Cystic Fibrosis

Information for schools and the community

Cystic Fibrosis WA

community

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Introduction

Having a child with cystic fibrosis in your care means that you are interacting with a child who has a chronic illness, a condition that has no effect on intelligence, does not interfere with a child's ability to participate fully and is not contagious.

You can have the same expectations in the classroom or social setting of a child with cystic fibrosis as you would of any other child.

There is no typical child with cystic fibrosis. The disease affects each child in different ways with varying degrees of severity, and each child's health can change considerably from month to month or day to day.

Almost all children with cystic fibrosis show some symptoms of the disease. Typically they live with mild, moderate or even severe lung disease and gastrointestinal problems. Children with mild or moderate cystic fibrosis usually appear healthy, but this appearance can be very misleading.

The best way to help children with cystic fibrosis at school or in the community is to treat them as individuals and to be receptive to their changing needs.

This booklet has been designed to assist teachers, teacher assistants, staff in primary and secondary schools, daycares and other community groups as well as other parents, to understand cystic fibrosis and how it affects the child concerned.

What is cystic fibrosis?

Cystic fibrosis is one of the most common life-threatening, recessive genetic conditions in Australia. It primarily affects the lungs, digestive system and sweat glands. CF can also affect the reproductive system in males and to a small degree in females.

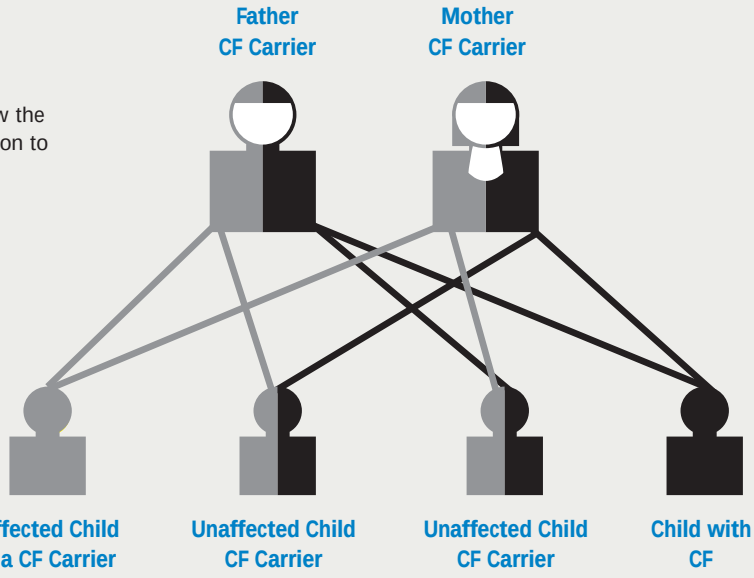
Cystic fibrosis (CF) is not curable at the moment, however a great deal of resources are being directed towards finding new and improved ways of treating it and working towards finding a cure.

CF is a condition in which there is considerable variation in the severity of symptoms. CF was first recognised as a specific condition in the 1930's at which time most babies born with the condition had a very short lifespan. Today with earlier diagnosis, greater understanding of the condition, improvements in treatment and better management, the majority of children live well into adulthood, many leading independent and successful lives.

How common is CF?

- In Australia there are approximately 80 babies born with CF each year.
- The incidence is 1 in every 2500 births.
- 1 in 25 people are carriers of the CF gene
- There is a 1 in 400 chance of two carriers forming a relationship
- If two people are genetic carriers for CF and they have a child, there is a 25% chance that their child will have CF.
- There are approximately 3000 people with CF in Australia.
- The are approximately 300 people with CF in Western Australia
- 50% of the CF population in WA are adults
- CF is most common amongst Caucasian races.

A diagram showing how the CF gene can be passed on to offspring



cystic fibrosis

How does cystic fibrosis affect the body?

A person who has cystic fibrosis will be missing a functional protein from his or her cells. This means that chloride and sodium cannot be properly transported across the cell membrane. This causes mucus (which everyone has in their body) to become thicker and stickier than normal. The mucus then builds up in organs like the lungs and pancreas. Other complications from CF may include fatigue, chronic sinusitis, asthma, late onset of puberty, CF-related diabetes, liver cirrhosis and male infertility.

