



LUNG TRANSPLANTATION:

What Every Patient with Idiopathic Pulmonary Fibrosis (IPF) Should Know

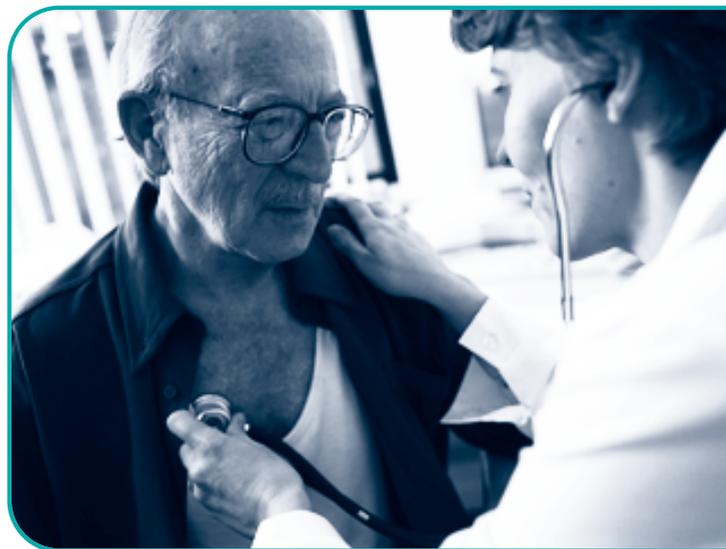


Education. Support. Hope.

WHEN YOUR LUNGS FAIL

Approximately 83,000 Americans are living with idiopathic pulmonary fibrosis (IPF), and more than 31,000 new cases develop each year. This debilitating condition involves scarring of the lungs. The lungs' air sacs are replaced by scar (fibrotic) tissue, gradually interfering with the body's ability to breathe and thus obtain enough oxygen for vital organs to function normally.

Although the course of IPF varies greatly from person to person, the disease usually develops slowly, sometimes over years. Management often includes drug therapy, oxygen therapy and pulmonary rehabilitation. For younger individuals (usually 65 years old, although program criteria may differ between transplant centers), lung transplantation may also be an appropriate therapy. Lung transplantation can both extend life and enhance the quality of life. It is an important treatment option that patients with IPF should discuss with their physician AS EARLY AS POSSIBLE. The need for considering a transplant early in a patient's disease course stems from the potentially long waiting time on the transplant list combined with the progressive nature of IPF. According to United Network for Organ Sharing (UNOS), approximately 30% of IPF patients currently listed for transplantation will succumb to their disease prior to transplantation.



WHAT IS A LUNG TRANSPLANT?

Lung transplantation is the placement of one (unilateral) or both lungs (bilateral) from a cadaveric donor (who has been declared brain-dead but remains on life support) to a recipient with end-stage lung disease. The donor lung must be matched for blood type and size to that of the recipient. The decision to transplant one or both lungs is dependent upon the extent of disease and the results of pre-transplant testing. Controversy exists regarding single versus bilateral lung transplantation for certain lung diseases, however, UNOS data has suggested superior survival outcomes with single lung transplantation for IPF. These issues should be discussed on an individual basis with the transplant center, in the context of each centers' experiences.



CADAVERIC LUNG TRANSPLANT

In a single lung transplant, an incision is made on the side of the chest. The patient's old lung is removed and the new lung is then sewn in, connecting the blood vessels to the lung and from the lung to the heart (the pulmonary artery and pulmonary vein) and the main airway (bronchus). At the end of the operation, the ribs are brought back together and the incision is closed with layers of stitches. Chest tubes are inserted to drain air, fluid and blood out of the chest for several days to allow the lungs to remain fully re-expanded.

In a double lung transplant, an incision is usually made side-to-side across the middle of the chest just under the nipple line and both chest cavities are entered between the ribs. The lung on one side is removed and the new lung sewn in place. The opposite lung is then removed and the second new lung sewn in place. At the end of the operation, the ribs are brought back together and the incision is closed in layers with stitches.

