

Cystic Fibrosis talking about transplant



Cystic fibrosis
and transplant

Fighting for a Life Unlimited

Sincere thanks to all the people with CF, their partners and families who generously shared their stories, offered guidance and were so enthusiastic about the development of this booklet. We are also immensely grateful to the professionals from the CF and transplant teams for giving up their time to talk about cystic fibrosis and transplant, answer questions, review drafts and generally share their expertise! Thank you all very much.

*This booklet provides general information about transplant and is not intended to replace any advice you may receive from your specialist CF or transplant centre.



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Introduction

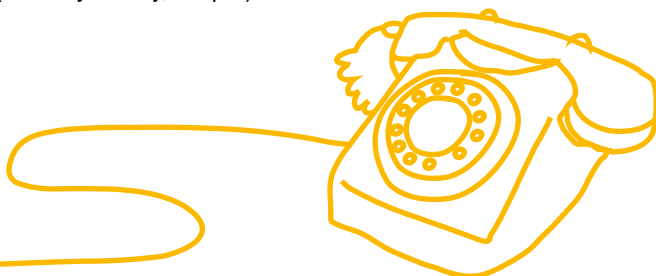
The Cystic Fibrosis Trust appreciates that while there is excitement and anticipation about new and emerging treatments, there are many people with cystic fibrosis (CF) who are struggling in the here and now, and for whom existing treatments are no longer having an impact.

Lung transplant is a possible option for people with CF who aren't responding to standard treatment. Transplantation of other organs, including liver, can also be necessary due to the damage the condition inflicts across multiple organs.

People with CF who've been through transplant have told us that you can never be fully prepared for what the transplant process is like. In part, this is because you just can't explain how some things really feel, and also because everyone will have a different experience. However, engaging with the information you will receive from your clinical teams, asking questions, and hearing about other peoples' experiences, can all contribute to increased feelings of empowerment and preparedness.

This booklet presents some difficult topics and challenging considerations but also inspiring stories showing a range of experiences relating to transplant, from which you may find elements that are relevant or interesting to you. We hope the information and stories will help you better prepare for the transplant process, encourage you to ask questions, and seek support.

If you have any concerns, the Cystic Fibrosis Trust helpline can provide support and a listening ear. If you would like to talk, please call us on **0300 373 1000** (Monday-Friday, 9-5pm).



Lung transplant

Lung transplant can offer hope to people with CF; however, it's a major operation and one that carries considerable risks. That's why lung transplant is only appropriate when all other forms of conventional CF treatment have been exhausted.

“Lung transplant isn't for everyone, it doesn't always feel like a good deal for some people. It's also important to be aware that it's not a forever solution; but it can really increase quality of life for people with CF when other treatments are no longer working.”

CF psychologist

Transplant is not a cure for CF; transplanted lungs will not have CF and will never develop the condition. However, as CF is a genetic condition, this means that you will still have CF in the rest of your body and may still need CF treatment, even after a successful lung transplant. Transplanted lungs need very specific care for the rest of your life and many people who've had lung transplants say that it feels like swapping one chronic condition for another, although with fewer symptoms and an improved quality of life.



Liver and other organ transplant

As you will know, CF is a multi-organ disease and some people require organs other than lungs to be transplanted because of the damage caused by the condition, most often the liver or pancreas.

The transplanted organ will not have CF and will not develop it, but as CF is a genetic condition, the rest of your body, including your lungs, will continue to have cystic fibrosis.

“I was diagnosed with cystic fibrosis-related diabetes and cystic fibrosis-related liver disease in my mid-teens. They were mostly under control, but the increasing damage to my liver began to throw up very difficult and dangerous symptoms, especially in relation to my diabetes; one not being able to feel sudden drops in blood sugars. I would collapse unconscious without warning. I was also struggling with severe lethargy, abdominal distension and oesophageal varices (bleeds) due to the liver damage. With all of this going on, I was absolutely over the moon to be accepted onto the transplant list, despite knowing all the risks.”

Laura, five years post-liver, pancreas and small bowel transplant

While the process for other organ transplant is largely the same as for lung transplant in terms of being assessed, listed and waiting for the call, people with CF who are going through other organ transplants can have very different experiences to those undergoing lung transplant.

- Symptoms are clearly different. As well as managing CF in terms of lung health you will also be dealing with symptoms related to the affected organ/s, such as oesophageal varices, abdominal distension (swelling), lethargy and/or diabetes.
- Assessment requires a careful balance to ensure that existing lungs are healthy enough to cope with major surgery. When existing lungs are not being replaced with donor lungs, they need to be healthy enough, not just to get through surgery, but also to recover well post-transplant.
- Recovering from major surgery and maintaining CF treatment for lungs can present challenges.
- The treatment burden is not usually as reduced with other organ transplant as it may be with lung transplant, and immunosuppressants medications that are critical to protect the transplanted organ potentially expose CF lungs to more infection.

"The prospect of transplant was quite scary, even though I'd known most of my life that it'd happen. It felt like a really fast progression, from feeling very well to six weeks later being listed for a liver transplant."

Aoife, four years post-liver transplant

"I had a liver, pancreas and small bowel transplant. Recovery was very tough. One of my lungs had collapsed and consolidated during the operation, and chest clearance was incredibly difficult and upsetting due to the pain of the surgical scars. I hadn't quite realised just how hard it would be recovering from major surgery, still having lungs affected by CF."

Laura, five years post-liver, pancreas and small bowel transplant

Although there are inevitably potential hurdles and risks with this major surgery, there can also be significant gains in quality of life.

"Not having to take Creon anymore has revolutionised my life! I don't have to think about what I eat, and for the first time in my life I have a normal BMI. When I was pancreatic insufficient, I was always thinking about what to eat and it's a constant pressure to gain, or at least maintain, your weight. Now I can have a salad, guilt free! Feeling healthy, and with an even better lung function than before, is incredible."

Laura, five years post-liver, pancreas and small bowel transplant

"It took me about 10 months after the operation to feel the benefit, up until then I really worried that I'd done the wrong thing. Even though realistically I had no choice, I'd have died without it. The benefit now is clear though; I took a year out to go travelling, I have energy, a full-time job and my lung function has also improved."

Aoife, four years post-liver transplant



Talking about transplant





"I found transplant quite a positive thing to think about, knowing that it could be a possibility for me if my health deteriorated."

Tor, six years post-lung transplant

Why now?

Although people with CF may have had transplant at the back of their minds for many years, it may not be actively considered or talked about until their CF team bring up the possibility of referral for assessment for transplant. Starting to really think about transplant and what it could mean, can be challenging.

“I’d never really given much thought to lung transplant. I knew of it, and the basics of what it involved, but it was always something that other people go through. I never believed, or maybe didn’t want to believe, that I’d ever end up needing a transplant.”

Richard, seven years post-lung transplant

“When my team suggested transplant, it felt like a slap in the face. I knew I was really ill; it was end-stage CF, but I was so used to the oxygen, breathlessness, tiredness and exacerbations that it had become my normal.”

Veronica, two years post-lung transplant

Some people with CF have told us that when their CF team suggested referral for transplant, they didn’t feel ‘sick enough’. Your CF team will explain in detail why they are suggesting the referral at this time and encourage you to think about changes in your health and quality of life. They are working to identify your window of opportunity for transplant; it’s a balance between being poorly enough to justify the risks of transplant but being well enough to ensure a good recovery from the operation.

