

The Liver Transplant Program
A Guide for Families

What to expect: Pre-Operative



The Children's Hospital of Philadelphia®

Hope lives here.

INTRODUCTION

Your child is ill. Your anxiety is high. Your questions are many. The good thing is you've come to the right place — The Children's Hospital of Philadelphia.

You are about to embark on a journey that, although difficult, can have a life-transforming result. Many families have already taken the liver transplant journey and our "alumni" group has grown strong. Children just like yours are now laughing, playing, attending school, spilling their milk and skinning their knees. Their dreams of earning a place on the varsity team, going to the prom and even going to college are now coming true. Their lives have been forever changed for the better thanks to the Liver Transplant Program.

Your family will not make this journey alone. The staff of Children's Hospital will be with you every step of the way. This booklet is designed to give you an idea of what to expect and how we can assist you at every stage of the journey — and beyond.

While your child waits for a liver, Children's Hospital's team will work to keep your child in the best health possible. Throughout this time, the Hospital's world-renowned physicians, nurses, transplant coordinators, social worker and others will answer questions, clarify medical terms and provide you with the information you need to make medical decisions for your child.

At the time of surgery and during the post-operative recovery period, that same team of experts will provide your child with the most advanced treatment available, be there to help you with practical matters such as insurance and do whatever they can to ease your emotional stress.

After recovery, your child will return to Children's Hospital for ongoing care where you can expect the same exemplary commitment to your child's health and the same attention to your family's well-being.

All of this information is explained in greater detail in this guide booklet. Please read it carefully and jot down any questions you might have. You may review your questions with your child's transplant coordinator.

Remember, Children's Hospital's team is here for you and your child. Should you need anything, just ask. No request is too big or too small. No question is too insignificant. And most importantly, no operation is more important than your child's. We know you feel that way — and we do, too.

Introduction	2
The Hospital Staff	3
What Your Liver Does	4
The Evaluation Process	4
Before the Transplant	5
Getting to the Hospital	6
At the Hospital	7
The Operation	7
After the Transplant Operation: The Pediatric Intensive Care Unit	8
After the Transplant Operation: the Recovery Period	9
Rejection	9
Conclusion	10

THE HOSPITAL STAFF

You will get to know many healthcare professionals through the transplant process. They are here to help answer your questions and to help you make decisions regarding your child's healthcare. 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.

Some of the people you will meet include:

Nurse Transplant Coordinator

The transplant coordinator is your main contact. The coordinator organizes the transplant process from pre-transplant evaluation to post-transplant care. The coordinator serves as a link between the transplant team and other healthcare professionals involved in your child's care.

Physicians

The physicians include hepatologists and transplant surgeons. The pediatric hepatologist specializes in diseases of the liver. The transplant surgeon performs the transplant operation.

Nursing Staff

The nursing staff plans and provides inpatient day-to-day care for your child, both before and after transplant. They will give you information regarding specific tests so you and your child will be well-prepared and will know what to expect during each test. They will also teach you how to care for your child after transplantation.

Social Worker

The social worker will provide emotional support and help for you, your child and your family. He or she may assist you with financial matters, housing, transportation and community resources.

Nutritionist

The nutritionist works with you to develop the best diet plan for your child. Many children with liver disease do not feel like eating and have problems growing. The nutritionist will work with your child to build healthy eating behaviors.

Child Life Specialist

The child life specialist works with your child to help him or her cope with the Hospital experience through play and other appropriate activities.

Psychologist

The psychologist assesses the ability of you and your child to cope with the transplant process and helps your child understand the process.

Case Manager/Discharge Planner

The Hospital case manager stays in contact with your insurance company. He or she will arrange for any equipment or services your child will need after his or her liver transplant.

What Your Liver Does

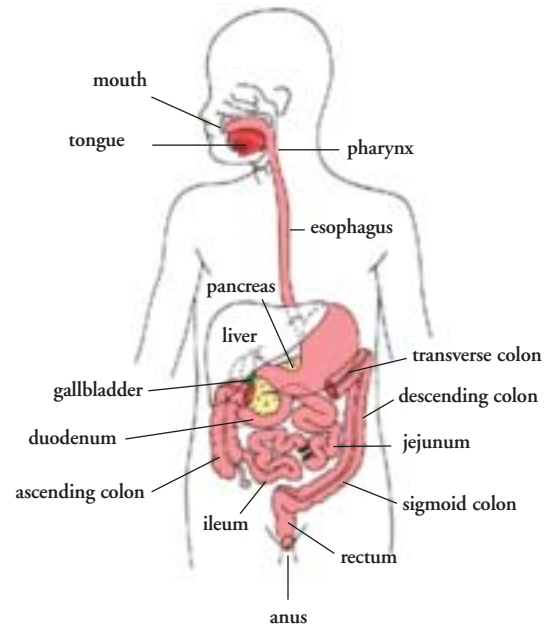
WHAT YOUR LIVER DOES

The liver is the largest organ in the body. It is located on the right side of the abdomen, underneath the ribs. It has many important functions. These include:

1. Changing food into chemicals needed for life and growth
2. Making proteins that are needed for normal blood clotting
3. Changing medications into forms the body can use
4. Cleaning poisons and drugs from the blood
5. Controlling blood sugar levels

The liver makes bile, which helps in the digestion and absorption of fat. Bile also aids in the absorption of vitamins A, D, E and K, and medications. Bile is stored in the gallbladder and is released into the intestines as needed.

The liver also filters the blood, removing many chemicals, drugs and waste products.



The Evaluation Process

THE EVALUATION PROCESS

Your child will need to go through an evaluation process before being placed on the transplant waiting list. This is an opportunity for you and your child to learn more about liver transplantation and to meet members of the liver transplant team. It also helps the medical team decide if a liver transplant is the best option for your child.