BENEFITS AND RISKS OF LUNG TRANSPLANTATION

Quality of life can be moderately to substantially enhanced by lung transplantation, and life may be extended beyond one's life expectancy prior to transplantation. It is impossible to predict how long a patient may survive after transplantation. Current survival rates are as high as 80% at 1 year following transplantation and 60% at 4 years. Further, survival statistics may vary between individual transplant centers. Individual institution's survival statistics are available for review at UNOS's web page (<http://www.unos.org>). The most critical period for survival of both the patient and the donor lung(s) is the first year after transplantation – when surgical complications, acute rejection and infection are the greatest threats to survival. Rejection and infection are the two potential major complications of lung transplantation. Adherence to treatment plan and maintaining a close alliance with the transplant team may serve to minimize these potential complications.



REJECTION

Because your transplanted lung(s) is "foreign" to your body, your body's immune system will try to destroy it, just as it tries to destroy "foreign" bacteria and viruses when they invade. This process of foreign-tissue rejection helps protect you from illness, but the process has to be "turned off" to keep your transplanted lung(s) from being destroyed.

Immunosuppressive (anti-rejection) medications prescribed by your doctors will help keep the rejection process "turned off." Other medications or an increase in the dosage of your current medications may be necessary to control and treat rejection if your immune system breaks through the immunosuppressive blockade. Following your doctors' orders and taking all medications as prescribed help to prevent or control rejection. However, sometimes rejection can begin despite your compliance with prescribed medication.

The most likely time for rejection to begin is during the first three months after transplantation surgery. Symptoms of rejection are non-specific and might include shortness of breath, changes on your breathing tests or chest X-ray, reduced oxygen saturation levels or in rare cases fever, chills and flu-like aches. The latter three symptoms are more likely to be indicative of infection. Rejection can also occur without any apparent symptoms. That is one of the reasons why regular check-ups are necessary and why some programs may elect to perform "surveillance" fiberoptic bronchoscopy with biopsies of your lung. Your transplant team will instruct you regarding whom to call to immediately report any symptoms.

INFECTION

Because you will be taking immunosuppressive medications, your immune system will be less able to fight off invading bacteria, fungi and viruses. You will be much more susceptible to infection, and infections are more likely to become severe.

You have an important role in the prevention of infection by following instructions to avoid exposure and immediately reporting any symptoms of infection to your physician. Symptoms might include fever, chills, cough, shortness of breath, headache, sore throat, redness or draining pus at the site of your surgical incision, pain during urination, vomiting, or diarrhea.

Are You a Candidate?

According to the American College of Chest Physicians, in order to be considered for lung transplantation, you must:

- Have a condition for which transplantation is considered an effective treatment.
- Have severe and progressive lung disease that no longer responds to medical treatment and may be fatal within two years.
- Be willing to accept the risks of surgery and subsequent medical treatment.
- Be physically capable of undergoing surgery and subsequent medical treatment including pulmonary rehabilitation.

Most single lung transplant candidates are 65 years or younger, and double lung transplant candidates are 60 years or younger. They also have an acceptable nutritional status, and a satisfactory psychological profile and support system.

There are a number of potential reasons that may preclude patients as appropriate transplant candidates. For example, severe, coexisting medical conditions may be worsened by the surgical procedure or by the powerful immunosuppressive drugs that are necessary after the transplant. In addition, patients who are acutely ill, or who have an unstable clinical status, those with an uncontrolled infection, those who continue to smoke cigarettes, and those with a drug or alcohol dependency may not be good candidates. The specific criteria (e.g. age limitations) and contraindications (e.g. prior cancer) to lung transplantation may vary between different centers. Therefore, motivated patients should consider obtaining a "second opinion consultation" if they are not deemed acceptable at a single center.

HOW THE TRANSPLANT SYSTEM WORKS

Under contract with a branch of the U.S. Department of Health & Human Services, Health Services & Resources Administration (HRSA), UNOS maintains a centralized computer network linking all organ procurement organizations and transplant centers. This computer network is accessible 24 hours a day, seven days a week, with organ placement specialists in the UNOS Organ Center always available to answer questions.

If a transplant center determines that a patient is a candidate for a lung transplant, it will add the patient's medical profile to the national patient waiting list for organ transplant. The patient is NOT placed on a ranked list at that time. Rather, the patient's name is added to the "pool" of patients waiting.

When a deceased organ donor is identified, a transplant coordinator from an organ procurement organization accesses the UNOS computer. Each patient in the "pool" is then matched by computer for the donor characteristics (and for lung donation, by waiting time). The computer then generates a ranked list of patients for each specific organ that can be procured from the donor in accordance with organ allocation policies. Other factors that help determine the suitability of a potential donor include blood type, height and in some cases, the immune status (e.g. tissue type).

