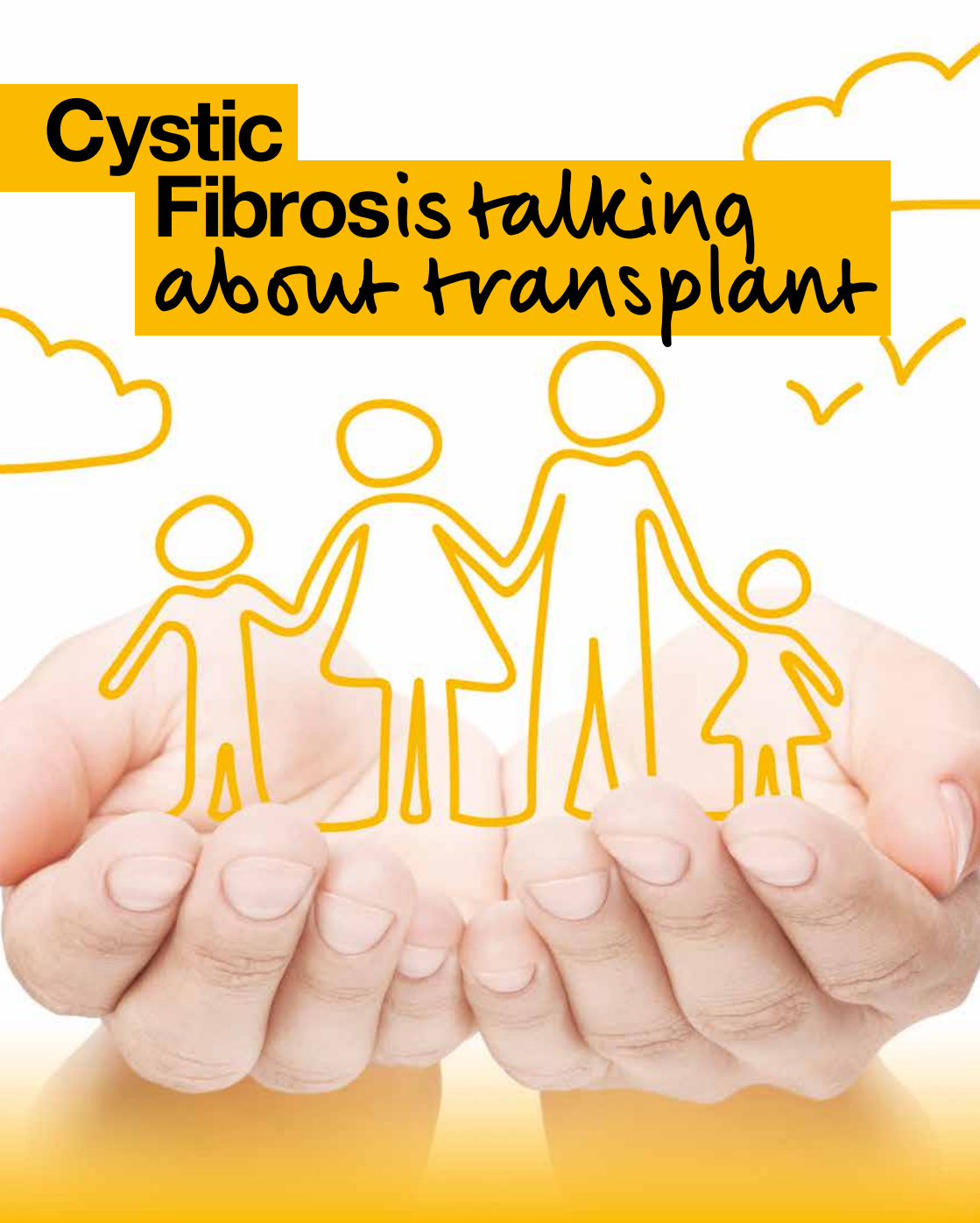
Cystic Fibrosis talking about transplant



Cystic fibrosis and lung transplant: an information booklet for parents

Fighting for a Life Unlimited

The Cystic Fibrosis Trust is very grateful to everyone who contributed to the development of this leaflet. The input and support from members of the paediatric transplant and paediatric cystic fibrosis teams was incredibly helpful. Sincere thanks also to our parent reviewer.

