

Cystic Fibrosis

New Diagnosis Information
For Parents

Advice from other parents

- Don't miss out on just loving and getting to know your new baby.
- Don't feel guilty if you miss a treatment occasionally.
- Take time to build your knowledge. Don't dive headfirst into the net and scare yourself with information that may be outdated, even untrue.
- The progress in Cystic Fibrosis (CF) treatment in the last 10 years is phenomenal and it can only get better and better. Have faith there are much better times ahead.
- When the time is right you will learn more and you will come to a more comfortable place with what is happening in CF treatment and science.

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Congratulations on your new baby!



After some shadowing was apparent on the bowel during the 19 week ultrasound of our second child, my husband and I decided to have an amniocentesis.

Forty-eight hours later, it was confirmed that our little girl (also confirmed) was carrying two genes for cystic fibrosis (CF).

Thus began the whirlwind.

I spoke to my obstetrician, he suggested I speak to a paediatrician at Princess Margaret Hospital (PMH) – who was very supportive, helping us understand that it was OK to feel as if we were grieving, for we in fact were going through a grieving process of sorts.

She made contact with the respiratory team and we were able to speak to a specialist within days, helping us understand what we could expect with regards to care, medication and the prospects for the future. We left the

meeting satisfied that, with support, we were going to be ok. The most important thing that the doctor reminded us was that she was still our girl and that the CF was just part of who she would be.

WOW was he right...

Four years on, Zara is about to begin kindy and is a firecracker.

My pregnancy continued without incident and the adventure began the moment she decided to arrive early, with the theatre erupting to calls of “she’s pooped!!” (As there were fears the shadowing on her bowel may have indicated risk of meconium ileus).

What followed were two precious weeks with her without medication or physiotherapy, before our induction days at PMH, and since then she takes it all in her stride and has always lit up the room.

I try to normalize her treatments as much as possible, like treating percussion time as cuddle (or skin-on-skin) time when she was tiny.

Yes, I still wake most days fearful of what may lay ahead.

Yes, CF treatment takes time.

Yes, it is frustrating always leaving the house weighed down with antibacterial agents.

Yes, it is hard to stay on top of regimens, especially when she is unwell.

Yes, I get tired of people saying, “but she looks so well” - because she does, even when she isn’t.

However, above all else, she is our precious little girl. Zara has been admitted to PMH twice, after weeks of extra physiotherapy and medication have not been able to shift coughs.

She has endured treatments and procedures that would rattle most and come through all of it with a smile and I have found a strength in myself I never knew I had (I now recognize it in other parents of CF kids that I have met).

Zara is already an inspiration to many - including her big brother, and best friend, Evan. He makes sure he reminds us about enzymes and other treatments, and he proudly defends his little sister, making sure anyone with a sniffle keeps

their distance. It has not been an easy journey.

My husband and I have endured many stresses and tears, but with the support of PMH and Cystic Fibrosis WA (CFWA) we deal with what we can to the best of our ability every day and look to the future with optimism.

CF can be an isolating condition but we have tried to “live life unaffected by CF”. We go camping and have taken the kids to Broome, Sydney and the Gold Coast and yes, there are precious moments when we forget altogether that Zara has CF.

Cherish your new little one. Try not to think of what could have been (better or worse). CF presents hurdles, most of them surmountable, but nothing compares to the joy your child brings every day.

Amongst the treatments, there are plenty of laughs, smiles and wonderful moments to keep you going.

Remember, you are never alone.

My thoughts are with you.

Michelle





Cystic Fibrosis

New diagnosis information for parents

Cystic Fibrosis (CF) is an inherited condition that affects many organs in the body. The organs most affected are the lungs and gut.

Although CF is life limiting, advances in our understanding regarding CF and our approaches to management have made life better for people with CF, most of whom will live well into adult life.

CF affects every family differently. Every child with CF is affected differently.

As a parent of a child with CF, you will still get to experience all the normal 'ups and downs' of parenting. Normal life will continue, your child will go to school and join in all normal activities whilst managing the daily challenges of living with CF.

This booklet will help you to learn about CF, and understand how it may affect your child and family. Many parents feel isolated when they receive the diagnosis. Our CF health care team would like to

work in partnership with you, and your child, in managing your child's CF.

Managing CF requires regular appointments to meet with your CF team. If you live in a rural area there are subsidies available to assist in travel required for hospital appointments.

Whom is affected by CF?