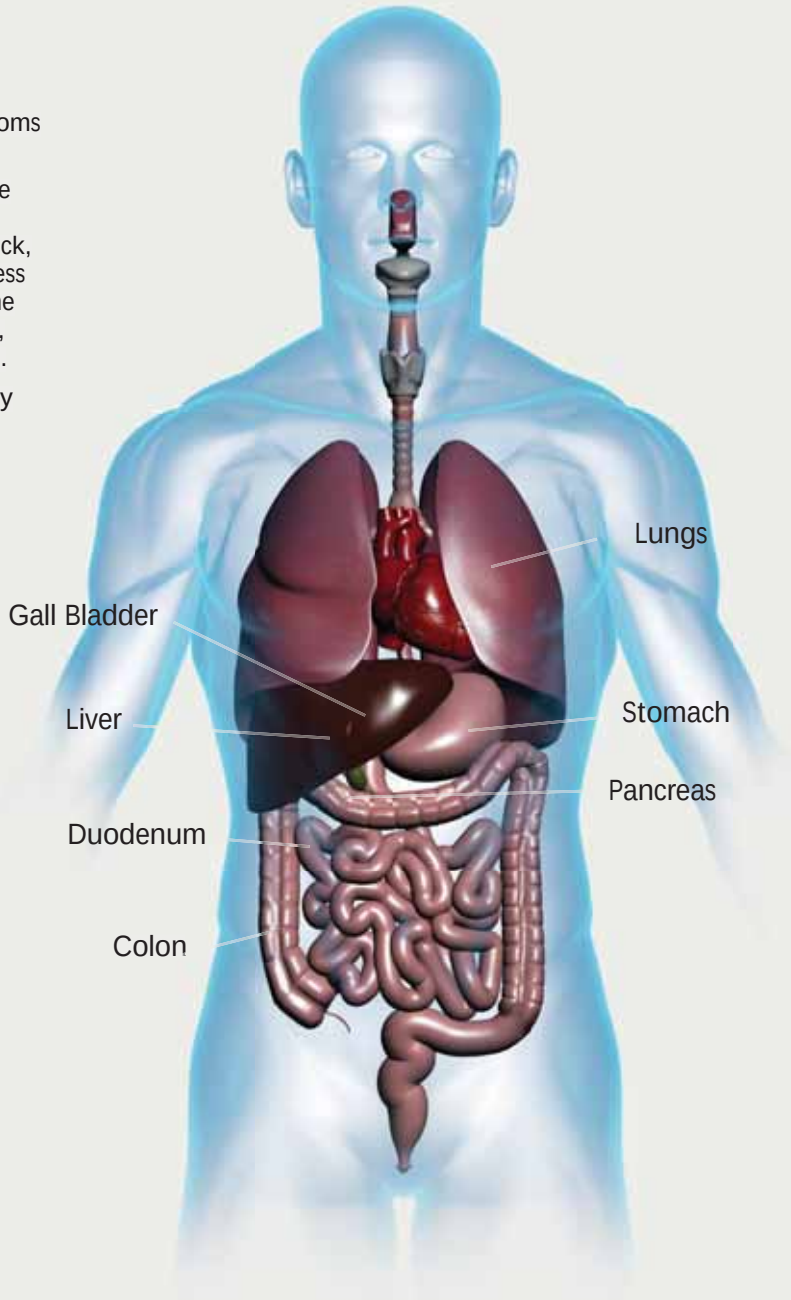
Respiratory system: lungs

The effects of cystic fibrosis are very individual. Not all people with CF have the same complications or symptoms or require the same care.

For the majority of people with CF, complications in the respiratory system are the most serious. Respiratory failure is usually the most common cause of death. Thick, sticky secretions interfere with the body's natural process of clearing infectious material away from the lungs. The mucus clogs the breathing passages and if not cleared, can lead to recurrent lung infections and lung damage.

A child with CF may have the following respiratory symptoms;

- chronic coughing (not contagious)
- mucus (sometimes tinged with blood)
- shortness of breath or wheezing (due to low lung function)
- pale appearance
- frequent respiratory infections or flu



Digestive system: pancreas

The pancreas is a gland just below the stomach and one of its functions is to produce enzymes which break down food so that it can be absorbed by the digestive system.

With CF the sticky mucus can make it difficult for the enzymes to reach the small intestines to break down food so nutrients can be absorbed. As a result fats and proteins are poorly absorbed and this is known as malabsorption. Children and adults with CF usually have great difficulty gaining weight. **A diet high in fat is recommended for a child who has CF.**

The pancreas also produces insulin. Later on, some people with CF may require insulin supplementation, without the need to change their diet. Symptoms of CF related diabetes may include thirst and frequent urination.

A child with cystic fibrosis may have:
- excessive or poor appetite
- poor weight gain, small size and distended belly
- occasional flatulence and stomach cramping
- foul-smelling, excessive or urgent stools
- a need for extra toilet privileges and extended time needed in the toilet

About 10% of people born with CF have relatively normal functioning digestive systems.

Sweat glands

People with CF lose **3-4 times more sodium (salt)** through the sweat glands than those without CF, so their daily need for this essential mineral is **high**, approximately 4000-6000mg minimum daily for adults, although this varies between individuals.

People with CF require a diet high in salt in order to limit the risk of dehydration and they need frequent drinks of water or salty drinks such as Gatorade.

Salt depletion can happen very quickly during a hot day with prolonged exertion. Drinking plenty of water and taking salt supplements is important especially during and after sports activities.

Symptoms of dehydration and salt depletion include:
- loss of appetite
- tiredness, listlessness
- grumpy or irritable
- difficulty concentrating
- headaches
- cramps
- thirst
- sunken eyes
- nausea

The main diagnostic test for CF is the sweat test which measures the level of salt in the sweat. If you were to lick the face or neck of a person with CF they would taste very salty!

