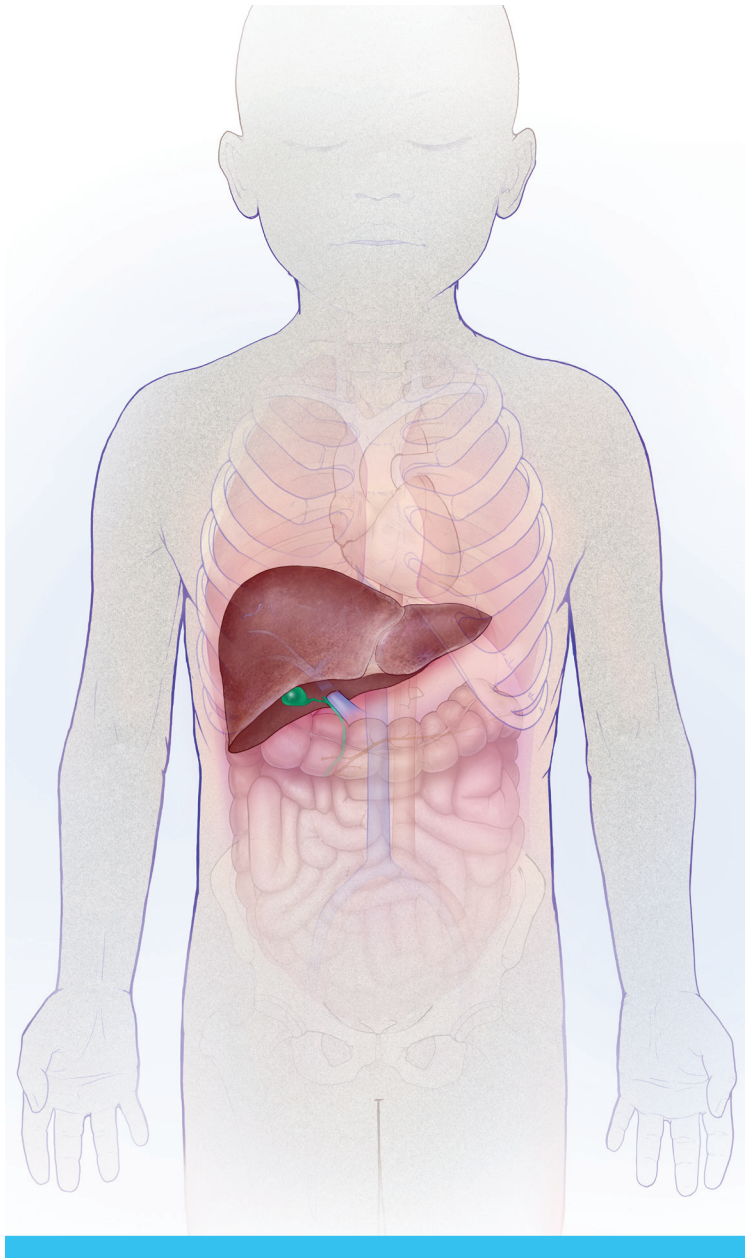


LIVER TRANSPLANT EDUCATION BOOKLET

Welcome to Children's
Hospital of Philadelphia

LIVER TRANSPLANT PROGRAM



This booklet was created to help you understand the liver transplant process at Children's Hospital of Philadelphia (CHOP). You will learn information about:

- Evaluation process for liver transplant
- Transplant surgery
- Recovery period
- Follow up
- Possible complications