The pre-transplant evaluation is usually done on an outpatient basis and is coordinated through the liver transplant office. The evaluation process begins with a review of your child's medical and surgical history and an office visit with the hepatologist. Specific blood tests will be done. Radiology tests may also be scheduled as part of the evaluation process.

If transplantation is indicated for your child, a family meeting with the liver transplant team will be arranged. Some of the members you will meet are the hepatologist, transplant surgeon(s), nurse coordinator and transplant social worker. The results of your child's evaluation will be discussed during the meeting. The team will also explain the transplant surgery, donor options and post-transplant care. This is a good time to ask the team members questions about transplantation.

BEFORE THE TRANSPLANT

Once evaluated and accepted as a liver transplant candidate, your child will be listed with the United Network for Organ Sharing (UNOS) for liver transplantation at The Children's Hospital of Philadelphia. UNOS is a national agency that maintains the list of people awaiting transplant. It is also responsible for distributing organs.

Your child's rank on the liver transplant list is determined by several factors, including body weight, blood type and severity of the liver disease. A computerized program using your child's laboratory results generates a score that reflects the severity of your child's liver disease.

Once a donor is identified, these factors are compared with those of other people awaiting liver transplants who have the same blood type and are an appropriate size match for that donated liver. Children with the highest priority are transplanted as soon as possible.

Waiting for an organ will probably be one of the hardest times for you and your family. It is impossible to predict when an organ will become available. A liver may become available quickly or the wait may take weeks, months or years. You may worry about your child's medical condition and you may feel helpless. Waiting may become stressful and frustrating.

There are a number of things you can do to make the time pass more easily. By concentrating on the care of your child, you can ensure that he or she will be in good health when an organ becomes available.

Your pediatrician should continue to see your child for routine visits. You will need to come to transplant clinic for ongoing evaluations. It is important that you maintain contact with the transplant coordinator. He or she will need to know about changes in your child's health status. Contact the transplant coordinator if your child is hospitalized.

Other strategies to ease the stress of waiting include maintaining your normal daily routine. Discuss your concerns with your family or supportive friends. You may also contact a member of your child's transplant team who is available to help you through this stressful time.

While waiting for an available organ, it is important that the transplant center can reach you if a suitable organ becomes available. You will be asked to leave phone numbers with the transplant coordinator of people who can easily locate and contact you. You will also be asked to carry a beeper by which the transplant coordinator can reach you. When you receive your beeper, you must contact the liver transplant coordinator with the beeper number as soon as possible. We ask that you return the beeper afterward so it may be passed on to another family.

Your child may travel while waiting for a liver transplant. You should contact the transplant coordinator with the destination, schedule and phone numbers before you leave.

Getting to the Hospital

GETTING TO THE HOSPITAL

When your beeper goes off, you must call the phone number in the display. If the coordinator cannot reach you, the liver may go to the next person on the list, so it is important for the coordinator to have phone numbers where you can be contacted. Remember to check the batteries in your beeper and replace them as needed.

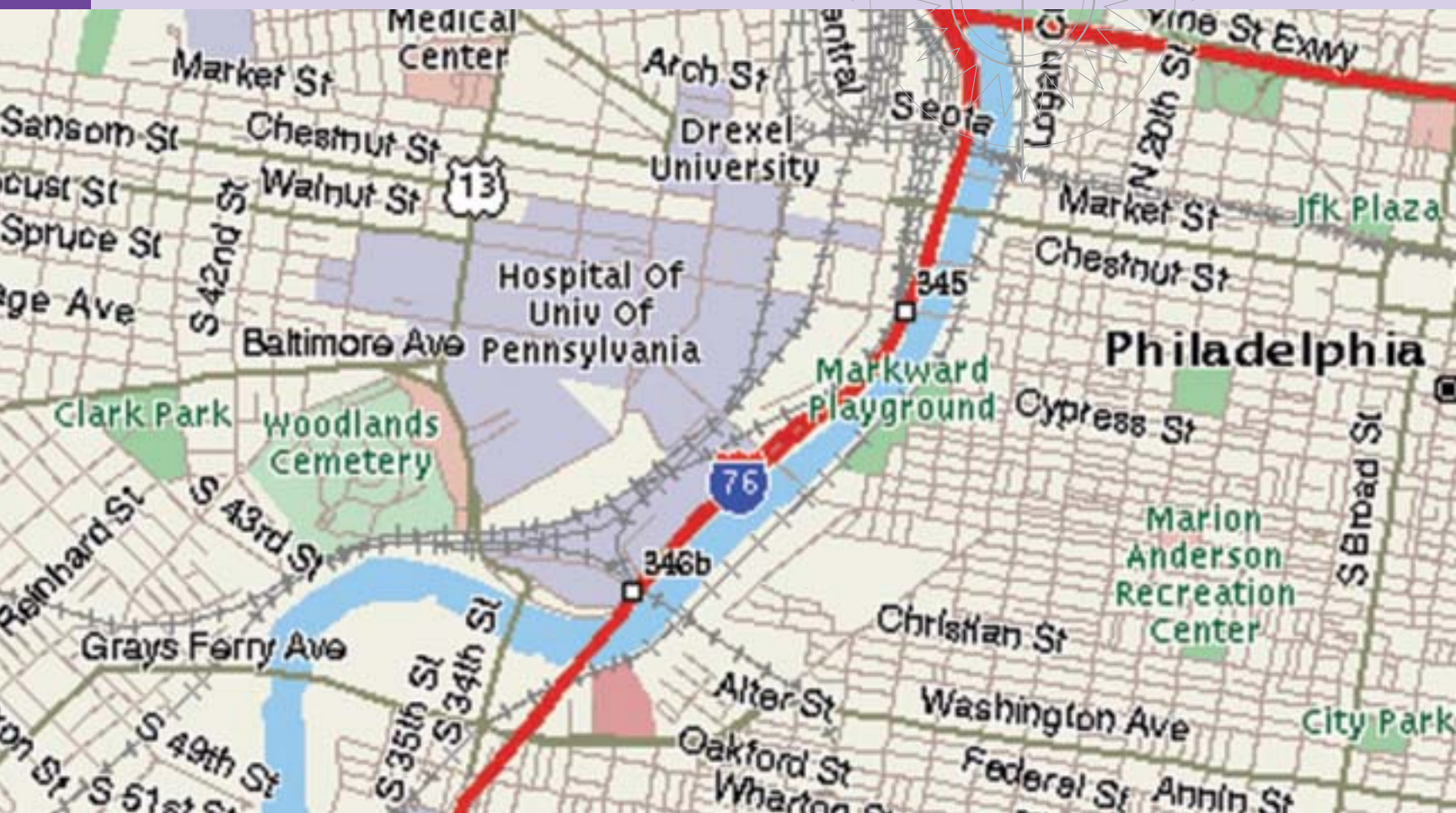
When you are contacted and told an organ is available, the transplant coordinator will ask you questions about the health of your child, such as if your child has had a recent cold, flu or fever, or if your child has been exposed to chicken pox or other infectious diseases. It is important that your child be in the best possible health for the transplant. If there are any concerns about your child's health, the transplant coordinator will discuss them with the transplant physicians.

