

Lung Transplantation In Children

Lung Transplantation Mini-Series #5

A lung transplant is a surgery where one or both of your child's lungs are replaced with the lung or lungs from a suitable organ donor. A lung transplant is considered only when other treatments are no longer successful and the person's own lungs have failed. This fact sheet focuses on lung transplantation in children. For information on adult lung transplantation, see the "ATS Patient Information Series—What is Lung Transplantation?"



Why does my child need a lung transplant?

There are many diseases that may lead to end-stage lung disease, and the need for a lung transplant. The most common diseases that may require lung transplantation in children are:

- cystic fibrosis (an inherited disease),
- pulmonary hypertension (high blood pressure in the lungs)
- bronchiolitis obliterans (lung damage from severe infection or lung injury)
- interstitial lung disease [rare diseases where the lungs become fibrous (stiff and scarred)]

Lung transplantation is an option to consider if the answer to the following questions is 'yes':

- Can the child's lung disease not be improved by other treatments?
- Is there is a strong possibility the child will die in the next 2 years?
- Would the child's lifespan and quality of life expect to be improved with transplant?

What is the process for a child to be considered for and get a lung transplant?

Step 1: Referral

In most cases, your child's primary specialist (specializing in lung, heart, or newborn care) will start the process by discussing lung transplantation with you and your child. He or she will then contact a transplant referral center. There are only a few hospitals in the United States that perform lung transplants for children.

Step 2: Evaluation

The usual evaluation process to be considered for a lung transplant is detailed and involves both testing and meetings with a number of transplant team members. They include transplant pulmonologists (lung specialists), cardiothoracic surgeons, transplant coordinators, dieticians, physical therapists, social workers, psychologists, and child life specialists. Testing can include pulmonary function tests,

blood work, tests to measure the ability to exercise and how much oxygen the child needs, and imaging.

Step 3: Transplant Listing and Waiting for a Transplant

If the team and you decide to proceed with lung transplantation, then your child will be placed on a waiting list (this is referred to as "being actively listed"). This means your child's name will be given to the United Network for Organ Sharing (UNOS). UNOS keeps the waiting list for people in the U.S. who need a lung transplant. Factors used to match a person to donor lungs include blood type, height, and location of donor lungs. For children who are 12 years of age and older, a Lung Allocation Score (LAS) is calculated. The LAS is based on factors that help decide how sick the child is and helps to allot available donor lungs to those on the waiting list. The LAS is not affected by the duration of time on the waiting list. However, for children less than 12 years of age, the amount of time spent on the waiting list is the main factor for allotting available donor lungs that match.

During the waiting time, it is important to stay in touch with your transplant coordinator and have a working phone. You should always notify the transplant team of any changes in your child's health or how to contact you.

Step 4: Lung Transplant Surgery

Most children who need a lung transplant will have both lungs replaced. When new lungs are donated and found to be a good match, you and your child are notified by your transplant coordinator. Usually the donor lungs have to be brought from another place to the transplant center. Transplant surgery can take five hours or longer. Your child will usually be placed on a cardio-pulmonary bypass machine while the old lungs are removed and new lungs are put in. Right after the surgery, your child will have a breathing tube in place and be on a ventilator (breathing machine). There will be several chest tubes in place to help drain fluid and air from around the new lungs. As your child recovers, these will all be removed.

What medications will my child need to take, and what is the duration of these medications?

There are two main types of medications that will need to be taken after transplantation.

Anti-rejection medications (also known as immunosuppressants) are medicines that decrease the function of the immune system, and help protect the new lungs against rejection. These medications will need to be taken for the rest of your child's life after a lung transplant. These medications are taken daily and include (generic names given):

- Prednisone or prednisolone (a steroid)
- Tacrolimus or cyclosporine
- Mycophenolate mofetil or azathioprine

Anti-infective medications are needed to protect against infection, especially those that can affect patients who are immunosuppressed (as in those who have received a lung transplant). These can include medications to protect against bacteria, certain viruses, and fungal infections.

There are other medications as well, such as vitamins and supplements, which may be prescribed. It is important to always follow the instructions given by your pharmacist and transplant team. Some medications will always need to be taken at the same time each day. Some medicines need to be taken on an empty stomach. Blood drug levels will be monitored regularly for certain medications.

How often does my child need to be followed after transplantation?

For the first few months following transplantation, your child will need to be seen very often (one to two times every week). For this reason, most transplant centers ask families to relocate to live near the transplant center from time of active listing until at least three months after transplantation. After that time period, you may be allowed to follow up with your regular lung specialist. You will be expected to return to the transplant center for follow-up every 3-6 months. If your child is doing well, the visits may become less frequent over time. When your child becomes an adult, the team will usually transfer care to an adult transplant team. Care of the new lungs is lifelong! During follow up evaluations, your child will likely need to get a combination of lab work, pulmonary function tests and imaging. Your transplant doctor may recommend a bronchoscopy, initially at regular intervals, and later (if needed) when your child has new symptoms (such as cough, wheezing, or decreased lung function). See the ATS Patient Information Series on "Flexible Bronchoscopy".

Can my child get immunizations after transplantation?

This is an important question. Most of the usual childhood immunizations can be given and your child will benefit from them. However, your child should not receive any LIVE vaccines, as these vaccines expose a child to the mild form of the virus. Live vaccines include the MMR and varicella (chickenpox) vaccines. The lung transplant team can give you advice about specific vaccines and what your child needs. All family members are encouraged to get the influenza vaccine yearly, usually given in September.

What complications can occur after a lung transplant?

A number of problems can develop after a lung transplant. Every person with new lungs has to have close monitoring to prevent or catch problems early. Some of the complications that can occur include primary graft dysfunction (injury to the newly transplanted lungs immediately after surgery), rejection, airway stenosis (narrowing) and infection. You can learn more about complications after lung transplantation at www.thoracic.org/patients.

What is the overall survival in children after a lung transplant?

The overall 5-year survival in children receiving a lung transplant is about 50%. This means that after five years of getting a transplant, about half the patients are alive. This number depends on many factors including the reason for transplant. Without new lungs, these children would have died much sooner. If the new donor lungs fail, a repeat transplant may be offered in some cases, after weighing the risks and benefits of another major surgery. Research continues on ways to help transplanted lungs stay healthy longer. While it is a lot of work to take care of new lungs, studies have shown an improvement in the quality of life for children who had end stage lung disease and were able to get lung transplant. You and your child have to decide if transplant is worth the risk and if you are prepared to commit to the work it takes to care for new lungs. Your lung transplant team will be there to help you in your decision.

Authors: Shailendra Das DO, Ernestina Melicoff-Portillo MD, Marianna Sockrider MD, DrPH

Reviewers: Stuart Sweet, MD, Hrishikesh Kulkarni MD

Rx Action Steps

- ✓ If your child has a severe lung disease, ask your primary specialist physician if he or she is a candidate for a lung transplant.
- ✓ Make a list of questions that you have for the lung transplant team and be honest about your concerns and feelings about the decision.
- ✓ Ensure good nutrition, regular physical activity, and avoiding infections to help your child stay as healthy as possible while waiting for new lungs.
- ✓ After transplantation, take all doses of medicines prescribed by the transplant team. If you have concerns about a medicine, call the transplant team right away.
- ✓ Keep all follow-up appointments and stay in touch with the transplant team with any change in your child's symptoms.

Healthcare Provider's Contact Number:

Resources:

United Network for Organ Sharing (UNOS)
https://www.unos.org/docs/Lung_Patient.pdf

American Thoracic Society Patient Information Series
www.thoracic.org/patients.

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