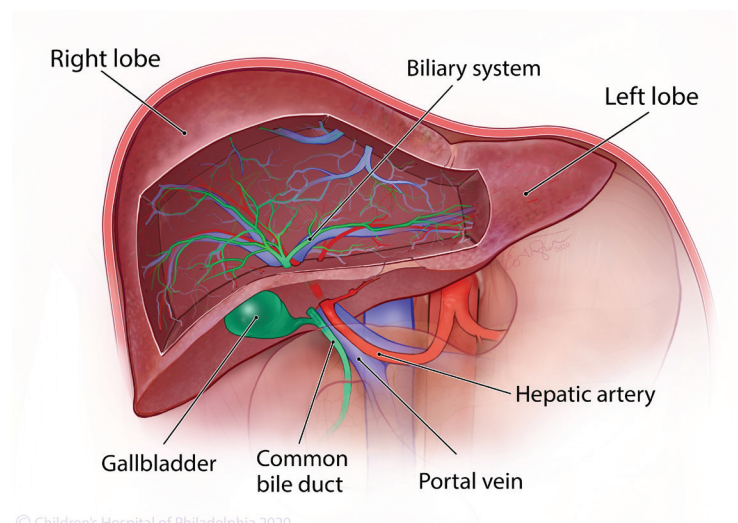
What does the liver do?

The liver is located in the upper right part of the abdomen (belly). It is the largest organ in the body. These are its functions:

- Changes things, like food and medicine, into forms that can be used to keep the body growing and healthy
- Makes proteins needed for normal blood clotting and growth
- Removes waste products from the blood
- Stores and releases sugar to keep blood sugar levels stable
- Helps to digest food
- Keeps blood flowing in the abdomen
- Fights infections in the intestines (gut)

The liver breaks down the food we eat and changes the nutrients into energy for the body. If the liver does not process nutrients, it can lead to poor growth and low blood sugar.

The liver builds proteins that help the blood to clot. These are called clotting factors. If the clotting factors do not function, this can lead to bruising and an increase risk for bleeding.



The liver makes a digestive juice called bile. Bile dissolves fats, certain vitamins and food that is absorbed by the gut. When the liver removes waste products from the blood stream, they go into the bile. These products leave the body through bowel movements. If the liver is not cleaning the blood or not making bile, waste products will build up in the blood. This can cause the skin and eyes to turn yellow. If there is not enough bile in the gut, this can lead to loose and/or pale-colored bowel movements and poor growth.

When the liver does not function, medical care may be needed. Medication and treatments can improve liver function. If these do not work, a liver transplant may be the next step. During the transplant evaluation, our team will discuss your child's medical history with you.

The Transplant Team

You will get to know many healthcare professionals during the transplant process. All team members receive training in pediatric transplant and are available to answer your questions and help you make decisions about your child's care. They are also here to provide emotional support during a difficult time. You may find it helpful to keep a written list of your questions and concerns. This team will care for your child in both the inpatient and outpatient settings.

Physicians

Together, these physicians will manage your child's medical care.

Transplant Hepatologist: A pediatrician with advanced training in gastroenterology (GI) disorders who specializes in liver disease and transplantation.

Transplant Surgeon: Performs the transplant operation.

Nursing Staff

This staff includes registered nurses (RNs) and nurse practitioners (NPs). Together, these nurses will provide care throughout the transplant process.

Inpatient Nursing Staff: These staff members provide the day-to-day bedside care when your child is admitted to the hospital. The nurses will provide education regarding medications and medical procedures. Each day, this team will outline the plan of care for your child.

Nurse Practitioner: Can prescribe medications and make medical decisions.

Anesthesiologist

The anesthesiologist is a doctor who puts your child to sleep for surgery. For the transplant surgery, your child will be placed on a breathing machine. Your child will not be aware of or have a memory of the surgery.

Child Life Specialist

The child life specialist can help your child and family cope with the hospital experience through play and age-appropriate activities. The child life specialist can prepare your child for transplant surgery and medical procedures.

Financial Advisor

The financial advisor works with your insurance company to ensure that your child's transplant surgery is covered. The financial advisor can answer questions and provide counseling regarding healthcare costs.

Infectious Disease Staff

The infectious disease team reviews your child's immunization history and risk factors. Age-appropriate immunizations are required before transplant. This team will make recommendations if additional immunizations are needed.

Pharmacist

The pharmacist is an expert in transplant medications. The pharmacist will provide education for the medication your child is prescribed and will help you create a medication plan.

Psychologist

The psychologist assesses the ability of you and your child to cope with the transplant process. The psychologist will help you and your child develop coping strategies.

