



Cystic Fibrosis Canada

Lung Transplantation and Cystic Fibrosis

An important treatment option

When medical management alone can no longer maintain lung health and physical function, a lung transplant can improve the length and quality of life for a person with cystic fibrosis.

Most transplant recipients report improved strength and energy, and freedom from symptoms such as coughing and shortness of breath. Transplantation is an important treatment option for damaged CF lungs, but unfortunately it is not a cure for CF.

The lungs that are transplanted into the recipient's body do not have cystic fibrosis because they have the DNA of the person who donated them, and not the DNA that the transplant recipient was born with. However, cystic fibrosis still occurs in other organs in the body, for example, in the pancreas. Following the transplant, the CF clinic team still helps to manage the recipient's other CF-related health issues.



Breathing life into the future®



“I’m no longer scared of going to sleep at night for fear I’ll stop breathing and I won’t wake up. No longer feeling like I’m drowning in mucus, breaking lungs from coughing up blood, or feeling like I run a marathon gasping for air just getting a glass of water. Life is no longer a struggle just to stay alive.

I have been breathing easy since July 5th 2010 and what a beautiful journey it has been.”

Hattie Dunstan, Toronto, Ontario
(double-lung transplant, 2010)

Lung transplantation

Transplantation is a surgical procedure that replaces severely damaged organs with healthier organs. Despite the progress made in the treatment of cystic fibrosis, it remains a life-shortening disease. For many who have cystic fibrosis, years of chronic lung infections have damaged and destroyed the lung tissue which, in turn, has led to decreased lung

function and the need for new lungs.

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Increasingly, lung transplantation is providing individuals with cystic fibrosis

whose disease has progressed to a critical stage with hope of living a longer and healthier life. It is a very exciting time for transplantation, as new advances are ongoing. Post-transplant outcomes have improved significantly in recent years.

Assessing candidates for lung transplantation

CF physicians follow general guidelines to determine whether an individual should consider transplantation. Advice to one person may differ from advice given to another.

Although various factors may affect a physician's decision to refer an individual with cystic fibrosis for transplantation, physicians generally consider the following two primary tests:

- ◆ Forced Expiratory Volume (FEV₁): This test indicates the amount of air that can be forcefully blown out of your lungs during the first second of expiration. Individuals may be considered for transplantation if their FEV₁ falls below 30 per cent OR if there is a sudden and rapid decline in FEV₁.
- ◆ Oxygen Saturation Level: This test indicates the extent to which red blood cells are saturated with oxygen. Individuals may be considered for transplantation if their levels fall below 88 percent or if they need to use additional oxygen on a regular basis.

Deciding to have a transplant is a significant decision that requires serious consideration because:

- ◆ Transplantation is a major surgical procedure which, in itself, includes a certain level of risk
- ◆ Transplantation requires a life-long commitment to the maintenance of a rigid post-transplant regimen
- ◆ Transplantation can mean trading one set of medical issues and complications for another

Making the decision

To help make the decision, individuals considering a transplant are advised to consult with family members, the CF clinic team, the transplant team, friends, and loved ones. The CF clinic and transplant

“The CF Clinic and Transplant teams can put individuals considering a transplant in contact with a support network.”

teams can put individuals considering a transplant in contact with a support network comprised of other people and/or families who have undergone lung transplantation.

While a transplant candidate is the only person who can make the final decision about having a transplant, those undergoing a transplant are advised to select a designated support person such as a parent, spouse, or friend to accompany them to appointments and to provide support throughout the entire process.

Referral and evaluation



Initially, the respirologist at their CF clinic will refer transplant candidates to a lung transplant program. A referral is not the same as being placed on a transplant waiting list. First, a formal and intensive

assessment which includes a variety of discussions, tests and an evaluation of the candidate's general health and well-being takes place. During this time, the transplant team determines the patient's current health status. In addition, the potential candidate and his or her support person(s) learn about all the procedural details to assist in making an informed decision about opting to be placed on the waiting list.

Pre-transplantation evaluation tests

A pre-transplantation assessment includes a number of tests that evaluate lung, heart, kidney and liver function, as well as an assessment of nutritional status. People with cystic fibrosis are familiar with many of the procedures such as blood tests, chest x-rays and pulmonary function tests. In addition to assessing physical health, a psychological assessment is made to determine the candidate's and his or her family's ability to cope with the stresses of a transplant.

