



# cystic fibrosis

**in education and  
children's services**

Planning and support guide for education  
and children's services

**2008**



**Government  
of South Australia**

Living with cystic fibrosis—planning and support guide for schools, preschools and childcare services.  
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## Further information

In South Australia, families and health professionals can seek further information about health support planning from the manager of the service in which the child or student is enrolled or planning to enrol.

Department of Education and Children's Services regional personnel can assist worksites to plan support for children and students with additional needs. These services can be contacted through region offices: [www.decs.sa.gov.au](http://www.decs.sa.gov.au). General enquiries can be directed through the Department's toll free telephone number on 1800 088 158.

General information about cystic fibrosis management and associated specialist services is available from Cystic Fibrosis South Australia (CFSA), telephone (08) 8221 5595, 1800 23 28 23 (free call SA only) or [www.cysticfibrosis.org.au](http://www.cysticfibrosis.org.au).

Copies of this book and related material, training programs and services can be accessed at [www.chess.sa.edu.au](http://www.chess.sa.edu.au). This site gives detailed information about the South Australian *child health and education support services (chess)*.

This icon indicates that the information can be accessed from the chess website:

[www.chess.sa.edu.au](http://www.chess.sa.edu.au)



## Acknowledgments

This book has been developed in consultation with a wide range of education, childcare and health professionals, families and communities. Their contribution to continuity of education and care for children and students is gratefully acknowledged.

## Disclaimer

The health-related information in this book is accurate at the time of going to print. Readers are encouraged to check with their doctor or local health service provider for more recent information.

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# 1 Living with cystic fibrosis

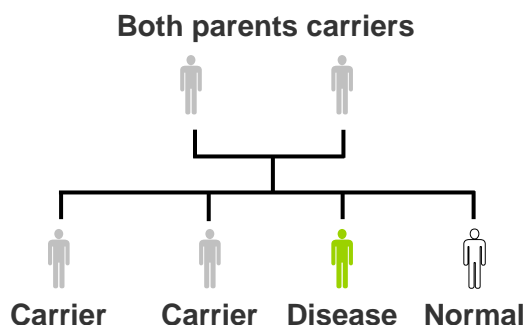
*“cystic fibrosis is a part of my life—it’s NOT my life”*

## 1.1 Cystic fibrosis (CF)

Cystic fibrosis (CF) is an inherited genetic condition, which mainly affects the lungs, digestive system and sweat glands. It is the most common life-threatening condition affecting Australian children.

In Australia one in 25 people are carriers of the CF gene. Carriers of the CF gene do not have any symptoms of the condition. If two people carry the gene and have a child, each pregnancy will have:

- a one in four chance that the child will have CF
- a two in four chance that the child will not have CF, but will carry the gene
- a one in four chance that the child will not have CF and will not be a carrier.



In Australia approximately 80 babies are born with CF each year. The incidence is one in every 2500 births. There are approximately 2500 people with CF in Australia and just under 300 in SA. CF is most common amongst Caucasian races.

The disorder's impact and severity varies considerably among individuals but its effects are generally progressive over time with increasing malfunction of the respiratory system. Recurring bouts of illness and the necessity to maintain a strict health routine interfere, at different ages for different individuals, with the individual's capacity to lead a normal life.

CF affects the exocrine glands which secrete body fluids such as sweat, mucus and enzymes. CF also affects a number of organs, but the most serious problems are found in the lungs and pancreas.

## Lungs

People with CF produce abnormally thick, sticky mucus which blocks small air passages in the lungs. This causes difficulty in clearing infections, which leads to lung damage over time. This results in a chronic cough, progressive breathlessness, the production of sputum (phlegm) and episodes of respiratory infections.

## Pancreas

The pancreas is a gland just below the stomach. One of its functions is to produce enzymes which break down food so that it can be absorbed by the digestive system. The sticky mucus can make it difficult for the enzymes to reach the digestive system and food cannot be fully digested. Children with CF may therefore have difficulty in gaining weight and the undigested food results in large, bulky and often foul smelling bowel actions. In late adolescence and early adulthood, some young people with cystic fibrosis may also develop diabetes.

## Sweat glands

People with CF do not sweat more than other people, but they do lose more salt and potassium in their sweat. The main diagnostic test for CF is the Sweat Test, which measures the level of salt in the sweat. The test cannot differentiate between mild and severe cases.

## Symptoms

People with CF can have the following symptoms:

- persistent cough, particularly with physical effort
- tiredness, lethargy or an impaired exercise ability
- frequent visits to the toilet
- salt loss in hot weather with consequent muscle cramps or weakness
- poor appetite.

