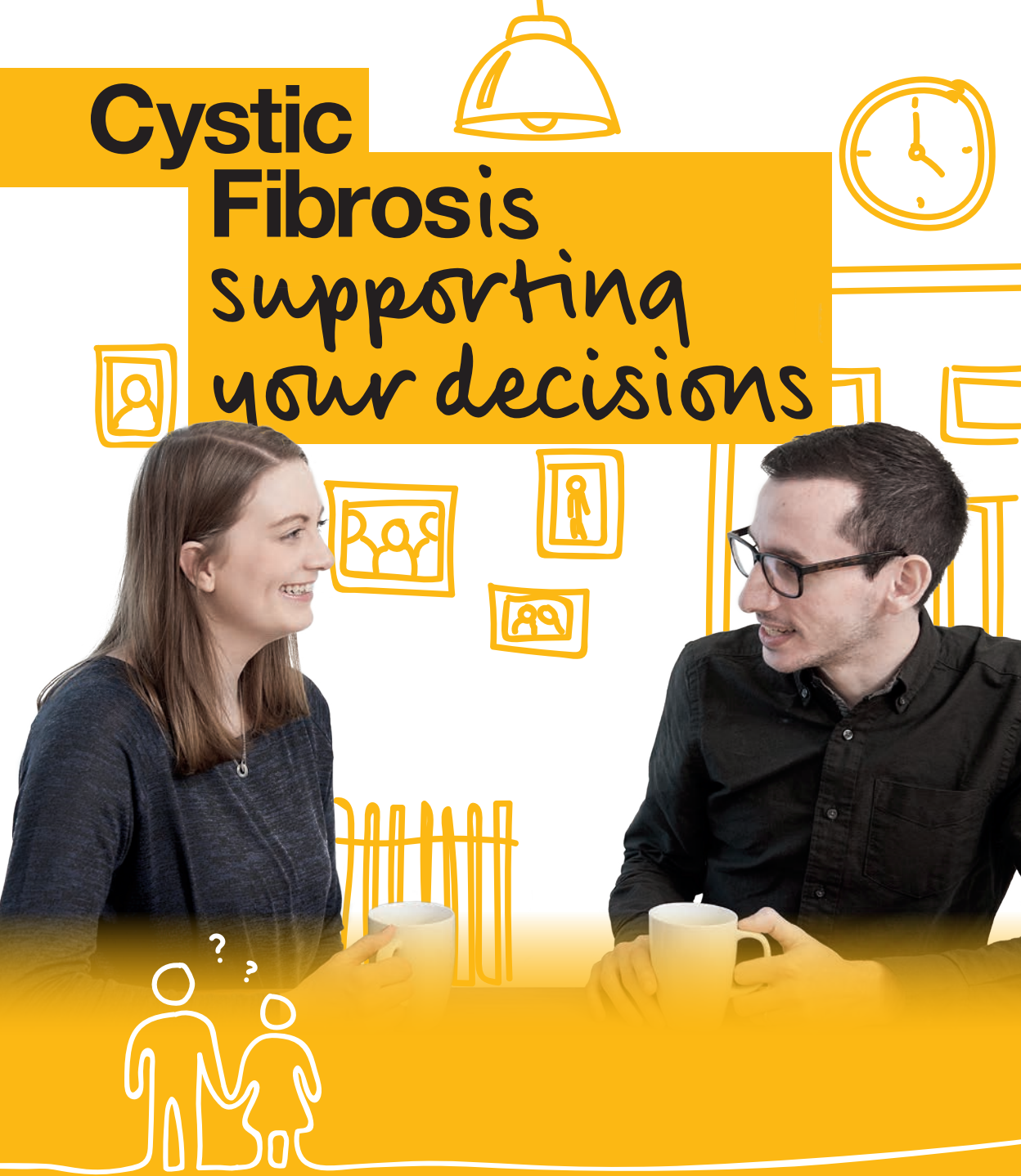
Cystic Fibrosis supporting your decisions



Thinking of starting a family?
A guide for adults with cystic fibrosis and their partners

Fighting for a Life Unlimited

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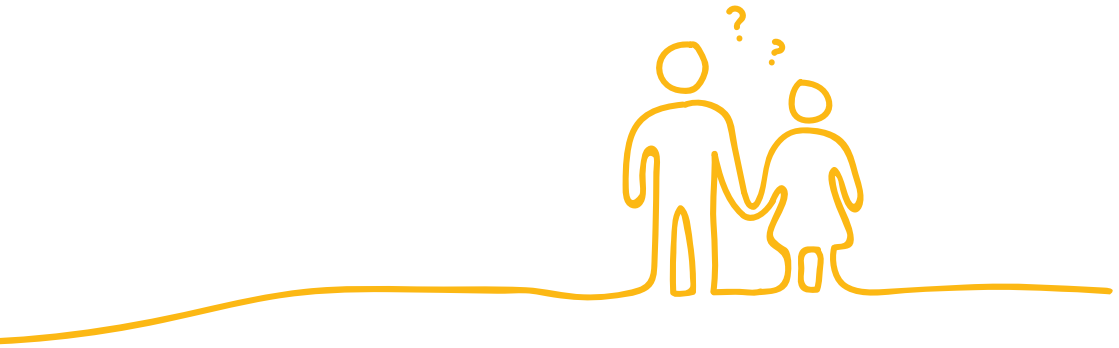


Introduction

If you have cystic fibrosis (CF), or have a partner with CF, and are thinking about having children, you might be grappling with some of the many issues covered here. We'd love to give you a factsheet with all the answers, nice and simple: job done. However, many of these important questions need tailored answers and will be very personal to you. This booklet contains information to support you in thinking through the issues in relation to you; your health, your life, your hopes and fears.

In developing this resource, the Cystic Fibrosis Trust has spoken with and listened to men and women with CF and their partners. Their stories are told alongside the facts to help put it all in context. You might find it helpful, for example, to hear from a mum with CF who has twins, or to read about what fertility treatment was like for one dad. The photographs in the booklet are all of real people who are sharing their stories with you. They have done this because they, and the Cystic Fibrosis Trust, feel that sharing these stories alongside clinical information will help people with CF and their partners make important life decisions that are right for them.

CF nurses, doctors, physiotherapists, psychologists, social workers and dietitians have worked with us to ensure the information provided is accurate and up to date. If you need any further information, please contact your CF team, or the Cystic Fibrosis Trust on **0300 373 1000** or at **helpline@cysticfibrosis.org.uk**

We are here to support you.

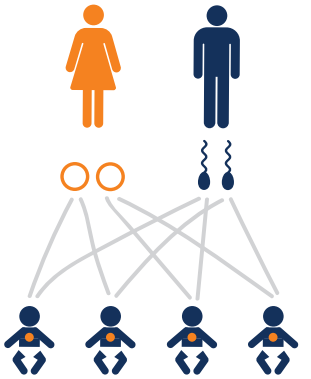
Emma and Geoff

Would our baby have CF?