Treatment for cystic fibrosis

The treatment for cystic fibrosis involves a team of medical and allied health professionals including doctors, nurses, physiotherapists, dieticians, pharmacists, psychologists and social workers. In between periods of hospitalisation and clinic visits, families take on the role of managing routine treatment and therapy. Once a child reaches adolescence he or she is encouraged to manage their daily home treatment independently.

Treatment for cystic fibrosis can be intensive and time consuming. It is ongoing and lifelong. At present, treatment is aimed at slowing the progression of the condition. The type of treatment required may vary during the year, depending on whether the child is having an exacerbation (a worsening of lung condition) or a "well" period. Children or adults with CF may be on anti-inflaminmatory medications, antibiotics, steroids and/or bronchodilators. These medications may be given orally, intravenously, through metered dose inhalers or in aerosol form using a small-volume nebuliser.

Treatment for the lungs

Each day, most people with CF follow a treatment routine to control the accumulation of potentially harmful mucus on the lungs. Since CF affects each individual differently, the therapy prescribed for one child by his/her physician may not be appropriate for another person with the same condition.

The treatments that help to clear abnormal mucus include airway clearance and inhalation of medications. In airway clearance, a mixture of percussion, vibes and breathing exercises are used to clear the lungs and dislodge small mucus plugs in the airways. Chest therapy is usually performed twice a day; once before school and perhaps upon arriving home or before bedtime.

Increased physical education participation is medically beneficial, even though some children may cough a lot. This coughing helps to remove the mucus from their lungs so exercise tolerance may be decreased and needs to be taken into consideration.

Intravenous (IV) medications

Most children or adults with CF periodically require intravenous (IV) medications in addition to their daily health regime. Quite often the child's parents are able to administer the IV antibiotics at home, through a Peripherally Inserted Central Catheter (PICC) line in the childs arm. The line is sealed and covered by a bandage which enables the child to continue at school instead of staying in hospital.

Hospitalisations

Most adults and children with CF in Western Australia attend regular clinic sessions at Princess Margaret Hospital (for children) and Sir Charles Gairdner Hospital (for adults). It is also common for people with CF to visit the hospital for a two week period for what is often called a "tune up". During the two weeks the person will receive extra IV antibiotics, chest therapy and their diet may be assessed by a dietician.

Digestion

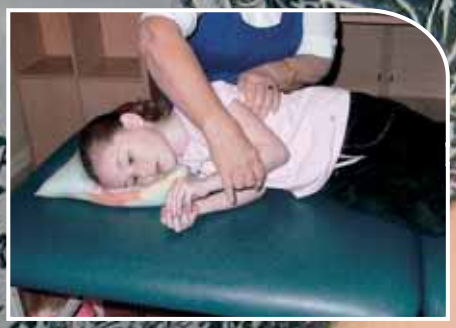
To improve absorption of food most people with CF require enzyme replacement capsules with meals and snacks. These enzymes pass into the stomach and act like the body's natural enzymes by breaking down food and increasing absorption in the small intestine.

Enzymes are not drugs, they are supplements. Enzymes need to be taken before eating a meal. They are not dangerous to other children.

To improve nutrition and maintain body weight a diet higher in fat and calories is required. Some people may also require other supplementary vitamins.

Gastrostomy tube

Some children have a gastrostomy tube to help with severe malnutrition. This is a feeding tube that goes directly through the abdominal wall into the stomach. A plastic button sits on the outside of the abdomen. Liquid nourishment and certain enzymes can be administered through this button. This is rarely done when the child is at school. Please note that in some instances the tube can become dislodged.



Enzyme capsules

A child or adult with cystic fibrosis may take capsules containing pancreatic enzymes with their meals and snacks. These enzymes help the body to absorb nutrients from food and to reduce the number and bulk of stools, the amount of flatulence and abdominal pain and distension.

Most children who need to take enzyme capsules have been taking them since infancy and can take them on their own. There is no need to lock these medications in a desk or store them in an office, however they do need to be kept in a cool, dry place under 25°C. In most cases, depending on the child's age and maturity, he or she can carry the day's supply in their lunch box and take them with their lunch and snacks. Parents should be responsible for providing the number of capsules required and a proper storage container to keep them in.

Children with CF are encouraged to manage their own pancreatic enzyme replacement therapy once they reach the age of six or seven, depending on the child. Young children may require a level of supervision to ensure they take their enzymes at the appropriate times.

The enzyme capsules are not habit forming or life threatening if taken by another child and will not alter the child's attitude or emotional behaviour. A person with CF may need to make a quick exit to the toilet and flatulence can be odorous and embarrassing.

The capsules are only effective for 30 minutes. If a meal is longer than 30 minutes additional enzymes must be taken. (It is preferable to take the capsules prior to eating and to finish the meal within half an hour, but they can be taken after the meal)

If enzymes are not taken, the immediate consequence is reduced absorption of food resulting in stomach pains, wind and diarrhoea.