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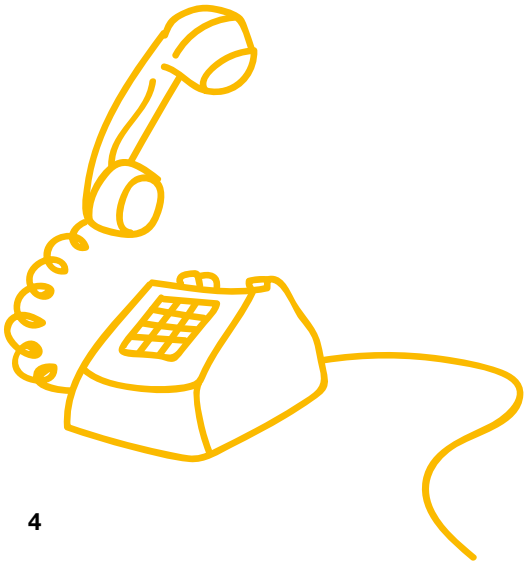
Introduction

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 at far too young an age. If you are reading this booklet, it's likely that your CF team has discussed the possibility of a lung transplant for your child. We understand that thinking about your child going through transplant must be very difficult.

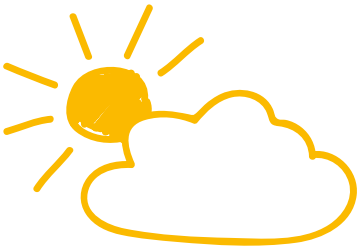
This booklet complements the information you will receive from your CF and transplant teams, and will help you and your family prepare for the process, as well as giving you details of where to go for further information and support. The information is intended to be clear and honest, and while some of the messages may be hard to hear it is not our intention to cause distress, only to give you the most accurate information possible.

The Cystic Fibrosis Trust helpline can provide a listening ear – if you want to talk, please call us on **0300 373 1000** (Monday-Friday, 9-5pm).



Lung transplant and CF

It is rare for a child with CF to require a lung transplant. A referral for assessment indicates that the child must be deteriorating and not responding to the usual array of CF treatments. Lung transplant is not an option that either the CF team or the transplant team take lightly.



Lung transplant is not a cure for CF – transplanted lungs will not have CF and will never develop the condition, but because CF is genetic your child will still have the condition in the rest of their body. This means that some CF treatments will still need to continue for life, even after a successful lung transplant.

The Cystic Fibrosis Trust works hard to encourage organ donation, but sadly there are still not enough lungs for everyone who is listed for a transplant. This is why your child, and all others on the list, has to wait, and why no-one can give a timescale for when lungs will be available. Transplant teams work hard to make sure that assessment is timed correctly to ensure that if lungs do become available, the child is prepared and ready to go.

Transplant assessment

Preparation

If your child's CF team feel that other treatment options have been exhausted, they will talk to you about the possibility of a lung transplant. It's at this point that you may have been given this booklet.

Your CF team will talk you through the transplant assessment. You'll receive detailed information about this process, including what tests will be done and how the team will decide if your child is suitable for a transplant. It's important to remember that this first appointment will only be for an assessment – your child is not going for a lung transplant or going on the transplant list at this point.

This may be a lot for you and your family to take in. We suggest that you take your time to read and think about the information, possibly writing down any questions you may have so you can discuss them with your CF team, or with the transplant team, when you attend your child's assessment.

How you talk to your child about their assessment will obviously depend on their age, personality and how you've discussed CF with them in the past.

You may find that having CF has made them quite used to hospitals and clinical tests, and that they take the conversation in their stride. Again, depending on age, it's likely that your child will have already been involved in conversations with the CF team about the assessment and what it means.

Do try to answer any questions your child has about the assessment honestly and clearly, and avoid making promises that you can't keep, for example, "you'll be getting new lungs" or "this operation would cure your CF".

You can always contact your CF team, particularly your CF psychologist, if you need more support with these conversations.

At assessment – parents

Your child's assessment will be at either Great Ormond Street Hospital (GOSH) in London, or the Freeman Hospital in Newcastle. These are currently the only two centres in the UK where paediatric lung transplants are carried out.

The assessment process can be very difficult for parents. The multidisciplinary transplant team understands this and will support you, often with input from the transplant psychologist, to explore coping mechanisms and identify specific concerns or worries. Social workers may also be involved during transplant assessment, helping to identify areas where additional support may be needed, such as with childcare or financial issues.