Registered Dietitian/Nutritionist

Because children with liver disease have difficulty eating and absorbing all the calories and nutrients needed for growth and development, the dietitian works with you and the medical team to develop a diet for your child.

Social Worker

The social worker will provide emotional and psychosocial support for you, your child and your family. The social worker may assist you with insurance issues, housing, transportation and community resources.

Transplant Coordinator

The coordinator is responsible for organizing the hospital admission when a liver is available. The coordinator will walk you through the step-by-step process.

THE TRANSPLANT PROCESS

There are many steps in the transplant process: evaluation, waitlist, waiting for transplant, transplant surgery, hospital stay and care at home after surgery. The following sections will provide you with information on each step of the transplant process.

The Transplant Evaluation

Your child will go through an evaluation process before being placed on the transplant waiting list. You and your child will learn about liver transplant and meet members of the transplant team.

The evaluation will take place in the outpatient clinic at CHOP. It is coordinated through the liver transplant office. During this evaluation, the transplant team will review your child's medical and surgical history.

Your child will need blood tests, imaging (including ultrasounds and an echocardiogram of the heart) and a CT scan. Your child will be scheduled for these tests on another day.

After the evaluation, the liver transplant team will meet to discuss your child's medical history. During this discussion, the team will determine if your child would benefit from a liver transplant. A member of the transplant team will call you to discuss their recommendations. During this phone call, you will be given information regarding the next steps of your child's care.

The Waitlist

Your child will be placed on the United Network for Organ Sharing (UNOS) list. UNOS is a private nonprofit organization that is under contract with the federal government. UNOS maintains the list of patients awaiting transplant in the United States. This group is also responsible for distributing organs to individuals on the waitlist.

Your child will be listed according to height, weight, blood type and lab results. A computerized program generates a score that reflects the severity of each person's liver disease. The sicker a child is, the higher the score will be. This score is called a PELD or MELD score.

- PELD (Pediatric End-stage Liver Disease): children under 12
- MELD (Model for End-stage Liver Disease): children 12 and older

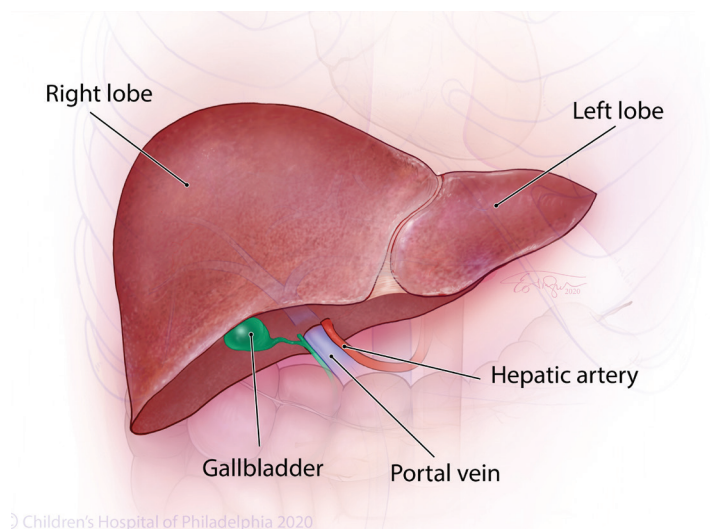
When a liver becomes available, this score determines who will receive the donated liver.

The MELD or PELD score does not always reflect the severity of a child's liver disease. Our team may determine that the transplant is more urgent. In

this case, we are able to request a higher score. This is known as an exception. A board of transplant professionals reviews the request. If approved, the patient's score is increased.

About the Donors

Liver grafts can come from deceased or living donors. The UNOS waitlist is for livers from deceased donors. Deceased donors are brain dead with no chance of recovery from the brain injury. The family decided to donate the organs. The transplanted organs are kept alive using medications and medical technology. The liver transplant team reviews the donor's medical information to determine if the liver is a good match for your child. Livers that come from a deceased donor can be a whole liver from a donor that is a similar size to your child or a piece of the liver from a larger donor. The liver can be split into two segments. A smaller piece of a liver can be used for a small child and the larger portion for an older child or adult.



