Chronic Illness Alliance

Invisible Illness

—an online resource about children and young people with chronic conditions for school communities

Cystic Fibrosis Entry

Organisations who wrote / approved the information

Cystic Fibrosis Victoria

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www.chronicillness.org.au

A: FACTS ON THE CONDITION

1. General description, including different types, causes, prevalence, signs and symptoms

What is CF?

Cystic Fibrosis (CF) is the most common inherited life-shortening condition affecting Australians. When someone has CF, his or her cells are missing an essential protein so that chloride and sodium can not be properly transported across the cell membrane. This cellular defect affects the body in many significant ways. In the lungs, mucus secretions are thicker, stickier and hard to move. Blockages in the airways trap bacteria, causing repeated lung infections and localised inflammation that eventually produces scarring and irreversible damage to the lungs. Respiratory failure is the major cause of premature death. CF also directly affects the pancreatic and gastrointestinal systems. Around 90% of people with CF need to consume enzyme capsules when eating. This is because the pancreas is unable to produce sufficient enzymes to digest food. Furthermore, thickened secretions block the pancreatic duct, meaning that naturally occurring enzymes can not reach the small intestine to break down food so that nutrients can be absorbed. CF can also affect the sinuses, liver, spleen and reproductive systems. Around 95% of males with CF are sterile as they either do not have or experience shrinkage of the vas deferens (the tubes which carry sperm from the testes to the ejaculatory ducts). However, the existence of sperm in the testes means that many are able to father children using IVF techniques. Women with CF may also have reduced fertility but in most instances can conceive naturally.

How does CF vary?

Both the severity and symptoms of the condition vary greatly between individuals. Some people experience more problems with their lungs, others with their pancreas and digestive system. Typically, they live with mild, moderate or severe lung disease and gastrointestinal problems. People with mild or moderate CF usually appear healthy but this appearance can be misleading as they must adhere to a rigorous health regimen and see specialist consultants. Life expectancy has improved dramatically for all persons with CF since it was first recognised as a specific condition in the 1930's and infants with CF rarely lived to be toddlers. Today, with early diagnosis, greater understanding of CF, improvements in treatment and better management, people with CF can live successful, happy and fulfilled lives. The median age of survival (half live longer, half die younger) is now 37 years. Many people who have CF live well into their 30's, 40's and 50's.

How do you get CF?

CF is not contagious. It is an inherited, recessive condition that equally affects both males and females. To be born with CF a child must receive a faulty copy of the 'CFTR gene' from both parents. A child that receives a faulty copy from one parent and a normal copy from the other is born a 'carrier' and will not have CF. Approximately one in every 25 people in the community is a healthy carrier of the faulty gene, equating to around 1 million carriers in Australia.

If both parents are carriers then every pregnancy has a one in four chance of producing a child with CF. Accordingly, around one in every 2,500 babies in Australia is born with CF . Since 1989 the National Newborn Screening program has tested all babies born in Victoria for CF.

Approximately 3,000 Australians have CF of which more than 600 live in Victoria.

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2. Treatments, including role of specialists, effects of treatments, use of devices, daily routines

People with Cystic Fibrosis (CF) undertake a daily health regime which increases longevity and maximises quality of life. Each person's regimen is individual and is informed by and agreed with his or her treating consultants. It may include physiotherapy for airway clearance, exercise, attention to specific dietary requirements, nutritional supplementation and medications

Airway clearance
Airway clearance techniques are used daily to help remove the thick secretions in the lungs, to decrease infections and improve air exchange. Inhaled medications are often used to open the airways either by an inhaler or nebuliser. Then a mixture of percussion physiotherapy, vibes and breathing exercises are used to dislodge small mucus plugs in the airways. This usually causes coughing, enabling the person to 'cough up' the mucus. Alternatively, breathing into a device such as a PEP mask or flutter device exerts pressure to loosen mucus. Airway clearance is commonly undertaken between 1-4 times daily at home, but may sometimes be needed in the school or workplace environment. Exercise further assists airway clearance by loosening mucus and promoting deep breathing.

