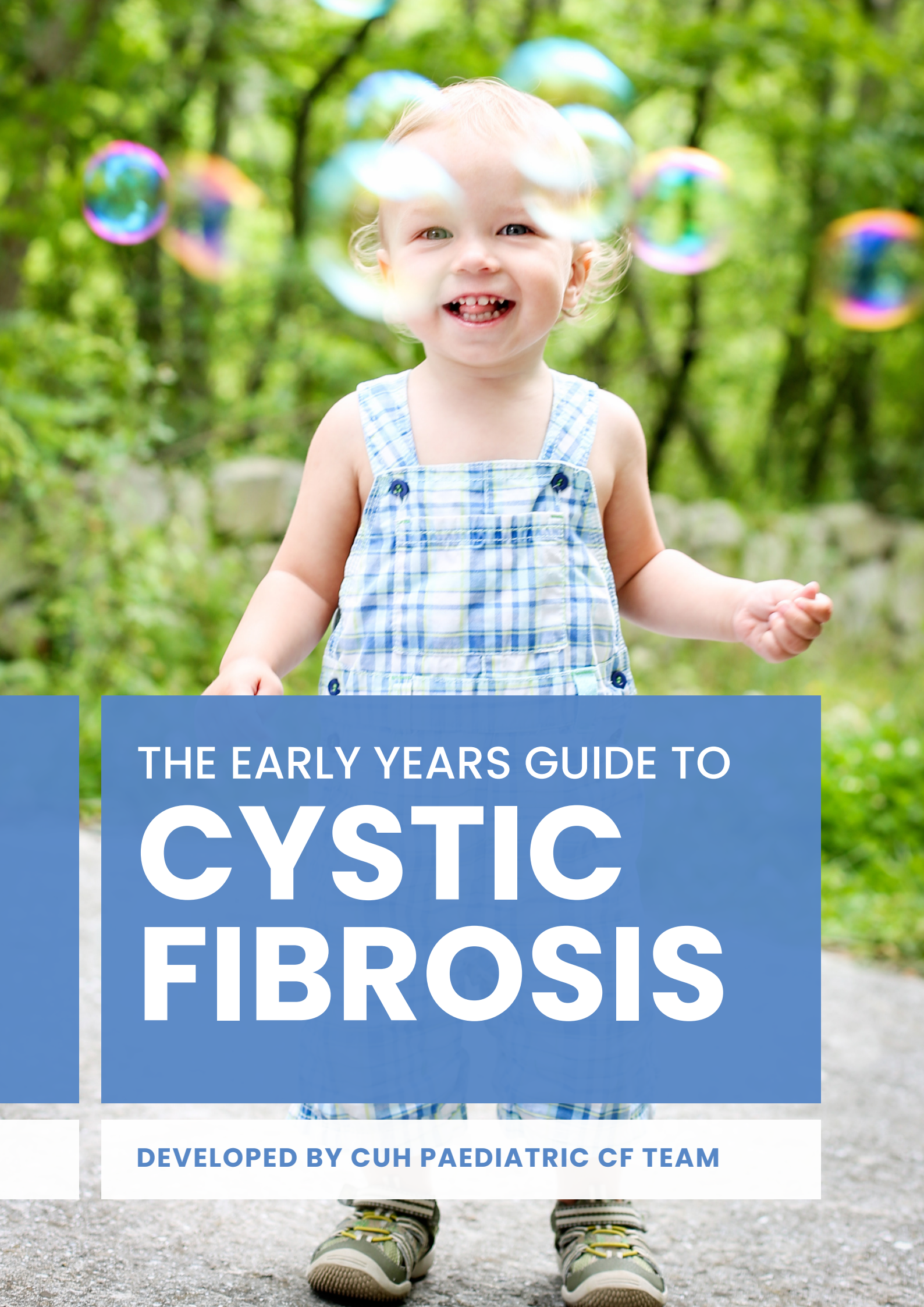

THE EARLY YEARS GUIDE TO CYSTIC FIBROSIS

DEVELOPED BY CUH PAEDIATRIC CF TEAM

CONGRATULATIONS ON YOUR GORGEOUS NEW BABY!

It is our privilege to work as part of the CF multidisciplinary team. We look forward to working with you and your child to ensure he/she has the best outcome with this condition. We are committed to implementing international best practice here at Cork University Hospital.

The CF Team

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CUH
Ospidéal Ollscoile Chorcaí
Cork University Hospital

Feidhmeannacht na Seirbhíse Sláinte
Health Service Executive

YOUR CUH CF TEAM

Below are the members of the CUH multi disciplinary CF team:

MEDICAL TEAM

The CUH paediatric consultants are Dr. Muireann Ní Chróinín, Dr. David Mullane and Dr Gavin Stone. These consultants manage the care of all paediatric CF patients. You will have met one of these doctors when receiving your child's diagnosis. You will usually meet one of these doctors at your clinic appointments, day unit reviews, annual assessments and if your child ever has to stay as an inpatient in CUH. Non-consultant hospital doctors (NCHDs) work as part of the CF team and will also be present at CF clinic.

NURSING

The CF nursing team will be your point of contact regarding appointments and if you have any concerns about your child. You will meet one of the nurses at your clinic appointments, day unit reviews, annual assessments and if your child ever has to stay as an inpatient in CUH. In some cases, the nursing team may liaise with your local pharmacy to ensure the correct medication is in stock to allow you to fill your prescription. You can contact the nursing team between the hours of 9am - 4pm Monday to Friday. Queries that do not need an immediate reply can be emailed to CUH.PaediatricCFNurses@hse.ie. You can contact nursing team on 0879683245 for any Cystic Fibrosis Related query. Weekly clinic takes place on Tuesday afternoon 1.30pm – 4.30pm and Wednesday mornings 9am-12 midday, calls may not be answered during this time but leave a voicemail or text message and we will return your call as soon as possible.

If possible please ring the nurses with any queries before lunch to allow the nurses to deal with the queries in a timely manner.

PHYSIOTHERAPY

You will meet the physiotherapist at your clinic appointments, day unit reviews, annual assessments and if your child ever has to stay as an inpatient in CUH. They will review how you and your child are getting on with their airway clearance routine. They will discuss how your child is exercising and will offer advice if needed.They will check your equipment and answer any questions you may have. The physiotherapy team can be contacted on 087-3602979 between 08.00-16.30 Monday to Friday.

CLINICAL PYSCHOLOGY

The clinical psychologist provides psychological support to you as parents and eventually your child (and any siblings he/she may have) following a diagnosis of CF. Receiving an unexpected diagnosis can be difficult for families. Having a chronic health condition can have an emotional and psychological impact and can be stressful in many different ways. The clinical psychologist is here to help you make sense of any difficulties you may be experiencing and support you to find a way forward. They offer input from the point of diagnosis throughout your journey as a family through the Paediatric CF Service until your child transitions to the Adult CF Service. The clinical psychologist also supports your family with preparing for this transition when the time comes.

DIETITIAN

The CF dietitian's role is to monitor your child's growth and development, to provide advice and guidance to maintain a healthy weight and body mass index for their age, and to ensure they are getting sufficient nutrients for growth.

Our CF dietitian will monitor your child's:

1. Weight, height/length, BMI
2. Relevant blood results such as fat soluble vitamin levels (A, D, E and K) and iron.
3. Urinary sodium – this is measured by taking a urine sample. We do this to ensure there is adequate sodium in your child's diet.
4. Faecal elastase – this is measured by taking a poo sample. We do this to monitor your child's pancreatic function.
5. Dietary intake. The dietitian may suggest you complete a food diary.