Liver grafts can come from a living person. The liver can be split into different segments. A portion of the liver remains inside the donor and a portion is used for the liver transplant. To proceed with a living donor transplant, an appropriate donor needs to be identified and evaluated. The living donor does not need to be a blood relative of the recipient. The donor needs to have a compatible blood type, be the right size and be in good health. The living donor undergoes a head-to-toe evaluation by our living donor team to ensure the donation is safe for the donor and that the liver is appropriate for the recipient. The living donation process will be discussed during your transplant education session. It is important to know that if you choose to consider live donor transplant, your child will still be placed on the deceased donor waiting list and will remain eligible for deceased donor organ offers.

Waiting for Transplant

Waiting for a liver can be a difficult time for you and your family. It is impossible to predict when a liver will become available. The wait may be weeks, months or years. You may worry about your child's medical condition and may feel helpless. Waiting can become stressful and frustrating. If you find yourself having difficulty managing these feelings, we have resources available to assist you.

Your child should continue to see your pediatrician for routine visits and receive immunizations as required. The hepatologist and other specialty teams will continue to monitor and care for your child. Maintain contact with the transplant team to let us know of any changes.

While on the waitlist, it is important that the transplant center be able to reach you if a liver becomes available. You will be asked to provide contact phone numbers. It is important to keep your phone with you and on at all times. If your contact information changes, call the liver transplant program. If the coordinator cannot reach you, we may have to turn down a liver offer.

Your child can travel while waiting for a liver transplant if approved by the medical team. Before you leave, contact the transplant team with the destination, schedule and phone numbers.

When a liver is offered for your child, the transplant surgeons and physicians will review the donor's history to determine if the donor is a suitable match. If it is determined that the donor is suitable, you will receive a phone call from the liver transplant coordinator. You will be given instructions about when your child should stop eating and drinking, when to arrive at CHOP, and where to go. You will not be given any specific personal or medical information about the donor.

THE TRANSPLANT SURGERY

When you arrive at the hospital, the transplant coordinator will tell you where to go depending upon the time of day. Once the admission process has been completed, you and your child will be taken to a room on an inpatient unit.

When you arrive on the unit, an intravenous line (IV) will be placed and blood will be drawn. Additional imaging such as a chest X-ray may be required as well as an EKG. Your child may not be able to eat or drink depending on the timing of transplant. Your child will remain hydrated with IV fluids.

Once the liver is determined suitable for transplant, your child will be brought into the Operating Room (OR). The OR team consists of anesthesiologists, surgeons, nurses and technicians. The surgery usually takes six to eight hours. Medication to help your child sleep will be given to start anesthesia. Once your child is asleep, additional IV lines will be inserted. A larger IV line is placed in a large vein in the neck. This is called an internal jugular line. Medications can be given and blood can be drawn through this line after surgery. A Foley catheter will be placed in the bladder to monitor urine output. A blood pressure cuff, EKG leads, a temperature probe and a pulse oximeter will be used to monitor your child's vital signs during surgery.

The surgeon will begin the procedure by cleaning your child's skin. The area being operated on will be protected by drapes. The diseased liver will be removed and sent to the laboratory for examination. The healthy liver will be sewn in place by the surgeons. Your child's gallbladder will be removed with the diseased liver.

During the surgery, you and your family members will wait in the surgical waiting area. The transplant team and operating room nurses will provide updates.



Olivia,
*liver transplant
at 8 months*

AFTER SURGERY

Intensive Care Unit

Your child will be admitted to the Pediatric Intensive Care Unit (PICU) after the transplant surgery. The PICU is a busy place with many alarms, sounds and different types of equipment. When your child arrives in the PICU, there will be a 30- to 60-minute delay before you can see your child. This allows the PICU staff to receive information about the surgery from the OR staff and get your child settled. The staff will place your child on monitors that check heart rate, respiratory rate, blood pressure and oxygen level. Intravenous pumps will be used to give IV fluids and medications. The monitors and pumps have alarms that may sound. The nurses will check your child and the equipment when this happens.

