

Cystic Fibrosis Patient Charter

This Patient Charter describes the specialist health care services people with Cystic Fibrosis (CF) have the right to expect, and how patients (children, teenagers and adults) and their families/carers can contribute to gain the best results from these healthcare services.

People with CF are encouraged to become confident and able to look after their own health on a day to day basis with the support of their families, friends, the specialist CF teams and other services who may become involved in their care.

In South Australia specialist services for people with CF are provided by the Women' and Children's Hospital (for infants, children and teenagers) and the Royal Adelaide Hospital (for adults).

CFSA (Cystic Fibrosis South Australia) also provides support and assistance to people with CF and their families, including information about living with CF, peer support, parent dinners, sibling activities and financial assistance.

What is Cystic Fibrosis?

CF is a genetically inherited condition which is usually diagnosed by screening tests done on babies when they are about 2 days old. CF is caused by a defect in the CF Conductance Regulator (CFTR) gene which makes a protein that controls the movement of salt and water in and out of the body's cells. Both parents need to be a carrier of a faulty CFTR gene for their child to inherit CF.

The disease causes the production of abnormally thick mucus. This can clog up the lungs, making it hard to breath and increasing the risk of lung infection. CF can also affect other body organs, in particular the digestive system, making it difficult to digest food and for the body to get all the vitamins and nutrients it needs.

While there are different degrees of severity of the disease, managing this lifelong condition can be difficult and time consuming. People with CF generally need to manage a daily therapy routine and be regularly reviewed by a specialist multidisciplinary health care team.

Significant advances in treatment means that children with CF can usually manage the normal childhood milestones and go on to finish school and take on tertiary study or employment. Life expectancy continues to increase so that now people diagnosed with CF are generally expected to live full and productive lives well into their adult years. People living with CF have many inspiring stories of courage and determination.

How is treatment provided?

Specialist multidisciplinary CF team management

People with CF are referred to a specialist multidisciplinary health care service for ongoing support and treatment management. Specialist

multidisciplinary CF team management of care has been shown to provide the best outcomes for people with CF. The team includes a:

- > respiratory physician (lung doctor);
- > registered nurse;
- > physiotherapist;
- > dietician;
- > social worker;
- > psychologist;
- > admin support officer.

The team has access to gastroenterologists (stomach and bowel doctors), endocrinologists (body glands and hormones doctors) and pharmacists as required. The multidisciplinary team is lead by a respiratory physician (lung doctor).

What you can expect

- > You/your child's health care needs will be routinely assessed by a specialist CF respiratory physician who will take overall responsibility for the management of your care.
- > You will be provided with information about you/your child's CF condition, the latest research and the recommended treatments.
- > You will be provided with training, support and equipment to help you manage you/your child's care at home.
- > You will be able to access supportive counselling and be assisted with referral and linkages to other services.
- > You/your child will be allocated a key contact person.
- > You will work out a care plan with your Care Coordinator. The care plan will describe you/your child's individual health care needs and how these will be managed in partnership with the CF team.



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What you can expect (continued)

- > The specialist CF team will be a source of information and support to other health services who may be involved with you/your child's care.

What you can contribute

- > Do not be afraid to ask members of the team who they are and what their role is if they do not introduce themselves to you. Staff expect you to do this if they have overlooked telling you who they are.
- > Ask questions and seek the information you need to make the best decisions for you/your child's healthcare.
- > Engage with the multidisciplinary team and be available as agreed.
- > Talk with your key contact person about your support needs and make sure these are properly reflected in you/your child's health care plan.
- > Let your key contact person know if you are having trouble in managing you/your child's care, so that this can be reviewed in consultation with you.
- > Contact your key contact person when you first notice changes in you/your child's symptoms or if you have any concerns about you/your child's health to discuss if any changes in management may be needed.

Outpatient clinics

It is recommended that people with CF attend outpatient clinics (usually a minimum of 4 times a year) to be reviewed by the specialist multidisciplinary health team. This helps you monitor you/your child's health and to attend to any issues or concerns.

What you can expect

- > You will usually be given a follow-up appointment time each time you attend an outpatient CF clinic. Otherwise the hospital will contact you to arrange a time to attend a specialist CF outpatient clinic. These may include outreach and out of hours clinics if needed.
- > You/your child will have a routine physical examination including lung function testing (if over 5 years of age). Growth measurements in children (height and weight) and weight loss/gain in adults is also reviewed.
- > From time basic screening tests such as a blood test will be done for conditions commonly associated with CF, such as CF Related Diabetes (CFRD).
- > All the CF team specialists (doctors, nurse, dietician, physiotherapist, psychologist, social worker) will be available to provide information, support and advice.
- > You/your child's care plan will be reviewed and updated as needed in consultation with you.
- > You/your child will be provided a comprehensive annual review covering diet, lung testing (conducted within a lung function laboratory), exercise and physiotherapy, medication management.
- > The specialist CF team will arrange referrals to other services as needed.