The dietitian will assess your child's dietary intake and compare this to your child's nutritional requirements and the National Healthy Eating Guidelines and provide dietary suggestions specific to your child as required. The dietitian will also advise on vitamin supplementation based on your child's requirements, dietary intake, and blood results.
You will meet the dietitian at your clinic visits and at your annual assessment. You can also mention any diet related questions to the CF nurses and a dietitian will contact you.

SOCIAL WORK
Following a diagnosis of CF all children and families are referred to the Medical Social Worker by your child's multidisciplinary team. Within the CF Team the role of the medical social worker is to support patients and their families through the emotional and practical challenges of a diagnosis of CF. Social workers can provide practical advice and counselling to children, parents and siblings and can also assist you in making contact with community services that may offer additional support or services. You may also wish to avail of additional social work support at different times in your CF journey and that support is available.

The Social Worker can meet you during hospital appointments, inpatient stays or alternatively you can arrange an appointment by contacting the Medical Social Work Department Monday to Friday 8am to 4pm on 021 4922488.

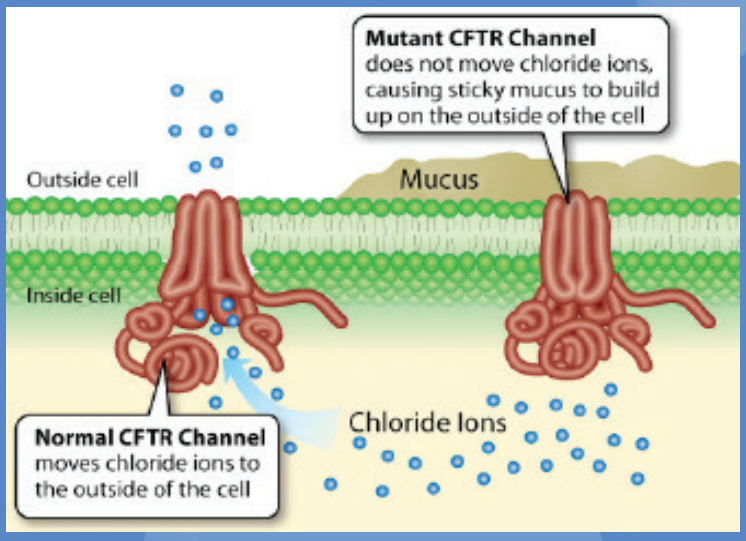
What is cystic fibrosis?

Cystic fibrosis is a genetic inherited disease affecting mainly the lungs and digestive system. CF occurs when a child inherits a CF gene from both parents. CF can affect both males and females equally.

The CFTR (Cystic Fibrosis Transmembrane Conductance Regulator) gene normally makes a protein that controls the movement of salt and water in and out of our cells. If this gene is impaired it can cause thickened secretions in the lungs and pancreas. In people with CF, this gene is altered so the protein does not function correctly. The ability of this gene can vary from person to person.

In some cases no CFTR protein is produced. In others the chloride channel opening may be abnormal. While in others the CFTR protein at the cell surface has sub-optimal function, thus allowing some transfer of salt and water but not enough to ensure normal secretions. Everyone contains two copies of this gene. For someone to have cystic fibrosis both copies of this gene must be altered.

There are new medications for children with CF with specific mutations called CFTR modulators. These therapies are targeted therapies and therefore an individual's genetic mutation determines which modulator the child can receive. Your consultant will discuss with you if your child is suitable for one of these medications.



Genetics

A child needs to have two copies of a mutated gene to have CF, one inherited from each parent. If a person has only one copy of a CF mutation, they are carriers of the CF gene and will not show any symptoms.

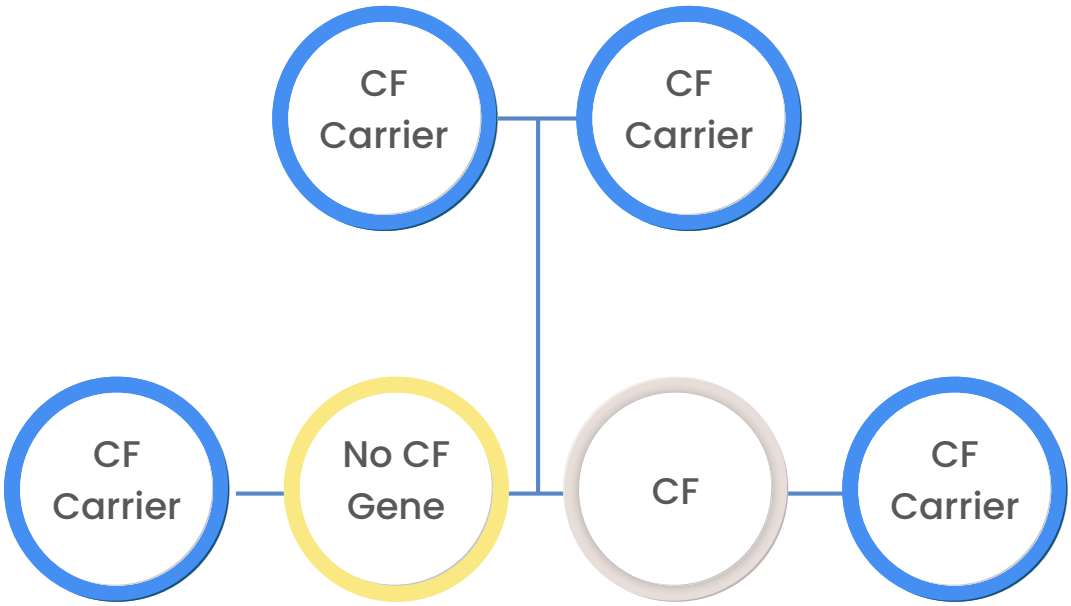
If both parents are carriers of the CFTR mutation; then for every pregnancy;

- There is a 1 in 4 chance (25%) that their child will have CF.
- There is a 1 in 2 (50%) chance that their child will be a carrier of the CFTR mutation.
- There is a 1 in 4 chance (25%) that their child will not have CF or be a carrier of the CFTR mutation.

This is explained in the diagram below
If both parents are carriers of the CFTR mutation, the chances of having a child with CF are exactly the same with each pregnancy.

Both boys and girls have an equal chance of being affected. If one parent has CF and one parent has two normal genes, then all of the children will be carriers.

You will be referred by your child's doctor for a consultation with genetic counselling. This appointment will help you understand the implications of a having a child with a diagnosis of CF in terms of future pregnancies and how it may implicate other family members.The appointment usually takes place in Cork University Maternity Hospital in the year following diagnosis.



Cystic Fibrosis Modulator Therapy

Modulator Therapy is a ground-breaking treatment for Cystic Fibrosis that targets the underlying cause of the disease rather than just managing the symptoms.

CF is caused by the mutations in the CFTR (cystic fibrosis transmembrane conductance regulator) gene. This gene codes for the CFTR channel, which is a transporter protein present in the cell membrane of particular parts of the body, like the lungs and digestive system. In CF this channel is absent, reduced or not working properly. These channels are crucial for regulating the movement of salt and water in and out of the cells. When they don't function correctly, thick sticky mucus builds in various organs particularly the lungs and the digestive system.

WHAT ARE MODULATORS?
CFTR Modulators are medications designed to correct the function of the defective CFTR protein channel. There are different types of modulators.

1. **Potentiators:** Enhance the function of CFTR channel that is already present in cell surface but is not functioning correctly.
   Eg Ivacaftor (**Kalydeco**).

2. **Correctors:** Help CFTR proteins fold correctly so they can reach the cell surface. Eg Lumcaftor and Tezacaftor (components of **Orkambi** and **Symkevi**).

3. **Combination Therapies:** Use more than one modulator to improve CFTR protein function. Eg Elexacaftor/Tezacaftor/Ivacaftor (**Kaftrio**).