Medications
Respiratory medications aim to clear thickened secretions, open airways and prevent or control respiratory infections. Medications vary depending on the individual and the severity of his or her CF. Typically they are numerous, time-consuming and vary throughout the year depending on whether the person is experiencing an exacerbation (a worsening of lung condition) or a 'well' period. Some common types of medications include:

- Bronchodilators - these can induce overactivity, with a fast heartbeat and trembling hands
- Anti-inflammatory medications
- Antibiotics
- Steroids - these can cause mood swings, irritability and an increased appetite

These medications may be given orally, intravenously, through metered dose inhalers or in aerosol form using a nebuliser. Intravenous medications may be administered either in hospital or at home.

Enzymes and other supplements
Pancreatic enzyme capsules are supplements and do not pose a risk if accidentally consumed by someone who does not have CF. The capsules contain a combination of several body-friendly enzymes to help digest and absorb nutrients. When these enzymes pass into the stomach they act like the body's natural enzymes by breaking down the fats, protein and starch contained in food and increasing absorption in the small intestine. Most people with CF must take enzymes when eating foods and may consume as many as 40 capsules each day. Young children with CF who are unable to swallow capsules consume the enzyme beads from opened capsules mixed together with a pureed fruit such as apple. Additional dietary supplements such as vitamins and calcium tablets may also be taken. Daily salt supplements may also need to be taken as people with CF lose a particularly high level of salt through their sweat. This increases their risk of dehydration and electrolyte imbalance. Alternatively, salty foods may be eaten.

Diet
People with CF are encouraged to consume a diet that is high in salt, protein, fats and calories to help provide their high-energy requirement and meet nutritional needs. In general they require an energy intake of between 120% - 150% of the recommended daily allowance (RDA). This may mean consuming large meals and additional snacks throughout the day to maintain proper nutrition. Some people with CF 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. Supplemental liquid nourishment and some medications can be administered through this button. This is rarely undertaken in the school or work environment.

Children and adolescents with CF can spend large amounts of time in hospital and can miss a lot of school. As they grow older people with CF may need to have leave from their employment to attend appointments at their CF clinic or ‘tune ups’ in hospital.

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B: THE CONDITION'S EFFECT ON THE CHILD/YOUNG PERSON

1. Effects on the individual

Cystic Fibrosis (CF) affects a person's life in many ways and at times considerably challenges his or her health and lifestyle. People with CF may find it difficult to study or work full-time while coping with hospitalisations, fatigue and a demanding treatment regimen. Some adults choose to relinquish full-time employment to better manage their health and this may impact seriously on personal income. Despite these pressures, research has shown that people with CF are no more likely than those without the condition to suffer from depression or mental illness.

Hospitalisations
Time away from school or work is inevitable for people with CF. Many will have 2 week 'tune-ups' to have intravenous antibiotics and extra chest therapy. Some children and adolescents with CF do not experience periods of hospitalisation until later in life while others and adults may require frequent admissions. Being in hospital and not being with family and friends is very distressing for the person with CF. While in hospital, children strive to keep abreast of their schoolwork and parents may request that teachers provide work for the child. Aside from admissions, all people with CF require hospital based check-ups that may take a full day.

Fatigue and Endurance
A person with CF may tire more easily than their peers for numerous reasons including:

- Waking early for morning treatments - this may in itself be very tiring for someone with CF, especially if they have low lung function

- Having a chest infection - which may require longer sessions of airway clearance and more coughing. Coughing absorbs energy and can be very frustrating, especially if the person is trying to maintain quiet in a classroom or workplace.

- Low lung function - a person with CF who has low lung function will tire more easily than if they had high lung function. This is due to: burning up energy with increased breathing; having lower oxygen saturation levels; coughing more; spending more time doing airway clearance techniques; and having a lower appetite due to tiredness. Someone with low lung function may find it harder to exercise but should be encouraged to do so because of numerous benefits.