Once a potential organ recipient is selected and contacted, he or she is instructed to quickly and safely make their way to the hospital. After routine pre-operative testing is performed, the transplant then proceeds. This entails a highly coordinated team effort with the timing of events at the recipient hospital dependent on events at the donor hospital. You can find more information about UNOS on its website at www.unos.org, and more information about organ donation on the U.S. Department of Health and Human Services website for Organ Donation at www.organdonor.gov.

THE WAITING LIST

Unfortunately, the waiting times for organ transplants are long and can vary from region to region. Waiting times may vary depending on the institution that you choose to perform your transplant surgery. Transplant centers also may differ in their "acceptable criteria" in choosing suitable donors for lung transplantation. Overall, the average waiting time for lung transplant was 275 days in the United States in 2002. As of March, 2004, there were over 3,900 people waiting for a lung transplant. Currently, the only factor that affects a patient's ranking on the transplant wait list for lungs is the length of time on the waiting list and the location of the Transplant Center in relation to the donor. The only group of patients that are afforded any priority are IPF transplant candidates who are allotted a three month "head start". Therefore a patient with IPF who is listed will automatically be credited with 90 days of wait list time.

PATIENT EVALUATION

Before you can qualify for a lung transplant, you will receive an evaluation from a lung transplant team, which typically consists of transplant pulmonologists, transplant surgeons, transplant coordinators, a social worker and a pulmonary rehabilitation specialist.

During the evaluation, you will undergo a number of testing procedures that are used to assess your physical and medical condition. These tests may include:

Ventilation-perfusion lung scan – This test is designed to determine blood and air supply to the lungs (and requires injection of "tracer" into a vein).

Pulmonary function tests – These are designed to measure lung capacity. The patient is required to breathe in and out of a tube connected to a measuring device that records various volumes of air within the lung, air flow and the ability of the lungs to take up oxygen.

Chest CT Scan – This imaging test provides detailed images of the lungs.

Echocardiography – A test designed to evaluate the function of the heart and to estimate the pressure of the pulmonary artery.

Cardiac catheterization – A study of the vessels and measurements of pressures of the heart may be required in some candidates to exclude the presence of coronary artery disease.

All information from tests, interviews and your medical history will be considered in determining whether you are a candidate for lung transplantation. Different transplant centers may have additional criteria for transplantation evaluation.

THE TRANSPLANT SURGERY

In a cadaveric transplant, once a donor lung becomes available, time is critical. The lung must be transplanted into the patient receiving the organ within four to six hours. A team of surgeons and anesthesiologists performs an operation to remove the lung from the donor. Additional surgical teams may be present to remove other organs.

After the lung is removed from the donor, it is preserved and packed for transport. Although the donor is brain dead, this procedure is treated like any other operation using standard surgical practices and sterile techniques. Once the operation is complete and the incisions are closed, the donor's body is prepared for funeral or cremation. Organ procurement surgery respects the body and an open casket funeral is possible if desired.

Typically, both lungs are removed from the donor together. If the recipient is in need of a double lung transplant, both lungs will be transplanted. Otherwise, the lungs are usually separated after they are removed from the donor and used for two single lung transplant recipients.

In the meantime, the recipient is contacted and then prepared for surgery. Preparation involves administration of general anesthesia, and placement on an artificial breathing machine (e.g. ventilator). Lung transplant surgery may require the use of a "heart-lung machine" to oxygenate blood and maintain adequate blood flow to the patient's vital organs. The transplant of the lung begins with removal of the diseased lung and the blood vessel attachments to the heart and large airway called the bronchus. When the lung is placed within the recipient, the blood vessels and bronchus from the donor lung are connected to the recipient's corresponding blood vessels and bronchus. Next, the blood flow and airflow are restored. After the transplant is complete, the incision is closed. The patient then recovers in the hospital's intensive care unit.

Interestingly, when a double lung transplant is performed, it is much like two single lung transplants. The lung that is more diseased is transplanted first and then the less diseased lung is transplanted.





AFTER TRANSPLANT SURGERY

Following lung transplant surgery, the patient may remain on an artificial breathing machine for the first 12 hours of recovery. However, if the donor lung is functioning properly, the ventilator may be removed at the end of surgery.

Depending on their progress, some patients are moved out of the ICU within a few days. Generally, they will also begin eating within the week following surgery. The total hospital stay is usually 10 to 14 days.