The Cystic Fibrosis Trust works hard to encourage organ donation, but sadly there are not enough organs for everyone on the transplant list. For this reason waiting times on the transplant list may run into many months or years, and during this time your health may deteriorate further.



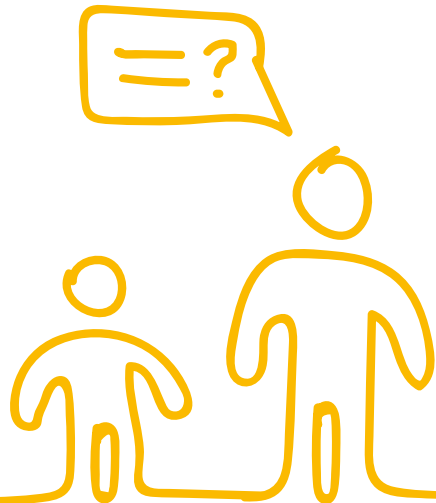
What does transplant mean for me and my family?

Transplant means different things to different people; some view it as a positive development, particularly if they are critically ill, whereas others feel that it's an unwelcome sign of how much their health has deteriorated. Your own life priorities, beliefs and circumstances come into play when considering this life-altering operation.

“The first time I was recommended for transplant, I felt quite devastated, I really didn't realise I was that ill. It kind of crept up on me and I didn't see the deterioration. Even though all my life I'd known this was likely to be the outcome, I still found it hard to accept. My health actually improved to the extent that I was removed from the list. Several years later I was assessed again and it felt very different this time, I was really poorly and I felt that I was dying so I didn't hesitate to go forward for assessment.”

Vicky, 18 years post-double lung transplant

It is your decision whether or not to accept the referral for assessment. If it's helpful, talk to your family, friends and CF team, use them to help work out the pros and cons, but keep in mind that the decision is yours. Even if you decide to go for assessment, it doesn't commit you to being listed and do remember that the assessment may indicate that transplant is not a possible option for you.



Assessment

The transplant assessment most often takes place at a transplant unit and can take up to five days. The assessment includes a range of tests such as X-rays, blood tests, full lung function tests, exercise tests, as well as scans of your heart, abdomen and bones. Because the transplant process is so demanding, every aspect of people's functioning is looked at to ensure they have the best chance of doing well. This may also include assessment with a psychologist and a social worker. As well as finding out whether you can cope with the transplant process, this is an opportunity to get to know the new team who may be looking after you, and to plan any support you may need post-transplant with them.

There is detailed information about assessment and every aspect of transplant in the patient information booklet, which those being assessed for lung transplant should receive in advance of the assessment.



"At assessment we really emphasise that transplant is a team effort; we need patients to take responsibility for their part of the partnership, which is to engage with their care and work with us."

Transplant Psychologist

"I found the assessment process emotionally and physically gruelling. No stone was left unturned – it was, quite rightly, utterly comprehensive."

Aoife, four years post-liver transplant

"The assessment process was much more intense and in-depth than I'd expected."

Oli, 10 years post-lung transplant

The assessment process 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. It's important to recognise that if you are found to be suitable for transplant, the decision whether or not to be listed is yours.

Decision making

You might feel very unsure about what to do for the best. This is completely understandable – it's a very hard decision. Some people feel under pressure from their family, or feel in some way that they are letting others down if they choose not to be listed; try to focus on yourself and remember that you are not obligated to anyone else. It's your decision and you are the one who has to go through the operation and the recovery.

“The decision-making process around transplant messes with your head. Right now I can do stuff with my kids, participate, enjoy life, but I'm ill and restricted in the types of activities I can do. It feels like a gamble right now; if I get the call right now I'm risking what I have today, but if I don't take it I'm risking what I might have tomorrow. My focus is my family and they are at the heart of my decision making.”

James, currently on the list for a lung transplant

Often people find it helpful to have a partner or family member with them during assessment; there's a lot of information to take in and it can feel overwhelming.

“Don't be afraid to ask questions, no matter how silly you might think they are, it's better to know the right information. Don't let things fester in your mind and start worrying.”

Libby, five years post-lung transplant

“If you ask me what worries me most about being listed: pain – no. Survival – no, not so much. How will it affect our day-to-day life – yes, absolutely. Who will look after the children, will my wife be able to get time off work, what about the costs of travel and accommodation? It's those practical issues that concern me.”

James, currently on the list for a lung transplant

Psychologists have told us that it's useful to think about your life as lanes of a road. You can continue straight ahead, with no transplant, and eventually reach the end of your road with cystic fibrosis. We all reach the end of our roads, whether that's from CF or something else, and sometimes the road that is familiar to us is less scary. However, you can decide to take the exit onto another road for transplant. That road might take you onto different locations such as employment, relationships or other new opportunities, it might extend the length of your road, or it might not.

The difficulty is, if you decide that you want to take the exit when you've already driven past it, it may not be possible. It's important to talk to your transplant team about the right time to be listed.

“Sometimes people decide that transplant's not for them and they don't want to go through it and then when their health becomes intolerable they change their mind, at which point it can be too late. It does happen and it's incredibly difficult for everyone involved.”

Transplant Nurse Consultant

Advance care planning

The supportive and palliative care team are sometimes introduced at transplant assessment, although in some cases you will have already met them via your CF team. This team are experts in symptom management (such as breathlessness, pain and anxiety) and will also focus on practical support.

It is important to note that involvement from palliative care doesn't mean that people around you think you are going to die imminently; their role is to support you and your family at a time of declining health or challenging symptoms. Their involvement will be in addition to the active CF care that your team will provide.

"I struggled with the idea that my daughter, Tor, was being referred to palliative care. Hospice care meant only one thing to me; that she was going to die and, of course, I baulked against it. But the hospice team were lovely and supported us as a family at a very challenging time. Tor is now six years post-transplant."

Christine, mum to Tor,
six years post-lung transplant

They may suggest some 'emotional homework', for example, memory boxes and letters for loved ones. This can be challenging and it is your decision whether you wish to do it or not. The team will help you to maintain hope for the best outcome, but also to make appropriate plans in case your transplant does not happen.

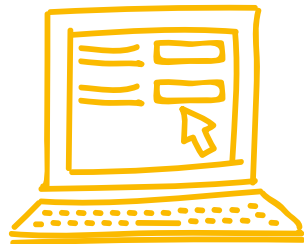
"I was advised at assessment to 'arrange my affairs'. I didn't think this was negative, I was so poorly then that it was a realistic possibility that I could die. My mum found it really hard, she thought I had a negative mindset, but I didn't, I just felt strongly that I wanted plans in place should the worst case happen."

Veronica, two years post-lung transplant

"I found the assessment process emotionally harrowing, there's so much information to take on board and difficult conversations to have. I'm a very positive person and even though I was hoping for the best, I was also making plans for the worst, including making a memory box for my son and planning my funeral. It's different for everyone, but I wanted to do this, it was all for my son and husband."