Once it is determined that your child is in the best possible health for transplantation, you will be asked to come to the Hospital as soon as possible. The

transplant coordinator should be able to give you an estimated time when the transplant will take place. Your child should not eat or drink anything from this time on in preparation for surgery.

It is important to be prepared to travel to the Hospital. Travel arrangements should be planned beforehand. Whether you are traveling by car, plane or train, make your arrangements in advance. Keep a bag of your belongings packed. Make a list of things you want to bring with you.

Occasionally, children and their families are called into the Hospital for transplant and the donor organ is later found not to be usable for the patient. Though this is disappointing, your child will only be transplanted if the surgeons are confident the organ is perfect for your child.



AT THE HOSPITAL

When you arrive at Children's Hospital, you should go to the Admissions Office, located on the first floor of the Main Hospital Building, or the Emergency Department if it is after midnight. Once the admissions process has been completed, you and your child will be taken to a room on a patient unit.

When you arrive on the patient unit, an intravenous (IV) line will be placed in your child, and blood will be drawn. X-rays will be taken and a urine sample will be obtained. Your child will receive antibiotics through the IV. The anesthesiologist (the doctor who will help your child sleep during the surgery) and the transplant surgeon or surgical resident will perform physical examinations to determine your child's health status and review the surgical procedure. They will ask you to sign the surgical and anesthesia consent forms. These are standard procedures before any surgery.

THE OPERATION

The Operating Room team consists of anesthesiologists, surgeons, registered nurses and technicians. The entire liver transplant may take four to eight hours. Operating Room staff will call for your child about one hour before the surgery in order for the anesthesiologist and nurses to prepare your child for the operation. Medication to help your child sleep will be given through the IV line. Once your child is sleeping, two more IV lines and an arterial line will be inserted, as well as a Broviac catheter. A Broviac is a catheter that is inserted into a large vein. Blood can be drawn from this line, and it may remain in place even after your child is discharged from the Hospital. A Foley catheter will be used to monitor urine output, and a blood pressure cuff, EKG leads, a temperature probe and a pulse oximeter will be used to monitor your child's vital signs.

The surgeon will begin the procedure by cleaning your child's skin. After this is completed, the area operated on will be protected by drapes. Next, an incision in the shape of an upside-down "Y" will be made in the middle of the abdomen. The diseased liver will be removed and sent to the laboratory for examination, and the healthy liver will be sewn in place by the surgeons. Blood products may be given during the operation.

AFTER THE TRANSPLANT OPERATION *THE PEDIATRIC 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 different types of equipment, alarms and sounds. When your child arrives in the PICU, the nurses and physicians will ask you to wait to see your child so they can get him or her 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 various IV fluids and medications. The monitors and pumps have alarms that may go off. The nurses will check your child and equipment when this happens.

Your child will have an endotracheal (ET) tube coming out of his or her nose or mouth. This tube is placed in the trachea (windpipe) during the operation and connected to a ventilator to help your child breathe. Your child will not be able to talk, eat or drink with the ET tube in place. Many children communicate using hand signals or by writing on a pad and the ET tube is removed when your child no longer needs help breathing. Once the tube is removed, the nurse will have your child do deep breathing and coughing exercises to keep his or her lungs clear and to prevent atelectasis (lung collapse).

Other tubes and drains will be placed during surgery while your child is asleep in the Operating Room. A nasogastric tube (NG tube), which passes through the nose and into the stomach, helps to drain secretions from the stomach. There will be several Jackson-Pratt drains in your child's abdomen to help remove extra fluids from the operation. The NG tube and drains will stay in place for several days. A Foley catheter will drain urine from the bladder. It is used to measure urine output and to monitor kidney function.

There will be multiple intravenous lines and an arterial line. The arterial line looks like an IV, except it is in an artery. 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 a surgical unit.

A Broviac catheter may also be placed while your child is asleep in the Operating Room. It might be used to give IV fluids and medications, or to draw blood. Your child may go home with the Broviac catheter. If this occurs, you will be taught how to care for the catheter.

Your child will have many routine studies and tests following the transplant surgery. These will include ultrasounds, X-rays and blood tests. An ultrasound is done either the day of or the day after surgery to check blood flow to the liver. It may be repeated during the Hospital stay. Chest X-rays are done to assess your child's lungs. Blood tests are done daily throughout the Hospital stay.