- Malnutrition - people with CF require a high energy diet of between 120-150% of the recommended daily allowance (RDA). They are encouraged to consume foods high in calories, protein, fat, salt and sometimes sugar. Meeting this requirement can be a struggle for some and many people with CF look underweight. Some may require a feeding tube to meet their dietary needs.

Health regimen
People with CF are encouraged to take responsibility for their health regimen from a young age. Children are encouraged to be independent with all their medications and to recognise the signs and symptoms of their condition. Some of the medication self-administered in early life may include:

- Inhaling medication such as Ventolin
- Taking enzyme capsules with food and salt tablets to prevent dehydration
- Monitoring blood sugar levels and injecting insulin for CF related diabetes (CFRD)

The length and complexity of treatment regimens varies greatly. People with lengthy and or complicated regimens may often have difficulty finding sufficient time each day for treatment.

Image
Many people with CF are concerned about symptoms that affect their image, particularly during adolescence. Young people with reduced height and weight or delayed puberty can have a hard time. They often look younger than their peers. Worse, they may be treated as though they are younger than

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they actually are. Another concern is that some are tempted to skip taking enzymes to lose weight. Adolescents and adults with CF sometimes find it difficult to do the things their friends enjoy, such as joining late night parties and attending events held in smoky venues. Smoke filled rooms are particularly hazardous for people with CF. All people with CF face the difficult decision of whether and when to tell others about their condition.

2. Effects on those close to the child/young person

Caring for a child or adolescent with Cystic Fibrosis (CF) can be very demanding for the parents and caregivers. They facilitate their child's health regimen and provide the education and the motivation needed for the person with CF to take responsibility for their own treatment. In addition, parents take the child to regular hospital appointments at specialist CF centres based in Melbourne. At times people with CF will be admitted to hospital for intensive treatment and the parents will support their child's care.

Caring for a child with CF is also expensive. Families must meet the high cost associated with ensuring that their child is kept as well as possible. Ongoing expenses include medication, enzymes, vitamins and other supplements, high calorie foods, physiotherapy equipment and nebuliser pumps. Some couples find that one parent needs to relinquish their job in order to care for the needs of their child. This places an additional financial burden on the family.

A recent study has shown that the primary care-giver of a young child with CF has a one in three likelihood of suffering from depression, compared to the national average of one in five. Carers often seek the support of others who are dealing with the same or similar issues. They may also seek opportunities for respite from the daily demands of their child's health regimen.

Research has shown that siblings of a child with CF often feel the effect of growing up with a brother or sister with a progressive condition. Some display anxiety over the future of their sibling or feel overshadowed by their brother or sister. They can become angry and may resent the attention given to their sick sibling by their parents. Yet, some siblings develop considerable independence while spending time in the care of friends or relatives when their brother or sister is in hospital. Other siblings display a greater understanding of the needs of people who are sick or disabled.

Siblings of a child with CF need to have particular attention paid to their needs and require help when they experience difficulty dealing with the condition.

3. "In Their Shoes" - stories from children/young people with the condition

Dean, aged 18

"Having CF didn't really affect me when I was little. I was only in hospital once a year. I don't think it starts to matter until you really understand it. For me that was when I turned twelve. I started going to high school and suddenly I was around all these different people. At primary school there were less people and everyone was kind of close to each other and I didn't think about it much. It changed when I went to high school.

CF began to affect me seriously when I was sixteen. It was my last year at school. It was a bad year, my worst. I was getting really bad colds and lots of chest infections mainly. The antibiotics would relieve it for a while. I'd be three weeks in hospital, one week at home and then back into hospital. It was hard because I kept missing school, lots of day off, and I got behind in my school work.