Vicky, 18 years post-lung transplant

The Cystic Fibrosis Trust has a booklet on planning for end of life, which you might find helpful. It is aimed at anyone, regardless of their current state of health, who wishes to have plans in place for the end of their life, [cysticfibrosis.org.uk/endoflifeplanning](https://www.cysticfibrosis.org.uk/endoflifeplanning)



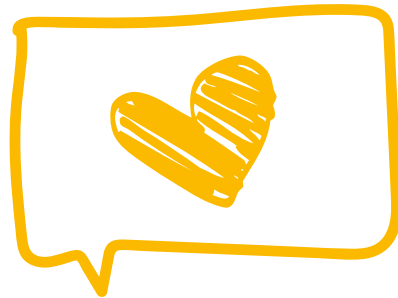
When transplant is not an option for you

For some people, the complications of CF and the risks of surgery may be greater than the risks of not having the operation. Transplant would then not be a suitable option for you. If you fall into this group, then you will be given the most appropriate advice for your clinical management and the chance to talk about what care you want for the future.

This can, of course, be incredibly difficult for the person with CF, and for their partner or family. The CF team will be there to provide support, and input from the palliative care team at this time can be invaluable. The Cystic Fibrosis Trust helpline provides a listening ear. You can call our friendly team on **0300 373 1000** (Monday-Friday 9am-5pm).

"Unfortunately, there are times when people are not suitable for transplant. Sometimes this is because they are just too ill to survive the operation or the risk of transplant is felt to outweigh the potential benefits."

Transplant Nurse Consultant



Waiting

Waiting





Waiting

The Cystic Fibrosis Trust works hard to encourage organ donation, although sadly there are still not enough organs for everyone listed for a transplant. This is why people who are listed don't know when, or if, they will get a transplant. The average wait for a lung transplant is 18 months, but it can be shorter, longer, or might not happen at all.

Cystic fibrosis and transplant teams work in partnership, drawing on all of their expertise and knowledge, to make sure that transplant assessment is timed correctly, so that if an organ does become available, the person with CF is prepared, in good enough health and ready to go.

Although most people wait for many months or longer, it's not always the case, so you do need to be prepared as soon as you are accepted onto the list.

It is vital that you can be contacted immediately should suitable donor organs become available. If your contact details change or you go on holiday, you must contact the transplant coordinator. You also need to keep your transplant coordinator informed if you are admitted to hospital. Having your mobile phone with you is mandatory and it should be kept with you at all times, with the battery charged.

Other important arrangements to remember are:

- Keep a bag packed
- Remember to take your medications and NIV with you
- Have transport and domestic (childcare etc) arrangements in place
- Don't take non-steroidal anti-inflammatory drugs (such as aspirin and ibuprofen) once you are accepted onto the list; speak to your team for more information about this and to explore other options for pain management.

Prehab

Prehab refers to the work you do before transplant, in order to stay well and in the best possible shape for the operation. Even when people are very poorly, there can sometimes still be opportunities to maintain or improve health, such as armchair exercises or dietary changes. As well as focusing on your physical health, you may also work with a psychologist to optimise your psychological health, by developing effective ways of coping with the worry and sadness that can sometimes affect people with CF who are very unwell.

Engaging with prehab is a key part of your responsibility to the transplant process. The whole process of transplant requires a partnership approach; everyone needs to do their bit to get the best possible outcome, and prehab is your part of that team effort while you are on the waiting list.

“I really didn’t want to have the PEG fitted, but my weight was low and I was advised that this was the best way forward. I wanted to be as healthy as possible in case the call came.”

Helen, 11 years post-lung and liver transplant

“I was told to get in the best shape possible for the operation, including to increase my weight and lung function as much as I could, and I worked very hard to do that. I increased my calorie intake, and would even be nebulising while on my exercise bike! If I’d known then how much it would benefit me, I’d have definitely tried to do even more.”

Laura, five years post-liver, pancreas and small bowel transplant



Life on the list

While you're on the transplant list, it is sensible to continue living life as normally as you can. Of course this isn't easy, particularly in the early days. However, people with CF who've been through transplant have said that you do find ways to put it to the back of your mind, although it's always there lingering.

"I waited for 11 months for my call. Although you try to live life normally, it's always there at the back of your mind and you can't plan anything for the future or turn your phone off. It's very much taking each day as it comes."

Aoife, four years post-liver transplant

"When I went on the list I thought about all the pros and cons, practicalities and how I'd cope with a false call. I gradually got my head round it all. But the one thing I didn't really think about was how I'd feel if I didn't get any calls, which is exactly what's happened. Fortunately I've managed to stay relatively well and stable whilst on the list, but it's now been two years without a single call. It's made me feel very forgotten, even though I know I'm not, because that's not how it works. It's becoming harder to keep believing the call will still come, but I know I'm lucky to have remained stable, so I just have to try and think positively."


Katy, currently on the transplant waiting list

Do participate in life as fully as you can, and try not to be tempted to put things off until 'after the transplant', because unfortunately, you cannot predict when that might be.

It can help to have a focus; some people are well enough to remain at work (particularly where working from home is an option), while others have the distraction of family. Having a goal or aim to achieve something that you can't currently do might help you to focus on what you are aiming for in the long run.

"I immersed myself in work and campaigning – I wrote a blog about being on the list, appeared on a couple of TV shows, spoke on radio programmes and campaigned on organ donation issues too. For me, it was important to keep busy and also to keep working. I didn't want to have a gap on my CV, so even though I wasn't in a paid job at this time, I was still doing work relevant to my career."

Mark, 12 years post-lung transplant

A man with short brown hair and blue eyes is sitting in a hospital bed. He is wearing a dark blue long-sleeved shirt with thin yellow horizontal stripes. He has a nasal cannula in his nose, connected to a clear tube. He is holding an open book and looking at it. He has a blue flame tattoo on his left forearm and a red flame tattoo on his right forearm. The background is a plain, light-colored wall.

"It's hard waiting for the call. I worry that I might be out of signal range, or if I take the kids swimming I need to phone my father-in-law so he can 'man the phone'. I did come off the list for a few days because we were taking the children to Thomas Land and I wanted to enjoy it and not be worried about the call."

James, currently on the transplant waiting list

“My focus was, and continues to be, my son and husband. When I was ill I couldn’t be the mum and wife I wanted to be. I couldn’t participate. For me, transplant was my only option to get that family life that I so desperately wanted for our future. Before my transplant I compiled a list of all the things I planned to do post-transplant; go round the block with my son on his go-cart, have a night out with my husband and arrange for him to have a weekend away. I started arranging that trip for him while I was still in hospital recovering.”

Vicky, 18 years post-lung transplant

You might find that people around you ask well-meaning, but rather frustrating questions, such as “When are you getting your transplant?” or “Is there any update on your transplant?” It can be useful to have a ‘stock answer’ to these types of questions. Alternatively, the Cystic Fibrosis Trust have developed a webpage that explains a little about the transplant process, including why timescales are unknown, cysticfibrosis.org.uk/abouttransplant

“I found it hard because people would say things like ‘still no call?’ or ‘why is it taking so long?’, but it was just because they didn’t really understand.”

Libby, five years post-lung transplant



For some people, particularly if they are very poorly, a long wait offers up time to think and worry. If you are struggling with the stress of being on the list, it may be an idea to discuss it with a member of your CF or transplant team, or with friends and family.

“When I was on the list, I just felt that I didn’t have anything to talk about. My friends would visit, but my life was so dominated by treatment that I had nothing to talk about. I wanted to hear about their lives and news, but at the same time, that also made me feel sad and frustrated that I couldn’t be part of it.”

