



Heart/Lung Transplantation

A Handbook for Families

ST. LOUIS CHILDREN'S HOSPITAL

WELCOME

The heart/lung transplant team at St. Louis Children’s Hospital would like to welcome you to our hospital and transplant program. As you know, patients are considered for heart/lung transplantation when either medical therapy or surgery cannot significantly improve their heart/lung disease and consequently, there is a high probability of death. Heart/lung transplantation is not a cure, but rather a treatment. We strive to increase the quality of life for your child. It is our greatest hope that by having a heart/lung transplant your child will have the opportunity to achieve life goals that are important to them.

This booklet has been written as a resource for you. It contains basic information regarding heart/lung transplantation that is important for you and your child to understand. Please remember that each child who is transplanted is an individual who comes with their own complex medical history and will respond to transplantation in their own unique way. The transplant team is committed to providing the best possible care based on your child’s individual needs.

This booklet is provided through the generosity of contributors to the St. Louis Children’s Hospital Foundation.

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ST. LOUIS CHILDREN'S HOSPITAL

GENERAL INFORMATION

During your child's stay in our transplant unit, the heart/lung transplant team will be your main contacts for the overall care and treatment of your child. Your child's primary pulmonary physician will work closely with the heart and lung transplant teams and remain involved in the medical care of your child. You may need to call a team member during the transplant process. Following are some important numbers:

IMPORTANT NAMES AND NUMBERS	
St. Louis Children's Hospital (SLCH)	314.454.6000
SLCH Toll Free Number	877.578.4449
PRIMARY NAMES AND NUMBERS	
Heart/Lung Transplant Social Worker	314.454.6068
Transplant Financial Coordinators	314.454.2091 314.454.6191
Heart/Lung Transplant Program Fax	314.454.4280
ADDITIONAL HELPFUL NUMBERS	
Ambulatory Procedure Center	314.454.2514
Chaplain	314.454.6211
Child Life Services	314.454.6178
Family Resource Center	314.454.2350
Information Desk	314.454.2615
Outpatient Lab	314.454.4130
Outpatient Pharmacy	314.454.6123

ADDITIONAL HELPFUL NUMBERS	
Patient Accounts	314.286.2384
Patient Registration Services	314.286.2700
Psychologist	314.454.6069
Pulmonary Function Lab	314.454.2695
Same Day Surgery	314.454.2290
School Liaison	314.454.2173
Security (escort to parking)	314.454.2660
<i>* If you are calling from within the hospital, dial only the last 5 digits of the number.</i>	

LODGING

Transplantation is a long and often complex process. You will be asked to live in the St. Louis area pre- and post-transplant. Please plan on establishing a place to stay, whether with relatives, friends, local hotel, or if available, Ronald McDonald House post-transplant.

Ronald McDonald Houses: St. Louis currently has three Ronald McDonald Houses with long-term apartments that serve as lodging for families. These accommodations cost \$5 per night. First-time guests must work with the social worker to arrange for this lodging. Return guests may work directly with the local Ronald McDonald House.

Your social worker can provide more detailed information regarding reserving a place in one of these houses. He/she will also suggest other lodging options. Please be aware that there is often a waiting list for families, you should plan ahead and anticipate lodging needs.

PARKING

Parking is available in the visitor's garage across from the entrance to the hospital on Children's Place. Only parents and grandparents of inpatients may have their parking tickets validated at the information desk.

FOOD SERVICES

- **Edison Center Atrium Café**, the hospital's cafeteria, located on the lower level, is open nearly 24 hours a day. You may order guest trays (call 42752) that will be delivered with patient trays. Vending machines are also available on the lower level near the cafeteria.
- **Barnes-Jewish Hospital (BJH)**, North & South each have a cafeteria. BJH, North & South are both connected to our hospital via an enclosed walkway. Our hospital operator can provide the appropriate phone numbers to enable you to check hours of operation and selection.
- **Special food needs or requests** can be accommodated. If you have any dietary restrictions, please inform your child's nurse, dietitian, or social worker. Please also tell them of any significant family dates such as birthdays or anniversaries, so we may try to assist you in celebrating them.

THE CENTRAL WEST END

Our hospital is within walking distance of a popular area of St. Louis called the Central West End. Within several blocks are many dining choices, including ice cream shops, sandwich shops and full-service restaurants. There are also art galleries, antique shops, novelty shops, a movie theatre and a bookstore. However as in any large metropolitan area, one must be aware of one's surroundings.

OTHER SERVICES IN OR NEAR THE HOSPITAL

For information about available services in this hospital, please refer to the St. Louis Children's Hospital Family & Patient Guide, which you will receive with your initial evaluation packet. You will find information about our Family Resource Center, banking, postal services, the hospital chapel, the laundromat and other available services.

UNDERSTANDING

HEART/LUNG TRANSPLANTATION

HISTORY

The first successful heart/lung transplantation was performed in 1981. Ongoing research led to the development of single and double heart/lung transplantation in 1983. St. Louis Children's Hospital performed its first lung transplant in 1990, its first heart/lung transplant in 1993, and its first living donor lobar transplant in 1994.

St. Louis Children's Hospital operates the largest and most active pediatric heart/lung transplant program in the world. The heart/lung transplant program is incorporated in the lung transplant program. It attracts pediatric patients from across the country and has provided transplants for patients from Israel, Mexico, Japan, Saudi Arabia, Canada, and Australia. In general, outcomes comparable to lung transplant survival.

Alternative treatment therapies may be available for your child's medical condition. Please discuss your child's condition and any possible alternative therapies with your health care team.

COMMON INDICATIONS FOR A COMBINED HEART/LUNG TRANSPLANTATION:

- Pulmonary hypertension with structural heart defect(s) not amenable to surgical repair
- Patients with severe heart failure involving the left ventricle in conjunction with the heart/lung disease due to any of the conditions listed above
- Potential Candidates for heart/lung transplantation must meet the following criteria:
- End-stage heart and lung disease with evidence of progression of disease and limited life expectancy or stable requirement for mechanical ventilation.

- No other active systemic disease or irreversible organ dysfunction likely to progress or complicate the transplant.
- Low likelihood of recurrence of the original disease.
- Committed family and psychologically stable patient with a record of adherence to medical regimen, and stable, adequate support structure.
- Adequate potential to participate in follow-up testing cardiopulmonary rehabilitation.
- Adequate insurance coverage or means to pay for services and medications.
- Access to routine and pulmonary follow-up care in home environment and means to travel to St. Louis for transplant follow-up when necessary.

ABSOLUTE CONTRAINDICATIONS TO HEART/LUNG TRANSPLANT:

- Severe scoliosis.
- Intractable systemic infection.
- Malignancy (unless in remission for greater than two years with low likelihood of relapse.)
- Acquired Immunodeficiency Syndrome.
- Chronic Renal Failure requiring dialysis.
- Active Tuberculosis.
- Irreversible Central Nervous System Injury leading to respiratory compromise.
- B.Cenocepacia (B.Cepacia Complex Genomovar III) colonization.

SURVIVAL STATISTICS

Hearth/lung transplantation is extremely rare. Only a few centers in the world perform heart/lung transplants, and most of those patients are adolescents. In 2018, St. Louis Children's Hospital was the first center to successfully perform a heart/lung transplant on an infant in the last decade.

Due to the small numbers of heart/lung transplants performed in the United States, there are not currently SRTR specific reports for heart/lung transplants. Children's Hospital statistics show that our heart/lung transplant patient survival statistics are comparable to our lung transplant patient survival statistics, which are approximately 90% at 1 year and approximately 50% at 3 years.

These survival statistics are a reminder that heart/lung transplantation is not as successful as kidney or liver transplantation. Therefore, heart/lung transplant should be considered an exchange of one set of medical problems for another. In many ways, life after heart/lung transplant can be an emotional roller coaster, filled with uncertainty. The main complications that affect long-term survival, including Bronchiolitis Obliterans and post-transplant lymphoproliferative disease, are described at length in this teaching manual.

CADAVERIC TRANSPLANT

Where Do Donor Organs Come From?

Organ donors are individuals in whom all brain function has ceased due to severe brain injury, (also known as "brain death") and consent has been obtained for organ donation from family members. The evaluation of potential organ donors includes their medical history, blood tests, and studies looking at function of the specific organs (e.g. heart, lungs, kidneys, liver and pancreas). These tests are done by protocol at the site where the donor is located. In most cases, the tests are performed twice, with several hours in between to ensure an accurate result.

Brain death should not be confused with coma or vegetative state. A patient in a coma is medically and legally alive and may breathe on their own when the ventilator is removed. A patient in vegetative state retains motor reflexes, and has a natural sleep-wake cycle, but is not aware of any activity. Organs are only retrieved from donors who have suffered brain death.

The lungs in brain-dead donors are particularly vulnerable. Function may rapidly deteriorate due to fluid buildup in the lung tissue, infection, or trauma. In only 20-25 percent of organ donors are the lungs healthy enough to be recovered for transplantation. Therefore, the number of donor lungs available is lower than the number of donor kidneys, livers, or hearts.

REFERRAL PROCESS

The majority of our referrals come from a primary physician or medical specialist caring for the child. Information is gathered from the referring physician. This information includes a clinical summary, trends in laboratory results, PFTs (Pulmonary Function Test), blood gases and/or cardiac catheterization reports, surgical notes, a complete patient and family evaluation by a social worker, and insurance information. The multi-disciplinary heart/lung transplant team reviews this information and decides if heart/lung transplantation evaluation is appropriate for the child and if so, an evaluation is scheduled.

FINANCIAL

Who pays for an organ transplant?

Many insurance companies now offer at least partial coverage for transplant costs although the terms and extent of coverage vary widely. The transplant team includes staff that is able to assist with exploring your transplant medical coverage. Additionally, they can assist with exploring your options for fund-raising or other financial assistance programs. These individuals will meet with you and your family during the transplant evaluation to help you understand the financial aspects of transplantation. There are instances when we experience long delays with authorization from out-of-state Medicaid agencies to bring a child in for evaluation. We make every effort to work with those agencies on behalf of the patient; however there may be instances when it is not possible.

Your insurance coverage or financial situation may change over time. It is extremely important to notify the financial coordinator or social worker of these changes.

It is the family's responsibility to check insurance benefits about the coverage provided for transplant surgery, hospitalization, and follow-up care. After your child has had a heart/lung transplant, health insurance companies may consider your child to

have a pre-existing condition and refuse payment for medical care, treatments, or procedures.

After the surgery, your health insurance and life insurance premiums may increase and remain higher. In the future, insurance companies could refuse to insure you. The Financial Coordinators at St. Louis Children's Hospital will discuss any financial concerns you may have.

MEDICARE APPROVED TRANSPLANT CENTER

St. Louis Children's Hospital is a Medicare approved transplant center. If your transplant is not provided in a Medicare approved transplant center, it could affect your ability to have immunosuppressive drugs paid for under Medicare Part B.

NOTIFICATION OF MEDICARE OUTCOME

Requirements not being met by Center

Specific outcome requirements need to be met by transplant centers and the transplant program is required to notify you if those requirements are not met. Currently, St. Louis Children's Hospital meets all requirements for transplant centers.

TRANSPLANT TEAM MEMBERS/ROLES

Transplant Surgeon – The transplant surgeon is a surgeon who has had additional specialized education and training in transplant surgery and medical management of transplant recipients.

The transplant surgeon will provide information to you and your child about how the transplant is performed and the potential benefits and risks of transplant surgery. Additionally, the transplant surgeon participates in the immediate post-operative management of the transplant recipient, including performing any necessary re-operations.

Transplant Medical Staff – We have a specialized team of heart/lung transplant physicians dedicated to caring for children undergoing heart/lung transplantation. The transplant physician coordinates the overall functioning and policy making of the heart/lung transplant program, including pre- and post-operative care. You and your child will meet with the transplant physician during the evaluation process. Your child's medical history will be reviewed and a physical exam will be performed. Information will be provided regarding the potential risks and benefits of transplantation. As well long-term outcomes will be discussed. Other transplant medical staff

members may be caring for your child during their transplant hospitalization. These attending physicians have had extensive training in pulmonary and cardiology medicine and supervise fellows and house staff physicians. A fellow is a pediatrician who is specializing in pulmonary and cardiology medicine and is very important for your child's care during the nights and on weekends.

Pediatric Nurse Practitioners are nurses who have an advanced level of education and experience. They have a master's degree in nursing and are trained as nurse practitioners. They are licensed nationally both as registered nurses, and as nurses in advanced practice. They are board-certified by the state of Missouri to practice as pediatric nurse practitioners. Nurse practitioners work under the direction and supervision of your heart/lung doctors.

Some of the responsibilities of the nurse practitioner:

- Take care of children in the clinic or the hospital
- Perform physical exams
- Write immunosuppression orders/write prescriptions
- Diagnose minor illnesses in children and prescribe treatment
- Perform procedures such as suture/staple removals and drainage tube removals
- Monitor the side effects of immunosuppression medications and provide necessary treatment
- Admit your child to the hospital if needed
- Educate children and families about their disease
- Do extensive discharge planning with local physicians, hospitals, and home health agencies
- Contact your local doctor and communicate the plan of care
- Coordinate your child's care among all services involved (transplant surgery, other specialists)
- Work closely with other team members (transplant coordinators, social worker, psychology, child life specialist, dietitians, pharmacy, etc.)
- Refer your child to another specialist if needed (dermatology, allergist, etc.)
- Ensure a smooth transition between inpatient and outpatient care

Transplant Nurse Coordinators are nurses with experience in caring for transplant patients.

They will teach you about your child's disease, transplantation, and how to care for your child after transplantation. Your transplant coordinator will be your main contact with the transplant team. They work under the direction and supervision of the doctors and nurse practitioner. Some of the responsibilities of the transplant nurse coordinator are:

- Educate children and families about their disease.
- Coordinate the heart/lung transplant evaluation including scheduling tests and consultations with members of the transplant team (surgeons, medical doctors, social worker, etc.).
- Add your child onto the national heart/lung transplant waiting list.
- Make the necessary arrangements at the time of heart/lung transplant including contacting you about an organ being available, arranging for OR time, contacting the hospital staff and transplant team members about your child's admission, etc.
- Educate children and their families about post-transplant care including medications, signs and symptoms of rejection and infection, follow-up visits, etc.
- Do extensive discharge planning with local physicians, hospitals, and home health agencies.
- Work closely with all other team members (surgeons, medical doctors, nurse practitioner, social worker, etc).
- Perform telephone triage of any illnesses your child may experience and refer to the appropriate team member (medical doctor, nurse practitioner, local physician).
- Participate in your child's outpatient clinic visits (review medications, provide ongoing education, coordinate visits/referrals to other services).
- Perform procedures such as staple or suture removal.
- Contact your local doctor and communicate the plan of care.

Child Life Specialists help children and adolescents cope with the stress of the medical setting and treatment by providing developmentally appropriate social, emotional and educational support and activities. Services include:

- Providing materials and guidance for play and adapting activities according to a child's strengths and limitations.
- Preparing children for hospitalization, surgery, and medical procedures through hands-on activities and providing support during procedures.
- Emotional support to children, siblings, and parents.
- Advocating for an age-appropriate and child-focused environment and treatment experience.

Psychologists – Staff from the St. Louis Children's Hospital Department of Psychology specialize in the psychological issues associated with chronic illnesses and disease. They work directly with children and families and provide consultation to the medical team. New patients are seen by a psychologist during the transplant evaluation. After this initial evaluation, follow-up services during hospitalization and/or on an outpatient basis may include:

- Emotional support and counseling.
- Individual therapy with patients, siblings, and/or parents
- Family counseling.
- Behavioral techniques to manage pain, anxiety, and/or reactions to treatment side effects.
- Interventions to assist with illness management.
- Medication compliance.

Social Workers – St. Louis Children's Hospital social workers are members of the diverse team who serve as a liaison between families and medical staff. During the transplant evaluation, a psychosocial assessment is completed by the social worker with the patients and their families to help the medical team begin to understand your family and any social concerns there may be. Social workers also assist you and your child with stress as well as the emotional and practical problems associated with your child's illness. Assistance is also available to connect you

with community resources that may be helpful to your family. Other services they can help with are:

- Advice about resources potentially available to families, including financial assistance and lodging alternatives for families from out-of-town.
- Assistance with acquiring those resources.
- Family support.

Chaplain – St. Louis Children's Hospital is committed to meeting the spiritual needs of all our patients. We have two full-time chaplains on staff. Please notify your transplant team if you wish to meet with the chaplain before or during your transplant hospitalization. We can provide a religious practitioner of your choice, i.e. Roman Catholic Priest, Rabbi, Mormon Elder, etc. upon your request.

- 24 hour on-call service. Call 314.454.6000

Registered dietitians are members of the team who provide recommendations to support adequate and appropriate growth both pre- and post-transplant. Registered dietitians work with physicians, nurse practitioners, nurses and other team members, as well as patients and their families to ensure adequate caloric intake and to promote balanced and varied diets. Services include:

- Meeting with patients and families during hospital admissions
- Clinic visits as requested by staff, patients or families
- Help in obtaining formulas or specific nutrition products for outpatients
- Assistance with any food allergies, intolerance, preferences, or special needs related to diet while inpatient.

Pharmacist – The transplant pharmacist is an important member of your transplant team. A pharmacist is a health care professional who consults with and sometimes advises the licensed practitioner concerning drugs.

Services include:

- Assisting the physician in medication selection and dosing.
- Monitoring and managing side effects
- Monitoring for adherence concerns; offering tools and alternatives as appropriate
- Educating patients and families on medications

Nursing Staff – Direct patient care for transplant patients will be provided by nursing staff that have special training in the care of transplant recipients.

ADHERENCE

Adherence is defined as how a patient follows through with medical advice given to them by their medical team. Your transplant team considers adherence very seriously because we believe it will affect your child's outcome after transplant. Transplant patients are required to follow a complex self-care regimen before and after transplant. Patients are expected to participate in and cooperate with medical recommendations throughout the transplant experience. Families are expected to support and monitor their child's self-care program as directed by the medical team. Different types of self-care and self-monitoring are required depending on your child's transplant. Your transplant team will outline adherence guidelines they would like you to follow.

POTENTIAL MEDICAL AND PSYCHOSOCIAL RISKS

Hearth/lung transplantation can have potential medical risks and psychosocial risks. Medical risks include a) wound infection, b) pneumonia, c) blood clot formation, d) organ rejection, failure, or re-transplant, e) lifetime immunosuppressant therapy, f) arrhythmias (slowing or fastening of the heart rate or heart rate skipping beats), g) cardiovascular collapse (heart attack), h) multi-organ failure, and i) death. Psychosocial risks include depression, post traumatic stress disorder (PTSD), generalized anxiety, anxiety regarding dependence on others, and feelings of guilt.

WHAT YOU CAN EXPECT FROM US

- We will treat you in a respectful manner
- We will make every effort to be on time for appointments with you
- We will return your calls within one business day
- We will be honest, up front and keep you informed to the best of our abilities
- We will not discuss your child with other families
- We will treat your child and family as individuals and tailor a plan of care to address your needs
- Hospital personnel who are involved in the course of your child's care may review the medical records. They are required to maintain confidentiality as per law and the policy of the hospital. If your child is accepted for transplantation, data about your case which includes identity will be sent to UNOS and may be sent to other places involved in the transplant process as permitted by law.
- St. Louis Children's Hospital is a teaching hospital, so there may be other doctors, nurses, and/or technicians in the operating room observing during the time of the operation and/or participating in post-operative care.

WHAT WE EXPECT FROM YOU

- We expect you to be on time for your appointments, biopsies, lab draws
- We expect you to get testing done when it is requested
- We expect you to call us with questions, and leave a message if we are not immediately available
- We expect at least 7 days notice to refill prescriptions and 24 hour notification of cancelled appointment or tests
- We expect you to keep us updated on phone/address/insurance information
- Upon relocation to St. Louis for transplant, we expect you to have a cell phone so that we may contact you when a lung offer is available.
- We expect you to have a working phone with voice mail.
- We expect you to discuss questions/concerns about your child with the transplant team and not other families
- We expect you to respect the privacy of other families

THE TRANSPLANT EVALUATION

The evaluation usually takes three to four days to complete. It is most often done in an outpatient setting. During the evaluation, the child and family will meet with members of the heart/lung transplant team. Teaching regarding all aspects of transplantation will be done including, but not limited to, risks and benefits, the operation, medications, long term outlook and survival, psychological issues, financial issues, social issues and commitment required of the patient and family.

