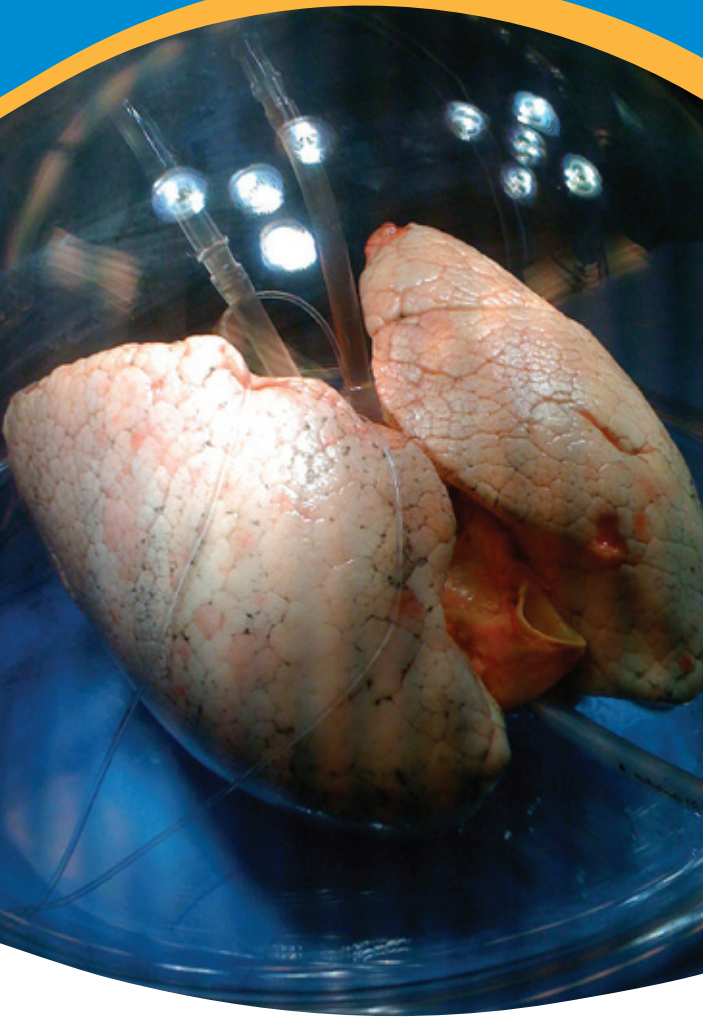
# Cystic Fibrosis and Lung Transplantation



Cystic Fibrosis Canada

*Breathing life into the future®*

Margaret Benson, who had a double-lung transplant in 1999, celebrates a gold-medal victory at the 2009 Australian World Transplant Games.

"It is an honour to represent Canada at the games and it is fun to win medals, but my biggest joy is to give them to donor families to honour their loved ones.  If it weren't for them I wouldn't be here living the life I am living."

## Introduction

Transplantation is a major surgical procedure that replaces severely damaged organs with healthy organs when medical management alone can no longer maintain a person's health or organ function. The transplanted lungs will not develop cystic fibrosis, but CF will still be present in other body cells, such as the cells of the pancreas. Although transplantation offers new hope, it also presents new challenges and responsibilities.

Individuals with cystic fibrosis in need of a transplant usually require new lungs, but may also need liver, kidney, or heart transplants. This brochure focuses on lung transplantation. Please note that programs offered at transplant centres may differ slightly, and each centre will provide details specific to its program.

## Some benefits of lung transplantation

Post-transplant, most individuals with cystic fibrosis report improved strength, energy, and exercise capacity, and freedom from symptoms like constant coughing, and shortness of breath. Ongoing advances in transplantation procedures and care are significantly improving post-transplant outcomes.

## Considering candidates for lung transplantation

Cystic fibrosis physicians follow general guidelines to determine who should be considered for transplantation. The main indicator for transplantation is the deterioration of lung function. Other indicators include an inability to maintain weight and the frequency of hospital admissions. To qualify for transplantation, individuals with CF must be ill enough to need a transplant, but well enough to endure the surgery.

**Margaret Benson**

*Double-lung transplant recipient:* 1999

*Post-transplant:* spent 18 days in the intensive care unit

*2003–Present:* Margaret has participated in four World Transplant Games and three National Transplant Games. She holds two world records, and is the national record holder, in her age category for the race walk and 200m run.