Tor, six years post-lung transplant

“If you’re struggling with the wait, just ring your coordinator. I did several times and she reassured me that they were still looking for a donor and they hadn’t forgotten about me (as sometimes that’s how you might feel).”

Libby, five years post-lung transplant

It is important to keep in touch with your transplant coordinator about significant changes in your health, including weight change, chest infections, IV antibiotics and increases in steroid doses.

Coming off the list

You can come off the list at any time. This could be a temporary measure, perhaps for a holiday or other event that takes you beyond the reach of a call for transplant. You can also come off the list entirely, should

you change your mind about going through transplant. If you wish to be reinstated on the list, a review of your health or full reassessment may be necessary.

Children

If you have children, it's important to think about how the transplant process might affect them. It can really help children cope with this uncertain period in life if they are involved in the process in some way; helping pack your hospital bag, or packing their own if they are going to family or friends if you get the call.

“Antony knew the CF centre and the staff, but he didn't know the transplant centre or team. The social worker suggested taking him to the ward where I would be in recovery, so that he could picture where I was. We made it a special day and went shopping for Lego as well as visiting the ward. He also had a photograph of the ward so he could picture me there.”

Vicky, 18 years post-lung transplant

It's useful not just to plan out childcare, but to also think about actual logistics. For example, if the call comes in the middle of the night will you wake your child/children? What if it comes during the day and they are at school, would you go to the school to tell them that you're going to the hospital?

There is no right or wrong way to approach these situations; it's just important to have a plan and make sure that the children are fully aware, so that there are no surprises. For children, knowledge and predictability is power. Do try to be honest with them and answer their questions as clearly as you can. While it may be tempting, it's not helpful to make promises you may not be able to keep.

In hospital, you can continue involving your child/children through regular phone contact, but also think about other ways you can engage with your family. Children often like getting post, so you might like to have a batch of postcards ready in your suitcase that you can send to them as you recover. It helps them to feel part of the process and keeps them up to date.

Some families do choose to do memory boxes together, and this is something you could discuss with any of the teams involved in your care, particularly with the palliative care and support team.

False alarms

The transplant team want to make sure that the lungs (or other organ) you receive give you the very best possible chance of good recovery and an improved quality of life. Donor lungs are only viable for a certain amount of time, and so often you will need to be in the hospital, ready, while the lungs are being visually checked. Doctors will be looking at the lungs for any sign of disease, infection or bleeding. Unfortunately, it's not unusual to be called for transplant, only to find on arrival that the lungs are not suitable.

Although these 'false alarms' can be very stressful and disappointing, some people also find a degree of reassurance from these calls, just knowing that they are 'on the radar' and that lungs are actively being sought for them.

"I was absolutely high as a kite with the first call. But after several hours of waiting, I was sent home because the lungs weren't suitable. It was the next day that I really dropped, my mood was very low for some time. I was worried it could have been my only chance and that maybe it'd passed me by."

Vicky, 18 years post-lung transplant



"My first false alarm was only six weeks after I was listed and I got as far as the theatre door. In a strange way I was relieved it didn't go ahead, I didn't feel ready."

Oli, 10 years post-lung transplant

"I was generally very positive throughout the transplant process and kept my eye on the prize. I found the false alarms really hard though. I was on the list for three years and had four false alarms. Twice I made it to the hospital only to be told it wasn't happening. It's a real plummeting feeling and it took my wife and me several days to get back on an even keel."

Paul, six years post-lung transplant

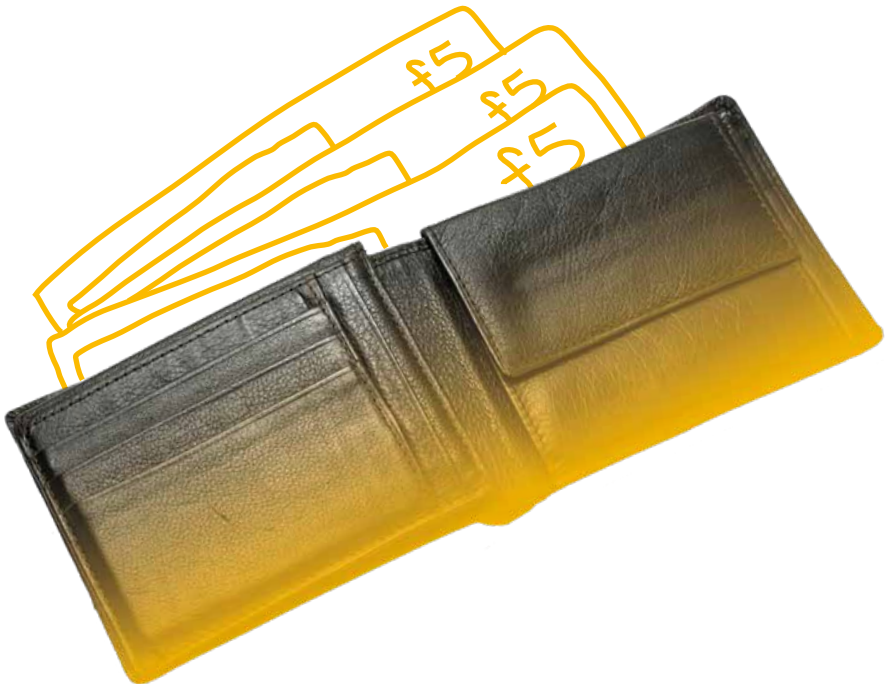
"The first few calls I had were quite nerve-racking. I was fully expecting to undergo the operation each time, and then when I didn't, I felt a mixture of emotions – both relief that I got to go home and carry on as normal, and at the same time dejection that my health was still bad and getting worse all the time"

Richard, seven years post-lung transplant

Financial support

There can be costs associated with going through transplant, particularly in relation to travel and accommodation costs during assessment and following the operation. Ask your transplant team about financial support - some teams have social workers or other members of staff who will be able to provide advice. Accommodation is sometimes available for a partner or close family, usually with a charge, at the hospital, so do ask your transplant team for more information.

The Cystic Fibrosis Trust has a welfare and benefits specialist, who may be able to support you with financial queries, and a transplant grant is also available to help families to cope with the financial burden of this process. You can find out more about these support services by contacting our helpline on **0300 373 1000** or via **helpline@cysticfibrosis.org.uk**



Operation and recovery



"I think having CF sort of prepares you a bit for transplant. I'm used to invasive medical procedures, I'm used to talking to clinicians and I know all about treatment adherence, so I felt pretty well prepared for that side of things."

Aoife, four years post-liver transplant



The operation

When you get the call for transplant, which is often in the evening, you'll be asked to make your way to the transplant centre and it's at this point that your plans should all kick in as smoothly as possible.

"When the call came, the nurse said to take my time. So my first reaction was to take a bath – I don't know why! Maybe I just needed some space to process what was happening. It was hard to say bye to my dad and sister, I didn't know if I'd see them again."

Veronica, two years post-lung transplant

"I don't consider myself to be a lucky person, I never win raffles or prizes or anything like that, but it feels that all my luck was centred on that day."

Helen, 11 years post-lung and liver transplant

On arrival at hospital you'll undergo a variety of tests to make sure you are well enough for the operation; these will include blood tests, urine samples, swabs and temperature checks. If the donor organ is confirmed as a suitable match, and your test results confirm that you're healthy enough to proceed, you'll be taken to the operating theatre.