Once your child is medically stable and does not require close monitoring and assessment, he or she will be transferred from the PICU to a surgical unit. The nurses will continue to monitor and assess your child while on the surgical unit.

If you have any questions about your child's care, please feel free to call the Liver Transplant Program at 215-590-4281.

AFTER THE TRANSPLANT OPERATION
THE RECOVERY PERIOD

The recovery period begins in the PICU and continues throughout your child's Hospital stay. All of your child's bodily functions will be monitored by the physicians and nurses. When medically stable, your child will be transferred to a private room on a surgical unit. The surroundings will be more relaxed because patients on the surgical unit no longer need the continuous monitoring they required in the PICU.

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

Walking

Take your child for walks around the room and hallway. This helps build strength. Your child will become more active every day. Walking also helps to improve your child's digestive function and allows for better expansion of the lungs. When your child is able to better expand his or her lungs, it decreases the risk of developing pneumonia or atelectasis, which are common concerns after surgery.

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 him or her take deeper breaths. This will also help prevent pneumonia and atelectasis.

Eating

When your child is allowed to eat, he or she will advance slowly from clear liquids to solid foods. The body needs many calories to heal and grow. It is important that your child eat well-balanced meals. If your child is unable to eat enough to provide the necessary nutrition, he or she may be fed through an NG tube or the IV. The nutritionists 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. A child life specialist will help your child find toys and games and suggest appropriate play activities during the recovery period.

REJECTION

Organ rejection may occur any time after transplant. It may occur immediately after surgery or even months or years later. The first year after liver transplantation is the hardest. Rejection can be treated most of the time and does not necessarily mean your child will need another transplant.

Early signs of rejection are:

- Fever
- Increase in liver enzymes

Late signs of rejection are:

- Jaundice, or a yellow color of the skin and eyes
- Changes in stool or urine color

Some or none of these symptoms may occur during rejection. Often, changes in blood tests are the only sign of rejection. It is important to keep your appointments for transplant clinic and lab work after your child is discharged from the Hospital. A liver biopsy will be done if physicians think your child had rejection. Rejection can only be diagnosed by a liver biopsy, which allows the physicians to see how much your child's liver has been affected.

Your child will need to be hospitalized for rejection. Rejection is treated with an intravenous steroid called Solu-Medrol™, an IV form of Prednisone. If Solu-Medrol does not work, other, stronger anti-rejection medications may be used.



CONCLUSION

If all goes well, you can expect a Hospital stay of two to four weeks. When your child is discharged, you will need to come to the transplant clinic once a week for at least the first month. Your child will be examined and blood will be drawn. This is the time when rejection and most problems occur. It is important that your child be monitored closely by the physicians and nurses on the transplant team.

As your child's condition stabilizes, clinic visits will become less frequent. If there are any medical problems or you have any questions, you should call the Liver Transplant Office, Monday through Friday, 8 a.m. to 5 p.m. In case of emergency, call the Hospital's main number, 215-590-1000, and have the operator page the GI fellow on call.

Watch your child. This is the best way of knowing how he or she is doing. Your child's play and activities are the best measure of recovery.

Please close book
then flip over to learn
“What to expect
after transplant.”



The Liver Transplant Program
A Guide for Families

What to expect: Post-Operative



The Children's Hospital of Philadelphia®

Hope lives here.

INTRODUCTION

Soon your child will be ready to leave The Children’s Hospital of Philadelphia. This is an exciting time. However, you may be feeling anxious. This booklet provides information about caring for your child at home. Some information from the pre-transplant section will be reviewed.

The first month after transplant is a busy time. Your child will come to the outpatient Liver Transplant Clinic frequently for blood work and clinic visits. You may need to take your child for blood work in between clinic visits. This is also a time of transition, learning to care for your child at home and adjusting to a more normal lifestyle after hospitalization.

We hope this booklet answers some of your questions. If you have additional concerns or questions, please do not hesitate to call the Liver Transplant Program at 215-590-4281. Remember that you have a right to information and should not be afraid to call. It might be helpful to write down questions before calling.

Introduction	2
Medications	3
Anti-rejection Medications	4
Medications to Prevent Infections	5
Additional Medications	5
When to Contact the Liver Transplant Team	6
Follow-up after Discharge	7
Rejection	7
Activity	8
Nutrition	8
Immunizations	8
Chicken Pox	9
Dental Care	9
Glossary	10
Resource List	13

MEDICATIONS

It is important for you and your child to understand why and how each medication is given. In the Hospital, your child is taking many different medications, and most of these will still be needed after discharge. When your child no longer needs a specific medication, the Transplant Team will discontinue it. The medications to prevent rejection will need to be taken for the rest of your child's life.

Getting children to cooperate in taking medication can sometimes be difficult. Parents and nurses often need to set firm limits with children about taking medication. It is important that children take their medicine as prescribed and on time. You may allow your child some control by giving choices such as which medication to take first, what to take with the medication and how to take the medication.

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. Many times a school nurse will also be available and can give your child medication during school hours.

Never let your child's medications run out. Call your pharmacy to refill prescriptions at least one week before the medication supply will run out. During the week, if there are no refills left on the prescription, call the Liver Transplant office. You will need to provide the name(s) of the medication(s), if it is in pill or liquid form, the strength and dose, and the phone number of the pharmacy.

Contact the liver transplant coordinator if another doctor prescribes medications for your child. Some medications affect the blood levels of anti-rejection medications. Your child's medication dose may need to be adjusted to prevent low or high blood levels of anti-rejection medications.

