

Lung transplantation is a difficult and personal decision. The Cystic Fibrosis Foundation has prepared this fact sheet to provide general information about lung transplantation and to help guide discussions between people with cystic fibrosis (CF) and their CF care teams. Recently, the United Network for Organ Sharing (UNOS) approved changes to the lung allocation process. These changes, which become effective in the spring of 2005, are outlined in more detail below.

When is it time for a lung transplant? What is involved in the evaluation process?

When someone with CF develops severe lung disease, the CF care team may discuss the option of lung transplantation with the person. The doctor can refer the person to a lung transplant center for evaluation. The transplant center evaluates the person's health to determine if a lung transplant is necessary and timely. Tests examine how well the lungs, heart, and kidneys function, the types of bacteria in the lungs, and, because of the serious health care implications of transplantation, the person's psychological well being. The transplant center also will evaluate the person's social support system including family, friends, and professional support. Most components of the evaluation are standard, but each center can have some specific requirements. The staff's decision to accept a person for a transplant is specific to that center.

How many people with CF have had lung transplants?

Approximately 900 lung transplants are performed each year in the United States. According to the CF Patient Registry, nearly 1,600 people with CF have received lung transplants since 1991; this is about 120 to 150 people per year. Unfortunately, more people apply to get transplants than can receive them, due to a shortage of available organs. In 2003, 524 people with CF were evaluated for lung transplants, and 368 were accepted for the procedure. Living donor transplants—when lobes, or sections, of a lung are transplanted from two living individuals to the person needing new lungs—are much less common than standard cadaveric transplants, which are transplants using lungs from people who have died. For example, in 2003, only 17 people with CF received transplants from living donors, while 126 people received cadaveric lung transplants.

What are the current success rates and recovery times?

The success of lung transplantation is determined by the recipients' average length of survival after the operation. For people with CF, the success of lung transplantation is as good as or better than for those with other lung diseases. As many as 90 percent of people with CF are alive one year after transplantation, and 50 percent are alive after five years. Following surgery, a person may be discharged from the hospital in a few days or a few months, depending on the person's health and complications. The average time in the hospital after transplant is two weeks.

What is the difference between a "cadaveric" lung transplant and a "living-donor" lung transplant?

A cadaveric lung transplant is when the lungs being transplanted are from someone who is "brain dead." A "living donor" transplant is the surgical removal of a right lower lobe from one person and a left lower lobe from another person, and the placement of these lobes into the person receiving the transplant. The person who is determined to be "brain dead" or giving a lobe of a lung is called a "donor". The person who is getting the new lungs is called the "recipient." Donors must be a "match" with the recipient. A match is when enough medical and biological information about the donor and recipient are the same, e.g., blood type.

Living donor transplantation is more complicated than cadaveric transplantation, because three people must have major surgery. These donors must be in good health and will have a slight decrease in their lung function following surgery. This should not limit their usual activities, however. People with CF can receive either a cadaveric or living donor lung transplant, and it is best to know what each procedure entails before deciding.

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Are cadaveric transplants more successful than living donor transplants?

In transplant centers that perform both types of lung transplants in people with CF, the success rate for cadaveric transplants is approximately the same as the success rate for living donor transplants. It is important that patients know the center's experience in each procedure and their success in CF when choosing a center.

What happens to the body after the transplant?

The immune system protects the body from foreign material, which is anything not belonging in the body, such as germs or transplanted organs. Therefore, the immune system naturally reacts against—or rejects—the foreign organs. Drugs to stop the immune system from rejecting the organ, called immunosuppressive drugs, must be taken daily for life. These drugs may cause side effects such as diabetes, kidney problems, cancer-like tumors and osteoporosis (i.e., thinning of the bones). Research on immunosuppressive drugs shows promise in helping people live longer with lung transplants.

Once the person has received a lung transplant, does CF "go away?"

Transplanted lungs come from people who do not have CF, so the new lungs do not have CF. However, after the transplant, the recipient still has CF in the sinuses, pancreas, intestines, sweat glands, and reproductive tract. The new lungs do not “get” CF, but immunosuppressive drugs may decrease the ability to fight germs like *Pseudomonas aeruginosa* and *Burkholderia cepacia* (*B. cepacia*). These germs may stay in the upper airways after a transplant and can infect the new lungs. The risks of infection are highest right after the transplant operation when immunosuppressive drugs are given at the highest doses so the body will not reject the new lungs. However, these drugs also make it hard for the body to fight infections, which can lead to lung disease.

Can a person with CF who has *B. cepacia* receive a lung transplant?

Survival for people with *B. cepacia* after a transplant is not as good as for those without these bacteria. Many lung transplant centers will not accept people with CF who have *B. cepacia*. The lung transplant coordinator at the center will know if it will transplant someone with *B. cepacia*.

How does someone choose a lung transplant center?

