to adolescents who participate in high school activities. We have been a leader in pediatric living-related liver donation and recently reported a 100 percent five-year survival in our children who undergo this operation.

Who needs a transplant?
Liver transplantation is usually recommended to a child with acute or chronic liver disease when medications and routine surgeries have not helped and when the likelihood of survival without transplant is expected to be lower than the likelihood of survival with a transplant. The procedure may be appropriate for children with certain conditions, including biliary atresia, autoimmune hepatitis, viral hepatitis, primary sclerosing cholangitis, acute liver failure and tumors of the liver.

How successful is transplantation?
Currently, approximately 90 percent of all patients receiving liver transplants survive one year or more, and 85 percent live five years or more. Most patients return to school or work and lead active lives. New therapies, our ability to learn about the body’s response to donor organs and our capability to recognize the early signs of rejection all brighten the outlook for patients. Our transplant physicians are highly skilled and aggressive in their pursuit of innovations to improve outcomes.

How will my child be evaluated for transplant?
The evaluation period for a pediatric liver transplant can be very stressful on a family and very time-consuming, usually involving a few outpatient visits. Johns Hopkins tries to ease this process with dedicated staff members who assist families every step of the way. If a child is acutely ill with liver disease, the process can occur more quickly. Parents and children should feel free to discuss any concerns or questions with members of the transplant team.

What should I expect at our first appointment?
First, we’ll evaluate your child’s past and present medical status. Next, we’ll perform a physical examination and blood tests, and we’ll plan the rest of the workup, including electrocardiogram, chest X-ray, ultrasound and CAT scan of the liver. Other tests may be included depending on your child’s condition. A psychosocial evaluation will also be conducted. Don’t forget to bring your child’s:
- Original Social Security card
- Medical and vaccination records
- Insurance information
- Name and phone number of referring doctor/primary care physician

Liver transplant surgeon Andrew Cameron with his young patient, Isabella, and organ-donor dad, Stephen Sindler. Through Johns Hopkins’ living donor transplant program, Stephen successfully donated a portion of his liver to Isabella.
Who are the members of my child’s liver transplant team?

Pediatric Hepatologists
These physicians manage your child’s care before transplant surgery, in coordination with the transplant surgeon after the transplant and after discharge from the hospital.

Transplant Surgeons
These physicians perform your child’s liver transplant operation and oversee your child’s care in the hospital following the surgery.

Anesthesiologists
These doctors administer anesthetic medicine to your child and monitor him or her during the transplant surgery.

Transplant Coordinator/ Nurse Practitioner
This person arranges your child’s care before and after transplant, helps answer your questions, and provides support for you and your family.

Pharmacy Clinical Specialist
This pharmacist, who has specialized training with transplant-related medicines, arranges for your child’s medicines and provides education about them.

Nutritionist
As many children with liver disease are not able to maintain their weight, a nutritionist evaluates your child’s diet and growth before and after transplantation.

Social Workers
Social workers help patients and families with the pretransplant waiting time, the hospital stay, discharge and care after your child returns home. Along the way, you will also meet many other people who will provide care for your child and your family, including critical care physicians, critical care and general pediatric nurses, physical and occupational therapists, child life specialists, and chaplains. Taken together, these individuals will help you and your family get through this stressful time.

What makes someone a good candidate for a liver transplant?
We look for motivation, adherence to previous medical therapies and strong family support. We also discuss financial planning and other arrangements. When the workup is complete, a letter of medical necessity and all medical information is sent to your insurance provider to request authorization to place your child on the United Network for Organ Sharing, or UNOS, waiting list.

After reviewing all of the information, we will recommend one of the following:
• Provide medication and/or standard surgical treatment.
• Accept your child into the transplant program but defer activation until necessary.
• Accept your child into the transplant program and actively seek a donor organ.

This will likely be a stressful time for your child, and he or she may share your emotions but show them in different ways. At the same time, your child may not feel well, and this will add to the fears and frustrations. The pediatric transplant team tries hard to ease the way and reduce stress as much as possible.

Types of Liver Transplants
For a child to receive a new liver, the donated organ must be the correct size to fit into the child’s abdomen. Because of this size restriction, there are several variations of pediatric liver transplants: whole-liver transplant, reduced-size transplant and living-donor transplant.