ANTI-REJECTION MEDICATIONS

Tacrolimus (FK506, Prograf™) is an immunosuppressive drug that is used to prevent rejection of transplanted organs. It is taken in the morning and at night, with 12 hours between each dose. It should be given at the same time every day. The dose is determined according to the amount of tacrolimus in your child's bloodstream, kidney function and side effects. It is important to give the drug exactly as directed. Tacrolimus comes in 0.5 mg, 1 mg and 5 mg capsules, and as a liquid suspension.

Side effects may include high blood pressure, headache, infection, high blood sugar, tremors, elevated creatinine levels, low magnesium level and diarrhea.

If it is a clinic or blood-draw day, do not give Tacrolimus until after the blood tests are finished.

Cyclosporine (Neoral™, Gengraf™) is an immunosuppressive medication that is used to prevent rejection of transplanted organs. It is taken in the morning and at night, with 12 hours between each dose. The dose is chosen according to the amount of Cyclosporine in your child's bloodstream. It is important to give this drug exactly as directed. Cyclosporine comes in 25 mg and 100 mg capsules and liquid.

If the liquid form of Neoral™ is prescribed, it is important that you follow a few instructions. The liquid form will taste better if mixed with chocolate milk, orange juice or apple juice. Do not mix or take Cyclosporine with grapefruit juice. Mix the medicine in a glass or hard plastic container with a metal spoon. Rinse the container with additional juice or chocolate milk to get the full dose. Do not rinse the dropper with water or other cleaning agents. Note: store capsules below 77° F, and the liquid below 86° F.

Side effects may include high blood pressure, headache, infection, elevated creatinine, increased body hair, overgrowth of gums and upset stomach.

If it is a clinic or blood-draw day, do not give Cyclosporine until after blood tests are finished.

Sirolimus (Rapamune, Rapamycin) is an immunosuppressive medication that is used to prevent rejection of transplanted organs. It is taken in the morning only. The dose is chosen according to the amount of Sirolimus in your child's bloodstream. It is important to give this drug exactly as directed. Sirolimus comes in capsules and liquid.

If it is a clinic or blood-draw day, do not give Sirolimus until after blood tests are done.

MEDICATION FOR TREATMENT OF REJECTION

Prednisone, Prednisolone (Solu-Medrol™, Pediapred™, Prelone™) is a steroid that decreases the body's ability to reject the transplanted liver. It is given once a day in the morning. The dose will be decreased over a period of three to six months and eventually stopped. The side effects of Prednisone are usually from high doses. Note: always take Prednisone with milk or food to prevent an upset stomach.

Side effects may include hypertension, upset stomach, increased appetite, weight gain, puffy face, acne, high blood sugar, emotional changes, insomnia and fluid retention.

Mycophenolate Mofetil (Cellcept, MMF) is an immunosuppressive drug that is used in addition to Prograf and Prednisone to treat rejection of the transplanted organ. It is taken in the morning and at night, with 12 hours between each dose. It should be given at the same time every day. Cellcept comes in 250 mg capsules and 500 mg pills. A liquid form is available. Note: Cellcept should be taken on an empty stomach. However, if this gives your child an upset stomach, the medication may be taken after a meal. If your child cannot swallow the capsule whole and the liquid form is not available from your pharmacy, the capsules may be opened and the powder mixed in water, fruit juice or apple sauce.

Side effects may include upset stomach, diarrhea, infection and low white blood cell count.

MEDICATIONS TO PREVENT INFECTIONS

Co-trimoxazole (Bactrim™, trimethoprim and sulfamethoxazole, Septra™) is an antibiotic to prevent pneumocystis pneumonia. It is taken once a day on Monday, Wednesday and Friday. Your child will be on this medication until the first anniversary of his or her transplant.

Side effects may include:
skin rash, low white blood cell count

Nystatin is an antifungal medication to prevent thrush. It is taken four times a day. Your child should swish the liquid in his or her mouth and swallow it. Your child should not eat or drink for 30 minutes after taking Nystatin. Your child will be on Nystatin for as long as he or she is on Prednisone.

Side effects may include:
nausea, vomiting, diarrhea

Zantac (Ranitidine) is an acid blocker that will protect your child's GI tract from ulcers, which may develop as a result of being on the Prednisone. It comes in a tablet or a liquid. It is taken two to three times per day. Your child will be on this medication for at least as long as he or she is on Prednisone.

Side effects may include:
headache, dizziness, vomiting, constipation

Ganciclovir is an antiviral medicine that is given intravenously to prevent cytomegalovirus (CMV). Your child will receive this once a day while in the Hospital and may go home on it. Ganciclovir is also used to treat CMV infection.

Side effects may include:
low platelet count, low white blood cell count, rash, elevated creatinine

Acyclovir (Zovirax) is an oral antiviral medicine used to prevent CMV. Your child may have to take this medication after he or she leaves the Hospital. It is taken three times a day for 100 days after transplant.

Side effects may include:
rash, upset stomach, vomiting, dizziness, headache

ADDITIONAL MEDICATIONS

Aspirin is given to prevent blood clots from forming in the blood vessels in the liver. It is taken once a day in the morning. Your child will be on aspirin until the one year anniversary of his or her transplant.

Side effects may include:
bruising, upset stomach, vomiting, dizziness, headache

Magnesium supplements may be prescribed for your child. These are usually taken two to four times a day. Magnesium supplements are available as magnesium gluconate and magnesium sulfate, and they come as pills or liquid.

Side effects may include:
diarrhea, upset stomach, vomiting

Calcium supplements may be prescribed if your child has low bone density because of his or her liver disease or medications. Calcium supplements are available in many forms over the counter.