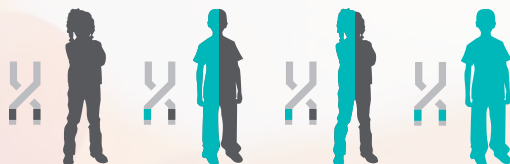
Cystic fibrosis occurs across the world. About 3000 people in Australia have CF including approximately 390 in Western Australia. PMH looks after approximately 200 children aged from birth to 18 years.

How is CF inherited?

In the cells that make up our bodies, each person has a set of instruction packets called genes. Genes determine things such as our eye, hair and skin colour. Every child inherits two sets of genes from their parents, one set from their mother and one set from their

Father
(carrier of CF gene)

Mother
(carrier of CF gene)



Unaffected child
(Not a CF carrier)

Unaffected child
(CF carrier)

Unaffected child
(CF carrier)

Child with CF
(CF carrier)

father. Sometimes variations in the genes occur. An altered gene is changed in a way that alters the instructions it gives.

This variation is often referred to as a genetic mutation. The CF gene carries instructions for the production of a protein called CFTR.

CFTR protein controls the movement of salt and water in and out of the body's cells. Some mutations of the CF gene result in problems with how CFTR works, causing thick and sticky mucus that affects the lungs and digestive system.

When each parent is a carrier of a CF gene mutation there is a:

- 1 in 4 chance of a having a child with CF.
- 2 in 4 chance of having a child who is a carrier of the CF gene.
- 1 in 4 chance of having child without any altered CF genes.

- 3 in 4 chance of having a child that does not have CF.

This is the case for every pregnancy.

How is CF diagnosed?

Newborn screening

Most children are diagnosed with CF as newborns from the heel prick blood test (Guthrie Test). Diagnosing CF from this blood sample is a multiple step process.

The first test will show if a baby has a high level of an enzyme that occurs in CF - immunoreactive trypsinogen (IRT).

High levels of this enzyme can also occur for other reasons. If the IRT enzyme is high, a second test, a DNA test, will look for the most common mutations of the CF gene. Both tests are done with the same blood sample.

If no CF genetic mutations are found, no further testing is done, as CF is very unlikely.

If two CF gene mutations are found on the DNA test, a diagnosis of CF is almost certain but needs to be confirmed with a sweat test. If one mutation on the CF gene is found the child may or may not have CF.

The second gene may be a rare CF gene mutation. In this case, a sweat test will be organized. Fewer than 1 in 10 babies who have a high IRT level turn out to have CF.

Clinical indicators

Heel prick testing does not detect all babies with CF, about 1 in 20 can be missed. Babies born with a bowel blockage due to meconium ileus are likely to have CF.

Your doctor may suspect CF in a child with poor weight gain, oily/greasy poos or regular chest infections.

Sweat test plus/minus DNA testing is required to confirm the diagnosis of CF in these cases.

Sweat testing

Most people with CF have high levels of salt in their sweat. A sweat test analyses a person's sweat for salt content.

- An abnormal (high) sweat test result confirms the diagnosis of CF.
- A normal sweat test result means the child does not have CF.
- Occasionally a sweat test may yield

results in the intermediate range.

This is called an equivocal result. In this case, DNA testing, clinical examination and follow-up may be required to confirm or exclude a diagnosis of CF. Sometimes more than one sweat test is required.

You and your family will be offered an appointment with the genetic counselor.

More about the sweat test:

www.cfwau.org.au/what-we-offer/resources



The effects of CF on the body

Respiratory system (lungs)

The lungs have two main functions:

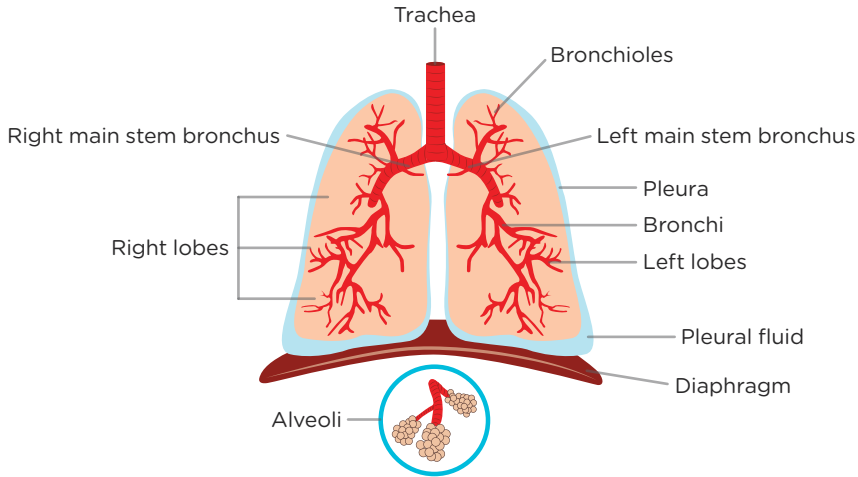
- To get oxygen from the air we breathe in, and get it into our blood.
- To remove carbon dioxide (waste) from the body through the air we breathe out.

