ACI NSW Agency for Clinical Innovation

MODEL OF CARE

Cystic fibrosis

Respiratory Network

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Abbreviations

ABF	Activity based funding
ACD	Advance care directive
ACFDR	Australian Cystic Fibrosis Data Registry
BMI	Body mass index
CF	Cystic fibrosis
CFNSW	Cystic Fibrosis NSW
CFRD	Cystic fibrosis related diabetes
CFSS	Cystic fibrosis specialist service
ChIPs	Chronic Illness Peer Support
ED	Emergency department
ENT	Ear nose and throat
FEV1	Forced expiratory volume in 1 second
GP	General practitioner
HITH	Hospital in the home
ICD 10	International Statistical Classification of Diseases and Related Health Problems, 10th revision
IPTAAS	Isolated Patients Travel and Accommodation Assistance Scheme
IRT	Immunoreactive trypsin
IVAD	Intravenous access devices
IVF	In-vitro fertilisation
KPI	Key performance indicator
LHD	Local health district
MDT	Multidisciplinary team
MRSA	Methicillin-resistant *Staphylococcus aureus*
PEG	Percutaneous endoscopic gastrostomy
SaO_2	Oxygen saturation
SHN	Specialty health network

Executive summary

There has been a rapid increase in survival of people with cystic fibrosis (CF) in recent decades. This has resulted in a change in the demand for, and type of, healthcare services required; namely an increased number of adult patients with CF and an increase in hospitalisation rates as disease severity progresses. As CF patients age, the complexity of health care increases, as well as the need to focus on multiple aspects of preventive care.

The *Cystic fibrosis model of care* aims to address the lifelong healthcare needs of people with CF in NSW – now and into the future. This means addressing how CF services are provided to ensure timely access to care, improved patient reported outcomes and reduced rates of unplanned hospitalisation for all CF patients.

Between November 2012 and April 2013, the Agency for Clinical Innovation (ACI) Cystic Fibrosis Working Group undertook a comprehensive diagnostic process to review how services were being provided to people with CF and to understand the patient and staff experience of care delivery. This involved a gap analysis of current services against the *Cystic fibrosis standards of care, Australia 2008*; site visits to review identified gaps in care; data review of NSW patients with CF; a review of literature relating to CF and multidisciplinary care; and interviews and surveys to assess CF patient experiences. Building upon this data, the *Cystic fibrosis model of care* was developed.

The *Cystic fibrosis model of care* supports integration of care across a designated network of skilled CF care providers. It recommends that the lifelong care of people with CF be led by a cystic fibrosis specialist service (CFSS). Patient centred care planning and effective communication and information sharing between the CFSS and a network of care providers is required. This network may include the patient's general practitioner, Cystic Fibrosis NSW, CF satellite facilities, hospital in the home (HITH) services, transplant units, palliative care services and local service providers such as paediatricians and psychologists. Clinically appropriate care settings and care providers are determined through shared decision-making on an individual basis. The model of care highlights where the use of telehealth can support episodes of shared care and enable patients to access their care closer to home.

The model defines a range of ways in which planned and unplanned care can be delivered in an annual cycle of care. In addition to the annual comprehensive review, all people with CF require regular, planned outpatient review by a CFSS multidisciplinary team with the frequency (minimum three times each year) determined by individual need.

To effectively manage acute episodes, people with CF and their care providers require a regularly updated exacerbation management plan. The plan clearly identifies individual early signs and symptoms, who to contact, any therapies that can be initiated outside the CFSS and urgent actions if not responding as expected. Decisions are made on an individual clinical basis about where acute CF episodes can be managed, which may include CFSS, a CF satellite centre, local HITH service or day only admission.

The model also identifies that CF patients have specific care needs at each life stage. For example, those who are newly diagnosed need education and referral to an appropriate CFSS to commence lifelong care. Children need support with things like commencing school and accepting daily therapies. Adolescents require help moving towards self-care and autonomy and transitioning to adult services. Adults on the other hand need additional support with travel, relationships, family planning and pregnancy. Those with severe disease require pain and symptom management, assessment for transplantation, psychosocial support and end-of-life care planning. These needs should be addressed by the CFSS, in collaboration with the patient, carer and members of their CF care network.

Implementation of the *Cystic fibrosis model of care* will involve collaboration between clinicians and managers within relevant local health districts (LHDs) and specialty health networks (SHNs). Additionally, a CF implementation advisory group will provide guidance for the development and implementation of statewide solutions.

The next steps involve supporting LHDs and SHNs to review local systems to ensure the activity based funding resource stream is attracting service delivery in line with evidence-based care. A needs assessment within each service will be undertaken in light of the *Cystic fibrosis model of care* recommendations. An implementation plan will be developed by LHDs and SHNs in partnership and with support from the ACI and the CF implementation advisory group. Finally, systems will be established to monitor progress and patient outcomes.

With appropriate implementation, assessment and review, it is hoped the *Cystic fibrosis model of care* will help to deliver improvements to service delivery across both the CF annual cycle of care and at specific life stages. Importantly it will help to better meet patient needs in terms of access to care, timeliness of care and providing components of care closer to home, especially for those living significant distances from the CFSS.

Contents

Introduction

There has been a rapid increase in survival for people with cystic fibrosis (CF) over recent decades, which has significantly changed the demographic of the CF population and demand for healthcare services in NSW. Clinicians and consumers raised the need to understand the current healthcare needs of people with CF and to identify ways that effective and patient-centred healthcare services could be delivered now and into the future.

Prior to the 1990's, CF was a rapidly progressive paediatric disease with few people reaching adulthood. Today, the average age of people with CF in Australia is 19 years and almost half of those with CF are now adults.[1] Over the last decade there has been a 35% increase in the proportion of the Australian CF population who are adults. The dramatic increase in survival is attributed to the provision of lifelong care by CF specialist multidisciplinary teams at designated CF centres and improved respiratory therapies and nutritional support. [2]

Cystic fibrosis specialist services (CFSS) provide the majority of inpatient and outpatient care episodes for people with CF in NSW. While paediatric CFSS have been established for several decades, the adult CFSS have developed informally and largely within existing resources to meet local patient needs. [3]

Despite significantly increased survival, the health status of people with CF will decline with age due to chronic lung infections and the impact of CF associated comorbidities which include CF related diabetes, pancreatic insufficiency, liver disease, renal impairment, anxiety and depression. The age at which health status deteriorates and the rate of decline are variable and amenable to change.

While people with CF live full and productive lives, they frequently report high levels of daily treatment burden and health-related costs that impact on their quality of life.[9]

In Australia in 2012, the median age at death for people with CF was 29 years.[1]

A 7-year analysis of admissions to NSW public hospitals for people with CF based on ICD 10 (International Statistical Classification of Diseases and Related Health Problems, 10th revision) codes (2005/6 – 2011/12) has shown:

- total admissions and bed days for all people with CF have increased by 3% per year
- the increase in overnight bed days for CF is mainly attributed to those aged 18 years or older
- admissions coded as CF with severe complications and comorbidities have increased by 20% per year
- overnight hospital bed days for people with CF aged 18 years and over, with severe complications or comorbidities, have increased 25% per year
- the greatest increase in the rate of hospitalisation in NSW has occurred in young females aged 15-25 years (34%) compared to young males (15%).

The *Cystic fibrosis model of care*

The aim of the model of care is to ensure that the lifelong healthcare needs of people with CF in NSW can be better met now and into the future. The *Cystic fibrosis model of care* provides a flexible framework for establishing an expanded service model in which the CFSS deliver the majority of CF care episodes and/or directly coordinate and oversee shared care within a designated network of care providers including CF satellite facilities, HITH, transplant units, palliative care services, general practitioners (GP) and local service providers across NSW.

The *Cystic fibrosis model of care* defines opportunities across both the CF annual cycle of care and at specific life stages where patient-centred, flexible service delivery formats may improve access to care, timeliness of care, or better meet individual patient needs especially for those living a significant distance from the CFSS.

Implementation of an expanded service model will require formal clinical and governance arrangements between local health districts (LHD) and clinicians, increased clinical capability and capacity within targeted services, efficient processes for knowledge and skills transfer, enhanced clinical information sharing and the use of telehealth technologies.

What are the consequences of not changing?

Demand projections show that existing CFSS, and especially adult services, will be increasingly challenged to provide all components of inpatient and ambulatory care for people with CF. Without service planning and changes to the way services are organised and delivered, clinicians and consumers anticipate people with CF in NSW will be increasingly likely to experience:

- reduced timeliness and access to care
- poorer health outcomes
- reduced quality of life
- increased rates of hospitalisation.

Section 1

Cystic fibrosis background

Cystic fibrosis is the most common inherited life-shortening disease affecting people in Australia with an incidence of 1 in 3000 births.[3] In 2013, there were 967 people living with CF in NSW, of whom 30% were living in regional and rural areas and 49% were adults.[1]

The prevalence of the CF gene mutation varies in different ethnic populations with the highest carrier frequency (1 in 25) in people of Caucasian or Ashkenazi Jewish descent versus a lower carrier frequency rate (1 in 160) in people from South East Asia. The incidence of CF is rare in Australian Aboriginal, Torres Strait Islander and Maori people.

When both parents are carriers of a CF gene mutation, there is a one in four chance that their child will inherit two abnormal CF genes and so will have CF. In NSW, CF carrier testing and genetic counselling is offered to first degree relatives and the partners of individuals diagnosed with CF. For those found to be carriers, further cascade CF carrier testing is then recommended for their first degree relatives.

While population level CF carrier screening is currently not recommended,[4] couples intending to have children, or those who are pregnant should be made aware of the availability of prenatal CF carrier screening. Antenatal screening and pre-implantation genetic diagnosis for CF may be offered to known CF carriers, targeted ethnic groups and within assisted fertility services.

The majority of new diagnoses of CF (> 80%) occur in infants within the first year of life and predominantly as a result of identification through the NSW Newborn Screening Program and subsequent testing. The remainder are diagnosed as children, adolescents or adults as the result of an acute clinical presentation or following the diagnosis of a sibling.

Cystic fibrosis affects the regulation of sodium and chloride on epithelial cell surfaces within the body which causes abnormally thick mucus to develop. This leads to progressive systemic disease predominantly within the lungs and pancreas, and to varying degrees in the gastrointestinal tract, kidneys, liver, bones and reproductive organs. People with CF have a far greater risk of developing associated comorbidities including CF related diabetes, vascular complications, drug toxicity, depression, anxiety, cancer, arthritis and organ failure.

People with CF have complex and evolving multi-system healthcare needs across their life span. To achieve optimal health outcomes, they require regular planned specialised multidisciplinary team based care, rigorous daily therapy regimes and psychosocial support across their life span. Evidence shows that CF care is best delivered and coordinated through CFSS.[2,5] In NSW, there are three paediatric CFSS and three adult CFSS.

People with CF are frequently admitted to hospital, with one in five infants and children, one in two adolescents and one in four adults admitted to a NSW hospital at least once during 2012.[1] People with CF have relatively long episodes of hospitalisation with the average length of stay for those with CF and severe complications or comorbidities being 12 days (three times the average length of stay for all patients admitted in NSW public hospitals) and 9 days for those hospitalisations for CF without severe complications or comorbidities.[6]

There is no cure for CF and the severity of disease and burden of disease increase at a variable rate with age. Genotype specific therapies are an emerging field in CF management which have been shown to reduce disease progression within the specific patient groups. Current treatments aim to delay the decline in organ failure and to prevent and manage comorbidities across the life span.

1.1 Severe disease and respiratory failure

For a significant proportion of people with CF, predominantly adults but also some children and adolescents, their disease will progress to severe stage lung disease with associated respiratory failure. Patients approaching this stage are referred by CFSS for transplantation assessment, primarily bilateral lung transplantation but also liver transplantation. Adults are referred to the St Vincent's Hospital Heart and Lung Transplant program and children are referred to the National Paediatric Transplant Unit at the Alfred Hospital in Victoria.

To date, over 200 people with CF have received lung transplantation at St Vincent's Heart and Lung Transplant program in NSW. Median survival for people with CF receiving a bilateral lung transplant in NSW is now greater than 10 years.

Improved post-transplant survival has resulted in another important patient demographic change, with an increasing cohort of CF patients surviving a decade or more post-transplant and requiring ongoing complex care and follow-up by both their transplant team and with their respective CFSS.

Despite advances in therapy and transplantation, end stage airflow obstruction and bronchiectasis leading to respiratory failure remain the predominant cause of premature death in people with CF.

1.2 Cystic Fibrosis NSW

Cystic Fibrosis NSW is the peak consumer organisation supporting and representing people with CF and their families in NSW.

CFNSW provides specialist social work, counselling and practical support services as part of integrated care plans developed in collaboration with the CFSS. The services are partially funded by a grant provided by NSW Health. Additional supports provided by CFNSW include travel costs associated with outreach services, ongoing CFSS professional development and administrative assistance with data collection for the Australian Cystic Fibrosis Data Registry (ACFDR).