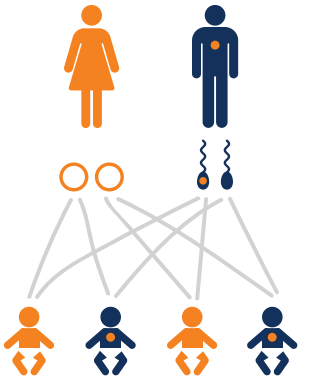
For some couples, asking this question might be what gets the ball rolling.

To be born with CF, the baby needs to have two copies of the faulty gene that causes cystic fibrosis. The baby will definitely receive one copy from the parent that has cystic fibrosis; whether they inherit a second copy depends on the carrier status of the other parent and the chance of them passing the gene on.

CF Parent - Non Carrier Parent



CF Parent - Carrier Parent



Please note: The CF parent in the diagram is female, of course the CF parent could also be male.

: with CF : CF carrier

To find out the likelihood of a baby having CF, the partner who doesn't have CF needs to be tested for CF carrier status using a simple blood test. There are over 2,000 known mutations of the CF gene, and the blood test cannot test for them all. This means that the test is not 100% accurate. If your partner receives a negative result for the CF gene, there is still a very small chance (approximately 1 in 500) that your baby will have cystic fibrosis.

If your partner is thought not to be a carrier of the CF gene it will no doubt be reassuring. However, you should be aware that your baby will be a carrier of the CF gene. This only has implications as your son or daughter gets older and perhaps wants to start a family of their own. There are no health problems associated with being a carrier of one copy of the CF gene. As the diagram above shows, if your partner is found to be a carrier of the CF gene, there is a 50% chance that your baby would be born with cystic fibrosis.

Some couples feel very strongly that if there was a high chance their baby would have CF, they may either choose not to have biological children, or to look at other options, including pre-implantation genetic diagnosis (PGD), which tests the embryo to see if it has CF (see next page for further information). Other couples have said the outcome of carrier status testing would not affect their decision to start a family.

To find out about getting tested for carrier status, you should contact your CF centre or GP in the first instance. Waiting times for carrier testing vary up to a few months, so it's advisable to start the process as soon as you can.

Some commercial companies offer home genetic testing kits. These kits are less reliable than clinical tests, as they search for fewer of the genetic mutations that can cause cystic fibrosis. They also do not come with the genetic counselling that is available in a clinical setting.

"If Tim had tested positive for the CF gene, we'd have opted for pre-implantation genetic diagnosis because CF has caused me so much suffering, I wouldn't want to put my child through that."

Jess

"Before my wife, Amy, became pregnant we talked about what it would mean if she was found to be a carrier of the CF gene. We decided that we would still want to have a baby if she was a carrier. I was fairly at ease with the idea that our baby could have CF. Amy was found not to be a carrier and fell pregnant following successful IVF. When Oscar arrived my feelings changed and I had a really strong fear that he would have CF – this caused me huge anxiety which I hadn't anticipated. I was desperately worried even though I knew the chances were really slim."

Michael

"Joe and I decided that we wanted to find out about his carrier status even though we weren't planning a family at that time. We thought it'd be better than finding out when we actually wanted to start a family. Joe was found to be a carrier and I was devastated. Years later when we were planning a family we decided, in the event of me falling pregnant, we would have chorionic villus sampling (CVS) to test the baby for cystic fibrosis. For us, a positive CF result would have resulted in me having a termination. It was an incredibly hard decision for us to come to and not one that we took lightly."

Martha

"My partner was tested for his carrier status and found not to be a CF carrier. Even if he had been found to be a carrier we still would have tried to have children. We both knew that any children we had would be carriers of the CF gene."

Tracy

"My husband has CF and so I always knew that if we were planning to have children there could be complications. I underwent carrier testing before we were even thinking about children, really just to get it out the way – tick it off the list. Chris, my husband, felt very strongly that if I was a carrier of the CF gene, we would need to re-consider our options because he didn't want to put a baby through CF."

Rachel

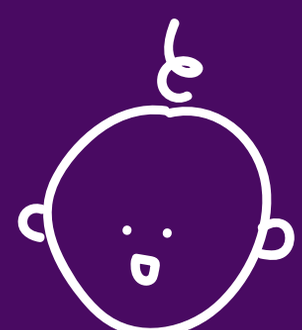
PGD is a way of screening embryos (fertilised eggs) for genetic diseases such as cystic fibrosis. This procedure is undertaken within an in-vitro fertilisation (IVF) cycle (see page 42 for more information) as the embryos need to be tested in a laboratory before being implanted in the uterus. Only unaffected embryos will be implanted. There are, of course, risks with this procedure, such as the embryo being damaged in the testing process. If you are considering PGD, speak with your CF team and GP to find out about funding and getting a referral.

"I was fully aware of the statistics about CF carriers but I never thought for a moment that Lee would be a carrier. I couldn't believe that we would be that unlucky. I was really shocked when we found out that he was a carrier of the CF gene. We talked a lot about our options. It was really hard, there were moral dilemmas that Lee and I had to work through. We decided that we would use PGD to have a baby without CF. That was our decision and the best one for us. I wouldn't judge other people's family planning decisions. It's complicated and very personal."

Katie

It's your decision

Emma and Geoff



Tracy, Mike, Baden and Jamie

Should we have a baby?

This is obviously a decision for you and your partner – although you may find that lots of other people have a view! It is **your** decision.

There are different considerations and concerns depending on whether you are a man or a woman with CF or a partner of someone with cystic fibrosis.

For women with CF, having a baby can pose a more direct risk to your health than for a man. Because of this possible threat to health, women with CF have told us that sometimes their families can be very worried about them getting pregnant, particularly their parents and partners. You may find that family and friends have strong views about whether or not you should have a baby and this can cause tension and distress. Talking over these worries, explaining the medical advice and showing that you have thought through plans for child care and maintaining your own health may alleviate some concerns.

Men and women with CF know that, despite great progress in cystic fibrosis treatment, the condition is life-shortening. This can raise important and difficult questions for the couple as well as their family and friends. Thinking about the possibility that a partner could die leaving the other with a child/children and a child without a parent is obviously very difficult, but it's a conversation that many couples affected by CF have.

"I get upset by people who judge whether or not we should have children because of my cystic fibrosis. It's not a decision that we, and I'm sure others with CF, take lightly. It was a huge decision for David and I and we spoke at great lengths about the 'what-ifs'. I know I could die young and leave David with the children but equally lots of things can happen in life – I could get hit by a bus! We're hopeful but prepared and we don't want to live in regret."

Clair

"I know I could die younger if I have a baby. Tim knows this too and recognises that he'd be left to care for the child. But we focus on the positive and I really believe that even if my health deteriorated, I'd be glad that I'd had a baby."

Jess

Tracy

Advice for women

"My CF centre have been really supportive and have worked with me to get me well enough during my 'window of opportunity'."

Emma

I'm a woman with CF, can I have a baby?