The worst part about being in hospital is that it's a totally different lifestyle. In hospital everything's a routine whereas at home you're pretty much free to do what you want. I found missing my family the hardest. When you're away from them, it's like they're not there, you know? You get separated from normal life, from what's going on in their lives. It's really good when they visit. At the start of the year they were really worried about me, but later on it almost became a routine, 'Oh, he's back in hospital again.' Everyone got sick of it. …"

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Leesa, aged 15

"People can tell there's something wrong with me because I cough a lot. I used to say I had asthma, but now I say it's complicated; asthma and something else. My closest friends know I've got CF, but others don't. Most just assume it's asthma. I read that Cameron, another CF patient who died, always kept it a secret, so I'm not the only one who does. I'd like everyone to know I've got CF but it's so hard to sit down and explain."

Jason, aged 17

"Doing everything I have to for my treatment takes a lot of time each day. Too long. I get angry when I'm told to do things, especially when I don't want to. I shut down and don't talk to anyone. I come right after a while; time's the only thing that brings me out again. I don't get angry about being sick. What's the point? It's my way of life.

The hardest thing about having CF is not being able to do things, like contact sports. I used to play a lot of sport. I loved footy before I was twelve, but I had to stop because I got a portacath put in. … I go to the gym every day and I enjoy it although I get sick of it too. I usually feel better after I've been. The gym instructors are okay."

The last three story extracts are reproduced with permission from:
Cameron, Heather (ed.), "Different but the same. Young people talk about living with serious illness." Lothian, Port Melbourne, 1998

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C: STRATEGIES FOR SCHOOLS/TEACHERS

1. Overview of strategies for schools.

Here are some suggestions to help teachers make school a positive environment for students with Cystic Fibrosis (CF).

Building relationships

As each student with CF is affected differently by the condition it is imperative that teachers speak with the student and his or her parents to determine that student's particular needs. Building relationships with the student and parents will enable a frame-work of care to be established and provide clear lines of communication. Essential information such as the name of the treating consultant should be exchanged in case an emergency arises.

Promoting hygiene

Viral infections such as the common cold are the most common cause of chest infections for people with CF. One of the most important things that they and the people they have contact with can do is to minimise exposure to harmful germs and bacteria. Coughing and sneezing are common modes of transmission. Simple hand washing and/or using antibacterial hand gel and covering one's mouth when coughing or sneezing prevents virus transmission. It is imperative that all students and teachers maintain this level of hygiene.

Supporting the 'CF diet'

When teaching your class about nutrition, be sensitive to the student with CF who has a diet that may appear unhealthy by most nutritional standards. Students with CF are encouraged to consume large meals that are high in calories, fats, protein and salt. Some struggle to maintain this daily intake of between 120-150% of the recommended daily allowance and will benefit from support. A young child torn between eating and playing may want to eat a little and run to the playground. He or she needs time and encouragement to eat every meal. Some students need additional snacks to help maintain proper nutrition.

Students with CF are taught to self-administer enzyme capsules when eating and should be allowed to do so. A discussion with the child's care-giver will determine whether the student is sufficiently old and capable of self-administering. As enzyme capsules are supplements they do not pose a risk to other students if accidentally consumed.

Supporting the need for exercise and combating dehydration

Exercise should be encouraged as much as possible for students with CF. It is fundamental to their treatment, helping to strengthen the lungs, muscles and bones and assists in clearing mucus from the lungs. Exercise is also of emotional benefit. It can help lower stress levels, promote self esteem and assist in building friendships. How much he or she is able to participate will depend on that individual's level of health and how they feel from day to day.

The student with CF may cough more during exercise but should always be encouraged to participate in the activity. This may be embarrassing to him or her, and if so a discussion between the student, parents and teacher can help by agreeing a strategy to manage the embarrassment.

Most people with CF have a reduced tolerance to heat, especially when exercising. Their loss of salt and risk of dehydration and electrolyte imbalance should be managed by the student carrying water or a sports drink, eating salty snacks or taking salt tablets during hot weather.

Dehydration can lead to tiredness, difficulty concentrating, feeling grumpy or irritable, loss of appetite, nausea or vomiting, headaches, cramps, thirst and sunken eyes. It can also make mucus harder to cough up as it becomes even stickier. Bowl blockages can also occur.