Upon completion of the evaluation, the heart/lung transplant team will meet to decide whether the child is a suitable candidate for heart/lung transplantation. The transplant team will let you know the result of the heart/lung meeting. If the child is approved, prior to the procedure, the surgeon will discuss the operation and risks and you will be asked to sign a consent form for the operation. At all times, the child's health and safety will be the primary focus.

At any time during the evaluation process, or prior to transplant, you are free to decide for any reason your child no longer wishes to have a transplant.

Testing may include the following. Please refer to the glossary for definitions of the tests described:

- **Blood tests** – A number of blood samples are taken to evaluate your child's liver, kidneys, blood counts, prior exposure to certain viruses, blood typing and tissue typing.
- **Pulmonary function testing (also called PFT's)** – These tests are used to evaluate your child's lungs and airways. Different breathing maneuvers such as breathing deeply and rapidly, blowing out rapidly after taking a deep breath in, and breath holding are measured.
- **Chest X-ray** – Determines the size of your child's heart and lungs as well as how well your child's lungs inflate and if there is any fluid or infection in their lungs.
- **CT scan of the chest** – A special computerized X-ray of the chest, which gives a more detailed three-dimensional picture of the lung tissue and the lining of the chest wall or pleura.
- **Physical therapy evaluation** – Assessment of strength, posture and stamina. Measurements are taken of walking capability and oxygen level during six minutes of supervised walking.
- **Electrocardiogram (EKG)** – Records your child's heart rate and rhythm.
- **Echocardiogram (ECHO)** – Uses sound waves to look at the size of your child's heart and how the heart muscle and valves are working.
- **Cardiac catheterization** – Patients with pulmonary hypertension may require a cardiac catheterization. This test measures the specific pressures in the heart and lungs. Sedation is administered and a pediatric cardiologist inserts a catheter into the large vein or artery in the groin. The catheter is advanced into the heart. Pressure readings are recorded. If the pressures are high, medication may be given to see if the pressures can be lowered. The procedure is done in the "cath lab" and takes one to two hours. Your child will need to lie flat for at least six hours after the procedure to prevent bleeding at the catheter insertion point and may require an overnight hospital stay.
- **Clinic visit with the transplant physician and coordinator** – A brief clinical history will be taken. A physical exam will be performed. Your child's current medication regimen will be reviewed. You will meet some of the transplant team at this time. During a portion of the clinic visit parents of adolescents will be asked to leave the exam room. We believe that a teenager must feel as though they have some control over the transplant process and decisions regarding transplantation. This time will allow the teenage heart/lung transplant candidate an opportunity to ask questions they may not feel comfortable asking in the presence of their parents.
- **Neuropsychology and /or Developmental Testing** – (not applicable for infants and toddlers) developmental testing completed by the psychology staff to determine if your child is at his/her age appropriate development level. For children and teenagers, school performance and emotional and social development are evaluated.

At the end of your visit an evaluation review or “exit interview” will be scheduled. This is the time when the family and the heart/lung transplant physician and nurse can sit down and go over available test results from the evaluation. It also provides an opportunity for the family to ask questions which may have come to mind during the evaluation process. After you return home, the SLCH heart/lung transplant team will review the results of the studies and present your child to decide whether your child is a candidate for heart/lung transplantation. We ask that you go home and carefully consider all aspects of heart/lung transplantation, including the risks and the commitment required.

Because the evaluation typically runs from early Monday morning through late Wednesday afternoon we ask that you arrive in St. Louis on Sunday evening. A number of hotels and motels offer discounts to families of patients of St. Louis Children's Hospital. For assistance in making hotel reservations and for families interested in staying at the Ronald McDonald House, please call the heart/lung transplant social worker.

LISTING INFORMATION

Once your child is evaluated for heart/lung transplantation, we review all the medical information and present your child to the entire heart/lung transplant team where a decision is made regarding their candidacy for heart/lung transplant. Once your child is approved for listing by the heart/lung transplant team, we will call you and ask if you have made a decision whether you would like your child to be listed on the national waiting list. We will list your child only with your permission along with insurance approval. Insurance approval may take a few weeks, therefore we will call you when we actually list your child. Your child will be listed on the national transplant waiting list called UNOS. UNOS is a not-for-profit organization that maintains the nation's organ transplant list. UNOS brings together medical professionals, transplant recipients and donor families to develop organ transplant policy.

The heart/lung transplant candidate listing is the result of combining the lung requirements with the heart requirements. This is explained in detail below.

In addition to the heart/lung listing, your child will also be listed for heart transplant and a lung transplant in regulation with UNOS.

For listing a lung candidate age 12 and older they will receive an individualized lung allocation score (LAS). The system will determine the order of everyone awaiting a lung transplant by their LAS, blood type, and geographic distance between the candidates and the hospital where the lung donor is located. Lungs from pediatric and adolescent donors will be offered to pediatric and adolescent transplant candidates before they are offered to adults. Since 2010, every lung transplant candidate age 11 and under has been given a status score based on their medical urgency.

The status system for heart listing is:

Status 1A – A patient meets at least one of the following conditions:

- needs continuous help breathing with a ventilator and is admitted to the listing hospital
- has a balloon pump and is admitted to the listing hospital
- has a blood flow to the body or lungs that is dependent on a stent or prostaglandin infusion and is admitted to the listing hospital
- was born with significant heart disease and requires certain IV medications, known as inotropes, at high dose or more than one inotrope and is admitted to the listing hospital
- assistance of a mechanical device such as ECMO or a ventricular assist device (VAD). May or may not be admitted to the hospital

Status 1B –

- needs IV medications, known as inotropes, but does not meet Status 1A criteria
- is less than one year old and has a diagnosis of hypertrophic or restrictive cardiomyopathy

Status 2 – A patient is listed for a transplant but does not meet the criteria for status 1A or 1B.

RELOCATION INFORMATION

We have patients relocate to St. Louis when we believe they are likely to receive an offer. When your child has received one or more primary offers, your transplant physician and transplant coordinator will discuss having you and your child relocate to St. Louis and actively await heart/lung transplant. The decision to relocate is based on medical need, quality of life, frequency of hospitalization, and patient readiness to move forward.

PREPARING FOR THE CALL

There is no way to know when the heart/lungs will become available; however, there are things you can do to help prepare your child and family:

- Make a list of things you and your child will need at the hospital: comfortable clothes, toiletries, your child's favorite stuffed animal or toys, book/magazines to read, etc.

- Should your child develop a fever, infection, or contagious illness (i.e. chickenpox, influenza, etc.) while on the waiting list, contact your transplant coordinator regarding treatment. Your child's transplant may be temporarily postponed until your child has recovered.
- If you have other children, arrange childcare for them ahead of time.
- Have cash available for gas money, cab fare, etc., so you will be able to leave at a moment's notice.
- Call your transplant coordinator if your family or your child has any questions or concerns about proceeding with transplantation.
- Each family is expected to have a cell phone to carry with them 24 hours a day so that they can be contacted when a donor becomes available.

THE ACTUAL TRANSPLANT

When heart/lungs are offered to your child, the offer is made to the organ procurement organization (OPO) for St. Louis. The OPO for St. Louis is MidAmerica Transplant Services (MTS). A coordinator from MTS calls a heart/lung transplant coordinator at St Louis Children's Hospital. The heart/lung transplant coordinator collects pertinent information about the donor, such as cause of death, medical information, height, blood type etc. They call the surgeon, cardiologist and pulmonologist. The surgeon evaluates the information and decides whether or not to accept the organ offer. Many factors play into this decision, such as: donor or recipient stability, height match and distance to travel. Once our center has accepted the offer for your child, you will be notified by phone. This first call is often met with many emotions by you and your child. We will tell you during this first call that we have a "potential" offer for you, meaning we are very early in the process and the transplant could be cancelled, if the conditions warrant. The transplant coordinator will give you the necessary information about coming to the hospital. You will be told to stop feeding your child, to come to admitting or directly to the patient floor.

It is at this point the surgeon has a clear picture of risks associated with this particular organ versus the risk of waiting for the next available donor and can tell you specific recommendations. You always have the option to decline an organ.

Certain conditions in the donor may affect the success of the heart/lung transplant, such as the donor's history and the condition of the organ when it is received in the operating room for the transplant operation. Additionally, there is a potential risk of contracting HIV, hepatitis, and/or other infectious diseases if these infections cannot be detected in the donor.

The nurses on the patient floor will be prepared for your arrival. Your child will have required blood tests, chest X-ray and other necessary tests to assure that there are no current medical reasons not to proceed, such as an active infection. While you are on the floor, the waiting can be difficult and anxiety producing. We encourage you and your child to get as much rest as possible. The coordinator will keep you updated as much as possible by phone as to the timing of when the donor heart/lungs are due to arrive at the hospital. The timing is dependent on many factors;

one important factor is that other medical centers may be sending physicians to retrieve organs from this donor and attention is paid to this process. Our surgeons always travel to where the donor is located to inspect the heart and lungs both inside and outside. Your child will go to the operating room at least two hours before the donor heart/lungs arrive here. There are times when the condition of the donor or recipient warrants canceling the transplant. The physicians make this decision, and it is not done without careful consideration. If the transplant is cancelled, this is called a "dry run" and is very disappointing for you and your child. In the event of a cancellation, you will be discharged from the hospital and resume normal activities. Your child's listing status will not change.

Should the transplant proceed, you will be able to stay with your child until the time they go into the actual operating suite. The cardiothoracic surgeon and anesthesiologist will speak with you and your child before they go to the operating room. They can answer any last minute questions for you. The cardiothoracic surgeon will explain the operative procedure, risks, etc., with you during your child's evaluation for listing and again the day of transplant.

Your child will be in the operating room approximately six to eight hours on average. The operating room nurses will provide you with updates as often as possible. Your child may bring a camera to the OR if they would like to have photos of the surgery.

During the transplant operation, your child will receive general anesthesia, which means your child will be given medications to put him/her to sleep, block pain, and paralyze parts of his/her body. A machine (ventilator) to help with breathing will also be used. The anesthesiologist will talk about the risks of anesthesia.

The transplant surgeon will make an incision between the ribs or through the breastbone. Through this incision, your child's heart and lungs will be removed, and the donated heart/lungs will be placed into the chest. Your child will likely need blood or blood product transfusions during the surgery. There is a separate form for consenting to transfusions. You will be asked to sign this consent before the operation.

Once your child is in the operating room, you can expect the organs to arrive within two to three hours. When the surgery is finished, your child will be taken to the Cardiac Intensive Care Unit (CICU) in the Heart

Center, where they will be stabilized and you will be permitted to see them. Your child will be sedated and have many tubes visible to you. This is often difficult for you as the parent, but remember that your child is sedated and will have adequate pain management.

Our transplant team is here to provide the best care possible to all our patients. Careful screening of organ donors is always done before we call any patient or family with an organ offer.

The chance of an infection from a donor organ is very low, but not zero. Examples of possible infections are undetected Human Immunodeficiency Virus (HIV), Hepatitis B Virus (HBV), and Hepatitis C Virus (HCV) infection. If one of these infections is found, there are treatments that can help. Our team will let you know the chance of an infection at the time of organ offer. We will be there to answer questions.

Testing is done before and after transplant to screen for HIV, Hepatitis B, and Hepatitis C. Any positive results will be told to patients and families. If you decide not to accept an organ from a donor that has a possible infection, this will not affect your care with the transplant program, and it will not change your position on the waiting list.

YOUR STAY IN THE HEART CENTER

Your child will go to the Cardiothoracic Intensive Care Unit (CICU) in the Heart Center immediately following surgery. The intensive care team will require an hour or more to stabilize your child before you will be allowed to visit. Your child will be on a respirator (also known as a ventilator, or breathing machine) and will have many monitoring lines and drainage tubes present. While on the respirator your child will have a tube in his throat (called an endotracheal tube) and will not be able to speak. Your child will have four large tubes in their chest (called chest tubes) to allow drainage of fluid to help re-expand their lungs, and help healing. There will be a tube in the nose that goes to the stomach (called a nasogastric or NG tube). This tube helps to keep air from accumulating in the stomach and helps to drain stomach acid. This tube is usually removed when the breathing tube comes out. A urinary tube will be in place for several days after the transplant. Special mechanical boots or sleeves around the legs may be used to keep blood flowing through the legs to prevent blood clots. Vital signs, such as blood pressure and heart rate will be monitored closely. Lab tests will be drawn frequently

from an arterial line (A-line). This special line (a type of IV that is actually placed in an artery rather than a vein) is placed during the surgery. The A-line not only allows for blood draws, but also helps to monitor blood pressure. The ICU team will keep your child as comfortable as possible. As your child recovers, the tubes and monitoring lines will be removed. Each child is unique and the length of time your child is in the CICU will depend on many factors.

Please check with your nurse regarding the visitation policy. We realize that there will be many concerned and relieved family members and friends, however **your child is vulnerable to infection and we strongly discourage visiting by all but immediate family (parents, guardians or appropriate siblings) at this time.** All visitors should not have any signs or symptoms of illness. Good handwashing by all visitors is strongly advised to protect your child. The staff can only give patient information over the phone to parents. It is suggested that you designate a friend or family member as a contact person to receive updates from you.

Your child will be in an isolation room. Staff having direct contact with your child will wear a mask, gloves and gown. All individuals entering the room should wear a mask regardless of whether direct patient contact is made. The family will also abide by isolation protocol.

Within the first 24 hours of transplant, a perfusion scan and a bronchoscopy will be performed. During a bronchoscopy a small scope will be passed into your child's airway and into the lungs. The scope has a small video camera lens on the tip. The physician can actually check the airway and surgical connections for any signs of redness, swelling, narrowing and for the presence of mucous. Approximately one week after transplant a bronchoscopy and biopsy of the heart and lungs will be performed. During a biopsy, a fine

wire will be passed through the bronchoscope. The wire has very small pinchers on the tip. Approximately six to ten pieces of lung tissue will be "snipped" from the transplanted lungs and sent to the pathology department to look for microscopic evidence of rejection and infection. Typically the first heart biopsy is performed one week after the transplant along with the lung biopsy.

Your transplant team will work together with the Heart Center team of doctors and nurses to provide your child with the best possible care.

During your stay in the Heart Center, the nurses will educate you and your child about transplant medications and blood pressure measurements. It is important to become comfortable with the medications and equipment needed for home care. Your transplant coordinator can also serve as a resource person during this educational process.

TRANSITIONING TO OUTPATIENT SERVICES

Preparations for discharge home begin after your child is transferred out of the CICU to the Transitional Care Unit (TCU). It will seem like an overwhelming prospect: there is a lot to learn and you will be concerned about your child's new vulnerability. Most parents feel exhausted by the time their child is ready for transfer to the floor. During the time on the TCU, it is important that family members reestablish a routine that allows time to take care of themselves, including getting plenty of sleep.

Once your child is transferred to the TCU, your coordinator will arrange several sessions to teach you what you need to know about caring for your child. Please be assured that we will not discharge your child from the hospital until they are medically stable, and you feel comfortable with the new responsibilities related to your child's care.

POST- TRANSPLANT

Once your child has been transplanted, their progress will determine when they are released from the hospital. This varies with each person, for all cases are different. You will be asked to remain in St. Louis for a three-month period following transplant. This period is very important for several reasons. During this time, teaching will take place to enable you and your child to perform all aspects of required care and home monitoring. Also during this time, we will closely monitor your child's progress, with recommendations being made to help achieve positive outcomes.

Once your child has stabilized after their transplant surgery, the transplant nurse coordinators will provide you with written instructions concerning how to care for your child after transplant. It will include the following information that is located in this booklet:

- Signs and symptoms of rejection
- Signs and symptoms of infection
- Information about medications your child will be taking post-transplant
- Reasons to call the transplant nurse coordinator or transplant physician
- General health information about immunizations, dental health, skin care, etc.

The transplant nurse coordinator will arrange times to meet with you and your child (if appropriate) to review all of these instructions. If there are other family members (i.e., grandparents, stepparents) who will be taking care of your child, you may want to have them included in the teaching sessions, especially if they will be administering medications to your child.

The staff nurses will also be working with you throughout your child's hospitalization to teach you any special skills you may need to take care of your child. Some of these skills include:

- How to take a blood pressure
- How to administer medications
- Wound care and dressing changes

Arrangements will be made with a home health agency for any equipment or additional home nursing care that your child may need.

Once you are released from the hospital and living locally, your child will be required to come to the hospital two times a week to be seen in transplant clinic. Your child will also perform spirometry (PFTS). Your child will be required to participate in physical therapy at least three times a week for the immediate post-transplant period. This therapy is essential in helping your child recover from surgery and resume activities of daily living. As your child progresses, your visits here will be less frequent, and as you approach discharge from St. Louis, you will be seen in clinic once a week or two times a month. Again, this is all individual and depends on your child's progress.

SCHEDULED ROUTINE EVALUATIONS

We ask that you respect the fact that we schedule many families after transplant, and that flexibility in scheduling is not always available. We ask that you give special attention to trying to keep scheduled appointments. We realize that it is often hard for your child to miss school, etc., while they are in St. Louis for the required testing, but this is inevitable at some times. Please call four months in advance to schedule your child's appointment. Availability depends on holidays, doctor's schedules, appointment availability, and availability of testing equipment (X-ray etc.). Schedules are not finalized until the bronchoscopy is scheduled. Appointments are subject to change if an emergency occurs.

During the first three months, the bronchoscopies, heart biopsies and testing will be set up automatically and you will be notified of your child's scheduled tests.

It is the families responsibility to arrange all physical therapy appointments.

At two months post-transplant a pH probe and V/Q scan will be performed. Please refer to page 58 for further explanation of a V/Q scan. Please refer to page 57 for further explanation of pH Probe.

Evaluation procedures that will be completed

3 months post-transplant, every 3 months the first year post-transplant, and every 6 months thereafter. These procedures include:

- Clinic visit with physician and nurse coordinator
- Bloodwork

- High resolution chest CT
- Chest X-ray, EKG, Echocardiograms
- Ventilation Perfusion Scan
- Physical Therapy
- Pulmonary Function Testing (see page 21 for definition)
- Bronchoscopy with transbronchial biopsy (this may be deferred after the 18 month visit) (see page 23 for definition)
- Bone Densitometry (once a year) (see page 53 for definition)
- A cardiac catheterization with coronary angiogram is at the first year anniversary after the heart/lung transplant and then every two years or as needed.

A cardiac catheterization shows precise assessment of the function of the heart. To perform the procedure, a small area of skin over a vessel (usually the right femoral artery in the groin area) is numbed with a local anesthetic and a catheter (long narrow plastic tube) is advanced through a small skin puncture, with fluoroscopic assistance, through the body to the base of the heart. The catheter tip is inserted into the origins of the right and left coronary arteries (one at a time) and dye is injected. As the dye flows down the arteries and opacifies them, movies (angiograms) are made and, when developed, demonstrate any areas of narrowing in the walls of these vessels. The catheter is also advanced through the aortic valve into the main pumping chamber, or left ventricle, where an injection of dye allows filming of the contours of this chamber as it beats. Both coronary and cardiac angiographic studies are compared every other year to detect any changes that may have occurred.

There may be an occasion when additional testing is required or ordered by your physician. You will be notified of this.

Scheduling can be done by phone or by email. Patients should call the Ambulatory Procedure Center (APC) or Same Day Surgery unit the Friday prior to the evaluation to ask questions or prepare for any sedated procedures.

The three-month evaluation will be automatically set up by the post-transplant scheduler.

Before the patient leaves to return home, the scheduler will meet with the family to answer any questions regarding scheduling and to provide the information to contact the scheduler. Patients return for follow-up evaluation every three months the first year and every six months thereafter.

HIGHLY IMPORTANT: All patients need to call the pre-arrival number and post-transplant scheduler when any personal information has changed, such as insurance information, address, name, phone numbers, etc. This information is needed to receive authorization from your insurance company. Without the proper information, approval for the evaluation will not occur and you will be responsible for the medical charges.

EXERCISE

Daily exercise is strongly recommended post-transplant. Exercise helps to maintain weight, reduce loss of calcium from bones, improve the proportion of muscle to fat body stores, lower blood pressure and reduce stress. In addition, exercise may lower blood pressure and help reduce stress. Physical activity should essentially be unrestricted with the exception of avoiding contact sports such as football and rugby. Your child will probably feel more tired than usual for one to two months after surgery. There will be a healing period of approximately four weeks during which your child should not attend school or work or lift more than ten pounds. He/she also should not lift more than 20 to 25 pounds, participate in gym class, or exercise excessively until three months after surgery or until the doctor allows it.