When we breathe, air travels through the windpipe (trachea) down the stem of the airways (bronchi), ending at tiny, grape shaped air sacs (alveoli).

These tiny air sacs expand and relax every time we breathe. Small hairs called cilia line the windpipe and airways.

The job of cilia is to move germs and dust out of the lungs. The lungs are lined with a thin layer of liquid (mucus). This liquid assists the cilia to remove unwanted particles including dust and bacteria.

THE HUMAN LUNGS



How does CF affect my child's lungs?

In CF the mucus in the airways gets thick and sticky. This makes it hard for the cilia to do their job (moving out unwanted particles). Germs can get trapped.

Mucus that stays in the lungs can lead to inflammation and infection. Without treatment, this can lead to more rapid lung damage.

How will I know if my child has a lung infection?

It is important to learn the early warning signs that may indicate that your child has a lung infection. Changes to monitor:

- **Cough** - Your child might start to cough more than normal or their cough may sound different. They may start to cough at night.

- **Mucus** - Your child may start coughing up mucus. You may notice the amount and colour is different to normal. Sometimes a mucus sample is collected. This may be referred to as a sputum sample.
- **Wheezing** - Your child may start making a whistling noise while breathing.
- **Difficulty breathing** - Babies may have difficulty with feeding. You may find your child cannot keep up with other children when playing. They may also be breathless when feeding, talking or walking.
- **Decreased energy or lethargy and/or reduced appetite.**
- **Fevers and night sweats can occur**

Some children with CF will not appear to be sick. Changes may still be happening in the lungs of these children, so it is important to continue your treatments and clinic visits even if your child seems well.

What can I do to help keep my child's lungs healthy?

1. Practice good infection prevention

Germs are small organisms that people cannot see but can make them sick. For you and me, most germs are not a problem. These same germs may cause problems for people with CF because they can get trapped in their lungs.

Our team is very concerned about germs that travel between people with CF. We recommend that people with CF do not have close contact with other people with CF, this includes babies. Check with the person in charge that there are no other children with CF enrolled before joining playgroups, child care centres etc.

During clinic appointments, red wristbands are available from the Respiratory Medicine Department if you wish to identify your child as having CF to allow families to minimize close contact with others with CF.

To protect your child from germs you will not need to keep them away from all people. Your child can do most normal activities. They will be able to go to school, the shop, the supermarket, and to the playground. If you have any questions about infection prevention talk to your CF nurse or doctor.

Some simple things you and your child can do to reduce contact with germs:

- Wash your hands well (liquid soap and water) or use alcohol rub
- Teach your child to wash their own hands after each cough, before eating food and after toileting

- Use tissues, throw them away after use and wash your hands
- Cough into your sleeve/elbow
- Stay away from people with colds
- Keep your child's immunizations up-to-date
- Clean your child's medical equipment daily and dry properly
- Do not share eating utensils or cups
- Keep your house dry and aired out
- Do not leave containers of water lying around. These may breed types of germs that can cause lung infections with CF. It's okay to keep water in flowerpots!

2. *Educate family and friends on the impact of passive smoking on your child's health.* Encourage them to seek assistance to stop smoking.



3. Do regular chest physiotherapy

Chest physiotherapy will help your child's lungs by keeping the airways clear of mucus. Physiotherapy will also help your child recover if they become unwell.

Every child will have an individualised physiotherapy program that will include two sessions of physiotherapy a day. Physiotherapy is an everyday activity,



just like brushing your teeth. We like to think of it as lung hygiene.

Even if your child does not have a cough, physiotherapy will help to prevent the buildup of thick and sticky mucus in your child's lungs. It will also help to move germs and dust out of the lungs, reducing the risk of chest infections.

Some of the physio techniques we use are:

- Airway clearance techniques
- Inhalation therapy
- Exercise

Your child's CF physiotherapist will help create an individualized physiotherapy program for your child. As your child grows their physiotherapy program will change to suit their needs. If you have any questions talk to your child's CF physiotherapist.

What should I do if I am concerned about my child's lungs?

During office hours: contact the CF nurses. The nurse will be able to offer you advice on management. If needed, the nurse will arrange an appointment with the doctor. You will also receive a follow up phone call a few days later to see if things are better.

After hours: call the hospital, state that your child has CF, and ask to speak to the Respiratory Physician On-Call.