## 1.2 Diagnosis

In South Australia, all babies are screened at birth for several different genetic disorders, one of which is CF. This is done through a heel prick test called the Guthrie Test. In instances where this blood test shows a positive result for CF, a Sweat Test is undertaken to confirm the diagnosis.

There are over 500 mutations of the CF defective gene. Initial testing only covers the most common mutations and so the Guthrie Test will miss approximately 11% of cases each year. These children are likely to present at an older age with failure to thrive due to malabsorption, recurrent chest infections, chronic liver disease, or, in males, infertility.

Sometimes, diagnosis is made after an intestinal blockage called meconium ileus has been detected.

## 1.3 Common associated health issues

Treatment of CF involves a team of medical and allied health professionals including doctors, nurses, physiotherapists, dietitians, pharmacists and social workers. In between periods of hospitalisation and clinic visits, families take on the role of managing routine treatment and therapy. During adolescence, people with CF are encouraged to take over responsibility for the various elements of their daily home treatment. This maximises independence and enhances their quality of life.

The management of CF is ongoing and lifelong. It involves daily physiotherapy, exercise, diet adjustments (children with CF require a high fat, high calorie and high salt diet), enzyme replacements, inhaled medication to thin the mucus, and the use of antibiotics to fight chest infections. Many people with CF also have asthma and there is a higher incidence of (cystic-fibrosis related) diabetes than in the population in general. In South Australia, children have regular appointments at the Monday morning CF clinic at the Women's and Children's Hospital. Severe chest infections are generally treated in hospital with intravenous antibiotics over a two to three week period.

Living with CF means living with uncertainty. This has a huge impact on the child, the parents and siblings, and on many aspects of family life. Adolescence in particular is a time when children with CF are at risk of depression. They may go through periods of grief both for themselves and their families.

Children with CF are likely to develop friendships, through the hospital, with others who are very sick with this condition. The reaction of children with CF to their life circumstance varies greatly. Teachers and other carers have a key role in supporting these children to maximise their participation in, and enjoyment of, all aspects of their learning and living.

# 2 Treatment for cystic fibrosis

*“cystic fibrosis is a part of my life—it’s NOT my life”*

Treatments now available for CF can improve symptoms and slow the deterioration in health, leading to significantly improved quality of life. Teachers and other care providers play a major role in minimising the focus on illness and treatment, and maximising access and participation in learning and recreation.

## 2.1 Respiratory system

*In Year 11 I was really worried about missing school. I was due for an admission, but was trying to delay it till the school holidays. I had always been good at sport, but my lung function was bad at this point. The PE teacher told us to run round the oval three times. I did one and stopped. I told him I couldn’t go on. I didn’t mention CF, I felt he should have known. (Student)*

The respiratory system is made up of the lungs, air passages and sinuses. These are lined with glands called exocrine glands which produce mucus to help lubricate the body. In people with CF these glands produce abnormal mucus that blocks air passages and sinuses and leads to repeated lung infections. Some children will take a preventative antibiotic either as a tablet or syrup. This helps the body fight off the bacteria that cause their chest infections. Over time, the infections cause permanent lung damage and are the cause of most disability and death for people with CF.

Physiotherapy helps to move the mucus into the large airways so it can be cleared by coughing. Sometimes inhaled medications are given before physiotherapy to open the airways and/or loosen the thick mucus. Physiotherapy may be required during care or schooling. This will be undertaken by a community nurse but as the child gets older they can do it themselves.

As children become unwell with a chest infection, the frequency of their physiotherapy increases, and the amount and form of antibiotics change. Sometimes another oral antibiotic is included in their daily regime; sometimes inhaled antibiotics are given. When the infection cannot be treated by these means, a two to three week hospital admission is required for intravenous antibiotics and intensive physiotherapy. Some children will be admitted three or four times a year, others more or less, depending on the severity of their condition.

Children with CF can also have asthma and/or hayfever, which both further irritate the airways and can cause serious breathing problems. Teachers can help by avoiding the

- ✦ triggers identified on the student's health care plan, and by being alert and responsive to any distress.

The care of all South Australian children with CF is managed primarily by the Cystic Fibrosis Clinic at the Women's and Children's Hospital in Adelaide. All children attend the Monday morning clinic as frequently as is required by their current health status. Attending clinic may mean missing time from school but their condition requires close monitoring and early effective treatment to lengthen life span, slow progression of the disease and maximise quality of life.