The transplant team do have to give you difficult information and ensure that you've understood all the risks and benefits of this procedure, and there is no easy way to deliver this. Life expectancy is a key concern for parents. It's important to understand that a lung transplant won't give a child with CF a 'normal' life expectancy. However, lung transplant can prolong life beyond what a child would have had without the transplant, and can give a better quality of life.

In terms of life expectancy post-transplant, GOSH reports 95% survival one year post-transplant. This percentage decreases over time, with approximately 50% of patients still alive 10 years after transplant.

However, statistics are only averages and are not personalised to your child. While it's understandable that you might want this information, it's important to discuss your child's own situation with your team and understand the limitations of life expectancy statistics.

At assessment – children

Children are sometimes frightened at this first meeting, particularly as the environment and team are new to them. Depending on their age, they may have questions for the team. We know that sometimes children, especially older children, want to protect their parents, and might like to ask questions without you being present. You may feel it would be beneficial to your child to give them this opportunity.

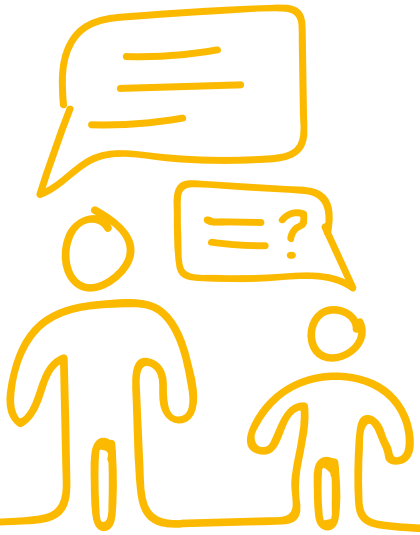
The specialists involved in assessment, including nurses, psychologists, doctors and social workers, are experienced in communicating with and supporting children of all ages. They will use different approaches according to the age and needs of your child.

Some teams use personalised stories to help younger children understand and prepare for the transplant procedure. These stories include pictures and photographs, and aim to introduce them gently to the wards and staff, as well as to some of the medical procedures that they may have.

Usually children get over their fearful feelings and become quite accepting of the idea of transplant, sometimes even feeling excitement at the prospect of getting new lungs. Some children start to plan wish lists of what they'll do post-transplant, which is helpful in terms of setting goals and developing a positive outlook. It is useful to ensure their expectations are realistic though – keep in your mind that they will continue to have CF, even with new lungs.

Some children will ask where the new lungs come from and this conversation can be challenging for parents. Your transplant team will be able to support you with these conversations, and it may be useful to have thought through how you might explain organ donation in a way that is comfortable for you and your family, in advance of any questions.

Some parents may experience feelings of guilt for the person who has died and whose organs are being used for transplant. While these feelings are understandable, you should try to remember that the individual wanted to donate their organs and that, in some cases, doing so provides some bereaved families with a feeling that there is a positive aspect to their otherwise sad and challenging situation.



Assessment process

Your child will undergo a variety of tests during the assessment process, including blood tests, lung function tests, exercise tests, CT scans and ECGs – these will be explained to you in detail by the transplant team. It's very likely that because of your child's routine CF care, they will already be very familiar with most of these tests.

The team will be measuring all aspects of your child's health to help them decide if your child needs a transplant and, if so, whether they are well enough to have it. There are some circumstances where a child may not be eligible for a transplant – if this is the case, the transplant team will explain the reasons for this decision. Infection with Burkholderia cenocepacia is considered to be a complete contraindication to transplant because of very poor outcomes after transplant. Other very resistant organisms, such as Mycobacterium abscessus, may be a contraindication to transplant, but this is decided on an individual basis.

Your CF or transplant team might suggest that you meet with the palliative care team at some point. This is often done early in the process, and you will be able to discuss controlling difficult symptoms, such as breathlessness, if your child is deteriorating. It should not be seen as a withdrawal of active care, as this often runs in parallel to waiting for a transplant.

We understand that some parents feel overwhelmed by the amount of information given at assessment – there's a lot to take in. If you don't feel fully informed, or if you have any questions at all, please ask any member of the transplant team. Remember, the transplant team are experts and work with families like yours day in, day out. They will help you with any queries or concerns that you and your child have.

Not being listed

It is quite common for children not to be listed for transplant at their first assessment. This is because the transplant team want to ensure that every child's life is prolonged by transplant rather than shortened. A child with CF must be poorly enough to warrant the risks of transplant, but not too poorly to go through the procedure. It's a fine balance, and the team would always prefer to ask your child to come for re-assessment later, even if that's only a few months down the line, to try and make sure that the timing is absolutely right.