Digestive system

The functions of the digestive system are to:

- Break down food into smaller parts that the body can then use for fuel
- Remove waste

The digestive system is made up of a long, twisting tube from the mouth to the anus. It also includes the stomach, liver and the pancreas.

The pancreas produces digestive juices (enzymes) that reach the intestine through small tubes. Enzymes break down food so that it can be absorbed. Our body has different enzymes to break down different parts of the food we eat.

The pancreas

The functions of the pancreas are to:

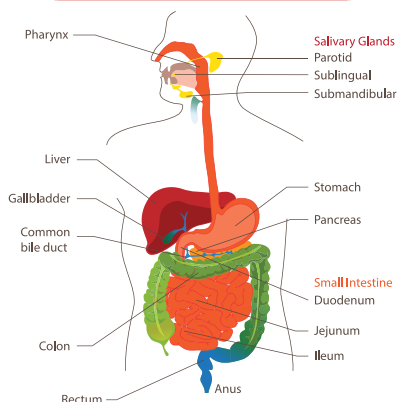
- Produce enzymes that help break down the food we eat
- Control the amount of sugar in our blood

How does CF affect my child's pancreas?

We will test to see if your child's pancreas works well, 85% of those with



• Human digestive system •



CF will have issues with their pancreas. Children with CF who have problems with their pancreas have trouble breaking down their food.

Their digestive system also contains thick and sticky fluid. This fluid blocks the small tubes that connect the intestine to the pancreas.

Digestive enzymes cannot work when these tubes are blocked. People with CF may need to take extra enzymes (granules or capsules) to be able to absorb fat and fat-soluble vitamins.

The dose of enzymes will change as your child grows and as their diet changes,

so check regularly with the dietitian to review enzyme doses, including babies who are breast fed. We want to manage this well so your child grows normally.

What can I do to help my child grow?

It is important to learn the warning signs that may indicate that your child has problems digesting their food.

Changes that you might notice or should monitor:

- Tummy pain
- Discomfort
- Excessive smelly wind
- Greasy, fatty poos
- Diarrhea and/or constipation
- Hunger despite eating lots of food
- Poor weight gain, poor growth

Your dietitian and doctor will work together with you, to work out the best plan to improve your child's food absorption. This will help ensure optimal nutrition and weight gain.

Your dietitian will also be able to advise you on the type of food your child should eat (usually a high energy diet). This will help ensure normal nutrition and weight gain.

The liver

Not all children will have liver involvement. The liver is a very complex organ, and it is the largest organ in the body. It is located in the upper abdomen on the right, up under the lower ribs. It has multiple functions, some of the most important include:

- Cleaning the blood
- Creating and releasing bile that helps digest fat from your food
- Controlling blood clotting
- Helping our body fight infection
- Storing iron

How will I know if there is something wrong with my child's liver?

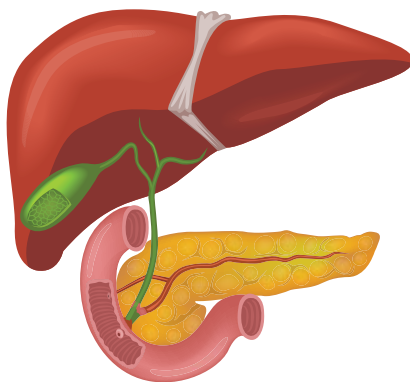
When there is early damage to the liver, you may not notice anything different. As the liver damage worsens, some of the following things might happen:

- Enzyme capsules not working as well as they had been
- Poor appetite
- Enlarged tummy
- A yellow colour may develop in the whites of your child's eyes or skin
- Itchy skin

If you are concerned about your child's liver talk to your CF nurse.

The sweat glands

Our bodies have many sweat glands all over our skin. These sweat glands make sweat to keep our body at a normal temperature (around 37°C). Sweat is mostly water, but also contains some salt (sodium chloride).



When a person sweats, the moisture on their skin evaporates. This cools the body.

How does CF affect my child's sweat glands?

Sweat is usually very salty in people with CF. A person with CF loses large amounts of salt and water if they sweat too much. This can lead to dehydration.

This is a bigger problem over summer with the hotter weather. It is also a problem with exercise, fevers, or infection. Water loss makes your child's mucus thicker and harder to cough up.

How will I notice if my child has not had enough salt (dehydrated)?

People with CF lose about 2-5 times as much salt in their sweat than other people. Loss of salt can cause dehydration and also make the mucus in the lungs more difficult to clear.

You should always consult with your baby's CF care team about his or her individual salt requirements, as their needs will differ depending on factors

such as symptoms, body size, dietary intake and the climate you are living in.