Your health

Many women with CF have babies and they all have different stories and experiences. Some will fall pregnant very quickly, others require fertility treatment; some are very ill during pregnancy, while others report improved lung function and a feeling of well being; some never manage to conceive. There is no standard experience and there are no definites.

In planning a family, it's important to link in with your CF team at the very outset. They will be able to advise you on what a pregnancy could mean for your health. They will generally advise what lung function and weight they recommend for you to support a pregnancy. Some research suggests that the stability of lung function is also an important consideration; for example, does an exacerbation usually cause your lung function to plummet? Perhaps you have lower lung function, but it is very stable? As well as optimising your health for pregnancy, your CF team will also review all your CF medications to make sure they are safe for the baby.

"My CF centre told me that they'd only recommend a pregnancy if I could get my lung function above 50%. This has been a great incentive to get well. I used to miss medications, didn't do my exercises or physio, drank alcohol regularly and was actually borderline anorexic. Then I met Tim and we started talking about having a family. I've stopped drinking, put on weight and am trying really hard to stick to my treatment regime. He wants me to live longer and be healthy so we can have a baby, and so do I."

Jess

It's not just about your physical health. Having people in your life who can support you through pregnancy and motherhood can be incredibly helpful both practically and emotionally. Being able to call on family and friends for help if you are feeling poorly gives you time to recover and look after yourself and your baby.

Working with your CF team in planning a pregnancy from the outset is the best way to ensure the best health for you and your baby. However, life can be surprising and many women with CF do have unplanned pregnancies. In this event, the key thing is to speak to your CF team as soon as possible.

There are some circumstances where a CF team may feel that a woman is too poorly to safely become pregnant; for example women with poor lung function, respiratory failure, pulmonary hypertension or heart disease. Women with severe liver disease or the bacteria Burkholderia cenocepacia will also be strongly advised against pregnancy. For some women and their partners this can, understandably, be extremely difficult to hear and some women feel a sense of grief at losing the hope of having a child. Your CF team may be able to help, or to direct you to other sources of psychological or emotional support.

"I understand the risks of being pregnant with CF and I've heard of girls who've had babies and died when their baby was young. But I've always wanted to be a mum. I'm an auntie to 11 nieces and nephews, but it's not the same as having your own baby. I want to experience that."

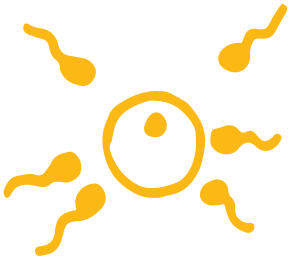
Jess

Your fertility

Many women with CF have babies without undergoing any fertility treatment. If you are reading this and are not sure whether you want a family right now, it's best to use contraception until you've decided! As already mentioned, planned pregnancies give you and your CF team a chance to work on ensuring your best possible health.

However, it is thought that women with CF are more likely to experience fertility problems than women who don't have CF, due to a couple of factors. Firstly, women with CF are more likely to experience irregular periods or the absence of their period altogether, particularly if ill or very underweight. This means they may not ovulate (produce an egg for fertilisation) each month. Women with CF may also have thicker vaginal mucus, which can make it harder for sperm to reach the egg.

Fertility issues in women with CF are treated in the same way as they are for women without cystic fibrosis. There is a range of treatments available, including medications to boost egg production, procedures to introduce sperm directly to the womb (intrauterine insemination or IUI) or full IVF techniques. IVF is explained in more detail later in this booklet.



Pregnancy

As well as the obvious growth of 'the bump', a woman will experience many other significant changes to her body during pregnancy. As well as increased metabolism, the expectant mum's lungs and heart work differently to accommodate the pregnancy. For some women with CF, particularly those with a low or unstable lung function or who have poor nutrition or low body weight, these changes can cause significant and irreversible health complications.

For this reason, pregnant women with CF will be monitored much more regularly than women without cystic fibrosis.

This extra involvement can sometimes result in improved compliance with treatment and even reports of improved lung function. However, research does suggest that overall, pregnant women with CF have a greater number of hospital admissions, increased diagnoses of insulin-dependent diabetes and reduced lung function.

"I had CF-related diabetes before I fell pregnant. However, it got much worse with pregnancy and I went from needing insulin very rarely (usually around once a month) to now injecting insulin several times a day."

Harriet

The way that your lungs react to pregnancy varies a lot between individuals. Hormonal changes, and a growing baby making less space for lung expansion, can cause breathlessness for many women, including those without cystic fibrosis. For women with CF, breathlessness can be more of an issue particularly during any physical activity. It's important for women with CF to be active and maximise their fitness prior to and in the early stages of pregnancy. The aim is to maintain fitness as much as possible in later pregnancy. If breathlessness becomes a problem your CF physiotherapist may look at ways of increasing lung expansion using different devices and positions and will help you recognise what's normal and what needs treatment.

Continence issues are far more common in women with CF than other women, even before they have children. Working with your CF physiotherapist on continence management techniques, such as pelvic floor exercise, can help treat continence problems. This is particularly important before and after childbirth to ensure your muscles are in the best shape possible! Understandably, women can become reluctant to cough effectively if they are worried about continence and this can lead to deterioration in lung function.

Your CF physiotherapist is likely to see you frequently during pregnancy to ensure your exercises are effective as your body grows and changes.

Calorie requirements will vary during pregnancy, however it is generally thought that a pregnant woman will need around 200 extra calories each day in the last trimester of pregnancy. This amount will vary for women with CF according to her health and nutritional status. For some women with CF this may be hard to achieve and nutritional supplements or additional feeding via a naso-gastric or gastrostomy tube may be needed. Your CF dietitian will be able to advise on this and on any additional vitamins you may need.

"I was really large by 27 weeks because I was pregnant with twins. My little girl was pushing on my ribs and one evening as I coughed in an awkward position I felt terrible pain in my ribs. I was admitted to hospital and found to have broken ribs. I was told I'd need to stay in hospital until the babies were born and that they might have to be delivered early. I cried every time I coughed because it was so painful. The CF physio came every day and used a cough assist machine to help ease my pain. I don't think I would have managed without it.

By 30 weeks, I was exhausted with the pain, size of the bump and the pressure on my lungs. I was also on a feed for extra calories and I was worried about the impact of the morphine on the babies so I stopped pain relief. I was really struggling. My doctor suggested they deliver early and I knew this needed to happen but felt devastated because I felt that I was choosing to have premature babies. It was an incredibly hard pregnancy from 27 weeks through to delivery, but my beautiful Ben and Mia arrived safely."

Clair

Tracy and Mike

"I developed gestational diabetes while I was pregnant with the twins. Quite a while after they were born I was diagnosed with CF-related diabetes and this has affected my health."

Tracy

Pregnancy and transplant

It is relatively rare for a woman who's had a lung transplant to also become pregnant. This means there isn't a lot of research to draw on and what research there is inevitably is based on small numbers of women. We do know that pregnancy for women with CF who've had a lung transplant is high risk, for both the mother and the baby.