"Deciding to get a lung transplant was difficult. I knew that someone would have to die for me to live, and I struggled with that injustice. However, transplantation was a chance I had to take.

If you had told me, when I had end-stage lung disease or when I had a full-body seizure caused by a stroke post-transplant, that I would be winning medals at the World Transplant Games in running and rowing events, I would've thought you were crazy! There were many bumps in my road to recovery. My struggle was not nearly the fight that some have had, but it was up there with the worst of them.

I often forget what it was like to have the lungs of someone with cystic fibrosis. I remain forever grateful to my donor family who, in a time of tragedy, was able to think of others in need."

Some of the test results considered by the CF clinic team when referring an individual to a transplant centre are:

- *Forced Expiratory Volume ($FEV_1$):* the maximal amount of air that can be forcefully exhaled in one second. Transplantation may be considered if $FEV_1$ falls below 30 per cent OR if there is a sudden, rapid decline in $FEV_1$.
- *Hypoxemia < 55 mmHg:* a measure of lung damage due to the lungs' inability to provide enough oxygen for the body to function.
- *Hypercapnia > 45 mmHg:* a measure of the lungs' inability to get rid of carbon dioxide.

Life-threatening events, such as repeated episodes of hemoptysis (coughing up blood), frequent infections and/or pneumothoraces (a collection of air between the outside surface of the lungs and the inside surface of the chest wall) may call for earlier referral.

## Making the decision

To help make a decision about transplantation, individuals with cystic fibrosis considering a transplant may want to consult with family members, their CF clinic and transplant teams, or use the online *Lung Transplant Decision Aid for People with Cystic Fibrosis*.[1] Candidates can be referred to a "support network," comprised of other individuals and/or families who have undergone lung transplantation. However, the final decision on whether to proceed with a transplant must ultimately be made by each individual.

[1] decisionaid.ohri.ca

## Referral process

To qualify for a lung transplant and to be added to a transplant waiting list, individuals with cystic fibrosis need to go through a referral and assessment process.

Generally, the CF clinic respirologist refers candidates to the lung transplant program by submitting clinical notes and most recent test results. A transplant centre respirologist reviews the information and books the candidate for an appointment. If an individual is deemed a good candidate, a decision is made by the transplant team on whether to proceed with the complete pre-lung transplant assessment.

The transplant team usually includes the transplant physician, transplant coordinator, social worker, psychiatrist, physiotherapist and dietitian.

## Pre-lung transplant assessment

The pre-lung transplant assessment evaluates lung, heart, kidney and liver functions, as well as nutritional status. On average the assessment process lasts one week.  In addition to assessing physical health, psychological assessments are made to determine a candidate's and his/her family's ability to cope with the stresses of a transplant.

## Assessment meeting

Approximately two to four weeks after a pre-transplant assessment is completed, the transplant team meets to discuss the candidate's case and decide if the time is right to place this candidate on the transplant waiting list.

**Kelly Sheppard**

*Double-lung transplant recipient:* 2008

*Post-transplant:* after two false alarms and over 18 hours of surgery Kelly vividly remembers the excitement of taking a huge, deep, clear breath!

*2009-Present:* advocates tirelessly for the cystic fibrosis community, is a member of Cystic Fibrosis Canada's Board and Adult CF Committee, and takes every opportunity to promote organ donation.

"My decision to have a lung transplant was easy because surgery really was the only chance I had at living. One morning I awoke struggling to breathe and turned to my husband and told him it was time, 'I have to do it now'.

Leaving my family, friends and pets behind in Newfoundland was the hardest part of my journey. I was placed on the waiting list in Toronto on December 19th, 2007, received my new lungs on February 22nd, 2008, but didn't return home for over a year and a half!

The waiting and "false alarms" were also quite difficult, not to mention the financial toll of having to relocate. After three years we are still struggling to get our finances back on track.

I am extremely happy with my decision. I have to be careful with infection control, follow a strict treatment regime, and endure follow-up tests and assessments, but I have a LIFE again!"

## Valérie Mouton

*Double-lung transplant recipient:* 2004

*Post-transplant:* was left completely deaf, and is currently receiving hemodialysis while awaiting a kidney transplant.

*2011:* completed university and works for a non-profit organization.