People choose transplant centers for different reasons, including location and experience in transplanting people with CF. Another consideration has been the length of a center's waiting list; however, the rules regarding distribution of donated lungs are changing (see below), so the waiting list will no longer apply for teens and adults. To learn more about which centers perform lung transplants, talk to your CF care center team.

Once a person is listed, how is a person chosen to get new lungs?

In the past, to receive donated lungs, the person must be near the top of a lung transplant waiting list and must be a “match” for the donated organs. Waiting time varied from a few weeks to a few years, but the average wait was nearly two years.

In spring 2005, a new lung distribution policy will eliminate the waiting list for people over age 12.

According to the new policy, people awaiting transplantation will be assessed periodically to determine how sick they are and how likely they are to stay healthy after a transplant. This allows donor lungs to be given to the person who needs them most at that time.

The new system uses a mathematical formula, called a “lung allocation score,” to determine when someone should receive new lungs. The score uses medical information about the person's health before the transplant, and projected health after the transplant. The medical information gathered to determine this score includes disease diagnosis (CF or other lung disease), lung function tests, health factors such as diabetes, the use of oxygen or a ventilator to help the person breathe, and other factors. Individuals will be re-evaluated every six months and given a score between 0 and 100; those with a higher score have a better chance of obtaining lungs sooner.

How will the new system to distribute lungs affect people with CF?

The new system was put into place by the United Network for Organ Sharing (UNOS), on behalf of the federal government. The CF Foundation and its experts reviewed and commented on the proposals to ensure the needs of people with CF were considered. Although the system does not favor people with certain lung diseases, CF experts believe the system will make more lungs available to people with CF. People with CF generally do well after lung transplantation – often better than people with other lung diseases. Therefore, they should score well under the new system and receive new lungs more quickly than people with other lung diseases. People who are currently on the waiting list for lung transplants should speak with their transplant team and their CF care center team to determine how this new system will impact them.

Does the person have to live close to the lung transplant center?

All transplant centers require the person to get to the center within a certain amount of time, usually within a few hours, so that the person is there in time for the surgery to be performed. Sometimes, a person must become a legal resident in the state where the transplant center is located, which is often based on the type of insurance coverage the person has, such as state-based Medicaid.

Are lung transplants covered by insurance?

Most health insurance and government programs, including most Medicaid programs, will pay for a lung transplant. However, insurance companies may require the person to obtain the lung transplant at a certain center. Check with your CF care center team to determine which centers would be good for you and then talk to your insurance company regarding coverage. The coordinator at the lung transplant center may have information on private and public insurance options, and on ways to raise funds for the transplant and other expenses. It also is important to learn about the insurance coverage for medications needed after transplantation, such as immunosuppressive drugs. For example, at this time, Medicare covers these medications, but only if the transplant surgery was paid for by Medicare. Under the Medicare Prescription Drug Act of 2003, it will cover these medications; check with your Medicare office to determine how this new law can help you through drug discount cards in 2004 and full benefits in 2006.

What is the future of lung transplantation for people with CF?

Lung transplantation was first performed in the United States in the late 1980s and has improved over time. At first, heart-lung transplants were the recommended surgery, but double lung transplants became possible as surgical techniques improved. Most recently, there have been improvements in the techniques to enable living donor lung transplants. In addition, studies show some advances in the use of immunosuppressive drugs for lung transplants, which hold great hope in improving survival even further. Based on progress made in the past 10 to 15 years, lung transplantation surgery, health care, and survival, will continue to improve.

What is the CF Foundation's role in lung transplantation?

The CF Foundation is supportive of organ donation and the importance of lung transplantation for people with CF. The CF Foundation continues to push for further advances in the transplant field, for research on CF and transplantation, and for policies that help people with CF obtain lung transplants. It is hoped that the CF Foundation's efforts to develop new drugs for CF will reduce the need for lung transplants. For more information, please visit the CF Foundation's Web site at www.cff.org or call (800) FIGHT CF.

Resources

(for informational purposes only)

Organ Donation and Transplantation

- United States Department of Health and Human Services (government initiative to increase awareness of organ donation, and Secretary’s Advisory Committee on Organ Transplantation):
www.organdonor.gov
- Coalition on Sharing Life: www.shareyourlife.org, (804) 782-4920
- United Network for Organ Sharing (data by center): www.unos.org, www.transplantliving.org, (888) 894-6361
- Organ Procurement and Transplantation Network: www.optn.org/members/search.asp, (To search for lung transplant centers, specify “Transplant Centers by Organ” and then use “lung” for the organ, and “all states” for the region.)

Financial Assistance

- National Transplant Assistance Fund: www.transplantfund.org, (800) 642-8399
- Children’s Organ Transplant Association: www.cota.org, (800) 366-2682
- Medicare: www.medicare.gov, (800) MEDICARE