DIET

Diet recommendations will depend on your child's disease process and nutritional status. Prednisone may increase your child's appetite and thus your child's weight. We will follow weight gain and weight loss in clinic. A dietitian will be available for recommendations and concerns. Older children will benefit from drinking plenty of water or non-caffeine type drinks to keep their kidney function at an optimum. Remember, several of the medications your child will be taking post-transplant are hard on the kidneys. Dehydration can be harmful to the kidneys; drinking plenty of fluids will be beneficial.

SKIN CARE

Skin problems (usually minor) are very common in transplant patients. Patients who take immunosuppressant medication are at an increased risk for skin and lip cancers. There are things you can do to help decrease your child's risk.

- Your child needs to apply lip balm and sunscreen lotion with a SPF (sun protection factor) of at least 30 **every day, rain or shine**, April through October. Apply the lotion to all exposed areas, especially the face, ears, neck and hands. Remember that sunscreen lotions wash off. Reapply the lotion as needed, especially after swimming.
- Avoid midday (10 a.m. to 3 p.m.) sun when ultraviolet rays are the strongest.
- Wear a hat and long sleeves when outdoors.

Oily Skin or Acne: Your child, especially in pubertal or post-pubertal stages, may develop acne on the face, chest, shoulders, or back. Acne post-transplant is mostly due to the medication Prednisone. Primary measures used to control the acne are aimed at removing the excess oil and preventing formation of "white heads" or "black heads". Wash the areas of acne three times a day, scrubbing gently with a soapy washcloth to remove the oil, dead skin and bacteria. Neutrogena soap works well, but any drying soap is fine. Rinse soap off the skin completely to leave pores open and clean. Use a clean wash cloth each time. Shampoo hair and scalp frequently and keep the hair away from areas of acne. Benzoyl peroxide cream or lotion (5-10 percent), can be purchased over the counter and is helpful in drying the acne. When acne is present, it is best not to use cosmetics, but if you must, use them sparingly. If the skin becomes dry, consider reducing the frequency of washing and application of medication. Remember that drying the skin is the objective of the treatment. Do not apply skin lotions to treat dry skin. If acne persists, we will refer you to a dermatologist. Please check with your transplant nurse or doctor.

Dry Skin: Use a mild soap like Dove for bathing and apply body lotion after bathing if your child has problems with dry skin without acne. Keri Lotion, Curel, and Moisturel are good over the counter lotions.

Skin Lesions: Warts may be particularly difficult to treat after transplant since they are caused by a viral infection. Moles that are changing, raised

skin lesions, or sores that do not heal, should be brought to the attention of the transplant nurse or doctor. We will refer you to a dermatologist for these problems.

Cuts and Scratches: Prednisone can cause the skin to become thin, making it tear, scratch, or bruise easily. Keep the skin clean so such areas do not become infected. Keep minor cuts and scratches clean and dry by washing with soap and water. Cleanse with hydrogen peroxide if desired. For large cuts, animal bites, or cuts that appear infected (redness, swelling, pus, tenderness), contact the transplant nurse. Antibiotic therapy may be indicated.

HAIR CARE

Prednisone and Tacrolimus will affect the condition of your child's hair. Older children may want to avoid tints, dyes, bleaches, and permanent wave solutions because they may make your child's hair break. We recommend that your child wait until their Prednisone dose is less than 20mg/day before they have a permanent wave or other hair treatment.

DENTAL

Dental visits are recommended every six months. Wait until six months after transplant to schedule the first visit. Your child will require prophylactic antibiotic therapy before any dental cleaning or work. The mouth is a warm, moist area where bacteria exist in high numbers. Bacteria may enter the blood stream during dental work, therefore we recommend following the "American Heart Association Guidelines for Dental Prophylaxis."

Good mouth care, including brushing teeth, tongue, palate, and flossing should be done regularly to prevent yeast infections in the mouth and painful swollen gums. Overgrowth of gum tissue on the teeth (called gingival hyperplasia) is a side effect of Cyclosporine and seems to be exacerbated by poor oral hygiene. Oral surgery to cut the gums back off the teeth may be necessary in some cases.

EYES

We recommend yearly eye exams by a pediatric ophthalmologist because your child is susceptible to cataracts and glaucoma from long term use of Prednisone.

GYNECOLOGY

Adolescent girls should have a yearly gynecological examination. St. Louis Children's Hospital Adolescent Medicine offers comprehensive services for adolescent special needs. Please discuss with your transplant coordinator if you would like a referral.

IMMUNIZATION INFORMATION

Vaccines are developed to prevent certain illnesses and contain either live or killed organisms.

Because your child's body has been purposefully immunosuppressed to prevent rejection, your child is at risk for infection. The introduction of live organisms through immunizations/vaccines may lead to an illness similar to that which the medicine was intended to prevent. Following a transplant, **NO LIVE VACCINES** should be given.

It is important that we receive a list of vaccines that you or your child has had during the evaluation and listing process. The Immunization Action Coalition provides an updated handout on the Internet detailing vaccines your child should have received and which ones MAY be due. This website is updated yearly or as new regulations occur. The website address for the Immunization Action Coalition is www.immunize.org. There is no copyright approval necessary; you may print directly from the website for updated schedules. This should be used as a guideline only. Your child's immunization schedule may differ.

DO NOT GET THESE VACCINES POST-TRANSPLANT:

- Mumps, Measles, Rubella (MMR)
- Polio (oral) or OPV
- Varicella (Chickenpox)
- Yellow Fever
- Typhoid (oral)
- BCG
- Rotavirus
- Flu Mist (influenza vaccine live, intranasal)

YOU MAY GET THESE VACCINES POST-TRANSPLANT:

- Tetanus
- Diphtheria, Pertussis, Tetanus (DPT)

- Polio (injectable) or IPV
- Hemophilus influenza B (HIB)
- Hepatitis B (Hep B-series of three)
- Flu vaccine
- Typhoid (injectable)
- Pneumonia vaccine – Pneumovax and Prevnar
- HPV
- Meningococcal

While not a vaccine, your child may receive the:

- Mantoux Tuberculin skin test
- Synagis

Keep in mind that if immunizations are given too soon after transplant (a month or so) the vaccinations may not take effect as they would later when your child's immunosuppressive medicines are not as high. We will make vaccine recommendations after transplant. We recommend a flu shot in the fall. We suggest children under two years of age receive Synagis.

SMOKE

Your child must stay away from all forms of tobacco smoke. Smoke causes inflammation within the lungs whether first-hand or second-hand, making it easier to develop a lung infection. Your child should stay away from smoked filled areas as much as possible. If family members smoke they should smoke outside of the home and the family car.

MEDIC ALERT JEWELRY

We advise all transplant recipients to wear a medical identification bracelet or necklace. Information about Medic-Alert jewelry can be obtained by calling 800.ID.ALERT, or ask your transplant nurse for an application.

We suggest the following information:

- List the type of transplant your child received, "Immunosuppressed", and any allergies. Include St. Louis Children's Hospital, 314.454.6000 and your local primary physician in the emergency contact section.

PETS

If you have a pet, it is important that it visits the veterinarian on a regular basis. Your child should

wash his hands after touching or playing with your pet. The child should never change kitty litter because of the potential infectious agent toxoplasmosis. We recommend against birds as pets because of the potential airborne infectious agents in their stool. Some reptiles carry salmonella in their stool, therefore if your child handles such animals they need to wash their hands thoroughly afterwards.

TRAVEL

When packing for your vacation, make sure you have an adequate supply of your child's medications, including some extra in the event you experience delays in your travel plans. If you are flying, carry the medications with you on the plane. Make sure you carry your child's insurance card and the phone numbers to the hospital in the event your child becomes ill.

If you and your child are traveling outside of the U.S., you will need a letter from your child's transplant physician verifying all of your child's medications to show with their passport as they go through customs. Use caution in what your child eats and drinks. For instance, only water and ice from adequately chlorinated sources can be considered truly safe. If you are unsure, it's best to have your child drink only bottled or canned water and soft drinks. Select food with care to avoid illness. Avoid unpasteurized milk and milk products and eat only what can be peeled or has been cooked and is still hot. If your child needs medical care while abroad, contact your travel agent, American Embassy or Consulate for names of physicians or hospitals.

TRANSITIONING TO AN ADULT CENTER

Once your child turns 21 years of age, we feel that they are best served in an adult transplant unit. We can help you identify a reputable transplant center if you wish. When your child is ready to transfer we will schedule a final appointment with us at St. Louis Children's Hospital. We will have you sign a release of medical information form so we can forward your child's medical records to the new transplant center. Once the adult center has received a copy of your child's records, we will have you make your first appointment. We will continue to follow your child until they have been seen at the adult center so that there is no interruption in your child's care.

As your child moves toward adulthood it is our responsibility as health care providers and your

responsibility as parents to prepare them for transition into the adult world of medicine.

We have outlined critical milestones for patients to achieve prior to transfer to adult care and will work with you and your child to meet these important goals. We realize this time of transition to independence is a complex process and will take several years. Outpatient clinic appointments will help your child develop skills in communicating with medical providers, and therefore at approximately age 14 we will ask that your child be seen alone and then invite the parent into the clinic room to review the plan of care. We realize maturity and developmental readiness is a better indicator of capabilities versus chronological age and will work with each family individually. As your child turns 18 years old they will be seen alone, and you may join him/her at the end of the visit if they consent.

Critical Milestones:

- Understanding of and ability to describe original cause of organ failure and need for transplant.
- Awareness of long and short-term implications of the transplant condition on their overall health and other aspects of their life:
 - Infection prevention
 - Routine surveillance
 - Academic and vocational goals
- Comprehension of the impact of their illness on their sexuality and reproductive health
 - Impact of pregnancy on their own well being
 - Effect of medication on fertility and potential teratogenicity
 - Role of genetic counseling
 - Risk of STD
- Demonstration of a sense of responsibility of their own healthcare
 - Knowledge of medications: indications, doses
 - Call for own prescription refills
 - Prepare own medication boxes
 - Independently communicate their own healthcare needs to their providers

- Know when and how to seek urgent medical attention
- Ability to make, keep, and follow through with their own health care appointments.
- Understanding of their medical insurance coverage and eligibility requirements.
- Verbalize importance of adherence to medical regimen
- Capacity to provide most self care independently
- An expressed readiness to move into adult world
- Ownership of their medical information in a concise portable accessible summary

PULMONARY FUNCTION TESTS

One of the most important ways to understand how well the lungs are functioning is to perform pulmonary function tests, often called PFTs. This is not just one test, but a series of tests which measure the size of the lungs, how fast air can move in and out of the lungs, and how well air (oxygen) travels to the blood supply. Not only are pulmonary function tests important for evaluation before a lung transplant is performed, but they are critically important after lung transplantation. PFTs cannot be used to make a diagnosis like CF, or pneumonia, or asthma. They do, however, help your doctor know if the lungs are functioning normally, or better or worse than previous tests. They are a simple, non-invasive way to trend lung performance. PFTs are, therefore, tests that are repeated quite often, and the trending of data is closely monitored. Most insurance companies no longer cover the cost of the home spirometer equipment needed. We ask that each family be prepared to cover the cost of this piece of equipment. You may speak with the financial coordinators and/or social worker to discuss any questions you may have regarding this cost.

To better understand what the various PFT measurements involve, descriptions are provided below:

Spirometry – This test is usually performed on every visit to the hospital or doctor. The purpose is to tell your doctor how big a breath your child can inhale and exhale, and how fast the lungs empty. Think of blowing up a balloon as far as possible. The bigger the balloon gets, when released, the more air that comes out and the faster the balloon deflates. The volume and speed of air is measured by a flowsensor that is placed near the mouth. A filter is placed on the flowsensor to trap viruses or bacteria from the mouth. Soft nose clips are placed on the nose to prevent any leakage of air

in or out of the nose. The measurements made are compared to the PFTs of children that do not have any lung problems. If your child has obstruction of their airways due to mucus, inflammation, or spasm of the bronchioles, it is more difficult to blow air out quickly. Think of trying to blow as hard as possible through a narrow straw. It is very important for children to learn how to perform this test properly and with maximal effort, and for parents to learn also. The patient will be asked to repeat this test several times (five to eight times) during each session to insure the best possible effort is recorded. The lab technologists help teach the patient and parents what the most important parameters in the test mean and what the graph should look like. This spirometric tracing, or flow-volume loop, gives valuable information in a visual form, and is sometimes easier to understand than the results in numerical or percentage form. Before transplantation, spirometry is performed with each physician visit. Following transplantation, this test is performed once or twice a week for the three-month period until discharge home. A spirometer is provided for each child to take home with complete instructions from a PFT lab technologist about the equipment. Hopefully, all the practice performing spirometry prior to 'going home' prepares the patient and parent. As with measurements of blood pressure, temperature, oximetry, etc, spirometry needs to become a routine part of the day. The child is initially asked to "blow PFTs" twice a day for approximately one year after transplant. Depending on how well the patient is doing, the physician may then adjust the frequency of testing. It will be necessary, however, for the patient to continue to perform spirometry throughout his/her lifetime, so its importance cannot be overstated.

Plethysmography or 'The Body Box' – A body plethysmograph is a scientific name for a large see-

through chamber that your child will be asked to sit in and perform more breathing tests. We call it the **body box**, because your entire body sits in the chamber, and it is a much easier term to use. Nose clips are again used, but the mouthpiece appears more like that of a 'scuba-diver's mouthpiece'. The measurements made in this chamber add more information to the spirometry previously obtained. As explained above, during spirometry, the child blows out as much as possible. The lungs, however, do not completely empty, or they would collapse and be very hard to re-expand. The body box indirectly measures the volume of air that is remaining in the lungs after exhalation. This is accomplished by pressure changes within the chamber and at the patient's mouthpiece. Since very small changes in pressure are recorded, the body box must be completely closed during the test. This is rarely a problem for the child. Most children do not experience claustrophobia. A microphone in the box allows the child to hear instructions or to speak to the technologist. The door is opened after each test, and the child can relax for a moment, or put an oxygen cannula on as needed. During the initial visit to the lab, the plethysmograph, or box, **looks** much scarier than it really is. Every effort is made by the technologist to first gain the child's confidence, and not force the child if he or she is fearful. Once the child realizes he or she can successfully do PFTs without fear of being hurt, the testing proceeds very easily. Measurements in the body box are not made during each visit, but are part of a **full** evaluation performed pre-transplant and with each complete evaluation post-transplant (3 months, 6 months, 9 months, 12 months, 18 months, 2 years, etc).

Diffusion capacity – This PFT is a measure of gas transfer in the most distant portion of the lung, where oxygen diffuses into the bloodstream. The patient is asked to take a maximal breath of a special test gas, hold his/her breath for ten seconds, and then smoothly blow the breath out. This can be a difficult test for young children because they have to follow instructions very carefully. However, with practice, this test can be successfully accomplished. The DLCO is also a test that is performed with each full or complete evaluation.

Arterial blood gases (ABG) – This is the one test performed in the PFT lab that does not involve breathing or blowing into a machine. It involves taking a small sample of blood (approximately 1 teaspoon) from an arterial blood vessel. Several arteries are

available; however, the easiest is usually the radial artery. It is important to understand that this sample comes from an artery and NOT a vein. Therefore, the venous blood that may be drawn at a different time during the evaluation cannot be used for this test. The amount of oxygen present in arterial blood is much higher than the oxygen level in venous blood. Additionally, carbon dioxide levels are lower in arterial blood. Obtaining an arterial blood gas sample is the most accurate way to determine the levels of oxygen and carbon dioxide in the body. The procedure, like any blood draw, can cause pain and discomfort. The procedure is saved for the end of all the PFT testing. It is not unusual for a child to get upset and cry when faced with a needle stick. The lab will attempt to minimize any discomfort by applying a local anesthetic (EMLA cream) and/or injecting 1 percent lidocaine just under the skin. The choice of anesthetic is up to the patient. In our hospital the technologists in the PFT lab are most experienced in drawing arterial blood. Therefore, the sample is usually drawn during the PFT evaluation. Arterial blood gases are ordered as part of pre-transplant evaluation, but are not routinely performed after transplantation. Your child's physician may request an ABG whenever oxygen or carbon dioxide status is in question.

Infant Pulmonary Function Tests – Our smallest patients, newborns through toddler age, can also have pulmonary function tests performed. They are not capable of following instructions so an alternate method of obtaining pulmonary functions is necessary. This requires sedation of the infant and more complex techniques to obtain pulmonary function results. Infant PFTs may not be attempted prior to transplant if your child is very ill and at increased risk from sedation. Infants (post lung transplant) are generally sedated in the APC, or Ambulatory Procedure Center. Often several procedures for follow-up (V/Q scan and CT scans) that also require sedation are scheduled on the same day. There are several possible options for sedation. The choice of sedating agent is based on the child's age, medical condition, and at the anesthesiologist's discretion. A sedation nurse and/or anesthesiologist will administer the sedation and remain with your child throughout all testing procedures and recovery. Although parents are permitted to observe many procedures, the PFT lab requests that parents **NOT** be present for infant PFTs. The technicians, nurses and doctors need to closely monitor your child, and this is best accomplished if

distractions are kept to a minimum. Additionally, the equipment is very sensitive to air currents, and even movement around the room must be minimized. Parents may sit in a waiting room adjacent to the testing room and are welcome to view the equipment before or after the child arrives.

A mask and putty seal is applied to the baby's face and remains in place throughout all the testing. The infant receives fresh air (and oxygen if necessary), and breathing is monitored at all times. For a short period, a see-through plexiglas tube slides over the baby's body to make measurements of lung volume. This method of lung volume measurement is called whole body plethysmography, and is very similar to the technique performed in older children. The child is observed through the glass for any movement, and this tube is retracted once the measurements are complete. A second type of test involves placing an inflatable air bladder on the child's chest that is kept in place by a cloth vest. During this procedure, the child takes several deep breaths, followed by a rapid exhalation as the air bladder inflates compressing the chest. This procedure is not at all harmful, and actually relaxes the child and improves oxygen levels. Remember, your child is asleep during this entire test. If necessary, an albuterol treatment may be given to your child and testing repeated. Total testing time takes approximately one to 1-1/2 hours. Results of the PFTs will be available from your doctor.

It is important to understand that not all children are capable of performing the PFTs discussed above. We strongly encourage the child to try to the best of his or her ability, and we accept and utilize whatever measurements or information we obtain. We understand the stress that your child and family are experiencing, and will try to accommodate requests that are reasonable and do not interrupt flow of patients in the lab. Infection control is also an issue that is particularly relevant in the pulmonary function lab. The laboratory observes the guidelines established by the hospital infection control team. New filters are placed on all breathing circuits and changed between patients. Children are asked to wash their hands with disinfecting foam prior to touching the equipment. The surfaces of the breathing tubes are also wiped with a disinfecting cloth between patients. Additional special precautions may be taken with those children that have infections with potential for spread to other ill patients.

Home spirometry is crucial in evaluating your child's lung function after transplant. We expect each child to perform home spirometry 3-5 times per week, and report findings to their transplant coordinator on a monthly basis as a minimum. You will be responsible to purchase the PFT device. Your coordinator will discuss this process and cost with you.

BRONCHOSCOPIES AND BIOPSIES

After transplant, your child will undergo a procedure called **bronchoscopy**. This test is done to determine the presence of infection and/or organ rejection. This test is routinely performed within the first day after transplant, at seven days post-transplant, again at one month, two months and three months post-transplant. The bronchoscopy performed within the first 24 hours is to check only for infection and inspect the area where the donor lungs are attached to your child's airway, (called the anastomosis sites). Subsequent bronchs are to rule out both rejection and/or infection. Should your child exhibit symptoms of rejection or infection at any time after their transplant, we may opt to perform a bronchoscopy with transbronchial biopsy. After three months post transplant, we perform routine bronchoscopies every three months the first year. After the first year post transplant, we generally do bronchs every six months depending on your child's progress.

To determine whether infection is present, a BAL specimen is sent to the lab from the bronchoscopy. A BAL (bronchoalveolar lavage) is a saline lavage from the lungs that is sent to the lab for culture to rule out infection. We have results usually in 24 to 48 hours.