What you can contribute

- > Attend regular outpatient clinic appointments as arranged with you.
- > Participate in the annual review of you/your child's health at a time arranged with you.
- > Contact your key contact person (or the 24/7 contact numbers if afterhours) if you are unable to attend a booked outpatient clinic appointment time. Another time will be arranged for you.
- > Ask questions and seek the information and support that you need to manage you/your child's health care.
- > If attending as a parent/carer, support your child to ask questions and participate in the care they are receiving.
- > Contact your key contact person if you have any concerns about you/your child's health. If necessary, they can arrange an earlier clinic appointment.
- > Let your key contact person know if you are having trouble in managing you/your child's care, so that this can be reviewed in consultation with you.
- > Contact your key contact person when you first notice changes in you/your child's symptoms or if you have any concerns about you/your child's health to discuss if any changes in management may be needed.

Hospital in the home

If your child or you need to have intravenous antibiotic therapy, this can be provided at home. Hospital in the Home (HITH) (known as the Hospital@home at the RAH) is preferred by many patients and their families. It is less disruptive, allowing people to continue to go to school, work etc and it also minimises the risk of being exposed to other germs in hospital when people are unwell. HITH/Hospital@home is only available where care can be provided safely at home.

What you can expect

- > If intravenous antibiotics are needed to fight infections in the lungs, they are given through a special line usually inserted into a vein in the upper arm which remains in place for the period of treatment. Insertion of the line is done in hospital.
- > If HITH is agreed between you and the CF team as a suitable option, you will receive:
 - education and training about how to administer the medication, use of equipment and monitoring of symptoms;
 - education and training in allergy first aid management;
 - a home care management plan including monitoring of medication levels;
 - home nursing and physiotherapy visits;
 - contact with other members of the specialist team as needed;
 - weekly review by respiratory physician (lung doctor).
- > Your HITH/Hospital@home Co-ordinator will arrange the supply of:
 - all the necessary medicines and equipment;

- any other home support services that are needed.
- > You will have a 24/7 CF number that you can contact after-hours and on weekends.
- > Information about you/your child's HITH/Hospital@home treatment will be provided to your General Practitioner and/or other services as agreed by you.

What you can contribute

- > Talk with the CF team about whether HITH/Hospital@home could be an option if you/your child needs intravenous antibiotic therapy.
- > Talk with your HITH/ Hospital@home Co-ordinator about any additional supports you need to successfully manage intravenous antibiotic therapy at home.
- > To be available at agreed times for home visit contact by members of the specialist CF team.
- > To be available for medical review as requested by the CF team.
- > If you are a parent/carer: maintain a current CFR (Cardiopulmonary Resuscitation)/Emergency First Aid certificate (you will be given information about how to get this, free training is provided). Carers (parents, partners etc.) are required to have a current CFR certificate to be able to provide HITH/Hospital@home.
- > Call the CF contact number if you have any concerns.

Inpatient care

If you/your child's condition gets worse quickly, hospital is usually the best place to be. Planned discharge aims to get you/your child back home as soon as you/they are well enough, with the supports that you need to manage you/your child's ongoing care.

What you can expect

- > You/your child's admission will be streamlined and where possible you/your child's planned admissions will be arranged directly with the appropriate hospital ward.
- > The specialist CF team will co-ordinate and monitor you/your child's care during their hospital stay for example conducting regular Lung Function Testing.
- > A CF specialist respiratory physician (lung doctor) will review your care and visit you/your child during your hospital stay.
- > Hospital staff will encourage you/your child to participate in care as appropriate.
- > A discharge plan will be developed in consultation with you, including:
 - proposed date/time;
 - any home support needs;
 - discharge medications;
 - follow-up appointments or referrals;
 - changes to you/your child's care plan as needed.
- > If appropriate services are available, it may be possible to complete you/your child's inpatient care at another hospital, for example one closer to where you live. This needs to be discussed and arranged between the hospital and the specialist CF team before you/your child's inpatient admission.

- > Information about you/your child's inpatient care will be provided to your General Practitioner and/or other services as agreed by you.