Side effects may include:
constipation, headache, upset stomach, vomiting

Name of Drug Concentration	Dosage	Morning 8 a.m.	Noon	Evening 4 p.m.	Morning 8 a.m.	Special Instructions
Prograf (Tacrolimus/FK506) 1mg/ml	mg ml	•			•	Twice per day * For clinic/labs hold until after labs are drawn
Ganciclovir (Cytovene) 100mg/ml	mg ml	•		•	•	Three times per day
Prednisolone 3mg/ml	mg ml	•				Once in the AM
Aspirin tab 81mg	mg tab	•				Once in the AM * Take with food
Bactrim (Co-trimoxazole/trimethoprim/sulfamethoxazole) 40/200/5ml	mg ml	•				Once per day * Only on M W F
Nystatin 100,000 U/ml	ml	•	•	•	•	Four times per day
Zantac (Ranitidine) 15mg/ml	mg ml	•			•	Two times per day

WHEN TO CONTACT THE LIVER TRANSPLANT TEAM

- If your child has a temperature greater than 101.5° F at any time or a low grade fever (99.5° to 100.5° F) for more than three days. Do not give Tylenol or acetaminophen without contacting the Liver Transplant Team first.
- If your child is having diarrhea or vomiting. Both can affect the level of medication in your child's blood.
- If your child has redness, pain, swelling or pus draining from the surgical incision.
- If your child has a cough or cold that won't go away.
- If your child is exposed to diseases such as chicken pox or measles.
- If your child develops mouth sores.
- If your child misses several doses of medication(s).
- If your child repeatedly vomits the medication after it is given.
- If too much medicine is given.
- If your child has any side effects from the medication.
- If your child has any signs of rejection (see Page 9).
- If your child does not feel well.
- If any other physician prescribes a new medication, even if it is a cough medicine.

Remember to call the office at 215-590-4281 during normal business hours (8 a.m. to 5 p.m.). Paging should be reserved for emergencies only, and even in the event of an emergency, you should call the office first.

If it is after 5 p.m. or on a weekend or holiday, call the operator at 215-590-1000 and ask for the GI fellow on call.

FOLLOW-UP AFTER DISCHARGE

Your child's care does not end after discharge from the Hospital. He or she will need to come to the outpatient Liver Transplant Clinic for regular visits and lab tests. This is a good time to ask questions regarding your child's health and medications. The Liver Transplant Clinic is located in the GI clinic on the third floor of the Richard D. Wood Pediatric Ambulatory Care Center. Your child will be seen weekly for the first month after his or her transplant and then every other week for one month. The frequency of the clinic visits will decrease depending on how your child is doing. Sometimes blood tests may need to be performed in between clinic visits. These may be done either at the Hospital or at a lab close to your home.

When you bring your child to the clinic, you will need to bring any referrals that your insurance provider requires. After your child is registered, he or she will go to the laboratory to have blood drawn. Do not give your child Prograf (FK506), Cyclosporine (Neoral) or Rapamycin (Sirolimus) before his or her blood is drawn. Please bring the medication with you to the clinic visit so you can give it to your child after his or her tests.

The transplant team will review your child's lab results on the Tuesday following clinic. The transplant coordinator will contact you to let you know if any medication changes need to be made or if additional blood tests need to be done. The transplant coordinator will call you only if there are any changes.

If you have any problems or questions, call the Liver Transplant Program at 215-590-4281 between 8 a.m. and 5 p.m. For emergencies after 5 p.m., and on weekends or holidays, call the Hospital operator at 215-590-1000 and ask for the GI fellow on call.

REJECTION

The immune system is the body's natural defense mechanism against infection. It protects the body from foreign substances, such as bacteria and viruses, by destroying them. Organ rejection occurs because the immune system considers the transplanted liver to be a foreign body and tries to attack and destroy it. To prevent this, your child is placed on immunosuppressive, or anti-rejection, medications. These medications decrease the activity level of the immune system, making it less active. Your child will have to take immunosuppressive medication for the rest of his or her life. Rejection, however, may still occur even while your child is taking immunosuppressive medication.

Almost all patients will experience an episode of rejection. It most often occurs seven to 10 days after the liver transplant. The chance of rejection is less over time, but it may occur at any time after transplantation. Signs of rejection include:

- Fever
- Clay-colored stools
- Increased liver enzymes
- Dark-colored urine
- Yellow eyes and skin

Some or none of these symptoms may occur during a rejection episode. Often, changes in liver tests are the only sign of rejection. This is why it is important to keep scheduled appointments for Liver Transplant Clinic and lab studies. If rejection is suspected, your child may need a liver biopsy to confirm the diagnosis of rejection and to allow the physicians to see how much of your child's liver has been affected.

Most rejection episodes can be successfully treated and rejection rarely results in irreversible damage to the transplanted liver. Your child may be admitted to the Hospital depending on the severity of the rejection. Intravenous steroids and adjustments to the immunosuppressive medications are the usual treatments for rejection. OKT3, which is a stronger anti-rejection medicine, may also be used to treat rejection. Remember rejection does not mean your child will need to be re-transplanted. However, chronic rejection (repeated episodes of rejection) may lead to re-transplantation, so it is important to take the medications every day as directed.

Notify the Liver Transplant Team immediately if your child has any signs of rejection.

ACTIVITY

Your child may return to his or her regular activities after a liver transplant. There may be restrictions at first to allow the incision to heal, however after that (while wearing protective gear), your child may participate in gym class, ride a bike, skateboard, etc. He or she should start exercising slowly and gradually to build up strength. Your child may require rest periods. Most parents report that their children have more energy after transplant. This is because they now have healthy livers that can appropriately metabolize nutrients and waste products.

NUTRITION

Nutrition plays an important role in your child's recovery after transplant. Some children have a difficult time maintaining good nutrition before transplant due to liver disease. This may result in a longer time period before they are in this best state of health.