To determine whether rejection is present, several tiny biopsies are taken from the lung tissue using a small biopsy forceps that is inserted through the bronchoscope tube. The tissue is taken to the pathology lab and examined under a microscope for the presence of rejection. See page 44 for further explanation. The amount of rejection present is graded as such: A0 is no acute rejection, A1 is minimal, A2 is mild, A3 is moderate and A4 is severe rejection. With A0 and A1, no further treatment is necessary, with A2 or greater, we treat with three doses (one each day) of high dose intravenous steroid called Solumedrol. The side effects can include high blood glucose levels, high blood pressure, and increased moodiness. Due to the possibility of side effects, we will admit your child to the hospital during this therapy. Children will

be admitted for rejection and may be admitted for certain types of infection. We ask you to make plans to stay overnight in St. Louis after a bronchoscopy due to potential complications that may occur, and in case any treatment is needed based on test results. Your child will continue to be prescribed the oral dose of steroids that they have been taking since transplant. After the three day therapy of intravenous steroids, your child will be discharged from the hospital and return to normal activities. Follow-up biopsy will be scheduled two weeks after treatment.

HEART BIOPSIES

Rejection of the transplanted heart is classified as mild, moderate, or severe. It is characterized by swelling of the tiny cardiac (heart) cells with destruction, and finally, if left untreated, cellular death. Often such changes are subtle and limited. Many people do not experience any symptoms or feel any different during a rejection episode. Ideally, we like to detect the process of rejection before any damage occurs. Therefore, periodically, a biopsy is performed, as it is the most accurate and reliable method currently available to diagnose rejection in the transplanted

heart. An IV is placed in the right internal jugular vein in which the doctor places a biptome (biopsy forceps). The forceps are passed through the vein and into the upper chamber of the heart (right atrium) through the valve and into the lower chamber (right ventricle). The biptome has tiny pinchers on the end to take pieces of the myocardial tissue from the right ventricle. Six little pieces are sent to the pathology lab where a pathologist looks at the heart tissue under a microscope. Once the biopsy is performed, an EKG, echo and chest x-ray will be done. Once your child is awake and able to eat and drink, and we have all the lab results, your child will be discharged to home.

The amount of rejection present is graded as such: 1R, mild; 2R, moderate; 3R, severe. 1R mild requires no additional treatment. 2R and greater requires a three day course of high dose IV Solumedrol given to the child in the hospital.

Cardiac catheterizations and heart biopsies are done routinely at one week, (give or take several days), one month, three and six months and one year. After that, they are done every other year or as needed.

DIET AND NUTRITION

After your child goes through transplant surgery, it is very important that he/she follows a healthy diet. During surgery and recovery, your child may have also lost some weight or had a poor appetite. Your child's body needs enough energy (calories & protein) to help recover after surgery.

A registered dietitian is available to you as part of your child's transplant team. Dietitians are there to help you plan your child's diet to meet his/her specific needs and tastes. Just a few simple changes in your child's diet can help keep your child and your child's transplant healthy for a long time.

Follow the simple food guide pyramid to ensure good nutrition:

- Depending on age and weight, eat enough servings from each of the five basic food groups everyday.

For Meat and Protein Foods, CHOOSE MORE OF:

- fresh fish, chicken and turkey without skin

- ground turkey
- lean, well-trimmed beef, veal, lamb, pork
- meatless protein-dried beans, lentils, peas, tofu
- egg whites
- water-packed tuna

LIMIT

- fried meats
- fatty cuts of meat
- bacon, sausage, lunch meats, frankfurters
- liver and organ meats
- cured and smoked meats, fish
- egg yolks to 3 per week
- canned meats and fish

For Breads and Cereals,**CHOOSE MORE OF:**

- plain breads, English muffins, bagels
- plain pasta and rice
- hot or cold cereals
- popcorn, rice cakes
- low-fat baked goods—angel food cake, graham crackers, fruit cookies, gingersnaps, fortune cookies

LIMIT

- high-fat, high-sugar baked goods – Danish pastry, croissants

For Milk, Cheese and Dairy Products,**CHOOSE MORE OF:**

- skim or 1 percent milk
- evaporated skim milk, or nonfat dry milk powder
- yogurt
- cheese

LIMIT

- creams, half & half, non-dairy creamers
- whipped cream, non-dairy whipped toppings
- whole-milk yogurt, sour cream and cheeses

For Fruits and Vegetables,**CHOOSE MORE OF:**

- several servings a day, with plenty of variety
- all fresh and sundried fruits
- all fruit juices (preferably unsweetened)
- raw or frozen vegetables
- vegetable juices

LIMIT

- coconut
- deep-fat fried vegetables
- vegetables with cream or butter sauces

For Fat, Choose a Minimal Amount:**CHOOSE MORE OF:**

- unsalted margarine
- vegetable oils

- low-fat diet salad dressings
- reduced calorie mayonnaise
- cook with oils which are low in saturated fats, such as olive oil and canola oil

LIMIT

- saturated and trans fat
- foods containing salt pork, lard, meat fat, and hydrogenated or partially hydrogenated solid vegetable shortening; products made with coconut or palm oil
- regular salad dressings and those made with sour cream

For Sweets, Choose a Minimal Amount:**CHOOSE MORE OF:**

- fruit, ices, gelatin, sherbets low-fat frozen yogurt ice milk

LIMIT

- doughnuts, pastries, cakes, cookies, and pies unless prepared with low-fat ingredients

FIBER

It is very important to eat a diet that is high in fiber. High fiber foods help to lower cholesterol, maintain bowel regularity, and may help reduce your blood sugars. There are two different types of fiber:

1. INSOLUBLE FIBER – Insoluble fiber is found in foods such as wheat bran, whole grain breads, bran cereals and vegetables. These foods help prevent constipation. They can also help fight against colon cancer.

2. SOLUBLE FIBER – Soluble fiber is found in foods such as oat bran, oatmeal, citrus fruits, apples, strawberries, beans and barley. They help lower your blood cholesterol, too.

Other tips...

- Limit fats such as margarine, gravy, cream, oil, mayonnaise, salad dressing, and fried foods.
- Limit sugar intake of high-sugar sweets such as candy, cookies, cakes and regular sodas. Try fruits or popsicles for dessert.
- Limit portions. Select only one entrée at mealtimes. Add a salad or vegetable if still hungry.

- Eat slowly and eat a variety of foods.
- Drink plenty of water and low-calorie beverages. Limit high-calorie beverages between meals (soda, Kool-Aid, sweetened juices).
- Trim all visible fat from meat including skin from poultry.
- Avoid frying meat. Instead roast, bake or broil meats.
- Steam or stir-fry vegetables in acceptable oils.
- Avoid using whole eggs in recipes. Instead, replace with two egg whites for one egg, or use 1/4 cup of egg substitute for one egg.
- Use applesauce in baked goods instead of oil or butter. One cup of applesauce equals 1 cup oil/butter.
- Replace low-fat for whole versions of yogurt, ice cream, sour cream, cheese and milk.
- Limit intake of high-sodium foods and use of table salt.

Can I keep my weight under control?

Weight gain after transplantation is commonly due to steroid treatment, which greatly increases your child's appetite. You can successfully control your child's weight by:

1. Limiting the amount of high calorie foods your child eats.
2. Gradually increasing your child's physical activity to burn off calories.

If you are not sure if your child's weight is healthy, ask your child's doctor. He/she can determine which weight is good for your child's height and body size.

My doctor said I need to lose weight ... what is considered safe weight loss?

- Set out to lose weight slowly. A weight loss of one half to one pound per week is generally considered a safe goal.
- Avoid yo-yo dieting (losing and gaining weight over and over) that can permanently slow down your metabolism and increase the fat stores in your body, making it even more difficult to lose weight.

- Eat regularly. Skipping meals may increase your appetite further.
- Include exercise in your program. Regular exercise helps you lose pounds and control weight. Regular physical activity may help to better control your blood pressure and reduce your risk of many other diseases.

CALCIUM

Steroids (Prednisone, Solumedrol) may cause bones to lose calcium particularly if dietary calcium is inadequate. Osteoporosis is a common consequence of long-term steroid use. Dairy products are the main sources of calcium and should be included in the diet to help keep bones strong.

Your daily calcium needs change as you age.

- Birth – 6 months: 400mg/day
- 6 months – 1 year: 600mg/day
- 1 – 5 years: 800 mg/day
- 6 – 10 years: 800-1200mg/day
- 11 – 24 years: 1200-1500mg/day

Ways to increase calcium in your diet:

- Choose at least 2 servings of low-fat dairy products per day, such as 2 eight-ounce glasses of 1 percent low-fat or skim milk. An 8-ounce glass of milk contains 300mg of calcium
- Eat a healthy, balanced diet. Salt, excess protein and phosphorus increase the loss of calcium in urine. Too many fibers reduce the amount of calcium that can be absorbed.
- Don't drink caffeine. This causes calcium to be secreted into the urine.
- Don't smoke! (Enough said!)
- Exercise to work your bones and help to build bone mass. Regular activity is vital to preventing bone loss.

FOOD SAFETY

Because of your child's medicines, he/she is more susceptible to germs in the foods they eat and bacteria from improper food handling and storage. Use the following tips to improve food safety.

Keep A Clean Kitchen

- Clean work surfaces often, and remove all food particles. Sanitize cutting boards after each use with a bleach and water solution and let them air dry. Also clean sinks, counters, kitchen tables with a bleach solution or with anti-bacterial soaps or sprays. The recipe for a homemade bleach solution is to mix one teaspoon of bleach with one quart of water.
- Change towels and dish cloths often. Toss sponges and scrub brushes into the dishwasher each night. Boil them with a small amount of bleach if you don't use a dishwasher. Replace sponges often.
- Wipe up spills in the refrigerator right away and keep shelves, sides and doors sanitized.
- Don't let dishes soak in the sink. The mixture of food, warm water and soap provides the perfect conditions for bacterial growth.

Proper Food Handling

- Wash your hands in warm, soapy water before and after every step in the food preparation process. Be especially vigilant immediately after preparing meat or poultry.
- If you have a cut or open sore on your hand,
- use plastic gloves or a plastic-sealing bandage. Wounds are easy areas for bacteria to enter the body.
- Be sure to wash all fresh fruits and vegetables with soap and water and peel before eating.
- Thaw meat, poultry or fish in the refrigerator. Never thaw on the counter. Bacteria thrive in food at room temperature.
- Use one cutting board for raw meat and poultry, another for chopping food that won't be cooked. Plastic boards that can be tossed in the dishwasher after each use are best.

- If you use a wooden cutting board, be sure to wash and scrub it after each use with soap and hot water, then sanitize with bleach solution or anti-bacterial spray designed for this purpose.
- Beware of cross-contamination. For example, don't carry the cooked meat to the table in the same dish used to carry the raw meat to the grill.
- Avoid raw or under-cooked meat, poultry or seafood. Cook beef and lamb to at least 140 degrees F, pork to at least 150 degrees F and poultry to at least 165 degrees F. Use a meat thermometer to ensure complete cooking.
- Cook eggs thoroughly until both the yolk and white are firm, not runny. Consider using pasteurized eggs instead of shell eggs whenever possible.
- Avoid sushi, raw oysters, and raw eggs in any form (cookie dough, eggnog, Caesar salad dressing).
- Stuff chicken or turkey just before roasting. This keeps the bacteria in raw poultry from invading the starchy stuffing. Once cooked, poultry and stuffing should be stored separately in the refrigerator.
- Treat cracked eggs carefully. If you find a cracked egg in a carton, don't use it. Cracked eggs can harbor disease-carrying organisms.

Proper Food Storage

- Keep any left-over foods out of the temperature zone: 40 to 140 degrees F. Foods left out for more than 2 hours (even in heated serving units) invite bacteria to grow. Keep refrigerator temperatures between 40 to 45 degrees F and freezer at zero degrees.
- Put raw seafood, poultry and meat in plastic bags so drippings can't contaminate other foods.
- Put eggs in their original carton in the main section of the refrigerator. Don't put them in the egg section of the door because the temperature there is higher.
- Keep the cupboard or pantry clean, dry, dark and cool. The ideal temperature is 50 to 70 degrees F. Temperatures over 100 degrees F are harmful to canned goods.

TRANSPLANT MEDICATIONS

OVERVIEW

This next section includes an overview of your child's medications, plus specific information about the medicines he or she will likely be taking. Please read this section carefully and ask any questions you may have. Following is a list of dosage forms and abbreviations that you may see. This is for informational purposes only and need not be memorized.

Your child's medications are prescribed in grams, milligrams or micrograms:

- One gram (g) = 1000 milligrams (mg)
- One milligram (mg) = 1000 micrograms (µg)
- One teaspoon (tsp) = 5 milliliter (mL)
- One tablespoon (tbsp) = 15 milliliter (mL)

Your child's medications are administered according to a preset schedule:

- Daily (one a day)
- BID (twice a day)
- TID (three times a day)
- QID (four times a day)

Your child may take his/her medications via different routes:

- PO (by mouth)
- NG (by nasogastric tube)
- SL (sublingually—under the tongue)
- IV (intravenously)
- Inhaled

MEDICATIONS

Immunosuppressive therapy is necessary to prevent or decrease the body's ability to reject the new organ. The goal of therapy is to induce the body's tolerance of the new organ while leaving the immune system intact to provide protection against infection. A combination of drugs is utilized to most effectively meet this goal. The type, dosage, and frequency of these medications are prescribed on an individual

basis. Each person's medications are prescribed according to his or her blood levels, body size, absorption, and tolerance of the drug.

These are powerful medications that have side effects. Therefore, they must be taken **EXACTLY** as prescribed to maintain the delicate balance in your child's body. Taking too little of the drugs may allow the immune system to destroy the new organ. However, too much of these drugs may alter the body's ability to fight off infections. Any alteration in medications that is not prescribed by your doctor may lead to serious consequences! If you are concerned about any medication side effects please contact your transplant nurse coordinator. There are several medication "rules" which you and your child must follow:

1. Never stop taking medication unless your doctor tells you to do so.
2. Do not skip a dose of medicine. If your child misses a dose, do not double the following dose. Please call your transplant coordinator for further instructions.
3. Do not take medicine that the doctor has not prescribed. Tylenol is acceptable in moderation.
4. Do not take medications at times other than what has been written on the schedule unless you have discussed this with your child's doctor. Make a schedule and get into the habit of taking medications at the proper times.
5. If your child becomes ill and cannot take medication or if he/she is throwing up or having diarrhea, please call your transplant coordinator for further instructions. If your child throws up within 15 minutes of taking medications, please repeat dose. If it has been greater than 15 minutes **DO NOT REPEAT DOSE.**
6. Do not allow your child's medicine to run out. Refill your child's medication before the bottle is empty. It is important to not miss a dose. Allow 7 days for your pharmacy to refill prescription.
7. Please check the expiration date on all medications. Do not take expired medications.
8. Please check liquid medication concentrations when refilled at the pharmacy. Liquid medications

come in many different concentrations so check carefully for proper dosage.

9. Please learn medications by brand and generic name. It is also very important that you know medication doses by concentration or strength (gram, milligram, microgram, or units) as well as form (capsule, tablets, liquid)

For children too young to take pills, we will prescribe a liquid form of the medication. All doses of the liquid medication will be measured by a syringe. The nurses will teach you how to use different size syringes and draw up the correct dose of medicine.

It is best to get in the practice of giving your child the medications directly out of the syringe so it is not spilled or wasted. We will send you home with a large supply of syringes. You may wash and reuse the syringes as long as the numbers are visible. When you need more, you can refill the prescription at the hospital or your local pharmacy.

Since children on immunosuppressant medicines are at a higher risk for infections it is important to treat each infection appropriately. Antibiotics are important medicines used to treat infections caused by bacteria. If antibiotics are used inappropriately they may cause unnecessary side effects and may not work as well in the future. This is called “antibiotic resistance.” Bacteria can become “super bugs” which are not able to be killed by antibiotics. You can help to make sure your child’s infection is treated appropriately:

- Ask if the infection is caused by a bacteria or virus. Viruses should not be treated with antibiotics.

- Take medications as prescribed by your doctor. Do not skip doses or stop the course early if your child is feeling better (unless your doctor tells you to do so).
- Never save antibiotics for the next time your child is sick.
- Never give your child antibiotics prescribed for someone else.

Store all medications out of the reach of young children. If accidental swallowing of any medication occurs, take the child and the medication to the nearest Emergency Center immediately.

Never stop any medication unless instructed by your child’s doctor.

DRUG INTERACTIONS

Many medications (both prescription and over the counter) can interact with your child’s immunosuppression (anti-rejection) in an undesirable way. For instance, some antibiotics affect the absorption or metabolism of these medicines, either causing the drug levels to become too high or too low. The following is a brief list of medications that are known to interact with your child’s transplant medications.

Prior to starting your child on any of the following medications, you must notify the transplant team:

This is just a small list of medications known to interact with your child’s transplant medicines; there are many others that interact as well.

Medications used to treat infections (bacterial, viral, fungal, or other) including:	Over the counter medications to treat pain, cough/cold, or gastrointestinal (GI) issues:	Other medications with known drug interactions:
Biaxin (clarithromycin) Ciprofloxacin Fluconazole Itraconazole Voriconazole Rifampin	Ibuprofen (Advil, Motrin) Naproxen (Aleve) Nyquil Pepto Bismol Pseudoephedrine (Sudafed)	Dilantin (phenytoin) Phenobarbital Reglan Tegretol Herbal supplements

Do not give your child any over-the-counter medications or medications prescribed by a physician who is unfamiliar with your child's medical history, without first consulting the transplant team.

Medications are essential to the success of your child's transplant. Your child will be on many different medications after his/her transplant. Three of these will be immunosuppression (anti-rejection) medications. Without these medications, the body will reject the new organ. Each medication is prescribed for your child's individual needs and is adjusted according to the body's response. The medications must be taken as directed; DO NOT change or quit giving your child's medications unless instructed by a member of the transplant team. We expect parents, and eventually the child, to understand the actions of each medication; so please be sure to ask questions to clarify any information given to you. Understanding the medications enables you to better identify the side effects when they occur.

Prior to discharge, your family will begin to administer medications to your child, with the assistance of your nurse. Before discharge from the hospital, we will teach you:

- the generic and brand names of each medication;
- what each looks like;
- the purpose and action of each drug;
- how to determine the correct dosage;
- precautions required for each one;
- how and when to take the drugs; and
- the common and uncommon side effects for each drug.

Before discharge, your transplant nurse coordinator will give you a medication schedule for your child

listing the names of the drugs, dose, any special instructions, and times to administer the medications. Each time you or your child is getting ready to take his/her medications, you should have the medication schedule in front of you as you get the medications ready.

Each time a change is made in medication (either the dose or the frequency), you should make the change on the schedule. Medications should be given by your schedule, not according to the instructions written on the medication bottle. If you have any questions, please contact your transplant nurse coordinator.

Be sure to store all the medications in their original containers. This will be helpful in keeping up with the expiration dates and who prescribed each drug. Store all medications away from heat, direct light and moisture, each of which can cause deterioration of the drug. Always consistently administer medications at the same time of day. Each of the routine medications needs to achieve a stable level in the blood. Monitoring of these blood levels will be done at intervals and the test results are used to make adjustments in medication dosages. Therefore, **it is crucial that medications are given in a consistent manner.**

Always carry your child's medication schedule with you so changes may be made if necessary. You may also wish to provide your child a copy of medications for his/her wallet. Should your child require hospital admission, please bring all your medications.

The pharmacy may change brands or liquid concentrations without your knowledge resulting in an improper dosage. Pay close attention to your label and the appearance of your medication. Ask the pharmacy or transplant team if the medication looks different than usual.

IMMUNOSUPPRESSANT MEDICATIONS

PROGRAF® (Tacrolimus)

Action:

Tacrolimus is an immunosuppressant drug that decreases the number of specific white blood cells to help prevent rejection. Should not be given with Cyclosporine.

Preparation:

Available in 5 mg, 1 mg, and 0.5 mg capsules. Also available as a liquid preparation.

Frequency:

Twice a day (bid) at 12 hour intervals.

Possible Side Effects:

- Infection: Because this drug suppresses the immune system, infection is one of the leading potential risks.
- High blood sugar: Tacrolimus may cause high blood sugar and progress to diabetes.
- Kidney Impairment: Kidney function is carefully monitored in patients on this drug because kidney damage can occur.
- Tremor: Fine shaking of the hands may occur during the first month of using this medicine. If it worsens and/or interferes with the child's daily activities, contact your child's doctor. Seizures are a rare complication seen in the first several months.
- High Blood Pressure: This may be a common early side effect and can be controlled with blood pressure medicine.
- Hair Loss: Sometimes occurs but usually not permanent. Hair will grow back.