Small studies have shown that around half of these pregnancies result in the baby being born prematurely. Premature birth carries a number of risks for the baby, which vary according to how early a baby is born, including breathing difficulties, heart problems and developmental delay. For the mum, the main risk is rejection of the transplanted lungs.

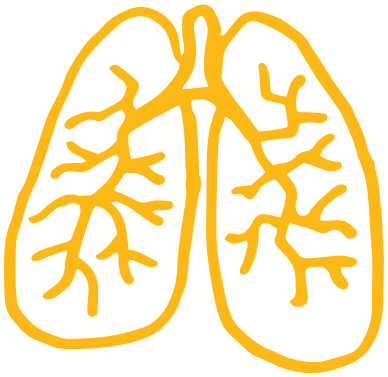
Some studies have found that 40% of women in this group died within three years of their baby being born.

The medical recommendation is that women who have had a lung transplant should wait for a minimum of two years before planning a pregnancy.

These are hard statistics and give only a picture of the 'average'. There will be women who've had a transplant and, subsequently, children, who don't suffer extreme ill effects, but this booklet needs to give you all the information to help you make your own choice. To understand what the risk could mean for you personally, speak to your transplant and CF teams.

"CF wasn't the main concern for me or my medical team, it was my transplanted lungs. The worry was that the pregnancy could result in my body rejecting the lungs. I was told that there was a 33% chance that I could die or suffer and a 33% chance that my baby would be adversely affected. These odds were worse than my husband and I had thought. But who knows what life can bring? So we decided to go ahead. My team had to alter my anti-rejection drugs because of concerns that they could harm the baby. Luckily this didn't cause health problems for me. We were delighted, after many years of trying, to welcome George into our family in 2014."

Harriet



Tracy, Mike, Baden and Jamie

Childbirth

As part of antenatal care, a birth plan is usually completed that encourages expectant mums to think about what type of birth she would prefer and her preferences for pain control. For many women with CF, the plan will be to have a normal vaginal delivery with the baby at full term. However, sometimes that plan will need to change to ensure the best outcome for both the mum and baby – and sometimes the baby may have its own plan! Research suggests that around 25% of babies born to women with CF are premature, either because they have been induced early (usually due to concerns about the mum's health) or have arrived early spontaneously. Ultimately, whatever plan keeps mum and baby safe and healthy is the best birth plan.

Managing pain in childbirth is important for all women but can have particular significance for women with cystic fibrosis. Pain and fear naturally can cause fast and shallow breathing (hyperventilation). For some women with CF, this type of breathing during labour can cause a reduction in the amount of oxygen getting into the bloodstream. Low oxygen levels will result in closer monitoring of the mum and baby and possibly the need for oxygen or a change of birthplan. Pain relief and breathing techniques can help with hyperventilation. Unmanaged pain can also make it harder to cough effectively during or between contractions, so some assistance may be needed from your CF physiotherapist.

An epidural is very effective in pain control and therefore can greatly reduce the stress on the heart and lungs that intense pain can bring and might therefore be suggested for some labouring women with cystic fibrosis. Your CF team may also have advice on what pain relief would be most appropriate for your own circumstances.

"I planned what I wanted for my C-section. I brought my own nightie and cut it up the back to make a gown because I really didn't want to wear a hospital one because they remind me of all the medical procedures I've had. I wanted to make this experience more personal. I had a CD which I wanted them to play during the delivery, it was music that I usually listen to and I really hoped the babies would hear it and feel familiar. The theatre was quite overwhelming because there were three teams present, one for each baby and one for my CF care but everyone was lovely and very supportive."

Clair

If a caesarean section is needed, it is likely that the team will prefer to use a spinal anaesthesia. However, this would always be decided according to the mum's own specific circumstances.

On rare occasions, a general anaesthetic may be used but there are concerns around this if the woman has severe respiratory disease and infection.

"I was booked in for a C-section at 36 weeks because the team were worried that I wouldn't be strong enough to push the babies out. I was anxious because the specialist had highlighted how difficult delivery could be for me so I was happy to go with the section. However, the boys had other plans and I went into labour at 33 weeks. I delivered my boys naturally with gas and air and had no complications."

Tracy

Tracy, Baden and Jamie

Being a mum

We think it's fair to say that, regardless of CF, very few women can prepare themselves for what it's really like being a mum. Mums and dads will experience a range of emotions and levels of tiredness! There are thousands of publications that give great detail on what those postnatal, early days can be like and these are as applicable to a mum with CF as any other mum. However, women with CF have an additional challenge: to look after themselves and to maintain their treatment regime to stay well.

"You almost have to think of the CF as being an additional child to look after and manage in your life."

Emma H

Mums, dads and CF professionals say that the mum's CF care routine will often take a back seat, with the baby's needs prioritised. This is absolutely understandable. It's difficult to find time and energy in a day occupied with a newborn's often frequent demands. However, it's a short-term strategy as the mum's health is important to the baby's long-term wellbeing.

"I have a really heavy treatment regime and my husband supports me full time. I don't think we would have chosen to have a baby without this full time support."

Emma H

Other mums with CF advise you to think through your plans carefully and make sure you have family or friends who are available to support you. Think about who will look after your baby if you are unwell or if you have a hospital appointment or admisson. How will you make time for your treatments? Do you have someone who can give you a break if you are tired? Can someone help with housework for a little while? If family and friends offer help, our advice is: take it! Mums with CF often say that the hard work begins when you get home with the baby. As the intense support from clinical teams that is so present during pregnancy decreases, and the stream of visitors wanes, some mums (even without CF) find the reality of motherhood surprisingly hard. Do speak to your health visitor, CF team or the Cystic Fibrosis Trust helpline if you need support.

"I've been really lucky and had 15 months' maternity leave and support from my sister who lives nearby. But even then, I just don't always find the time to do my treatments. It took me a year to get back to the gym after my little girl was born so it is hard to maintain the treatment routine."

Martha

"It's hard caring for the twins. They are great children and I'm so, so happy to have them, but it can be so exhausting! I know most new mums will feel this but I think having CF adds to the challenge. My husband is a policeman and works shifts and my mum works too so I have to manage on my own a lot. That's been really hard when I've been ill and had to organise childcare to fit in hospital visits. It's difficult being on IVs and feeling really poorly and trying to look after the children. I try to hold off illness and battle on but when I was really poorly earlier this year I was literally having to peel myself off the sofa; it turned out I had a chest infection and flu and had to be admitted. Mum took a month off work to help me which was amazing. My kids really are great, I love them to bits and wouldn't change my decisions, it's just I didn't realise how hard caring for children (especially twins!) and having CF would be."

Clair

"My lung function decreased dramatically after Arran was born. I think this happened because I just didn't have the time to dedicate to caring for myself, I was really focused on looking after Arran. By the time he was 18 months old, I was really thin and not very well and that's when I had my 'eureka' moment. I knew I had to look after myself so that I could look after him."

Julie

Breastfeeding

Breastfeeding is good for your baby, but it can be very tiring. It is important to look after yourself, so you may have to find a compromise that suits both you and your baby.