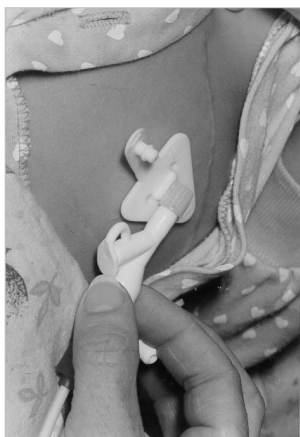
*Routine physiotherapy*

Treatments are improving all the time. Lung transplants are performed on people with severe lung disease. The wait for a suitable donor and the

potential in some cases for organ rejection can be devastating. Lung transplants are not a cure for CF but they can greatly improve quality of life.

## 2.2 Digestive system

*I recall when Jason started school in my reception class. We went to have pancakes in the primary section and we forgot his enzymes. I made him wait until we got back to our room to have his pancake and enzymes. We both got upset. We're better organised now. (Teacher)*



*A gastrostomy button*

The digestive system is made up of the stomach, intestines and pancreas. These are also lined with cells that are affected by the defective exocrine glands and therefore also produce thick mucus that often blocks off the passage from the pancreas to the small intestine.

The pancreas is a small gland that supplies enzymes to the small intestine that break down fat and protein so they can be absorbed to provide the body with essential nutrients needed to grow and develop. If this enzyme is not released, fat and protein are not absorbed. They are instead, discarded in foul smelling, fatty and frequent stools. Sometimes malabsorption causes constipation. There are also four fat-soluble vitamins that are not absorbed: A, D, E

and K. Children with CF replace these with vitamin supplements to help their bodies grow and develop.

Children with CF have considerable problems maintaining and putting on weight, and are often quite underweight and smaller than others in the class. Their malabsorption can cause increased appetite. They need extra food and calories, and a diet high in fat, protein, calories and additional salt. The feeling of fullness that thick mucus can create can cause a decreased appetite in some children.

A low and unhealthy body weight can affect the CF child's lung disease and, in turn, speed up the progression of their condition. They are therefore given high calorie food supplements and eventually, if their weight does not improve, an operation to form a gastrostomy. In the operation a small opening is made through the skin directly into the stomach. A gastrostomy button is placed into the opening, fitting flush with the skin. It has a plug that can be opened to connect tubing to give high calorie feeds directly into the stomach. When the feed is finished the tube is removed, and the plug closed. These feeds are usually given at night, when the child is asleep, through a pump that constantly delivers a set amount over a set period of time. At all other times the gastrostomy button is closed. This does not affect the child's ability to play sport, swim or perform other activities. Even so, the child may not wish others to know they have a gastrostomy button. A nurse or credentialed careworker can assist with this procedure if required (eg on camps or sleepovers).

Children with CF can develop problems with their liver as the disease progresses. Treatment is by medication and is monitored regularly. The passage between the liver and the small intestine becomes blocked with thick mucus and bile (the substance that helps to break up fats so they can be absorbed in the body) can no longer be passed from the liver. This also reduces absorption of fats.



Enzymes



Enzyme case

Children with CF are given pancreatic enzymes to help them breakdown and absorb fats and proteins. These enzymes come in capsule form but are not drugs and do not require a medication authority for taking at pre/school. They must be taken before all meals, snacks and milk drinks. Sometimes if the enzyme dose is too small or not taken, the child will suffer stomach cramps, stomach distension and smelly, frequent stools. Enzymes can be safely stored in the classroom/eating area or child's bag to enable ready access for the student prior to meal times.

Children with CF may be embarrassed about passing smelly wind in the classroom and in the toilet block: it can cause them to withdraw from classmates. They may want to go to the toilet during class time to obtain some privacy.

*I hate going to camp, mainly because of going to the toilet. I suppose it's because with CF you don't absorb your food properly, but I need to go to the loo lots and it's often hard to flush away. It's so embarrassing, especially as they are really smelly compared to other people. (Student)*

## 2.3 Body temperature control

*The school teachers are like an extra family... at camp, one teacher sat with my child throughout his self-physio treatments and made sure there was an extra drink around at all times so he wouldn't get dehydrated. (Parent)*

Children with CF lose considerable amounts of salt in their sweat. They need a diet high in salt and, because of the risk of dehydration, should have frequent drinks, especially in warm weather and/or when playing sport.

Children with CF should have access to air-conditioned classrooms. Fans are not recommended as an alternative.

## 2.4 Intravenous drug therapy

*I get a bit scared that my arm will get knocked at school with the drip in. But I want to be at school a hundred times more than hospital. (Student)*

Intravenous lines or 'drips' administer medication directly to the bloodstream, often through a short line (cannula) inserted into a small vein in the arm. An arm board used as a splint prevents damage from excessive movement. The line, which is stiff, is taped to the skin and covered to prevent infection and the whole lot is bandaged for security. Children with a short line cannot play contact sport but can participate in all learning activities which do not threaten to dislodge the line.