Special Instructions:

- There are a number of medications that interact in various ways with Tacrolimus either increasing or decreasing Tacrolimus levels in the bloodstream. Your child should not take any other medications (over-the-counter or prescription) without checking with the transplant team first.
- Do not take Tacrolimus with grapefruit or grapefruit juice.
- While taking this medicine, lab work will be obtained to monitor the drug level. It is important to have this

done as scheduled so dosages can be correctly adjusted according to your child's needs.

- Capsules should not be opened or crushed.
- Shake the bottle well before administering the liquid.

PREDNISONE or PREDNISOLONE (Orapred®) – Steroid

Action:

Prednisone decreases the body's response to foreign cells by altering the immune system, to help prevent rejection.

Preparation:

Available in liquid preparation and various pill strengths.

Frequency:

Usually given once per day.

Possible Side Effects:

- Infection: Because this drug suppresses the immune system, infection is one of the leading potential risks.
- Increased appetite and weight gain: Transplant patients taking Prednisone often have an increased appetite leading to weight gain. Refer to sections on Diet and Nutrition (see page 24) and Obesity (see page 48).
- Acne: Most often occurs on the face but can also occur on the chest and back. Keep affected areas clean and dry. Refer to section on Acne (see page 18).
- Increased sensitivity to the sun: Always apply sunscreen to exposed skin when in direct sunlight. A minimum SPF (sun protection factor) of 30 is recommended.
- Delayed growth: Slows growth and delays puberty.
- Muscle weakness or cramps: These can be decreased by walking, biking, or doing exercises that strengthen muscles.
- High blood sugar: Prednisone may cause high blood sugar and progress to diabetes.
- Change in Vision: Cataracts may occur in some patients who receive Prednisone in high doses for a long time.

- Mood Changes: Very “up” or very “down”. Children may cry easily, be giggly, or have increased irritability. This usually improves as the child adjusts to the medication and the dose is decreased.
- Decreased bone density: Thinning of the bones.
- Insomnia: May cause sleep disturbance.

Special Instructions:

- Never stop this medication unless instructed by your child's doctor.
- Take this medication in the morning if possible. This may help alleviate sleep disturbances.

CELLCEPT® (Mycophenolate Mofetil, MMF) or MYFORTIC® (Mycophenolic acid)

Action:

CellCept is an immunosuppressant drug which decreases the number of specific white blood cells which are responsible for rejection.

Preparation:

Mycophenolate mofetil (CellCept) available in 250 mg and 500 mg capsules. Liquid preparation is also available as 200 mg/mL. Myfortic (mycophenolic acid) available in 180 mg and 360 mg delayed release tablets. Preparations are not interchangeable.

Frequency:

Twice a day (bid) at 12 hour intervals.

Possible Side Effects:

- Infection: Because this drug suppresses the immune system, infection is one of the leading potential risks.
- Bone Marrow Suppression: May decrease white blood cell counts leading to increased risk of infection and decreased platelets, causing bruising. Dosage adjustment is required if this occurs.
- GI Distress: Signs and symptoms include nausea/vomiting/diarrhea/cramping. Generally this resolves in time.

Special Instructions:

- Capsules should not be opened or crushed.

IMURAN® (Azathioprine)

Action:

Imuran is an immunosuppressant drug which decreases the number of specific white blood cells which are responsible for rejection.

Preparation:

Available in 50 mg tablets; can be prepared as liquid.

Frequency:

Once per day, usually at bedtime; need to take consistently at the same time every day.

Possible Side Effects:

- Infection: Because this drug suppresses the immune system, infection is one of the leading potential risks.
- Bone Marrow Suppression: May decrease white blood cell counts leading to increased risk of infection and decreased platelets, causing bruising. Dosage adjustment is required if this occurs.
- Hair Loss: Sometimes occurs, but usually not permanent. Hair will grow back.

RAPAMUNE® (Sirolimus) or ZORTRESS® (Everolimus)

Action:

Sirolimus and everolimus are immunosuppressants used to prevent transplant rejection.

Preparation:

Sirolimus is available in tablet and liquid form. Everolimus is available in tablet form.

Special Instructions:

- The liquid form of Rapamune must be mixed in 2 ounces (60 mL) of water or orange juice in a plastic or glass cup. Stir the solution well and drink immediately. The cup should then be refilled with 4 ounces (120 mL) of water or orange juice, stirred, and the patient should drink this as well.
- **DO NOT** use Rapamune with apple juice, grapefruit juice or milk products. Patients who have taken Rapamune undiluted have developed mouth ulcers.

Possible Side Effects:

- Infections.
- Impaired wound healing.

- High cholesterol and high triglycerides.
- High blood pressure, edema (swelling), fever, headache, acne, upset stomach, decrease in white blood cell and platelet counts, and joint pain.
- Renal impairment.

Special Instructions:

- While taking this medicine, lab work will be done to monitor the drug level. It is important to have this done as scheduled so dosages can be correctly adjusted according to your child's needs.
- Take consistently with regards to food.

INFECTION PREVENTION MEDICATIONS

BACTRIM/SEPTRA® (Sulfamethoxazole/ trimethoprim – (SMZ-TMP)

Action:

Bactrim is given to children for the treatment and/or prevention of urinary tract infections (kidney/bladder infections). In the case of transplant patients, it is given to prevent a specific type of pneumonia called pneumocystis pneumonia.

Preparation:

Bactrim is supplied in both liquid and tablet form. The strength of the liquid is: Sulfamethoxazole 200 mg and Trimethoprim 40 mg per 5 mL (1 teaspoon). A single strength tablet contains Sulfamethoxazole 400 mg and Trimethoprim 80 mg. A double strength tablet contains Sulfamethoxazole 800 mg and Trimethoprim 160 mg.

Possible Side Effects:

- Rash
- Bone Marrow Suppression: May decrease white blood cell counts leading to increased risk of infection and decreased platelets, causing bruising. Dosage adjustment is required if this occurs.
- Sun sensitivity

Special Instructions:

- Please notify your physician if your child has sulfa allergy

MYCOSTATIN® (Nystatin)

Action:

Mycostatin is an antifungal used to prevent or treat yeast infections (thrush) of the mouth and throat.

Preparation:

Supplied as liquid or in a lozenge form.

Possible Side Effects:

- Side effects are very rare

Special Instructions:

- For infants, apply liquid Nystatin directly to gums and inside cheeks.
- Older children should swish and then swallow.
- Do not eat or drink anything for a least 20 minutes after taking Mycostatin.

ZOVIRAX® (Acyclovir)

Action:

Acyclovir is an antiviral drug that fights or helps prevent viruses of the Herpes family i.e. Herpes Simplex (virus that causes "cold sores"), Varicella (chickenpox), etc.

Preparation:

Available in 200mg/5 ml liquid preparation. Available in 200mg, 400mg, and 800mg capsules/tablets. Also available as an IV preparation.

Frequency:

Variable, depending on if it's being used for prevention or for treatment.

Possible Side Effects:

- Bone Marrow Suppression: May decrease white blood cell counts leading to increased risk of infection and decreased platelets causing bruising. Dosage adjustments are required if this occurs.
- Kidney Impairment: Because this drug is cleared through the kidneys, possible damage may occur. Kidney function is carefully monitored in patients on this drug.

Special Instructions:

- Important to increase oral fluid intake while on this medicine.

VALCYTE® (Valganciclovir) or CYTOVENE® (Ganciclovir)

Action:

Valganciclovir and ganciclovir are anti-viral drugs used to prevent and/or treat Cytomegalovirus (CMV).

Preparation:

Valganciclovir is available in 450mg scored tablets. Liquid preparation is also available. Ganciclovir is available in IV form.

Frequency:

Usually once or twice a day (BID). Depends on if used for prevention or treatment of infection.

Possible Side Effects:

- GI Distress: Signs and symptoms include nausea/vomiting/diarrhea/cramping. Generally this resolves in time.
- Headaches, Confusion
- Bone Marrow Suppression: May decrease white blood cell counts leading to increased risk of infection and decreased platelets, causing bruising. Dosage adjustment is required if this occurs.
- Kidney Impairment: Because this drug is cleared through the kidneys, possible damage may occur. Kidney function is carefully monitored in patients on this drug.

Special Instructions:

- Wash hands thoroughly after handling this medication.
- Capsules should not be opened or crushed.

ANTI-HYPERTENSION/ DIURETIC

NORVASC® (Amlodipine)

Action:

Norvasc is given for the treatment of high blood pressure.

Preparation:

Available in 2.5 mg, 5 mg and 10 mg tablets. Liquid preparation also available.

Possible Side Effects:

- Headache, dizziness
- Swelling of hands and feet

Special Instructions:

- Always tell your child's doctor that he/she is taking Norvasc.

LASIX® (Furosemide)

Action:

Stimulates the kidney to remove excess water from the body. There is usually an increase in urine output starting about one hour after the dose and lasting from four to eight hours.

Preparation:

Lasix Oral Solution is supplied as 10 mg/mL liquid. Lasix is also supplied in 20 mg, 40 mg, and 80 mg tablets.

Possible Side Effects:

- Weakness or unusual tiredness
- Irritability or listlessness
- Sudden weight change
- GI Distress: Signs and symptoms include nausea/vomiting/diarrhea/loss of appetite. Generally this resolves in time.
- Dizziness, headaches, blurred vision
- Ringing in the ears or hearing loss
- Light-headedness upon standing
- Signs of dehydration: May include inability to make tears, dry mouth, decreased urine output, and sunken eyes.

Special Instructions:

- Once opened, Lasix liquid is good for 90 days.
- When Lasix stimulates the kidneys to release the excessive water in the body, an important body element, potassium, is also washed out of the body. To help replace the potassium you need to include foods from this list in your child's diet:
 - All Baby Formulas
 - Orange Juice

- Bananas
- Peaches
- Beans (pinto or kidney)
- Potatoes
- Citrus fruits
- Prunes
- Dried dates
- Prune Juice
- Contact the dietitian for further nutritional information.

ALDACTONE® (Spironolactone)

Action:

Stimulates the kidney to remove excess “water” from the body without losing potassium.

Preparation:

Available as 25 mg, 50 mg, and 100 mg tablets. Liquid preparation is also available.

Possible Side Effects:

- Weakness, unusual tiredness
- Irritability or listlessness
- Sudden weight change
- Abdominal cramping and diarrhea
- Dizziness, headaches, mental confusion
- Irregular menstrual cycle, or swelling of breast tissue
- Deepening of the voice
- Signs of dehydration: May include inability to make tears, dry mouth, decreased urine output, and sunken eyes.

Special Instructions:

- Shake the bottle well before measuring.

MEDICATIONS GIVEN FOR TREATMENT OF REJECTION

THYMOGLOBULIN® (Antithymocyte Globulin – Rabbit) or ATGAM® (Antithymocyte Globulin – Equine)

Action:

Thymoglobulin and ATGAM are very potent immunosuppressants only used in the hospital for prevention or treatment of transplant rejection.

Preparation:

IV formulation

Possible Side Effects:

- Fever and chills
- Bone marrow suppression: Decrease in white blood cell count and decreased platelet count.
- Pain
- Headache
- Diarrhea
- Increased blood pressure
- Severe allergic reactions can occur in rare instances. Patients are pre-treated with acetaminophen, antihistamines, and steroids to reduce the severity of adverse reactions. Vital signs and blood work will be used to monitor for adverse effects.

SIMULECT® (Basiliximab)

Action:

Basiliximab is an intravenous medication used to prevent organ rejection.

Preparation:

IV formulation

Frequency:

Given prior to transplant, and then 96 hours post-transplant.

Possible Side Effects:

Because this drug suppresses the immune system, infection is one of the leading potential risks.

IMMUNE GLOBULIN or IVIG (Gamunex®)

Action:

Immune Globulin works on specific cells to treat antibody mediated rejection. It can also be given before and after transplant in patients with high antibody levels to help prevent rejection.

Preparation:

Available only as an IV infusion and is usually given at the hospital, in clinic or by a home care agency.

Possible Side Effects:

- Infusion related reactions: chills, flushing, headache, itching, muscle pain, fever and fast heart rate.

Special Instructions:

- • Acetaminophen (Tylenol®) and diphenhydramine (Benadryl®) may be given prior to the infusion.
- • The nurse giving the medication will start the infusion slowly and increase the rate as tolerated.

OTHER IMPORTANT MEDICATIONS

MAGNESIUM

Action:

Magnesium is an important element used by the body to complete many enzyme reactions. It is vital for muscle function including heart function. Several immunosuppressant drugs can decrease magnesium levels.

Preparation:

Many forms of magnesium are available. Please check with your physician for recommendations.

Possible Side Effects:

- GI Distress: Signs and symptoms include nausea/vomiting/diarrhea/loss of appetite. Generally this resolves in time.
- Muscle weakness

PHOSPHORUS (Neutraphos®/Osmoprep®)

Action:

Phosphorus is an important electrolyte in the body. It works together with calcium to keep bones strong and healthy. In patients who are urinating large amounts (i.e. new kidney transplant recipient), too much phosphorus is lost in the urine so it needs to be replaced.

Preparation:

- Available as a capsule or individual packets.

Possible Side Effects:

- GI Distress: Signs and symptoms include diarrhea and nausea. Generally resolves in time.

Special Instructions:

- Neutraphos capsules and packets must be mixed with fluid. Open capsule or packet and dissolve in 75mL of fluid of choice (i.e. water, juice, punch, etc) and drink. **DO NOT SWALLOW CAPSULES WHOLE – MUST BE DILUTED.**
- Do not take magnesium or other antacids at the same time as Neutraphos. Allow at least 2 hours between these medications.

CALCIUM

Action:

Calcium is an important mineral used by the body. It is vital to proper bone structure and many functions of the nerve and muscle systems. For transplant patients, calcium is used to help prevent calcium loss from bone due to steroids (Prednisone).

Preparation:

Many brands of calcium are available. Please check with your physician or dietitian for recommendations.

Possible Side Effects:

- Constipation

Special Instructions:

- None

POTASSIUM CHLORIDE

Action:

Potassium is sometimes given to children who are taking diuretics ("water medicine"). Potassium is a body element, which is very important for proper functioning of the heart and body cells. Although diet usually provides adequate amounts of potassium, children taking diuretics may require more.

Preparation:

Potassium comes in various liquid and tablet forms.

Possible Side Effects:

- Weakness, unusual tiredness, irritability or listlessness
- GI Distress: Signs and symptoms include nausea/vomiting/diarrhea/loss of appetite. Generally this resolves in time.
- Numbness in hands or feet

Special Instructions:

- Potassium has a bitter taste that cannot be completely hidden, but mixing it in different juices and having something to drink immediately afterward helps. It is usually better to take it at the beginning of the meal when the child is hungry. Potassium can also cause an upset stomach; giving it at mealtime helps avoid this.

- **DO NOT REPEAT ANY VOMITED DOSES. IF YOUR CHILD HAS THE "FLU" AND IS FREQUENTLY VOMITING OR HAS DIARRHEA, CONSULT YOUR CHILD'S DOCTOR.**

Many medications will affect the absorption of the immunosuppressive drugs or may interact with them in an undesirable way. Therefore, do not give your child any over-the-counter medications or medications prescribed by a physician who is unfamiliar with your child's medical history without first consulting the transplant team.

WHAT TO REPORT

If you have any questions about your child's care, call your transplant nurse coordinator during daytime hours (Monday through Friday 8 a.m. to 4 p.m.). Before 8 a.m. and after 4 p.m., and on weekends and holidays, call the hospital operator at 314.454.6000 and ask for the pulmonologist on call to be paged.

For emergency situations, call 911.

Call with any of the following signs or symptoms:

- Temperature over 100.4°F or 38.5°C
- Flu-like symptoms such as nausea, vomiting or diarrhea
- Chills
- Cough or shortness of breath
- Any questions regarding medication dose, how to give, if started on a new medication, or a missed dose

- Blood in the stool
- Redness, swelling, drainage, pus or pain at the incision or Broviac site
- Home spirometry FEV1 decrease by 10 percent over two days
- Heart-failure symptoms: decreased exercise tolerance, fluid retention, difficulty breathing, nausea and vomiting
- Exposure to contagious disease: especially chicken pox

Often you will be directed to the primary care doctor if the issue is not transplant related. The transplant team is always willing to help you get the best care for your child.

READMISSION TO THE HOSPITAL

READMISSION

Readmission to the hospital may occur for several reasons:

- **Fever** – especially if your child has a Broviac (central line)
- **Infections** – such as pneumonia or CMV, etc. because your child may need antibiotics.
- **Vomiting and Diarrhea** – especially if your child is unable to keep down fluids and/or their medications
- **Observation after a procedure** – i.e. after a biopsy
- **Treatment for organ rejection**

In the event your child is to be admitted to the hospital after transplant, please bring the following with you:

- All current medications
- Medical records (including X-ray films) – if your child was initially seen at an outside physician office or hospital for this current illness or problem
- Current insurance card

In most cases, your child will be admitted on the same floor as they were on post-transplant. However, if your child has an infectious illness such as chickenpox or influenza, they may be on the general medicine or infectious disease floor to decrease exposure to other children who have been transplanted.

TRANSPLANT COMPLICATIONS

There are many complications that are associated with transplantation. Complications may be a result of the surgical procedure itself or due to side effects from the many medications your child will be taking after transplant. There is no way to predict if your child will develop any of these problems or how severe they may be. Some complications are more severe than others; some are temporary and some are long term. Most complications are treatable, however, some complications may result in permanent conditions. If complications do occur, the transplant team will take the necessary steps to treat and/or minimize the problem.

SURGICAL COMPLICATIONS

POST-OPERATIVE BLEEDING

Bleeding may occur in the post-operative from blood vessels that are exposed when the old heart and lungs are removed. Signs of bleeding include a rapid or ongoing drop in hemoglobin and hematocrit, increased heart rate, low blood pressure, and increased bleeding from chest tubes. If there is a significant amount of bleeding, your child will need to be taken back to the operating room so the surgeon can find the cause of bleeding and stop it. Rarely, diffuse life-threatening bleeding may occur that is difficult or impossible to control. Your child may require additional blood transfusions to help replace the blood that is being lost.

AIRWAY COMPLICATIONS OF THE BRONCHIAL TUBES

The breathing tubes leading to the new lungs are connected to the recipient by a connection called an "anastomosis". Two connections are made, one on each side. Problems that can occur in the area of the anastomoses include: airway narrowing (stenosis), collapse (malacia) and dehiscence (failure of the connection to heal and close).

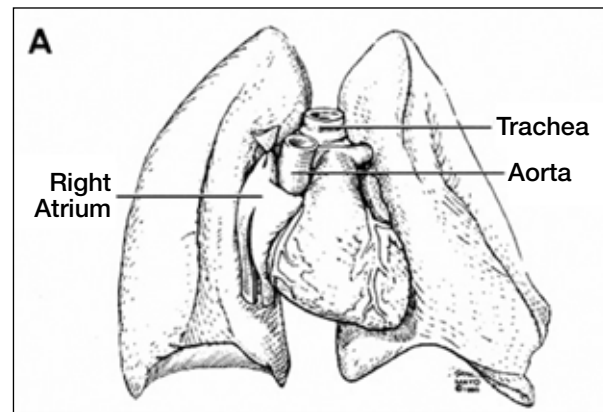
Airway narrowing or stenosis: Sometimes as the anastomoses heal, scar tissue called "granulation tissue" can develop. If a large amount of granulation tissue develops, the breathing tube can be narrowed at that point. Symptoms of airway narrowing may include wheezing, and decreased pulmonary

functions, particularly the FEV1. Generally, airway narrowing is evaluated with a procedure called "rigid bronchoscopy" which is performed in the operating room and may be treated by dilation of the affected region with a small balloon catheter.

Airway collapse or malacia: The stiffness of the breathing tubes depends on the development of a part of the underlying tissue called cartilage. In infants, the airways may be more "floppy" until the cartilage becomes fully developed. This is called "tracheomalacia" if it involves the main breathing tube or "bronchomalacia" if it involves the breathing tubes leading into one or both lungs. Other than supportive care as necessary, there is no specific treatment. Usually the malacia resolves on its own as the child grows and the cartilage develops.

Airway dehiscence: If the connection (anastomosis) between the donor and recipient breathing tubes does not heal completely, air and/or fluid can leak in to the space outside the lung. Depending on the size and timing, this can be a minor problem or a catastrophe. Advances in surgical techniques have made airway dehiscence a relatively rare occurrence.