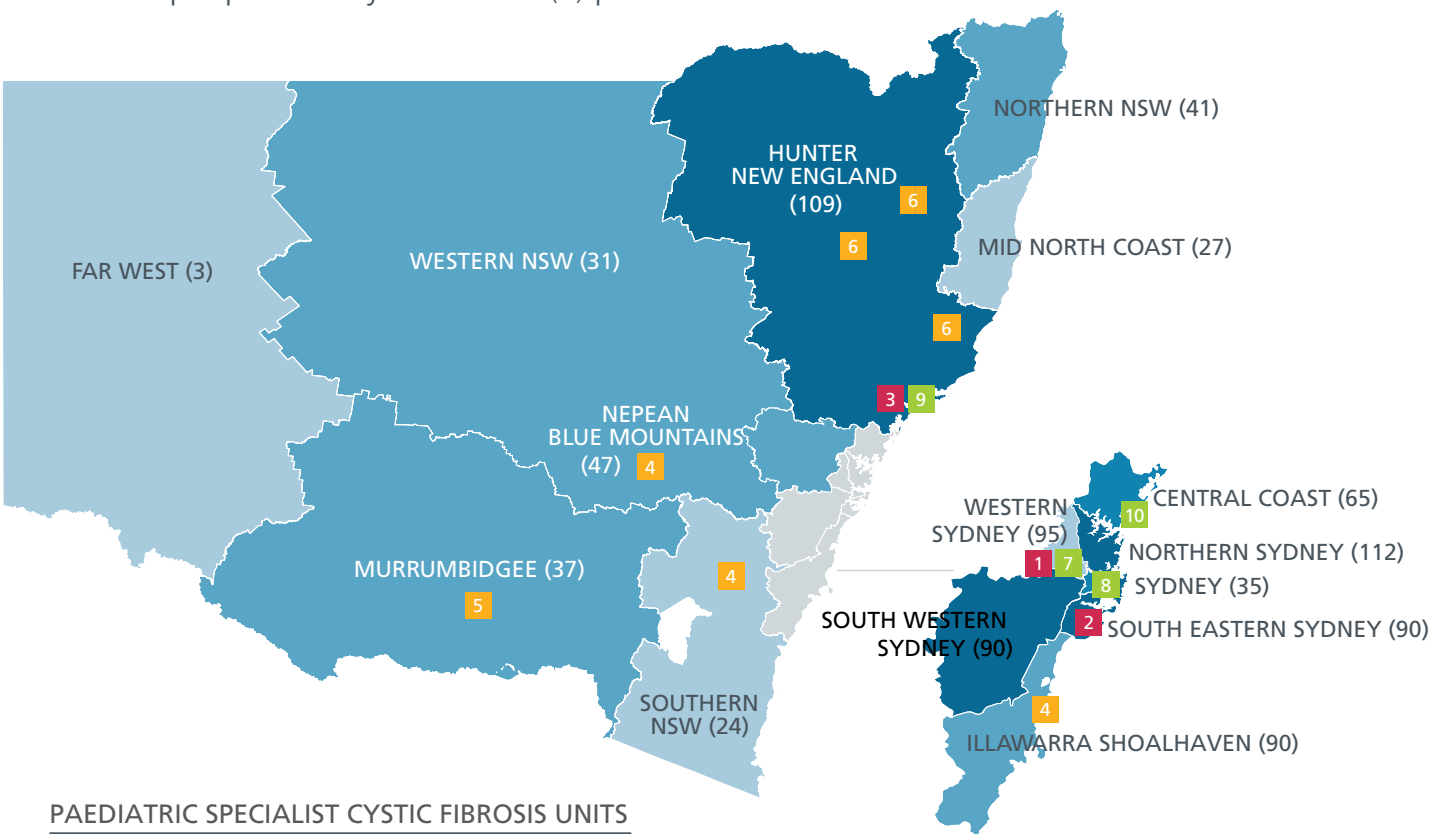
1.3 Cystic fibrosis standards of care in Australia

The *Cystic fibrosis standards of care, Australia 2008* have been endorsed by the Thoracic Society of Australia and New Zealand, the ACI Respiratory Network and CFSS across Australia as best practice guidelines.[7] Additionally there is a national independent peer review process in place which enables aspects of CF care provided by CFSS centres in Australia to be periodically assessed against the standards of care.

1.4 Australian Cystic Fibrosis Data Registry

All CFSS in Australia collect and submit standardised, de-identified patient data reflecting service delivery, health status and markers of quality of life to the ACFDR annually. The ACFDR collated data at individual CFSS level is publically available and used to monitor trends in patient outcomes and care processes and to enable benchmarking between all the CFSS within Australia and internationally.

Number of people with cystic fibrosis (n) per local health district*



PAEDIATRIC SPECIALIST CYSTIC FIBROSIS UNITS

1. Children's Hospital Westmead (WSLHD)
2. Sydney Children's Hospital, Randwick (SESLHD)
3. John Hunter Children's Hospital, Newcastle (HNELHD)

PAEDIATRIC OUTREACH SERVICES

4. Goulburn, Orange, Nowra (Children's Hospital Westmead)
5. Wagga Wagga (Sydney Children's Hospital)
6. Armidale, Tamworth, Taree (John Hunter Children's Hospital)

ADULT SPECIALIST CYSTIC FIBROSIS UNITS

7. Westmead Hospital (WSLHD)
8. Royal Prince Alfred Hospital, Camperdown (SLHD)
9. John Hunter Hospital, Newcastle (HNELHD)
10. Gosford Hospital (Satellite service)

KEY

- Small population (<30)
- Moderate population (30–59)
- Large population (60–89)
- Largest population (90–120)

** Based on the Australian Cystic Fibrosis Data Registry December 2012*

Figure 1. Number of people with cystic fibrosis per LHD

Section 2

Case for change

Analysis of NSW admitted patient data showed the majority of outpatient and hospital care for people with CF is provided by the three paediatric and three adult designated CFSS: Sydney Children's Hospital, Children's Hospital Westmead, John Hunter Children's Hospital, Royal Prince Alfred Hospital, Westmead Hospital and John Hunter Adult Hospital. There is one satellite service at Gosford which provides shared care episodes for selected local patients under the direction of the patient's CFSS.

Cystic fibrosis admissions to other hospitals occurred across 63 hospitals, with the majority having less than 10 admissions over the 7-year period. Only a small proportion of the NSW CF population (2%) are admitted to hospitals in other states and 2% of patients with CF who are admitted to NSW facilities are interstate residents.

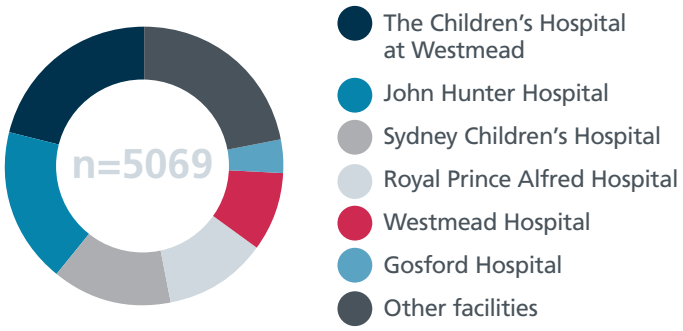


Figure 2. Number of CF hospital separations, by facility, between 2005-06 and 2011-12

Source: *ACI Financial and Service Utilisation Analysis of Admitted Patients with Cystic Fibrosis in the NSW Public Hospital System*

While the role of CFSS as the provider of all care for people with CF has been pivotal in achieving the improvements in health outcomes for people with CF, it is increasingly evident that an effective and sustainable model is required to maintain patient outcomes in the context of increasing demand and finite CFSS resources. To date there has been no statewide approach to service planning for the relatively small CF population who have evolving complex lifelong healthcare needs and relatively high healthcare utilisation and both patient and system costs (see Section 3.4).

2.1 Change in healthcare demand

The rapid increase in the life expectancy of people with CF over recent decades has led to significant change in the demand for healthcare services in NSW. The proportion of CF patients who were adults in NSW in 2013 was 49% and this is projected to increase in line with trends in life expectancy. The changes in healthcare demand arise from:

- increasing life years of healthcare required by people with CF
- increased focus on preventive care to avoid or delay deterioration in health status
- increasing number of adults living with CF
- increasing complexity of care as people with CF age
- increased hospitalisation rates as disease severity progresses.

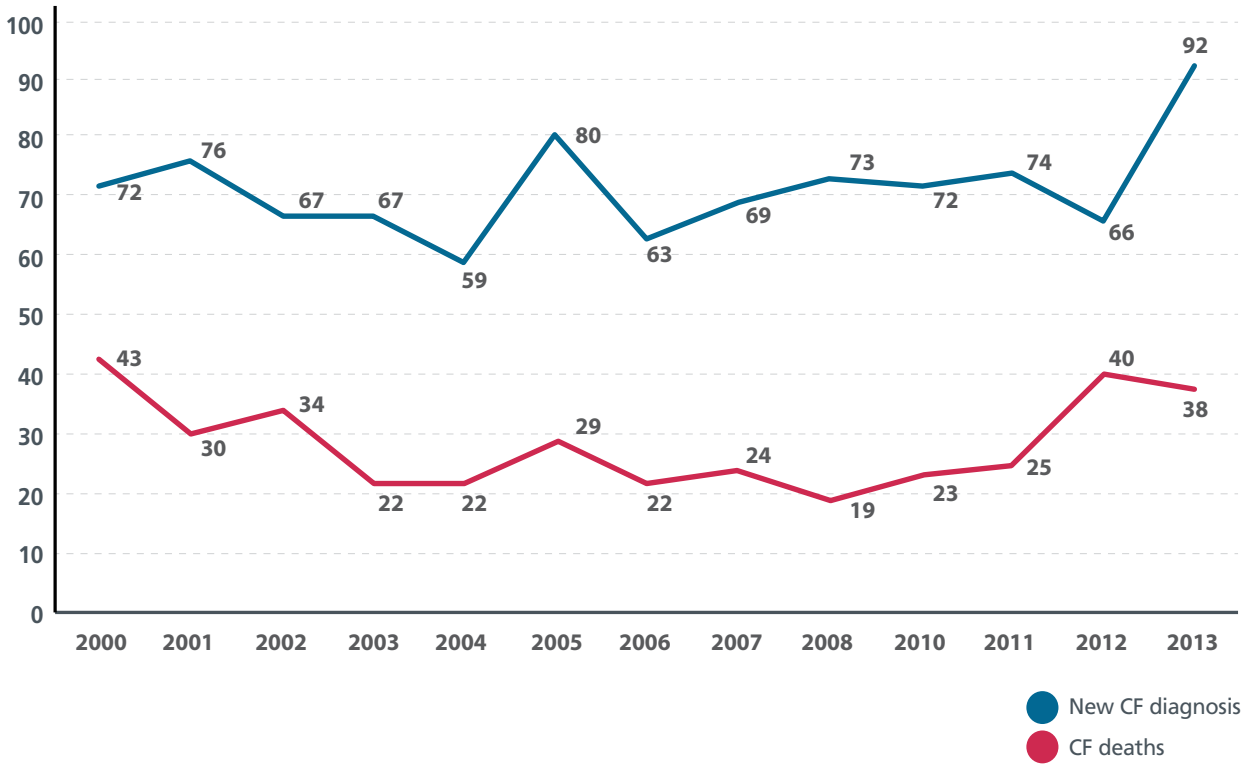


Figure 3. Cystic fibrosis new cases compared to deaths, Australia 2000-2013
Source: *Australian Cystic Fibrosis Data Registry Reports 2000-2013*

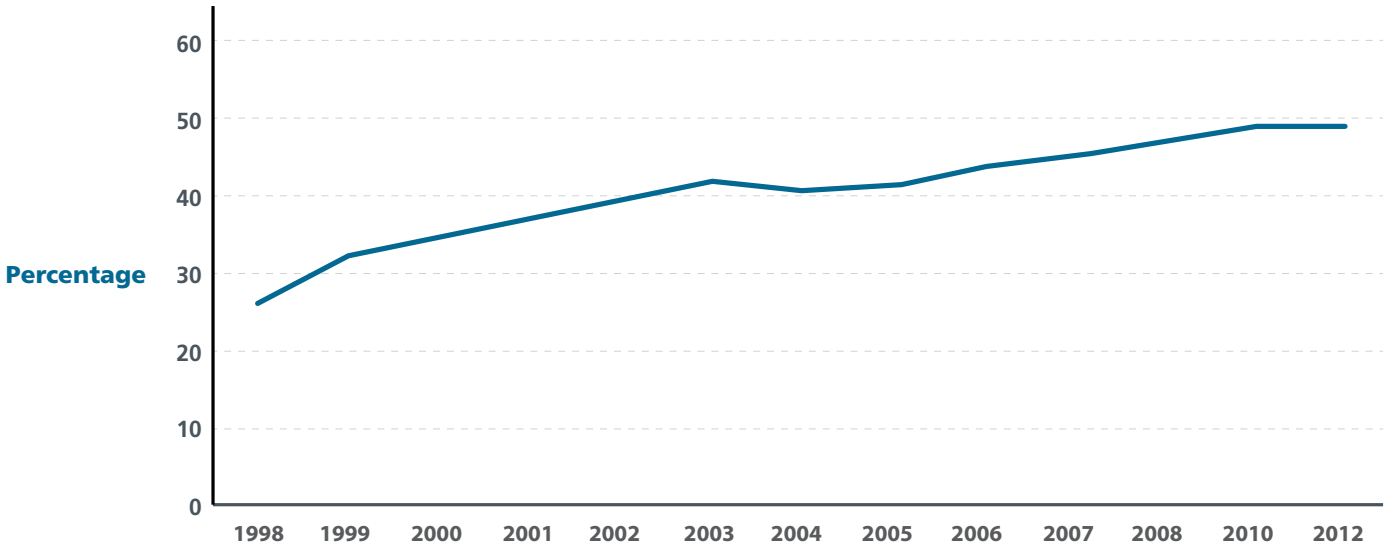


Figure 4. Proportion of adults in Australian CF population
Source: *Australian Cystic Fibrosis Data Registry as at December 2012*

In 2012 the median age of death was 29 years. The current life expectancy for an Australian person with CF is 37 years which reflects that most people with CF today will need more life years of healthcare to be provided by an adult CF service than a paediatric CF service.

2.2 Projected demand

The ACI health economics and evaluation team projected the service utilisation within NSW public hospitals for the CF population over a 10-year period. Projections show the overall number of hospital separations and bed days for people with CF will grow at approximately 2% per annum.

The most significant increase in utilisation of hospital based care is expected to occur for those patients aged 18 years or older, with severe CF complications and comorbidities, with the current trend in separations and bed days in this group expected to continue to grow by approximately 7% each year for the next 10 years.