Some children will have a different sort of line called a PICC (peripherally inserted central catheter). This is a long, flexible line that is threaded, usually through an arm vein, into the central vein system. They are inserted under anaesthetic and may remain in place for a two to four week period of treatment. As the line is flexible, the arm does not need to be restrained. Contact sports and activities could dislodge the line and should therefore not be undertaken. Swimming is also considered unsafe.

Some children will have a permanent port implanted under their skin to enable administration of medication into the vein system. When the port is not being used for medication, all that can be seen is a slight bulge under the skin in the chest wall. When medication is administered, a 90 degree angle needle is inserted through the port. It stays in for the two to four weeks that the medication is administered. A special dressing is applied over the needle to keep it securely in place and to prevent infection. Children with ports cannot play contact sports, and, if they have the needle in, they must not go swimming.

Children can have a port protector individually made for them, by the hospital, to protect the port from being accidentally knocked or traumatised. Schools and centres should seek advice through the family from the child's medical team about whether a port protector should be worn at the school or centre.



*Accessing a port for intravenous drug therapy. The port is covered in a dressing to keep the needle in place and to protect against infection.*

# 3 Health support

*“I was brought up and encouraged to do anything I wanted”*

School, preschool and childcare staff are trained and expected to care for children/students in a manner which enhances their learning and independence, and which respects their privacy, dignity and right to feel and be safe at all times.

- ★ Health care management training for education and childcare staff is generally limited to supervision for safety and first aid. Additional care required by children/students should be written by the doctor (or other relevant health professional) in a health care plan detailing routine and potential emergency care recommendations within the education and care setting. Staff can then use this plan to develop a health support plan, detailing how staff will address the child/student’s health care support needs.
- ★
- ★

The health care plan and support plan must reflect respect for the child/student's privacy, dignity, safety and comfort. Confidentiality issues should be explicitly negotiated with the family.

## 3.1 Confidentiality, privacy, dignity and safety

Young children often enjoy sharing the news and their experiences of living with CF with their classmates. The degree and nature of this sharing should be discussed with parents so that they can support their child in this process.

Older children and adolescents often wish to keep their illness and experiences private or confined to a very small group. At this age they can be particularly wary of teachers' respect for their privacy. They may also be fearful that by sharing the information they will be treated differently by their teachers and peers.

Information exchange between the family and health professionals and school, preschool or childcare is essential to support academic progress and enhance peer support. The sharing of information needs to be assessed and negotiated for each child with CF, with due consideration to their needs. Staff need information about routine and predictable emergency care as it affects the child/student's access to curriculum and their safety.

The following guidelines have been adapted from material written by Dr Christine Simons (Women’s and Children’s Hospital South Australia, 1996).

*Confidentiality means that the information provided to medical personnel by patients, or their parents, is done so in confidence. Such information remains the property of the child and their parents.*

This means that, except in circumstances where there is a legal obligation to do so (for example in child protection situations) medical personnel are not free to divulge personal information about children/students without their parents' or their consent. They may provide general information about, for example, the nature of CF and care for people with CF – information generally available to the community.

Families of chronically ill children of uncertain prognosis are chronically stressed families. It would not be surprising if even the most stable and well-adjusted parents and children were, at times, anxious and touchy.

The most undermining threat to the trust between family and the school, preschool or childcare service is the feeling of things happening behind the family's back. Staff need to be acutely conscious of this in their dealings with the family and with/between other members of staff. Liaison between education and careworkers and medical personnel is usually welcomed. However some very private people may find any discussion of their lives an intrusion.

Families may also be reluctant for liaison for a range of other reasons, for example:

- feeling embarrassed, guilty, ambivalent or overwhelmed by the diagnosis/condition
- uncertain whether they can trust the people who are seeking the information
- negative experiences, if such consultations have 'gone wrong' in the past.

Generally, the situation can be clarified through a sensitive exploration of the concerns and clear identification and agreement of what will and will not be discussed and with whom, and the anticipated benefit to the child.

Adolescents are particularly sensitive about information distribution. In particular, at the beginning of secondary schooling, it may require a great deal of work to persuade them that anyone at school be told anything. Early contact with school staff may determine how they resolve this issue in the longer term.