YOUR CHILD'S ANASTOMOSIS



NERVE INJURY

Many important nerves run through the area that the surgeons are working in during the lung transplant operation. These include nerves that go to the vocal cords (voice box), the diaphragm (an important muscle for breathing that separates chest cavity from the abdominal cavity), and the gastrointestinal tract (stomach and intestines).

If there is a lot of scarring in the chest cavity (often present if the child has had previous heart surgery), these nerves may be difficult to identify during the dissection and become injured.

Vocal Cords: The vocal cords are a critical component of voice production. The vocal cords may become paralyzed if the nerves controlling them are injured during transplant. Patients with paralyzed vocal cords may have a change in voice and have an increased risk of aspiration. Vocal cord paralysis often resolves in the first several months after transplant.

Diaphragm: The diaphragm is a large muscle that separates the abdominal cavity (stomach and intestines) from the thoracic cavity (heart and lungs). The diaphragm is a very important muscle that facilitates moving air in and out of the lungs. There are two parts to the diaphragm, one on each side. Each is controlled by a separate nerve. Injury to one or both nerves may lead to weakness or paralysis of one or both sides of the diaphragm. Diaphragm hemi-paralysis is not a life threatening complication and is usually well tolerated. However, it may lead to infants remaining on oxygen longer and older children having difficulty weaning from the ventilator. If these difficulties persist, an operation called *plication of the diaphragm* may be performed.

Gastrointestinal Tract: Injury to the nerves serving the gastrointestinal tract may lead to slowing of the muscle activity that moves food through the esophagus, stomach and intestines. Narcotics used for pain in the early post-transplant period may further decrease motility. Symptoms associated with this “gastric dysmotility” may include nausea, vomiting, and reflux. Generally these symptoms improve as the pain medications are stopped and resolve with time. Gastric dysmotility can predispose patients with cystic fibrosis to have “distal intestinal obstruction syndrome” (DIOS). (See page 47.) A very small percentage of patients who develop an intestinal obstruction require surgery to relieve the obstruction.

TRACHEOSTOMY

A tracheostomy may be recommended after heart-lung transplant if your child is experiencing an extended length of time on the ventilator. Instead of the breathing tube that goes through the mouth or nose, a tracheostomy is a surgical procedure (performed in the operating room under general anesthesia) that allows the breathing tube to be

inserted into the trachea (windpipe) through a hole in the neck called a stoma. With a tracheostomy, your child will be able to be more awake and alert and may be able to wean from the ventilator more quickly and more comfortably. Most children who require a tracheostomy post-transplant are able to have it removed prior to discharge.

LUNG GRAFT DYSFUNCTION

There are several potential injuries that can affect the lungs that come from a deceased donor. These include the events that led to the death of the donor, the period of cold storage during which the lungs do not receive blood or oxygen, and the “reperfusion injury” that occurs after the lungs are sewn into the recipient, restoring ventilation and blood supply. In a small percentage of transplants, during the first 24 hours following the transplant these injuries lead to poor functioning of the transplanted lungs. While this “early graft dysfunction” may be tolerated with transplanted kidneys or livers, immediate effective functioning of a transplanted heart or lung is critical to survival of the patient.

Early graft dysfunction results from leakage of inflammatory cells and fluid into the lungs. The lungs then become less flexible and are more difficult to inflate. This, in turn, leads to difficulty in maintaining effective uptake of oxygen and removal of carbon dioxide. Patients with early graft dysfunction may have increased frothy secretions coming from the breathing tube and increased infiltrates (whiteness) on their chest X-rays.

Treatment for early graft dysfunction is primarily supportive, including the use of mechanical ventilation and in rare cases, a temporary heart lung bypass machine called an extra-corporeal membrane oxygenator (ECMO). Most patients slowly recover from early graft dysfunction. However, in rare cases it can be fatal. Because of poor outcomes, patients with severe early graft dysfunction are not considered candidates for retransplantation in our program.

HEART GRAFT DYSFUNCTION

Primary graft non-function is a very rare but life threatening complication. Malfunction of the new heart results from an intense reaction by your child's body against the new heart graft (hyperacute rejection). Should this occur, your child will have to be treated with increased doses of medications

and/or additional therapies to treat this rejection. In rare instances, the new heart may not respond to treatment and will likely result in death.

HEART FAILURE

Primary graft non-function and rejection may cause your child to develop heart failure. Signs and symptoms of heart failure may be similar to those your child was experiencing prior to transplant including increased water retention, more difficulty breathing, and decreased energy level. Signs of heart failure will be closely monitored for post-operatively and if they develop, will be treated aggressively with medications.

CARDIAC ALLOGRAFT VASCULOPATHY (CAV)

CAV occurs when the coronary arteries (small blood vessels that carry blood) thicken and narrow, making it harder for blood to get to the heart. This causes parts of the heart muscle to die when they do not get enough oxygen.

This problem can happen at any time after transplant, the exact cause is unknown. It is typically a long term complication and continues to be the major reason people eventually die or need another heart transplant.

CAV is often difficult to diagnose. Adult patients with blocked coronary arteries have chest pain, but this often does not happen to patients after transplant because the heart does not have any nerve signals.

Coronary angiography is performed at one year after transplant and then every other year thereafter to assess for CAV.

In some cases, nothing can be done to stop CAV from developing. A number of things thought to be helpful include: regular exercise, following a healthy diet, taking medication regularly and not missing any doses, maintaining a normal body weight, not smoking.

At the moment, there is no cure for CAV. A number of medications may be started to try and slow down the narrowing in the coronary arteries and prevent blood clots from forming in the small coronary arteries. In some cases, your child may have surgery to open up the blocked arteries. In very severe cases, your child may need another transplant.

POST-TRANSPLANT INFECTIONS

The immunosuppressant medications that your child will be taking after transplant to prevent rejection will also interfere with your child's ability to fight off infections. Your child will not necessarily be more prone to catching colds or other illnesses but when they are ill, it may take them longer to get over the illness. Additionally, your child is more prone to "opportunistic" infections; these are infections that your child has already been exposed to either prior to transplant or from the transplant itself that reactivate when your child's immune system is suppressed.

It's very important for you to re-establish your child with their pediatrician as well as a pediatric pulmonologist and pediatric cardiologist if you do not live in the St. Louis area, once they return home from the hospital. We recommend that you take your child in for a well visit appointment within the first one to two weeks after returning home. This will give your pediatrician the opportunity to see your child when they are healthy as well as review your child's transplant course with you.

BACTERIAL INFECTIONS

Bacteria can be found everywhere. We all have bacteria on our skin, in our mouth, and in our intestine. When we are healthy, this bacteria does not hurt us or make us sick. However, children who have had surgery, such as a transplant, or who are on immunosuppressant medications, can develop infections from their normal body bacteria. Bacterial infections are usually treated with oral or IV antibiotics, depending on the severity of the infection. The following are signs that your child may have a bacterial infection:

- Fever – temperature greater than 100.4°F or 38°C
- Sore throat – may be a strep throat infection
- Redness around your child's incision and/or pus draining from incision, drainage tubes, or central line site
- Pain or burning with urination – may be a urinary tract (bladder) infection
- New cough
- Difficulty breathing or shortness of breath
- Decrease in PFTs

If your child develops any of these symptoms, you need to call your child's transplant nurse coordinator or transplant physician on call for further instructions. For minor illnesses such as earache, we will most likely have you take your child to their pediatrician for exam and treatment. However, before starting your child on any new medications, contact your transplant nurse coordinator to make sure these medications are compatible with your transplant medications.

If your child has been transplanted for less than three months, has a central line (broviac), and develops any of the above symptoms, we may ask you to come to the transplant clinic for further evaluation. You should bring extra clothes, as your child may need to be admitted to the hospital for evaluation and treatment.

VIRAL INFECTIONS

Viruses most often cause illnesses such as the common cold, stomach flu, and influenza. In most cases, these illnesses are caught from other people in the community who are currently sick. There is no "cure" for these types of viral illnesses – it just takes time for the virus to go away. There are things you and your child can do however, to help prevent them from getting these infections:

- Practice good hand washing! Most cold germs are passed from your hands to the mucous membranes in your nose and eyes.
- Avoid friends and relatives you know to be ill. If an immediate family member is ill, use common sense: have them use separate drinking glasses, cover their mouth when coughing and sneezing, etc.
- Instruct your child to not share drinking glasses or eating utensils with others, at home and at school.

Should your child develop any of the following symptoms, please contact your child's transplant nurse coordinator or physician on call:

- Fever – temperature greater than 100.4°F or 38°C
- Vomiting and diarrhea – especially if your child is unable to keep fluids and medication down or is having persistent diarrhea (watery and/or more than six stools per day) for more than two days
- Cold symptoms such as cough, earache, sore throat, or runny nose
- Low grade fever, muscle aches, excessive fatigue

In addition to being transmitted by coming into contact with another person who is infected, viruses can affect transplant recipients by two other mechanisms:

- For a first time infection, the virus could have been transmitted from the donor organ or from a blood transfusion.
- Reactivation of a person's own virus – in other words, the patient had been exposed to the virus prior to transplant and the virus "reactivated" when the patient was immunosuppressed with medication.

Blood tests are performed on your child and the organ donor prior to transplant to see if they have been exposed to these viruses. Exposure to these viruses does not exclude either the patient or the donor from transplant; it merely helps the transplant team to plan for surveillance of developing infection and treatment after transplant.

CYTOMEGALOVIRUS (CMV)

CMV is a type of virus in the **Herpes Virus Family**.

It causes flu-like symptoms such as fever (usually spike a fever at the same time every day), muscle ache, and fatigue. CMV can also affect different organs in the body as well. For instance, the virus can settle in the retina of the eye, cause pneumonia, or cause hepatitis (inflammation of the liver).

A medicine called Ganciclovir is used to try to prevent an active CMV infection immediately after transplant; this same medicine is also used to treat patients who develop active CMV. Unfortunately, it does not get rid of the virus completely; it merely returns it to an inactive state. Some transplant recipients have multiple CMV infections.

EPSTEIN-BARR VIRUS (EBV)

EBV is also a member of the **Herpes Virus Family**.

It is the same virus that causes mononucleosis ("mono"). It is transmitted the same as the CMV virus – either reactivation of the virus or transmission from the donor organ. Signs and symptoms of an EBV infection include low grade fever, extreme fatigue, sore throat, and swollen lymph glands in the neck. Treatment of EBV infections consists of supportive care and often, a reduction in your child's immunosuppressant medications. Reducing your child's anti-rejection medicines will allow your child's natural defenses to combat the EBV infection. In some cases, the EBV

virus continues to grow or proliferate within your child's lymph system causing swollen lymph nodes throughout your child's body. This condition is called **Post-Transplant Lymphoproliferative Disease (PTLD)**.

PTLD can be pre-cancerous but it can also develop into a lymphoma, which is a type of cancer. In some cases, chemotherapy is given to treat this condition.

HERPES SIMPLEX VIRUS (HSV)

There are 2 types of HSV – Type I and Type II. HSV

Type I is more commonly known as the virus that causes fever blisters or cold sores on the lips and in the mouth. **HSV Type II** is more commonly known as the virus that causes genital herpes and is spread through sexual intercourse. In both cases, the virus initially causes a tingling sensation followed by redness then small blisters. The fluid in the blisters is very contagious, and if broken open, can spread to other areas. HSV can be treated with a medication called Acyclovir. Should your child develop cold sores, please contact your transplant nurse coordinator.

VARICELLA ZOSTER (VZV, CHICKENPOX, SHINGLES)

Chickenpox is a very common childhood illness that most children tolerate well. For children who get chickenpox after they've received a transplant however, it can be very serious, even fatal. During your child's transplant evaluation, if it's determined that your child has not had chickenpox, we will ask you to have your child vaccinated with the Varivax, which is the vaccine that prevents chickenpox. If your child is too young to receive the vaccine and/or is transplanted prior to receiving the vaccine, you must notify the transplant office in the event your child is exposed to chickenpox so he/she can receive appropriate treatment, ideally within 72 hours of exposure.

This can sometimes either prevent or lessen the severity of chickenpox. If your child should develop active chickenpox, contact your transplant nurse coordinator immediately or the physician on call if after normal business hours so treatment with Acyclovir can begin. Your child will need to be admitted to the hospital.

In some people who have had chickenpox, the virus stays in the body and settles along nerves. When patients are stressed or immunosuppressed with medications, the virus can "wake up" and cause what's known as shingles. When shingles occurs, the patient will develop pain, itching, or tingling followed

by the development of small blisters. It usually is found on the chest, back, or hip but can occur on the face, arm or leg. It is usually only present on one side of the body. The fluid in blisters is contagious to people who have not had chickenpox. Treatment for shingles is the same as for chickenpox – Acyclovir until the lesions crust over. Your child may also need pain medicine until the shingles resolve.

FUNGAL INFECTIONS

The most common type of fungal infection that affects transplant recipients is **candida albicans**, which is a type of yeast. Most often this presents as **oral thrush**. Thrush is thick, white patches that are present on the tongue and inside cheeks. It can also be present in the throat and esophagus. Oral thrush is most often treated with a medication called Nystatin; in severe cases, it may be treated with another medication called Diflucan. Girls can also get **vaginal yeast (candida)** infections. This presents as white or yellow vaginal discharge and they may complain of itching and burning in the genital area. In most cases, vaginal yeast infections are treated with Diflucan.

There are many other types of fungus and molds that are in the environment that can be dangerous to transplant recipients if inhaled. These types of fungus and molds are most often found in construction areas (in the dust), barns, compost piles, and in bird droppings. If you are remodeling your home, check with your transplant nurse coordinator to determine if your child needs to stay elsewhere while the construction is being done. We do not recommend birds as pets.

SURGICAL COMPLICATIONS

REJECTION

What is Rejection?

There are five classes of rejection in lung transplantation:

- Hyperacute Rejection (generally preventable)
- Acute Rejection (most common)
- Chronic airway rejection (also known as Bronchiolitis Obliterans – see page 45)
- Chronic vascular rejection
- Antibody mediated rejection, also known as humoral rejection

HYPERACUTE REJECTION

Hyperacute rejection is caused by preformed antibodies (a type of protein in the blood that is responsible for recognizing and removing bacteria and abnormal cells). In this case the antibodies, called “HLA antibodies” recognize the proteins on the surface of the cells lining the blood vessels of the transplanted lungs. The resulting injury to the blood vessels may compromise the blood supply to the transplanted lungs.

- Hyperacute rejection is rare. However, the most severe form can cause complete rejection of the lungs within 24 hours.
- Patients may develop HLA antibodies in response to blood transfusions (antibodies to the cells from the blood donor), previous transplants, or during pregnancy (antibodies to cell surface proteins in the baby that aren't shared with the mother). Therefore patients who have had multiple blood transfusions or multiple pregnancies are at highest risk.
- A special blood test called the “PRA” is done prior to transplant to test for the presence of HLA antibodies.
- In cases where the PRA detects significant HLA antibodies, a crossmatch to the donor can be done at the time of transplant.
- Treatment of hyperacute rejection involves plasmapheresis, a process that removes dangerous proteins, including antibodies, from the bloodstream.

ACUTE CELLULAR REJECTION

Acute rejection occurs when immune system cells called T-lymphocytes identify foreign proteins on the surface of cells in the transplanted lung and cause injury to those cells. This is a normal function of the immune system and also occurs in response to tumors and to viral infections. The immunosuppression medicines (usually tacrolimus, mycophenolate and prednisone) your child will take after transplant inhibit this reaction. The T-lymphocytes become activated and multiply when they find foreign cells.

- Acute rejection usually occurs within the first six months and is usually detected at a mild stage.
- We obtain surveillance biopsies to check for acute rejection at one week after transplant, monthly for the first three months, every three months the first year, then every six months.

- Acute rejection is usually always treatable by adjusting your child's current immunosuppressant medications or by adding additional medications and treatments.
- The chance of your child developing rejection will likely decrease over time, however, it can still occur many years after transplant.

To review what we know about Acute Rejection:

- Acute rejection is a normal response of the immune system where lymphocytes recognize the transplanted lungs and invade and damage the lung tissue. The immunosuppression medications your child takes help prevent acute rejection.

How do I know if I have Acute Rejection?

Acute rejection may be without symptoms or have symptoms similar to a lung infection including:

- Fever, cough, difficulty breathing
- Decrease in pulmonary function tests (FEV1, FVC)
- Elevated Blood Count (WBC)
- Infiltrates and effusion (fluid outside the lung) on chest X-ray that may be confused with pneumonia.

What will happen if I get Acute Rejection?

- Treatment – Initial treatment of acute rejection grade A2 and greater is “pulse” doses of Solumedrol (an IV steroid) each day for three days.
- Two weeks after completion of treatment we repeat a biopsy to make sure the rejection has cleared.
- If the rejection is still there on the second biopsy and it is not mild or improving, we will treat with a special IV antibody called ATGAM that attacks and destroys the T-lymphocytes.
- Most patients have at least one episode of acute rejection and in most patients the acute rejection resolves after treatment with Solumedrol.

HUMORAL REJECTION

Humoral rejection occurs when immune system cells called B cells and plasma cells produce antibodies against the transplanted organ (new heart and lungs). These antibodies are called donor specific antibodies and bind to blood vessels in the new heart and lungs. In general antibodies are proteins found in the blood that detect and destroy foreign organisms.

During Humoral Rejection antibodies are directed against the new heart and lungs.

How do I know if I have Humoral Rejection?

We perform regular screening for Humoral Rejection. This is detected by blood work and biopsy tissue from the heart and lungs. Donor Specific Antibodies (DSA) blood work will be performed frequently in the first three months after transplant and during routine evaluations. If a patient is DSA positive, meaning blood testing reveals DSA in blood, screening may be performed more often. Another screening tool used in the diagnosis of humoral rejection is special staining of biopsied tissue from the heart or the lungs. Every time a biopsy is performed a special test called a stain is performed looking for Humoral Rejection.

What will happen if I get Humoral Rejection?

Some patients have DSA and remain symptom free and require no treatment.

However, patients that develop DSA in blood and have biopsy stain positive for humoral rejection will require plasmapheresis to remove the antibodies from their blood. Treatment requires admission to the hospital for minimum of five days. Depending on the severity of the rejection, other treatments to remove B-cells and plasma cells may also be considered. The treatment regimen will be discussed with you further should your child develop Humoral Rejection.

BRONCHIOLITIS OBLITERANS

What is Bronchiolitis Obliterans?

Bronchiolitis Obliterans is the most important disease that limits long term survival after lung transplantation. It is also the most common complication occurring more than a year after lung transplant. Bronchiolitis Obliterans goes by different names (OB, BO, chronic lung rejection) that refer to lung damage seen through the microscope on a biopsy of the lung. Some patients have symptoms of BO, but have not had a biopsy that confirms the diagnosis. Such patients are said to have **Bronchiolitis Obliterans Syndrome (BOS)**.

The damage to the lungs in BO occurs in the bronchioles (hence the name bronchiolitis). The bronchioles are the smallest airways in the lung. Air going into the lungs goes first to the trachea, then to the bronchi, then to the bronchioles, before finally reaching the alveoli. It is in the alveoli where gas

exchange occurs (oxygen is brought in and carbon dioxide is removed).

Under the microscope the bronchioles of patients with BO initially show signs of injury, then scarring. In advanced cases, the bronchioles are destroyed (hence the name "obliterans"). As this occurs, lung function deteriorates. When enough of the bronchioles have been lost symptoms of shortness of breath and low oxygen saturation occur. The exact cause of the lung injury is unknown (and there are probably multiple causes). Our experience suggests that patients who have prolonged ischemic times (the amount of time the lungs are out of the donor and surgically placed in the recipient) during their transplant, multiple episodes of acute rejection, or chronic infections (either bacterial or viral) are at the highest risk. Children who receive their transplant before 3 years appear to be at lower risk. Patients with GERD and/or donor specific antibodies may be at higher risk.