### Denis and Christiane Mouton, parents of Valérie

"When Valérie told us she was on the lung transplant list, it was a moment of great joy for our family. However, she developed pulmonary hypertension confining her to the hospital, and had part of her intestine removed due to a blockage. We remained by her side day and night.

On July 16th, Valérie's doctors delivered great news – her long-awaited lungs were finally available! We knew that even if she didn't make it, she would die happy because what she wanted most in the world was to receive her transplant.

The surgery was a success and her ability to breathe without assistance made the recovery easier to bear. Only days following surgery Valérie became completely deaf. Being a musician since the age of four made this difficult to endure. Thankfully, she received a cochlear implant and her hearing has been restored!"

## Listing and waiting for the surgery

If a candidate decides to be put on a waiting list, other requirements may have to be completed, such as designating a support person, signing the consent form, or securing medication and/or oxygen funding. Candidates who live far away from their transplant centre may need to relocate so their health can be monitored while they wait, and to ensure they can participate in an exercise program.

Many candidates consider the waiting period the most stressful part of the transplantation process. Prior to being called for a transplant, candidates may experience a variety of emotions, including fear, anxiety and uncertainty. Members of the CF clinic team and the transplant team can help candidates cope with their concerns.

Waiting times can range from a few weeks, to several months or years. Waiting is influenced by organ availability, a candidate's blood type and size, and the number of transplants done at a transplant centre. In Canada, the average wait for lung transplant surgery is approximately six to 18 months.

## Preparing for a transplant

Candidates are required to participate in a physical rehabilitation program designed by a CF physiotherapist and to eat a healthy diet as recommended by a CF dietitian. Adequate body weight and good physical health will help candidates during surgery, reduce the risk of complications, and help regain strength post-transplant. Many transplant centres also offer pre-transplant education programs and support groups.

## Proximity to transplant centre

Candidates on waiting lists are often required to find accommodation within a 2.5 hour limit of their transplant centre. A pager/phone may be required to ensure they can be contacted as soon as lungs become available.

## Donor organs and assignment of organs to candidates

Organs accepted for donation must meet certain criteria to ensure they are in good condition following the death of an organ donor. A variety of factors determine how organs are assigned including blood type, size of the available organ, length of time on a waiting list, and, in some centres, the degree of urgency. Race and gender have no bearing on the match.

## The call

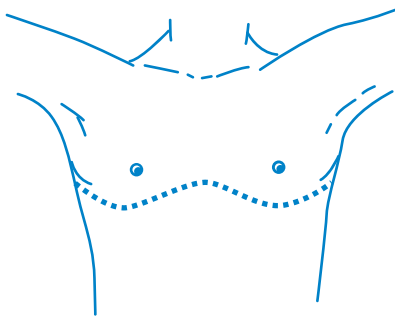
When suitable lungs become available, candidates will receive a call or page. As the call may come at any time day or night, candidates are advised to have a readiness plan.

## A “dry run” or “false alarm”

Once lungs have been removed from a donor, transplantation must occur as quickly as possible. The condition of donor lungs is not known until the lungs are retrieved - candidates may be notified that lungs are available, but arrive at the hospital to learn the lungs are not suitable for transplantation. This is known as a “dry run” or “false alarm”. Many candidates report that having a “false alarm” helped prepare them for the real call.

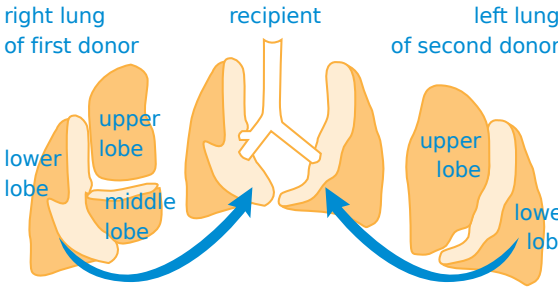
## The surgery

Individuals with cystic fibrosis undergoing lung transplant surgery always receive double-lung transplants to reduce the risk of infection. The surgery may take anywhere from five to 10 hours and is performed through a “transverse sternectomy incision” (across the chest). Following the operation, lung transplant recipients are placed on a ventilator to assist with breathing, and are moved to the Intensive Care Unit (ICU).



## Living donor lung transplantation

Due to the shortage of available organs, living donor lung transplants may be an option. In this procedure, lobes (portions of lung tissue) are removed from two separate donors for transplantation into the recipient. Age limits for living donors vary from centre to centre. Donors must be in excellent health and either a family member or longtime friend of the recipient.