Maintaining good nutrition and hydration is really important if you are breastfeeding; you are advised to drink an extra two litres of fluid and consume 500 additional calories each day. Speak to your CF dietitian about breastfeeding, ideally before the baby arrives.

"My babies were in special care because they were born prematurely. I expressed milk for them for the first few weeks while they were in hospital but I was really struggling to keep my weight up and my diabetes was becoming difficult to control. I was advised to stop breastfeeding."

Clair

"I still breastfeed my 22 month old daughter. I did have to have my antibiotics changed and also extra vitamin D but because I'm pancreatic sufficient, there were no dietary changes."

Martha

Many drugs are passed from you, into your breastmilk and onto your baby so it's important to tell your CF team if you are planning to breastfeed. Try to do this before the baby arrives so the CF pharmacist has time to check the safety of your medications. Drugs that are safe during pregnancy are not necessarily safe for breastfeeding and vice versa.

Organisations providing support for women wishing to breastfeed are listed at the back of this booklet.

Studies have found that CF breast milk has the same levels of sodium as non-CF breastmilk and is safe for baby.

"It's hard to breastfeed twins, regardless of CF. I was finding it hard to reach the 3,000 calories a day I needed so to increase that further was not realistic, so my boys were bottle-fed."

Tracy

"I never thought being a dad would happen. Growing up as a young teenager and reading into CF gave me the impression that this could never happen. Fast forward to 2013 and our fertility journey to Oscar being born began and what a journey it has been."

Michael

Michael and Oscar

Advice for men

I'm a man with CF - can I be a dad?

Your fertility

Most men with CF (around 98%) will not be able to father a child biologically without assistance from fertility specialists. This is because the tube which carries sperm from the testicles to the penis (called the vas deferens) is either missing or blocked. You may hear this referred to as congenital absence of the vas deferens (CAVD). There's nothing 'wrong' with the sperm, they just can't get to the semen. This is called obstructive azoospermia.

However, it is possible for men with this CF-related problem to father biological children through fertility treatment including microsurgical epididymal sperm aspiration (MESA).

MESA is commonly offered to men with CF and is the procedure that extracts the greatest number of sperm. However, unlike the other procedures it is usually carried out under general anaesthetic and involves a small incision being made in the scrotum from where a sample is extracted from the epididymis. This sample will be tested microscopically and, if present, the sperm can be extracted and either used straight away or if necessary, frozen. The incision to the scrotum is closed using dissolvable stiches and you may need to remain in hospital for several hours to be monitored before going home.

There are other similar procedures that you may hear about, which are described in the table below and usually occur under local anaesthetic.

Procedure	Description
Percutaneous Epididymal Sperm Aspiration (PESA)	A fine needle is placed into the epididymis to extract sperm via a syringe.
Testicular Epididymal Sperm Aspiration (TESA)	A needle attached to a syringe is inserted directly into the testicles to extract fluid.
Percutaneous biopsy of the testicles	Similar to TESA, but the needle is larger and is inserted into the testicles to collect testicular tissues from which larger amounts of sperm can be extracted.

To ensure the sperm sample is used efficiently, a procedure called intra-cytoplasmic sperm injection (ICSI) is usually offered. This differs from conventional IVF in that instead of fertilisation (the joining of the sperm and egg) taking place in a dish where many sperm are placed near an egg, a single sperm is selected and injected directly into an egg. The success rate for ICSI depends on many factors, including the age of the woman. As a very approximate guide, around a third of women age 35 or younger having IVF with ICSI will go onto have a baby. It's an average measure so difficult to apply to individual circumstances. There is some evidence to suggest that where there are no fertility problems with the woman and IVF is needed to overcome a male fertility problem, the likelihood of a pregnancy may be higher.

Although your partner may not have any fertility issues, she will still have to undergo the IVF process, which is described later in this booklet.

"I had TESA. It was uncomfortable and faintly embarrassing but I wouldn't call it painful and it's quickly over. There was a lot of bruising afterwards which looked bad but was hardly sore at all, just a bit uncomfortable for a few days. I'd been expecting it to be more painful afterwards than it was."

John

John and Joshua

Your health

For men with CF, becoming a dad will not have a **direct** impact on your cystic fibrosis. There are, however, health implications to think about. Firstly, it is highly likely that you will have to undergo fertility treatment and, as with any medical procedure, there is a small risk of infection, injury or bleeding.

Arguably, the more significant health impact comes from actually being a dad and incorporating your own treatment into a new and possibly quite disruptive life. In amongst the excitement and exhaustion of new family life, it's important that your treatment regime doesn't suffer. This has implications for both you as a dad with CF and for your partner who may find they need to carry more of the baby care responsibility to ensure you have the space to continue with your treatments.

"My wife and I are proud parents to a happy, bubbly, bouncy, smiley, adventurous 20-month-old son. We were supported so much by my CF team at the hospital before he was born and in the months after his birth. It's a difficult time to balance everything you need to do as new parents and keep as healthy as possible. It was a busy time trying to adapt to our new life and routines, and spend as much time as possible with my family. I had less time to exercise and my lung function definitely took a hit; I had to find time to fit in the exercise. I successfully applied to the Cystic Fibrosis Trust for a grant for a rowing machine, meaning I had an easy, indoor, efficient way of exercising. I'm really starting to feel the benefits of having it."

Steve

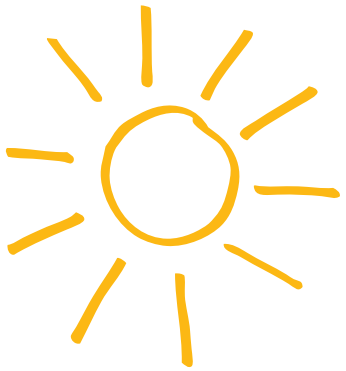
John, Ellen and Joshua

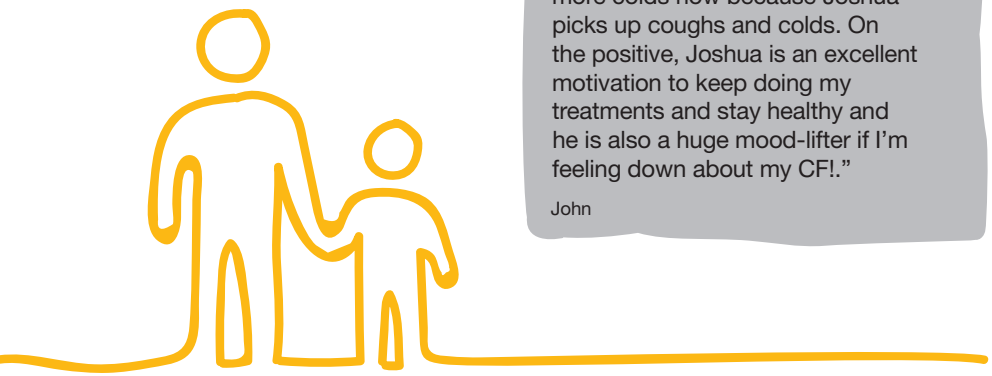
"I had been warned that having a baby could make John's health worse due to the extra demands and the viruses that babies and children pick up. I decided to try and do all the newborn nights to allow John to sleep as he's more likely to become ill if he's tired. I knew I would need to do most of the childcare if John was unwell and that was quite a daunting prospect."

