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# Australasian Clinical Practice Guidelines for Social Work in Cystic Fibrosis

## 2017

Australian Cystic Fibrosis Social Work Interest Group

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# Executive summary

The Australasian Clinical Practice Guidelines for Social Work in Cystic Fibrosis project was proposed in 2013 by the Australian Cystic Fibrosis Social Work Interest Group and supported by the Australian Association of Social Workers (AASW) Lyra Taylor Grant and Cystic Fibrosis Community Care with the provision of funding in 2017. Senior cystic fibrosis social worker Anne Li and cystic fibrosis social workers Tamar Peleg and Stephanie Chen constituted the initial steering committee and writing group for this project. This document represents the first set of comprehensive clinical guidelines for social work in cystic fibrosis (CF) in Australia and New Zealand.

The project scope included an analysis of the current literature on the psychosocial needs of CF patients and best practice social work in this field. The expert reference group was comprised of social workers practising in CF care in Australia and New Zealand. Consultation was undertaken with the reference group to ensure professional consensus. This document was additionally reviewed by members of the multi-disciplinary team, representatives from CF advisory groups

from Australia and New Zealand and people with CF and their caregivers.

The specific areas of CF social work practice covered by this document include:

1. Diagnosis
2. Infancy and early childhood
3. Childhood
4. Adolescence
5. Transfer of care
6. Adulthood
7. Transplant
8. End of life care

Each chapter provides recommendations for social work practice at development stages across the lifespan. These recommendations endeavour to accommodate the needs of each individual patient and the practices and systems of each CF care centre. The recommendations are based on current evidence and expert consensus. It is the intention of this document to provide a clear guide to best practice for social work in CF care.

# Foreword

## Purpose and scope

The writing of these standards commenced in 2017, with the establishment of the steering committee and reference group. A need was identified by the Australian Cystic Fibrosis Social Work Interest Group for comprehensive clinical practice guidelines in CF social work. The purpose of which is to provide clear standards of best practice for social workers in CF care in Australia and New Zealand and align social work with the other allied health disciplines (physiotherapy and dietetics) in the multidisciplinary CF care team.

These guidelines cover the psychosocial needs of CF patients across the lifespan and provide recommendations for social work practice based on review of the current literature and consensus of expert opinion. These guidelines were written for the specific health care delivery systems, demographics and social care systems in Australia and New Zealand. The expert reference group included representatives from both Australia and New Zealand.

While these guidelines are structured in a linear format along the lifespan, individual development will differ and these stages should not be interpreted as discrete but as existing along a spectrum. Patients will progress through these stages in a unique way and time. Social/cultural indicators and transitions in CF care have been used to distinguish each life stage.

These guidelines should be read in conjunction with the Cystic Fibrosis Standards of Care, Australia (2008) and the Standards of Care for Cystic Fibrosis in New Zealand (2010).

## The membership of the Steering Committee included:

**Stephanie Chen**- BA, MSW, Paediatric and Adult Cystic Fibrosis Social Worker, Monash Health.

**Emily Armstrong**- B.HSc, B.SW, M. Public Policy and Management, Research Assistant, Monash Health.

**Associate Professor Fiona McDermott**- BA dip Soc Studs., M. Urban Planning, PhD, Monash University/ Monash Health.

**Marie Emmeline Bosquet**- BA, B.SW (candidate), Student Research Assistant, Monash Health.

## The membership of the expert reference included:

**Nicole Anderson**- Social Worker, Regional Cystic Fibrosis Liaison/ Representative Victoria, Central Gippsland Health.

**Kate Leahy**- Adult Cystic Fibrosis Social Worker, Royal Adelaide Hospital, Adelaide.

**Courtney Cronin**- Community Health Worker, Cystic Fibrosis Queensland.

**Julia Langrehr**- Executive Officer, Cystic Fibrosis South Australia.

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Special thanks to the membership of the stakeholder advisory group:

**Scott Bell**  
**Nettie Burke**  
**Charlotte Kenny**  
**Lucy Holland**  
**Sherril McCleary**  
**Holly Murphy**  
**Jodie Williams**

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No potential conflicts of interest were identified by the membership of the steering committee or reference group.