Salt supplementation is important for breastfed babies as human milk has a lower salt content than formula. Salt supplementation is required in WA all year round.

For infants, salt solution is usually recommended. This can be added to expressed breast milk or formula or can be syringed straight into the mouth.

Signs of dehydration

If your baby is lacking salt they can become dehydrated. Signs of dehydration include:

- Fewer/lighter wet nappies than usual
- Dark sunken eyes
- Dry skin or lips
- Tear less crying
- Dark yellow urine
- Lethargic and drowsy
- Rapid breathing
- Salt crystals on the skin

If you think your baby is dehydrated contact the CF clinic. Try giving them smaller amounts of milk feeds more frequently.

Medications

Assistance with costs of medications will be available to you as your child will be eligible for a Health Care Card. The pharmacist will explain the Pharmaceutical Benefits Scheme (PBS) that offers subsidized medications. Your Social Worker will be able to help you with the application for a Health Care Card.

Reproductive system

How does CF affect the reproductive system?

MALES

Men with CF make sperm normally. However, most men with CF will be infertile. The tube that carries the sperm from the testes to the penis is often blocked or missing, therefore the sperm gets reabsorbed. Most men with CF will require medical help to have babies.

FEMALES

Women with CF have a normal reproductive system, however their fertility may be variable.

The only difference in CF is that the mucus in the cervix is thick and sticky. This can make it hard for the sperm to reach the egg. However, many women with CF have no problems falling pregnant.

Future pregnancies


Reproduction and sexuality is often a difficult subject to talk about with your health care team. You may want to ask questions regarding future pregnancies and the genetic implications for your extended family.

For further information contact:

Genetic Services WA, King Edward Memorial Hospital, (08) 9340 1652

For information on carrier screening for family members go to:

www.cfscreening.com.au/



Who will be there to help me?

WA has a team of highly qualified health professionals that will help you keep your child healthy. Children with CF need to visit the clinic every three months to see all members of the health care team.

If your child is unwell, we are able to see them more often. Our team will be working with you and your child until your child reaches adulthood when they will be transferred to adult care.

Feel free to ask any member of the CF team any questions you need answers to.

The CF health care team

PARENT AND CHILD

You and your child are valued members of our health care team. You have a unique view on how CF affects your child and everyday life. We create treatment plans with your input.

CLINICAL NURSES

The CF nurse is the primary contact person for questions and concerns. CF nurses work at PMH and at Sir Charles Gardiner Hospital (SCGH) for adults.

You can contact the PMH CF nurses during office hours by calling the Respiratory Medicine Department: 9340 8830

If you are concerned about your child outside normal business hours, please ring PMH on 08 9340 8222 & ask to speak to the respiratory doctor on call or go to our Emergency Department (ED).

The CF nurse will help you to navigate the health care system and promote your child and family's views about health. He/she will also teach you how to keep your child in a safe environment.

Your CF nurses oversee outpatient and inpatient admissions in WA. They always make sure that the CF team is providing the best care available for your child.

RESPIRATORY PHYSICIANS

There are a number of respiratory physicians working within the clinic at PMH. When your child is diagnosed with CF you will have one main physician who will oversee your child's care, called a primary physician.

This doctor will be involved in making major decisions regarding your child's care. During your clinic visit your child



may be reviewed by another member of the medical team. If any major decisions are to be made, where possible, your child's primary physician will be involved.

GASTROENTEROLOGIST

We have a Gastroenterologist attached to the CF team and your child will be seen by the Gastroenterologist usually at diagnosis and then at least every year. Some children who have problems with their digestive system are seen more frequently on the same day as their respiratory clinic appointment.

DIETITIAN

The dietitian will work with you to make sure your child gets the fuel they need to grow normally. He/she will teach you about the specialist nutritional needs of most people with CF. *These often include:*

- Pancreatic enzymes
- A high energy diet
- Extra salt to prevent dehydration

PHYSIOTHERAPIST

The physiotherapist will teach you a variety of techniques to keep your child's lungs healthy.

These include:

- Airway clearance techniques
- Inhalation therapy
- Exercise

They will provide you with an individualized physiotherapy program for your child. The physiotherapist will see you at clinic reviews to monitor how your child is going with their physiotherapy program. The physiotherapist will also assess your child's posture and development.

SOCIAL WORKER

The social worker will meet you and learn more about your particular family. He/she will assist you in your adjustment to your child's diagnosis of CF. Each family and child are unique, and have different strengths and challenges.