## 3.2 Health care planning

*Our son has been missing a considerable amount of school. This means he is falling behind in his work and has to try to catch up with more homework. This comes at a cost, ie more homework means either less sleep or less free time or less treatment. (Parent)*



A health care plan for a child/student with cystic fibrosis should address the following components:

- overall wellness
- diet
- therapy and care
- internal body temperature control
- curriculum participation issues
- potential emergency/first aid situations.

This information should focus on what education and care staff need to know to provide routine and emergency care. It will be used by staff in planning support for the child.

### Overall wellness

#### Fluctuations in wellness/recent hospitalisation

Schools, preschools and childcare staff need to know if recent/frequent hospitalisation and/or general unwellness mean additional care and consideration. They also need to know of any infection control issues in addition to standard precautions. It is important to the future health of a child with CF to minimise the risk of cross infection of bacteria and viruses from others. This must be balanced with efforts to encourage children with CF to lead as normal lives as possible. Teachers can help by understanding if parents wish to keep their child with CF at home when a particularly virulent strain of virus is present in the classroom. All children in the class should be encouraged to not sneeze or cough on or near each other, as a normal public health measure. If possible, a child with CF should, discretely, not be partnered or sit next to another child with an obvious cold or cough.

#### Cough management

Children with CF have a persistent and necessary cough or wheeze. Their coughing is essential to help clear the lungs of mucus and fight chest infections. It is not contagious. Teachers can help by not drawing attention to the coughing. Often children with CF are so used to it, they are not even aware of it. A more frequent and moist or wet cough can be an early indication of a chest infection.

### **Management of port**

A child with a port cannot play contact sports. If knocked, a port may be very tender to touch.

Staff need to inform parents if children report any redness, swelling or bruising in the vicinity of the port. Injury to the port should be reported to the child's parents as soon as possible.

### **Management of an intravenous (IV) line**

If a child/student has a short intravenous line attached, the line will be covered, and the forearm splinted and wrapped to protect the line from being dislodged or damaged. If a child/student has a long flexible line, the forearm will be wrapped but not splinted. Education and centre staff do not have to manage these protective measures: they will be put on at home or by a nurse and will remain on during schooling/care.

### **Mental health issues**

Living with an illness like CF, with on-going treatment regimes and hospitalisations, can have social, emotional and behavioural impacts on the student. Other family members are also affected. Siblings may feel sad, scared or resentful of the extra attention the child with CF receives. Parents have the task of striving to maintain a normal lifestyle for themselves and their family while adjusting to the meaning of the illness and all it entails for the family.

Children with CF who have been hospitalised have often been exposed to illness, treatments and death (actual or through discussion) at an early age. They may speak about death in ways unexpected for their age. Talk of death can also be an indicator of the meaning their illness has for them and so should be dealt with sensitively by all concerned.

Younger children commonly exhibit their emotions through their behaviour. If so they require understanding and the maintenance of clear firm limits. Older students sometimes express their feelings in more direct ways and present with a depressed mood, and decreased motivation or focus on their studies.

It is not uncommon for a child to become uncooperative and poorly motivated to attend to treatments. This is worrying to parents, education and careworkers and the health care team and requires a coordinated approach to help the child meet the demands of treatment.

All students with CF experience interruption to their studies and their relationships with peers and teachers. The centre or school can help to minimise the effects of this by an understanding approach with realistic expectations. Sensitive, clear communication with the student, their family and the health team will facilitate this support.

## Could you stomach up to 60 tablets a day?

### Some children with cystic fibrosis do.

⊕⊖⊖⊖⊗⊗⊗⊗	Before breakfast	⊗	Around 4pm
⊕⊕⊕⊕⊕⊕⊕⊕⊕⊕	With breakfast	⊕⊕⊕⊕	Snack foods
⊗	After breakfast	⊕⊕⊕⊕⊕⊕⊕⊕⊕⊕	Tea time
⊕⊗⊖	Before lunch	⊕⊗⊖	After tea
⊕⊕⊕⊕⊕⊕⊕⊕⊕⊕	With lunch	⊕⊕⊕⊕	Supper
⊕⊕⊕⊕	Afternoon tea	⊗	Before bed

⊕ enzymes   ⊗ vitamins   ⊖ antibiotics

## Diet

### Special dietary requirements

Children with CF have difficulty maintaining their weight and growth patterns as they cannot absorb essential vitamins, minerals, fat and proteins. They lose large amounts of salt in their sweat, suffer stomach cramping, and have frequent, sometimes foul smelling stools. They take vitamin and mineral supplements, and pancreatic enzymes to help the body digest fat. Their diet therefore needs to be high in fat, protein, salt and calories as well as incorporating all essential food groups.