## Glossary of terms

AASW	Australian Association of Social Workers
ANZASW	Aotearoa New Zealand Association for Social Workers
ANZCOTR	Australia and New Zealand Cardiothoracic Organ Transplant Registry
CF	Cystic Fibrosis
CFTR	Cystic Fibrosis transmembrane conductance regulator protein
CFSCA	Cystic Fibrosis Standards of Care, Australia
HRQOL	Health Related Quality of Life
IVF	In Vitro Fertilisation
MDT	Multidisciplinary team
PTSD	Posttraumatic stress disorder
Patients	Different CF care teams/ organisations may use alternative terms e.g. service users, consumers or clients.
SRH	Sexual and reproductive health
SCCFNZ	Standards of Care for Cystic Fibrosis in New Zealand.

# Methods

## Search strategies and literature review

The literature search for these guidelines was conducted using Google Scholar, PubMed, CINAHL, PsycINFO and the Cochrane Library. Electronic searches were conducted by the research student and research assistant. Manual searches of the reference lists were conducted. Searches were limited to articles in English and literature up to and including 20/7/2017 was included. Literature published before 1/1/2010 was excluded. Due to the limited evidence in many areas of CF social work practice systematic reviews, meta-analyses and less robust research designs were included.

Each article was reviewed in detail by both the research student and research assistant and made available to the reference group. Published clinical guidelines associated with CF care were also reviewed and included where appropriate. A review template was developed to guide the search and analysis. Standards were then derived from analysis of the literature.

Where there was no appropriate evidence available for areas of practice, a consensus of the reference group (expert opinion) was provided. This is the case unless otherwise stated.

## Consultation

To ensure a consensus of expert opinion the reference group reviewed and edited this document at two stages throughout the development and conducted a final review on the 17/11/2017. The stakeholder advisory group, which included medical consultants, CF advocates, patients and parents/caregivers reviewed the guidelines on the 17/11/2017.

## Endorsement

These guidelines have been endorsed by the Aotearoa New Zealand Association of Social Workers, Cystic Fibrosis Australia, and Cystic Fibrosis New Zealand. Endorsement is being sought by the Australian Association of Social Workers.

# Introduction

## Disease overview

Cystic Fibrosis (CF) is a genetic disorder affecting roughly 70,000 people worldwide (Cutting 2015). People of European descent are most commonly affected by CF. The disease is caused by a mutation in the cystic fibrosis transmembrane conductance regulator protein (CFTR) gene. This protein controls the movement of salt (sodium chloride) in and out of cells in the body. When a mutation occurs in the CF gene, the function of the CFTR is compromised, resulting in abnormal movement of salt in or out of cells (Cutting 2015). There are more than 2,000 different identified mutations in the CF gene, the most common of which in Australia and New Zealand is called F508del.

CF affects multiple systems and organs in the body including:

**The lungs/ respiratory system-** people with CF commonly have difficulties with their lungs. They may have a persistent cough and wheezing or difficulty breathing.

**The pancreas/ digestive system-** CF affects enzyme production in the pancreas making it difficult to absorb food. Insulin deficiency may become an issue for CF patients over time and may lead to a diagnosis of CF related diabetes.

**Reproductive system-** Most males with CF are infertile, because of an absence or blockage in the sperm canal. Women with

CF may experience fertility issues primarily due to nutritional problems caused by CF. CF was previously a fatal paediatric illness. Although there is currently no cure for CF, advances in treatments continue to increase the median life expectancy which is currently around 40 years (Cutting 2015). There is no way to predict life expectancy at diagnosis.

Living with CF involves a high level of care and time-consuming treatments. Managing CF involves lifelong regular review and treatment at a specialist multidisciplinary CF centre in a major metropolitan hospital. A daily self-care regime of airway clearance, exercise, nebulised and oral medications, enzyme replacement and vitamin and nutritional supplements are required to maintain optimal health.

Families with a child that has CF are more likely to have a lower socioeconomic status and labour supply, increased residential mobility and increased rates of parental separation when compared to families without a child with CF. Maternal and child psychological stress and marital discord is significantly associated with increased CF related health-care use (Douglas et al. 2016).