The majority of our families adjust to the emotional challenges and practical tasks required to parent a child with CF within the first six months. Of course, there will be stressful times for all families along the way.

The social worker will assist you with any challenges that may limit your ability to parent or manage the CF care of your child. They will also assist with information and resources such as

Centrelink benefits, and Patient Assistant Transport Scheme (PATs) for country families. This support continues until your child is 18.

CLINICAL PSYCHOLOGIST

Our Clinical Psychologist is available to assist your child with any CF related psychological concerns from birth to age 18. You will meet the clinical psychologist briefly as part of your education. He/she is also available to work with parents if anxiety, depression and/the impact of the diagnosis is affecting the enjoyment and relationship with your baby.

PHARMACIST

The [pharmacist will help you](#) understand your child's medications, including how and when to take medications and what assistance is provided by the government towards the cost of medications. They will explain the Pharmaceutical Benefit Scheme (PBS).

Other useful health professionals

GENETIC COUNSELOR

The genetic counselor can help you understand why your child has CF.

They can assist you and other family members with carrier testing, and will provide you with information about your options for future pregnancies.

GENERAL PRACTITIONER

Your child's general practitioner (GP) is a key member of your healthcare team. Infants with CF need basic medical care as well as CF care. Your GP will take care of some of your child's health needs.

The CF team will provide care specifically related to CF. You will receive individual information on when you should contact your CF team and when to contact your GP.

CHILD HEALTH NURSE

The child health nurse will complete your child's growth and development assessments.

FAMILY AND FRIENDS

Encourage close family members to become familiar with your child's treatment. This can be valuable if you are unwell. Some grandparents / family members may assist with airway clearance so in time you may be able to have a break.

PARENTS OF OTHER CHILDREN WITH CF

There are other families out there, who have been through the same thing. If you would like be in touch with other CF families ask your CF health care team or CFWA. For infection control reasons the best way to contact other CF families is by phone or internet.

CYSTIC FIBROSIS WESTERN AUSTRALIA (CFWA)

CFWA offers a range of services for CF member families. Please refer to the CFWA membership link

www.cfw.org.au/get-involved/become-a-member

A community nurse or physiotherapist from CFWA can visit at home.

Immunizations

Everyone should have the immunizations recommended by the Australian Government. These will help your family prevent the flu and other infections. Children with CF should have their immunizations according to the normal schedule. Children with CF also need:

- Pneumococcal vaccine

- Influenza (annual flu shot)

Pneumococcal Vaccine

At 12 months of age, all children with CF should receive a booster of the pneumococcal vaccine. There is a space to record this on the personal health record. When your child is four or five, they should also have a further booster dose. This booster dose is a slightly different type of pneumococcal vaccine.

These booster doses will help prevent your child from developing infections caused by the pneumococcal organism (bacteria).

Influenza (flu) Vaccine

The influenza vaccination is often referred to as the 'flu shot'. The flu (influenza) can be life threatening for people with CF. All children with CF should receive this vaccine from 6 months of age and they should have it every year just before the flu season.

Each year, the most common viruses that cause influenza are made into a vaccine. Having this vaccine will not prevent your child from the usual coughs and colds, but will help prevent a serious illness.

We recommend that all family members and close contacts also have the annual influenza vaccine. The more people protected against the flu, the less risk of your child getting sick.

Please review the infection control section on *page 9* which offers some helpful hints on how to protect your child from germs. If you have any concerns or questions about immunizations, please talk to your CF team, GP or immunization nurse.

Personal adjustment

Your child

Having CF will affect every family and child differently. Your child will be able to do most things. Our team would like children with CF to:

- Lead a normal and active life.
- Have a positive and hopeful attitude to life.
- Develop the skills to manage their life by themselves (including their CF).
- Go to school regularly.
- Get a job.
- Plan for their future, including positive relationships and having a family of their own.

Parents

In time, you will get to know how CF affects your child and family. You will most likely feel overwhelmed at first. It may seem that only your child's health care team has the answers.

As a parent, you will know if there is something wrong with your child. Your CF health care team wants to know your concerns. We will always take these seriously no matter how small you think they are. Feel free to ask us questions and learn more about CF.

There may also come a time when you feel sad or angry. This can happen at any time in many different ways. Everyday living can seem more difficult.

There are people on our team that can talk to you about this and offer you some advice. You may need to seek help from your GP.

If you are feeling overwhelmed take some time to look after yourself. If you are not coping, it will be harder to look after your child.



Please Remember:

The PMH Pediatric Cystic Fibrosis Team is always here to support your child and help your family manage the daily challenges of living with CF.

