

TRANSPLANTATION

Lung Transplantation for Patients with Cystic Fibrosis

*Published by the Cystic Fibrosis Trust
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This factsheet has been written to assist you and your medical advisers. It is not intended to replace any advice you may receive from your Specialist CF Centre or CF Clinic.

The Cystic Fibrosis Trust funds medical and scientific research aimed at understanding, treating and curing Cystic Fibrosis. It also aims to ensure that people with Cystic Fibrosis receive the best possible care and support in all aspects of their lives.

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Introduction

Since the first heart transplant by Christian Barnard on 3 December 1967, organ transplants have emerged as one of the main medical advances of the 20th century.

The first successful heart-lung transplant for Cystic Fibrosis (CF) was performed in 1985. Since then, hundreds of transplants have been performed on patients with Cystic Fibrosis.

This guide has been written to help all patients with CF and their families who are seriously considering organ transplantation. After you have read this factsheet, there may be questions you think have been left unanswered. Please speak to your CF team at your Specialist CF Centre who will be able to answer any queries that you or your family may have.

Lung transplantation for Cystic Fibrosis

Many of the patients with CF who have undergone successful organ transplantation since 1985 were so breathless that they could hardly walk before surgery. Others were dependant totally on oxygen. Many have now returned to work or education.

These statistics are encouraging. Although deaths can still occur, 74% of patients survive one year after lung transplantation.

Some patients still receive heart and lung transplants but most patients with CF keep their own heart and only lungs are transplanted. The choice is recommended by the surgeon, depending on the availability of suitable organs and transplant centre.

Major surgery with hope – but also with risk

Lung transplantation is major surgery and therefore carries considerable risks. This is why it is appropriate only if you are severely ill and have received all other forms of conventional treatment without improvement. Lung transplantation offers new hope of life - a life which is more active and of better quality than before.

But a word of caution. It must be emphasised that lung transplantation is not a magic cure for Cystic Fibrosis. Although you will have new lungs, you will have to continue with all your medication for other parts of your body as you will still have Cystic Fibrosis. Your new lungs will need very specific care for the rest of your life, because transplanted lungs are liable to episodes of infection and rejection.

Lung transplantation is not, by any means, the most appropriate form of treatment for all patients who are severely ill with Cystic Fibrosis. Nor is it a type of treatment that all patients would want to consider. Therefore, on no account should you be persuaded to ask for a lung transplant against your wishes. An assessment for suitability for transplantation will be offered to you and if you are suitable and understand fully the risks as well as the potential advantages, then maybe you will be put on the list. It must also be pointed out that there are

many more patients needing or wanting this form of treatment than there are donor organs available.

The assessment

If you would like to be considered for lung transplantation you will need a period of intensive assessment, which usually takes 3-5 days in hospital. Some local CF Centres carry out a preliminary assessment before the assessment by the transplantation team. The purpose of this assessment is to gauge your medical suitability, how you will cope with the treatment that will be necessary for the rest of your life, as well as providing you and your family with sufficient information to allow you to make a fully informed decision.

The assessment includes x-rays, blood tests and tests of respiratory (breathing) function, both at rest and with exercise. If you are diabetic, take steroids, have liver or kidney disease, or have had previous chest surgery, you will need special assessment and preparation. For some people with CF, the risks of surgery may be greater than the risks of not having the operation. If you fall into this category then you will be given the most appropriate advice for your future management for Cystic Fibrosis. It is also important to mention that some types of infection may make people unsuitable for transplant.

You may be offered regular assessment with the transplant team while you wait for your transplant or your care may be from your Specialist CF Centre and they will keep the transplant team informed of your health.

During the assessment period, you may be introduced to somebody by email or telephone who has successfully had a lung transplant. You will have the opportunity to talk to the medical and nursing staff involved with lung transplantation, so that you can find out what actually happens.

If you still want to proceed, and the results of the medical tests indicate that this may be an appropriate treatment, you will meet the transplant surgeon. Your assessment will be discussed at a multidisciplinary team meeting and if you are accepted you will be placed on the waiting list.

The waiting time

Once you are on the waiting list, it may be many months or longer before suitable donor organs become available. During this waiting time it is important that you stay as fit as possible - **and** you must notify the Specialist CF Centre at once if there is any change in your state of health, if you are not at home or if you go on holiday. You must also contact the transplant co-ordinator if you go on holiday or your contact details change.