Whole-Liver Transplantation
A whole-liver transplant occurs when an entire liver is transplanted from one individual to another. The donated liver most likely comes from a child donor or small adult. This is the traditional way pediatric liver transplants are managed, but it means that critically ill children may spend a long time waiting for an organ that’s the proper size to become available.

Reduced-Size Liver Transplantation
If an adult liver becomes available and is a match for a pediatric patient, a segment of the liver may be used. This procedure has significantly lessened the waiting time for small children and infants, as more organ options are accessible. Using liver segments also means that the same donor organ can be used on multiple pediatric patients.

Recommendation
We strongly encourage you to keep your child current on his immunizations.

Live-Donor Transplantation
Healthy adults, most often relatives, may donate a segment of their own liver to a pediatric patient. In both the adult donor and the pediatric recipient, the liver will regenerate over time to form a completely whole liver. If a matching adult is identified, the child will no longer need to wait for an organ to become available, and the surgery can be scheduled.

Johns Hopkins has been a leader in this type of donation and has reported a 100 percent five-year survival rate. While the risk to the donor is minimal, live liver donation is a major surgery. Donors can expect to stay in the hospital for six to eight days, with a full recovery period of up to eight weeks. Despite this hardship, in two decades of performing this surgery, all donors have said they would repeat this experience.

How can I help my child?
This will likely be a stressful time for your child and your family, and children share their feelings in a variety of ways. Keep your child’s lifestyle and daily activities as normal as possible, and maintain your usual standards of behavior.

• Let your child know it’s okay to talk about his or her thoughts and feelings.
• Explain that he or she will be going to the hospital to get help for an illness. For preschoolers, begin this discussion the day before or the morning of your visit. For school-age children or adolescents, begin as soon as they hear the subject discussed in your doctor’s office.
• Avoid overprotecting your child or keeping important information from him or her.
• Make a list of questions to ask and things to take to the hospital, such as a favorite toy.
• Take pictures of friends, family or pets to enjoy after the operation.
• Assure your child that you will be there when the child goes in for the operation and when the operation is over.

What should we do while awaiting a donor liver?
Once your child has been officially listed with UNOS as a candidate for a liver transplant, a waiting period follows until a suitable donor becomes available. The waiting time varies for each child based on organ availability, and the Pediatric End-Stage Liver Disease (PELD) score for children under 12 or the Model for End-Stage Liver Disease (MELD) score for individuals 12 and older. The PELD/MELD score prioritizes the waitlist such that higher scores typically correspond with more severe liver disease, allowing for sicker patients to be ranked higher on the waitlist. As a parent, this waiting time may be a difficult step in the process. Parents often feel a sense of helplessness and loss of control. The pediatric transplant team will do whatever it can to decrease your anxiety and allow you to continue your normal activities.

You must be ready to come to the hospital at any time and be there within three to four hours. Please make sure our pediatric liver transplant coordinator can reach you at all times by providing cell, school, work, family and vacation telephone numbers. Let us know if your phone number or address changes. If we can’t contact you, the liver will be given to another patient.

Meanwhile, we strongly encourage you to keep your child current on his or her immunizations. If your child’s medical status changes, or if your child is admitted to another hospital, please call us. Have a plan for family or friends to take care of other children and siblings during the transplant.

Waiting for a donor liver, which can range from days to months, can be difficult for you and your child. The emotions patients and families experience include:
• Hope: Help is available for my child.
• Fear: Will my child die before a donor organ is found?
• Anticipation: Is that the hospital on the phone? Did my cellphone drop a call?
• Frustration: Why is this taking so long?
• Guilt: Another child must die before mine has a chance to live.

What do I tell my child?
You may want to talk to your child about what he or she wants other children to know. One option is simply to say that he or she is waiting to have an operation to fix his or her liver.

You can help your child handle questions like:
Where will my new liver come from?
Most likely, it will come from a person who dies in a car accident.

What will happen to my old liver?
Doctors will examine it to determine why it made your child sick, which can help other children.