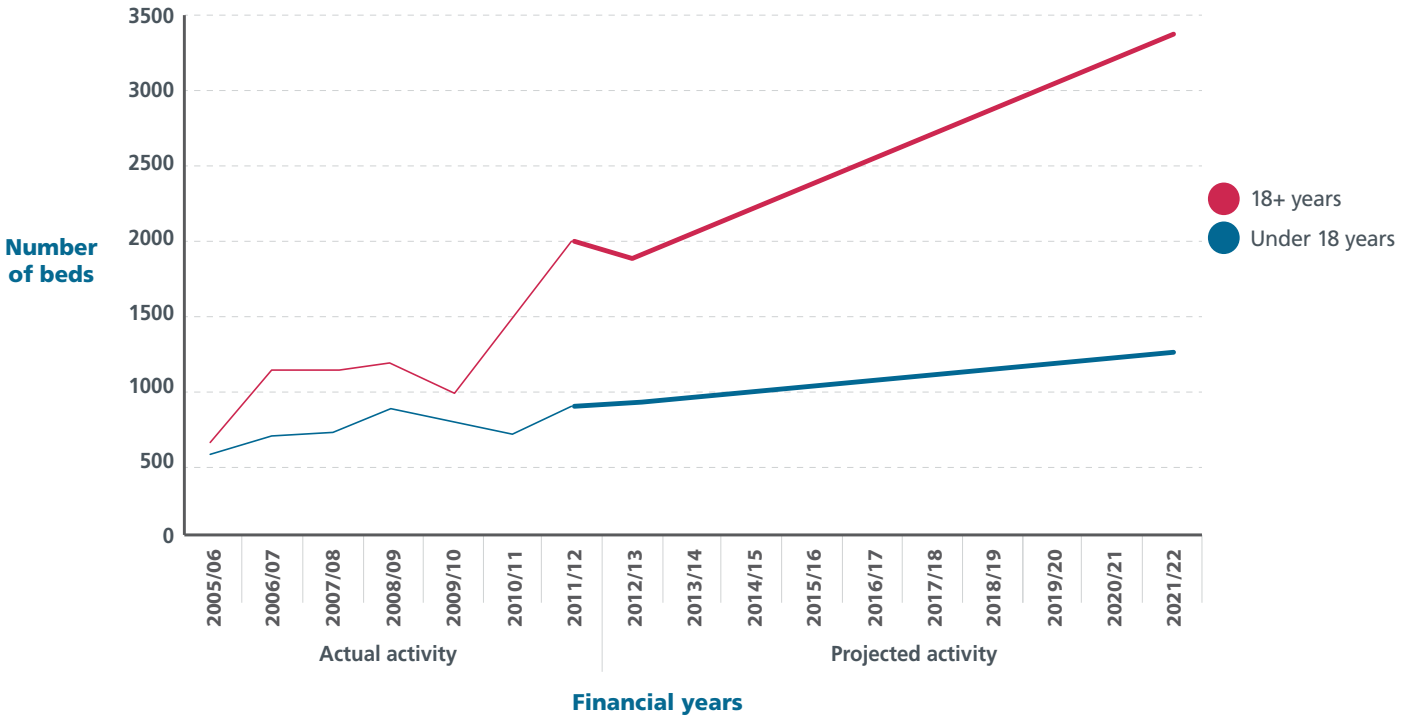


Figure 5. Projected hospital bed day utilisation for patients with CF (aged under 18 years; and 18 years or older) with severe complications and comorbidities

2.3 Hospitalisation trends

The analysis of admitted patient data for all people with CF over the past seven years (2005-6 to 2011-12) in NSW public hospitals has shown:

- total hospital admissions and bed days for all people with CF have increased by 3% per year
- the increase in overnight bed days for CF is mainly attributed to those aged 18 years or older
- admissions coded as CF with severe complications and comorbidities have increased by 20% per year
- overnight hospital bed days for people with CF aged 18 years or older, with severe complications or comorbidities have increased 25% per year
- the greatest increase in the rate of hospitalisation in NSW has occurred in young females aged 15-25 years (34%) compared to young males (15%).

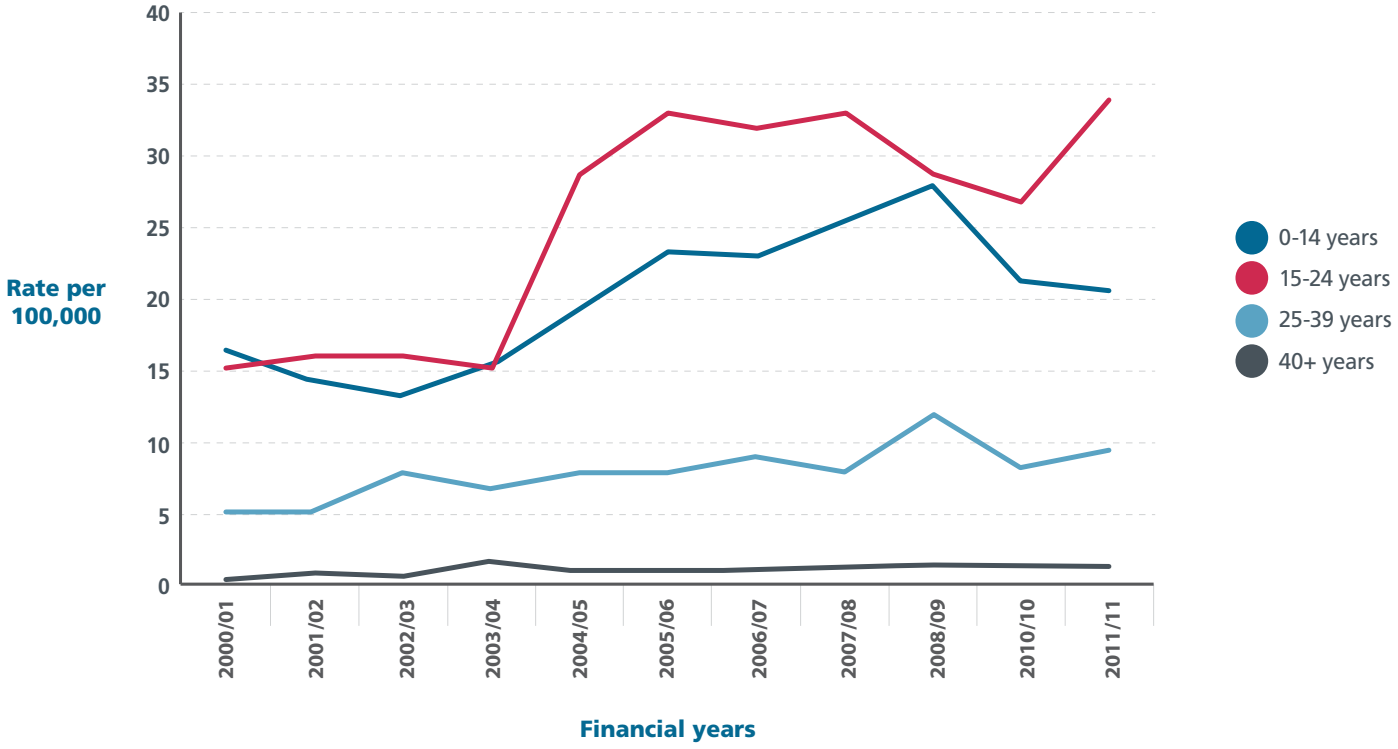


Figure 6. Age specific hospital rates of CF hospital separations (unadjusted) in females, by financial year and broad age groups
Source: *ACI Financial and Service Utilisation Analysis of Admitted patients with cystic fibrosis in the NSW public hospital system.*

2.4 Healthcare utilisation and costs by disease severity

Healthcare costs for people with CF in Australia from 2003-2005 showed hospital based care and total healthcare costs increase in line with the severity of lung disease. The greatest increase on healthcare costs occurred when CF disease severity progressed from severity level 1 to severity level 2, which predominantly occurs in young adults aged 18-29 years. Hospital sector costs as a proportion of total costs increased from 50% to 77% between those with mild disease classified as severity 1 and those who were post transplantation classified as severity 4.[8]

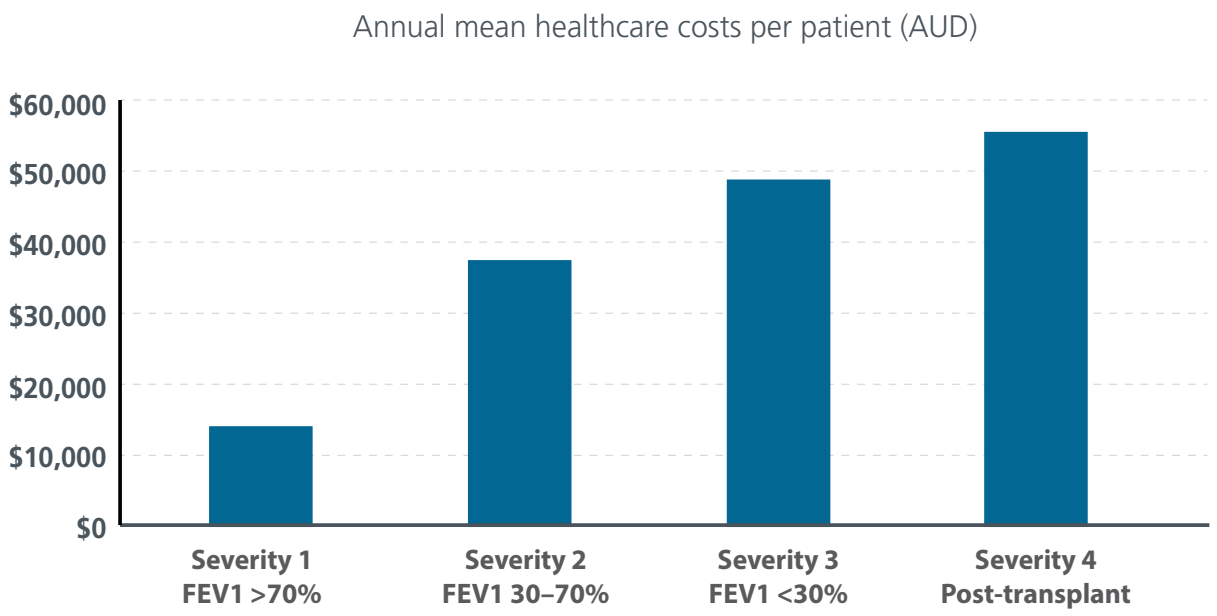


Figure 7. Annual mean healthcare costs per patient based on severity of disease

Adapted from: *Understanding the costs of care for cystic fibrosis: an analysis by age and severity.* March 2011, Centre for Health Economics Research and Evaluation.[8]

2.5 Quality of CF care in NSW

National standardised data from the ACFDR for 2011 was extracted to compare the key clinical indicators of health status: forced expiratory volume in 1 second (FEV1) for lung function; and body mass index (BMI) for adults or BMI z-score in children for weight, across age bands for people with CF in NSW with those in other Australian states.

Overall, NSW patient outcomes compared favourably with other states, with NSW being one of the two best performing states for both weight and lung function measures across the majority of age groups. The outcomes for CF patients in NSW that were less favourable than other states were lung function (FEV1) in males 30 years and over and weight (BMI z-score) for adolescents aged 12-17 years.

2.6 Patient needs and expectations

People with CF today can expect to live full and productive lives. People with CF and parents of children with CF require healthcare services that are effective, provided at the optimal time and organised in ways that work for them.

In 2012, over a third of Australian adults with CF were married or in a de-facto relationship and one in seven were living with their parents. Most people with CF complete school and one in five adults have attained university qualifications. Two thirds of adults with CF are in full or part-time employment.[1] While people with CF live full and productive lives, they frequently report high levels of daily treatment burden and health-related costs that impact on their quality of life.[9] Additionally people with CF experience frequent health-related disruptions to their family, school, study, work and social life.

For adults with deteriorating health status, the ability to maintain employment becomes increasingly difficult at the same time as their medical needs increase and associated costs steadily rise. These changing circumstances present considerable and increasing financial and psychosocial burden and an increased need for support services.

2.6.1 Patient experience

Patient experience tracker devices were used to survey people with CF and parents of children with CF (n = 62) during scheduled CF clinic visits in 2013 at multiple adult and paediatric CFSS in NSW. The devices only allow for five questions and one response per question.

They were asked the following.

'The time I am expected to spend managing CF each day is?'

- A total of 6% responded 'very little'.
- A total of 32% responded 'little'.
- A total of 52% responded 'a lot'.
- A total of 10% responded 'too much'.

'I would like more options for care to be provided by'.

- A total of 44% identified hospital in the home services.
- A total of 23% indicated their GP.
- A total of 19% identified more options for care via computer.

'Over the last year I have missed some CF appointments, treatments or medications mainly due to?'

Of those surveyed, 60% had missed appointments, treatments or medications over the last year mainly due to the following.

- A total of 35% identified that they were too busy.
- A total of 10% were due to the distance required to travel.
- A total of 8% were due to how much it costs.
- A total of 6% were not convinced they needed it.

'Does my CF healthcare team understand and focus on my needs and priorities?'

- A total of 56% responded 'always'.
- A total of 32% responded 'most of the time'.
- A total of 6% responded 'sometimes'.
- A total of 2% responded 'never'.

'Overall, how would you rate your healthcare experience?'

- The majority (84%) rated their healthcare experience as excellent or good, 13% average and 2% very poor.

Semi structured interviews were conducted with individuals with CF or the parent of a child with CF, which reflected that changes in demand for CF services have impacted on their perceptions of care.

The common consumer themes identified were:

- limited access to, and time available with, the specialist CF health professionals
- timeliness of care and increasing wait times, especially for planned admission
- gaps in care including post discharge support and home based care
- need for flexibility of where, when and how care could be provided.

> "The CF specialist service lacks enough staff – the quality is great but not enough of them for all the adults. This means you are limited as to when and for how long you can see them."
>
> **Young adult**

> CF clinic care is not what it used to be and you feel processed as there are more adult patients each year. The care is rushed and not as personal.
>
> **Adult**

> CF clinic – a whole day is lost. A lot more could be done over the computer.
>
> **Mother of a 2.5 year old**

> I am out of pocket $60/fortnight just for travel and parking for CF care. I would value more options for care that does not involve me having to travel.
>
> **Adult, on the transplant wait list, living in regional NSW**

> My priority is to live a full life – I work full time and it is difficult to organise things when I need to come into hospital for tune ups four times a year. Sometimes I wait 2-3 weeks to get a bed.
>
> **A 30 year old**

> My son is lucky; he has just started as an apprentice but feels he doesn't want to jeopardise his job by asking the boss for multiple days off to come to Sydney for all his CF clinic reviews.
>
> **Mother of a 17-year-old boy in regional NSW**

Section 3

Methodology

3.1 Project initiation

Consumers and clinicians identified the need for a statewide review of CF service provision to inform a planned and coordinated approach to statewide service delivery, with the aim to ensure the health system can meet the healthcare needs of people with CF now and into the future. In response ACI convened a Cystic Fibrosis Working Group to provide expert advice and guidance for the development of a *Cystic fibrosis model of care*.

The aim of the *Cystic fibrosis model of care* is to improve timely access to effective and patient-centred care for people with CF who live in NSW. The *Cystic fibrosis model of care* describes the healthcare needs of people with CF across the life span and provides a flexible service delivery framework which details how healthcare services may be organised and delivered to people with CF. All care described within the model of care is consistent with evidence based guidelines and standards which have been endorsed by clinicians and peak professional organisations across Australia including:

- *Cystic fibrosis standards of care, Australia* (2008)
- *Infection control guidelines for cystic fibrosis patients and carers* (2012)
- *Physiotherapy for Cystic Fibrosis in Australia and New Zealand: A clinical practice guideline 2016*
- *Australian clinical practice guidelines for nutrition in cystic fibrosis* (2006)
- Australian and New Zealand Cystic Fibrosis Nurses Group. *Australia and New Zealand Nursing Standards of Care for Cystic Fibrosis 2015*.