Appreciating the effects of CF on the student

Having CF may affect a student in many personal ways that should be appreciated by teachers and are described fully under 'Effects of the Condition on the Individual'. In summary, these include:

- the need for time away from study for hospitalisations and medical appointments - parents may request that teachers provide work for the student to undertake in hospital.
- fatigue and reduced endurance - a student with CF may tire more easily than their peers for a number of reasons including: waking early for morning treatments, having a chest infection; having low lung function and malnutrition.
- health regimen - students are encouraged to be active participants in their regimen including the self-administration of medications. Aside from consuming enzyme capsules, students may

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inhale medications like Ventolin or monitor blood sugar levels and inject insulin for CF related diabetes. They may also need to conduct airway clearance techniques at school including the use of inhaled medications,

• personal image - delayed puberty and reduced height and weight affect body image. Teachers can assist by emphasising with the student who has CF and expressing to other students that appearance is only one thing that "makes you who you are". It is the student's choice whether or not to tell others about their condition and should be supported and respected in their decision.

• infertility - this is common in boys with CF. It is particularly important during sex education that males with CF still be encouraged to use protection with an emphasis on STD's.

Avoiding an over-protective attitude

Children with CF wish to be treated the same as any other child. There are just some things they can't do and some things they must do which are different to their school friends.

Intervening to ensure a positive school environment

Students with a condition may be a target of bullying and teasing. There can be many reasons why they may be a target such as being small in stature, low in weight, taking enzyme capsules, eating extra food including high calorie or "junk" food, persistent coughing and spitting out mucus or having time away from school. They may have a feeding tube, portacath or PICC line. Unfortunately some students may make jokes about CF being a life-shortening condition and this can be traumatic for the individual. Some students with CF may already have self esteem issues about their condition and bullying on top of that can disrupt their education, reduce enjoyment of school and compound further esteem issues.

For additional comprehensive information it is highly recommended that school staff contact Cystic Fibrosis Victoria for a copy of their Schools Information Pack. You may also wish to request the brochure:" Cystic Fibrosis and School: A Guide for Teachers".

2. Link(s) to useful other online resources for schools on strategies in responding to a child/young person with this condition

• Association for Children with a Disability: www.acd.org.au

• Royal District Nursing Service: www.rdns.asn.au

• Disability Online: www.disability.vic.gov.au

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D: FURTHER INFORMATION

1. Organisations, including services and resources available

Cystic Fibrosis Victoria Inc

Cystic Fibrosis Victoria was established in 1974 to support parents of children with Cystic Fibrosis. Since that time, the organisation has grown to provide services and support across Victoria for children, adolescents and adults with Cystic Fibrosis and their families and carers. At any one time, there are over 600 Victorian families using our services.

Cystic Fibrosis Victoria can be contacted at:
80 Dodds Street
Southbank Vic 3006
Phone: (03) 9686 1811 Toll free: 1800 633 685
Email: admin@cfv.org.au Website: www.cfv.org.au

Cystic Fibrosis Victoria programs and services include:
Support, information and education, advocacy, and fundraising

Recommended reading/viewing:

Websites:

CFV website: www.cfv.org.au

Cystic Fibrosis Worldwide: www.cfww.org

Cystic – L: www.cystic-l.org

CF Care: www.cfcare.com

CF Foundation USA: www.cff.org

CF UK Trust: www.cftrust.org.uk

Boomer Esiason Foundation: www.esiason.org

Videos

'Speaking from Experience' Versions 1, 2 & 3: Newly Diagnosed, Adolescents, Adults

Talking About CF 1

Talking About CF 2

Understanding Cystic Fibrosis (2002)

Books

Cystic Fibrosis: Medical Care: Author: D Orenstein et al:
Publishers, Lippincott Williams & Wilkins 2000

CFV Handbook (available from Cystic Fibrosis Victoria - see above for contact details)