Ellen

Being a dad

Many men with CF that we spoke to grew up thinking they could never be a dad. Some dads told us that having a child has given them an extra incentive to stay well, although they found it was tricky to keep to their treatment regime in the early days.





"Looking at him now, a three-month-old, healthy and I must say handsome little boy, is almost indescribable. He has changed my outlook on everything and put my health to the forefront of everything I do and think about. I need to be here to see him grow up and to enjoy my family for as long as I possibly can. 'My family' was not an option 15 years ago but now, coming home to a wife and baby is remarkable and I couldn't be happier or prouder that we took on the challenge of fertility treatment."

Michael

"Becoming a dad is a dream come true for me, but fatherhood is not without its challenges. Fitting in my treatments has become harder, but for me the main challenge is that I'm generally more tired and there's less chance to relax and recover if I'm feeling a bit unwell. It takes more planning to have downtime and if I'm resting then Ellen is looking after Joshua as well as doing everything else. Another challenge is that I get more colds now because Joshua picks up coughs and colds. On the positive, Joshua is an excellent motivation to keep doing my treatments and stay healthy and he is also a huge mood-lifter if I'm feeling down about my CF!."

John

John, Ellen and Joshua

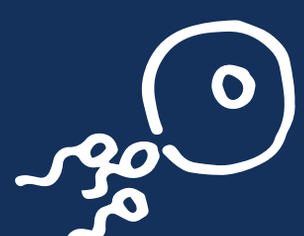
"It feels extra special to have been able to have a baby with John – he grew up thinking he wouldn't be able to have children, and he is so happy to be a dad! He takes a day off work each week to look after Joshua and he appreciates this father and son time so much!"

Ellen

"Staying healthy and sticking to my regime is now that much more important - I compromised a little bit more than usual in the first couple of months, leading to a course of IVs at home but now we are in the swing of things I think treatment will get back on track and I will probably take on more to ensure that I stay healthier for longer."

Michael

IVF and other options



"I've been going through the IVF process for more than a year now and recently completed my first cycle which, very sadly, didn't work. When the process started last year I suffered from hyper-stimulation syndrome which is when the body is producing too many eggs. Unfortunately it also can cause fluid on the lungs. I had to stop the treatment for a while until I was well enough to start again."

Emma

Fertility treatment and other options

IVF

IVF essentially means to attempt to fertilise an egg with sperm outside the human body, 'in glass' (in vitro). Britain was home to the world's first baby born through IVF in 1978. Since then, many thousands of couples have undergone IVF each year in the UK with varying rates of success. Key determinants in the likelihood of a successful pregnancy include the age of the woman and health behaviours (eg smoking, weight and alcohol consumption).

The IVF process may vary from clinic to clinic, but is likely to follow a similar schedule. The first step involves taking daily medications for around two weeks, either by injection or nasal spray, to suppress the natural menstrual cycle. Once the cycle is suppressed, daily injections of follicle stimulating hormone (FSH), which stimulates the ovaries to produce more eggs, are required.

It's important that your CF team is aware that you are undertaking IVF, as CF can have an impact on how well you absorb these medications and hormones.

The fertility team will monitor the development of the eggs using ultrasound scans and blood tests.

"I had to have my IVF drugs altered because of absorption issues due to CF. This meant I would produce fewer eggs."

Harriet

The eggs are collected using a fine needle which is passed through the vagina into the ovaries, guided by an ultrasound scan. The patient will be sedated for this short procedure. The harvested eggs will be fertilised by either mixing sperm with the eggs or through ICSI, which involves a single sperm being injected into the egg. This process is described fully on page 33.

"Suppressing my natural cycle gave me some pretty difficult side effects – poor memory, bloating, mood swings, dry skin and blurred vision – not nice!"

Emma

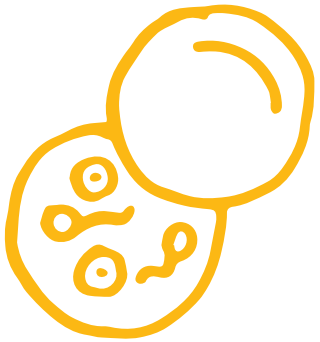
Once there are fertilised eggs (now called embryos), a hormone is given to prepare the uterus for the implantation of the embryo. It's possible that there will be more than one viable embryo and decisions will need to be made about how many to implant. Remaining embryos can be

frozen. The implantation procedure is quite straightforward and doesn't require the patient to be sedated; a catheter (small tube) containing the embryos is passed through the vagina into the uterus.

A pregnancy test can be taken around two weeks after the procedure. The likelihood of a pregnancy varies but for women under 35 around 33% of IVF treatments resulted in the birth of a baby. It's a very involved procedure and couples can find it extremely emotional and difficult, especially if it doesn't result in a pregnancy.

"I had five rounds of IVF. After the first round failed, I was devastated and even more determined to become a mum. It was incredibly hard to deal with successive rounds failing. I fell pregnant on the third round but unfortunately I suffered a miscarriage. On the fifth round of IVF I fell pregnant with twins. My husband and I were over the moon. My CF clinic was concerned though, they hadn't dealt with a woman with CF having twins so it was new territory. IVF can be a heart-breaking journey and one that can change you as a person but I didn't want life without a baby so it was all worth it for me."

Clair



"I had IVF after trying to conceive for six years, that was an emotional rollercoaster. We were successful with the IVF and found that we were expecting twins!"

Tracy

"In some ways IVF wasn't as bad as we expected – we just had to accept the many appointments, the injections, the invasive procedures, and got on with it. The hardest bit was knowing that the odds of success were quite low so we kept our expectations quite low. We were enormously lucky to be successful with our first cycle. We couldn't quite believe it."

John and Ellen

The IVF process can be physically and emotionally demanding, with multiple appointments, bureaucracy and a sense of living in limbo. It can also take a long time; some patients can wait up to two years from initial referral to implantation and this period can be longer if there are complications. Obviously, this can all be very stressful and frustrating. Undergoing fertility treatment can be a very personal experience, but if you feel comfortable talking to friends or family about what you're experiencing, having their support can be really valuable.

Funding and IVF

You may already be aware that funding for IVF is under pressure and that availability of funds can vary depending on where you live.

The decision-making on funding for fertility treatments in England falls to Clinical Commissioning Groups (CCGs). These groups are responsible for planning and financing health services for their local area. Although there is guidance on access to fertility treatment which has been set out by the National Institute for Health and Care Excellence (NICE), in practice this is just guidance and most CCGs set their own criteria based on this. Few CCGs offer the recommended three cycles of IVF, and some only fund one or two cycles or do not fund IVF at all. It's important to remember that the criteria apply to both of you, and you must both qualify for treatment, not just the person with cystic fibrosis. Your GP should be able to explain the criteria for access to IVF funding in your CCG area, or you can download the full fertility treatment policy from your CCG's website.