Sometimes children won't be listed because they are not well enough. This can be incredibly difficult for families, and your transplant team, with input from the CF psychology team, will be there to support you and help you understand why this decision has been made.

The Cystic Fibrosis Trust helpline provides a listening ear; if you want to talk, please call us on **0300 373 1000**.

Life on the list

If your child is eligible for a transplant, you and your child will be asked to decide if you want them to go on the transplant list. There is no pressure for you to do so.

If your child is listed for a transplant, you may be advised to try to keep life as normal as possible. While it can be difficult to try to live life 'normally' under these circumstances, trying to stick to your usual routine and living your lives as fully as possible can help everyone in your family cope and enjoy life. If you have other children, they may need some support to understand what is happening to their sibling and how they can be involved or help. Try not to hold off doing things until 'after the transplant,' because unfortunately you don't know when that might be.

It's useful to be prepared for well-meaning people around you who may ask "When's the transplant happening?" or "Any update on the transplant yet?" and similar questions. It can be helpful to think about a 'stock answer' for these types of questions. Some people find social media can help them to share information without having to repeat themselves again and again, but this is very much a personal choice.

If your child is called for transplant it is important to be mindful of the donor family (who may not want to see things in the public domain at such a difficult time). If this happens, your transplant team will ask that you don't share information about a possible transplant on any social media forums that can be viewed by the public. It is, of course, fine to share the information with your close family.

Your transplant team will give you detailed information about how to prepare for the call. In short, it's all about being ready to go quickly and calmly, so having plans in place for childcare, pet care, work absence and any other commitments you may have, is really important. Think about if you get a call in the middle of the night, would you wake other siblings to say goodbye or just get your planned childcare in place and go? If the call came during the day, would you disrupt siblings at school to say goodbye, or not? Whatever you decide, just make sure everyone knows the plan, so that no one is upset, disappointed or confused by what's happening.

Transplant process

Arriving in hospital following the call for a possible lung transplant can be completely overwhelming. It's a good idea to get any questions out of the way before this time if you can.

The transplant team are, of course, very used to supporting families in your situation. Everyone will be very calm and the process will be very clear. The team will be focused on ensuring that your child understands and consents to the transplant. Even young children must be able to demonstrate that they know what's happening so that the team is confident that your child is 'informed'.

Because of the critical timing for transplant, patients are called to hospital before the donor lungs are visually checked by doctors. This means that there is always a chance that your child will be called to the hospital and sent home if the visual examination shows that the lungs are not suitable. This may be because the doctor sees signs of bleeding, infection or other damage. These 'false alarms' are unavoidable, but nonetheless difficult and disappointing for both children and parents. You may find it helpful to think about this in advance, so that you are able to be positive for your child in this circumstance.

If the lungs are suitable, your child will go to theatre for the operation. It usually takes around seven to eight hours, but this can vary. You might be interested to know that some children with CF want to see photos of their old lungs and the new ones – it can help them to understand 'the enemy they've been fighting'. Some parents also find that seeing the old lungs helps them to justify their decision to pursue transplant for their child. This is obviously a very personal decision.

Intensive Care Unit (ICU)

It's hard to prepare anyone for the ICU, but the transplant team will talk to you and your child about what will be happening, what the machines are, what they're doing and how it might feel. It is common for there to be a few problems and complications after transplant, and it can feel like two steps forwards and one step back at times. This is normal and the transplant team will keep you informed of your child's progress.

If the transplant operation has been straightforward, then recovery in the ICU tends to be fairly short, usually between one and two weeks. Longer-term ICU stays can be harder for children and their families to cope with. The ICU team will take great care to address any pain your child may feel, but will also ensure that their pain medication is managed correctly to minimise the risk of hallucinations, as these can sometimes occur after surgery.

After transplant

Immediate period

The immediate period after transplant is busy with rehabilitation, getting your child back on their feet and used to their new lungs, and a new treatment regimen. The anti-rejection medications that are essential to stop your child's body from rejecting the new lungs must be taken as prescribed. This is very important, as there isn't the leeway that you may have had with timings for their CF medications. The transplant team will ensure that you understand all the medications and how they need to be taken.