During the waiting time it is vital that you can be contacted immediately should suitable donor organs become available. You should stay where you can be contacted by telephone or, when you go out and the telephone is not manned, you should carry a pager. Having your

mobile phone with you is mandatory and should it be kept with you all times with the battery charged. In some Centres mobile phones are used instead of pagers.

Other important arrangements to remember are:

- Keep a bag packed.
- Have transport arrangements in place.
- Do not take aspirin or ibuprofen once you are accepted onto the list.

Sometimes you may be called whilst tests on the donor organs are being made. Often they are found unsuitable. These calls are disappointing but always a possibility.

It must be remembered that donor organs are matched with the patient, particularly in terms of blood group and size of transplant organs. There are, though, other factors which have to be matched and so unfortunately it is not always possible, naturally, to guarantee that every patient accepted on to the waiting list will receive suitable organs.

The operation

On the day of surgery (although this is often at night) the transplant centre will have identified organs which appear to match a patient with CF waiting for a transplant operation. If you are suitable and the best match then you will be contacted and asked to go as quickly as possible to the transplant centre.

Again, normal practice can often mean calling you in whilst tests on the donor organs are still being completed. Sometimes they are found to be unsuitable. This causes further disappointment, of course, but you need to understand that this is always a possibility. A limited amount of time is available and you must start being prepared for the operation even though all the tests on the donor organs are not complete.

When a correct match of donor organs to you is confirmed, you will be taken to the operating theatre.

Most patients with CF have double lung transplantation which means you keep your own heart and have two new lungs transplanted. (For those patients undergoing heart-lung transplantation, their own lungs and heart are removed and replaced with a new heart and two new lungs. Most patients receiving heart-lung transplantation donate their own heart to another patient. This enables the surgeons in a nearby theatre to save the life of a patient dying of heart failure. This procedure is called the *domino operation*.)

You will wake up in the Intensive Care Unit (ICU) and will be on a breathing machine, with a number of tubes in your chest. If things go smoothly, you may be breathing by yourself within one day of surgery. You may be out of the ICU within a few days. However you may need to stay on ICU longer until the transplant team is happy with your progress.

Whilst you are in the ICU, medical staff and relatives may wear masks, gowns and gloves. This is to protect you from infection.

Post operative care and follow up

You will be moved from the ICU to a single room, where on average you will need to stay for another four to six weeks. During that time you will exercise regularly, using fixed pedal cycles, treadmills and walking, so that your new lungs start working as well as possible.

After transplantation, you will have to take drugs for the rest of your life, so that your body does not reject the new heart and lungs or lungs. Most patients will have to take steroids as well. If rejection or infection of the lungs occurs, it may be detected by simple lung function tests, such as:

- The forced expiratory volume in one second (FEV₁).
- The forced vital capacity (FVC).
- Record daily temperature and weight.

Therefore, you have to measure your FEV₁ and FVC **every day** at home with a device called a spirometer, which is a small instrument for measuring the volume of air entering and leaving the lungs. These are the same measurements that you have been performing in clinic for years. If there appears to be evidence of rejection you may need a small biopsy taken of your transplanted lungs – this is called a bronchoscopy. This is carried out under light anaesthetic or other sedation and you may only need to be in hospital for one night. It is a very routine procedure with minimal risks attached.

You may also experience lung infections from time to time and these infections will need prompt diagnosis and treatment.

Self-treatment and tests

After the operation you will learn to take the necessary medicines and to complete a daily diary. This diary records which medicines you have taken, your temperature, lung function and weight.

After you have been discharged from hospital, for the next three months you need regular blood tests, x-rays and ECGs (tests to gauge the state of your heart). Although some of these tests can be done at the local hospital, you will need to return to the transplant centre quite often. As time goes by, hospital visits will be less frequent and, after a year, you will probably need to see a doctor only once a month.

After a lung transplant operation most patients take six to nine months to get the maximum benefit and will then be thinking about going back to education or work.

Quality of life

After lung transplantation, your quality of life is nearly always better than before the operation. Following successful transplantation many people will be in full time employment, some will be at school or in higher education and some will have married.