To review what we know about BO:

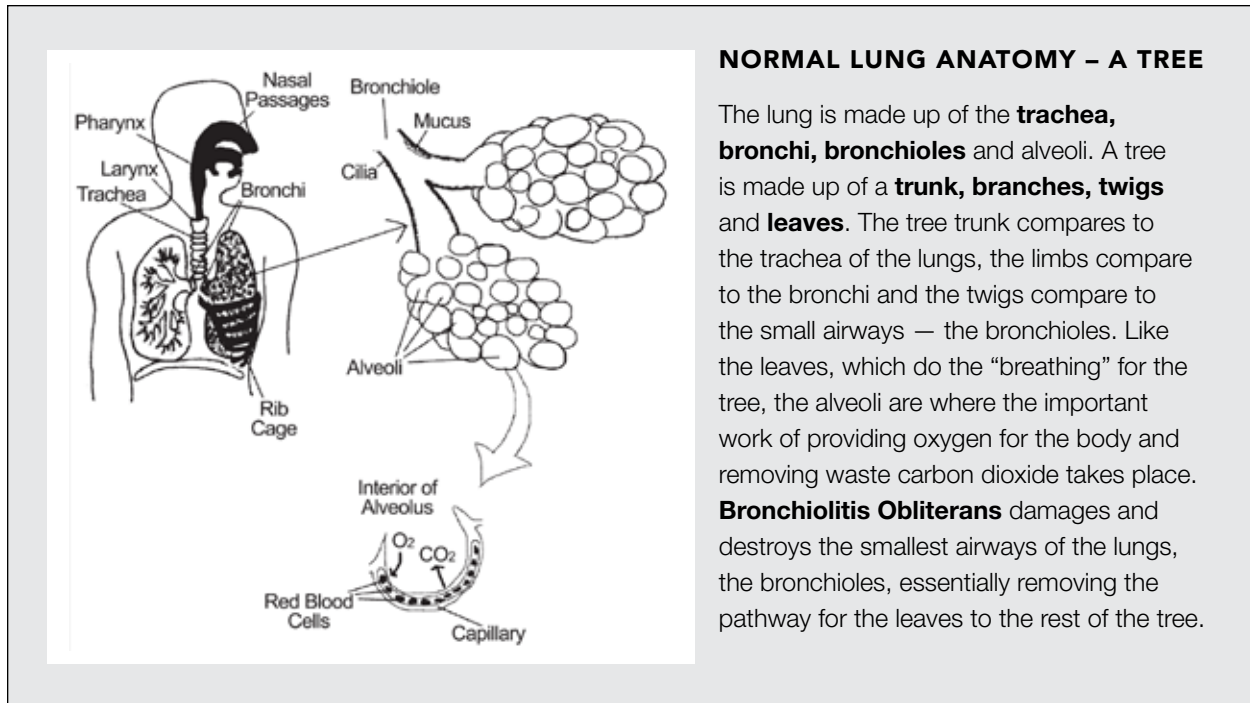
- BO is the result of damage to the smallest breathing tubes in the lungs that cannot be repaired.
- BO is poorly understood but thought to be the result of repeated lung injury, perhaps from different causes.

How do I know if I have BO?

Patients who develop BO may have chest cold symptoms or initially may have no symptoms at all. The most useful EARLY indicator of BO is a drop in lung function (FEV1) that is otherwise unexplained. Symptoms that develop once there is significant progression of BO include:

- dry cough or a cough that produces sputum not related to a lung infection
- new onset of shortness of breath with or without exercise
- decrease in oxygen saturations usually occurs after other symptoms, this is usually a late sign
- decrease in lung function (FEV1) that is otherwise unexplained

The diagnosis of BO is usually made by more than just one test. If there is an unexplained drop in lung function (FEV1) we will obtain a ventilation/ perfusion (VQ) scan and a high-resolution chest CT scan. If these show findings suspicious for BO, a lung biopsy



is necessary to confirm the diagnosis. The pieces of tissue obtained from transbronchial biopsy (the biopsy done during a routine bronchoscopy in the APC) may not be large enough to detect BO. Therefore an open lung biopsy may be necessary. An open lung biopsy is done in the operating room under general anesthesia. The incision is usually made through the lung transplant scar.

What is BOS?

BOS is short for Bronchiolitis Obliterans Syndrome. Because BO often requires an open lung biopsy to diagnose, some centers prefer to use a clinical scheme to guide therapy. A clinical scheme means diagnosis not based on biopsy of lung tissue, but rather diagnosis based on symptoms and pulmonary function testing. BOS is a decrease in FEV1 for which there is no other identified cause (such as acute rejection, infection or airway narrowing). BOS is graded from 0 (no evidence of BOS) to 3 (severe decrease in FEV1).

What will happen if I get BO?

There appear to be three patterns of progression in BO (from most to least common):

1. Slow onset and slow progression.
2. Rapid onset with initial decline in lung function and then stabilization of lung function for months to years.

3. Rapid onset and relentless progression.

Treatment does not cure BO but it may halt the progression of the lung damage. The treatment is tailored to each individual patient based on his or her lung transplant course. Your physician may feel that it is necessary to perform a pH probe at this time, even if your child has had it done in the past. Your child may also be started on Azithromycin. Another change in medication may include removing Cellcept, and adding a drug called Methotrexate. Please refer to medications section for information on these drugs. The first step is generally a treatment with an antibody medicine called ATGAM. However, depending on your child's unique circumstances this may not be the treatment chosen. Before beginning therapy your physician will speak directly to you about the treatment plan for your child and answer all of your questions.

In the event that ATGAM therapy does not work, some patients may undergo a treatment called photopheresis. This process acts by targeting the T cells and irradiating them in the process. This treatment will not cure BO.

GASTROESOPHAGEAL REFLUX

Gastroesophageal reflux disease (GERD) is a condition in which the stomach contents (food or liquid) leak backwards from the stomach into the esophagus (the tube from the mouth to the stomach) and possibly the lungs (called aspiration). This action

can irritate the esophagus, causing heartburn and other symptoms. It may occur without any symptoms.

A pH probe will be performed 2 months after transplant to assess for GERD.

A pH probe measures the pH in the esophagus to determine if acid is coming up from the stomach. A pH probe study is helpful in determining if you have gastroesophageal reflux disease (GERD). GERD and aspiration are contributing factors in the development of Bronchiolitis Obliterans. Bronchiolitis Obliterans is the most important disease that limits long term survival after lung transplantation. For the pH probe study, you will be admitted to the hospital for a 23 hour observation stay. A thin plastic tube is passed through one of your nostrils and advanced into the esophagus. The esophagus is the tube from the mouth to the stomach. At the end of the tube that is placed in the esophagus is a sensor that measures the pH. The other end of the tube is connected to a monitor. A chest x-ray will be done after insertion of the tube to confirm correct placement. The tube will be taped to your cheek for securement.

If this study is positive for GERD, you will be referred to a general surgeon for evaluation of a Nissen Fundoplication procedure.

This procedure is done to prevent stomach contents from returning to the esophagus and to prevent these contents from aspirating to your lungs. The upper portion of the stomach is wrapped around the lower portion of the esophagus. This is done to tighten the lower esophagus so food contents and acid can go down into the stomach, but cannot return to the esophagus. This procedure helps to prevent you from having gastroesophageal reflux. Your surgeon will discuss with you how this procedure is done. You will be hospitalized for this procedure until you have recovered. You will need to be on a soft diet after this procedure for approximately 2 months.

SEIZURES

A frequent and often worrisome side effect of some immunosuppressants is **tremor**. Tremors are an uncontrollable trembling or shaking of the limbs that is sometimes accompanied by numbness.

Seizures, however, are a less common but more serious side effect of Cyclosporine and Prograf. Seizures may be tonic/clonic in nature (jerking of arms or legs or whole body) or focal in nature (staring, eye

deviation, or unable to speak). These medicines can lower the “seizure threshold” in the brain and then allow the seizures to happen. These seizures are not usually life threatening, however, if they occur outside of the hospital, call 911 for emergency medical assistance. If seizures occur, your child would have to be admitted to the hospital for medical treatment including observation and anti-seizure medications, if needed. Often the anti-seizure medicines can be stopped after several months.

DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS)

Distal Intestinal Obstruction Syndrome (DIOS) is a disorder that can occur in children with Cystic Fibrosis (CF). With DIOS, stool becomes hardened within the small intestine and the child is unable to pass the stool. Additionally, some patients have slow intestinal motility (movement) that contributes to the problem. This slowed motility can also be caused by medications, especially pain medications.

Children with DIOS can have acute or chronic symptoms including:

- Crampy lower abdominal pain
- Decreased frequency of bowel movements
- Abdominal distention
- Vomiting (with severe obstruction)

Treatment is often ongoing, using laxatives to keep stool soft as well as to keep bowel movements regular. In more severe cases, DIOS may need to be treated with either stronger medication such as Go-Lytely, with enemas to relieve the obstruction, and in rare cases, surgery.

PTLD – POST-TRANSPLANT LYMPHOPROLIFERATIVE DISEASE (PTLD)

Transplant recipients are at risk of developing **Post Transplant Lymphoproliferative Disease (PTLD)**. PTLD is a form of cancer that occurs more frequently in children who have never been exposed to the Epstein-Barr Virus (EBV) prior to transplantation, but who are exposed after transplant. The exposure can be from the heart and lungs of a donor who was EBV positive or from exposure to EBV in the general environment. EBV is the virus that causes mononucleosis. Children who have compromised immune systems due to powerful anti-rejection drugs

are vulnerable to EBV infection complications. An individual who has had EBV in the past will always “test positive” for the virus. This is because once the initial infection with the virus is over, the virus lies dormant in their system. EBV can still be detected in the blood of a person who had the infection, even if it was many years ago. PTLD tends to develop in children who are newly infected with EBV some time after transplant although a child who has had EBV in the past may develop PTLD. The EBV may reactivate because the child is immunosuppressed. The Epstein-Barr virus causes “B cells” to proliferate, or grow. B cells are normally occurring cells found in the body but they can become “hyperactive” in response to an EBV infection and cause tumor growth.

It most commonly occurs in the first two to three months following transplantation but may occur many years after transplant. Some patients do not have symptoms at all and the tumor is found during routine examination. Other patients have fever or flu-type symptoms. Some patients complain of symptoms related to the organ impairment at the site that the tumor is found. The prognosis of the disease depends on the amount and location of tumor formation and the disease's response to treatment. In a small number of cases PTLD may be fatal.

There are a number of treatment options for PTLD. Decreasing the dose of immunosuppression drugs may cause the tumor to shrink or disappear. Some forms may be treated with Rituximab, an antibody that removes B-cells. The risk of organ rejection increases when immunosuppression doses are decreased. The transplant team will follow your child closely for signs of rejection. Decreasing the dose of immunosuppression drugs may not be effective in the treatment of PTLD. Surgery, chemotherapy and/or radiation therapy may be necessary to treat PTLD. The lung transplant team will ask for help with the management of PTLD from medical doctors who specialize in cancer treatment (Oncologists).

HYPERTENSION

Hypertension, also known as high blood pressure, can be very common after solid organ transplant. Most often this is due to the amount of IV fluids needed during and after surgery but may also be due to medications that are needed after transplant. Sometimes your child's blood pressure will return to normal before discharge. If not, there are many

different medications available to treat hypertension. It is important to remember that diet and exercise are important factors in helping to keep blood pressure in good control.

RENAL INSUFFICIENCY

A common post-transplant complication is renal insufficiency, or decreased function of the kidneys. Decreased kidney function is a known side effect of the anti-rejection medicines Cyclosporine and Tacrolimus. Chronic antibiotic therapy (especially aminoglycosides) and post-operative complications such as bleeding and hypotension (blood pressure that is too low) can also contribute to decreased kidney function. Signs and symptoms of renal insufficiency include hypertension and elevated kidney function tests (i.e. elevated creatinine and BUN). Most transplant recipients have some degree of renal insufficiency, especially in the first few months post-transplant when their anti-rejection drug levels are at their highest. In many cases, once these drug levels decrease, the kidney function improves. Some patients, however, have more significant and progressive renal insufficiency requiring ongoing monitoring by a kidney doctor (nephrologist). In rare cases, these patients may have such severe renal insufficiency that they will need dialysis. There isn't any way to determine how much renal insufficiency your child will experience after transplant; your transplant team will continually monitor your child's kidney function post-transplant for evidence of decreasing kidney function.

OBESITY

A large number of patients gain weight during the first six to twelve months after transplant. Your medications do not cause the weight gain. Medications such as Prednisone cause an increased appetite so you are tempted to eat more calories than needed. It is important to begin (with permission from your transplant team) an exercise program after transplant. We encourage exercise and have never “lost” an organ due to activity. Your transplant team includes a dietitian who may guide you with a healthy eating and exercise plan.

DIABETES (Diabetes Mellitus)

A condition characterized by high blood sugar resulting from the body's inability to use sugar (glucose) efficiently. In Type 1 diabetes, the pancreas

is not able to make enough insulin; in Type 2 diabetes, the body is resistant to the effects of available insulin. Diabetes is one of the leading causes of kidney disease.

DIABETES POST-TRANSPLANT

Post Transplant Diabetes Mellitus (PTDM) is seen in approximately 5 to 40 percent of post-transplant patients. The leading cause is medication. Steroids increase glucose levels and cause the body to resist insulin. Cyclosporine and Tacrolimus inhibit the release of insulin and add to the effect of the steroids by causing elevated blood sugar. Certain factors predispose patients to developing PTDM: increase in age, family history of diabetes, and cystic fibrosis. African Americans are at higher risk.

The treatment of PTDM may depend on the severity of the hyperglycemia. A change in diet, decrease in steroids, and/or insulin may be used to treat elevated blood sugar.

HAIR

Excess hair growth on both the face and body in the transplant population is an unfortunate side effect of some of the anti-rejection medications. This can pose emotional problems in the pediatric and female patients. Prednisone can cause increased hair growth. Children that are on higher doses of Prednisone will see more changes than children on low doses of those medicines. The increased growth will be seen about two to four weeks after beginning the medicines. Patients that have not seen the increase in hair growth by 3 months after the transplant are less likely to have this side effect at all.

Most children remove hair with depilatory cream, shaving or waxing. Every method has drawbacks. Shaving or clipper cutting seems to be more preferred method for older children and boys. With waxing, the results last longer, however, it is often more expensive and painful than the other options.

Our dermatology department recommends **Nair Gentler Formula Cream Hair Remover with Baby Oil, For Face Too**. We have a special protocol to remove the hair. Be sure to test a skin area according to the directions on the bottle. This cream can cause irritation to the eyes and mucous membranes, so apply it carefully.

Instructions for Hair Removal:

- Apply Nair to the affected area. Leave on for 5 minutes **ONLY**.
- Remove with a wet wash cloth, rub with the hair and press firmly.
- Wash skin with soap and water after removing hair. Dry the area and then apply Hydrocortisone ointment 0.5 percent to the area.
- You only need to apply the Hydrocortisone once after removing the hair.
- It is very important to only leave Nair on the skin for 5 minutes, or severe burn may develop.

If the hair growth becomes psychologically debilitating, talk to your transplant team as there may be other medication options for your child.

HEARING IMPAIRMENT AND LOSS

Hearth-lung transplant recipients are at risk for hearing loss after transplant from medication side effects. Hearing loss can be caused by high doses of diuretics needed by infants requiring immediate ventilator support at birth, particularly those who go on an oscillating ventilator. Children with Cystic Fibrosis who have received frequent or prolonged courses of aminoglycoside antibiotics are also at risk for hearing loss. We have a low threshold for hearing evaluation when concerns arise. Treatment includes hearing aids and in rare cases cochlear implants.

GROWTH AND DEVELOPMENT

Most patients who require heart/lung transplant have growth impairment as a result of their underlying disease. The use of steroids after transplant will further impair growth. Although patients will remain on steroids indefinitely, we attempt to wean the dose over time in order to minimize the growth impairment. Your child's nutritional status will be monitored closely. If your child is not gaining weight appropriately, dietary supplements such as special formulas or nutrition additives, may be added to your child's daily diet. In some cases, it may be necessary to give extra nutrition to your child through a nasogastric or gastrostomy tube or intravenously using Total Parenteral Nutrition (TPN). Our team includes a dietitian who will make nutrition recommendations both before and after transplant.

Your child's normal childhood development may also be delayed due to their chronic illness. There are many resources available to help your child reach developmental milestones. We will monitor your child's development and recommend services as needed.

PSYCHOSOCIAL RISKS

Psychosocial risks of transplant may include depression, post traumatic stress disorder (PTSD), generalized anxiety, anxiety regarding dependence on others, and feelings of guilt.

TRANSITIONING HOME AFTER EXTENDED STAY IN ST. LOUIS POST-TRANSPLANT

After discharge from the transplant hospitalization, the next milestone families look forward to comes three months after transplant. If everything is going well following the full evaluation scheduled at that time, patients and their families are encouraged to return to their home community.

Moving home is usually a time of mixed emotions for our patients and their families (and the transplant team too!). For many families this is a reunion, as one parent often remains at home. Regardless, it is generally a return to a more extended support system. On the other hand, leaving the security of frequent visits and close proximity to the transplant center can be intimidating.

Our goal is to teach you enough about heart/lung transplant during your stay in St. Louis so that you are

well prepared to care for your child upon return to your home community. Rest assured, however, that we are only a phone call away!

We ask you to do the following in preparation for the move back to your home:

1. Identify a pediatrician, a pediatric cardiologist and a pediatric pulmonologist for your child. Please provide us with the name, address and phone numbers for these individuals so that we may contact them prior to your move. Make arrangements to see them within a few weeks after your arrival home so that they may become familiar with your child's condition.
2. Purchase a copy of most recent chest X-ray, front view only. X-rays are available from the film library. The front desk in the radiology department can direct you there.
3. There are a few laboratory tests (particularly immunosuppression drug levels) that we will ask you to obtain monthly and send to St. Louis to be run. We will provide you with secure laboratory tubes and mailer kits. Before you leave St. Louis, you will be given a summary of the labs to be obtained, which ones need to be mailed, and how frequently they should be obtained.
4. Don't forget that we will be eagerly looking forward to seeing you back in St. Louis three months later for your six month evaluation! We will be happy to schedule that evaluation prior to your departure.

COMMUNICATIONS AND MEDIA RELATIONS

CONTACTING THE DONOR FAMILY

Following your child's transplant, we will provide you with information about contacting the donor family. The decision to write to the donor family is a very personal one; we urge you to write a letter when you feel the time is right for you.

MEDIA RELATIONS

The overall purpose of media relations is to increase and manage the public awareness of St. Louis Children's Hospital (SLCH) and its role in the community. This requires a delicate balance between the needs of reporters, patients, parents, physicians, and SLCH employees while maintaining a high level of privacy and adherence to our mission to "Do what's right for kids."

In addition to protecting the health and general welfare of its patients, a hospital's responsibilities include protecting the patient's legal rights as well. That includes the legal right to privacy concerning medical information. To ensure patient privacy and confidentiality, we require a media relation staff escort anytime the media come into SLCH or interview a patient. We also require a signed consent form from each patient and/or parent.

When working with transplant patients and their families, the most common theme is the need for organ donations and creating awareness of the need for organ donations. When a child is listed for a transplant at St. Louis Children's Hospital, there are several reasons why the family might want to contact the media relations department:

- If the family is working with COTA or another fundraising group, and would like to contact the local media in their hometown, or the media in St. Louis.
- If the family has already had some media attention in their hometown, and their local media would like to interview the patient at St. Louis Children's Hospital.
- If a reporter calls the family and would like to set up an interview with their transplant physician.
- If a reporter would like to know the patient's current condition.

If you would like further information, please contact the Media Relations Department at 314.286.0416 or 314.286.0304.

RESEARCH AND PATHWAYS TO THE FUTURE

St. Louis Children's Hospital and Washington University are committed to making heart/lung transplant outcomes better. We can only do so with your help. You and your child will join a team that also includes your physicians and scientists here at Washington University and around the world. As a member of that team, you and your child will be asked to participate in both clinical and basic research studies. As with all research studies, participation is entirely voluntary and will not impact your ability to receive standard care.

Each research project you and your child are asked to participate in will have been reviewed and approved by the Washington University Human Studies Committee (an institutional review board). The Human Studies Committee is responsible for ensuring the protection of any patient participating in a research study. Before enrolling in any study, you / your child will be asked to review and sign a consent form that outlines the potential benefits of the study and the potential risks to you and your child. Minor children will be asked for their assent as well. The Human Studies Committee is also responsible for monitoring all ongoing studies.

It has the authority to halt or require modifications to any study in order to ensure the safety of participants.