Other possible pre-transplant tests

Tests that may also be administered pre-transplant include:

- ◆ Pulmonary Function Test (PFT): Measures how well the lungs work
- ◆ Sinus and chest X-ray: Determines the extent of lung damage, and sinus disease
- ◆ Arterial blood gas: Shows how well the lungs are able to bring oxygen in and remove carbon dioxide from the bloodstream
- ◆ Ventilation-Perfusion (VQ) scan: Shows the amount of blood flow to each lung
- ◆ CT scan of chest: Looks at the heart and lungs in more detail
- ◆ Abdominal ultrasound: Monitors the abdominal organs such as the liver and kidneys
- ◆ Bloodwork: Evaluates other systems including the kidney, liver, and other tests, as well as antibodies to prior infections and immunizations.
- ◆ MUGA scan: Evaluates the function of the right and left side of the heart.
- ◆ 2D Echo: Evaluates the effectiveness of the heart valves and muscle and their pumping capabilities
- ◆ Electrocardiogram (ECG): Shows the patterns of electrical waves in the heart
- ◆ Thallium Persantine Scan: Assesses blood circulation in the heart muscle.
- ◆ Bone Mineral Density Scan: Evaluates the strength of the bones
- ◆ Sputum analysis: Evaluates presence of infections in the lungs
- ◆ Six-minute walk test: Walking as fast as possible for six minutes, to test level of fitness
- ◆ Oximetry test: Determines the amount of oxygen required at rest and during exercise.

Generally, approximately three to four weeks after pre-testing is completed, the transplant team (respirologist, thoracic surgeon, transplant coordinator, social worker, dietician, psychiatrist, physiotherapist and nurses) meets to discuss a candidate's case and to decide if the candidate meets the criteria for transplantation.

If the candidate meets the criteria for transplantation, the transplant team will discuss the transplant process and implications with them. The candidate must then decide if he/she would like to be placed on a transplant waiting list.

Waiting for the surgery

If a candidate meets the criteria for transplantation and decides to have a transplant, he or she is placed on a waiting list.

Some candidates consider the waiting period to be the most difficult part of the transplantation process. Understandably, prior to being called for a transplant, candidates may experience many perfectly normal emotions such as fear, anxiety and uncertainty. Throughout the transplantation process, help is available to assist candidates in coping with stress, managing pain, financial burdens, temporary housing, and other concerns.

There is no way to predict when appropriate organs will be available, and waiting times can range from a few weeks, to many months.

“The waiting period can be the most difficult part of the transplantation process.”

Organs available for transplantation cannot survive outside the human body for an extended period of time. For

that reason, those on waiting lists are required to live within an acceptable distance of the transplant centre, to allow the candidate to travel to the centre and be prepped for surgery in a minimal time, to increase the chances of a successful transplant. The specific guidelines for acceptable distance vary between transplant programs. It is impossible to know when a candidate will receive the call for transplant surgery, so candidates must always be ready, and a support person must always know how to reach the candidate.

Assignment of organs to candidates

A variety of factors are used to determine how organs are assigned. Factors include, but are not limited to: blood type, size of the available organ, the length of time on the waiting list, and, most importantly, the current degree of urgency. If a candidate on the waiting list is extremely ill, it is possible to move the person up on the waiting list. As well, candidates with particularly rare blood types may have to wait longer to find a match. Race and sex of the donor or the transplant candidate have no bearing on the match.

Preparing for a transplant

To ensure that all candidates are as healthy and strong as possible prior to transplantation, individuals participate in a physical rehabilitation program and a healthy diet. Adequate body weight and good physical health are important as both can help a candidate during surgery and help to reduce the risk of post-transplant complications. Many transplant centres offer pre-transplant education programs and support groups. A dietitian at the transplant centre or in the CF program assists candidates in achieving and maintaining a healthy weight prior to the surgery.

Exercise is also very important. Inactivity can cause fatigue and loss of muscle mass and strength. Pre-transplant exercise can significantly assist a person in regaining strength after the transplant. Candidates are advised to exercise as much as possible to maintain or improve current abilities. The transplant team designs individual fitness programs suitable to each candidate's needs and abilities. Studies have shown that a candidate's physical condition prior to transplant surgery can assist recovery.

The call

When suitable lungs become available, a candidate waiting for surgery will receive a call to come to the hospital. The call may be made at any time of the day or night, and candidates are advised to establish a readiness plan that includes how to get to the hospital, who to call, and how to inform loved ones.

A 'dry run'

Timing and sequence of events can be critical factors for transplantation. Once organs have been removed from a donor, transplantation must occur as quickly as possible. However, the condition of the donor lungs is

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not known until the lungs are assessed by the surgical team. Given that candidates often need to be prepared

for surgery before the donor lungs have been examined, it is possible for a candidate to be notified that lungs are available, and, due to unforeseen circumstances, after the candidate arrives at the hospital, the lungs are not suitable for transplant and the transplant is cancelled. This turn of events is known as a false alarm or 'dry run'.

Availability of organs

The primary obstacle in obtaining a transplant is the availability of suitable donor organs. Although donation rates in Canada are slowly increasing, the number of patients listed for transplantation grows at a faster rate.

The surgery



Lung transplantation surgery may take anywhere from five to 10 hours. Double-lung transplant surgery is usually performed through a “transverse sternotomy incision” (across the chest), and the two lungs are replaced one after the other. The recipient is placed on a ventilator which allows one lung to breathe while the other lung is replaced. If necessary, the recipient is put on a bypass machine which functions for the heart and lungs, and keeps the patient’s blood oxygenated and pumping through the body. The recipient’s first lung is removed. Then, the first donor lung is attached, and blood flow is restored to the new lung. The procedure is repeated for the second lung. The incision is then closed and the patient is placed on a ventilator to assist in breathing, and moved to the Intensive Care Unit (ICU).