## Post-transplant

In the ICU, recipients will remain on a ventilator until the new lungs are functioning well, which can take anywhere from one day to many weeks. Generally, recipients achieve close to normal lung function, but it can take several months to achieve full-lung capacity.

Immunosuppressive drugs, also called anti-rejection drugs, must be taken daily for the rest of a recipient's life to reduce the immune system's ability to attack and reject the new organs. These drugs will cause a life-long reduction in the ability to fight infection.

Two types of rejection can occur post-transplant. Acute rejection most often occurs within the first 12 months, even when anti-rejection drugs are taken faithfully. The transplant team teaches recipients how to recognize and monitor for signs of rejection so treatment can begin early.

The second type of rejection is called chronic rejection, or obliterative bronchiolitis (OB). It results from the progressive loss of lung function due to inflammation and irreversible scarring of the smaller airways. Chronic rejection occurs in more than 50 per cent of recipients surviving lung transplantation for more than five years, and is the main cause of death in long-term recipients surviving lung transplantation. Treatment is very difficult and aims to avoid further decreases in lung function by altering the dose of anti-rejection drugs, and, in very selected cases, re-transplantation.

In the weeks following transplantation, recipients can expect the following:

- *Monitoring for rejection of organs:* recipients must have regular pulmonary function tests, chest X-rays, and bronchoscopies.
- *Adjusting to the immunosuppressive drugs:* as each transplant recipient is unique, different combinations of drugs are required.
- *Education:* it may take time to learn new, post-transplant routines. Some recipients,

particularly those who are pancreatic insufficient, develop diabetes after lung transplantation. A nurse and dietitian will help these recipients learn to monitor and control their blood sugar.

- *Rehabilitation:* for several weeks following discharge from hospital, transplant recipients will usually undergo an exercise rehabilitation program. Soon after recovery, recipients can feel a difference in breathing and exercise abilities.

After being released from hospital, follow-up by the recipient's transplant centre and/or CF clinic is fairly rigorous. Initially, recipients attend weekly appointments. These appointments eventually taper off to monthly, and then yearly, as long as a recipient's health is stable.

## Lung transplant statistics in Canada

Survival rates for people with CF after lung transplantation are approximately 91 per cent (for persons without *Burkholderia cepacia* complex) and 64 per cent (for persons with *B. cepacia* complex), one year after transplantation. Survival rates five years after transplantation are 68 per cent (for persons without *B. cepacia* complex) and 34 per cent (for persons with *B. cepacia* complex).[2]

## Cystic Fibrosis Canada and Lung Transplant Programs

Cystic Fibrosis Canada supports organ and tissue donor awareness and supports every Canadian's decision to register to be an organ donor. In Canada, five institutions host lung transplant programs for individuals with CF and receive Transplant Centre Incentive Grants. They include:

- Vancouver General Hospital, Vancouver, BC
- University of Alberta Hospitals, Edmonton, AB
- Health Sciences Centre, Winnipeg, MB
- Toronto General Hospital, Toronto, ON
- Notre-Dame Hospital, Montreal, QC

[2] Statistics from Toronto General Hospital, 2010

## Additional resources

*The Guide: Resources for the CF Community*, www.cysticfibrosis.ca

*Financial Resources for Individuals Seeking a Lung Transplant*, www.cysticfibrosis.ca

*Lung Transplant Decision Aid for People with Cystic Fibrosis*, decisionaid.ohri.ca/

CF Education – Lung Transplantation, www.cfeducation.ca

BC Transplant – Lung Transplant Program, www.transplantbc.bc.ca


## Acknowledgements

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## What is Cystic Fibrosis Canada?

Cystic Fibrosis Canada (formerly known as the Canadian Cystic Fibrosis Foundation) is a national health charity established in 1960, with volunteers in more than 50 chapters across Canada.

Cystic Fibrosis Canada's mission is to help people with cystic fibrosis by: funding research towards the goal of a cure or control for cystic fibrosis; supporting high quality cystic fibrosis care; promoting public awareness of cystic fibrosis; and raising and allocating funds for these purposes.

For more information about cystic fibrosis, please contact your local chapter or:


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Cystic Fibrosis Quebec is a provincial association of Cystic Fibrosis Canada.

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