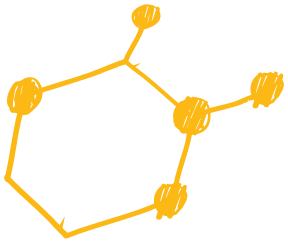
Currently in Wales there is a set of standardised criteria which should apply across all health board areas.

At present, if you meet the criteria, two cycles of IVF can be funded. In Scotland, there are also standardised criteria and two cycles on offer. In Northern Ireland, eligible couples can access one full IVF cycle and a frozen embryo transfer.

Waiting lists vary and are ever-changing, but it is currently fair to say that a minimum wait of a year should be expected, although it can be considerably more. It's important to check your national policy as it's subject to regular review and change.

If you have the resources, you can pay for IVF privately and this is likely to cost in the region of £5,000 or more per cycle. It's advisable to research your clinic carefully. The Human Fertilisation and Embryo Authority (HFEA) has a useful search page for NHS and private clinics.

www.hfea.gov.uk



This information was correct at time of printing. Please contact the Cystic Fibrosis Trust helpline on 0300 373 1000 or at helpline@cysticfibrosis.org.uk for the latest information

Adoption and surrogacy

For some people, adoption is their preference for creating a family, while others turn to adoption having not been able to conceive their own child.

The adoption process is very involved and does require a series of suitability checks, including a health assessment. This assessment will be organised by the adoption agency, which is either part of the local authority or a voluntary organisation. It is not possible to say whether a person with cystic fibrosis would be approved as an adoptive parent or not. As a first port of call, we recommend that you contact local adoption agencies and ask them about their criteria for the health assessment. You can find your local agency here:

www.adoptionuk.org

I knew from a young age that I was unlikely to be able to carry a baby. My husband and I were very keen to adopt but were devastated to be told that because I have CF we wouldn't be able to; "you won't be well enough", "you won't keep up" are some of the reasons we were given to explain why we weren't suitable. With support from the CF mothers group, we decided to try again with a different local authority. The process is intense and included talking about CF, what impact it has and what would happen if one of us died. It was really emotional. One year after our initial enquiry about adopting, we went to a panel for a final decision. It was nerve-wracking...but worth it when we brought our daughter home! She's amazing and I would do it all again for her in a heart-beat."

Laura

"It was incredibly hard for me to come to the realisation that I couldn't carry a child because my lung function was too low and I had other health complications. We started to think about surrogacy and found a lot of support through the CF mothers group. Surrogacy is a long, complicated process and there's a lot to think about; finding a surrogate, carrier testing, fertility treatment, finances and the legalities of surrogacy. We decided to opt for host surrogacy which we describe as 'our bun, her oven!'. A friend offered to be a surrogate for us but sadly she and her partner had second thoughts and the arrangement fell through. The silver-lining to that was my Aunt, seeing how massively disappointed we were, offered to be our surrogate… our little girl arrives in April 2016!"

Tamsin

Surrogacy is another option. This involves another woman carrying and giving birth to a baby for you. There are different possibilities with surrogacy:

- An embryo created by the couple's own egg and sperm
- An embryo created with a donor egg and the intended father's own sperm
- An embryo created with donor egg and donor sperm
- An embryo created with own egg and donor sperm

There are lots of issues to consider with surrogacy and we strongly advise that you research it carefully, particularly around the legalities. You might find the Human Fertilisation and Embryology Authority (HFEA) webpages on surrogacy useful:

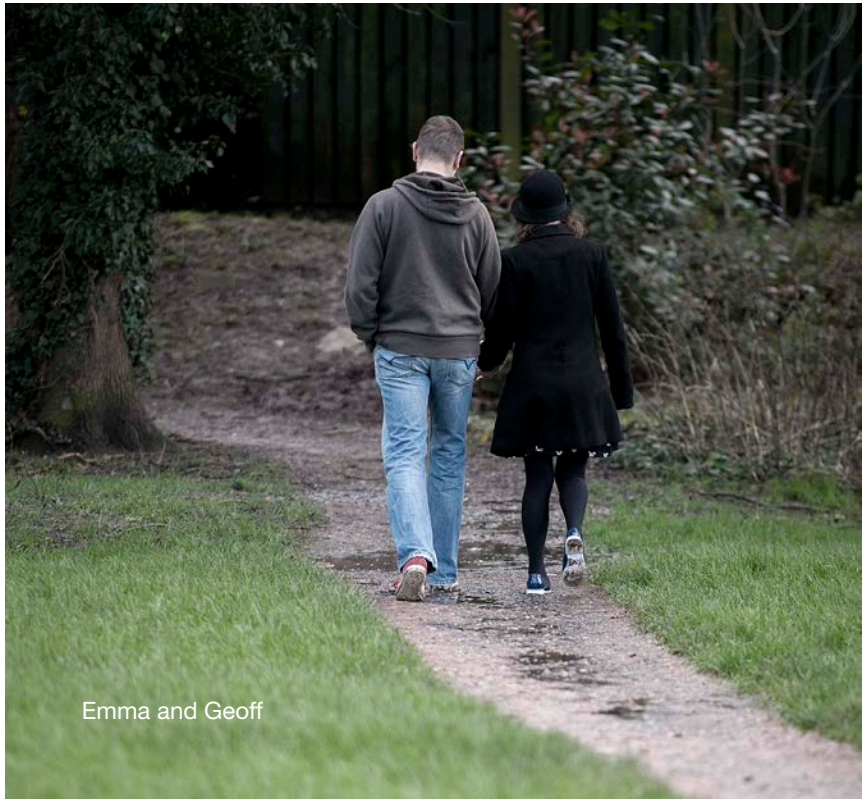
www.hfea.gov.uk

"Lee and I had agreed that we would only have three IVF attempts - we didn't want to get into the trap of 'just one more try'. We were going through PGD and for the first two attempts they couldn't tell if the embyos had CF or not. So it was only on our third attempt that an embryo was actually implanted. We were overjoyed and for those few days had a lovely feeling of 'maybe this is it'. Sadly, it wasn't to be. I can't deny that I want to be a mum and we were both obviously incredibly disappointed that it didn't work. But we are happy. We have nieces and nephews that we're really involved with, and that's fantastic."

Katie

"Of course we were disappointed, very disappointed. But there is more to life and you have to find other ways to be happy."

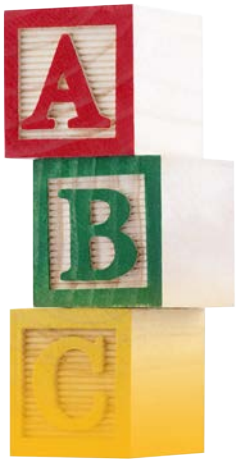
Lee



Emma and Geoff

Not having children

For some people, having children is not an option either because they don't want to or aren't able to. Cystic fibrosis can be a contributing factor in people deciding not to have children, however others may just not wish to be a parent.