CFWA newly diagnosed support services

Become a member to access these services—follow the link—membership is free;

www.cfwaweb.org.au/get-involved/become-a-member

HOME CARE WORKER (HCW) SUPPORT: Metro & Bunbury:	HCWs provide in-home airway clearance support, exercise programs, occasional respite and home duties. All workers receive ongoing training and supervision by the CFWA physiotherapist. The CFWA physiotherapist will liaise with your child's primary PMH physiotherapist to ensure that the support provided by the HCW is the same as your child's regular physiotherapy program.
SOCIAL WORK & GENERAL SUPPORT:	Having a newborn diagnosed with CF can raise some emotional issues for families. CFWA can provide general support, social work, nursing and physiotherapy support.
THE COMMUNITY NURSE:	Can offer support in the early weeks by meeting with you in the comfort of your home to answer questions, offer support information and individual care advice given at diagnosis. The nurse can assist in establishing a routine in consultation with the PMH CF team.
PHYSIOTHERAPY:	In home and phone support.
EQUIPMENT:	CFWA have a variety of equipment available for loan such as nebulisers and exercise equipment.

EDUCATION, INFORMATION & NETWORKING:	<p>Education sessions are available for extended family, daycare services, schools, community groups both in the metro area and regionally www.cfsmart.org</p> <p>Education resources: CFfood: nutrition information. A range of fact sheets on various CF related topics www.cfwa.org.au/what-we-offer/resources</p> <p>Seminars and other education events are held throughout the year</p> <p>RED E-news: email containing latest news and information from CFWA (fortnightly)</p> <p>RED Magazine: with latest research, stories and news (quarterly)</p> <p>Rozee Magazine: for CF children and siblings (annual)</p> <p>Facebook Page: https://www.facebook.com/CysticFibrosisWA</p> <p>To become a member: www.cfwa.org.au/get-involved/become-a-member</p>
RECREATION & SUPPORT EVENTS:	<p>Keep up to date with CFWA's recreation and support events. www.cfwa.org.au/what-we-offer/events/</p>
SUBSIDIES:	<p>(www.cfwa.org.au/what-we-offer/events)</p> <p>Country Patient Travel Subsidy (regional members only)</p> <p>Patient Support Subsidy to assist with equipment, exercise and other expenses.</p>
REGIONAL SUPPORT:	<p>Country regions are visited as required to provide education to schools, community groups and health professionals, and link up members within their local area. www.cfwa.org.au/what-we-offer/</p>
COMMUNITY FUNDRAISING EVENTS:	<p>A number of community fundraising events are held throughout the year to raise awareness and funds for specific programs. www.cfwa.org.au/get-involved/fundraising</p>
CONTACT DETAILS:	<p>Phone: 08 6457 7333</p> <p>Fax: 08 6457 7344</p> <p>Email: info@cfwa.org.au</p> <p>Address: The Niche Building, Suite C, 11 Aberdare Road, cnr Hospital Avenue Nedlands, WA 6009</p> <p>www.cfwa.org.au</p>

Useful Definitions

Alveoli: Tiny air sacs in the lungs.

Bile: A digestive juice, that helps digest fat, secreted by the liver.

Bronchi: Small airways in the lungs.

CF Gene: A gene we all have. This gene provides information so that the body can transport sodium and chloride in our cells.

Cilia: Tiny hair like structures that line the windpipe and airways.

Cystic Fibrosis Transmembrane

Reactive Protein (CFTR): Protein that controls the movement of salt and water in and out of the body's cells.

Dehydration: Losing too much water and salt from the body.

Deoxyribonucleic acid (DNA): A substance that holds genetic information in the cells.

Enzyme: Usually a protein that can cause, help or speed up a reaction. Enzymes play many roles in our bodies including digesting food and transmitting nerve impulses.

Genetic mutation: An altered form of the CF gene we all have. Two copies of this faulty gene results in an individual having CF.

Gene: A set of instructions that decide things such as eye, hair and skin colour.

Germes: Organisms that can cause infections, like bacteria, viruses and fungi.

Heel prick blood test (Guthrie Test):

A heel prick is a common procedure for taking a blood sample from the heel of a newborn infant. The blood collected is then tested for a variety of genetic conditions (including CF).

Immunisation: A vaccine given to protect children, adolescents and adults against harmful germs.

Immunoreactive trypsinogen (IRT): An enzyme that is elevated in babies with CF. This test is done on the heel prick blood test (Guthrie Test).

Influenza: An acute, highly contagious infection of the respiratory tract, often called 'the flu'.

Infection: The invasion and multiplication of microorganisms (germs) e.g. bacteria and viruses that are not normally present in the body that hurt the body.