Children with CF need much more food than a routine healthy diet (as described in the Australian Dietary Guidelines). They are encouraged to eat foods like chips, lollies, hamburgers, biscuits and highly salty foods and add foods such as cream, salt, ice-cream and butter to many of their everyday meals. They eat more food, more frequently than other children. Extra food will need to be provided for school activities and camps. The high calorie, high protein diet they need to fit in each day is often not easy.

Children with CF often report being teased by other children about having poor eating habits. Education and childcare workers can help by encouraging the child with CF to eat during all breaks, and to normalise their eating pattern to other children.

### Gastrostomy button

Children who need additional food supplements will receive them through a gastrostomy button located in their stomach. During school, preschool and childcare this button is closed and does not affect the child's ability to participate in any activity. A credentialed careworker can assist with the procedures for supplementary feeding at night. There are no routine care issues associated with a gastrostomy button for education and childcare workers. If the area becomes red or inflamed, parents should be informed as soon as possible. Staff should be aware that many children/students do not want others at school to know about their gastrostomy button.

## Enzyme supplements

Children with CF take pancreatic enzyme supplements daily by mouth with food. They come as powder-filled capsules, beads or tablets.

Most children have learnt to swallow tablets by the age of six or seven. For younger children, capsules need to be opened and the beads or powder inside mixed with one or two tablespoons of soft food. Children need to drink after taking enzymes this way, as enzyme powder will irritate the gums and tongue if it remains in the mouth.

Children should have their enzymes with them at all times. For younger children it may be useful for the education or childcare worker to carry extra enzymes. For older children it is important for them to be allowed to take their enzymes when they see fit and not to draw attention to adherence.

Enzymes are not classed as pharmaceutical drugs. Children with CF may also be taking pharmaceutical drugs such as antibiotics in addition to their enzymes. Enzyme supplements are not considered to be dangerous substances and therefore can be carried by the children and stored where the child can access them when needed.

## Therapy and care

### Nursing and physiotherapy

Some children/students with CF may require complex/invasive health support, such as physiotherapy, while attending school, preschool or childcare. This support should be provided by a visiting nurse or therapist.

### Nebuliser treatments

Some children with CF require nebulised medication prior to physiotherapy. While education and childcare staff can supervise nebulised medication, this will generally be managed by the child/student themselves or the visiting health worker. Education and childcare staff need training before supervising administration of medication via a nebuliser.

### Home based care

In South Australia families can sometimes opt for their children to receive intravenous (IV) drug medication through the Women's and Children's Hospital home IV program. This program works to minimise disruption to the child and their family caused by frequent or lengthy hospital admissions. The child/student is able to attend school during home IV treatment but will have an intravenous access line or an accessed port (see section 2.4 Intravenous drug therapy). The child will be supported by daily visits from a physiotherapist who may need to visit during school time.

The child/student will not be able to take part in any sport or physical activity during this time.

Because this IV treatment is to counteract an infection, the child/student might be more tired than usual and not be able to attend programs for the whole day. Staff should be sensitive and flexible in response to this episodic attendance.

## Body temperature control

### Clothing

Children/students may need to be reminded to adjust their clothing to help their internal body temperature control. Special allowances may be needed regarding school uniform requirements.

### Environmental management

A child with CF will have problems with internal temperature control and should be kept at a steady temperature in winter and summer. It is beneficial to place the child with CF in classrooms that have heating and cooling.

### Salt tablets/powder

Salt tablets are taken during warm weather and before playing sport. Teachers should be informed about the required timing and amount of salt tablets and ensure the child has access to fluids at all times. This may mean keeping a drink bottle on their desk - a healthy practice for all students. Families need to provide a medication authority detailing required dosage and timing of salt tablets.

## Curriculum/workplace participation

### Tiredness

An increase in fatigue or feeling tired is common for a child with CF. A lot of effort is required of a person with CF, on top of normal childhood activities, to maintain their health. They sometimes tire more easily during physical activity.

Because children with CF have problems with malabsorption, they may be shorter or thinner than their classmates with potentially reduced body strength.

Children also need to fit at least two sessions of intensive physiotherapy into their day. These often take place early in the morning and in the evening.

There will automatically be an increase in tiredness with any worsening of lung condition.

### Shortness of breath

During the onset of infections, children with CF may experience difficulty breathing or catching breath. This is caused by thick mucus secretions blocking the airways. It often goes hand in hand with increased coughing to clear the airways. Staff should be aware that, as with other children, breathing difficulties also can be asthma related.