3.2 Diagnostic

A comprehensive diagnostic investigation was undertaken between November 2012 and April 2013 to review how services are provided to people with CF and to understand the patient and staff experience of care delivery. The following diagnostic methods of inquiry were undertaken.

3.2.1 Gap analysis

Cystic Fibrosis Working Group representatives reviewed their respective service against each domain of the *Cystic fibrosis standards of care* to identify areas where evidence based care was currently not able to be consistently provided.

3.2.2 Site visits

The gaps that were identified in care were specifically targeted for review during the subsequent site visits. Extensive consultation occurred with respiratory physicians and paediatricians, nurses, genetic counsellors, adolescent health physicians, physiotherapists, dietitians, pharmacists, social workers and LHD service managers during visits to all CFSS in NSW, CFNSW, St Vincent's Heart and Lung Transplant Program, Trapeze service, NSW Newborn Screening and regional physicians and paediatric inpatient facilities involved in CF care.

3.2.3 Data review

Data was extracted and a retrospective review of NSW admitted patient activity for people with CF between 2005-6 and 2011-12 was undertaken. The ACFDR provided a 2011 report which compared NSW with other Australian states.

3.2.4 Literature review

A recent review of literature (2000-2013) was undertaken in key areas related to CF and multidisciplinary care.

3.2.5 Patient experience

To understand the current healthcare experience of people with CF both semi structured interviews and anonymous surveys using patient experience trackers were used with adolescents, adults and parents of children with CF across multiple sites.

3.3 Solution design

The Cystic Fibrosis Working Group members participated in a workshop where all the issues identified within the diagnostic phase were reviewed, themed and prioritised. High level solutions for each prioritised issue were identified through a brainstorming and collective mind mapping process. A solution matrix was developed which identified common solutions across the identified priority issues which informed the development of the *Cystic fibrosis model of care*.

Section 4

The Cystic fibrosis model of care

The *Cystic fibrosis model of care* details the service providers, where services can be provided and the key service deliverables from both an annual care perspective and across the specific life stage care needs of a person with CF.

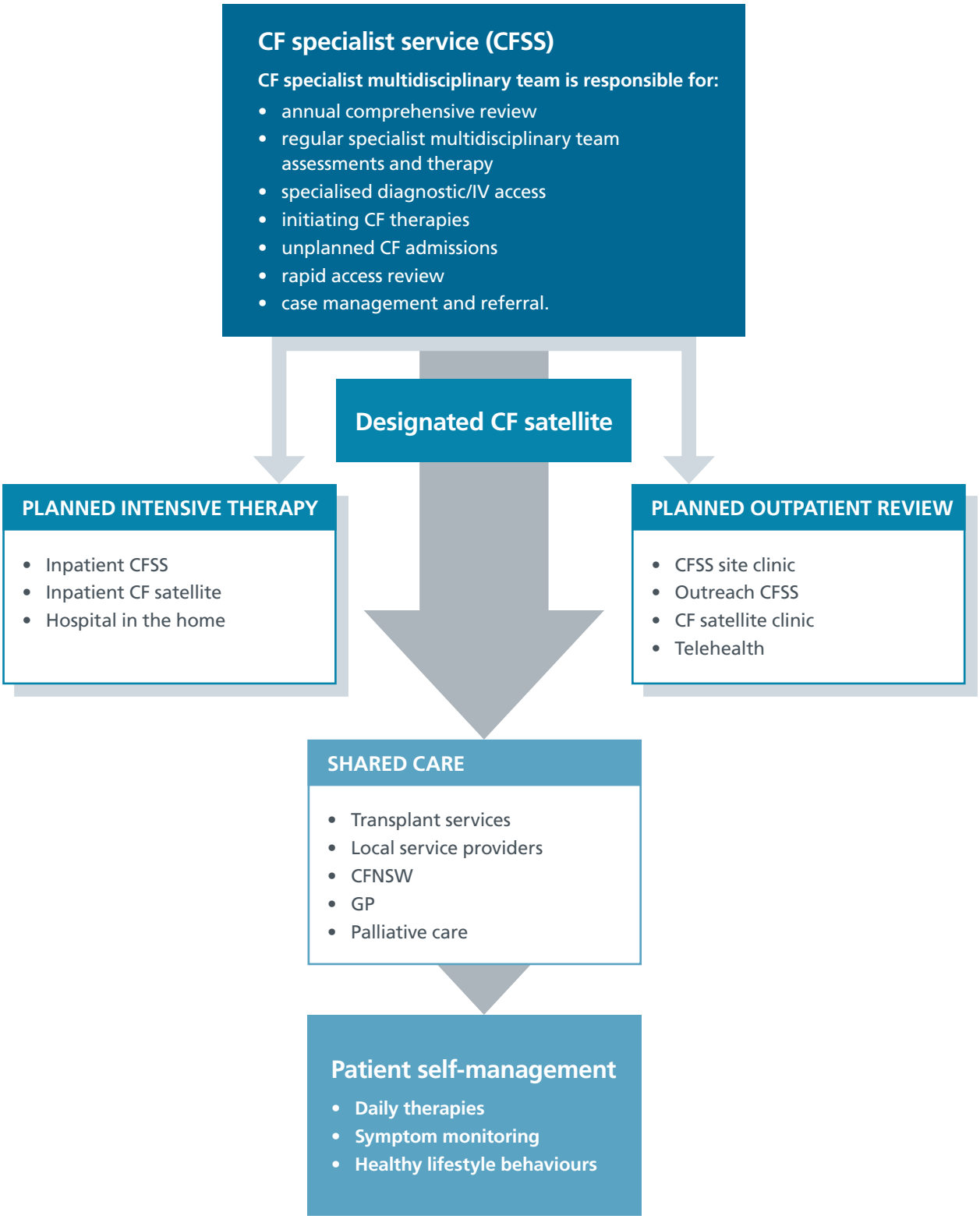


Figure 8. Cystic fibrosis model of care

4.1 Aims

The *Cystic fibrosis model of care* aims to implement a flexible service delivery framework which will:

- maintain or improve the health and wellbeing of people with CF
- increase access to the CF specialist multidisciplinary care team
- provide consumers with greater choice for how and where healthcare services can be safely and effectively delivered
- improve the care and support provided for people with CF across the life span, with additional focus on young people aged 15-24 years and those with severe disease.

4.2 Key principles

The *Cystic fibrosis model of care* is underpinned by four principles that will support integration of care.

- The lifelong care of people with CF is led by a CFSS.
- Care is provided by a multidisciplinary team (MDT) with knowledge and skills in the management of CF.
- Clinically appropriate care settings are determined on an individual basis.
- Clinical indicators are monitored to ensure status is maintained or improved throughout the implementation of the model of care.

4.3 Infection control

Chronic lung infections and colonisation by specific bacteria are the greatest predictor of deterioration in health status and premature death in people with CF. Therefore all areas within health facilities and HITH services that provide care for people with CF are required to have adequate facilities, plans and procedures established to prevent patient-to-patient contact or risk of cross infection. The CF specific infection control requirements are in addition to NSW policies and guidelines, *Hand hygiene* PD 2012_061; *Infection control* PD 2007_036; *Multi resistant organisms* PD 2007_084; *Environmental cleaning* PD 2012_061; and *Peripheral intravenous cannula insertion and post Insertion care in adult patients* GL2013_013.

Specific infection control procedures are required to be in place for:

- CF patients managed in ward areas with sufficient single rooms or the ability to nurse patients with CF in a room free of other patients with CF or respiratory infection
- physiotherapy treatment spaces
- outpatient clinic areas including sufficient single consulting rooms and ability to schedule patients with specific lung infections to designated clinics and times
- shared patient waiting areas
- treatment rooms used for procedures
- rapid review assessment areas
- pulmonary function laboratories.

4.4 Working with Aboriginal people

The impact of CF on the NSW Aboriginal population has been considered in the development of the model of care and summarised in an Aboriginal health impact statement.

Aboriginal people are less likely to carry the gene mutation, have CF and need for the services detailed in the model of care than the general population.

Nonetheless, there are Aboriginal people with CF in NSW and it is important that all steps are taken to ensure that services are delivered in culturally safe and competent ways across the life span.

To achieve optimal health outcomes for Aboriginal people with CF, the CFSS will need to undertake a cultural audit to identify and address the barriers for Aboriginal people related to access to care and ongoing self-management.

A cultural audit should specifically focus on ensuring:

- there are clear opportunities for Aboriginal people to be identified
- a demonstrated commitment (understanding, respect, listening and communication skills and sufficient time) to the development of trust with individual Aboriginal clients and Aboriginal communities
- screening and assessment processes are used that recognise the holistic approach to health that is shared by most Aboriginal people and communities
- education processes and resources are used that will build the literacy, engagement and empowerment of Aboriginal people
- linkages and referral processes are established with appropriate health and support agencies, with a particular focus on the unique role that Aboriginal medical services can provide for Aboriginal people within a network of service providers
- follow-up arrangements, both formal and informal, are individually tailored to ensure Aboriginal people are receiving the services required and are being supported to effectively manage their health.

To overcome the specific and evolving barriers to lifelong care that Aboriginal people with CF may experience, CFSS will need to work in partnership with individual Aboriginal healthcare providers (for example, Aboriginal health workers and Aboriginal liaison officers), Aboriginal health services and primary care services; and individually tailor care planning to achieve optimal health outcomes for Aboriginal people with CF.

For further guidance, refer to the Chronic care for Aboriginal People model of care.

4.5 Cystic fibrosis specialist services

The lifelong care of all people with CF should be provided by, or directly coordinated by, a CFSS as this service model has been shown to result in better clinical outcomes for patients based on lung function and BMI.[2] All people with CF are linked to a CFSS from the point of diagnosis, as evidence shows that they will have optimal health outcomes and increased survival when care is managed by a CFSS.[9-11]

A CFSS is a clinical service located within a tertiary referral hospital which has access to staff and facilities capable of managing all the systemic manifestations and complications of CF (see Appendix 1). To maintain a high level of knowledge and skills, a CFSS requires a minimum caseload of 50 patients. The *Cystic fibrosis standards of care* provides recommended staffing levels (based on 50-75, 75-150 and > 150 patients) for adult and paediatric CFSS (see Appendix 1).

Typically, a CFSS would need the following.

- It should have a designated MDT who are highly experienced in CF care and manage a large caseload of CF patients. The designated CF team members should meet regularly to plan and review the care of patients attending the service.
- A CFSS centre should have access to consults and ongoing review from medical and surgical specialties including gastroenterology, endocrinology, rheumatology, infectious diseases, psychiatry, ENT (ear nose and throat), obstetrics and gynaecology, adolescent health and palliative care – located within the tertiary facility to support the management of CF non-respiratory comorbidities and complications.
- Ready access to specialised diagnostics and procedures is needed.

- Ready access to telehealth technology is needed.
- Ready access to referral centres for lung and liver transplant services is needed.
- A CFSS centre should have an active research and audit program that allows evaluation of both the effectiveness of current care and outcomes of new therapies.

In NSW, there are three paediatric and three adult CFSS established at tertiary hospitals which oversee the lifelong care of people with CF.

4.5.1 Staffing

It is essential that all members of CFSS MDT maintain up-to-date specialised knowledge and expertise in the management of all manifestations of CF and are abreast of the latest CF research. The staffing compliment for a CFSS should include the following.

- A CF medical specialist has extensive CF specific experience, knowledge and the clinical skills that are required to assess and manage all manifestations and comorbidities of CF; and provides clinical leadership and governance for the MDT. The CF medical specialist is involved in national and international CF clinical meetings and provides leadership for the delivery of best practice across all aspects of patient care.
- A specialist nurse provides advocacy and psychosocial support for patients and families, as well as case management, especially at key times including at diagnosis, commencing new treatments, hospital admissions, onset of comorbidities, reproductive issues, transition, pre- and post-transplant and end-of-life care planning.
- A specialist physiotherapist provides expert knowledge and skills in inhalation therapy, airway clearance techniques, non-invasive ventilation, musculoskeletal care and exercise including pelvic floor strengthening.
- A specialist dietitian provides expert knowledge and skills in hydration, glucose tolerance, pancreatic enzymes, dietary supplements, nasogastric or gastrostomy feeding and energy demands.
- A clinical psychologist provides cognitive, behavioural and family therapies and referral to mental health services where appropriate.
- A social worker provides grief and loss counselling; information and assistance with social security entitlements; referral to social, housing and support services; liaison with schools, universities and employers on behalf of people with CF; health coaching; support for patients and families at key milestones and at end-of-life; and access to equipment and emergency financial assistance.
- A geneticist/genetic counsellor assists with specific CF mutation diagnosis, cascade screening, counselling families who have a child with CF or relatives of a person with CF at key stages including diagnosis and when considering family planning.
- A respiratory scientist provides complex lung function testing and specialised assessments.
- A clinical pharmacist assists in the assessment of pharmaceutical aspects of therapy, potential drug interactions, allergic phenomena and adverse drug reactions. The pharmacist provides ongoing education for patients and clinicians and strategies to optimise adherence to therapy.
- Specialist interventional radiology should be available on-site to perform bronchial artery embolisation.

4.5.2 CFSS responsibilities

The CFSS is responsible for providing overarching CF management and highly specialised CF patient care across the life span, and where considered clinically appropriate, to coordinate access to a designated network of shared care providers and directly supervise the delivery of episodes of shared care for people closer to home.

The CFSS is exclusively responsible for:

- initial care of all newly diagnosed patients
- monitoring and clinical management of all systemic manifestations of CF
- comprehensive annual review
- specialised diagnostics§ and procedures¥
- specialised MDT assessments and the initiation of therapy
- complex admissions
- rapid review clinic
- telehealth CF consultations
- data collection and reporting to ACFDR
- research
- establishing clear roles and responsibilities with relevant shared care providers.

4.5.3 Patient access to CFSS

At diagnosis, each person with CF is linked to a designated CFSS for ongoing CF case management, preventive care and management of acute CF episodes and related comorbidities. A person with CF, or parent of a child, requires a 7-day per week point of contact with a designated member of their CFSS team for clinical advice and ongoing education and self-management support. Contact may occur in a variety of formats including face-to-face, telehealth, phone, SMS or via computer.

Telehealth is the delivery of healthcare at a distance, using information and communications technology.[12]

4.5.4 Travel and accommodation assistance

People with CF who live in rural and regional locations may be eligible for financial assistance toward their travel and accommodation costs to attend CFSS clinic appointments. This assistance is available through NSW Government Isolated Patients Travel and Accommodation Assistance Scheme (IPTAAS).