Your child may have an endotracheal (ET) tube coming out of either the nose or mouth. This tube is placed in the trachea (windpipe) during the operation and is connected to a ventilator to breathe for your child during surgery. The ET tube is removed when your child no longer needs help breathing. Sometimes this can be done in the operating room, and sometimes the breathing tube needs to stay in for a little longer. Your child will not be able to talk, eat or drink with the ET tube in place. Once the tube is removed, the nurse will have your child do deep breathing and coughing exercises to keep the lungs clear.

Other tubes and drains will be placed during surgery while your child is asleep in the operating room. A salem sump tube, which passes through the nose and into the stomach, helps drain secretions from the stomach. There will be three Jackson-Pratt (JP) drains in your child's abdomen to drain fluid. The sump and drains will stay in place for several days and help the team monitor for bleeding or bile leaks. A Foley catheter will drain urine from the bladder to measure urine output.

There will be multiple intravenous lines and an arterial line in your child's blood vessels. The arterial line looks like an IV but is placed in an artery in the wrist instead of a vein. It is used to monitor blood pressure and to draw blood samples. The arterial line will be removed before your child is transferred from the PICU to the GI inpatient unit.

Your child will have many studies and tests following the transplant surgery. Your child may have several ultrasounds after transplant to check the blood flow to the liver. It may be repeated throughout the hospital stay. Blood tests are done daily throughout the hospital stay.

Inpatient Unit

When your child is medically stable and does not require ICU-level care, your child will be transferred from the PICU to the GI unit. The nurses will continue to monitor and assess your child. The surroundings will be more relaxed because patients on the inpatient unit no longer need continuous monitoring. This is where your child will continue to recover from surgery and you will receive education about caring for your child after transplant.

Some things you can do to help during the recovery period include:

- *Walking:* This helps your child build strength. Take your child for walks around the room and hallway. Your child will become more active every day.
- *Coughing and deep-breathing exercises:* Your nurse will help your child with these exercises. Your child may use an incentive spirometer, a plastic container that helps your child take deeper breaths.
- *Eating:* When allowed to eat, your child will advance slowly from clear liquids to solid foods. The body needs many calories to heal and grow. If unable to eat enough to provide the necessary nutrition, your child may be fed through a nasogastric (NG) tube or the IV. The dietitians will meet with you to help you develop a good diet for your child.
- *Relaxation and play:* Play is an important part of your child's physical and emotional recovery. The child life specialist will help your child find toys and games to play with during the recovery period.

During hospitalization, the liver transplant team will visit your child every day. Initially, we will see your child several times per day. As recovery progresses, your child will be seen by our team daily. During this time, we will review your child's current condition and the plan for the day. As your child's clinical condition changes, the plan for the day may change. The bedside nurse and our team will work together to keep you informed of changes. If you have any questions about your child's care during your hospital stay, ask to speak to a member of our team.

Your child will be ready for discharge when they are on a stable medication regimen, have improved liver function and are tolerating a regular diet. Prior to sending your child home, we provide education on new medications, reasons to call our office and our follow-up plan. Members of the transplant team including the pharmacist, dietitian, nurses and nurse practitioners will complete your education.

AFTER YOU GO HOME

Clinic Visits

Your child's care does not end after discharge from the hospital. You and your child will need to come to the Liver Transplant Clinic for regular visits and blood work. This is a good time to ask questions about your child's health and medications. Your child will be seen weekly for the first month after the transplant and then every other week for one month. The number of clinic visits may decrease, depending on how your child is doing. Your child may need blood tests in between clinic visits. These can be done either at the CHOP lab or at a lab close to your home.

The transplant team will review your child's lab results after clinic. A nurse or nurse practitioner will contact you to let you know if your child needs any medication changes or more blood tests.

Physical Activity

Your child may return to regular activities four to six weeks after liver transplant. Your child may have restrictions at first to allow the incision to heal. The staples at the incision site are typically removed two to three weeks after the surgery. After the incision has healed, your child can participate in gym class, ride a bike, go skateboarding and so forth. Introduce exercise slowly to gradually build up strength. Your child may need rest periods. Many parents say that their child has more energy after transplant than before transplant. This is because the child now has a healthy liver.