ELIGIBILITY FOR MODULATOR THERAPY
The eligibility for modulator therapy depends on the specific CFTR mutations your child has. Not all CF patients will benefit from modulators as some mutations are not currently targetable by these therapies. For example (at time of publication):

G551D: Patients with this mutation are eligible for Kalydeco (Ivacaftor) from one month of age.

DF508: Patients with two copies of this mutation are eligible for Orkambi (Ivacaftor/Lumcaftor) from 1yr of age and Kaftrio at 2yrs of age. Patients that have one copy of DF508 will be eligible for Kaftrio from 2yrs of age.

BENEFITS OF MODULATOR THERAPY:
Respiratory Benefits: Reduce mucus build up, decrease infections and improve lung function.
Digestive Benefits: Enhance nutrient absorption and may lead to the reduction in pancreatic enzyme supplementation.
Quality of Life: Improved energy levels, weight gain, and overall well-being.

All children on modulators are closely monitored by the CF team. During the first year of treatment on any new modulator patients are required to have three monthly bloods.

Cystic Fibrosis Modulator Therapy

REQUIREMENTS BEFORE COMMENCING TREATMENT

- Eye exam to rule out opacities in the lens of the eye
- Baseline liver function blood tests to observe liver function – which will be checked every three months for the first year of modulator therapy and if you change to a different modulator they will be repeated again every three months for that first year
- Quality of life questionnaire
- Member of the Cystic Fibrosis Registry to allow outcome data nationally to be measured.

CONSIDERATIONS AND SIDE EFFECTS

While modulators offer substantial benefits, like all medications they may have side effects. Most children tolerate modulators well, and every child is different and assessed individually.

Common side effects reported include:

Headaches, Nasal Congestion, Cough, Abdominal Discomfort, Increased Liver Enzymes, Rash, Mood Changes, and Sleep Disturbance.

All of this will be discussed in clinic visits prior to commencement of any modulator.

Modulators are a long term treatment. Establishing a routine with medication adherence is crucial for ensuring your child receives maximum benefits. Receiving a diagnosis of Cystic Fibrosis for your child can be overwhelming, but understanding modulators and their role in managing CF can bring hope and positivity. The CF team will offer you support, information and guidance on using modulators effectively as the time for prescribing approaches.

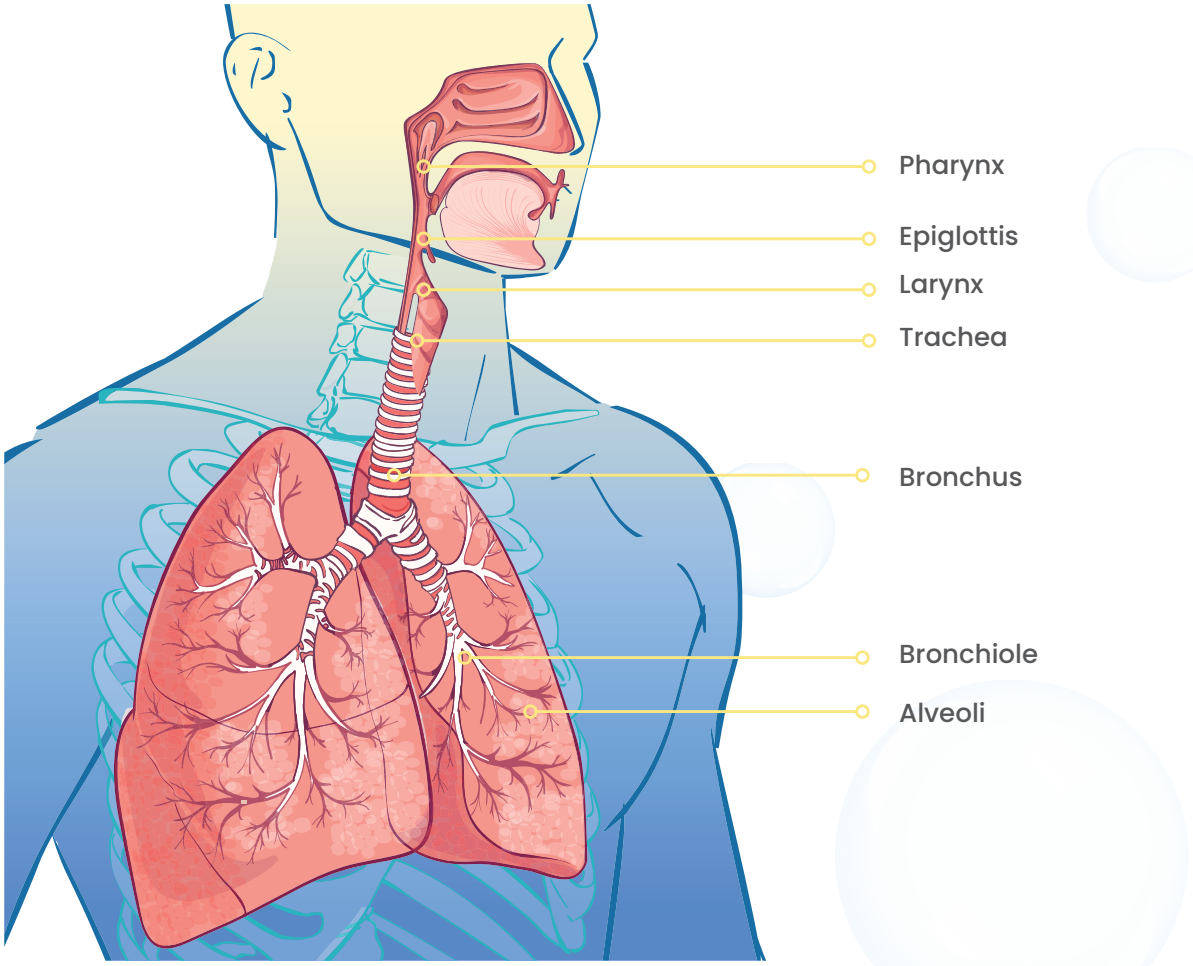
How do the lungs work? What do the lungs look like?

The human body has two lungs. They lie inside our chest cavity and are protected by our ribs. Our lungs transfer oxygen from the air we breathe into our blood. This oxygen is transported, by our blood, to the tissues and organs of our bodies. These tissues and organs need oxygen to work.

The lungs are made up of a system of tubes or airways. This system starts with the biggest airway called the trachea or windpipe. The trachea divides into two smaller airways called bronchi. The bronchi then divides into smaller airways called the bronchioles. Each bronchiole then divides into small air sacs at the end of the airway, these are called alveoli.

Bronchioles and alveoli are surrounded by small blood vessels called capillaries. These transfer oxygen into the blood and transfer waste such as carbon dioxide away from the blood into the airways to be breathed out. Everyone's lungs constantly produce secretions called mucus.

Mucus cleans and protects the lining of our airways, catches dust and germs in the air and allows us to breathe more easily. Our lungs are also lined with tiny hairs called cilia which help move mucus out of our lungs. Mucus can be cleared by coughing or huffing. This helps to keep our lungs free from infection.



Why does my child need physiotherapy?