## Services and facilities

In accordance with both the Australian and New Zealand national standards of care, the primary locus of care for people with CF should be in the nearest hospital with a CF clinic. Social work is part of the CF multidisciplinary team (MDT). The CF team support children and families who have CF from diagnosis through to the end of life.

The CF team should include:

- Centre Director
- Specialist / Consultant Physician
- Specialist Nurse / Centre Coordinator
- Specialist Physiotherapist
- Specialist Dietitian
- Social Worker
- Psychologist\*
- Clinical Pharmacist
- Clinical Microbiologist
- Administration Support Officer
- Audit, Research and Data Registry Coordinator

\*In some centres an experienced counsellor or an occupational therapist may undertake some functions of a psychologist

## Summary of the social work role

Due to the wide variation in genotypes and infection rates, the progression and severity of CF is highly variable. The social work role in the MDT offers a unique

understanding of the patient within his/her psychosocial context and the social determinants of health. Social work practice recognises the changing needs and capabilities of patients across the lifespan. Not only the potential risk factors but also each patients' individual capacity for resilience and strength.

The Cystic Fibrosis Standards of Care Australia describes the social work role:

*"...involves the provision of a range of practical and emotional support services to patients and their families. Regular patient review enables the identification of psychosocial issues such as emotional and relationship problems, financial stress, substance abuse problems, and educational and employment issues.*

*Specialised knowledge of key support services is required to enable advocacy on issues such as income support and concession card eligibility, reinstatement of cancelled income support payments, and access to travel and accommodation subsidies associated with travel to the CF centre. A key role is to provide emotional support to patients whose health is deteriorating and assist them to evaluate their short and long term options in terms of both the patient and their family reducing their working hours or stopping work and applying for income support and other benefits."* (Bell & Robinson 2008).

# Ethical practice

The social work role in CF must adhere to the core social work values of the Australian Association of Social Workers Code of Ethics and/or New Zealand Code of Conduct for Social Workers, including respect for persons, social justice and professional integrity. Social work practice must meet the objectives and standards of the Australian Association of Social Workers and/or Social Workers Registration Board of New Zealand.

These guidelines are intended to be read in conjunction with the following codes of ethics:

**ASSW Code of Ethics:**

<https://www.aasw.asn.au/document/item/1201>

**ANZASW Code of Ethics:**

<http://anzasw.nz/wp-content/uploads/Code-of-Ethics.pdf>

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Cutting, G 2015, 'Cystic fibrosis genetics: from molecular understanding to clinical application', *Nature Reviews: Genetics*, vol. 16, pp. 45-56, DOI: 10.1038/nrg3849

Douglas, T, Green, J, Park, J, Turkovic, L, Massie, J & Shields, L 2016, 'Psychological characteristics and predictors of health-care use in families with young children with cystic fibrosis in Western Australia', *Journal of Paediatrics and Child Health*, vol. 52, no. 1, pp.34-39, DOI: 10.1111/jpc.13011

Medical Advisory Committee of Cystic Fibrosis Association of New Zealand (MACCFANZ) 2010, *Standards of care for cystic fibrosis in New Zealand*, viewed 20 July 2017, <[https://www.thoracic.org.au/journal-publishing/command/download\\_file/id/18/filename/SOC\\_for\\_CF\\_in\\_NZ.pdf](https://www.thoracic.org.au/journal-publishing/command/download_file/id/18/filename/SOC_for_CF_in_NZ.pdf)>

# Social work practice across the lifespan

## Cystic fibrosis social work standards in context

Social workers may work with people with CF and their families in a range of contexts including community health settings and hospitals. The following guidelines are recommendations based on current literature and consensus of expert opinion. In some cases, recommended standards will not be applicable to all practice settings or organisations.

The make-up of CF care teams may also differ across settings and therefore some of the recommendations made here as part of the social work role may be fulfilled by other allied health workers (e.g. welfare workers, allied health support workers).

For social workers practicing in a hospital setting, practice may take place during an inpatient admission or outpatient clinic. During an inpatient admission patients and families may need additional emotional and logistical support depending on their circumstances.