4.5.5 CF satellite facility

A CFSS may identify the need for, and provide support to, establish and support a single or small number of designated CF satellite facilities in specific areas where there is a significant CF population with geographical limitations for accessing care at CFSS. A CF satellite facility requires a shared care caseload of > 20 patients and a designated multidisciplinary team (physician/paediatrician, nurse, physiotherapist and dietitian) with the demonstrated competency and capacity to deliver episodes of CF care in line with formal shared care arrangements with a CFSS. Processes for regular communication, information sharing and staff mentoring need to be established and maintained between CFSS and CF satellite.

A CF satellite facility, under the direction of CFSS, may be responsible for providing specific episodes of inpatient care or planned review for selected CF patients.

Establishment of telehealth capacity between a CFSS and CF satellite facility may further enhance opportunities for establishing and maintaining effective shared care arrangements and knowledge and skills transfer.

§ Diagnostics: bronchoscopy, lung function testing and interpretation of microbiology, sweat test or genotype analysis

¥ Procedures: Preserving intravenous access in CF patients is important for lifelong care and the CFSS is responsible for decision-making and planning for the insertion of temporary peripheral or central venous lines and implanted intravenous access devices (IVAD). As the majority of IV access procedures occur at the CFSS site, a dedicated service and theatre spaces should be available.

4.6 Shared care with GPs

While the CFSS are responsible for the majority of CF related care and treatment decisions, all patients with CF will need to establish a relationship with a GP. A GP caring for one or more CF patients will ideally develop a special interest in CF, agree clear roles and responsibilities with the relevant CFSS and maintain effective and timely two-way communication processes. Arrangements should be in place for CF patients to have accelerated access to their GP in the event that they are unwell and their access to the CFSS is delayed. The GP, especially in rural and regional areas, may be involved in pre-determined aspects of CF care in close consultation with the CFSS.

The GP should be kept informed of:

- CF related healthcare decisions
- CFSS/CF health summaries
- CF exacerbation management plans.

Key aspects of care provided by the GP include:

- holistic care of a person with CF
- providing annual referral and general health summary to CFSS and other specialists
- maintaining up-to-date vaccinations including influenza, pneumococcal vaccine and, where required, travel vaccinations
- early identification of individual psychosocial issues and referral to CFNSW or local services
- early identification of family issues or conflicts and referral for family therapy
- conducting general age appropriate preventive health assessments, for example cholesterol, BP, breast screening and pap smears
- re-enforcing adherence to CF treatment plan and liaising with CFSS where non-adherence is identified
- notifying CFSS promptly if a patient presents with acute respiratory or gut symptoms
- prescribing medications (CF related medications only under the direction of CFSS)
- initiating exacerbation investigations (for example, sputum culture) and management under the direction of CFSS when the CF patient is unable to rapidly access the CFSS
- providing contraception guidance as CF medication can interact with oral contraception methods
- identifying gynaecological issues and referring to a gynaecologist with CF experience
- heightened vigilance in screening for malignancies in CF patients post organ transplantation
- assisting with travel planning and letters for overseas care providers
- supporting end-of-life care, providing home visits and arranging 24-hour support in close collaboration with CFSS and/or local palliative care providers
- bereavement support for partners and families.

4.6.1 Shared care with other local care providers

Individual patient needs and circumstances may require specific shared care arrangements to be established between CFSS, CFNSW and local service providers, for example the physician, paediatrician, social worker, psychologist and palliative care service.

Shared decision-making by the CFSS team and the patient will determine if and when shared care arrangements are appropriate based on the patient needs; and that their clinical condition is considered sufficiently stable or predictable. The CFSS will need to ensure the local service provider has access to relevant clinical knowledge and detailed patient information for the relevant specific therapies, is provided with clinical education and support as required and agrees to accept the referral.

The referral should include a clear definition of responsibility for each service including medical governance, a management plan including escalation to CFSS, the appropriate CFSS contact details and a shared care plan review date.

Section 5

Annual cycle of care

Figure 9. Cycle of care

5.1 Self-management

Self-management is the cornerstone of effective CF management with recognition that patients and carers deliver the vast majority of therapy on a daily basis across their life span. Therefore all clinicians involved in the care of people with CF will require specific knowledge and skills to effectively engage with patients as partners and to educate and support them to effectively self-manage their health condition, recognise deterioration and access timely and appropriate review. Regular assessment of the individual patient's beliefs, goals and self-management capabilities should be embedded into all components of care.[13-15]

Where limitations to self-management, or an increased risk of non-adherence to therapies are identified, these patients and/or their carers should receive more frequent support from either the CFSS or be linked to local care providers or formal case management and health coaching services for additional support.

5.1.1 Treatment burden

People with CF experience a high treatment burden with daily nutritional intake requirements and supplements, respiratory therapies and airway clearance, medications and exercise regimes across the life span.[16-18] In a study of adults with CF, the median number of daily therapies was seven and the mean time that patients reported spending on treatment was 108 minutes.[17] Treatment burden is associated with decreased adherence to daily therapy. During intermittent acute episodes that are able to be managed at home, people with CF will have additional self-care expectations and increased frequency of their daily therapies. The majority (52%) of patients and carers surveyed during 2014 in NSW (n = 62) reported that they spent a lot of time managing their CF every day and 10% responded that they spent too much time.

It is important that consideration is given during all episodes of care to the burden versus benefit of specific therapies and that people with CF and their carers are supported to make informed treatment choices in the context of their priorities in life.

5.2 Annual comprehensive review

This is required for all patients with CF and must be provided at a CFSS facility where patients and carers have access to all members of the specialist MDT team. It involves assessment, team review and care planning.

The current needs of each person should be identified prior to their annual review (by phone or questionnaire) and be considered by the team during a case review and planning meeting prior to the annual comprehensive review visit. This will ensure that patient's concerns are addressed and wherever possible arrangements are made to schedule any additional investigations and consultations required during the annual review visit.

Pre-planning, coordination and scheduling of individual investigations and consultations by a designated clinic coordinator prior to the annual comprehensive review visit can significantly improve the efficiency of the process for both patients and clinicians.

Table 1: Routine assessments conducted at the annual comprehensive review

Assessment	Details
Respiratory status	Oropharyngeal or sputum culture for identification of *Pseudomonas aeruginosa, Burkholderia cepacia*, *Staphylococcus aureus*, methicillin-resistant *Staphylococcus aureus* (MRSA), mycobacteria and fungi such as *Aspergillus* and *Scedosporium* Spirometry Chest X-ray or chest CT scan as appropriate Review airway clearance techniques, inhaled respiratory therapies and cleaning of equipment Oximetry/arterial blood gases for those with moderate-to-severe respiratory disease Need for supplemental oxygen or non-invasive ventilation Exercise assessment
Biochemistry	Fat soluble vitamin levels (A, E, D), liver function, full blood count, urea, electrolytes, creatinine, total 'immunoglobulin E Oral glucose tolerance test (aged 10 years and over) or HBA1C where clinically indicated
Nutritional status	Assessment of dietary intake Review use of pancreatic enzymes, vitamins, salt replacement therapy Nutritional supplements Growth and BMI
Psychosocial	Depression Anxiety
Fertility and contraception	As appropriate
Procedures	Flushing IVAD Assessment of gastrostomy feeding tube
Preventive	Full medication review Musculoskeletal assessment and exercise programs including pelvic floor strengthening Audiology for patients receiving specific antibiotics Bone mineral density in adolescents and adults (repeated every 3 years) Enquiry as to presence of oral or vaginal candidiasis Transition preparation in age appropriate patients

5.2.1 Did not attend

In the event that a patient does not attend for their scheduled annual review, a follow-up phone call from the designated clinic coordinator is recommended to identify and resolve the contributing factors and to reschedule another appointment.

5.2.2 Involving patient and family in clinical management plans

It is important that people with CF and their family are equal partners in their healthcare and are supported to make informed decisions about their care. To achieve this, it is important that adequate time is provided for the patient and family to discuss all results, raise concerns and contribute to treatment and management decisions with all members of the MDT at the annual comprehensive review visit.

For regional and rural patients, identified by CFSS as clinically appropriate, specific discussion and decisions should be made in conjunction with patient and family regarding potential shared care arrangements for planned intensive therapy admissions or planned review to be provided by the multidisciplinary team at a networked CF satellite service during the next 12 months.

5.2.3 CF exacerbation management plan

As acute respiratory exacerbations occur frequently in people with CF, an individual exacerbation management plan developed in consultation with the patient and family is recommended. The exacerbation management plan is shared with local care providers (physician, GP) and where available uploaded to My Health Record.

The exacerbation plan clearly identifies:

- signs or symptoms that trigger the change in management
- specific changes to therapies and medications
- expected response and time frame
- further action and time frame required if there is insufficient response.

5.2.4 Educating the patient and family

Regular and age appropriate education, including written and online information will need to be provided to patients and their families in conjunction with all episodes of CFSS care. The core components of education include understanding their individual manifestations of CF, how to monitor their disease, setting treatment goals, preventing or controlling infections, correct use of medications and inhaled therapies, care and cleaning of their equipment, prevention and self-management strategies and how to access locally available support services.

Aboriginal and Torres Strait Islander people with CF should be offered the choice to involve a local Aboriginal Health organisation in their CF healthcare team. Local Aboriginal health workers or Aboriginal liaison workers can explore the needs of the individual patient, family and community and establish local culturally appropriate support systems.

People with CF from culturally and linguistically diverse backgrounds should be offered the choice to involve a translator or an appropriate bilingual health support person in their CF healthcare team.

5.2.5 Communication

At the completion of each annual comprehensive review clinic visit, a summary report is compiled, filed in the medical record and sent to the patient, their GP, CF satellite and other relevant local care providers.

5.3 Planned review: three or more per year based on clinical need

In addition to the annual comprehensive review, all people with CF will require regular planned outpatient review by a CFSS multidisciplinary team with the frequency (minimum three times each year) determined by individual need.

Where CFSS and designated CF satellite arrangements are established, a CFSS may coordinate access to the CF satellite for selected patients to receive specific planned reviews closer to home. Where CFSS and CF satellite arrangements are in place, the annual comprehensive review and at least one other planned review per year need to be provided at the CFSS.

Telehealth is increasingly being used in Australia and internationally to provide planned review for people with CF who have restricted access to CFSS clinics. Where a CFSS has available the technology and resources, access to telehealth supported planned reviews should be considered and offered to selected patients.

Table 2: Routine assessments and care planning conducted at a planned CF review

Assessment	Details
Respiratory status	Spirometry Sputum culture Oxygen saturation (SaO_2)
Biochemistry	Blood glucose monitoring Urinalysis if declining weight or taking oral steroids
Nutritional status	Growth in children BMI in adults
Psychosocial screening	As appropriate
Fertility and contraception	As appropriate
Procedures	Flushing IVAD Assessment of gastrostomy feeding tube
Preventive	As appropriate

5.3.1 Care planning performed during a planned CF review

Care planning performed during a planned CF review involves:

- clinical interpretation of results and longitudinal monitoring by the CF specialist team
- CF multidisciplinary case review
- review and amendment of the clinical management plan and written exacerbation action plan as required
- review and amendment of time to next planned review as clinically appropriate.

At each planned outpatient review, each patient has an individual consultation with a designated CF respiratory physician/paediatrician, nurse, physiotherapist and those with pancreatic insufficiency or nutritional issues will require consultation with a dietitian. Psychosocial and medication issues should be identified and processes established for timely access to a psychologist, social worker or clinical pharmacist for follow-up as required.

COMMUNICATION

A summary of each planned review visit is provided to the patient, family, patient's GP and where applicable, the CF satellite and local care providers.

5.3.2 Outreach clinic

Access to multiple planned reviews each year can be a significant issue for people with CF who live in rural and remote areas. Outreach clinics provided by CFSS staff in conjunction with CFNSW can improve access for the patient and their family to specialist CF clinician review and psychosocial support. In addition, outreach clinics provide CF specific clinical support and up-skilling opportunities for the local service providers. An outreach CF clinic should involve members of a CFSS team and CFNSW visiting a regional hospital or outpatient centre on an agreed regular annual schedule. Clinicians from both the CFSS and the local healthcare providers are present and involved in all the discipline specific aspects of the outreach clinic.

Standard requirements for a regional outreach clinic include:

- CF infection control standards being met
- local designated clinicians available and able to participate in consultations and case reviews
- access to microbiology testing
- access to spirometry
- availability of antibiotics and equipment commonly used in management of CF
- a point of contact for both CFSS and a local MDT team for patients and families
- access to follow-up psychosocial support
- scheduled time for the CFSS to provide CF clinical education to the local MDT at each visit
- sufficient patient numbers to justify the service: a minimum 10 CF patients is recommended.

COMMUNICATION

A process for ongoing two-way communication should be established with regular opportunities available for all health disciplines within the local MDT to participate in case reviews with the CFSS.

Telehealth is increasingly being used in Australia and internationally to provide planned review for people with CF who have restricted access to CFSS clinics. Virtual outreach clinic reviews can potentially reduce travel for both patients, families and clinicians and provide upskilling and mentoring support for local clinicians.

5.4 Managing acute CF episodes

People with CF experience relatively frequent acute episodes and may require planned and unplanned access to acute care interventions which may be provided in a variety of formats.