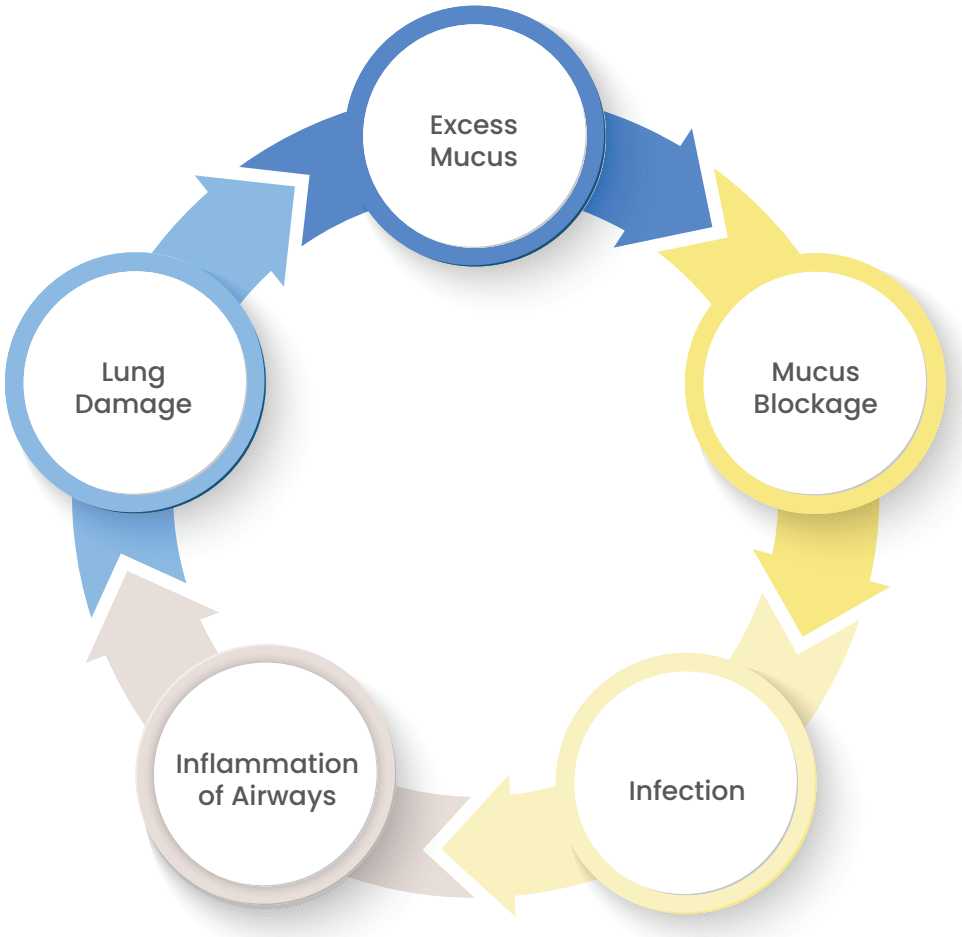
In CF mucus is thick, sticky and produced in larger amounts. Smaller airways can become easily blocked if this mucus is allowed build up.

This sticky environment is an ideal place for bacteria to grow and cause infection. An infection in the lungs can lead to swelling of the airways which makes the airways narrower. This makes it harder to breathe.

Repeated lung infections can cause permanent damage to the lung tissue called Bronchiectasis, which reduces how well the lungs work.

This damage can be lessened by clearing the mucus in the lungs with airway clearance techniques and exercise.

Physiotherapy is a combination of airway clearance techniques (see page 11) and exercise (see page 14). Alongside nebulisers and antibiotics, physiotherapy helps to move and clear excess mucus from the lungs and reduces the chances of infection and lung tissue damage. We want to get air into the lungs to clear mucus out of the lungs.



Breathing Assessment

Daily assessment of your child's chest and establishing your child's normal breathing pattern is very important. Once you are familiar with how your baby normally breathes, it will become easy to recognise if there are any changes. There are things that you can see, feel and hear if your baby has secretions or is unwell with a chest infection. It is beneficial to assess your child daily during a nappy change with their top or vest off.

Here are some tips for assessing your child's chest:

1. Ask

Ask yourself is your child:

- Unwell
- Tired
- Feeding
- Breathless
- Wheezy (sounds like a whistle when they breathe out)

2. Look

Does your baby look unwell?

- Are they breathing faster than normal? (normal for a baby under a year is 30-50 breaths per minute and for a baby 1-2 years of age is 20-40 breaths per minute)
- Is their chest being drawn in with breathing?
- Are there signs of a blocked nose i.e. struggling to feed because they can't breathe through their nose.
- Are their nostrils flaring more than normal?

3. Listen

- Does your child's breathing sound wheezy?
- Are they coughing more than normal or at all?
- Can you hear secretions/mucus when they cough? Is it a wet or dry cough?
- Can you hear any whistling or secretions coming from their nose?

4. Feel

- When you place your hands around their chest can you feel any secretions moving or rattling?
- Does the skin feel warmer than usual/ does your child have a temperature?

5. Treat

- When you are doing physiotherapy is your child coughing more than usual?
- Is it harder for them to clear their secretions?
- Is your child tiring more than usual during their treatment?

If you notice any changes from normal for your child, it may be a sign that your child has a chest infection. You may need to increase the frequency of your chest physiotherapy and contact a member of your CF team.

NASAL SYMPTOMS

Babies often have runny or blocked noses. This can be a concern for parents and make feeding difficult for the child. For babies with CF it is important to keep the nasal passages clear. To help nasal symptoms if your baby is experiencing them, you can use either saline drops (available from the chemist), **calpol nasal drops** or **sterimar drops.**

You can put 1-2 drops up the babies nose and this can be done up to 4 times a day but it is best to clear their nose 10-20 mins before they feed.

Airway Clearance

Airway clearance is part of your baby's physiotherapy treatment. It helps get air into the lungs and moves mucus out of the lungs. This keeps the smaller airways of your child's lungs healthy. Airway clearance should **always be carried out either before a feed or at least one hour after a feed** to prevent reflux of their food and to prevent your baby being sick.

How do we do airway clearance?
There are several different airway clearance techniques, but only some can be used on babies. This is because babies cannot cough or do breathing exercises to command, so treatment has to be given to them. The most common treatment techniques used are:

- Positioning
- PEP mask
- Bouncing on a gym ball +/- pep mask

Positioning
It is beneficial for your baby to spend some time on either one of their sides. This allows more air enter the uppermost lung, along with gravity, this will help clear any mucus in the lungs.

This can be done when changing your baby's nappy or during tummy time. It is recommended not to have your baby secured in a buggy/pram or high chair for more than one hour while they are awake.

Infant PEP
PEP stands for positive expiratory pressure. When using this technique with infants, a soft mask is placed on the baby's face. As the baby breathes out a small amount of back pressure is applied to the lungs. This will not be painful or uncomfortable for your baby.

This helps open up the airways, changes the distribution of air in the lungs and clears any mucus if there is any there. When you first start using the PEP mask your baby may wriggle or cry. Try to remain calm. If your child is very upset, stop and try at another time that day.

Airway Clearance

How to select the correct gym ball for you.

We often combine the PEP mask with bouncing on a gym ball. It is essential to use a ball that is suitable for your height and to look after your own posture when sitting on the gym ball with your baby. You can purchase an ABS gym ball at www.physioneeds.ie

When sitting on a gym ball:

1. Your feet should be flat on the floor and hip distance apart.
2. Your hips, knees and ankles should be at a right angle.
3. Your thighs should be parallel to the floor (not angled downwards).
4. Your spine should be in the "neutral" position meaning that your lower back should not be arched too far forwards or backwards.

45CM GYM BALL
– suitable for height 5'0" (152cm) or shorter

55CM GYM BALL
– suitable for height 5'1"–5'6" (155–167cm)

65CM GYM BALL
– suitable for height 5'7"–6'1" (170–185cm)

75CM GYM BALL
– suitable for height 6'2" (188cm) or taller

Airway Clearance

How to use a PEP mask with a gym ball?