# 1. Diagnosis

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Patients and/or parents/ caregivers who receive a diagnosis of CF should have prompt access to CF social worker and psychosocial support. An initial psychosocial assessment should be performed at this stage.
- Standard 2** Following diagnosis patients/parents/caregivers should receive assistance in managing practical and logistical challenges emerging from diagnosis and potential hospital admission.
- Standard 3** Parents/caregivers and families should be provided with appropriate referrals and educational materials based on individual needs assessment.
- Standard 4** Provide ongoing psychosocial / emotional support during this period based on individual needs assessment.

## 1.1 Overview

### New born screening and diagnosis

In Australia and New Zealand, all newborns are screened for CF 48-72 hours after birth by a heel prick blood test or Guthrie blood test (MACCFANZ 2010; Bell & Robinson 2008). Test results that return an elevated level of immunoreactive trypsin are tested for mutations in the CFTR gene. Newborns who are found with severe homozygous or compound heterozygotes gene mutation are assumed to have CF and referred to a CF centre or team. These newborns then undergo a sweat test for diagnosis.

The sweat test measures the level of chloride in the sweat and is considered the 'gold standard' in testing for cystic fibrosis, and this will provide a conclusive diagnosis.

### Late diagnosis

While most CF patients are diagnosed by neonatal screening, 10% of patients are diagnosed in adolescence or adulthood. These patients may have been born overseas, have been missed on newborn screening or born prior to the introduction of newborn screening. While patients may

feel relieved receiving a firm diagnosis, after a long process of medical reviews, some react with shock, anger and denial. Patients diagnosed in adolescence or adulthood are sometimes reluctant to adhere to the treatment requirements seeking curative treatment rather than ameliorative treatments. Issues for treatment can also arise in patients being reluctant to visit a care centre for fear of infection, preferring to see a previous physician or not wanting to be labelled as having CF or 'sick'.

Whether diagnosis occurs at newborn screening or in later life a diagnosis of CF can be distressing for patients, parents/ caregivers and other family members. There are genetic implications for both the patient and their family and significant impacts for the patient and their family's future. As outlined in the *Cystic Fibrosis Standards of Care, Australia* (Bell & Robinson 2008), it is important that appropriate counselling, support and education are provided to the patient and their families and that feelings of hope and optimism are conveyed clearly and strongly by the treatment team.

**Psychosocial challenges:**

- Shock and adjustment to CF diagnosis including grief and loss
- Practical and logistical concerns, e.g. income support/ return to work
- Emotional /mental health needs including anxiety and depression

## 1.2 Guidelines

### 1.2.1 Undertake initial psychosocial assessment at diagnosis

- a. An initial psychosocial assessment should be performed to identify risks to and protective factors for good CF management and wellbeing, including:
  - Emotional health and wellbeing of all family members including siblings
  - Screen for depression and anxiety symptoms
  - Family relationships and dynamics
  - Parental/ caregiver relationship stability (communication and coping style)
  - History of relevant losses or chronic health conditions in the family
  - Adjustment to diagnosis
  - Family and social network responses to diagnosis
  - Adjustment to parenthood and parenting coping styles
  - Child parent attachment
  - Genetic concerns/ future fertility
  - Other relevant psychosocial stressors (needs of other family members, during hospital admission, extraneous circumstances)
  - Existing knowledge of CF/ family history
  - Financial stressors (employment flexibility, income support, returning to work)
  - Family's health literacy, comprehension of information provided and capacity to administer treatments

### 1.2.2 Assistance with practical and logistical issues

- a. Financial assistance and income support applications (information, support and referral)
- b. Transport to and from hospital/ accommodation

### 1.2.3 Referral and education

- a. Referral to Cystic Fibrosis Australia or Cystic Fibrosis New Zealand
- b. Referral to appropriate external support services based on needs assessment

### 1.2.4 Provide psychosocial and emotional support regarding:

- a. Parental / caregiver adjustment to diagnosis
- b. Parental / caregiver grief and loss
- c. Developing and maintaining routine
- d. Parenting / caregiver style and communication