Inflammation: The body's response to an infection or injury. In the lungs, airways become swollen and narrow.

Inherited (genetics): Personal attributes

acquired when parents pass on genetic information in their DNA to their child.

Intestine: A tubular passage connecting the stomach to the anus. This is where digestion of food happens.

Liver: A large and complicated organ of the digestive system. Functions include: storing blood, creating bile to help with fat digestion, controlling blood clotting, fighting infection and storing iron.

Meconium ileus: A type of bowel blockage commonly seen in newborns with CF. The baby's first stool is thick and hard, blocking the small intestine.

Mucus: A thin layer of protective liquid. In the respiratory system, this liquid helps germs from penetrating the cells below. In the digestive system, it lubricates a passage for food, as well as protecting the cells.

New born screening test: See 'heel prick blood test'.

http://healthywa.wa.gov.au/Articles/U_Z/Your-newborn-babys-screening-test

Pancreas: A gland that sits behind the stomach that secretes enzymes and a hormone (insulin) that help to digest food.

Sputum Sample: Thick mucus that is expelled from the lower respiratory tract (bronchi and lungs) through coughing; it is not saliva or spit.

Stomach: A hollow muscular organ of the digestive system. This organ stores food in the process of digestion.

Sweat glands: Tiny structures in the skin that secrete sweat. Their function is to keep the body at a normal temperature (37°C).

Sweat test: A test that measures the concentration of chloride and sodium that is lost during sweating. This test is used to confirm a diagnosis of CF.

www.cfwa.org.au/what-we-offer/resources

Trachea: The windpipe leading from the throat to the lungs.

Vaccine: A preparation that helps to improve immunity to a particular disease.

Recommended websites:

www.cysticfibrosis.ca

www.cysticfibrosis.org.uk

www.cff.org



In the future

Do you plan to have another baby?

Your reproductive choices

You may be thinking about having another child, but, you know that there is a 1 in 4 chance that your next baby could also have CF. What do you do?

Your choices are to:

Conceive naturally and 'take the risk' so to speak. There would be a 1 in 4 chance that the baby would have CF. Testing of the new baby could be arranged at birth, using blood from the umbilical cord. You would know if your baby had CF within two weeks of the baby's birth.

Conceive naturally and have pre-natal testing. This can be done by chorionic villus sampling (CVS). This happens at around 11 weeks into a pregnancy.

A needle is passed through the wall of your uterus to remove a sample of the baby's placenta. The cells from the baby's placenta are then DNA tested. The test comes with a small risk of causing a


miscarriage (about 1 in 100). If the baby was found to be affected you would have the option of continuing with the pregnancy or terminating the pregnancy.

Pre-implantation genetic diagnosis (PGD). PGD is performed on embryos conceived by in vitro fertilization (IVF). When the embryos are very small (8 cells), one cell is removed from each embryo.

These cells undergo a DNA test to see which embryos have CF. Embryos with CF are discarded. Unaffected embryos can be implanted into your uterus.

As with all IVF procedures, there is no guarantee of a pregnancy. If private services are used you will need to research the costs carefully.

Use a sperm or egg donor. Although you can use an egg donor, it is much easier to use a sperm donor. The aim here is to avoid both parents being carriers of the CF gene.



Artificial insemination is available through private fertility practices.

All sperm and egg donors are screened for the most common CF genes.

Use a donor embryo. Donor embryos are sometimes available. Couples undergoing IVF may choose to donate their embryos, if they have more than they need.

Obviously, there is no 'right' choice here. Only you and your partner can decide which option is the right one. Talk to a genetic counselor at Genetic Services WA (08 9340 1525) for more information.

All discussions you have with a genetic counselor are confidential. Information will not be passed onto the CF team without your permission.

Disclaimer

This publication has been prepared solely as a general guide for use by patients and prospective patients in relation to information given to patients in hospitals managed by the Government of Western Australia and the Department of Health.

Although all due care has been taken in the preparation of the publication, it is only to be used as a guide and is not a substitute for advice from your surgeon or other medical expert who is treating you.

Possibilities

Each child with CF walks a different path, and only time will tell what your child's journey will be, try not to look too far ahead and don't be disheartened by stories you might hear or read.



Harry Coffey
20 yrs, jockey,
Swan Hill, Victoria

Nathan Charles
Western Force NRL



Holly Nicolas

17 yrs, Table Tennis
champion No.1 ranked
U.18 Girl in Oceania
and Australia.



Joseph Iaria

18 yrs, State basketball
league player,
Geraldton, WA



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Child and Adolescent Health Service

www.cfwa.org.au