The dose of enzymes required depends on the individual and the type of food eaten.

The way that parents calculate how many enzyme capsules to give to their child before a meal depends on the strength of the capsules. They come in strengths of 5000 which equates to 1 capsule per 4g of fat or 10000 which is 1 capsule per 8g of fat.

The capsules are only to be consumed with food that contains fat and proteins. There is no need for a child with CF to take them with the following foods:

- fruit juices
- jelly
- sugar
- cordial and soft drinks
- fruit
- salad and leafy vegetables
- lollies
- icy poles
- jam and honey

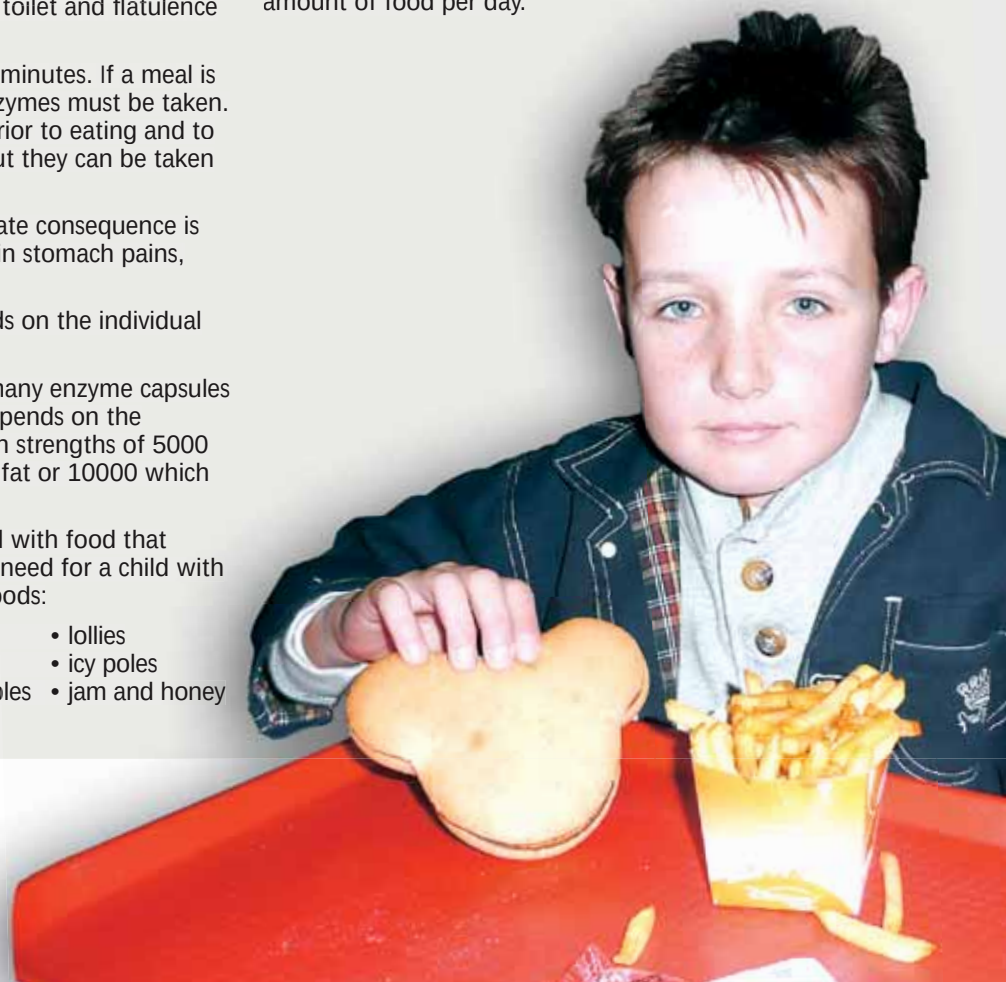
Some children and adults prefer to take their enzyme capsules with a drink.

Be aware that some children with CF, may feel embarrassed about taking their capsules in front of their classmates at school, or friends and may 'forget', hide or throw away their enzyme capsules or miss their meals.

Good communication with the parents of a child with CF is essential to put their minds at ease – it can be quite stressful managing their child's daily medication and CF treatment.

If a child takes too many capsules for the amount of food consumed in a couple of instances it is not dangerous, but in some cases, this could be harmful over a long period of time.

If the child doesn't eat their food or snacks they do not need to take the capsules, however parents should be notified of this, as a child with CF has to eat a certain amount of food per day.



There are a number of different ways that parents communicate with teachers and their children as to how many enzymes to take with which foods, while at school.

- Some parents write on labels the amount of enzymes needed and stick them on to the appropriate food items.
- Some parents may use a communication book, where the child or teacher (or both) record what was eaten and how many capsules were taken.
- Some children are able to take their capsules independently and require very little assistance. This should be encouraged.
- Some parents may provide a short list of the typical food items that a child may eat at school eg. ham and cheese sandwich, chocolate bar and packet of chips. Next to the different food items there will be the amount of capsules needed.
- Sometimes a teacher's assistant will monitor the student while he or she takes their capsules.
- Please take the time to discuss a plan with the parent as to what type of system will work for their child and for you as the teacher and your school.
- Consider what you will do if your student eats their lunch in an open area where there are different teachers doing lunch duty each day. How will you inform any relief teachers about the routine? Ask the parent about class parties, how many capsules will be needed?