## 1.3 References

Bell, S & Robinson 2008, *Cystic fibrosis standards of care in Australia*, Cystic Fibrosis Federation Australia, viewed 20 July 2017,  
<[http://www.cysticfibrosis.org.au/media/wysiwyg/CF-Australia/PDF\\_files/CFA\\_Standards\\_of\\_Care\\_journal\\_31\\_Mar\\_08.pdf](http://www.cysticfibrosis.org.au/media/wysiwyg/CF-Australia/PDF_files/CFA_Standards_of_Care_journal_31_Mar_08.pdf)>

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## 2. Infancy and early childhood

### RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Perform annual psychosocial assessment with parents/ caregivers.
- Standard 2** Identify any need for and provide relevant referrals, education and information resources.
- Standard 3** Assist with practical and logistical issues.
- Standard 4** Provide ongoing psychosocial/emotional support during this period based on individual needs assessment.

### 2.1 Overview

#### CF care burden of care

Parents/ caregivers of children with a chronic illness like CF are at an increased risk of psychosocial issues. Caring for chronically ill children can be challenging in a range of ways. Parents/caregivers of children with CF may be required to perform several additional hours of care each day including physiotherapy, diet management, administering medication and travelling to and from the hospital or medical appointments (Barker & Quittner 2016). Parents of chronically ill children also report significantly higher levels of parenting stress, not only due to the increased care responsibilities but potential difficulties in managing employment and social activities (Barker & Quittner 2016).

#### Parent/caregiver wellbeing and mental health

Parents/caregivers of chronically ill children are at risk of lower Health Related Quality of Life (HRQOL), experience more posttraumatic stress disorder (PTSD) symptoms and higher levels of distress (Van Oers et al. 2014). Parents/caregivers and particularly mothers, are also at an increased of anxiety and depression (Barker & Quittner 2016; Van Oers et al. 2014). The consequences of this are wide reaching and can impact parent-child relationships and the well-being and physical health of the patient (Van Oers et al. 2014).

The strongest factors associated with parental/caregiver anxiety and depression are practical problems in daily life such as

housing, work and parenting stress (Van Oers et al. 2014). Several studies have identified key factors associated with parental/caregiver anxiety and depression including increased risk for younger parents, parents with lower education levels and parents who lack supportive people around them.

#### Marital/ relationship breakdown

Couples with a child with CF are more likely to experience relationship strain and separation than those without (Douglas et al. 2016) Separated or divorced parents face distinct challenges in providing care for children with CF. To achieve the best standard of care for the child CF management and well-being parents/caregivers should be supported in developing effective co-parenting arrangements (Russell et al. 2016).

#### Family functioning and adjustment

Family members of children with CF will experience family life differently from those without a child with CF. Evidence suggests that most families with a child with CF will adjust well in relation to the parent-child relationship and parenting styles (Pinquart 2013). Family routines are strongly associated with increased adherence to treatment but parents often find it difficult to develop and maintain routines (Grossoehme et al. 2014). Providing anticipatory guidance to parents in developing routines and strategies to manage potential barriers may help foster the use of routines and improve CF management (Grossoehme et al. 2014).

Difficulties that emerge in parent-child attachment relationships have negative impacts on long term health status (Barker & Quittner 2016; Goldbeck, Fidika, Herle & Quittner 2015). Social work intervention should emphasise coping strategies and social support, provide practical support and appropriate therapeutic support or referrals for mental health concerns.

#### **Psychosocial challenges:**

- Adjustment to diagnosis
- Parental stress
- Family functioning / parenting style
- Practical and logistical challenges e.g. income support
- Emotional /mental health needs, high risk of PTSD symptoms, anxiety and depression

## 2.2 Guidelines

### 2.2.1 Annual psychosocial assessment

- a. A psychosocial assessment should be performed annually with parents/caregivers to identify risks to and protective factors for good CF management and wellbeing, including:
  - Family relationships and dynamics
  - Adjustment to diagnosis
  - Adjustment to parenthood, parenting coping styles
  - Other relevant psychosocial stressors (needs of other family members, during hospital admission, extraneous circumstances)
  - Emotional health and wellbeing of all family members
  - Screening for anxiety, depression and PTSD
  - Parental relationship stability (communication and coping style)
  - Financial stressors (employment flexibility, benefit payments, returning to work)
  - Family's health literacy, comprehension of information provided and capacity to administer treatments.