### **Difficulty in concentration**

These symptoms often make it very difficult for children with CF to concentrate in class. Children with CF are continually battling infections or recovering from them. With low energy levels come low levels of concentration. Excessive tiredness later in the day and in the evening can have a profound effect on concentration span.

Education and childcare workers may need to make special arrangements for activities scheduled late in the day and provide extensions for school assignments.

### **Fluctuating capabilities**

Most young people cope with their CF very well. However, their capability to function at what is considered a normal level for them will fluctuate with the onset of infection, during treatment and after hospitalisation. This is beyond the child/student's control and is a result of both physical and psychosocial issues. They can be hospitalised up to four or five times a year. Older children may need to negotiate school workload and curriculum during this time.

### **Need for frequent, self-monitored physical activity**

A regular exercise program is very beneficial to children with CF, because it helps loosen mucus, stimulates coughing and helps build up strength and endurance of the breathing muscles. Children with CF should be allowed and encouraged to take part in exercises such as swimming, bicycling, sports and games. However, it is important to remember that ability to perform will fluctuate with health status. Children who have a port will not be able to take part in contact sports. Children at school with an intravenous (IV) line will not be able to participate in strenuous physical activities.

Children with CF can become dehydrated much more quickly than other children. This is because the gene that causes CF affects the way cells work in the body. An imbalance in sodium chloride (salt) exchange means that cells will not hold water. Sweat is therefore very salty, and hot weather and exercise can lead to salt loss and dehydration. Education and childcare workers should:

- encourage frequent drinks during and after exercise, and on warm days
- ensure salt tablets are taken either before or after exercise on warm days
- avoid scheduling physical activity during temperature extremes
- ensure children with CF remain, as far as is practical, in a fairly constant temperature, neither too hot nor too cold.

Sports skills, fitness and energy levels will vary from time to time depending on growth spurts, onset of chest infections, hospital admissions, or deterioration of their condition and overall health. Some adolescents may find that they are unable to maintain their fitness levels against their peer group. They should be encouraged to maintain the degree of fitness with which they are comfortable and with which their body can cope at any one time. People with CF usually know their own physical capabilities. The teacher should respect the child/student's assessment of their capabilities.

## Need to plan for episodic attendance

Students may know some days in advance that they are due for a hospital admission. Occasionally they are admitted straight from a clinic appointment. It is important to plan with the student for hospital admissions. Secondary students appreciate getting work in advance so that they can make work plans around medical procedures and bouts of tiredness.

Hospital is not the easiest venue to work in and students will need all the support and motivation available to complete simple assignments. Students of all ages welcome contact from their school during a hospital admission. They may often feel 'cut off' and isolated from the daily environment and need reassurance that they are not forgotten. Children with a chronic illness, such as CF, who suffer regular and frequent admissions, often do not have time to initiate and keep up this level of contact. Yet it is vital to their psychological well-being that the school keeps in touch with them.

## Potential emergency situations

Emergency situations associated with CF are rare.

If children/students have an intravenous line for medication, there are specific standard first aid responses which may be anticipated:

- Child reports discomfort, nausea, rashes or general unwellness.  
*Call family emergency contact. If they cannot be reached, call the nominated cystic fibrosis nurse for advice.*
- Child reports redness, pain, inflammation or swelling at site.  
*Call nominated cystic fibrosis nurse for advice, and then advise family emergency contact.*
- There is a leakage of some sort from the site.  
*Call nominated cystic fibrosis nurse for advice, and then advise family emergency contact.*
- A needle or line falls out.  
*Use standard first aid and apply pressure to stop any bleeding, call nominated cystic fibrosis nurse for advice, then advise family emergency contact.*

If any action is required, in addition to standard first aid, this will be documented on the individual first aid plan.

## Additional information

### Medication

Children/students with CF need to take their enzymes before eating. These are essential dietary supplements.

Normal tablet intake may temporarily increase when infection is present, or when children need vitamin supplements. It may seem at times that these children are forever taking something.

In addition, some of the children may have medication for other conditions. This should be managed in the same manner as medication for any other child/student.

## 3.3 Health support planning

*Sometimes when studying if I'm not feeling 100 per cent then I find it very hard to concentrate (sic) on my studies. It's hard to improve in this area because there are always days in the week where I don't feel 100 percent. (Student)*

- ★ The health support plan for an individual child/student will be developed by the principal, director or home-based carer with the family. It will be based on the child's health support needs as identified in their CF care plan and other care information (for example if the child also has asthma or diabetes).

A health support plan documents individualised support which staff have agreed to provide in the areas of:

- first aid
- supervision for safety
- personal care, including infection control
- behaviour support
- special curriculum support to enable continuity of education.