Families can sometimes find the treatment burden post-transplant overwhelming, particularly in the early stages where follow-up at the transplant centre is on a weekly basis. For those who live a distance from the transplant centre, the logistics of regular check-ups (as well as the cost) may be challenging. As previously mentioned, some CF treatment is likely to still be required, particularly if your child is pancreatic insufficient, and this is in addition to all the post-transplant medications. The burden will ease as the check-ups become less frequent and everyone gets used to the new regimen. Do speak with someone from your transplant team if you are struggling to cope.

Although your child will still attend their CF clinic for some appointments (usually only annual reviews), their care will shift predominantly to the transplant centre. This can sometimes be difficult for families, particularly where close relationships have existed with the CF centre. However, new relationships will soon form and the intensity of the transplant process means that you'll find you get to know the transplant team quite quickly.

Life post-transplant can take a bit of getting used to – it can be a positive but strange adjustment for families, particularly when children have been poorly with their CF all of their lives. Parents may have always had the role of carers, with their child unable to progress with their independence in the same way as their peers.
So this fairly quick and monumental shift can sometimes upset the family dynamic for a little while.

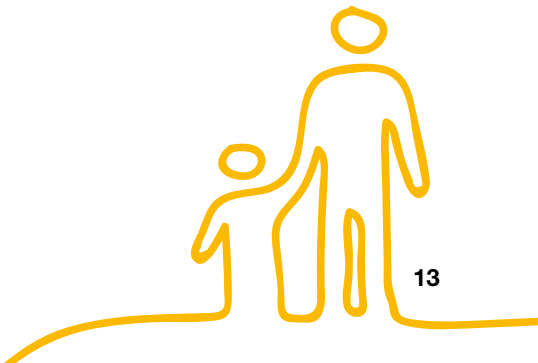
For some families there can also be a financial impact of transplant, through the loss of Disability Living Allowance (DLA). If you have concerns that this could affect you, please contact our Welfare and Rights Adviser through our helpline on **0300 373 1000** or at **helpline@cysticfibrosis.org.uk**. The Cystic Fibrosis Trust can also offer financial support through transplant grants. You can find more information about these grants at the back of this booklet.

Longer-term recovery

In the longer term, the transplant team, and later your GP, will be looking out for any signs of organ rejection, infection, renal dysfunction, diabetes or any other health issues associated with the transplant. Your transplant team will discuss these possible complications with you in detail when you go for assessment.

Depending on the age and sex of your child, you may need to think ahead and be aware that there can be implications for girls who've had transplants. There is limited evidence on the impact of pregnancy on women who've had transplants, but it's generally accepted that it is risky to a woman's health to become pregnant post-transplant, and many doctors will advise against it altogether.

If there are serious complications with the new lungs, it is possible for children to have second transplants. However, it very much depends on their overall health and whether they would be well enough for a second procedure.



In summary

You will receive a lot of information and support from both your CF team and the transplant team, and it may feel overwhelming. Take your time to read, digest and understand what transplant means for your child and your family. We would strongly encourage you to write down questions or concerns, both yours and your child's, and follow these up with your transplant or CF team.

If your child is listed for a transplant, do prepare for the call – both practically (childcare, work absence, etc), but also emotionally, including how you might explain a false alarm to your child and support them through that situation.

If you have support networks, family and friends for example, draw on them. They'll often be delighted to be able to be useful, even if that just means taking some of the burden away from you by helping with school runs or domestic tasks.

Finally, the multidisciplinary transplant team is there to support you and your child – draw on their expertise and knowledge to ask any questions or raise concerns.

More information and support

You can contact your child's CF centre if you have concerns about anything at all CF related, or if you are in the process of being referred or assessed.

You'll be allocated a transplant coordinator if your child is listed for transplant, and they are your first port of call for any concerns around transplant, even if you just need reassurance.

The Cystic Fibrosis Trust helpline provides a listening service. Our trained staff are here if you need to talk. You can call on **0300 373 1000** (Monday-Friday, 9am-5pm).

The Cystic Fibrosis Trust also offers small grants, specifically to support families who are going through the transplant process. You can apply for one grant at assessment, and a further grant can be applied for to support you through the transplant process. If you didn't apply for a grant and then find that you need financial support to help with transplant-related costs, such as those associated with regular follow-up clinic appointments post-discharge, please contact us to find out if we can support you.

Please contact our helpline on **0300 373 1000** or email **helpline@cysticfibrosis.org.uk** to find out more about our transplant grants.

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