Will the operation hurt?
Tell your child that he will be given special medicine that will allow him to be in a deep sleep so he cannot see, hear or feel anything, not even pain, during the operation, and that medicine will be given so that your child is comfortable after the procedure.

Could I die?
Even grown-ups think about dying when they have an operation. Your child is having this operation because doctors and nurses feel it gives him the best chance to live a normal life.

Will I be the same person after the operation?
Yes, and with time, your child will feel stronger and healthier because the new liver will give him more energy.

Will I be afraid?
When children are afraid, they need information to reassure them. Ask them what they are afraid of. If you don’t know the answer, ask a member of the transplant team to assist you.

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While we wait, what do I tell friends and teachers?
You may wish to discuss your plans with friends, your child’s friends, teachers and classmates. Let your child’s teacher know how long your child will be out of school and that sending homework to the hospital will help him or her feel a part of things, making returning to school easier. Among friends, some will show genuine caring, while others may ask intrusive questions or smother your child with attention. Many families let friends know how they can be helpful by:
• Caring for your other children, ideally in your home
• Bringing your other children to visit you at the hospital
• Planning special treats for your other children, who may be feeling lonely or worried, even jealous
• Bringing you and your family home-cooked meals while you are at the hospital and when you return home

Did you know
Johns Hopkins Pediatric Liver Center doctors are actively researching ways to improve transplant outcomes including better ways to detect infections and decrease liver scarring.

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What happens when a liver is found?

When a donor liver is located, you will be telephoned and given instructions on what to do. Once you get the call, do not give your child anything else to eat or drink. When you arrive at the hospital, go directly to the pediatric emergency room for surgery prep unless notified to go directly to a hospital floor. Blood work will be obtained, and an IV will be started to maintain hydration. Many doctors and nurses will come by to find out how your child is doing and to discuss the operation. This discussion will include obtaining your formal consent for the surgery once you have reviewed the risks, benefits and alternatives to the operation.

Also, please bring a list of your child’s medicines and drug allergies and health insurance card to the hospital.

As you wait for your child to be transported to the operating room, you can talk to him or her to help prepare for the exciting days ahead. Reassure your child that you will be there when he or she wakes up. You can let your child know that he or she will wake up in a hospital room with many nurses and doctors and will feel quite sore at first. Your child will see bright lights and bags that drip medicine into his or her body, and he or she may feel a breathing tube in the throat for a few hours, or perhaps longer if the child has had previous abdominal surgeries. The transplant coordinator, operating room nurse or child life specialist will update you and give instructions on what to do. Once you get to the operating room, you will be notified to go directly to a hospital floor. Blood work will be obtained, and an IV will be started to maintain hydration. Many doctors and nurses will come by to find out how your child is doing and to discuss the operation.

What happens after the operation?

Following the surgery, your child will go to the pediatric intensive care unit (PICU) at the Children’s Center for postoperative care and monitoring. Here’s what to expect:

- A breathing tube may be inserted into your child’s mouth or nose and down the windpipe and lungs to assist with breathing. Your child will be unable to talk while this tube is in place. Once the tube is removed, usually within one to two days, your child will receive oxygen from a face mask or nasal cannula. We will encourage your child to take deep breaths and to cough to bring up any mucus in the lungs.
- A feeding tube, or nasogastric (NG) tube, which is passed through the nose, down the throat and into the stomach, will remain in place for a few days. This tube helps prevent nausea and vomiting, since the stomach and intestines don’t work as well after a major surgery.
- A large dressing will cover the abdominal incisions. Abdominal tubes will drain excess blood and fluid, and they will be removed after the drainage subsides, usually within one to three days.
- In some instances, typically if the new liver is slightly larger for the abdominal cavity, the surgeons will delay closure of the muscle and skin for a few days while the abdominal cavity stretches to accommodate the larger size. In this case, the surgeons will place a sterile dressing over the abdomen so that your child doesn’t get an infection or lose extra fluid.
- Two or three intravenous (IV) lines and an arterial line will be in place to administer fluids and medications and to monitor blood pressure and blood oxygen content. These lines also allow doctors and nurses to draw blood to monitor drug levels and blood counts.
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Did you know

We recently published the results of our living-related liver program, showing 5-year survival at 100%.