Nutrition

Nutrition plays an important role in the recovery of your child after transplant. Before transplant, some children have a hard time eating enough calories and nutrients due to liver disease. This may result in a longer time period before children are in the best state of health. Sometimes children are not able to take in enough calories even after transplant and need other kinds of nutrition. Dietary supplements, such as high-calorie drinks or NG tube feeds, may be used to provide extra nutrition. A well-balanced diet with enough calories and a multivitamin are all your child needs. Other vitamins and minerals may also be prescribed. These may include vitamin D, calcium, magnesium and iron.

Your child's eating habits may change after the transplant. Intake and appetite may be better or worse. This happens for many reasons. It is important to know that every child is different and your child should not be forced to change habits. The dietitian can teach you about good food choices for your child. After transplant, most children will need a high-calorie diet rich in protein and fat as well as carbohydrates to promote healing.

Herbal supplements, natural food supplements and home remedies are popular. Do not give these to your child without checking with the transplant team. Some of these supplements interfere with the immunosuppressive medications and may stop them from working.

Regular Pediatric Care

All liver transplant recipients must continue care with their primary care provider. Your child will continue to need routine well childcare and ongoing care for general health concerns. Our team will work closely with you and your child's healthcare provider to determine which concerns are best to evaluate in the primary care office versus by the liver transplant team.

Liver transplant recipients should continue to receive their immunizations in the primary care setting. Timing of the vaccines will vary based on the patient's medication regimen. The liver transplant team will work with you and your child's primary care provider to determine the most appropriate timing for both inactivated and live virus vaccines.

Your child should receive the influenza (flu) vaccine every year. It is important for all household members to receive the flu vaccine every year as well.



MEDICATIONS AFTER TRANSPLANT

After transplant, it is important for you and your child to understand why and how each medication is given. Your child will be taking many different medications in the hospital. Most of these medications will still be needed at home after discharge. When your child no longer needs a specific medication, the transplant team will stop it. Do not stop any medications or change any doses without discussing with the transplant team first.

Before discharge, the nursing staff will help you schedule your child's medication around home and school activities. It is important that you mention any special situations at home or school that may affect the medication schedule. Medications need to be given at the same time every day. Many times, a school nurse will be available and can give your child medication during school hours.

Medications after transplant include:

- *Anti-rejection/immunosuppressive medications:* Immunosuppressive medications are used to prevent rejection of the transplanted liver. These medications are very important and should be taken exactly as directed. Immunosuppressive medication will likely be required for the rest of your child's life.
- *Anti-infection medications:* Medications to prevent infection will be given immediately post-transplant while your child is receiving multiple immunosuppressive medications. The transplant team will stop these medications after a few months.
- *Anti-coagulant medications:* Mild anti-coagulation medication (such as aspirin) will be given to prevent blood clots from forming in the blood vessels in the liver for up to one year after transplant.

- *Vitamins and supplements:* Your child may be prescribed oral vitamins or supplementation if needed. These may include magnesium, calcium, iron and vitamin D.
- *Acid blockade:* Some medications can irritate the lining of the stomach. An acid-blocking medication is used to protect the stomach from irritation.

COMPLICATIONS AFTER TRANSPLANT

As with most surgeries, there are potential complications following liver transplant. The complications that occur can happen soon after transplant (short-term) or after much time has passed (long-term). Some complications may require additional appointments, testing, medications, procedures, or surgical intervention for evaluation and treatment. In rare cases, there can be complications requiring re-transplant. It is important to know that the liver transplant team works to prevent and monitors closely for complications.

Listed below are a few of the common complications. These complications along with additional complications will be discussed in more detail during your transplant education with the transplant team.

Rejection

The immune system is the body's natural protection against infection. It protects the body from invaders, such as bacteria and viruses, by attacking and destroying them. Rejection occurs because the immune system considers the transplanted liver to be a foreign invader and tries to attack and destroy it. To prevent this, we give your child immunosuppressive, or anti-rejection, medications. These medications make the

immune system less active. Most children have to take immunosuppressive medication for the rest of their life. Rejection can occur even while your child is taking immunosuppressive medication.