Transplant operations take many hours, usually between six and 12 hours for a lung transplant, although this will vary from person to person, so ask your transplant team for more information.

"I was in theatre for eight and a half hours. It's a part of my life that's missing."

Veronica, two years post-lung transplant

"I think some patients are a bit shocked that they have to have their torso completely shaved for surgery."

CF Psychologist

After you are put under general anaesthetic, you will have no awareness of any of the process until later on when you wake in the ICU.



ICU

Waking up in ICU

Your transplant team will tell you about the intensive care unit (ICU) in detail, so that you're prepared for the unique surroundings and experiences. The noise of the machines, being connected to lots of devices, the lack of a sense of night or day, and difficulties in communicating can all feel a bit frightening for some people, so it's helpful to get as much information as you can in advance.

It's also important to think about your support networks. Who will visit? Who could act as a contact point for other concerned family and friends? Some people like to have their ICU experience documented, and you may like to ask family or a partner to keep a diary and take photos; do check this with your team well in advance, as some units have restrictions on photography.

The experience of waking up in the ICU is different for everyone; a few report feeling an instant 'wow' moment with new lungs, whilst others do not. It can be a very strange experience, partly because of all the physical changes and connection to devices, but also because of the cocktail of anaesthetic and pain-relieving medications. The ICU teams are experts in caring for patients post-surgery, and you will be very closely monitored.

"Waking up was confusing and disorientating. Looking back I have a jumble of memories all in the wrong order and snippets of things that happened."

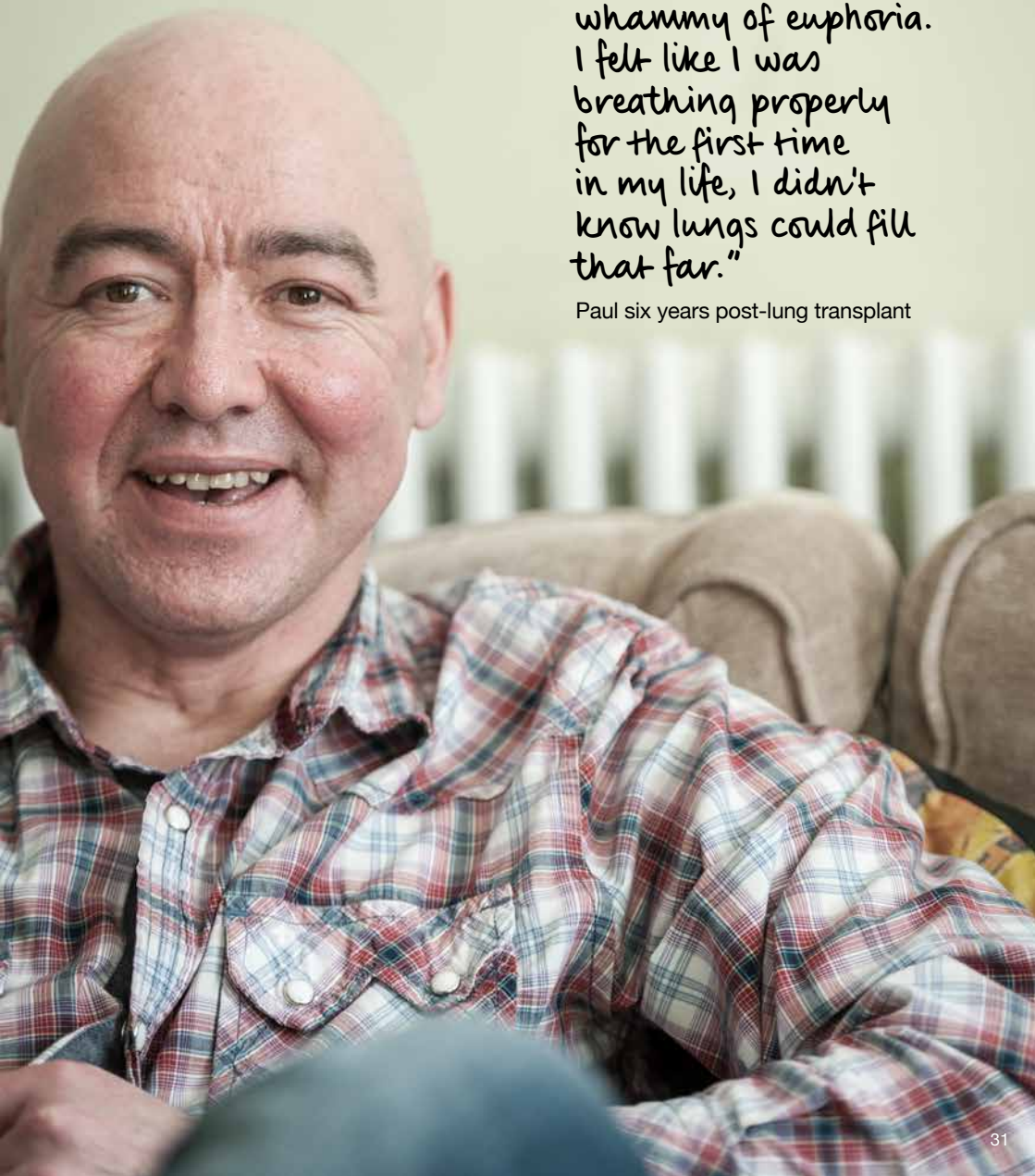
Oli, 10 years post-lung transplant

"My first breath was awful - rattily breathing and lots of pain. All I could think was, "oh my god, it hasn't worked". I remember being really distressed when I was coughing up sputum during my first week of recovery. I was crying and saying "these aren't my lungs, this isn't my sputum", it was such a lot to get my head around. I also struggled with constipation and that was very difficult to cope with."

Veronica, two years post-lung transplant

"Waking up in the ICU was quite shocking; I knew that I'd be hooked up to lots of machines, but I guess you can't really prepare for how it'll feel. To be honest though, the thing I most struggled with in ICU was getting a bed bath - I just hadn't thought about it and wasn't prepared for it."

Tor, six years post-lung transplant



"When I woke up, I had an instant feeling of new lungs, plus pure joy at being alive, so it was a double whammy of euphoria. I felt like I was breathing properly for the first time in my life, I didn't know lungs could fill that far."

Paul six years post-lung transplant

Hallucinations

The combination of anaesthetic drugs and painkillers may cause hallucinations for some people following transplant surgery, although not everyone will remember them. You may be lucky and your hallucinations will involve something pleasant, like a beach in the Maldives, but unfortunately some hallucinations can be more frightening or strange. This is quite normal and the ICU team are experts in dealing with such side-effects.

“People can naturally feel pretty anxious in ICU and some have told me about beliefs or hallucinations that they were at risk in the ICU – even that the staff might be doing them harm or doing things wrong. This is understandable as the mind is trying to deal with feeling under threat. The staff on the ICU understand if this happens due to drug effects.”

CF Psychologist

“Most of my hallucinations were quite pleasant actually, but I do remember feeling like I was lying in a pool of my own urine at one point, which was a bit weird. I also went through a period of being unsure if my lungs would continue working if I fell asleep so I was terrified of falling asleep.”

