

Cystic **Fibrosis** *our focus*

Lung transplantation in cystic fibrosis

Factsheet – July 2016

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This factsheet provides general information about lung transplantation in cystic fibrosis (CF). It is not intended to replace any advice you may receive from your specialist CF centre.

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 was carried out in 1985. Since then, hundreds of transplants have been performed on patients with cystic fibrosis.

This factsheet is designed to help people with CF, and their families, who are considering lung transplantation. After you have read this factsheet, you might still have questions. Please speak to your CF team, who know your individual circumstances and can give tailored answers to your questions.

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Lung transplantation for people with cystic fibrosis

Lung transplantation has been available to people with cystic fibrosis for more than 30 years. Techniques have changed as surgeons have become more technically skilled; today the most common operation carried out is called a double-lung transplant, or a bilateral sequential lung transplant.

Survival rates are constantly improving, with approximately 85% of patients surviving for at least one year following the operation, and many returning to full-time work or education. Although this is encouraging, sadly deaths do still occur.

Major surgery: with hope, but also with risk

Lung transplantation may offer new hope of life; however, it is a major operation and carries considerable risks. This is why transplant is only appropriate when all other forms of conventional CF treatment have been exhausted. It is essential that patients considering transplant and those accepted for transplant adhere to all prescribed treatments, and maintain good nutrition for transplant to be a realistic option.

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 the condition. Your new lungs will also need very specific care for the rest of your life, as transplanted lungs are at risk of infection and rejection.

Lung transplantation is not the most appropriate form of treatment for all people with cystic fibrosis. Nor is it a type of treatment that everyone would want to consider. You should not feel that you have to discuss transplant against your wishes.

If you want to be considered for transplant and your CF team agree, then an assessment for suitability will be arranged with you. Following the assessment process, if you still wish to proceed, if you are clinically suitable, and fully understand the risks and advantages then you will be put onto the active transplant waiting list. However, there are many more patients needing or wanting a transplant than there are donor organs available.

The assessment

The assessment takes place in hospital and usually lasts up to five days. Some CF centres carry out a preliminary assessment before the full assessment by the transplant team. The assessment allows the transplant team to make a decision about your suitability, how you will cope with the long term treatment requirements, and provides you and your family with information to enable you to make your own choices.

The assessment includes many tests such as X-rays, blood tests, full lung function, an exercise test, as well as scans of your heart, abdomen and bones. For some people, the complications of CF and the risks of surgery may be greater than the risks of not having the operation. If you fall into this group then you will be given the most appropriate advice for your future clinical management.

During the assessment period, if you're interested, you may be introduced to somebody who has successfully had a lung transplant so that you can ask questions by either email or over the phone. You will also have the opportunity to talk to the transplant team, including the transplant surgeon, so that you can find out what actually happens. Your assessment will be discussed at a multidisciplinary team meeting and if you are accepted you will be placed on the waiting list.

Once you have been accepted onto the transplant list you may be offered regular assessment with the transplant team while you wait for your transplant. Alternatively, your CF team may continue with your care and keep the transplant team informed of your health status.

The waiting time

Once you are on the waiting list it may be many months or longer before suitable donor organs become available. However, you need to be prepared as you could be offered an organ as soon as you go on to the waiting list. During this waiting time it is important that you stay as fit as possible – and you must notify your CF team immediately if there is any change in your state of health.

It is vital that you can be contacted immediately should suitable donor organs become available. If you are not at home, your contact details change or you go on holiday, you must contact the transplant coordinator as you may need to come off the list for the duration of holidays abroad. Having your mobile phone with you is mandatory and it should be kept with you at all times with the battery charged. In some centres, pagers may be given to patients.

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

Donor organs are matched to the individual, particularly in terms of blood group and size of transplant organs. There are other factors that also have to be matched and so unfortunately it is not always possible to guarantee that every person accepted on to the waiting list will receive suitable organs. Sometimes you may be called while tests on the donor organs are still being made, and occasionally they are found to be unsuitable. These calls are disappointing, but are a possibility so try to prepare for this.

The operation

On the day of surgery (although, it is often at night) the transplant centre will have potentially matched organs to a patient waiting on the transplant list. If you are the most suitable match then you will be contacted and asked to go as quickly as possible to the transplant centre.

When you arrive at the hospital a limited amount of time is available, so you will start preparing for the operation even though all the checks on the donor organs are not complete. To start with you will undergo a variety of tests, including blood tests, urine samples, swabs and temperature, to ensure that you are well enough to undergo the operation. When the donor lungs are confirmed as a suitable match and all your test results show you are fit enough, you will be taken to the operating theatre. If the organs are not a confirmed match, surgery will not go ahead despite the time you have spent in hospital.

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 and IV lines in your arms. If things go smoothly, you may be breathing by yourself within one day of surgery and may be out of the ICU within a few days. Occasionally though, you may need to stay in the ICU longer, until the transplant team is happy with your progress.