Enzymes for babies:

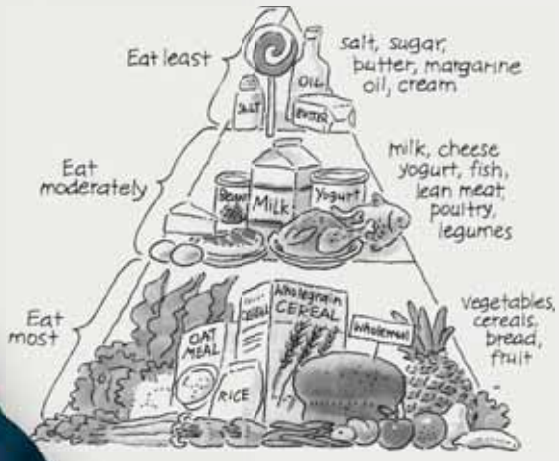
- To give enzymes to babies and young children who are not able to swallow capsules, parents open the capsules and mix the granules with apple puree or fruit gel. They use up to ½ a teaspoon of apple to cover the enzyme granules.
- It is advisable to check the baby's mouth and gums for left over granules at the end of each feed and remove them. Granules left in the mouth after a feed can cause mouth ulcers.

The cystic fibrosis diet

People with cystic fibrosis require extra calories for energy because of the demands that CF places on the body. The lungs need to work harder and the nutrients consumed are not always absorbed properly. Therefore people with CF are encouraged to eat a high energy, nutritious diet following the Dietary Guidelines for Children and Adolescents with CF.

The energy requirements of a person with cystic fibrosis can be 20-50% more than people of the same age to achieve normal growth. This does not mean that an adult or child with CF has to consume 20-50% more food in their diet, they need to find ways of adding extra calories or kilojoules into the foods they normally eat (such as adding more butter, cheese or cream to foods).

Children attending school often have great lunchboxes filled with chocolates and chips, which can be difficult in a school that follows a structured healthy eating program.



Healthy Eating Pyramid

Cystic Fibrosis Food Cube
From "More Please" (2007)

Social and psychological issues

Cystic fibrosis is a serious chronic illness. Like any other chronic illness, CF can cause social, emotional, and psychological problems. To a varying degree, CF may affect everyday life and activity. Regardless of the health of the individual, the mere fact that they have CF will influence and may even prejudice opportunities.

Children with cystic fibrosis may have side issues, such as; low self esteem, school failure and associated behavioural problems. Children with CF are often underweight and small for their age and along with persistent coughing and breathlessness during sport, they may be a target for teasing and bullying. Educating the school, sports team, or friends and family on the effects of CF with permission from the child and parents may help to alleviate this problem.

It is also helpful if schools who have healthy eating programs, acknowledge that some people have to follow a different eating plan to the healthy pyramid model to maintain their health. It can be very confusing for a child with CF who is told by their parents and doctors that they need to eat more fat and salt, whereas at school they may be told to cut down on foods with high salt and fat content.

Parents' ways of coping with children who are affected by CF differ as widely as the condition itself. Men and women also respond differently to how they live with a child who has CF. The whole family will be affected by the psychological pressures arising from the chronic nature of CF, the uncertainty about the future, genetic aspects, worry, depression, the tiring routines of physiotherapy and supervising medication. Although medical advice, support and counselling is available, the pressures of coping with CF places enormous strain on relationships and family life.

The diagnosis of CF showers a family with a whole host of feelings and emotions. These include:

- concern for the child's well-being
- worry about the future
- guilt for having a child who inherited this serious illness from his/her parents
- fear of the unknown (How do I cope? How sick will my child be? How long will my child live?)
- anger that this has happened to them
- resentment for the time and attention that the CF child requires

Brothers and sisters of a child with CF may feel they are being ignored or left out because of the extra attention given to the child with CF. They may then feel guilty because of their feelings of resentment. They may also feel anxious about their sibling's health.

Some siblings of children with CF may try to attract attention by misbehaving or withdrawing into themselves.

Often, siblings want to be included in providing care to the child with CF. Many older children become very protective of their brother or sister with CF and take responsibility for monitoring the child's needs and health.

When a child gets older, he/she will begin to experience some of the special problems surrounding adolescence. Two problems can be especially difficult: embarrassment at being different and resentment at having to follow the rigorous health regime.

Teenagers with CF may be self-conscious about their cough. They may be shorter or thinner than their classmates. They may tire more easily during physical activity. They may have to take medications or treatment at school.