Sometimes children are unable to take in enough calories and need other kinds of nutrition. Dietary supplements, such as high-calorie drinks or nasogastric tube feeds, may be used to provide the additional nutrition. Usually a well-balanced diet with enough calories and a standard 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. His or her appetite may be better or worse and this happens for many reasons. It is important to know that all children are different and that they should not be forced to change their habits. Your child should eat a reasonably balanced diet of fruits, vegetables and low-fat meats, such as poultry and fish, beans and grain products. Sugar and salt should be allowed in small amounts. The transplant nutritionist can help you plan good choices for your child.

Herbal supplements, natural food supplements and home remedies should not be given to your child without checking with the transplant team. Some of these supplements interfere with the immunosuppressive medications and may cause them to fail.

IMMUNIZATIONS

Children who have had chronic illness may not have received regularly scheduled immunizations. Most of these children may continue their immunization schedule after their transplant. Your pediatrician and the transplant team can tell you when this should happen.

All immunizations should be kept up to date, including live virus vaccines such as MMR (measles, mumps, rubella) and varicella (chicken pox). Sometimes due to the age of a child at transplant, he or she may not receive MMR or varicella before the transplant. Live virus vaccines will be postponed until after a child is off steroids. Please speak to the Liver Transplant Team for more information.

The recommended inactivated vaccines should also be given to your child. These include inactivated poliovirus vaccine (IPV), Haemophilus influenzae type b (Hib) conjugate vaccine, hepatitis B (Hep B) vaccine, DTP and pneumococcal vaccine. The meningococcal vaccine is recommended for adolescents and college-bound children.

Annual influenza vaccines, or flu shots, are recommended for all transplanted patients and anyone they live with. The vaccine usually becomes available in the fall. Call your child's pediatrician to arrange for your child to receive a flu shot.

CHICKEN POX

Chicken pox is a common childhood disease. It may develop at any age but occurs more frequently in children between the ages of 5 and 10. Chicken pox can be a serious illness for a child who is not immunized and has received a liver transplant.

Chicken pox is spread through the air or by close contact with someone who has the virus. A person with chicken pox is contagious two days before the rash ever occurs and until the rash has completely scabbed over. If your child is around someone during the contagious time, the virus may develop somewhere between 10 and 21 days after exposure (the incubation period).

Chicken pox often begins with a fever. The rash may begin on the head and spread downward, or on the trunk and spread outward. The rash consists of small watery blisters with red rings around them. The child may feel ill for a few days.

If at any time your child or any siblings have been exposed to chicken pox, be sure to contact a member of the transplant team right away. When exposed, your child may require a VZIG injection depending on his or her most recent varicella blood levels. This injection will lessen the effects of the chicken pox. Should your child actually develop a rash, it will be necessary to admit him or her to the hospital.

DENTAL CARE

All children should visit the dentist regularly. This is especially important for your child after his or her liver transplant.

Topical application of fluoride to the teeth may be necessary if the water supply in your home community does not contain fluoride. Fluoride helps to prevent tooth decay.

Any dental work, such as filling a cavity, may require antibiotics to prevent infection. Your dentist may call the transplant office about this. A dentist at Children's Hospital can be recommended if you do not have a dentist. Routine cleanings should not require antibiotics.

Usually, a dose of amoxicillin must be taken one hour before dental work.

You will also need to let your dentist know that your child is taking steroids every day.

If your child is taking Cyclosporine, he or she may develop gum overgrowth (hyperplasia). In order to prevent gum disease from this overgrowth, your child should visit the dentist at least three times a year for preventative dental care.

Alpha1 antitrypsin deficiency – An enzyme normally present in the blood. If the enzyme is decreased or absent, it may result in liver disease.

Arterial blood gas (ABG) – A blood test that measures how well the body is using oxygen.

Arterial line (A-Line) – A catheter placed in the artery. It is used to constantly check blood pressure without using a blood pressure cuff, and to withdraw blood for tests without having to stick the child with a needle each time.

Ascites – A buildup of fluid in the abdominal cavity.

Atelectasis – Collapsed air sacs in the lungs.

Atresia – A closure of a normal opening or absence of a normal anatomical opening present at birth.

Biliary Tree – A series of ducts that provide transport of bile from the liver to the duodenum (small intestine) where the bile is used to digest foods.

Bile – Secretion from liver cells; a thick fluid that passes from the bile ductules of the liver into the common bile duct and then into the duodenum. It helps in the digestion of fats. Bile is yellow in color. After it is stored in the gallbladder, the color varies from yellow to green to brown.

Bilirubin – A result of the breakdown of red blood cells. If the liver is functioning properly, the bilirubin is taken from the liver in the bile. When the liver does not excrete bilirubin, it builds up in the blood and causes jaundice.

Broviac – A catheter placed in a large vein or central vein located close to the heart. It allows for large volumes of fluid to be given and may be used to draw blood for studies. It is inserted in the operating room and may remain in place when the patient is discharged.

Cardiac Respiratory Monitor (CR Monitor) – A machine that measures the heart rate and breathing rate by placing three small, round, sticky dots on the chest. Thin, protected wires called leads are connected to the machine and the dots, allowing the staff to monitor the heart and breathing rates.

Cholangitis – Swelling of the bile ducts.

Chole – Refers to bile.

Cirrhosis – A chronic disease of the liver in which there is destruction and/or scarring of tissues and cells of the liver.

CT Scan (Computerized Tomography) – A test that combines an X-ray machine and a computer. X-rays passing through a part of the body are changed into signals that go to a computer. The computer takes the signals and forms an image.

DISIDA Scan – A radioactive scan in which a dye is injected into a vein and the normal course of bile flow is seen through X-ray techniques. This test helps identify whether there is an obstruction and, if so, where it is.

Electrocardiogram (EKG) – The measurement of the heart’s “electrical system.” It requires numerous small dots or bands on several parts of the body, usually hands and feet. This test does not hurt and is safe.