Because the recipient's immune system will identify the new organ as foreign, rejection of the transplanted lung is always a possibility. Powerful drugs called immunosuppressants are given starting at the time of lung transplant surgery to try to prevent rejection. Blood tests are done on an ongoing basis post-transplant to confirm that the patient is receiving the correct dosage of medication.

Prior to discharge, the transplant team reviews information with the patient, gives instructions for follow-up care and medications, informs patients of potential side effects and interactions of medications, and answers the patient's questions. A prescribed rehabilitation program will continue at home including physical activity, breathing exercises, nutrition, and the continuation of immunosuppressants and other medications. Signs and symptoms of rejection and infection are also discussed with the patient and family. Frequently, the patients are instructed to buy or are given small breathing machines called microspirometers, to monitor their own breathing capacity at home.

RETURNING TO THE HOME ENVIRONMENT

At-home rehabilitation for lung transplantation is a gradual process, and depends on the individual. The transplant team will give specific instructions. In general, walking is recommended to restore strength and prevent lung complications, but heavy lifting should be avoided for four to six weeks following transplant surgery. Other activities, such as driving, may usually begin when the incisions have healed. Sexual activity can resume when one is comfortable.

Follow-up visits are required for check-ups and begin soon after returning home. Initially, outpatient visits may occur weekly or even more often, and as time progresses the frequency of follow-up visits usually decreases.

As mentioned above, current survival rates are as high as 80% at one year following transplantation and 60% at four years. The most critical period for survival is the first year after transplantation, when patients are at highest risk of surgical complications, rejection and infection.

IMPACT OF LUNG TRANSPLANT ON FUNCTIONAL STATUS

The functional status of the majority of post-transplant patients is generally improved. More than 80% of adult patients who receive a lung transplant experience no limitations with regard to their activities one to five years post-transplant, while an additional 10-15% require only some assistance with their daily activities. Only a very small proportion of patients (less than 5%) require complete assistance. Patients may return to full or partial employment, and resume participation in sporting activities.

TALK TO YOUR DOCTOR ABOUT LUNG TRANSPLANTATION — TODAY

It is important to understand that IPF is a progressive disease for which there is no cure – with median survival rates between three and five years. You and your physician should discuss the possibility of lung transplantation as a treatment option AS SOON AS POSSIBLE during the course of your care. This is an important statistic that you should know:

According to UNOS, more than 30% of IPF patients who qualify for lung transplantation and who are placed on a transplant waiting list, will pass away before a lung becomes available; approximately 59% of IPF patients are transplanted within two years.

The American College of Chest Physicians provides the following questions that you should ask your physician and transplant center:

- Is transplantation my best option for treatment of my condition?
- What are organ and patient survival rates at this institution for the type of transplant I will need?
- How many transplants of this type are done every year at this institution? How many by my physicians and surgeons? How long have they been doing this type of transplant surgery?
- What costs of transplantation and rehabilitation are covered by my insurance? What out-of-pocket costs will I have to pay?
- Am I likely to get a donor lung more quickly if I get wait-listed at more than one medical center, in different parts of the country?

Lung transplantation may be a viable option for patients with IPF. But don't wait too long to become educated about the details of this procedure. Become proactive and find out more from your physician. This is a commitment you need to make to yourself – today.

RESOURCES FOR TRANSPLANT CANDIDATES AND RECIPIENTS

- United Network for Organ Sharing (UNOS)
www.unos.org
- Second Wind Lung Transplant Association, Inc.
www.2ndwind.org
- Coalition for Pulmonary Fibrosis
www.coalitionforpf.org
- American Lung Association
www2.lungusa.org
- Transplant Speakers International, Inc.
www.transplant-speakers.org
- Children's Organ Transplant Association, Inc.
www.cota.org
- National Foundation for Transplants
www.transplants.org
- National Transplant Assistance Fund
www.transplantfund.org
- Medicare Rights Center
www.medicarerights.org

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- The American College of Chest Physicians
- The American Society of Transplantation
- Duke University Medical Center & Duke Transplant Center
- Inova Fairfax Lung Transplant Program
- The Lung Transplantation Handbook
- The Merck Manual of Diagnosis and Therapy
- University of Pennsylvania Medical Center
- The Scientific Registry of Transplant Recipients
- Tampa General Hospital Transplant Center
- United Network for Organ Sharing (UNOS)

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