A teenager's embarrassment can be eased if he/she understands what CF is, how it causes these problems, and why the treatments and medications are necessary. It will help a great deal if he/she understands and can explain to friends that the cough is not contagious and that it helps to clear the lungs; that medications help to digest food, fight respiratory infection, or provide necessary vitamins. Removing the mystery will go a long way toward acceptance and understanding.

Teenagers and young adults want independence. They may rebel against schedules, treatments, medications and limitations. It is normal for adolescents to want to be independent of their parents. Learning independence and responsibility is an important part of growing up.

Adolescence is a difficult time of life for anyone. For an individual with a chronic disease like CF, there are extra problems regarding education, achieving independence, sexual and social maturity, and career planning.

As people with CF are living longer and healthier lives, new issues develop relating to independent living,

marriage, family planning and financial concerns. While the very existence of these issues represents a triumph for the adult with CF, they can, nonetheless, be difficult to deal with while managing a chronic illness. It is our hope and goal that many people with CF will be able to settle comfortably into an adult lifestyle, mixing good health care with independence, marriage, family and career.

Despite the relatively common occurrence of CF, most people remain surprisingly uninformed about the disease. This can make things a bit harder for parents of children with CF as they have to answer many questions from friends, family and others about CF and their child.

Cystic fibrosis and cross infection

Infections from other people

Children and adults with cystic fibrosis are more susceptible to respiratory illnesses due to their lungs being a haven for harbouring bacteria. **Therefore other children and adults with respiratory illnesses; tummy bugs (gastroenteritis) and chicken pox represent a risk.** Viral infections (eg. colds) are the most common cause of chest infections for people with CF and can be very serious.

Simple hand washing and/or use of antibacterial hand gel and covering one's mouth when coughing or sneezing, prevents the transmission of viruses. It is really important that all children and adults maintain this level of hygiene for the CF child and also for the benefit of the school or local community. **Children and adults, who have cold or flu like symtoms, should not be placed near a child with CF.**

Cross infection between people with cystic fibrosis

Cross infection between people with CF is now considered a risk. In the past children and adults with CF, were able to attend camps and other social activities together, whereas now this is discouraged. People with CF can harbour harmful viruses and bacteria that will not affect a person who does not have CF but can potentially be transmitted to other people with CF.

It is advised that people with CF avoid the following with other people who have cystic fibrosis:

- kissing
- intimate contact
- prolonged car rides
- fitness classes
- sharing drinking and food utensils
- handshaking
- sharing rooms or bathroom facilities
- sharing respiratory therapy equipment
- sharing toys

If there is more than one child with CF attending a school or a community group, it is highly recommended that they are not in the same class. A guide for Infection Control has been developed and is available from Cystic Fibrosis WA.

Considerations for schools

It is vital that the parents of a child with cystic fifrosis and the relevant teachers form a workable relationship where each party understands clearly the limitations and needs of the other party.

Communication between the school and the family

In some cases the student with cystic fibrosis and parents want to inform everyone, while others prefer total privacy. Ultimately it is up to the student and parents to determine whether or not they wish to notify teachers, other parents and students about their condition.

As the child nears adolescence it may be worthwhile to include him or her in any conferences held with the parents to discuss issues such as medication.

It will be more beneficial to teachers if parents of the child can regularly give notice if the child is feeling fatigued, not able to keep up, finds the workload too demanding, has been fighting a cold or has had a lot of after school medical appointments.

The more parents can do (early in the year) to anticipate the health needs of their child, the greater chance the child will be able to manage his or her class work successfully and also be safe and comfortable in the classroom.

Hospital school services at Princess Margaret Hospital (PMH)

Children in hospital should have educational provision from the Hospital School Services at PMH (Ph: 9340 8529). Staff at Hospital School Services are continuously establishing and maintaining the vital link between hospitals and schools that is necessary for a child's care. Academic exams can sometimes be arranged within the hospital setting. If the admission is pre-planned, an appropriate school work program can be arranged.

Intravenous (IV) Line

If a child comes to school or community activity with an (IV) line, the school administration, including the school nurse, should meet with the family to discuss who is caring for the line, if there will be any potential emergencies and what level of activity is appropriate for the child. The administration of the IV antibiotics in a school setting needs to be considered in the child's school healthcare plan.

Outings and overnight trips

Children with chronic illnesses are often denied social opportunities for fear of jeopardizing their treatment. Outings and overnight trips should be positively encouraged. The benefits usually outweigh any temporary lapse in optimal treatment. During school outings, teaching staff may be concerned about:

Physiotherapy – it is usually possible to train helpers to assist the child but the child may be able to perform an adequate physiotherapy program independently

Medication – the school will probably be familiar with enzyme administration, indeed the child is likely to be independent. It may be reassuring for the teacher to be given a list of medication and dosages and a contact telephone number

Equipment – most equipment, such as nebuliser compressors and feeding pumps, can be provided in a portable form

PhysED

Although children with CF are encouraged to play sport and keep active and fit, it is important to remember that CF may limit the extent to which the student can participate. The student's tolerance level can vary from time to time, even from one day to the next.