"I'm not planning to have a family in the traditional sense. This is partly because, for me, the risk to my health is too great. It's not just the CF though, I think I would always have a strong inclination to adopt or foster. However CF consolidates that standpoint. I have the maternal part of me that would like to have a role in raising a child, but I don't think I need to be a biological mother to do that. Who knows? Maybe I'll change my mind in the future. I'm mentally prepared for a range of possibilities; step-mum Kate, foster mum Kate or aunt Kate could all be possibilities. However I do know that CF will be a determining factor even in these options, I need to be well enough to cope with the emotional and financial commitment that raising a child demands."

Kate

"I know for a fact that having CF has been a contributing factor to me wishing to remain child-free, but it's certainly not the only reason. The older I get, the more I'm noticing the gradual decline in my health, and the more I feel like this has been the right decision for me. I feel like most of my time is already spent trying to perfect the fine balance of work, my health, and a home, that I can't imagine adding a child to the mix! "

Jessica

Being told that you can't have the family you wanted, for whatever reason, can be devastating. Your CF team may be able to provide emotional support or direct you to other sources of support. There are some useful organisations listed at the back of this booklet. You can also contact the Cystic Fibrosis Trust helpline on **0300 373 1000** or at **helpline@cysticfibrosis.org.uk**

And finally

People with CF have shared their stories and experiences generously and honestly for this booklet. They have done this because they may have been through something similar to you and they want you to feel supported and informed. Whatever decision you make about having a family, your CF team and the Cystic Fibrosis Trust are here to support you.

Producing this booklet has been a team effort. The Cystic Fibrosis Trust is very grateful to all the volunteers who shared their stories and offered invaluable feedback. We are also very grateful to the CF mothers group (www.cfmothers.com) for sharing their knowledge, personal experiences and enthusiasm for this project. Thanks also to all our clinical colleagues for their support and expertise.

Tracy and Mike

For more information

Useful websites

Fertility and IVF

www.fertilityfairness.co.uk
www.infertilitynetworkuk.com
www.pgd.org.uk

Parenthood

www.nct.org.uk
www.cfinfo.org
www.cfmothers.com

Breastfeeding

www.nationalbreastfeedinghelpline.org.uk
www.laleche.org.uk

Adoption and Surrogacy

www.first4adoption.org.uk
www.hfea.gov.uk/fertility-treatment-options-surrogacy.html
www.cfmothers.com
www.surrogacyuk.org
www.gov.uk/legal-rights-when-using-surrogates-and-donors
www.nhs.uk/Conditions/ivf

Research and policy

Edenborough FP, Borgo G, Knoop C, Lannefors, L, Mackenzie WE, Madge S, Morton AM, Oxley HC, Touw DJ, Benham M and Johannesson M (2008) "Guidelines for the management of pregnancy in women with cystic fibrosis" Journal of Cystic Fibrosis S2-S32 https://www.ecfs.eu/files/webfm/webfiles/File/documents/Pregnancy.pdf

Frayman KB and Sawyer SM (2015) "Sexual and reproductive health in cystic fibrosis: a life-course perspective" The Lancet Respiratory Medicine 3(1): 70-86

Goddard J and Bourke S (2009) "Cystic fibrosis and pregnancy" The Obstetrician and Gynaecologist 11(1): 19 – 24

NICE guidelines (CG156) Fertility problems; assessment and treatment http://www.nice.org.uk/guidance/cg156

Thorpe-Beeston JG (2009) "Contraception and pregnancy in cystic fibrosis" Journal of the Royal Society of Medicine 102: S3-S10

Welsh Health Specialised Service Committee (CP38) Specialised Fertility Services http://www.wales.nhs.uk/sites3/Documents/898/CP38%20Specialist%20Fertility%20Services%20v6%200.pdf

Glossary

Burkholderia cenocepacia is one of a group of bacteria called *Burkholderia cepacia* complex. These bacteria do not usually cause problems in healthy people but can be very harmful to people with cystic fibrosis.

Continence refers to control of the bladder or bowel. Urinary incontinence means when someone is unable to fully control their bladder, faecal incontinence refers to loss of control of the bowel.

Embryo is a baby in the very early stages of development (less than three months). Once the embryo reaches around 3 months it will usually be referred to as a foetus.

Epididymis is a coiled tube that is connected to and wraps along the side of each testicle. It connects to the vas deferens. Sperm mature and are stored in the epididymis.

Gastrostomy is a surgical opening into the stomach that is carried out to allow a feeding tube to be fitted.

Metabolism refers to all the chemical processes that take place in the body's cells. The conversion of food into energy to fuel the body is a key metabolic process.

Naso-gastric tube is a tube fitted through the nose into the stomach. This is usually a short-term solution to allow medicine or feeds to be easily delivered.

Ovaries are the female reproductive organs where eggs are produced. A women will usually have two ovaries, one on either side of the uterus.

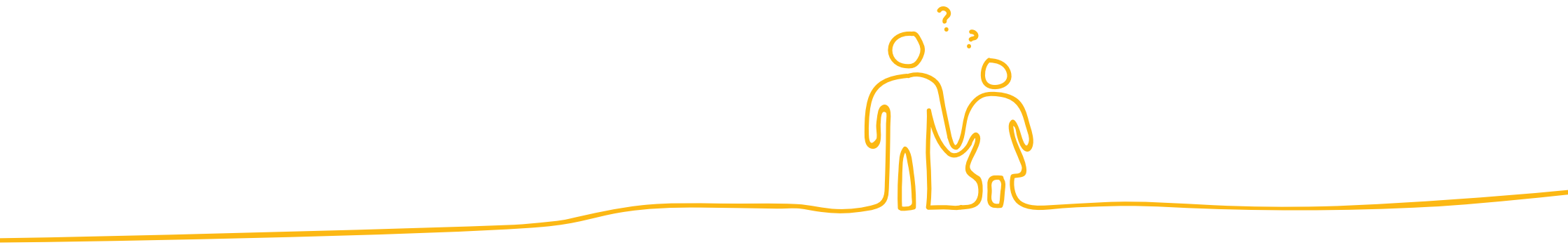
Semen is the fluid that mixes with sperm. For most men with CF, their semen will not contain any sperm due to the absence of or blockage in the vas deferens.

Sperm is the reproductive cell produced by a man, which combines with the woman's egg in reproduction.

Testicles are the organs enclosed in the scrotum behind the penis, which produce sperm.

Vas deferens is a tube that connects the testicles to the penis. In CF, this tube is often blocked or absent, which means the sperm can't get to the penis.

Intrauterine insemination (IUI) is a fertility treatment that involves inserting the sperm directly into the womb. The sample is prepared in a laboratory to ensure the fastest and healthiest sperm are used. The procedure doesn't take long and is usually painless.



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