In schools or centres, the support plan will identify a staff member who will be the contact person for the family and health service providers. This staff member will ensure all staff has information on a need to know basis, as negotiated with the family, while respecting the family's privacy.

### First aid

If any action is required, in addition to standard first aid, this will be documented on the individual care plan.

### Supervision for safety

The child/student's health support plan may include a range of routine accommodations so they can continue to access learning programs while effectively managing their health care. Accommodations could include:

- provision of additional time to support children managing their dietary requirements
- access to fluids and food, and the toilet, as needed
- rescheduling of physical activity to support body temperature control
- supportive and sensitive encouragement to participate in physical activity
- targeted social skills programs: frequent absences mean that some children with cystic fibrosis have difficulty making and retaining friends
- modification of the program and activities in response to the demands of therapy and treatment

- regular review of progress to keep everyone informed about changes and how best to plan for and support the child.

### Personal care, including infection control considerations

Education and childcare staff should plan to ensure that therapy and dietary management causes the least disruption to the learning program while maximising care. For example, the school timetable might be adjusted to ensure the same learning area is not always affected by therapy sessions. Personal care should not be routinely scheduled in recess and lunch breaks: recreation, socialisation and relaxation should also be accommodated.

Planning should respect students' privacy and dignity. For example, staff could organise discreet access to the toilet during lesson time to avoid embarrassment due to odour.

Staff should be aware that, where there is more than one family in the school or care community with CF, cross-infection is a serious health risk. Children with CF colonise different bacteria at different times. This affects their health status and future development of the disease. It is often necessary to take special precautions to segregate children with CF from different families. This matter should be addressed in confidence, with the families concerned.

### Behaviour support

As for all children, behaviour expectations for children/students with CF should be consistent and predictable, and also sufficiently flexible to accommodate periods of stress and other potential mental health issues.



The health support plan should encourage self-management. Children with CF have to learn to live with their condition for life. It is therefore important for education and childcare staff to work with the parents and child to encourage self-management of the condition as the child matures.

A child is on a continual learning curve in all aspects of their life and their self-care will not be perfect at first nor all the time. Self-management is not a smooth process. It is often hard for parents to stand back and hand over responsibility for management. Teenagers in particular are dealing with many new responsibilities, not just CF management. Remembering to take tablets and do their physiotherapy is tedious and boring. Children should not be punished for non-compliance. Mistakes help children learn, but feeling bad or guilty about a mistake makes learning harder.

Having CF is neither convenient nor fun. Children find it frustrating and unpleasant at times to behave responsibly. They need support to understand the importance of compliance with treatment.

## Curriculum

*One year our son missed 10 weeks of school. School sent work for him which was OK, but I don't know what will happen when he's in high school. (Parent)*

Planning and review processes should ensure continuity of access to education and care as the child moves between home, school/preschool/childcare and hospital.

While school students should not be expected to maintain the pace of curriculum participation when they are unwell, they should have the opportunity to choose to participate if and when they are able. Staff can assist by:

- providing course overviews, with key assignments and timelines highlighted, and negotiated as relevant
- developing an individual learning management plan to maximise continuity of access to curriculum, including additional curriculum support hours and links with the school's volunteer learning assistance program
- planning ways to continue communication with the student whatever the setting for learning, for example, fax, telephone, and e-mail
- liaising with hospital and/or open access teachers to maintain continuity of learning programs
- considering additional support to facilitate the ease of transition between the various learning settings, for example linking with a volunteer (learning assistance) support worker
- planning re-entry after periods of absence due to illness or hospitalisation, for example not requiring assessment tasks to be completed immediately upon return to school
- forward planning for camps, excursions and other special events to ensure the student does not miss out either through poor timing, inappropriate expectations of participation, or lack of sensitivity to health and personal care support needs.

The health support plan could also document curriculum issues for other students, for example:

- nutrition education including information on individual dietary needs which fall outside national dietary guidelines
- growth and development lessons taking into account the reasons many adolescents can be below average for height and weight
- drug education including reference to pharmaceutical drugs necessary for health and other substances, such as dietary supplements, which support health and well-being
- education about grief, loss and change
- anti-harassment sessions, as needed
- disability awareness sessions, as part of the overall strategy to support inclusion.

Any curriculum planning should ensure teachers do not make individual students, or their personal health issues, the content of the curriculum. Rather, confidentiality should be respected and issues raised in a one-step-removed, generic approach. If children/students choose to disclose personal issues, they should do so advisedly and teachers should ensure that the children concerned understand the potential impact of their disclosure.