Regular toilet breaks

Children with CF should be allowed to go to the toilet when they ask. Due to digestive problems, children with CF may have a short waiting time when they need the toilet. Teachers should make a plan with the student regarding leaving the classroom when necessary.

Should the child's peer group know?

Some parents may wish to maintain confidentiality about their child's condition. Whilst parents' wishes should always be respected, the following should be taken into account:

- Maintaining secrecy is extremely difficult, particularly when the child is requiring regular medication.
- A child who has to keep their illness a secret may feel shame, isolation and fear. The opportunities of staying overnight with friends, going to parties and school trips may be denied because of fear of disclosure.
- Other children are curious and notice differences in their classmates. Lack of an explanation from adults is more likely to lead to bullying and teasing.
- Even if a child is healthy enough to maintain secrecy in the short-term, the need for clinic visits and unpredictable acute medical complications make this an unrealistic policy.
- Most children affected by CF find the support of friends helpful at school and during hospital admissions.

Time off school

Time off school, if only to attend clinics, is inevitable. Many children will have two-week admissions for a "tune up" to have intravenous antibiotics and extra chest therapy. It is difficult returning to school and fitting back into the class room environment when the amount of time off school starts to create educational difficulties.

Good communication between parents and the school may allow work to be transferred to the home setting. Absenteeism for clinics can be anticipated allowing work to be rescheduled.

What about discipline?

Many parents of children with CF express a concern about discipline. Parents can feel hesitant or even guilty about disciplining their child. But discipline is necessary for any child (including children with CF) to be well adjusted and psychologically healthy.

It is important that parents and teachers apply the same discipline and behavioural standards to a child with CF as they would to any other child.

Further reading and resources

Websites for CF Information:

Cystic Fibrosis Road Map
www.cysticfibrosiswa.com.au

A portal for everything you need to know about cystic fibrosis.

Genetic Support Council of WA
www.geneticsupportcouncil.org.au

The GSCWA has established a resource centre which includes a library and online resources providing information on a range of topics involving genetic conditions.

Kids health
www.kidshealth.org

KidsHealth is the number one most visited website for children's health and development. There is information for young children, teenagers and also parents, available on this website.

Livewire
www.livewire.com

Livewire is a safe and fun online community designed for children living with a serious illness. Livewire is a supportive place to hang out, connect, share experiences, be creative and know that you are not alone.

Other organisations
There are other CF organisations in Australia and also around the world which have some great information such as;

www.cftrust.org.uk	England
www.cysticfibrosis.ca	Canada
www.cfv.org.au	Victoria

Books available in the CFWA library to loan.

(Contact CFWA for more details on borrowing from the library)

Monty: A story about Cystic Fibrosis Bubb, M (2004), Australia, Cystic Fibrosis Queensland

Monty is a whimsical canine, whose owner has CF. Through Monty we experience the daily routine of living with CF as well as heading off to hospital.

Mega Man Gardiner, Dominique (2007) Queensland, Cystic Fibrosis Queensland

Jack, his brother Joe and friend Tom love building robots. The Inter-school Robotics Challenge is on again and the boys reckon Mega Man, their latest idea, is going to win. But then their chief designer, Joe, who has CF, gets put into hospital for his "65 Roses tune-up" and building the robot doesn't go as they'd planned.

Living with Cystic Fibrosis Gray, SH, (2003) USA, The Child's World

This book is for primary school aged children and explains what CF is like for those who have it, what can be done to cope with it and when there might be a cure.

Cystic Fibrosis Everything You Need to Know Kepron, W, (2004) USA, Firefly Books

In this book, Dr Wayne Kepron explains this complex disease and the range of therapies available. Using case histories he describes ways of coping with the disorder.

Additional contacts:

Genetic Services of WA
Principal and General Enquiries
PH: (08) 9340 8828 and (08) 9340 8222
Fax: (08) 9370 7058

Hospital School Services
Principal and General Enquiries
PH: (08) 9340 8529
Fax: (08) 9382 2140
E-mail: hss@det.wa.edu.au
Web: www.hospitalschoolservices.wa.edu.au

Princess Margaret Hospital (PMH)
(where children with CF attend)
Department of Respiratory Medicine
PH: (08) 9340 8626
Fax: (08) 9340 8983

Sir Charles Gairdner Hospital (SCGH)
(where adults 18+ with CF attend)
Department of Respiratory Medicine
Respiratory Outpatient Clinic
PH: (08) 9346 1756
Fax: (08) 9346 1555

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James C. Cunningham M.D. & Lynn M. Taussig M.D. (1994) *An Introduction to Cystic Fibrosis for Patients and Families*, Maryland USA , Cystic Fibrosis Foundation

Appendix 1:

Example of things for teachers to consider in a medical healthcare plan

There are many factors that need consideration for primary and secondary students with CF. Below are examples of questions that need resolution in formulating a school medical plan to support a CF student.