1. Sit on an appropriately sized gym ball in the correct position
2. Hold you baby either in the crook of your arm if they are less than 6 months or sitting on your lap in front of you if they are older. It is important to support a baby's head if they have not yet developed head control (<6 months).
3. Start bouncing on the ball holding your baby in a safe position.
4. Once you and your baby are comfortable bouncing, introduce the infant PEP mask.
5. Place the mask gently over your baby's nose and mouth and continue to bounce. It is usually best to tuck the mask under the baby's chin first and then over their nose.
6. While bouncing, try and hold the mask over your baby's nose and mouth for 30 seconds to begin with. Remove the mask for 10-15 seconds. Repeat this on/off cycle for 10-15 minutes as tolerated.
   Do this 1-2 times a day as instructed by your physiotherapist.
7. Build the 30 seconds up to one minute as your baby tolerates the mask better.

If your baby coughs while the mask is on, remove the mask to allow your baby to cough properly.

As your child gets older

As your child grows older other methods of airway clearance can be introduced.

Blowing games such as blowing bubbles and blowing pieces of cotton wool can be used to help chest clearance and to strengthen their respiratory muscles. Forced expiratory Technique known as huffing or "crackly breaths" is a good way to help move secretions.

A fun way of getting children to do this is to fog up a mirror with an open mouth. This should be done during and after blowing games and exercise.

Exercising your baby

Exercise is an important part of your child's physiotherapy treatment. Exercise, jumping and laughing games make your child breathe more deeply, which allows air get to all the different parts of the lungs. If there is mucus in the lungs, it is often heard at this time.

Although babies can't exercise themselves, it is important that physical activity is introduced from an early stage. Being active as a family helps your child see exercise as a normal part of life.

How do I exercise my baby?

Positioning

Babies should be placed in different positions (i.e. on their side/tummy) throughout the day. Changes in position can help air distribution throughout their lungs and help them clear mucus. Being in different positions can also help your baby develop good muscle control. This can be done when you are changing their nappy/clothes or on a mat on the floor. It is recommended not to have your baby secured in a buggy/pram or high chair for more than one hour while they are awake.

Tummy time

Tummy Time should begin as soon as your baby comes home from the hospital. Tummy Time is when your baby lies on their tummy. Tummy Time is essential for an infants' core, motor, and sensory development. It helps to improve your baby's neck and head control, and strengthens their back, shoulder and core muscles. Being on their tummy also helps to prevent flat spots on baby's heads. Tummy Time should be done every day until a baby is able to crawl/move independently. If your baby doesn't like tummy time, don't get discouraged. Every bit of Tummy Time makes a difference!

How to do Tummy Time

Some babies do not enjoy being on their tummies and will not tolerate lying on their tummies for long. Build up tummy time in small time increments (1-2 minutes, 5 minutes etc). Aim for an hour of tummy time each day if your baby will tolerate it. You could build this into their routine by doing it every day after their nappy change.

Ways to encourage tummy time are:

- Lying your baby across your lap while distracting them with toys.
- Having a rolled up towel/blanket under their chest for support.
- Getting down to their level on the floor and encouraging eye contact with you.
- Placing your baby on your chest so your baby is face to face with you.
- Use rattles or other toys that make noise to encourage visual tracking.
- An interactive mat with mirrors / different colours and textures.
- Sing songs to calm and soothe your baby.
- Play peek-a-boo using a blanket or your hands
- Practice Tummy Time at baby's happiest time.

Exercising your baby

Once your baby is able to crawl/move independently on the floor you can add in more challenging activities such as:

- Crawling over various surfaces (steps, cushions etc)
- Reaching to either side with both hands for toys.
- Reaching overhead with both hands to bubbles/ toys.
- Rolling games.
- Pushing or pulling toys.
- Climbing up/down on to the couch/bed with your supervision.
- Crawling up and down the stairs with your supervision.
- Squatting down to get toys.
- Walking up and down the stairs while holding your hand.
- Ride on toys such as a tricycle.

THE FOLLOWING ARE SOME EXAMPLES OF **PLAY EXERCISE EQUIPMENT** THAT CAN HELP ENCOURAGE **FUN MOVEMENT WITH YOUR CHILD.**

Remember exercise/movement not only helps your baby's development, it is also an airway clearance tool to help keep their chest clear of mucus.

Tummy Time

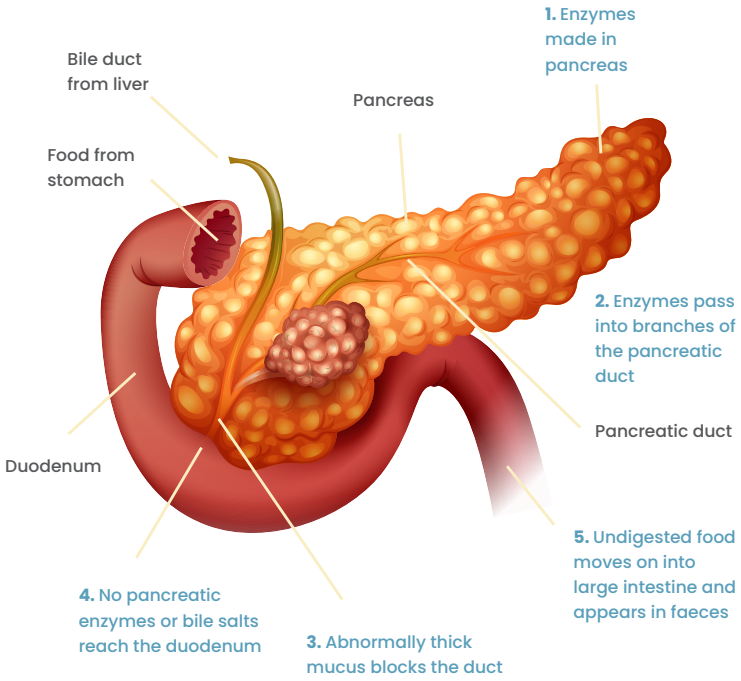
SOME TUMMY TIME AND SOME FLOOR PLAY POSITIONS.

PLAY IN SITTING - ENCOURAGING YOUR BABY TO TURN THEIR BODY AROUND AND TO REACH FOR OBJECTS.

Your baby's digestive system

How does CF affect the digestive system?
The pancreas is a gland in your abdomen that sits just behind your stomach. Glands are important organs, just like your heart or lungs. The pancreas makes substances called enzymes, which help your body break down food to give you the energy and nutrients you need to grow. It is common for people with CF to have issues with their pancreas.

In people with CF, the small channels where the digestive juices flow become blocked with sticky mucus. The enzymes then build up in the pancreas which causes the pancreas to become inflamed. This can lead to the formation of cysts and fibrosis (damage) of the pancreas. The effect of cystic fibrosis on the pancreas and digestion varies from person to person. It is now possible to replace most of the missing enzymes with enzymes taken by mouth. The most common enzyme prescribed in CUH is **Creon.**



Creon

Why do I need to take Creon?

Creon boosts the amount of enzymes in your child's body. This improves digestion and allows your child to absorb important fats and helps reduce wind. Creon comes in three different strengths- 5,000IU, 10,000IU and 25,000IU. Be sure to refer to the dosing schedule provided by your child's doctor or dietitian.

How much Creon does my child take and when?

The amount of creon your child needs to take depends on how much your child is eating and how much fat is in the food. You may find it useful to keep a food diary and track how much Creon your child is taking with their food. In order to work out the fat content in the food your child is eating, food labels or phone apps such as 'Carbs and Cals' and 'My Fitness Pal' can guide you at home. If your baby has not taken enough Creon for their food, you will notice their poo will be loose/runny/oily. The dietitian will provide you with written information regarding fat content in food and how much Creon to take at your regular clinic visits.

How does my child take Creon?

As babies are unable to swallow capsules their creon is in granule form. They take this sprinkled on top of a spoon of apple purée which is readily available in any supermarket. As your child gets older and they are able to swallow tablets they will take Creon in capsule form and will swallow them with water before their food.

