

# **Cystic** **Fibrosis** *our focus*

**Lung transplantation in cystic fibrosis**

Factsheet – March 2013

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# **Lung transplantation in cystic fibrosis**

## **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 factsheet has been written to help all patients with cystic fibrosis and their families who are seriously considering lung 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 or clinic who will be able to answer any queries that you or your family may have.

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## **Lung transplantation for people with cystic fibrosis**

Lung transplantation has been available to people with cystic fibrosis for almost 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 is 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 can still occur.

### **Major surgery with hope – but also with risk**

Lung transplantation may offer new hope of life; however, this is a major operation and carries considerable risks. It is therefore essential that patients considering transplant and those accepted for transplant adhere to all prescribed treatments, and maintain good nutrition if transplant is to be a realistic option. This is why it is only appropriate if you have received all other forms of conventional CF treatment without improvement.

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 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, by any means, 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. Therefore, you should not feel that you have to discuss transplant against your wishes. If you wish 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. Unfortunately there are many more patients needing or wanting a transplant than there are donor organs available.

### **The assessment**

If you would like to be considered for lung transplantation and your CF team agree that you are a suitable candidate, you will need a period of intensive assessment. The assessment takes place in hospital and usually lasts up to five days. Some local 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 sufficient information to allow 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.

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 specialist CF team may continue with your care and keep the transplant team informed of your health status.

During the assessment period, if you are interested you may be introduced to somebody who has successfully had a lung transplant so that you can ask questions either by email or over the phone. You will also have the opportunity to talk to the transplant team and 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.

## **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 specialist CF centre at once if there is any change in your state of health.

During the waiting time 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 then you must contact the transplant co-ordinator. Having your mobile phone with you is mandatory, 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, though, other factors which have to be matched 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 whilst 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.

## **The operation**

On the day of surgery (although this 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 which include 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.

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 on 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 on average you will need to stay for another 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)
- Recorded 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.

You may also get lung infections from time to time and 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 which medicines 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 regular basis – like your CF clinic visits.

On average it takes about 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 at once 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.

## Conclusion

Lung transplantation is a form of treatment for some people with cystic fibrosis and is available in the UK and other parts of the world for both children and adults.

The main aim, of course, is still to find an effective treatment for CF, so that the lungs do not 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.uktransplant.org.uk](http://www.uktransplant.org.uk) or call the NHS Organ Donor Line 0845 60 60 400).

## Answers to some frequently asked questions

### What do I do if I think I (or my child) 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 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, you may have other concerns about rejection and infection as described on page 7.

## **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. Unfortunately, due to the shortage of organs available, second transplants are not possible.

## **What support is available to me and my family?**

The assessment procedure is described on page 4 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 that can be met by the hospital or 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 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.

## **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. 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, the CF Trust Support Service is available to give you help, support and guidance. Their helpline is listed at the back of this factsheet.

## **I have heard about a new option for transplant called ex-vivo lung perfusion or EVLP. What is this and could I have the operation?**

EVLP is a process to evaluate and, if appropriate, improve lungs that are thought to have borderline function. This involves cleaning and aerating the donor lungs after they are removed from the donor. This is a research study which is co-ordinated by a team from Newcastle University and Newcastle upon Tyne NHS Foundation Trust, with all the adult lung transplant centres in England.

From April 2012 all patients listed for lung transplant in the UK have been offered the opportunity to take part in the DEVELOP study, as will all patients currently on the lung transplant list. This means that they have been considered as a potential match when donor lungs requiring EVLP assessment and reconditioning become available.

### **Further information**

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

Most of our publications can be downloaded from our website ordered using our online publications order form.

Visit **[www.cysticfibrosis.org.uk/publications](http://www.cysticfibrosis.org.uk/publications)**.

Alternatively, to order hard copies of our publications you can telephone the CF Trust on 020 8464 7211.

If you would like further information about cystic fibrosis please contact:

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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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