Rejection most often occurs 7 to 10 days after the liver transplant. The chance of rejection is less over time, but it may occur at any time after transplantation. Most of the time, rejection is easy to treat by adjusting medications.

Early signs and symptoms of rejection include:

- Increased liver enzymes
- Fever

Later signs and symptoms of rejection include:

- Dark-colored urine
- Clay-colored stools
- Yellow eyes and skin
- Itching
- Continued increased liver enzymes
- Tiredness

If your child has any of the above signs or symptoms, contact the transplant team. This is why it is important to keep scheduled appointments for the Liver Transplant Clinic and lab studies. If rejection is suspected, your child may need a liver biopsy to confirm the rejection and to allow the physicians to see how your child's liver has been affected by rejection.

Most rejection episodes can be treated. Rejection rarely causes irreversible damage to the transplanted liver. Your child may be admitted to the hospital. This depends on the severity of the rejection. We treat rejection with intravenous (IV) steroids and adjustments to the immunosuppressive medications. There are stronger medications we can use if needed. Repeated episodes of rejection can cause permanent liver injury and end-stage liver disease.

Notify the liver transplant team immediately if your child has any signs of rejection.

Infection

Since your child's immune system is being weakened by medication to prevent rejection, your child is at an increased risk for infection. The highest risk for infection is the first six months after transplant while your child is on a higher amount of immunosuppression. Most infections are common childhood illnesses, but there can be illnesses that are more serious. If your child develops symptoms of an infection, such as fever, call the transplant team. It is important to notify the transplant team so an appropriate plan for evaluation can be determined. During the first several months after transplant, if your child has a fever, we will want your child to be evaluated either in the emergency room, by the transplant team or by your child's primary care provider.



LIFE AFTER TRANSPLANT

Returning to School

School-age children return to school approximately four to six weeks after transplant. The transplant team will see your child on a weekly basis and will evaluate readiness to return to school or daycare.

Pets

Typical household pets are OK for liver transplant recipients. It is important to practice good hand hygiene. If you want to have an exotic pet, contact the transplant team to discuss. Some exotic animals and reptiles carry infections that do not affect the animal but could be harmful to your child.

Piercings and Tattoos

Both piercings and tattoos are a personal choice and can be done safely for liver transplant recipients. Because of the immunosuppression medication required to prevent rejection, the immune system is weaker and there is an increased risk of infection. In order to minimize risk for infection, it is important to talk to your team about the timing of tattoos and piercings and then make sure it is being done at a professional shop with licensed professionals.

Transition to Adult Care

There are many different aspects that affect the timing of graduating from CHOP and transitioning to adult care. We will start to prepare you and your child for transitioning to adult care at a young age by providing guidance on transitioning responsibility for medical management in addition to guiding the transition to an adult transplant facility.

LIVER TRANSPLANT CLINIC (NOTES)

CALL THE TRANSPLANT TEAM IF YOUR CHILD HAS:

- A temperature greater than 101.4 F once or a low-grade fever (100° – 101.4° F) twice over an 8-hour period
 - **Do not** give acetaminophen (Tylenol) without contacting the liver transplant team
 - **Do not** give ibuprofen (Motrin) without discussing with liver transplant team.
- Diarrhea or vomiting, because this can affect the level of medication in the blood
- Redness, pain, swelling or pus at the surgical incision
- A cough or cold that will not go away
- Been exposed to a disease (chicken pox, measles, etc.)
- Mouth sores
- Missed several doses of medication(s)
- Vomiting after the medication is given
- Side effects from the medication
- Signs of organ rejection

IMPORTANT PHONE NUMBERS

If you have questions or concerns:

- Monday to Friday, 8 a.m. to 4 p.m.: Call the Liver Transplant Office at **215-590-4281**
- After 4 p.m. and on weekends: Call the hospital operator at **215-590-1000** and ask for the gastroenterology (GI) fellow on call.

Learn more:
[CHOP.EDU/TRANSPLANT](https://chop.edu/transplant)