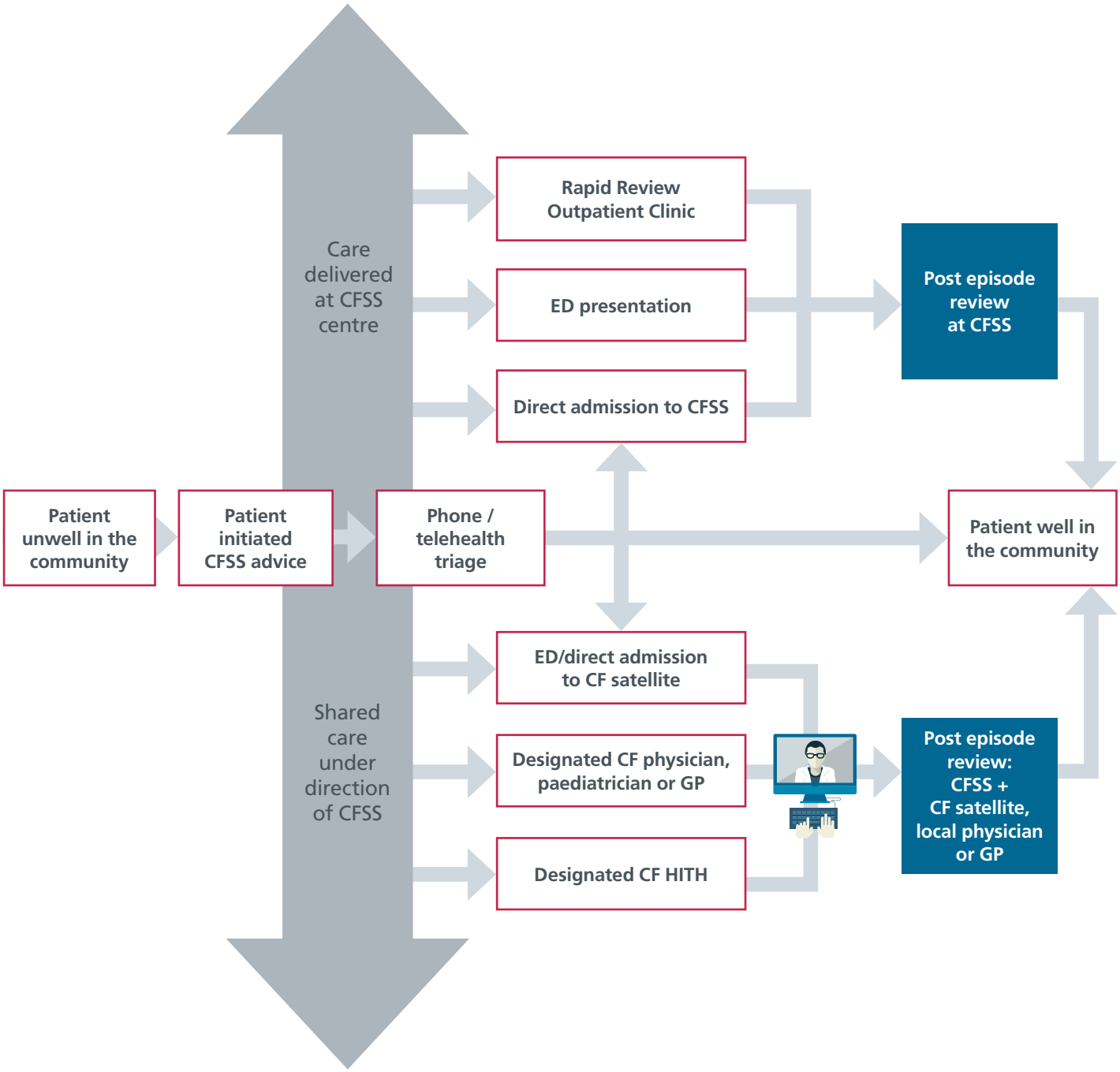


Figure 10. Managing an acute CF episode
ED = emergency department

5.4.1 CFSS advice

Patients experiencing early signs of an acute CF related episode (generally respiratory or gastrointestinal exacerbation) initiate contact with their designated clinician from their CFSS. An initial phone triage is undertaken to determine whether the patient's signs and symptoms can be managed by additional home based therapy or if they require further assessment at a rapid review clinic, direct admission to CFSS or presentation to an emergency department (ED).

5.4.2 Rapid review clinic

A rapid review clinic provides access to early triage for increasing signs and symptoms of acute episodes and may be provided as a CF specific service or as part of a broader respiratory rapid review service located at the CFSS facility. This service should be available at all CFSS.

On arrival at the rapid review clinic the patient undergoes a more thorough CF specialist nurse-led triage assessment process and if required a CF medical consult with further investigations and medication changes arranged as required. Patients who are able to return home should have arrangements made for phone follow-up and/or CFSS clinic review within 1 week.

CF clinicians involved in the existing rapid review clinics have reported that the majority of CF patients triaged are able to return home, the majority have responded well to early treatment and a significant number of admissions to hospital have been prevented.

> "I have taken my child to the CF rapid treatment room over 10 times now – it is really good, feels homely and my child likes it – we get sorted out and always go home."
>
> **Mother of a 2.5 year old**

5.4.3 Presentation to ED

The ED of hospitals that provide care for people with CF require the capability to manage CF complications and will have established processes in place that have been developed under the leadership of the relevant CFSS for:

- notification of CF medical specialist
- access to urgent CF specialist care
- investigations, treatment, appropriate IV access and monitoring of progress
- admission under a designated CF physician or joint admission for non-CF related health issues
- direct or fast track admission to designated ward/s
- access from ED to individual care plans, plans for IV access and other CFSS clinical information, either via electronic medical record or other means
- pre-arrival notification of the ED admitting officer, where CFSS refers a patient to ED, including specific initial assessment and treatment requirements and details for the admitting medical officer
- extended stays in ED due to bed availability – where CF patients are placed in a single room, preferably with protective isolation.

Where there is an extended stay in ED due to bed availability, the CFSS team should be notified, visit the patient and assume responsibility for care.

5.4.4 Inpatient care

The most common reason for a person with CF to have an admission to hospital is for intravenous antibiotics and intensive respiratory therapies for a respiratory tract infection. Other less frequent admissions are for pneumothorax, haemoptysis, gastrointestinal disease, liver failure, unstable diabetes, pre-operative care and psychological disorders. A significant proportion of admissions for people with CF are planned for intensive therapy and are often referred to as 'tune ups'.

Planned admissions may be considered appropriate for a direct admission process to a designated CF ward.

> "Sometimes when I am unwell I can only get a bed by coming to the emergency. You can be there for a day and even though I am diabetic, I did not get food regularly. My lungs were terrible and I asked them to get the physio to help me, but that didn't seem possible to organise in emergency."
>
> **Adult, on transplant wait list**

A person with CF who requires overnight hospital based care requires admission under a designated CF physician or paediatrician and should be assigned to a designated ward area within a CFSS tertiary facility or to a networked CF satellite hospital.

Requirements for a designated CF ward are:

- staff who have appropriate skills and are familiar with caring for people with CF
- availability of CF specific resources
- ability to provide appropriate infection control measures including single or cohorted room policy
- space for personal medical equipment that is required during admission
- space for the patient to exercise
- space and equipment for patients and families to store and prepare food
- access to members of a CF multidisciplinary team to provide ongoing input, especially a physiotherapist, dietitian and social worker.

5.4.5 Day only admissions

Selected patients may be suitable for day only treatment where a day only infusion centre or medi-hotel accommodation venue has been established which meets CF infection control requirements.

5.4.6 Self-administered intensive therapy

For a small number of selected patients, self-administered pre-prepared IV antibiotics and intensive respiratory therapy may be considered as a planned intensive therapy home treatment option. A formal documented protocol is recommended to ensure appropriate assessment for the individual patient's suitability to deliver IV antibiotics.

Patients or carers should receive appropriate training and supervision prior to commencing self-administered home treatment. Completion of a detailed written competency checklist is recommended to ensure all risks are assessed. Additionally the patient or carer should receive written information regarding possible complications, actions to follow and 24-hour emergency contact details.

Close monitoring by their CFSS is required to monitor clinical progress and biochemistry throughout the self-administered episode of care.

5.4.7 Hospital in the home

Hospital in the home services offer a potential alternative to inpatient hospital care for the management of less severe acute respiratory infections in individually selected adults and children with CF. A Cochrane review of home versus hospital intravenous antiobiotic therapy only for CF found limited evidence comparing HITH to inpatient care, with only one randomised controlled trial that showed there was no short term difference for clinical outcomes, adverse events or time to next admission. Patients treated at home reported less disruptions and overall costs, however they were more physically active and experienced greater levels of fatigue.[19, 20] Therefore a careful clinical judgement needs to be made as to whether a CF exacerbation should be treated as an inpatient or within HITH.

While home based therapy provided via HITH may be considered desirable and less disruptive for some patients, it is critical that the use of HITH services for adults and children with CF is planned, directly supervised and closely monitored by the CFSS. This is essential to ensure that all care provided is equivalent to inpatient care, that acute treatment outcomes are met and no long term deterioration in health status.

Potential advantages of care delivered by a HITH service are:

- reduced risk of cross infection
- less disruption to the patient and their family
- less treatment delay – as may happen when waiting for a non-urgent hospital bed
- money savings for the patient and family.

In 2012, a small number of HITH services in NSW had provided episodes of care for the management of acute CF respiratory infections. The successful use of a HITH service for acute CF care has been dependent on both the HITH service and CFSS agreeing to work in close partnership to establish appropriate clinical governance, formal shared care arrangements and ensure appropropriate clinical competencies.

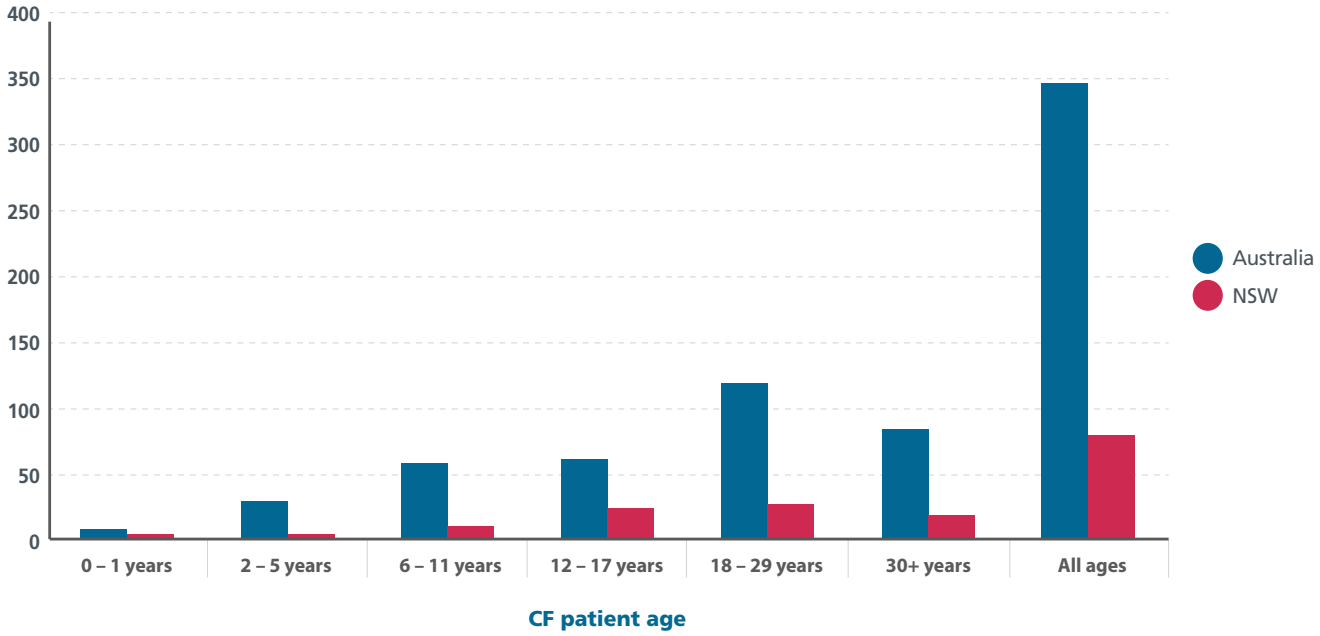


Figure 11. Episodes of hospital in the home by age group
Source: *Australia Cystic Fibrosis Data Registry Report*, 2011

NSW has the largest proportion (31%) of the Australian CF population. A comparison between NSW and other states in Australia for episodes of home based therapy reported to the ACFDR in 2011 showed that the relative use of HITH services in NSW was low across all age groups.

The number of episodes of care provided by HITH services to people with CF in NSW has increased in recent years. In 2012-13 there were 179 CF episodes of care with 3164 bed days that were able to be provided to patients with CF in their home (unpublished NSW Ministry of Health data). This equates to eight hospital beds (CF patients are usually managed in specialist respiratory or paediatric wards) across NSW that were available for other patients with more severe exacerbations or complications in that year.

A planned approach is needed to further develop the clinical capability and capacity within a small number of targeted HITH services to manage acute CF episodes in close association with designated CFSS. The CFSS and HITH teams should develop a close collaborative working relationship to support CF patients receiving care within HITH programs.

The CF working group identified a set of HITH capability requirements for potential referral for management of acute CF respiratory exacerbations which includes:

- a sufficiently large CF population within their service boundary to ensure a minimum caseload per year (recommended minimum 20 CF episodes of care per year)
- HITH staff with extensive experience in acute respiratory management with a regular case load of non-CF respiratory episodes of >30 per year
- access to medical review seven days each week
- access to urgent pathology and review of results seven days each week
- twice daily medication dosing seven days per week
- availability of an appropriate multidisciplinary team (specifically medical, nursing, physiotherapy and dietitian) with CF specific knowledge and skills (some components may be able to be provided through the relevant CFSS)
- medical, nursing and physiotherapy interventions that are available seven days per week to:
    - manage IVAD
    - manage port-a-cath
    - cannulate
    - perform spirometry
    - oversee sputum clearance techniques
- access to the required medications, nuitrional supplements and equipment
- processes that allow the CFSS to assume direct medical governance for HITH episodes of care
- processes for both the CFSS and HITH service staff to regularly meet to review patient outcomes and processes of care.

CF MANAGEMENT WITHIN HITH SERVICES

Hospital in the home services providing care to patients with CF should be formally networked with a CFSS or designated CF satellite facility and undertake regular review of their clinical practices and processes to ensure that an equivalent standard of care is provided and patient outcomes are maintained or improved. Hospital in the home service clinicians require ongoing access to individual clinicians within the CFSS team for advice, mentoring and further development of CF specific knowledge and skills.

Children and adults, identified by CFSS as clinically appropriate, are referred by CFSS to HITH services for the management of an uncomplicated respiratory infection. This may be as a complete substitution for inpatient hospital care in the case of a planned intensive therapy admission, or as an early discharge option following unplanned admission for an acute respiratory exacerbation. During the episode of care, the CFSS will directly supervise care through a formal shared care arrangement with the HITH service.

The selection of patients eligible for HITH should be made on an individual basis and in consultation with the patient and their CFSS. Patients who are considered not suitable for HITH include those:

- that feel unable to manage with intermittent daily care and support
- with recent or current instability that requires more regular monitoring
- with a history of poor adherence to therapy or response to treatment
- in unstable home situations
- with poor venous access
- residing in a remote area or who have anticipated difficulty returning for review
- who require a level of allied health intervention that cannot be provided through the HITH service (for example, multiple physiotherapist led interventions per day, psychosocial intervention or nutritional support).

IV ACCESS

Patients receiving care in the home will require stable venous access via a peripherally inserted central catheter (PICC) line, mid-line or implanted IVAD. The first dose of antibiotics should be administered in hospital (ED, CF clinic or inpatient setting).

CF patients should receive HITH care daily and be provided with a 24-hour point of contact for any unforeseen complications or emergencies.