Paul, six years post-lung transplant

“I remember waking up in the ICU and looking at all the little blue people - I wondered if they were oompa-loompas, and then I wondered if I'd died. It was very surreal, but I felt very calm and in no pain at all. I was just quite fascinated by all the tubes and buttons. Apparently I had been thrashing about a bit before and that was quite distressing for my family, but I didn't remember it at all.”

Helen, 11 years post-lung and liver transplant

“Sometimes people have some insight while they're hallucinating so you can talk to them about their experience and help them re-connect to reality. We use grounding objects (something familiar and soothing) to help them feel safe in their environment. It is important to reassure people that they are in a safe place. The presence of family members can be very helpful.”

Transplant Psychologist

In some ICUs, staff will keep a diary for patients to help them to fill in the blanks in their memories, and to separate what was real and what wasn't. Many people find this useful, and some ask their family and partners to also keep a diary and to take photographs to help them connect with this lost period of their life. If you are considering this, do check with the medical team beforehand, to ensure that photography will be allowed.

Recovery

New medications

After transplant you will have to take a number of new medications for the rest of your life. These immunosuppressant (anti-rejection) drugs play an essential role in the transplant process, because they prevent your body's immune system from rejecting the new transplanted organ. In addition to immunosuppressant drugs, most patients will also have to take steroids.

The transplant team will show you how to take your new medications and, for lung transplants, you will also be asked to complete a daily diary, which includes recording your temperature, lung function and weight.

Your team will talk to you (a great deal!) about adherence; this is because it's crucial to stick to the treatment plan post-transplant. With your previous CF medications you may have had some leeway on frequency, timings and missed doses. Over the years, you and your team will have worked in partnership to develop a treatment plan that delivered for you clinically, but which could also fit into your way of life. This is not the case with transplant medications. There is no flexibility in the treatment regimen.



It is essential that the medications are taken as prescribed, because not doing so will damage your transplanted organ and it's not an exaggeration to say that this could be life threatening. Your transplant team cannot fix lungs or other organs that have been damaged. Second transplants are possible, but very rare because you have to meet the same health criteria as in a first assessment. If your lungs are damaged due to non-adherence, your transplant team may not consider you suitable for a second transplant.



Rehab

When you are ready you will be moved from the ICU to a ward, where you will stay for an average of four to six weeks. During that time you will exercise regularly, both with the help of physiotherapists and independently; this helps to ensure that your new lungs start working as well as they can. Recovery is very much an active and cooperative process, and you'll be expected to fully participate in your rehabilitation, to help your own recovery.

"It was strange to walk around for the first time, with all the machines attached and a weird sensation in my lungs, almost like they were moving around. I was scared to death when they said they were going to take my oxygen off, I thought my sats would plummet, oxygen had been my security blanket for so long."

Helen, 11 years post-lung and liver transplant

"I remember exercising in a chair in hospital and I felt that I was getting out of breath so I stopped. My brother encouraged me to keep going and I realised that people with healthy lungs can get out of breath and keep going. My experience throughout my life was that getting out of breath was bad news, so it was quite difficult to adapt to a new reality."

Oli, 10 years post-lung transplant

"The ward was a bit like boot camp!"

Helen, 11 years post-lung and liver transplant

"I flew through my recovery, I was back on my feet quickly and had an almost instant improvement in my quality of life. I know it's not the same for everyone and my experience probably isn't typical."

Vicky, 18 years post-lung transplant

Although people with CF are often used to hospital stays, it can still be boring and frustrating being in hospital for a lengthy period of time particularly if you are far away from family and friends.

"I did get a bit sick of the food during my month in hospital. I experimented with the menu though, trying the halal and Afro-Caribbean options. It's worth doing, sometimes the other options have bigger portion sizes or are just tastier!"

Mark, 12 years post-lung transplant

After you have been discharged from hospital, you will need to return to the transplant centre very frequently for close monitoring of your health, including checking for signs of infection or rejection. The frequency of the visits to the transplant team will reduce as you recover and your health improves, and some tests may be done at your local hospital. Your transplant team will continue to monitor your health throughout your life with routine, but less regular, appointments similar to your CF clinic visits.

"I vividly remember walking up a slight hill from our local shop while I was recovering and getting out of breath, but I didn't need to cough. It was a remarkable moment for me. That's how my recovery felt, not one 'wow' moment, but lots of smaller moments of new abilities or feelings."

Oli, 10 years post-lung transplant

"I felt at my peak at discharge, about one month after the operation. I could cycle and jog, things I hadn't done for years."

Mark, 12 years post-transplant

Some people with CF find the rehabilitation process challenging. They are having to learn about a whole new 'condition' and treatment regimen, and going from being an 'expert CF patient' to a 'novice transplant patient' can be frustrating, particularly if the recovery isn't straightforward.





Complications

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

"I felt like everything was going wrong: acute rejection, infection and all the difficulties with trying to get my lung health back."

Laura, five years post-liver, pancreas and small bowel transplant

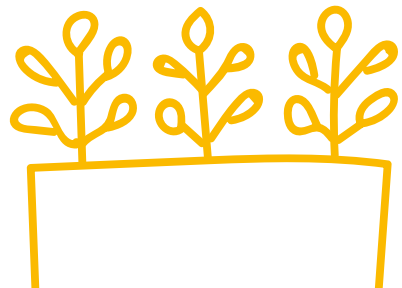
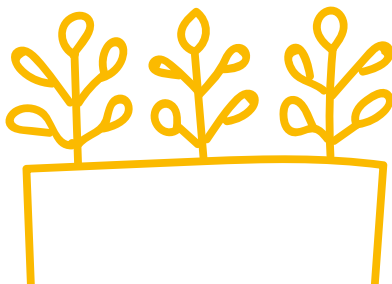
Acute rejection

Episodes of acute rejection are common post-transplant; symptoms can include raised temperature, flu-like symptoms, breathlessness and unexplained tiredness. Your team will be monitoring your new lungs for signs of rejection and will be supporting you in learning about identifying signs and monitoring your own health.

For people who've been through lung transplant, you may be asked to measure and record (in your diary as previously mentioned) your lung

function every day at home using 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 lung biopsy, which involves using a bronchoscope to get a small sample of your lung tissue. It is a very routine procedure with minimal risks attached that will be explained to you at the time.

Acute rejection is treated with steroids and altering the dose of immunosuppressants and is usually very effective.



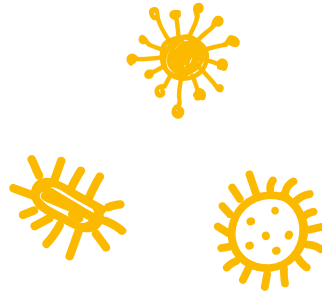
Chronic rejection

Chronic rejection, also called obliterative bronchiolitis (OB), usually develops over many months or years following transplant. In OB, the small airways become blocked with scar tissue, which can cause deterioration in lung function.

“The hardest part of the whole transplant experience was finding out that I have chronic rejection. When I found out I did feel that the whole thing had been a bit of a ‘con’. But the rejection is being managed, although it won’t be cured.”

Mark, 12 years post-lung transplant

There isn’t a cure for OB, but for some people a change in immunosuppressant can slow the damage. Your transplant team and the patient information booklet that lung transplant patients receive, can provide further information about chronic rejection.