What you can contribute

- > While staff at the hospital aim to make you/your child's hospital stay as comfortable as possible, time spent in hospital can be stressful. Carer (parent/partner etc.) support can be important in providing extra confidence and encouragement.
- > Ask questions about the care you/your child is receiving.
- > Participate/support your child to participate in care.
- > Be available to talk with the doctors and other members of the specialist CF team when they come to visit and assess your health care needs in hospital.
- > Talk with your key contact person about the discharge plan and any information and support needs that you have.

Self management

The development of self management skills is a gradual process that is learnt over the years with the support and guidance of parents/carers and the specialist CF team. We aim to provide all people with CF, the knowledge and skills to handle the physical, emotional and social aspects of living with CF and to become actively involved in managing their health care in partnership with the specialist team.

What you can expect

- > You/your child will be encouraged from a young age to learn about CF and to be involved in decision making about care in ways that are appropriate to age, ability and understanding.
- > If you are a parent/carer you will be given information and support from the specialist CF team on ways that you can help your child develop self management skills.
- > You/your child will be encouraged and supported to manage CF health care as part of life that includes family, friends, school, employment, holidays and travel, hobbies, sport and recreation, relationships and parenthood etc.
- > You/your child will be offered access to information, resources and supports provided by CF South Australia (CFSA), including access to peer and family support.

What you can contribute

- > Learn about CF and the symptoms you need to watch for.
- > Learn about how different treatments work and how you/your child responds to them.
- > Learn about how to develop/support your child to develop self management skills and take on the responsibility for developing these skills.
- > Ask for help whenever you need it.

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Transition from paediatric to adult services

While the overall management of CF is similar no matter what age you are, there are some differences that are related to different ages and stages of development. The role of families/carers too changes from being the total provider of day to day care for infants to progressively becoming an educator and support person. Preparing to move from paediatric health to adult services takes time and effort, much the same as education process from pre-school and primary school to high school and beyond.

What you can expect

- > The Women's and Children's Hospital (WCH) CF team will start to talk with you and your child (when they are around 12 years old) about planning for the time when they reach adult age and will be transferring their care to the specialist CF team at the Royal Adelaide Hospital (RAH).
- > Both the WCH and RAH CF teams will help your child develop the skills and confidence to successfully manage their own care as an adult (including for example managing their diet and exercise, making clinic appointments, keeping prescriptions filled) and provide ongoing support to families and carers.

Genetic counselling

Families with children with CF and adults with CF may wish to have genetic counselling. They can therefore, at any time, be referred to the SA Clinical Genetics Service by CF medical staff or their General Practitioner. This service will provide them with specialist genetic counselling and support.

Lung transplant and palliative care

When people with CF have very significant and deteriorating lung disease that cannot be improved, the potential for lung transplant will be explored. People will be referred to Palliative Care specialists who will work closely with the specialist CF team in providing care when referred for a lung transplant to when their health has significantly deteriorated.

What you can expect

- > Your respiratory physician (lung doctor) and the specialist CF team will discuss the possibility of being assessed for a lung transplant.
- > If a lung transplant is appropriate, you will be referred to the South Australian Lung Transplant Service who will work closely with the specialist CF team in assessing and preparing you for lung transplant.
- > All lung transplants are performed in hospitals interstate. Most people from Adelaide attend The Alfred Hospital in Melbourne.
- > After an intensive period of rehabilitation you will return to Adelaide where your care will continue to be managed by the South Australian Lung Transplant Service.

24/7 CF contact

From time to time unplanned things can happen and you need to be assured that you can contact someone who knows about CF at any time for support or information.

What you can expect

- > You will be able to contact someone with CF knowledge at any time during the day, at night, weekdays and weekends.
- > You will be given different contact numbers, depending on whether you attend the Women's and Children's Hospital or the Royal Adelaide Hospital.

Emergency care

If you/your child needs urgent medical attention you will need to go to a Hospital Emergency department as quickly as possible, usually by ambulance (make sure you discuss ambulance cover with the CF social worker).

What you can expect

- > You/your child will be assessed by the Emergency Medical Team.
- > The Emergency Department doctors will talk with one of the CF lung doctors for advice about you/your child's plan of care.
- > Your/your child's key contact person will follow-up with you and the CF specialist team.

For more information

Women's and Children's Health Network
72 King William Road
North Adelaide SA 5006
Phone (08) 8161 7000
www.wchn.sa.gov.au

Royal Adelaide Hospital
North Terrace SA 5000
Phone (08) 8222 4000
www.rah.sa.gov.au



www.ausgoal.gov.au/creative-commons



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