Do all babies with CF need enzymes (Creon)?

Some babies do not need enzymes (Creon) at first but may need them later on. Your baby's bowel function and growth will be monitored by the CF team. Most babies with CF need Creon from birth and most will need to take them for the rest of their lives. It becomes part of their routine and children soon learn how to take it themselves. Some children who are taking modulators may become pancreatic sufficient and may not require Creon, a poo sample is required to test for pancreatic function (This is known as a Faecal Elastase), The CF team will request this test as needed.

Other parts of the body that may be affected by CF

Fertility
Fertility is usually normal or slightly reduced in females but most males are infertile. Their sexual function is normal but it is rare for males with CF to father children naturally. However, significant advances in in-vitro fertilisation have allowed men with CF to father children.

Sinuses
People with CF can be prone to sinus infections, congestion, nasal polyps or hay fever. This can be managed with nasal sprays and medication which your team will discuss with you if indicated.

Liver
Some people with CF can develop CF related Liver Disease. Your child's liver function will be monitored using a blood test.

Diabetes Mellitus
Diabetes Mellitus develops in approximately 30% of adults with CF causing very high levels of sugar in the blood. Your child's blood sugars will be checked annually to monitor for any changes from the age of 10 onwards.

Bone Health
Traditionally people with CF were prone to developing osteoporosis due to their reduced absorption of nutrients. Due to advances in enzymes, monitoring and treatment, osteoporosis is less common in children diagnosed with CF today. Maintaining good bone health is important for everyone. This can be done by consuming calcium and vitamin D as well as doing weight bearing exercises (running, jumping, skipping) as your child gets older. Your team will discuss this with you at clinic. Your child's bone health will be monitored from the age of 10 using a DEXA scan.

The Emotional Impact of a CF Diagnosis

Having a newborn is a busy time with many highs and lows. Perhaps this is your first child or you may already have other children. You may still be recovering from the birth and adjusting to the changes to your life that a baby brings with her/him. Perhaps you feel confident and excited or perhaps you feel slightly terrified and exhausted. All of the above and more are typical feelings with a new baby.

Having a baby is a monumental change in itself. You have now also been given the news that your little one has CF, a life-long health condition. Each parent will react differently to this diagnosis. Parents often report feelings of sadness, loss, anger, and worry, especially given the uncertainties they face in terms of their child's future. You may be concerned for your child and for your family. You may be wondering what to say to others about the diagnosis or how to explain it to your child and their siblings as they get older. Everyone copes in different ways with these challenges. Some ways of coping are more helpful than others, but all are understandable in the context of the circumstances you find yourself in.

Parenting in the context of a child having CF can have its challenging moments and its own stresses. It is important that you are kind and compassionate towards yourself.

A meeting with the clinical psychologist is offered soon after diagnosis. This is to provide you with space for you to reflect on your thoughts and feelings about the new diagnosis. Some people look for practical strategies to manage the feelings that arise. Others simply like to know that the clinical psychologist is available should they need to speak with them at a future date when needed.

Managing the Psychological Impact of a Diagnosis of CF

When might it be useful to see the Clinical Psychologist?

Emotional support is available from many members of the CF team. Much of the time you may find that talking through your challenges with the team, or with family and friends will be enough. However sometimes further help is needed.
The clinical psychologist provides psychological assessment and intervention. Sometimes a one-off consultation is all that is needed. At other times, we may meet for a longer period. The approach the clinical psychologist takes depends on the needs and circumstances of each family.

In general the role of the clinical psychologist in the CF team is to support the young person and their family in the context of their long-term health condition. Some of the challenges that the clinical psychologist can help with include:

- Adjusting to and living with CF
- Difficulties managing the treatment plan
- Worries that you may have about treatment or about the future
- Anxiety about needles or procedures
- Managing pain
- Dealing with stress and difficult or overwhelming feelings
- How CF affects family life
- How CF affects your child's sense of self and identity, particularly as they grow up

What about confidentiality?

Information discussed during appointments will be kept confidential, within certain limits. The clinical psychologist will explain confidentiality further at your appointment. The clinical psychologist works as a member of the multi- disciplinary team and communicates regularly with them to provide the best care. We can discuss what will be shared with the team and what will not when we meet.
If there is a reason to think that there is a risk of harm to your child or others, this information will have to be shared. This is because it is our duty to keep people safe. Should this happen, we will talk to you about this first wherever possible.

When and how can I meet the clinical psychologist?

The psychologist attends as many clinic and annual assessments as possible. This is an opportunity to check in with families and offer advice and guidance. At annual assessment the clinical psychologist will also discuss your emotional wellbeing and that of your child with you, and consider whether there is any need for extra supports to be put in place. If your child is admitted to hospital, input from the clinical psychologist is available during their stay.

PSYCHOLOGY APPOINTMENTS

Individual appointments with the clinical psychologist can be requested through our CF Nurses or any other member of the MDT. If you opt for this extra support, our first appointment may last up to an hour and a half, either in person or by phone/video call. After initial assessments, appointments usually last an hour. In session we will talk about your concerns. The clinical psychologist will ask questions about how you are managing your child's CF care and how the diagnosis is impacting on your family. The clinical psychologist may use questionnaires to help with the assessment process and to monitor change over time.

They may also ask your permission to speak with other relevant people such as your GP. The reason we cover all of these areas in our assessment is to ensure we get the best picture of your presenting needs. This helps us determine what intervention might suit your family best. Figuring out the best approach is always a collaboration between the clinical psychologist and the family. The clinical psychologist is not medically trained, so there are no physical examinations, injections or other medical procedures during these appointments. A clinical psychologist does not prescribe medication.

Managing the Psychological Impact of a Diagnosis of CF

As your child grows there may be periods in their life where the clinical psychologist input is needed. During this time the clinical psychologist will try to meet with parents/caregivers by themselves, the child or young person by themselves, and the family altogether to discuss the child's needs and decide on an appropriate intervention.

What happens after our initial appointment?

After our first appointment we can decide together what to do next.

There are a few options:

- Some people decide that they do not need to meet with the clinical psychologist again. Sometimes one or two appointments is all that is needed
- Some people may need referral on to a different service
- We might identify some goals for working together and arrange to meet for further appointments.

Talking to your child about having CF

You have been given this booklet because your baby has just been diagnosed with CF. It may seem far too early as yet to think about how to explain their condition to them. We know from research, however, that talking about your child's CF and incorporating the terms 'cystic fibrosis' and 'CF' into your family life from the early days can be very helpful for your child (and any siblings they have) as well as for you, as their parents. We recommend using these terms in your everyday language when carrying out tasks and activities that are part of your child's CF care. In this way, as your baby grows and develops, s/he will come to know the terms 'CF' and 'cystic fibrosis' as familiar, non-threatening words and s/he will start to form a positive narrative about their condition. Our aim is for your child to understand that their CF is a part of them, but does not define them. We hope to empower your child, and you as their parents, to feel competent and confident to manage their CF. An important part of this is helping children to understand, in a child-appropriate way, why they receive certain treatments, carry out specific care tasks, and come for regular check-ups at the hospital. As your child's Paediatric CF Team, we will all use the terms 'CF' and 'cystic fibrosis' during your child's appointments.

You will likely find that as your little one gets older, s/he will have questions about their CF. We know that in families where CF has been named and spoken about openly, children tend to have more positive outcomes in terms of their emotional well-being and their sense of self in relation to their condition, as well as greater compliance with treatments. Sometimes parents try to protect their children by not telling them about their CF. We know that their intentions are loving, but unfortunately this can lead to children struggling to accept and understand their diagnosis at a later date when they are told about it. It can also result in some difficulties with treatment compliance as children do not understand why they have to carry out certain tasks while their sibling/peers do not have to do so.