Infection

The immunosuppressant medications that protect your new organ from rejection also make you more susceptible to infection, particularly in the early months following transplant. Repeated infections, as you know with your

CF, can cause irreparable damage, so it’s important to look out for any signs of infection and seek treatment promptly. Your team and patient information booklet (for lung transplant patients) will give more detail about the signs to look out for.

Diabetes

For people with cystic fibrosis-related diabetes (CFRD), transplant can upset glucose levels and changes may need to be made to treatment regimens. Steroids and other medications required post-

transplant can cause diabetes - sometimes this is temporary, other times permanent. You will be closely monitored for any changes in your blood sugars.

Other possible complications

Transplant can also increase the risk of damage or complications in other parts of your body. For example, the immunosuppressant medications that are essential to prevent rejection can cause damage to your kidneys. Your team will monitor your kidney function to look out for any signs of complication. These medications can also be linked to an increased risk of certain cancers, particularly cancers of the lymph nodes and skin.

Long-term steroid use can also be a cause of osteoporosis (lower-bone density), which causes increased susceptibility to fracture. Bone health can already be an issue for some people with cystic fibrosis.

Bone-protection therapy may be prescribed; you may already be taking medications to help with this.


It's important to remember that while these complications can be serious, not taking the prescribed medications post-transplant can cause rejection of the new organs leading to irreparable damage and death.

Do speak to your transplant team for more information about any of these complications.

Second transplants

Second transplants, although very rare, are possible. Assessment for second transplants follows the same format as the first and requires the same levels of health and fitness as the first time round. People who have had second transplants say that the physical and psychological recovery is even harder than for their first transplant.



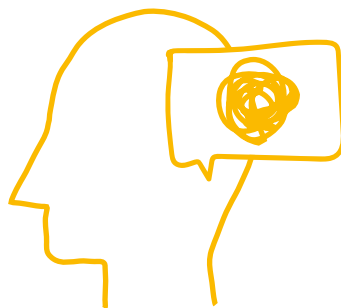
A photograph of a man and two children walking through a stone archway. The man is on the right, crouching and smiling, wearing a dark jacket and blue jeans. A young child in a red jacket and blue pants is in the center, walking away from the camera. Another child is partially visible behind them. The background shows a residential street with houses and trees under a clear sky.

"I remember thinking that I'd be grateful every day for my new life and feel this sense of euphoria at being alive. But actually there was a bit of a come down after about six months when the reality of normal life kicked in - the usual things, Monday mornings going to work ... in the rain. I try to keep perspective and appreciate every day, but I'm human, you can still get cheesed off at little things after your transplant."

Paul, six years post-lung transplant

Psychological impact

Given what people go through dealing with end-stage CF and then the transplant process, it's perhaps not surprising that many do experience psychological challenges while progressing through the recovery period and beyond.



Adapting to a new reality

Transplant is life-altering and this can be stressful, even when quality of life has actually improved. Some people may find that transplant doesn't meet all their expectations and may feel a sense of disappointment with the outcome.

The change in your quality and experience of life can take some getting used to; it's a 'new normal'.

"I felt really anxious after my transplant. I'd been dependent on oxygen and a wheelchair for so long that it took some time to trust my new lungs. My physical recovery was also quite slow, so I didn't feel the impact of my new lungs for some time."

Tor, six years post-lung transplant

"I felt vulnerable, it was all new to me and I didn't know what to do next – I asked myself, what is this new life all about? I felt a bit lost."

Helen, 11 years post-lung and liver transplant

"I'd been surviving for so long, I didn't know how to live."

Aoife, four years post-liver transplant

"I found it hard to go out alone after transplant, I'd been so used to being with my partner or my mum. I remember being on an escalator at the mall and just panicking; I was alone and crying."

Veronica, two years post-lung transplant

"It's hard getting to know your new body. Before my transplant, I was really good at working with my body and understanding what was going on. All of a sudden, you don't know anything about your body, you don't know what's an issue and what's not. It feels a bit scary."

Oli, 10 years post-lung transplant

Others are surprised by how quickly the reality of normal life kicks in.

Psychological challenges

It is not unusual for people to experience psychological challenges, including depression and post-traumatic stress disorder (PTSD), following transplant. Sometimes this can stem from trauma relating to experiences of end-stage CF or from the transplant recovery; medications can also lower mood; survivor guilt, or a pressure to be happy and inspiring, can also contribute to emotional difficulties.

For some, the idea that their transplant was a 'gift of life' can be laden with guilt, leading to feelings of immense pressure to make the most of this gift and sometimes feelings of not being worthy of it.

For others, the idea of the gift can be a motivating thought, and help them to focus on making the most of a second chance at life. It's important to try to keep in mind that the only hope or expectation associated with transplant is that you'll be able to enjoy a better quality of life. It's your life, not anyone else's.

"You've got to find the life that suits you – whatever that is. For some people after transplant they'll go off and climb the Himalayas, for others they'll just want to sit and watch a box set on telly."

Paul, six years post-lung transplant

"I celebrate my lungiversary and my donor is an intrinsic part of that celebration. I feel connected to my donor and positive about the donation and I do now consider my lungs to be my own."

Veronica, two years post-lung transplant

However, adopting a positive mindset is very often easier said than done, and if you are experiencing difficulties, seek support and help from your transplant team, or from your GP. You can also contact our helpline on **0300 373 1000** (Monday-Friday, 9am-5pm) or see the back of this booklet for additional sources of support.

"I started to feel amazing several weeks post-transplant; I had a new lease of life, but I was also confused and felt under pressure to 'live life to the full' to do justice to this amazing gift. I suffered with PTSD relating to my experience of end-stage CF and have also been depressed due to the medications I'm on and everything I've been through. Talking and time has helped; you need to create your 'new normal'. I've tried to take it a step at a time and set small goals. I realise now that to give credit to my donor means living my life as I want to and for me that's normal things like singing in the car, walking my dog and getting a job."

Veronica, two years post-lung transplant

"I experienced post-operative neuropsychiatric symptoms after my transplant. It was really frightening, I didn't know what was happening to me. It did subside, but I struggled afterwards and was subsequently diagnosed with PTSD. I've had a lot of support with that with a psychologist and using Cognitive Behavioural Therapy."

Aoife, four years post-liver transplant

Body image

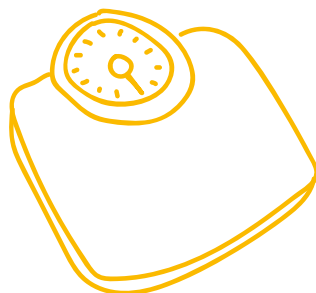
Transplant can also impact on your body image; the operation results in scarring, whilst the steroids cause increased appetite, as well as possible facial swelling. Some immuno-suppressants are known to increase hair growth all over the body, and this can be particularly difficult for women to cope with.

“I’ve put on weight since the transplant, it’s partly because the steroids increase your appetite, but also because they cause swelling. A lot of my old clothes don’t fit me anymore and I feel that I look like a completely different person.”

Veronica, two years post-lung transplant

However, transplant can sometimes have a positive impact on body image, particularly with liver transplant, where abdominal swelling can be reduced following successful transplant.

If you are struggling with body image, you might find it helpful to speak to your transplant psychologist or request a referral from your GP.



Changing roles in families and relationships

It’s different for everyone, but some people who’ve been through transplant have said that this major life event impacted on everyone close to them and changed the dynamic of some relationships. Caring roles can change following successful transplant, and while this is a positive development, it can take some time for relationships to readjust, where one partner was previously perceived as the carer and the other ‘cared for’.