HITH CLINICAL MANAGEMENT PLAN

As part of the referral process, a detailed CF summary and management plan should be developed and provided to the HITH service and the patient or their carer which includes:

- diagnosis and comorbidities
- baseline investigations required including sputum culture
- IV access requirements
- antibiotic prescription and duration
- monitoring requirements including lung function, drug levels and blood glucose levels
- physiotherapy airway clearance plan
- nutritional support plan including nasogastric or gastrostomy feeding.

The CFSS will continue to direct care during a HITH admission and determine the appropriate time for referral and planned discharge from the HITH service.

Patients will need to be regularly reviewed by their CFSS after discharge from a HITH episode of care via telehealth or at a planned clinic review.

More than 40% of CF patients or parents surveyed indicated that they would like more options for care to be provided by HITH service.

> We have had no experience with care at home but we and others have been asking about this option. Tune up admissions really disrupts school work – so it would be great to be able to get care before and after school.
>
> **Mother of an 11-year-old**

> I have had HITH for a tune up for the first time recently – they were great except they cannot always confirm the time they are coming especially for the first visit. They knew what they were doing and did it really well especially the CNS nurse. They were really good at taking blood too which is important for me as my veins are not good. It was so good for me to be able to stay at home.
>
> **Young adult**

Section 6

Cystic fibrosis life stage care needs

People with CF have specific care needs at key points in their disease trajectory and at specific life stages.

These specific care needs are detailed in the following section.

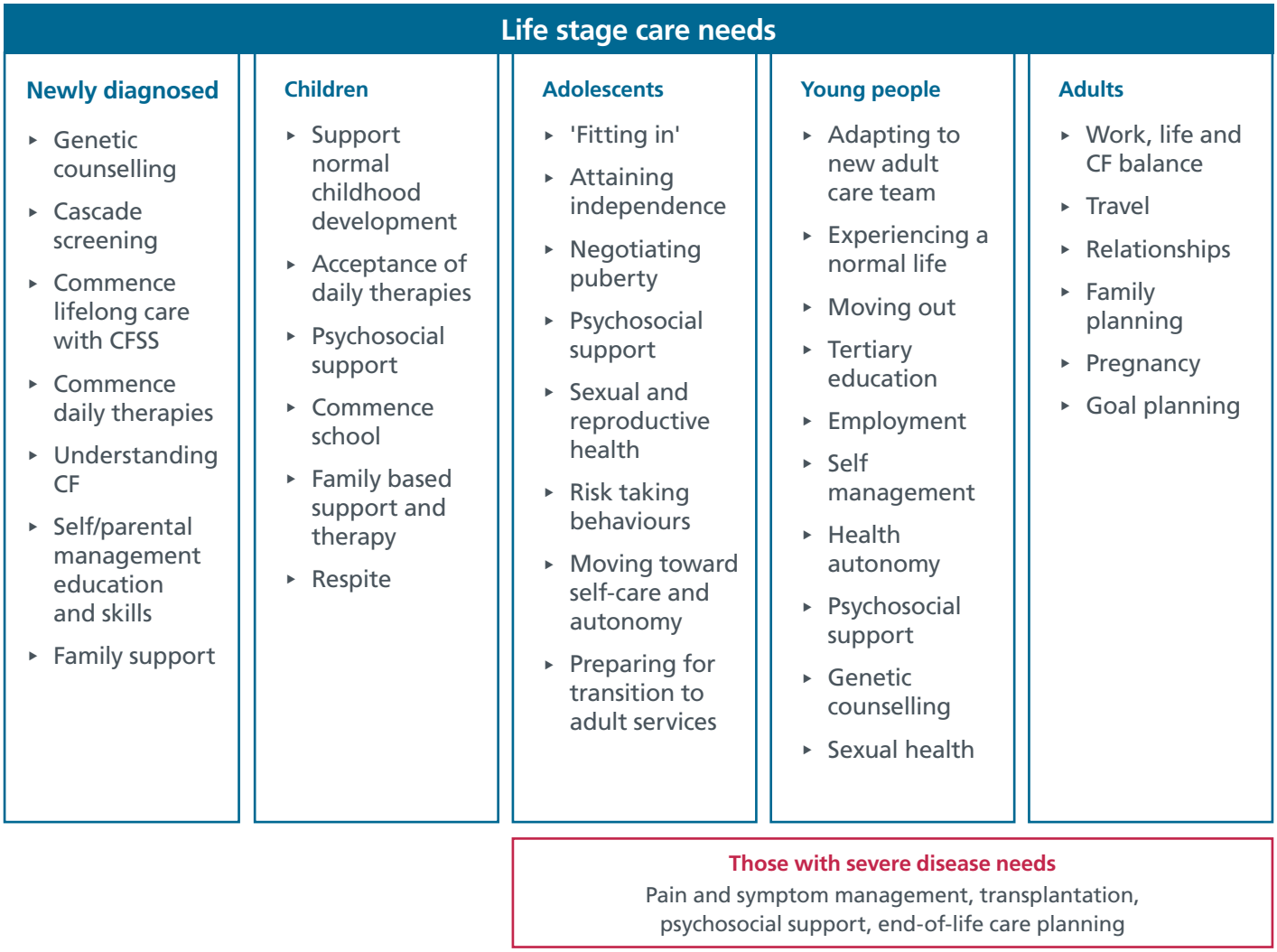


Figure 12. Life stage care needs

6.1 People newly diagnosed with CF

In Australia, the majority of new cases (> 80%) have the CF diagnosis made within the first months of life following identification through the newborn screening program. All (> 99%) babies born in NSW are offered newborn screening for CF as part of the routine Guthrie heel prick test which is generally conducted at 48-72 hours after birth. In a small number of people, a late diagnosis of CF is made following an acute clinical presentation or a CF diagnosis in a sibling.

In 2012, there were 96 babies born in NSW or ACT who were identified with cystic fibrosis transmembrane conductance regulator (CFTR) mutation of which 18 had a positive sweat test and diagnosis of CF. In the ACFDR Report 2013, there were 35 people newly diagnosed with CF in NSW, of which 26 were under 1 year, five were aged 1-4 years and four were aged 20 years or older.

Routine neonatal screening detects those infants with a raised level of immunoreactive trypsin (IRT) followed by genetic testing for three common CF mutations F508, G542X and G551D. Babies identified as positive through newborn screening gene testing may have either a single (heterozygous) or both genes (homozygous) affected. All babies with an identified CFTR genetic mutation and elevated IRT are notified to their neonatologist/obstetrician and referred on to a CFSS for a sweat test to confirm diagnosis.

There are a small number of cases where the diagnosis remains in doubt due to a discrepancy between genetic and sweat test results. These cases are classified as 'atypical CF' and these patients will require referral to CFSS for review of clinical signs and symptoms on an annual basis. Parents are highly anxious during the 'waiting for diagnosis' period and additional individually tailored support should be provided by a designated healthcare provider.

6.1.1 Genetic counselling

All adults and parents of infants and children diagnosed with CF or confirmed genetic carriers should be offered genetic counselling. Genetic counselling is essential for them to understand what their CF genetic mutation means and their options for future pregnancies including prenatal diagnosis or consideration of in-vitro fertilisation (IVF).

6.1.2 Cascade genetic testing

In addition, all adults or the parents of babies or children newly diagnosed with CF should be encouraged to inform their close relatives that there is a risk they may be a CF genetic carrier and the need to seek genetic counselling and genetic testing.

Table 3: Risk of being a CF genetic carrier by relation to person with CF

Relationship to person with CF	Risk of being CF genetic carrier	
Parent (biological)		100%
Brother or sister (not with CF)	2 in 3	66%
Half-brother or half-sister	1 in 2	50%
Uncle or aunt	1 in 2	50%
Cousin	1 in 4	25%

6.1.3 Initial care of newly diagnosed patients

Newly diagnosed people and parents of children with CF should be referred with minimal delay to an adult or paediatric CFSS as appropriate. The choice of CFSS should be made in conjunction with the patient or parents and include factors such as travel, parental accommodation and existing outreach or satellite services.

As part of initial assessment, patients or parents meet with all members of the CFSS team and receive intensive one-on-one education and self-management support to enable them to become confident with daily preventive therapies including:

- pancreatic enzyme replacement therapy
- vitamin supplements
- nutrition
- airway clearance
- antibiotic regimens.

The patient or parent is provided with phone or email contact details for the individual CFSS clinician/s and is encouraged to seek advice whenever needed. In addition, CFNSW contact and services information should be provided to all newly diagnosed patients and their family to ensure they have access to additional community services information and support.

Specific advice needs to be provided to patients and family concerning the variable accuracy and value of online information related to CF and links provided to verified useful sites.

After initial comprehensive assessment and education, newly diagnosed patients and families require an individually tailored period of frequent and regular review which is focused on CF management education, specific self-management skills and psychological support.

6.2 Care of children with CF

Care during childhood is focused on supporting the families to understand their child's manifestations of CF and to provide the daily therapies and regularly screening to prevent respiratory infections and CF related comorbidities. Tailored family based support may be required to ensure that children with CF experience normal development opportunities, achieve developmental milestones and are well integrated into the pre-school and school environment. In some instances, families may need assistance to access childcare or respite services which have an appropriate level of CF specific knowledge and skills.

6.3 Care of young people with CF

With adolescence and young adulthood, comes a significant time of change and development. During this life stage young people are acquiring skills for future independence including the ability to manage their healthcare needs.

Young people with CF face significant additional challenges related to increasing symptom burden, high daily treatment burden and frequent disruptions to their lives related to recurrent health appointments and hospitalisations. In addition, young people with chronic conditions are more likely to experience anxiety and depression and be involved in risky behaviours.[21-23]

> "I do not feel I am part of health and treatment decisions. I don't think they understand how little spare time I have between school, working in the holidays, doing a course at night, going to the gym. So it means I often miss nebulisers and physio sessions."
>
> **Adolescent**

Studies show that non-adherence with recommended therapy during this period is higher particularly in adolescent girls.[24] Depression and anxiety in people with CF are associated with poorer lung function and BMI.[23] It is during this period that young people currently experience a significant and irreversible decline in their lung function (ACFDR) and have the greatest utilisation of hospital based care.[6]

The deterioration in health status and increased utilisation of hospital based care that occurs in young people is considered attributable to:

- disease progression
- impact of hormonal changes in puberty
- unmet psychosocial and healthcare needs
- variable adherence to therapies and healthy lifestyles
- onset of risky behaviours.

YOUTH FRIENDLY SERVICES

Given the particular vulnerabilities of this age group outlined above, clinicians caring for young people with CF need to develop specific skills around engaging with young people and understanding their specific needs. The overall aim of care at this stage is to:

- avoid preventable deterioration
- identify and address their health, sexual and reproductive health needs
- identify and address psychosocial needs
- address and encourage autonomy and self-management skills
- build resilience and protective behaviours in the young person.

One of the strategies that can help clinicians to effectively engage with young people is to offer a variety of ways in which they can initiate contact, regularly communicate and receive support at their specific times of need from members of the CF healthcare team.[25-28]

ASSESS AND MANAGE YOUNG PEOPLE AT RISK

Due to the increased risks within this age group, regular assessment and referral pathways are specifically recommended for:

- anxiety and depression with referral for individual and family based therapy [22, 27]
- individual knowledge about CF and perceptions related to the effectiveness of CF medications and treatments
- monitoring during puberty (especially in girls) with referral for 'body image' related counselling and support as appropriate.

It is important to engage with young people identified at risk to find age appropriate case management and/or peer led support programs that best meet their needs. In NSW, young people with CF can be referred for additional health coaching and self-management support services provided by:

- Chronic Illness Peer Support (ChIPs)
- CFNSW counsellors/peer support program
- ACI transition coordinators
- The Sydney Children's Hospital's Network Trapeze service
- NSW Chronic Disease Management Program
- Adolescent medicine departments.

6.4 Transition from 'kids' to adult CFSS

With significantly increased survival, the majority of children with CF will transition from a paediatric CFSS to an adult CFSS. The transition process is a major milestone and is a potentially stressful time for both young people and their families.

The following strategies improve effectiveness of the transition process.

- A systematic and formal process exists that is underpinned by formal guidelines and policies.
- Transition is a process not an event. Education on transition and empowerment around self-management commences early – around age 14 years.
- There is a designated member of the healthcare team who partners with both the young person and their family to prepare for and facilitate the process. A relationship should be established or strengthened with their local primary care provider who is usually the GP.
- Communication processes and tools support person centred care – openness, transparency and willingness to work together underpins all good communication. All clinicians should have open discussion with young people regarding common concerns of young people such as growth and development, sexuality and reproduction, mood and mental health problems, substance use and health promoting versus damaging behaviours. [29]
- All young people should have an individualised transition plan which focuses on all aspects of their lives. The plan also covers details of clinical care, a record of needs assessment, goals and should include interagency agreements.
- The young person and their family need to be involved in the decisions regarding the timing of transition and in selecting an adult CFSS. Responsibility for decision-making should be increased gradually, and adolescent friendly transition services should be put in place. Where the young person cannot take full responsibility for self-management, it is particularly important to involve the family/carer.
- At least one pre-transition joint clinical review occurs involving the young person and clinicians from both the paediatric and adult CF teams. Follow-up may be required for several years to ensure that young people have engaged effectively with adult health services and evaluation of the process must be undertaken to inform future planning and policy.

Transition principles are available in the document, ACI Key Principles for transition of young people from paediatric to adult healthcare.

6.5 Care of adults with CF

Balancing a normal life with managing their CF can be an ongoing and increasing challenge for adults. New CF related complications are often emerging as well as progression of their disease which over time may require additional therapies and increased burden of care. At the same time, adults with CF are undertaking tertiary university education, gaining employment, establishing a career, forming and maintaining relationships, cohabiting, having children and planning for the future.

Adults with CF may require individually tailored additional clinical review and psychosocial support for multiple care needs including:

- maintaining lung function in the presence of acute and chronic respiratory infections
- maintaining optimal body weight with their increased energy demands and frequency of acute respiratory infections
- managing CF related diabetes throughout variations in their health status
- maintaining optimal bone density and muscle strength
- pain management
- dealing with diseases associated with ageing
- dealing with fertility issues (98% of males are infertile)
- contraception
- genetic counselling
- preparing for pregnancy
- dealing with prenatal/pre-implantation diagnosis
- parenting with a progressive life limiting condition
- drug and alcohol issues
- emotional and social issues
- outliving their parents who have been their lifelong carers and/or support people.

6.6 Care of pregnant women with CF

Pregnancy will have a likely impact on the health status of women with CF and poses a higher risk for the foetus. The level of risk is increased in those women with an FEV1 < 50% predicted, and those with CF related diabetes and chronic infections such as burkholderia cepacia.