If you feel you would like some help with talking to your child (or their siblings, or others) about CF, you can ask to speak to our clinical psychologist.

Infection Control

Chest infections are caused by “bugs” including viruses and bacteria that are harmless to people without CF but can cause damage to your baby’s lungs. Children with CF are advised not to mix with other children or adults with the condition.
This is because people with CF can carry certain bacteria in their lungs that are not harmful to people without the condition, but can be very harmful to others with cystic fibrosis. Personal hygiene is very important for children with CF. Children should be taught from a young age to regularly wash their hands.

Harmful bacteria can also be found in certain environments. Things to avoid are:

- Squirting bath toys
- Swimming in stagnant ponds & canals
- Fish tanks (especially warm tanks for tropical fish)
- Compost
- Mucking out stables
- Hot tubs, Whirlpools, Jacuzzis, Hydrotherapy pools
- Tropical greenhouses, butterfly houses, outdoor misting systems

Hospital Information

Where is the CF paediatric unit?

The CF paediatric unit is located in the Seahorse Day Unit. If you arrive at the front door of the hospital , walk straight past main reception. You will see a shop on your right. The Seahorse Day Unit will be on your left a little bit beyond the shop. You will walk through double doors. A member of the CF team will meet you here or ring the CF Nurses phone once you arrive on the ward.

Parking

Concession parking is available for patients/ visitors on site of CUH. This car park is at the front of the hospital. As you enter the main entrance of CUH from the road turn left and follow the road straight ahead. This road will bring you around to the right and the car park will be located on your right. Please discuss parking discs with nursing staff if your child is an inpatient.

ATM

There is no ATM on site in CUH. The closest ATM is across the road in Wilton Shopping Centre or in the Bank of Ireland outside Wilton Shopping Centre

Nappy changing facilities

You can change your baby's nappy in your private clinic room

Toilet

There will be a private toilet in your clinic room.

Cafés

There are two visitor cafés in CUH. The Fastnet café is located passed the Seahorse day unit to the left of the hospital lifts. The Coffee Doc café is located in the Cardiac Renal Centre , this is to the right as you enter the main door of the hospital. Coffee/ tea, cakes and sandwiches are available at both of these cafés. Visitors can not use the staff canteen.

Clinic Visits

What is a clinic?

A clinic is a review to discuss how your child is doing. It is an opportunity for the team to monitor your child and assess if any changes need to be made to their diet/enzymes/airway clearance/ exercise. These are particularly important for younger children as they are unable to tell us how they feel. Some clinic appointments may be virtual or over the phone.

Where do clincs take place?

If you are attending clinic in person, all clinic appointments take place in the CF Unit in the Seahorse Day Unit.

Who will I meet at the clinic visit?

You will meet the CF nurse, the physiotherapist, the dietitian, and the doctor. If you would like to see the social worker, this can be arranged.

How often do we need to go to clinics? How long will it take?

Your child will be seen every 2-3 months in clinic. A clinic visit takes a maximum of 90 minutes.

What do I need to bring?

Bring your child's airway clearance device (pep mask/aerobika etc). Your CF nurse will let you know if there is anything additional that you need to bring.

How do I arrange clinic appointments?

You will receive a letter with your appointment time and date. Please note the original time may change. You will receive a text confirming your time a few days prior to your appointment.

Infection control measures during clinic

Each child is allocated to a room, and all the members of the CF team (CF nurse, physiotherapist, dietitian, doctor) come to see him/her in that room. You will be shown directly to your room on arrival and will not have to wait in a waiting room. Staff members wear aprons and gloves and may have a mask on entering each room, these will be changed between patients. The room is thoroughly cleaned (desktops, chairs, sinks, surfaces) between appointments. This can sometimes delay your appointment, due to this we request that everyone rings the CF nursing phone before leaving their car.

Annual Assessment

What is an annual assessment?
An annual assessment consists of 2 appointments. The first of these is an appointment for tests to be carried out, the second will be when you get the results of these tests. Please see details below

Where will the annual assessment be?
All annual assessments take place in the CF Unit in the Seahorse Day Unit.

What tests will my child get?
- Blood tests checking their liver and kidney function, their vitamin levels, their bone health, their full blood count. Your child is not required to fast for these blood tests.
- Nose, throat, axilla and groin swabs.
- Lung function test (from aged 5).
- A walking test (from aged 6).
- Liver ultrasound from (aged 3 years).

Why does my child need these tests?
These tests are important to monitor your child's overall health. It is to ensure that your child is getting the appropriate treatment.

When will I get the results of these tests?
An appointment will be arranged for you to attend the CF unit on a Wednesday morning at 9am to receive your child's annual assessments results.

Who will I meet at the annual assessment?
You will meet the CF nurse, the physiotherapist, the dietitian, the doctor and the psychologist.

What do I need to bring to my results day?
- Your child's airway clearance device (pep mask/aerobika etc) and their medications.
- Your food diary.
- Your list of medications from your pharmacy.

Your CF nurse will let you know if there is anything additional that you need to bring.

How do I arrange an annual assessment?
You will receive a letter with your appointment time and date.

Hospital Admission

Why would my child need to be admitted to hospital?
Your child may develop a CF exacerbation (chest infection). If it can not be managed with oral antibiotics, they will be admitted to hospital for IV antibiotics. If a new baby is having significant feeding issues, they may require a short admission to help manage this and ensure adequate weight gain.

Where will they go?
When your child is under 2 years old they are admitted to Ladybird ward on the 1st floor of the hospital. When they are over 2 years old they are admitted to Puffin ward on the 5th floor of the hospital.

How long will they need to be in hospital?
This depends on why your child is in hospital. If your child requires IV antibiotics, they would likely need to stay for at least 14 days. As your child gets older, there will be a home IV option. Your CF team will discuss this with you if appropriate.

Can I stay with them?
Yes, one parent is welcome to stay with their child in hospital. Beds are provided in the rooms in Ladybird ward.

Will the CF team be looking after my child?
Yes your child will be reviewed every day by the CF team. Your CF consultant will be in charge of your child's care while they are in hospital.

Infection control measures while in hospital.
If your child is admitted to hospital they will be given a single room on the ward.

What do I need to bring to hospital?
- Medications
- Any airway clearance equipment you use at home.

Additional information/ Frequently asked questions

Are there different versions of CF? (i.e. mild/moderate/severe)

Some people are affected by CF more than others. There are a variety of factors that can contribute to this.

Will my GP and PHN be informed of my child's CF?

Yes the CUH team will inform your GP and PHN. Your GP and PHN will still be involved in your child's care for your child's weight checks, routine vaccinations and non CF illnesses or queries.

Do I still attend regular baby checks with the PHN?

Yes please attend all of your child's regular PHN appointments.

When do I ring the nurses if I am concerned about my child?

If you notice your child has a new respiratory problem (i.e. cough, nasal symptoms, wheeze, change of breathing pattern) ring the CF nurses phone. Please see physiotherapy assessment section to examine your child.

Who do I ring if there is an emergency with my child at the weekend?

You can contact your local out of hours GP service or else if you think your child needs to come to hospital please attend the paediatric ED department at CUH. There are paediatric staff available on call to deal with issues that may arise.

Which milk should I feed my baby?

Babies with CF can be fed by either breast or standard formula milk. We will support you whichever method you use to feed your baby. Your baby's weight gain will be monitored regularly after the diagnosis and in the first year.

What if I forget to give the enzymes (Creon)?

If a baby with CF does not receive enough enzymes their stool will be looser. If enzymes are missed regularly, the baby will not be able to digest food properly and their growth will suffer.

What if my child does not eat a meal after having the enzymes?