Endoscopy – An examination during which a physician places a narrow, lighted tube through the mouth to look at the esophagus and the stomach.

Endotracheal Tube (ET Tube or Artificial Airway) – A long, narrow plastic tube placed through the mouth or nose into the windpipe to provide an airway. It is taped in place.

Fibrosis – Abnormal formation of liver tissue.

Foley Catheter – A plastic tube that is inserted into the bladder. It is used to measure urine output.

Hepatic – About the liver.

Hepatitis – Swelling of the liver.

Hepatomegaly – Enlargement of the liver.

Hepatosplenomegaly – Enlargement of the spleen or liver.

Immune System – The body's natural defense system. It protects the body from foreign substances, such as bacteria and viruses.

Incentive Spirometer or Inspirometer – A plastic box with a flexible tube and a plastic ball. A patient is instructed to take large breaths through the tubing — as if using a straw — and move the plastic ball upwards. It helps prevent respiratory infections and pneumonia.

Intravenous Line – A catheter (tiny plastic tube) placed into a vein to give fluids or medication.

Jaundice – A yellow color in the skin and whites of the eyes due to the bile pigments that result from too much bilirubin in the blood and tissues.

Jackson-Pratt Drain – A plastic drain placed in the wound during surgery. At the end of the tubing is a bulb-shaped drain to collect the extra fluids.

Liver – The largest organ in the body, located in the upper-right portion of the abdomen under the rib cage. The liver makes and balances body chemicals; uses proteins, uses body fats; stores energy sources (proteins, fats, sugars), vitamins A, D, E, K and mineral (copper and iron); changes potentially harmful substances into forms the body can either use or get rid of; and filters bacteria and particles from the blood.

Liver Biopsy – A test in which a small sample of liver is removed. A needle is inserted through the abdominal wall into the liver. This test allows the physician to examine liver tissue under the microscope and to obtain samples for culture for the diagnosis of specific liver disorders.

MRI (Magnetic Resonance Imaging) – A scan that shows a detailed picture of the body without X-rays. This test uses magnets and radio waves that have no known side effects and are painless.

Nasogastric Tube (NG Tube) – A long, narrow plastic tube inserted in the nose and passed down into the stomach. May be used to empty the stomach of fluid and air, or to feed formula. It is taped in place.

Operative Cholangiography – A procedure in which dye is injected into the gallbladder and an X-ray is taken outlining the structure of the biliary tree. It requires an operation.

Percussion and Postural Drainage (PP&D) – A clapping on the back and chest with a cuffed hand (percussion) to loosen secretions and mucus and promote drainage of secretions (postural drainage). Helps to prevent respiratory infections and pneumonia.

Percutaneous Transhepatic Cholangiogram (PTC) – A procedure during which dye is injected through a catheter into a liver bile duct. X-rays are taken while the dye is injected. The PTC is done to check for leaks, blockages and other problems.

Pruritis – Severe itching believed to be caused by the buildup of bile salts and other by-products.

Pulse Oximeter – A machine used to monitor the oxygen levels in the bloodstream. This is done by taping a probe to a fingertip or toe.

Rectal Probe – A small, protected wire placed in the rectum to monitor a patient's body temperature.

STAT – To be done immediately.

Ultrasound – A test that uses sound waves to outline different organs and tissues in the body. These sound waves are sent to the area being tested and a picture is made as the sound returns to the machine. There is no radiation used during the test. A probe, which looks like a wand with a rounded tip, gently presses the skin near the area tested.

Ventilator – A large “breathing machine” that automatically provides breaths for someone with an artificial airway.

X-Ray – A picture of a part of the body using a small amount of radiation.

Laboratory Tests

To help monitor recovery and to evaluate medical therapy, blood will be drawn daily for testing while your child is in the Hospital. Many of these tests may be familiar to you; your doctor has used them to assess your child’s liver disease. These tests continue to assist in the monitoring of your child’s progress.

ALB (Albumin) – Measures the liver’s ability to make protein.

Alk Phos (Alkaline Phosphatase) – Measures bile duct function. Alk Phos is also found in bone. It may be higher during growth spurts.

ALT (SGPT) – Measures the amount of alkaline phosphatase, which is produced and stored by the liver.

Amy (Amylase) – Assesses the function of the pancreas. It is secreted by the pancreas into the intestine to assist in the digestion of carbohydrates.

AST (SGOT) – Measures the amount of amylase produced and stored by the liver. AST also is produced and affected by other organs.

Bilirubin – When red blood cells (RBCs) are broken down, hemoglobin is released. The hemoglobin is broken down into bilirubin, which is excreted by the liver.

BUN (Blood Urea Nitrogen) – Assesses the function of the kidney. The stomach changes protein into amino acids, which the liver breaks down into urea. Urea then enters the blood and is excreted by the kidneys.

Cr (Creatinine) – Assesses kidney function, but is more specific than BUN.

GGTP – Measures the amount of creatinine, which is produced in the bile ducts. It indicates bile duct function.

Hct (Hematocrit) – Measures the percentage of red blood cells circulating in the blood.

Hgb (Hemoglobin) – Carries oxygen from the lungs to other tissues.

K+ (Potassium) – Electrolyte found in the blood that has an important role in muscle contractions.

Na (Sodium) – Electrolyte found in the blood that reflects a balance between sodium intake and excretion by the kidneys. It is closely interrelated to the water in the body.

Plt (Platelets) – Component of blood that helps it to clot.

PT/PTT/INR (Prothrombin Time/Partial Thromboplastin Time) – Measures how long your child’s blood takes to clot.

TP (Total Protein) – Measures the amount of albumin (made in the liver) and globulin (made in other organs) in the blood to help assess liver function.

WBC (White Blood Cells) – Fight infection and react against foreign substances in the body.

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