It must be said, though, that many patients will not be free from worry. The concerns you may have had before surgery about infection of your CF lungs may be in the past but you may have similar worries about rejection or infection in your new lungs.

The problems

Life for you with transplanted lungs is not, unfortunately, trouble-free. For example:

- Most patients with transplanted lungs do, occasionally, get episodes of infection or rejection. That is why it is vital that you complete your daily diaries and that you contact the transplant centre at once if you have any unexplained temperature or deterioration in lung function.
- Some patients get chronic rejection, which leads to a condition called *obliterative bronchiolitis*. When this happens the transplanted lungs do not work as well and patients become more breathless, although there are treatments available to help with this.

Bilateral living donor lobe donation

There is a shortage of donor organs and approximately 40% of patients will die whilst on the waiting list. Doctors in the USA have pioneered a technique in which two people, usually close relatives, each donate a lung lobe to the patient with Cystic Fibrosis. The potential donors need intense pre-operative assessment, must be very fit and must understand the risks involved for themselves and the patient with Cystic Fibrosis. Although a small number of such operations have been performed in the UK since 1995, techniques are still being developed and this option is not available at all transplant centres, nor is it appropriate for all patients. Please talk to your transplant physician if you would like further information about this procedure.

Finally

Lung transplantation is a form of treatment for some patients with Cystic Fibrosis, and is being performed in the UK and other parts of the world in both children and adults.

The main aim, of course, is still to find a cure for CF, so that patients' lungs do not become damaged and therefore will not need transplantation at all. Until a cure is found, lung transplantation for CF patients remains a form of treatment that may save lives.

How many lives? Well, that depends largely on the number of donor organs available. At present the shortage of donor organs is the major limit to more widespread use of lung transplantation.

Encouraging more people to register with the national donor list and to carry a donor card would be helpful. (www.uktransplant.org.uk or NHS Organ Donor Line ☎ 0845 60 60 400)

Questions

Answers to some often asked questions

What do I do if I think my child or I might be a candidate for lung transplantation?

Your first stage should be to visit your Specialist CF Centre. If you are not in contact with a CF Centre, ask your hospital doctor or a GP to refer you. You can find out where your nearest Specialist CF Centre is by contacting the Cystic Fibrosis Trust.

Where are lung transplantation operations performed?

There are five main centres: Royal Brompton & Harefield NHS Trust, London; Great Ormond Street Hospital for Children, London; Freeman Hospital, Newcastle Upon Tyne; Wythenshawe Hospital, Manchester; and Papworth Hospital, Cambridge.

How is the decision made whether I go on the waiting list?

You will undergo a thorough assessment, as described on page 3 of this factsheet. The physician and the surgeon will advise you what is best and most appropriate for you.

Can Cystic Fibrosis return to a transplanted lung?

No, the good news is that it cannot return. Research has shown that your new lungs do not develop Cystic Fibrosis. However, you may have other concerns about rejection and infection as described on page 6.

What about a second transplant if the first does not work?

Second transplants have been carried out but they carry a higher risk than the first operation.

What support is available to me and my family?

The assessment procedure is described on page 3 of this factsheet. During the assessment period it may be decided that there are areas in which the hospital can help. The Cystic Fibrosis Trust can offer some financial help to patients undergoing lung transplantation (and their families) when their needs are greater than those which can be met by social services.

The time on the waiting list can be a great strain for both patients and families. During this time you should keep in contact with your Specialist CF Centre. You can also ring at any time and talk to a member of the transplant team.

When you are waiting for a transplant the hospital may have a community team including a nurse specialist, physiotherapist, occupational therapist and social worker who can support you and your family while you wait at home.

Can my family stay at the hospital during and after the operation?

Accommodation is available for the closest relative throughout the stay in hospital. Financial

assistance may be available for those who need it. You are free at any time to discuss any concerns you may have with doctors, nurses or social workers. Additionally, CF Trust Support Service is always available to give you help, support and guidance. Their Helpline is listed at the back of this factsheet.

Further information

The Cystic Fibrosis Trust Support Service has trained staff to help answer any question not covered in this factsheet. The telephone number is:

Cystic Fibrosis Helpline

☎ 0845 859 1000

For further general information and literature published by the Cystic Fibrosis Trust please contact:

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