Your child is like any other and may refuse to eat on occasion. Having no food after a full dose of enzymes will cause no harm to your child.
If this happens often you can give half the dose of enzymes at the beginning of the meal and half at the end of the meal. Mealtimes can be challenging for many young children as many go through periods when they do not want to eat much.
If mealtimes are becoming difficult, chat to your CF team as soon as possible.

How will coughs and colds affect my child?

All children, whether they have CF or not, suffer from numerous colds in their first few years of life. In children with CF the symptoms can last longer because of the increase in lung secretions.
Most colds are caused by viruses. If your child has ongoing symptoms they may need an antibiotic. Early and frequent antibiotic treatment can prevent or delay lung damage if your child is suffering from a chest infection. Extra physiotherapy is often needed if there is a lot of extra mucus.

Are cough medicines useful?

Cough medicines suppress the cough. If a child with CF is coughing it is important to find out what is causing the cough. Cough medicines should not be given to children with CF without discussing it with your CF team.

Why are swabs/sputum samples needed?

Throat swabs are performed regularly on all CF patients for surveillance. We do this to monitor if your child is growing any new bacteria. If your child has a new cough you will be asked to take a throat swab of your child and send it into the CF nurse at the following address: CF Nurses, Seahorse Day unit, CUH, Wilton Road, Cork. Swabs should be sent by post Monday to Wednesday only, but can be delivered in person Thursday and Friday.

Additional information/ Frequently asked questions

Can my child go to creche/ day care/ school as normal?

We would encourage attending a child minder for the first 2 years of life to avoid group child care settings. It is recommended to try and limit exposure to viruses for the first 1- 2 years of life if possible. Many schools and creches will have experience of caring for children with CF.
We recommend that you let your child's creche/ school know about your child's condition. It is important to familiarise staff with your child's Creon (if required). We encourage children to attend school as normal. The condition is part of their life, but it doesn't define them and shouldn't limit their school experience.

Are vaccines important?

We would encourage full vaccination of your child. It is advised that all CF patients get the yearly flu vaccine and additional pneumococcal vaccine at 2yrs of age.

Is financial help available?

Information can be found on the CF Ireland website. CF patients/parents are eligible to apply for the long term illness card which covers the majority of medications. Domiciliary carers allowance is also available for parents.

Things to remember in the warm weather:

The movement of salt and fluid through the cells in the body is altered in children with cystic fibrosis. This means high levels of salt can be lost through their sweat. This is especially important to consider in hot weather. Your child will sweat more, meaning the sodium level in the body can drop quite quickly, and therefore increases the risk of dehydration. When the weather is very warm it is important to give additional salt and fluids to ensure your child's sodium levels remain normal.

Here are some helpful tips to avoid this issue :

- Keep out of the sun between the hours of 11am and 3pm.
- Dioralyte (or equivalent) sachets are ideal. These can be bought over the counter in chemists and supermarkets.
- Ensure your child drinks plenty of fluid.
- Encourage your child to wear a hat and use usual precautions to avoid sunburn, including high factor sun cream.
- Add extra salt to your child's food and give them salty snacks. If your child starts on modulator therapy less salt is lost in the sweat so this precaution will not be necessary.

RESEARCH:
Your child may be invited to participate in research studies. If your child is suitable for a research study you will be informed by the CF Team.

Helpful Resources

- www.cfireland.ie
- www.cysticfibrosis.org.uk
- www2.hse.ie/babies-and-toddlers/
- Mobile phone apps including CF Daily can be useful to track nutrition, exercise and treatment routines.

Parents Perspective in the CF Service

Naturally enough we got a huge shock when we first heard of our baby's diagnosis. We were devastated for her and for us as first time parents. We assumed she was going to be a very unwell baby with lots of hospital admissions but this was certainly not the case as she was so well looked after by the team in CUH from the very beginning. Knowing and feeling the support from them made everything that bit easier and soon we became more relaxed about both the diagnosis and the administration of her medication which was very overwhelming at the beginning. Nowadays with the advancements of modulator drugs and their availability at such a young age, we can put our minds at rest that all will be okay and she can grow up living an ordinary life like any other child.

- PARENTS OF 13-MONTH-OLD

Fear of this new unknown is natural. Take it step by step. This will just be your normal as a family with cystic fibrosis. Don't let it define who you all are; it's just a part of your family.

Talk to each other as a couple, be open and honest about how you feel. This isn't the hope you had when having children but can navigate it together.

The team are there to help and support you, take their advice but you are mom and dad the experts living this day-to-day. Trips to clinic are a busy day but take it as a family day out together. You're a wee team and you can all get through this.
This isn't as scary as it first appears. The advances in CF medicine are massive. You can do this.

- PARENTS OF 10-YEAR-OLD AND 6-YEAR-OLD

It took us time to come to terms with our child's diagnosis, we kept waiting for something to go wrong. It took me about a year to see how strong she was and it was such a relief to see her getting bigger and to realise she was as much trouble as any other toddler. And sometimes we even forget she has CF- which we never expected at the start. We were concerned about any future children we would have even while our baby was a newborn. We had so many questions and fears. There were no quick or easy answers, no right or wrong decisions to be made. The genetic counsellor provided a very helpful insight into what our options were.

We received good support from the Seahorse CF team, the CF community and CF mammy groups.

- PARENTS OF 5-YEAR-OLD

Getting the diagnosis for us was a massive shock. He is the fifth of five boys with no history we know of, with anyone having Cystic Fibrosis. When we initially got a call from CF specialist nurse, I honestly thought they've got this wrong, having done the "quick Google", this couldn't be right with our 4 older boys not having CF.

We came in a day later for the sweat test and I think at this point we knew, even before the confirmation, that the result would be positive for CF. The CF team immediately put us at ease.

They explained everything and told us of the wonderful new breakthroughs in medication specifically to treat Cystic Fibrosis. We knew we

had a lot of homework to do with physio, pep masks, enzymes, etc.

Given our previous experience with children in hospital, we were a little upset, but also knew it was something we can treat, and he shouldn't suffer with this diagnosis. I suppose that's a parent's greatest fear, we don't want our children to suffer. Our boy is now four years old, in junior infants, the youngest (and toughest) of five boys and I can safely say he hasn't suffered with his diagnosis once in his four years. Although he's had a couple of hospitalisations for IV antibiotics, not once did he complain, we did all the complaining for him.

- PARENTS OF A 4-YEAR-OLD BOY

Our first two years were a constant struggle to try and fight off infection after infection with each sniffle and cough almost always developing into an issue along with a run of very bad luck.

In March 2023 we started her first modulator drug, Orkambi. This amazing drug transformed her life. From the moment she had her first dose, her body reacted very positively to it.

In the whole year of taking this drug, she only needed 1 non-maintenance antibiotic and was able to fight off colds and coughs on her own.

Her older brother had started primary school and a lot of different coughs and colds were brought into the house but never affected her.

In March 2024 she started her second 'Wonder Drug,' Kaftrio. She takes her granules in the morning and evening. This drug has given our little girl everything a parent could ever ask for. She is now well, and able to overcome a cough or cold on her own. She has a fantastic appetite and is a healthy, lucky child.

Whilst we are aware that she will no doubt have a few ups and downs along the way the new medication has protected her and given her the ability to protect herself.

PARENTS OF 3-YEAR-OLD GIRL

Acknowledgements

CF Ireland "Guide for Parents"

Cork University Hospital CF Team

Great Ormond Street Children's Hospital

Crumlin and Temple Street Children's Hospitals.

Royal Brompton Hospital

Parents of Children in the Cork CF Service

Notes

Version 2, December 2024

CUH
Ospidéal Ollscoile Chorcaí
Cork University Hospital

Feidhmeannacht na Seirbhíse Sláinte
Health Service Executive