All women considering becoming pregnant or who are pregnant will require opportunities for discussion with a genetic counsellor. Woman may have CF associated fertility issues and referral to specialist fertility services for advice, assessment and treatment may also be required. Pregnant women will require close monitoring by a CFSS obstetric service which is able to provide appropriate care in the situation of a high-risk pregnancy.

In women who are pregnant, shared care should be provided for the duration of the pregnancy and postnatal period by their CFSS and a specialist obstetrics service and within the context of a high-risk obstetric team. The nutritional and respiratory status of the woman are commonly adversely affected during pregnancy and regular review and input from both the CF dietitian and physiotherapist is necessary.

Women with CF are at high-risk of developing CF related diabetes, and those with diabetes will need enhanced diabetic control during their pregnancy and access to, as well as close monitoring by, a diabetic team experienced in CF and obstetric care.

Close monitoring in the last trimester and during the postnatal period is essential to ensure the woman achieves her pre-pregnant health status.

6.7 Care of those with severe disease

Despite significantly increased longevity, people with CF will experience declining health status and an increasing burden of disease as they age. The major factors associated with deterioration in health status are age, chronic lung infections, bacterial colonisation and presence of associated CF comorbidities. Severity of disease is based on lung function, and those who reach the severe stage of disease are identified as having <30% of their predicted lung function.

The CFSS will maintain the overarching clinical responsibility for the care of those with severe disease, with access to additional clinical care and support for the patient and family provided via a linked network of local service providers including GP, palliative care service and clinicians.

The needs of those with severe disease and their families are complex. Individuals with severe disease frequently experience an accelerated decline in health status and increasingly distressing symptoms including pain (commonly headache, neck and abdominal), breathlessness, cough, lack of sleep and depression. The majority of those with severe lung disease will be referred for transplant assessment and over half will be accepted onto the wait list for bilateral lung transplantation. Therefore for the majority of those with severe disease, there is a need for both active CF therapies to prevent further deterioration and holistic supportive care to be provided in parallel.

Additional support by a designated case manager is often required for the individual and family during this challenging phase to support them through increasing life threatening health crises and to resolve the multiple conflicting demands and expectations.

6.7.1 Goal planning

People with CF who are approaching or have reached the severe stage of their disease require honest and open communication with all members of the CFSS healthcare team and other specialist service providers. This is vital for the person to make informed decisions and undertake effective goal planning over the remainder of their life.

> “Honesty and openness was what I had been trying to achieve from an early age, yet was constantly met with denial from those around me who I felt needed to be aware of how I felt, my fears and my thoughts and my wishes… my questions…anything.”
>
> **Young adult**

A designated member of the team, who has an established rapport with the individual patient and their family, is nominated to undertake a series of conversations to understand their holistic care needs and to assist them to identify an enduring guardian and to develop an advance care directive (ACD) which reflects their wishes. The timing of goal planning discussions should occur early and before their life expectancy is considered to be less than 2 years.[30,31]

The goal planning discussions need to occur during periods when the person is relatively well and precede recurring life threatening health crises which commonly occur in the severe disease stage.

> “I remember being so exhausted and run down that speaking about anything that involved thought was just way too much by the time I hit that point.”
>
> **Young adult**

6.7.2 Transplantation

It is during the severe disease stage that individuals and families are faced with many challenging decisions including assessment for transplant. All patients with CF who reach the severe disease stage should be considered for transplantation. The CFSS team will determine the clinically appropriate timing to commence formal discussions with the patient and family regarding referral for transplantation.

Early discussions around the place for transplantation in CF care are often helpful to normalise this treatment in the context of managing end stage lung disease in CF. The CFSS should ensure that patients and their families have sufficient information to make these challenging decisions. Written CF specific transplant information and decision support tools should be available for patients and families which include:

- the range of symptoms and feelings they may experience from this stage onward
- additional support and services available for them
- personal stories from other people with CF
- options, benefits and risks for transplantation (CF Australia decision aid)
- preparation for decision-making in relation to transplantation
- goal planning.

Therefore, once a patient has made the decision to be assessed for transplant, intensive CF clinical management and patient-centred supportive care need to be provided in parallel.

It is important that discussions about specific aspects of end-of-life care planning occur well before the patient deteriorates (often rapidly) to the point where they require ventilatory support, as they may be increasingly unable to communicate their wishes and participate in decision-making.

PRE TRANSPLANT CARE

The CFSS is responsible for the care of CF patients throughout the period leading up to transplant which may extend over months or years and should be conducted in close consultation with the respective transplant service.

A comprehensive pre-transplant work up involving a suite of investigations and monitoring is required for the initial transplant assessment and to meet transplant eligibility criteria. During the pre-transplant work up, patients will have frequent medical appointments and travel requirements which can be both physically and financially difficult. Patients who live a considerable distance from the CFSS and the transplant service may even consider re-location to be able to readily access services during this period.

Once a patient has been accepted for transplant, the CFSS will provide close monitoring, aggressive CF management, care coordination across multiple service providers and psychosocial support for the patient and family. To achieve an effective and timely work-up a clinician from CFSS needs to be designated as the transplant co-coordinator, whose role is to liaise closely with the co-coordinator at the transplant referral centre. Close consultation with the transplantation team during this time is essential.

Patients who are deteriorating rapidly may be transferred to the transplant team prior to completion of pre-transplant work up if they require invasive ventilation or ECMO as a bridge to transplantation or to enable adequate time for the patient to develop a trusting relationship with the transplant team.

POST TRANSPLANT CARE

The post-transplant care unit is primarily responsible for the care of CF patients post-transplant. Optimal long-term patient outcomes post-transplant require ongoing clinical collaboration between the highly specialised transplant team and their specialist CFSS team to both preserve transplanted organs and manage systemic CF related comorbidities.

Effective communication and clinical information sharing processes need to be established to achieve effective clinical relationships across the two specialist teams required for the lifelong care of the post-transplant CF patients. Effective post-transplant clinical relationships across the teams will support the establishment of clearly defined clinical roles and responsibilities including planned scheduling of appointments to reduce duplication of investigations and/or gaps in ongoing monitoring of the systemic manifestations of CF. Effective clinical relationships and communication processes between the transplant and CFSS teams are essential to minimise the number of medical appointments and travel burden which has been identified as a significant issue by some post-transplant patients.

A more effective way of electronically sharing pathology results and medical imaging is urgently needed to reduce unnecessary or duplicate investigations, minimise patient travel and reduce the risk of errors that may occur as a result of missed investigation results when patients are managed by both transplant and CFSS teams.

PAIN AND SYMPTOM MANAGEMENT

All people with severe stage disease, including those on a wait list for transplant, should have access to ongoing individually tailored supportive care. The CFSS is responsible for determining the level of referral (consult or ongoing shared care with CFSS) and the timing of referral for each individual.

The care needs of people with severe CF are complex and the initial referral should be made to a palliative care service, generally located at the CFSS facility, which has extensive experience and expertise in managing CF related symptoms and life threatening crises. Following an initial assessment, shared care arrangements may need to be established with a local palliative care service which can arrange additional care and support services to be provided for the patient closer to, or within their home. A process for regular communication and ongoing mentoring between the local and CF specialist palliative care services is recommended.

The role of palliative care services in an ongoing shared model may include pain and symptom management, spiritual and psychosocial support, care coordination and referral to local health and community providers including home care, discharge planning and providing care in the home.

Patients who have received additional supportive care from palliative care services have indicated that they would have benefitted even more with an earlier referral.

PSYCHOSOCIAL SUPPORT

People with CF and their carers are at heightened risk of anxiety, depression and hopelessness.[32] Depression has been shown to lead to lower adherence to therapy in people with CF, which can subsequently impact on the rate of deterioration and ultimately their chance of receiving a transplant. Therefore, regular assessment for anxiety and depression should occur for patients and carers at this stage with referral to a psychologist or psychiatrist as appropriate. All people with severe disease should have access to a social worker to assist them to deal with the progressive loss of independence, changing financial needs, Centrelink entitlements and to establish ongoing support mechanisms for the patient, their carer and family.

CARER AND FAMILY SUPPORT

It is during this phase that people with CF may experience recurrent life threatening health crises. Information should be provided for the carer/family about potential health crises that may occur at home, the action they can take and who to contact both within and outside normal working hours. Carers also need to have clear instructions for who they will need to call in the event that their loved one dies at home. Patients waiting for transplant reported that the time they and their carers most need support is after 'false alarms' occur.

END-OF-LIFE CARE PLANNING

A proportion of people with CF will not receive a transplant or their transplanted organ/s may fail. As the health status of a person with severe stage disease continues to deteriorate and they approach end-of-life and death, specific medical treatments and the locations of care need to be discussed, initially with the patient, and subsequently involving their nominated support person/s. A series of conversations led by a member of the CFSS team, CFNSW or GP who has long-term rapport with the patient and family can assist them to identify their wishes specifically for the level of resuscitation, admission to ICU, intubation and ventilation and preferred place of death.

The patient's ACD should be reviewed and revised in line with their wishes and the ACD needs to be re-visited on a regular basis as each patient's wishes for specific treatments may change as their health status declines.[33-35]

BEREAVEMENT

Following the death of a patient with CF, members of the CFSS should have an opportunity to meet with the family and to provide input and assistance for the provision of ongoing bereavement support as required by each individual family.

Section 7

Cystic fibrosis model of care – next steps

The *Cystic fibrosis model of care* aims to better meet the healthcare needs of people with CF across the life span. The model of care provides direction for how and where effective patient-centred services can be provided for people with CF across NSW.

Implementation of the *Cystic fibrosis model of care* will involve a collaborative process led by clinicians and managers within relevant LHDs and SHNs. Implementation of statewide solutions will be supported by NSW Ministry of Health and pillars and with ongoing guidance from a CF implementation advisory group.

Key components required to support implementation will include the following.

7.1 Activity based funding assessment

ACI in collaboration with the Activity Based Funding (ABF) Taskforce, LHDs and SHNs, will undertake an ABF assessment of activity and associated ABF resource streams for episodes of CF care (inpatient and outpatients) provided in adult and paediatric CFSS, CF satellite facilities, outpatient clinics and via telehealth. A methodology will be designed to assess if the ABF resource stream is attracting service delivery in line with evidence based care.

7.2 Needs assessment

Cystic fibrosis specialist services will be supported by respective LHDs and SHNs to undertake an independent needs assessment against the *Cystic fibrosis model of care* to identify the elements required for implementation. The CF implementation advisory group will review needs assessments, establish statewide priorities for implementation and provide recommendations to LHDs and SHNs by January 2017.

7.3 Implementation plan

ACI and the CF implementation advisory group will develop a detailed implementation plan which will set out clear roles and responsibilities for LHDs and SHNs, aims and objectives and the support that ACI will provide to local teams for implementation. The implementation plan will provide flexibility for local teams to plan and change the way they deliver services based on local needs, priorities and local resources. The plan will outline Ministry of Health responsibilities for ongoing monitoring of performance and key performance indicators (KPIs).

7.4 Development of an evaluation framework

An evaluation framework has been developed to outline the key activities, outputs and outcomes related to implementation of the *Cystic fibrosis model of care*. Specific KPIs will be developed and systems established to monitor progress including short, medium and long-term outcomes within relevant time frames.

7.5 Strategic statewide governance

The *Cystic fibrosis model of care* will be used to shape discussions with the Ministry of Health and to further explore how statewide planning and performance monitoring can be developed strategically to ensure equitable access to care and outcomes for people with CF across NSW. This will allow planning of services in line with the projected needs for the CF population into the future. A framework needs to be developed in partnership to ensure there is an ongoing coordinated approach to CF service planning across NSW and to manage complex aspects of service delivery and sharing of health information within and between LHDs and SHNs.

7.6 Develop a communication plan

A comprehensive communication plan will guide strategies to raise awareness of the healthcare needs of people with CF and deliver key messages related to implementation across NSW. Communication will need to be targeted depending on solutions, audience and stakeholders. The plan will be distributed to all stakeholders, and will clearly define the key service components and service providers who are working in partnership to implement changes to service delivery.

7.7 Generate LHD and SHN executive sponsorship

Sponsorship will be sought from relevant LHDs and SHNs. The level of input required will vary depending on patient population and their role as CFSS or CF satellite facility. Local health districts will be asked to identify primary contacts within their organisations and establish a local implementation group.

7.8 Support LHDs and SHNs to monitor patient outcomes

Maintaining or improving patient outcomes is a key principle within the *Cystic fibrosis model of care*. The ACI, LHDs and SHNs will collaborate to establish a process for monitoring patient outcomes to be sourced from data collected at the service level and submitted to the ACFDR. The ACI will support LHDs and SHNs to share outcomes, learnings and improvement strategies to accelerate implementation across the NSW health system.

The ACI will undertake an evaluation of the *Cystic fibrosis model of care* at appropriate points in line with the formalised evaluation plan.

7.9 Possible enablers

Enablers identified to support implementation of the *Cystic fibrosis model of care* were:

- CF knowledge and skills workforce development
- formal shared care and outreach service agreements across sites and LHDs
- clinical responsibilities and governance arrangements across services providers
- ABF funding stream
- clinical policies and procedures
- information and communication technology to support information sharing
- telehealth capability and utilisation
- evaluation and monitoring of patient outcomes
- development of NSW service delivery models in collaboration with ACI to standardise procedures and policies and enhance the development of services especially in the areas of support to HITH, telehealth and transition to adult care.

This model of care aligns with the NSW Government strategies:

- *NSW State health plan: Towards 2021* [36]
- *The NSW Government plan to increase access to palliative care 2012-2016* [37]
- *Supporting Australians to live well at the end of life: National palliative care strategy 2010* [38]
- *NSW Integrated care strategy 2014-2017*.[39]

Appendix 1

Recommended staffing levels at adult CF centres in terms of full-time equivalent positions providing CF care

Table A1: Recommended staffing levels at adult CF centres in terms of full-time equivalent positions providing CF care

Positions	50-75 patients	75-100 patients	> 150 patients
Consultant 1	0.5	1	1
Consultant 2	0.3	0.5	1
Consultant 3	–	–	0.5
Clinical fellow/Advanced trainee	0.5	1	1
Registrar/RMO	0.4	0.8	1
Specialist nurse	2	3	6
Physiotherapist	2	4	6
Dietitian	0.5	1	2
Social worker	0.75	1	2
Psychologist	0.4	1	1.5
Secretary	0.5	1	2
Data clerk	0.4	0.8	1
Pharmacist	0.5	1	1

Source: The Thoracic Society of Australia and New Zealand. *Cystic Fibrosis* 2010.[44]

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