Transplant can also have financial implications too; this can be positive, enabling someone to work, or it may be more challenging, with benefits stopped because of the operation.

The Cystic Fibrosis Trust has a welfare and rights adviser, who can assist you with financial concerns; please contact our helpline for more information, on **0300 373 1000** or via **helpline@cysticfibrosis.org.uk**

“For some people who have a successful transplant, it can be challenging to develop confidence and independence, particularly for those whose illness has meant they have been dependent on others for a long time pre-transplant. This may also be a challenge for those close to them as roles and relationships change.”

CF Psychologist



Life post-transplant





Life post-transplant

When the intensive care and recovery is over, the well-wishing has stopped and normal life, albeit a 'new normal', begins – what next?

For the first few months, it can feel like treatment and anxiety, about what to worry about and what not to worry about, dominate your life. Your transplant team are still there to support you, so do pick up the phone to discuss your concerns.

“I remember feeling worried about every little thing that happened in my body. I'd have an itch, for example, and be worried that it might be 'a thing'. I stayed in touch with my transplant coordinator and would phone with these worries. That support was invaluable.”

Aoife, four years post-transplant

Over time, it's likely that although transplant care will continue to take up a section of your life, it will, as with CF, often become a part of your life, rather than the focus of your life. Clearly, this is not always the case; not all transplants are completely successful, some people never fully recover from transplant and very sadly, a proportion won't survive beyond the first few years.

Although it's not always easy to adapt, and many people who've been through transplant have had to make significant compromises, many also say that they've benefited in their quality of life.

“It was hard but I'd say around 10 months after transplant was when it all starting coming together and I really felt it had all been worth it. I went travelling after my transplant and then returned to London to work full time. I make sensible compromises around my transplant care and have moved on from those early days of feeling vulnerable.”

Aoife, four years post-liver transplant



“Transplant has given me everything that I wanted, but it’s not an easy ride or a cure. I feel great most days, rotten on others, but overall, I’m really proud of what I’ve achieved over the past six years, including becoming a dad.”

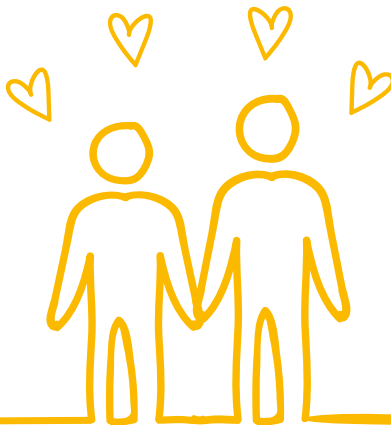
Paul, six years post-lung transplant

“I got involved with the British Transplant Games, an annual sports tournament for people who’ve been through transplant that celebrates life and promotes organ donation. I even met my wife there, three years after my transplant.”

Richard, seven years post-lung transplant

“I had to give up my job as a teacher because of CF and I’d always assumed I’d go back, but it’s not that easy. I’m concerned about absence, stress, infection, but I also miss my career and want to go back. I also had to come to terms with not having a baby, the risk to my health was too great. So at times, I’ve felt that the transplant has given me life, but taken away my opportunities.”

Helen, 11 years post-lung and liver transplant



Having a family

There is limited research available, but the evidence indicates that pregnancy following transplant is considered high risk, particularly if the pregnancy is unplanned or if the pregnancy is within two years of transplant. Many doctors will strongly advise against pregnancy altogether after lung transplant and suggest that couples consider other options, such as surrogacy.

If you do decide to pursue pregnancy following transplant despite the risks, it's very important that you inform your transplant team. Some of the transplant medications are dangerous to the foetus and must be altered if you are to have any chance of carrying a healthy baby.

"I didn't know that transplant would seriously affect my hopes of having a baby. I only found out at assessment and it took me a while to get my head around that."

Veronica, two years post-lung transplant

"I found it hard to come to terms with the fact that I wouldn't have a family and I wish this had been discussed in advance. I know the risks are just too great and I wouldn't want to jeopardise my lungs and my future."

Helen, 11 years post-lung and liver transplant

"Some women don't use contraception because they don't think they can get pregnant, because of their CF and transplant. But pregnancy is possible and can be dangerous post-transplant. We're careful to make sure our patients are aware of this."

CF Nurse

Do talk to your transplant team if you would like more information about the risks involved in pregnancy following transplant.



Restrictions

In the early period of recovery, you'll be advised to stay away from crowded areas, public transport or anyone who is unwell, to limit your exposure to infection risk. There are some other restrictions that your transplant team will give you more detailed information about, but the key areas of concern are:

- ensuring good food hygiene and preparation
- avoiding unpasteurised foods (such as some cheeses), raw fish (such as sushi or oysters), raw eggs and undercooked food (particularly undercooked meat, fish or eggs)

- keeping pets healthy and clean, and taking care when cleaning up any poo (litter trays etc)
- not smoking or being in smoky environments

For many people with CF, these recommendations may not feel especially restrictive, given their previous life-long experience of infection control.



Drugs and alcohol

People with a recent history of drug or alcohol misuse are generally not considered suitable candidates for a transplant. Misuse of drugs or alcohol following transplant can cause potentially life-threatening health problems. This is because drug use or excessive alcohol intake can affect the absorption of the immunosuppressant drugs that are critical in preventing the rejection of the new organ. If you are using drugs and/or alcohol, your body may not be absorbing the amount of immunosuppressant medication required to protect your transplanted organ.

Your team will explain the risks of excessive alcohol use or drug use to you. Their concern is with your health, getting support for you, and ensuring that your transplanted organ is safe and healthy, and so they will send a strong message on drugs and alcohol.

If you are drinking excessively or using drugs, it's really important to be honest about this with your team.

Finally

Everyone going through the transplant process will have a different experience – there is no set journey. We hope that this booklet gives you a useful overview of the process, highlighting some experiences and challenges that you might be able to relate to.

“It’s not for anyone else to tell others what to do. My personal experience has been life-changing; I’ve done things I’d never imagined doing, including meeting nieces and nephews. I’ve had all the up-sides of transplant. But I know others who’ve experienced many more challenges, and sadly have had friends who’ve died only months after transplant.”

Oli, 10 years post-lung transplant

Further information and support

The trained staff at the Cystic Fibrosis Trust helpline provide an information service and a listening ear, as well as further details about available grants, including a transplant grant. You can contact the helpline on **0300 373 1000** (Monday-Friday, 9am-5pm) or at **helpline@cysticfibrosis.org.uk**

■ Transplant-specific support

Details about support groups specifically for people going through liver transplant:

www.pscsupport.org.uk/support-groups-transplant-units

Harefield Hamsters provide support to people who have had, or are waiting to have, an organ transplant: **www.harefieldhamsters.org**

■ General psychological support

Butler and Hope (2007) ‘Manage your mind’ is published by OUP Oxford

Northumberland NHS Trust have developed a range of ‘self-help’ leaflets: **www.web.ntw.nhs.uk/selfhelp**

■ Post-transplant activities

Transplant Sport is the UK’s largest charity promoting active recovery for transplant recipients and increasing awareness of the benefits of organ donation: **www.transplantsport.org.uk**

Find us online

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