AUTOPSIES

When a transplant recipient dies, sometimes there are questions that remain unanswered for family members and physicians. The goal of an autopsy is to understand the reasons that lead to death. It is an opportunity for death to teach those who live, a gift comparable to the organ donation that gave the transplant recipient a second chance at life. For these reasons, we seek permission to perform an autopsy on all transplant recipients who die. An autopsy is a special type of surgical operation performed by an autopsy assistant and a pathologist (a specially trained physician). It may be performed on the whole body or on specific body parts. It is performed with respect and compassion as well as in accordance with religious beliefs. When completed, it allows for any type of funeral ceremony, including an open casket. It generally takes 8 to 12 weeks for the complete autopsy report to become available. When it is available, the results will be discussed with you by your child's physician.

GLOSSARY

A

ABO compatibility There are four blood types: O, A, B, and AB. Type O is the universal donor and type AB is the universal recipient. Type O can receive only type O blood, Type A can receive type A or O. Type B can receive type B or O. Type AB can receive A, B, AB or O.

Acquired Not caused by hereditary or developmental factors but by a reaction to environmental influences outside of the organism.

Acute Beginning abruptly.

Acute Rejection Attempt of body to destroy transplanted organ usually within the first year after transplant.

Adverse Reaction An unintended response from a drug.

Allocation System of ensuring that organs/tissues are distributed fairly to patients in need of transplant.

Allograft A graft between 2 individuals who are of the same species but have genetic differences, i.e., Human – Human.

Anaphylaxis A severe allergic reaction that can be fatal.

Anastomosis A surgical joining of two ducts, blood vessels, or bowel segments to allow flow from one to the other.

Anesthesia The absence of normal sensation, especially sensitivity to pain. Topical, local, regional, or general.

Antibody A substance that is produced by the immune system in response to specific antigens, helps the body fight infection and foreign substances.

Antigen Substances that trigger an immune response.

Antihypertensives Substance or procedure that lowers high blood pressure.

Antiviral Destructive to viruses.

Arterial Line A catheter inserted into an artery which allows for continuous direct blood pressure readings as well as access to the blood supply for monitoring labs.

Arteriogram An X-ray of the arteries taken with the aid of a dye.

Ascites An abnormal accumulation of fluid in the abdomen.

ATN – Acute Tubular Necrosis Reversible kidney damage resulting in delayed kidney function.

B

B-Cell A specialized white blood cell responsible for the body's immunity. B-cells function in antibody production.

Bacteria Tiny organisms (germs) that cause infection.

Bile A greenish-yellow fluid produced by the liver which is needed to help your body use fats and vitamins.

Biopsy The removal and examination of tissue to determine how well the organ is working or if it is rejecting.

Bladder Part of the urinary tract that receives and stores urine from the kidneys until you urinate.

Bloodwork Laboratory analysis performed on blood sample that is usually extracted from a vein in the arm using a needle, or via finger stick.

Bone Densitometry An enhanced form of x-ray technology that is used to measure bone loss.

Bronchoscopy Procedure used to diagnose infection and/or rejection of the lungs.

Broviac A type of IV that is placed through a large vein in the chest that allows for delivery of medicine and drawing of blood for labwork.

BUN – Blood, Urea, and Nitrogen A waste product normally excreted by the kidneys. The BUN, along with creatinine, will represent how well the kidney functions.

C

Cadaver A donor that has recently expired for reasons that do not affect the function of an organ to be transplanted.

Candida A type of yeast.

CAPD – Continuous Ambulatory Peritoneal Dialysis A cleansing fluid that fills a person's abdomen and then is drained to filter out wastes and excess fluid from the body.

Cardiologist A doctor who specializes in treating heart disorders and diseases.

Catheter A hollow, flexible tube that can be inserted into a vessel or cavity of the body to withdraw or instill fluids.

Central Line A type of IV that goes directly to the heart through a large vein in the shoulder or neck.

Chest X-ray A picture of the lungs and upper body taken by an X-ray machine.

Cholangiogram Dye is injected into the bile ducts of the liver to show leaking or blockage within the bile duct.

Cholangitis Inflammation of the bile ducts caused by bacteria from the bowel.

Cholestasis Stoppage or suppression of bile flow.

Cholesterol A fatty substance that comes partially from foods eaten.

Chronic Developing slowly and persisting for a long period of time.

Chronic Rejection Slow failure of the transplanted organ/tissue.

Cirrhosis A disease of the liver in which normal, healthy tissue is replaced with nonfunctioning tissue and healthy cells are lost.

CICU Cardiac Intensive Care Unit

CMV/Cytomegalovirus A viral infection common to immunosuppressed patients

Coagulation Blood clotting.

Compatible The degree to which the body's immune system will tolerate the presence of foreign material (organ, blood) without an immune reaction.

Compliance The degree to which someone follows medical instructions and protocols.

Congenital Present at birth.

Contraindication Prohibited.

Corticosteroids Hormones secreted by the adrenal gland. Can be man-made and given for immunosuppression.

Creatinine A substance found in blood and urine monitored to determine kidney function.

Cross Match A test which mixes a potential donor's blood with the recipient's blood and after several hours is examined under a microscope to determine compatibility. If there is cell death, the result is positive which means that the recipient has cells that attack the donor's blood. If there is no cell death, the result is negative and the recipient and donor are compatible.

CT Scan Special x-ray with sophisticated equipment and computers to produce multiple images or pictures of the inside of the body.

D

Dexascan A type of X-ray that measures the density of the bones.

Diabetes A disease in which patients have abnormally high sugar levels in their blood.

Dialysis Cleaning the body of waste by artificial means.

Diastolic Pressure The bottom number of the two blood pressure numbers which measures blood pressure when the heart is at rest.

Discharge To release from the hospital or from care.

Discontinue or D/C To stop.

Distention Visible increase in size of organ beyond normal.

Diuretic A drug given to promote the formation and excretion of urine.

Dormant An infection that is currently not active.

Drain A type of tube that may be attached to a collection device that allows an accumulation of fluid to be removed.

Dressing Change To remove an old covering of a wound or incision and replace, with clean or sterile technique.

E

Echocardiogram A test that uses sound waves to create pictures of the heart to measure size and function.

Edema A build-up of too much fluid in the body tissue resulting in swelling.

EKG/Electrocardiogram A measurement of the current through the heart that tells us how the heart is working.

Electrolytes Refers to the dissolved form of a mineral in the body, sodium, potassium, magnesium, etc.

Encephalopathy The liver no longer clears the wastes in the blood. Wastes build up and cause lethargy and coma.

Endocrinologist A doctor who specializes in treating diseases of the endocrine system (pancreas, thyroid, etc). These physicians manage the care of patients with diabetes.

Endotracheal Tube A breathing tube that is connected to a ventilator that helps you breathe.

Epstein-Barr Virus (EBV) – Mononucleosis A type of virus that causes fever, sore throat and swollen lymph nodes.

ERCP – Endoscopic Retrograde

Cholangiopancreatography An X-ray procedure that helps evaluate the liver and the bile ducts. Dye is injected into your biliary tree and X-rays are taken. A tube with a small light attached (endoscope) will look down your throat to examine the bile ducts.

Erythropoietin A hormone that helps make new red blood cells.

Evaluation A series of tests and meetings with the members of the transplant team to make sure that each candidate is ready for transplantation.

Extubate To remove a breathing tube.

F

Fibrosis Scarring caused by healing response to injury, infection or inflammation.

Foley Catheter A tube is inserted into the bladder via the ureter which is connected to a pouch for the collection and measurement of urine.

Fulminant Hepatic Failure A rapid, sudden and severe insult to the liver which can cause liver failure.

Fungal Infection An infection caused by a type of fungus. Can be life threatening in an immunosuppressed patient.

G

Gastroenterologist A physician who specializes in the treatment of disorders of the digestive tract.

Gastroesophageal Reflux When stomach contents leak backward from the stomach into the esophagus.

Generic A drug's chemical name.

Genetic Referring to hereditary, birth.

Gingival Hypertrophy Enlargement of the gums. A common side effect of Cyclosporine.

Glucose A type of sugar found in the blood.

Graft A transplanted tissue or organ (kidney, heart, lung, bone marrow or liver).

Graft Acceptance A transplanted organ or tissue that is accepted by the body and functions properly.

H

HCT – Hematocrit A measure of the amount of red blood cells in the blood.

Helper T-Cell A white blood cell that tells other parts of the immune system to fight infection or foreign material.

Hemodialysis A method of dialysis in which blood is cleaned of waste by circulating through a machine outside of the body.

Hemoglobin – Hgb A compound in the blood that carries oxygen to the cells.

Hemorrhage A rapid loss of blood/excessive bleeding.

Hepatic Having to do with the liver.

Hepatologist A physician who specializes in treating liver disease.

Hereditary A condition, characteristic or disease that is passed from parents to offspring.

HIDA Scan Dye is given through an IV and flows through the liver. An X-ray is taken to show the flow and possible blockages in the bile ducts that drain the liver.

Hirsutism An excessive increase in hair growth. A common side effect of Cyclosporine and steroids.

Histocompatibility The compatibility of the antigens of donor and recipient transplanted tissue.

HLA – Human Leukocyte Antigen Genetically determined series of antigens that are present on white blood cells and tissues.

Humoral Rejection B-cell mediated rejection that can occur at any time post transplant.

Hyperacute Rejection Very rare rejection that occurs very suddenly and unexpectedly. Usually occurs within the first few hours after surgery.

Hypertension High blood pressure.

I

Immune Response A defensive reaction to foreign material by the immune system.

Immunity Being able to resist a particular infectious disease.

Immunization Resistance to an infectious disease is induced by giving a vaccination.

Immunosuppression Prevention or suppression of the immune response either by drug therapy or by disease.

Intravenous (IV) Into or within a vein. Also refers to fluids or medications that are infused through a needle or catheter that is inserted into a vein.

Intubated A breathing tube inserted into the mouth or nose to the throat. The tube is connected to a breathing machine that will help him/her breathe until they are strong enough to breathe on their own.

Invasive A diagnostic or therapeutic technique that requires entering the body.

IVP – Intravenous Pyelogram Dye is injected into a vein. The dye concentrates in the kidneys and makes them show up on an X-ray. The doctor looks at the X-ray to see if there are two functioning kidneys with normal internal structure. Used in evaluating potential kidney donors.

J

Jackson-Pratt A small drain that is sometimes placed near an incision to drain any blood or fluid that may accumulate.

Jaundice Yellowing of the eyes and skin caused by an increased amount of bilirubin.

K

Kangaroo Pump A type of feeding pump used to deliver tube feedings.

Kidney An organ that rids body of waste materials and maintains fluid balance through the production of urine.

L

LRD – Living Related Donor A blood relative that donates an organ.

Lymphocyte A white blood cell.

M

Mononucleosis – (EBV – Epstein-Barr Virus)

A type of virus that causes fever, sore throat and swollen lymph nodes.

MRI – Magnetic Resonance Imaging A type of X-ray that uses magnetic waves to take pictures of tissues.

Myalgia Muscle aches and pains.

Myopathy Muscle disorder that causes severe weakness.

N

Nasogastric Tube A tube inserted through the nose that drains the stomach of excess bile to prevent nausea.

Nephrologist A doctor that specializes in treating problems involving the kidney.

Neuropathy A breakdown of the peripheral nerves. Symptoms are numbness and tingling in extremities.

Neutropenic Severe decrease in the amount of white blood cells.

Noncompliance Failure of the patient to cooperate by doing what is necessary or required for his/her medical care.

Noninvasive Does not require skin to be broken or body entered.

NPO – Nothing By Mouth Term used when someone is without food/drink prior to exam/procedure.

O

Oncologist A doctor who specializes in treating patients with cancer.

OPO – Organ Procurement Organization

Link between the potential recipient and donor. Responsible for retrieval, preservation and transportation of organs for transplantation.

Opportunistic Infection Infections that in healthy persons would not pose a threat but in immunocompromised persons can be very harmful.

Organ Part of body made of tissues specialized to perform a certain function.

Osteoporosis Weakening of the bones.

OTC – Over the Counter A type of medicine or product that does not require a prescription.

P

PACU Post Anesthesia Care Unit

Papilloma Virus Viruses that cause warts on hands, face and fingers.

PCP – Pneumocystis Carinii Pneumonia

A type of pneumonia that is seen primarily in immunocompromised patients.

PELD – Pediatric End-Stage Liver Disease A scoring system for pediatric liver patients. Based on certain lab values and growth failure; assigned a number 0-40 based on need.

Percutaneous Through the skin.

Peripheral IV A small IV that is placed in the arm, hand or foot for delivery of IV fluids or medicines. Usually placed during surgery.

PFT – Pulmonary Function Tests A series of tests used to determine the ability of the lungs to exchange oxygen and carbon dioxide.

pH Probe A tube inserted through the nose that is used to identify the presence of acid reflux.

Platelet A small blood cell needed for blood clotting.

P.O. By mouth.

Primary Care Physician Pediatrician or doctor that follows patient for primary illness.

PRN As needed.

Prophylactic An agent or regimen used to prevent an infection or disease.

PTLD (Post-Transplant Lymphoproliferative Disorder) A type of cancer that attacks the lymphatic system of some immunocompromised patients.

Pulmonologist A doctor who specializes in treating lung disease.

Pulmonary Functions Tests Measures size of lungs and how fast air can move in and out of them, and how well air (oxygen) travels to the blood supply.

R

Recurrence Reappearance of a sign or symptom of a disease after a period of remission.

Rejection An immune response against the transplanted tissue which if not successfully treated will result in graft failure.

Renal Having to do with the kidneys.

Resistance Ability of a virus to fight the effects of a treatment because the information in the virus changed. Can be caused from a person's noncompliance with certain medicines.

S

Sensitized Being immunized or able to mount an immune response against an antigen by previous exposure to that antigen.

Shingles A type of varicella zoster, characterized by a painful, blistering rash on one side of the body.

Stent A tube used to support openings and vessels during and after surgical procedures.

Stricture or Stenosis A narrowing or passage in the body.

Systolic The top number of the blood pressure, measures the maximum blood pressure as the blood is pumped out of the heart.

T

T-Cell White blood cell responsible for the body's immunity. Can destroy cells infected by viruses, graft cells and other altered cells.

T-Tube A tube placed in the bile duct to drain bile externally into a small bag.

Thrush A fungal infection in the mouth.

Tissue Typing A blood test that evaluates the closeness of tissue match between the donor's organ and the recipient's HLA antigens.

Total Parenteral Nutrition (TPN) – Parenteral (Intravenous) Nutrition A method of supplying nourishment to children unable to eat.

Transplant To transfer an organ or tissue from one person to another or from one body part to another to replace a diseased structure, restore function, or to change appearance.

U

Ultrasound A test that uses sound waves to create pictures of organs and tissues.

UNOS – United Network for Organ Sharing
Governing body that oversees organ transplantation in the U.S.

Ureter A tube that carries urine from kidney to the bladder.

URI Upper respiratory infection.

UTI Urinary tract infection.

V

Vaccine Protects a person against infection and/or disease. Made from killed or weakened forms of the disease and given to cause an immune response to create resistance to a certain disease.

Varicella Zoster Virus that causes chicken-pox and shingles.

VCUG – Voiding cystourethrogram A bladder and kidney X-ray.

Ventilator Breathing machine.

Virus Small disease causing germs that can only multiply when inside the cell of another organism.

Vital Signs Pulse, temperature, blood pressure, respiration.

V/Q Scans Ventilation perfusion scan of the lungs. Indicates air exchange and blood flow in the lungs.

W

Waiting List A nationwide computerized network called UNOS. All transplant centers in the U.S. belong. Supervised by the federal government to ensure that patients throughout the country receive organs as soon as they become available.

WBC – White Blood Cell Composed of several different types that all work to fight infection.

CONVERSION TABLES

WEIGHT (MASS)

Pounds to Kilograms (1 pound = 0.4536 kilograms)

Example: To obtain pounds equivalent to 33.2 kilograms, find 33.11 and 33.57 in table, read "70" on side scale and "3" or "4" on top scale. Equivalent is between 73 and 74 pounds, closer to 73 pounds.

WEIGHT (MASS)										
Pounds	0	1	2	3	4	5	6	7	8	9
0	0.00	0.45	0.91	1.36	1.81	2.27	2.72	3.18	3.63	4.08
10	4.54	4.99	5.44	5.90	6.35	6.82	7.26	7.71	8.16	8.62
20	9.07	9.53	9.98	10.43	10.89	11.34	11.79	12.25	12.70	13.15
30	13.61	14.06	14.51	14.97	15.42	15.88	16.33	16.78	17.24	17.69
40	18.14	18.60	19.05	19.50	19.96	20.41	20.87	21.35	21.77	22.23
50	22.68	23.13	23.59	24.04	24.49	24.95	25.40	25.85	26.31	26.76
60	27.22	27.67	28.12	28.58	29.03	29.48	29.94	30.39	30.84	31.30
70	31.75	32.21	32.66	33.11	33.57	34.02	34.47	34.93	35.38	35.83
80	36.29	36.74	37.19	37.65	38.10	38.56	39.01	39.46	39.92	40.37
90	40.82	41.28	41.73	42.18	42.64	43.09	43.54	44.0	44.45	44.91
100	45.36	45.81	46.27	46.72	47.17	47.63	48.08	48.53	48.99	49.44
110	49.90	50.35	50.80	51.26	51.71	52.16	52.62	53.07	53.52	53.98
120	54.43	54.88	55.34	55.79	56.25	56.70	57.15	57.61	58.06	58.51
130	58.97	59.42	59.87	60.33	60.78	61.23	61.69	62.14	62.60	63.05
140	63.50	63.96	64.41	64.86	65.32	65.77	66.22	66.68	67.13	67.59
150	68.04	68.49	68.95	69.40	69.85	70.31	70.76	71.21	71.67	72.12
160	72.57	73.03	73.48	73.94	74.39	74.84	75.30	75.75	76.20	76.66
170	77.11	77.56	78.02	78.47	78.93	79.38	79.83	80.29	80.74	81.19
180	81.65	82.10	82.55	83.01	83.46	83.91	84.37	84.82	85.28	85.73
190	86.18	86.64	87.09	87.54	88.00	88.90	89.90	89.36	89.81	90.26
200	90.72	91.17	91.63	92.08	92.53	92.99	93.44	93.89	94.35	94.80

CONVERSION TABLES

TEMPERATURE

Fahrenheit (°F) to Centigrade (°C): $C = (°F - 32) \times 5/9$

TEMPERATURE											
°F	°C	°F	°C	°F	°C	°F	°C	°F	°C	°F	°C
95.0	35.0	97.0	36.1	99.0	37.2	101.0	38.3	103.0	39.4	105.0	40.6
95.2	35.1	97.2	36.2	99.2	37.3	101.2	38.4	103.2	39.6	105.2	40.7
95.4	35.2	97.4	36.3	99.4	37.4	104.4	38.6	103.4	39.7	105.4	40.8
95.6	35.3	97.6	36.4	99.6	37.6	101.6	38.7	103.6	39.8	105.6	40.9
95.8	35.4	97.8	36.6	99.8	37.7	101.8	38.8	103.8	39.9	105.8	41.0
96.0	35.6	98.0	36.7	100.0	37.8	102.0	38.9	104.0	40.0	106.0	41.1
96.2	35.7	98.2	36.8	100.2	37.9	102.2	39.0	104.2	40.1	106.2	41.2
96.4	35.8	98.4	36.9	100.4	38.0	102.4	39.1	104.4	40.2	106.4	41.3
96.6	35.9	98.6	37.0	100.6	38.1	102.6	39.2	104.6	40.3	106.6	41.4

DAILY LOG

These pages are for you to record questions that arise, supplies needed, vital signs (blood pressure, temperature, etc.), medications and other information that is helpful for you to have at hand.

Date:

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ACKNOWLEDGEMENT OF RECEIPT AND REVIEW OF
**HEART/LUNG TRANSPLANT
 TEACHING BOOKLET**

I / we have read the entire Heart/Lung Transplant Teaching Booklet.
 I / we understand all of the information related to a heart/lung transplant.
 My coordinator, physician, and surgeon have discussed with me all of the information outlined in this booklet and have answered all of my questions.

PARENT / CHILD SIGNATURE	DATE
NURSE COORDINATOR SIGNATURE	DATE

 PARENT'S INITIALS



ST. LOUIS CHILDREN'S HOSPITAL

One Children's Place
St. Louis, Missouri 63110

**ST. LOUIS CHILDREN'S HOSPITAL
SPECIALTY CARE CENTER**

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Town and Country, Missouri 63017

314.454.KIDS (5437)
800.678.KIDS (5437)

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