Are there other things I should know following surgery?

Your child must take a combination of immunosuppressive medications, which like all drugs have side effects. We will monitor your child’s medications to achieve the best therapeutic effect with the least amount of side effects. With organ transplants, there are always risks of complications, but we will do everything we can to minimize these risks, which can include:

Infection

The immunosuppressive medications your child will take to prevent rejection suppress the immune system, the body’s natural defense against foreign substances. But these medications also decrease the body’s ability to fight infection.

To reduce your child’s risk of infection while in the hospital:
- • All family members will follow strict hand-washing procedures and, in some cases, wear protective masks.
- • Visitors will be limited.
- • Your child will wear a mask when well enough to leave his or her room.

Rejection

The body’s rejection of a new organ is a lifelong concern. Nearly every transplant patient experiences episodes of rejection, which are best detected by blood test measurements and a liver biopsy. Depending upon the degree of rejection, we may adjust the antirejection medication, which may require hospitalization. Other studies, such as abdominal ultrasounds, may be necessary.
Surgical and anatomic complications

Sometimes there can be a problem in one of the blood vessels that the surgeon connects between the new liver and your child’s body, or in the way that the bile drains from the liver. For example, although rare, a clot can form in the major vessels supplying blood to the liver, or there can be blockages in the flow of bile from the liver. For this reason, your child will be closely followed after the procedure and will likely need frequent blood tests and ultrasounds.

When may my child return to school?

Your child should be ready to go back to school about 12 weeks after discharge. Plan to meet with his or her teacher ahead of time to discuss your child’s medical needs, including the:

- Medication schedule
- Follow-up doctor appointments and biopsies
- Possible physical changes related to medications, like weight gain and puffy face

This may be a particularly challenging time for your child. Consider asking your child’s teacher, school nurse or psychologist to plan a group discussion with your child on his or her first day back in school. Prepare your child for questions such as:

- What was it like in the hospital?
- Was the surgery painful?
- Were you scared?
- Do you feel different now?

How should I prepare my child for return hospital visits?

Reassure your child that all people who have had transplants need to return to the hospital for checkups.

Talk about:

- What has helped him or her through challenging procedures in the past
- What he or she would like to do this time during the procedure
- What he or she would like to do with you after the procedure

When should I call you?

Naturally, you may feel very anxious in the weeks and months following surgery. You should call 911 in an emergency. You should call us if your child has any of these signs or symptoms:

- Burning sensation on urinating
- Change in activity level
- Cold or flu-like symptoms
- Cuts or scratches that do not heal
- Diarrhea
- Fever over 100 degrees Fahrenheit
- Poor appetite
- Shortness of breath
- Tiredness or fatigue
- Vomiting
- Any new or concerning symptoms

What about the future?

As your child grows, you may have other questions or concerns. We are happy to be a resource for you whenever the need arises. Periodically, we will want to re-examine your child and assess his or her progress to help improve transplant care. Our goal post-transplant is to help make your child’s life as normal as possible.

“During this trying time we could not have asked for better care. Abby was treated like a VIP from start to finish and that treatment has continued post-transplant.”

— Mother of pediatric transplant patient, Abby Bahr
Making an Appointment

For Patients and Families
Patients and families may make appointments with the Pediatric Liver Transplant Program by calling 410-955-8769. International patients should contact Johns Hopkins International Patient Services at 410-955-8032.

For Referring Physicians
The Pediatric Liver Transplant Program is committed to continually improving our partnership of care and communication with our health care colleagues. To refer a patient or to consult about a patient’s care, we encourage you to call the Hopkins Access Line, or HAL, a 24-hour hotline for physicians, at 1-800-765-5447 or 410-955-9444.

Pediatric Transport Services
The pediatric intensive care unit transport team travels by ambulance or helicopter to community hospitals across Maryland and some regions of Delaware, the District of Columbia, West Virginia and Pennsylvania to stabilize and transport critically ill patients to the Johns Hopkins Children’s Center, where they can receive a higher degree of specialized care. For critical care consultation or to request transport, call 410-955-5260 or 1-800-999-PICU (7428).

www.hopkinschildrens.org