Questions	Personnel required	Points to consider
What limitations are imposed on the student because of present medical status? E.g. Physio, hospital visits, family dynamics, avoidance	• school teacher • parents • Hospital School Services	• How much study time is available? • Does the medical condition vary? • Can assessment be delayed?
How can a student access relevant school work if unable to come to school, for either home or hospital school services?	• arrangement with parent to pick up? • teacher emails or faxes to parent or to Hospital Services • other siblings or peer collects	• Who should mark work? • When should student arrange for work to be handed back?
What plan is in place to inform teacher if student is sick at home or in hospital?	• teacher • parent • peer/sibling • Hospital School Services	• How to maintain communication between parent and teacher?
Plan for taking medication at school	• parents and student • teacher, support staff • principal • staff on duty at lunch	• Do all parties understand about enzymes e.g. when not to take etc?
Toilet breaks: what is the agreement?	• teacher • student • parent	• Does this need reviewing?
How to monitor intake of fluids etc to avoid dehydration especially in summer	• sports teacher • teacher • parent	• Education and understanding of CF
How to limit exposure of student to acquiring colds, flu etc from school environment	• parents and student • teacher, support staff • principal • whole school	• Educating the students about hand washing and good hygiene, using hand sanitisers in the classroom etc • Sending notes home to other parents

Appendix 2:
Information sheet for relief teachers

What is cystic fibrosis?

- **Cystic fibrosis (CF) is one of the most common life threatening, genetic conditions affecting children and adults in Australia.** CF affects primarily the lungs, digestive system and sweat glands. With CF the mucus glands in the body secrete very thick sticky mucus which clog air passages in the lungs and trap bacteria. The pancreas and sweat glands are also affected by the thick and sticky mucus.

Digestion and enzyme capsules

- A student with CF may take capsules containing pancreatic enzymes with their meals and snacks. These enzymes help the body to absorb nutrients from food and to reduce the number and bulk of stools, the amount of flatulence and abdominal pain and distension.
- **Enzyme capsules are not habit forming and will not alter the student's attitude or emotional behaviour.** Enzyme capsules are not harmful to other children.
- In most cases, depending on the student's age and maturity, he or she can carry the day's supply in their lunch box and take them with their lunch and snacks. Parents should be responsible for the number of capsules and a proper storage container to keep them in.
- The capsules are only to be consumed with food that contains fat, so there is no need for a CF student to take them with corn thins, water, fruit juice, cordial, fizzy drinks, fruit or vegetables (except avocado) or lollies (without chocolate or fat content).
- **The capsules only work within half an hour of food being consumed.** (It is preferable to take the capsules prior to eating and to finish the meal within half an hour, but they can be taken after the meal).
- In order for growth to occur, a diet that is 20% to 50% higher in fat and salt content is essential for a child with CF. (So his or her lunch box may contain food that is higher in fat or salt than the ordinary lunch box).
- **A student with CF may need to make frequent visits to the toilet.** Due to digestive problems, children with CF have a short waiting time when they need the toilet. Please make allowances for this.
- If a child takes too many capsules for the amount of food consumed in a couple of instances it is not dangerous, but this could be harmful over a long period of time.
- If the child doesn't eat their food or snacks they do not need to take the capsules, however parents should be notified of this, as a child with CF has to eat a certain amount of food per day.
- The way that parents calculate how many enzyme tablets to give to their child before a meal depends on the strength of the capsules.
- An example is: **4g of fat = 1 x 5,000 strength enzyme capsule or 8g of fat = 1 x 10,000 enzyme capsule**

Loss of salt through sweat

- People with CF lose **3-4 times more sodium (salt)** through the sweat glands than those without CF, so their daily need for this essential mineral is high. Salt depletion and dehydration can happen very quickly during a hot day with prolonged exertion. Drinking plenty of water and taking salt supplements is important especially during and after sports activities.

Catching viral infections etc

- **Children and adults with CF are more susceptible to respiratory illnesses, due to their lungs being a haven for harbouring bacteria. Therefore other children and adults with respiratory illnesses, tummy bugs (gastroenteritis) and chicken pox represent a risk.**

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Appendix 3:
Example of enzyme recording chart

Meal type	What was eaten	Number of enzyme capsules
Recess		
Lunch		
Afternoon Snack		
Party Food?		

Example of number of enzymes per food type if 4g fat = 1 x 5000 strength enzyme tablet

Food	Fat content	Number of enzymes
1 tbsp Nutella	6g	2
Pikelet average size	2g	1
Yogo snack pack 150g	5g	1
Polony 50g (2 slices)	9g	2
Margarine 1 level tsp	4g	1

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Cystic Fibrosis WA (CFWA)

The Niche
11 Aberdare Road
NEDLANDS WA 6009
PH: (08) 9346 7333
FX: (08) 9346 7344

Freecall: 1800 678 766 (Country WA)

E-mail:info@cysticfibrosiswa.org
educationofficer@cysticfibrosiswa.org
socialworker@cysticfibrosiswa.org
nurse@cysticfibrosiswa.org

Website: www.cfwa.info

Post:
Cystic Fibrosis WA
PO Box 959
NEDLANDS WA 6909