Post-operative care and follow up

When you are ready you will be moved from the ICU to a single room, where you will stay for an average of four to six weeks. During that time you will exercise regularly, using an exercise bike, treadmill and going for walks. This helps your new lungs start working as well as possible.

After transplantation you will have to take a number of new medicines for the rest of your life; these drugs are very important as they prevent your body rejecting the new lungs. In addition to the anti-rejection medicines, most patients will have to take steroids. If rejection or infection of the new lungs occurs, it may be detected by routine tests, such as:

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

To identify problems early you will be asked to measure your FEV₁ and FVC every day at home with a device called a spirometer. This is a small version of the machine that you have been measuring lung function with at CF clinic for years. If there appears to be evidence of rejection you may need a small biopsy taken of your transplanted lungs – this is done using a small tube with a camera and small forceps at the end, the procedure is called a bronchoscopy. This is carried out under 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 that will be explained to you at the time.

As well as careful monitoring of the new lungs, you may also get lung infections from time to time; these infections will need prompt diagnosis and treatment.

Treatment and tests

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

After you have been discharged from hospital, you will need regular blood tests and X-rays over the next three months. Although some of these tests can be done at your local hospital, you will need to return to the transplant centre quite often. As time goes by, hospital visits will be less frequent and gradually you will probably need to see a doctor on a routine but regular basis – like your CF clinic visits.

It takes an average of six to nine months after the operation before most people feel well enough to think about returning to work or education.

After the operation

After lung transplantation many people go on to enjoy a vastly improved quality of life and enjoy activities that previously may not have been possible. However, it is important to remember that your new lungs will need very specific care.

Unfortunately life with transplanted lungs is not always trouble free, and complications can occur post-transplant.

For example:

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

Finally

Lung transplantation is a form of treatment for some people, children and adults, with cystic fibrosis.

The main aim, of course, is to find an effective treatment for CF, so that the lungs don't become damaged and therefore will not need transplantation at all. Until this is found, lung transplantation for people with CF remains a form of treatment that can save lives.

How many lives? That depends largely on the number of donor organs available. At present, a shortage of suitable donor organs in the UK is the major limit to more widespread use of lung transplantation. One simple way in which people can help is by encouraging more people to register with the national donor list and to carry a donor card (visit www.organdonation.nhs.uk/ or call the NHS organ donation on 0300 123 23 23). As well as registering, it is also important for potential donors to inform their families of their decision, as their decision could otherwise be overruled.

Frequently asked questions

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

Your first stage should be to visit your specialist CF centre and discuss your (or your child's) options with your CF team.

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 4 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, rejection and infection as described on page 6, can affect your lungs. It's also important to note that you will also have to continue with your CF medication for other parts of your body as you will still have cystic fibrosis.

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

Unfortunately, due to the shortage of organs available, second transplants are not possible. Where they have been carried out in the past, they carried a higher risk than the first operation.

What support is available to me and my family?

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 CF team. You can also ring at any time and talk to a member of the transplant team.

When you are waiting for a transplant your CF centre 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.

The Cystic Fibrosis Trust can offer practical, financial and emotional support to patients undergoing lung transplant and their families. Please contact our helpline on 0300 373 1000 or at helpline@cysticfibrosis.org.uk for more information about financial assistance and our other support services.

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

Accommodation may be available for the closest relative throughout the stay in hospital. In addition, financial assistance may be available for those who need it. Do ask your transplant or CF team for more information about this. You should feel free to discuss any concerns you may have with doctors, nurses or social workers. Additionally, the Cystic Fibrosis Trust is here to offer support and guidance.

Further information

The Cystic Fibrosis Trust provides information about cystic fibrosis through our factsheets, leaflets and other publications.

Most of our publications are available through our helpline and can be downloaded from our website or ordered using our online publications order form. Visit cysticfibrosis.org.uk/publications.

The Cystic Fibrosis Trust helpline can help you with a range of issues, no matter how big or small. Our trained staff can provide a listening ear, practical advice, welfare/benefits information or direct you to other sources of support. The helpline can be contacted on 0300 373 1000 or helpline@cysticfibrosis.org.uk and is open Monday to Friday, 9am – 5pm.

Calls to 0300 numbers cost no more than 5p per minute from a standard BT residential landline. Charges from other landlines and mobile networks may vary, but will be no more than a standard geographic call and are included in all inclusive minutes and discount schemes. If you are worried about the cost of the call please let us know and we'll call you back.

You can also find more information at our website cysticfibrosis.org.uk.

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More factsheets available at:
cysticfibrosis.org.uk/publications

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The information included in this publication is not intended to replace any advice you may receive from your doctor or CF multidisciplinary team and it is important that you seek medical advice whenever considering a change of treatment.

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