Both lungs are transplanted to reduce the risk of infection spreading from the CF lung to the new lung. For this reason, it is most common for individuals with cystic fibrosis to receive double lung transplants.

Post-transplant

In the ICU, a patient is placed on a ventilator until the new lungs are able to function well on their own. Once the ventilator is removed, patients experience their first breaths with new lungs. For some, who have lived with cystic fibrosis, it may be the first big breath they have ever taken. Generally, after a double-lung transplant, individuals achieve close to normal lung function. It takes weeks to months for the lungs to fully recover to near-normal function, but most people notice a big improvement in their breathing right away.

A human’s immune system is designed to protect us and thus destroy anything foreign. Because transplant recipients now have new and “foreign” organs, they must take immunosuppressive drugs and antibiotics. The immunosuppressive drugs reduce the immune system’s ability to reject the new organs; the drugs must be taken every day for the rest of the recipient’s life. Individuals who have had a transplant, who must take these drugs, will have a reduced ability to fight-off infections caused by bacteria and viruses for the rest of their lives. It is very important to be up to date on recommended immunizations before transplantation, since some immunizations are not allowed or are less effective after transplantation. It is also important for close family members to get annual flu shots to reduce the risk that they will pass the flu on to the recipient.

Post-transplant (continued)

There are two types of rejection that can happen after a person receives a lung transplant.

- ◆ Acute rejection typically occurs in the first 12 months after transplantation. The immunosuppressive anti-rejection drugs that a recipient takes will help reduce the chances of this happening. Even when a person takes anti-rejection drugs faithfully, recipients can still experience rejection episodes that are usually temporary and reversible. The transplant team will educate recipients on the symptoms of rejection, and they will monitor transplant recipients for rejection.
- ◆ Chronic rejection usually occurs over a long period of time. This can also be called bronchiolitis obliterans syndrome (BOS) or chronic lung allograft dysfunction (CLAD). Chronic rejection is a drop in lung function due to irreversible scarring of the lung tissues or smaller airways. Treatment for chronic rejection includes an alteration or increase in anti-rejection drugs, and sometimes re-transplantation.

In the weeks following transplantation, a recipient can expect the following:

- ◆ Monitoring for rejection of organs.
- ◆ Adjusting to the immunosuppressive drugs: As each individual is different, different combinations of drugs are required.
- ◆ Education: Although transplant recipients are accustomed to drug and other treatment routines, it may take time to learn new post-transplant routines.
- ◆ Rehabilitation: Often, prior to lung transplantation, a CF patient is quite ill and out of shape. When lungs are replaced, the body must catch up with what the healthy lungs are able to do. Rehabilitation helps the patient improve physical fitness to achieve the most benefit from his or her new lungs.

The transplant process is different for each individual, and everyone who goes through the process will have a different story and outcome. Overall, 90 percent of individuals who have had a transplant report satisfaction with their decision. Immediately after the procedure, individuals can feel the difference in breathing and exercise abilities. Another dramatic change is that transplant recipients typically do not have a chronic cough or produce sputum; Most people do not feel that their breathing is limited in any way while doing normal activities. Lung transplant recipients can attend school, work and travel after they have recovered from the surgery.

After release from hospital, the transplant centre follow-up is fairly rigorous. Initially, individuals attend weekly appointments, which eventually taper off to much less frequent appointments every few months to annually, depending on the transplant clinic. Blood work and breathing tests must also be measured regularly.

Living donor lung transplantation

Due to the ever-increasing rates of lung transplantations, coupled with the shortage of available organs, living donor lung transplants may be considered. During this procedure, lobes (parts of a whole lung) are removed from two separate donors, and transplanted into a recipient. Significant assessment and coordination is required to ensure that both the recipient and the two potential donors are well prepared for the surgery.

Donors for this procedure must be in excellent health and must either be a family member or longtime friend of the recipient. The donors will have 20 per cent of their total lung volume removed, which typically does not affect their lifestyle. In Canada, living donor transplantations are performed at the following three centres: University of Alberta Hospital (Edmonton, AB), Health Sciences Centre (Winnipeg, MB), and Toronto General Hospital.

Connecting Canadians with cystic fibrosis

Join the My CF Canada Network, Canada's first-ever social network designed by CF patients specifically for CF patients to connect and share important information about living with this severe and potentially fatal genetic disease. Canadians with cystic fibrosis can discuss the treatments, programs and services available in their province/region, via video chat and instant message, share stories and personal issues, and receive support for advocacy and fundraising initiatives from a much broader community.

Cystic Fibrosis Canada acknowledges Novartis Pharmaceuticals Canada Inc. for the collaboration that has made the My CF Canada Network possible.

Sign-up today at www.mycfnetwork.com

For more information about cystic fibrosis, visit cysticfibrosis.ca

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