### 2.2.2 Provide education and Information resources

- a. Referral to external support services (e.g. respite services)
- b. Long term counselling.

### 2.2.3 Assist with practical and logistical issues

- a. Patient travel claims
- b. Transport to and from hospital
- c. Financial assistance and income support applications (information, support and referral)

### 2.2.4 Provide psychosocial and emotional support regarding:

- a. Parental / caregiver adjustment to diagnosis
- b. Parental / caregiver grief and loss
- c. Developing and maintaining routine
- d. Parenting / caregiver style and communication
- e. Coping styles and strategies
- f. Parental/ child attac

## 2.3 References

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Van Oers ,H, Haverman, L, Limperg, P, Van Dijk-Lokkart, E, Maurice-Stam, H & Grootenhuis M 2014, 'Anxiety and depression in mothers and fathers of a chronically ill child', *Maternal and Child Health Journal*, vol. 18, no. 8, pp.1993-2002, DOI: 10.1007/s10995-014-144

## 3. Childhood: primary school years

### RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Perform annual psychosocial assessment with patients and parents/caregivers.
- Standard 2** Identify any need for and provide relevant referrals, education and information resources.
- Standard 3** Referral to appropriate additional supports and programs.
- Standard 4** Provide ongoing psychosocial/ emotional support during this period based on individual needs

### 3.1 Overview

#### Wellbeing and mental health

Both parents/caregiver and patients with CF are at high risk of anxiety and depression (Ploessl, Pettit & Donaldson 2013; Quittner et al. 2016a). Depressive symptoms in children are significantly associated with low rates of adherence to treatment (Smith et al. 2010; Quittner et al. 2016b) Children with CF may experience a feeling of vulnerability, a lack of independence and opportunity, isolation and disempowerment (Jamieson et al. 2014). This will depend on each patient's adjustment to CF, health status and health literacy. Screening for symptoms is important for both patients and parents/caregivers.

#### Social wellbeing and school

Patients with CF who have a negative schooling experience during this stage can feel increasingly socially isolated. They also have a higher risk of mental health problems and this can impact their academic achievement (Branch-Smith et al. 2017). Patients can feel socially isolated due to absences from school and extracurricular activities because of CF (Jamieson et al. 2014). They may be bullied about their different physical appearance, symptoms like coughing and frequent toilet visits or by peers thinking that CF is contagious (Jamieson et al. 2014). Social workers can provide important support by helping young people develop the skills to talk about and explain CF to their peers.

### Family functioning

The wellbeing of young patients with chronic conditions like CF is heavily influenced by how well their families function (Leeman et al. 2016). Leeman et al.'s (2016) meta-analysis on the wellbeing of children with chronic illnesses and family functioning demonstrated a significant correlation between dimensions of family functioning and children's behaviour, social competence, quality of life and physical health. The family dimensions that had the most significant impact on child outcomes were family cohesion and conflict. Despite this well-established connection, there is little available research on the impact of specific interventions that target family functioning.

Siblings of children with CF have an important role in family functioning and tend to experience a higher incidence of emotional and behavioural problems than their peers. However, studies have also indicated that siblings can have increased maturity, empathy and involvement because of their sibling's CF (Fullerton et al. 2016). Siblings may also need support and education regarding disclosure. Social workers can provide support in helping siblings understand the impacts of disclosing their siblings CF without consent.

#### **Psychosocial challenges:**

- Parental stress
- Family functioning
- Practical and logistical challenges e.g. income support
- Emotional /mental health needs, high risk of anxiety and depression
- Social isolation and managing CF at school

## 3.2 Guidelines

### 3.2.1 Annual psychosocial assessment

- a. A psychosocial assessment should be performed annually to identify risks to and protective factors for good CF management and wellbeing, including:
  - Emotional health and wellbeing of all family members
  - Screening for anxiety, depression and PTSD
  - Family relationships and dynamics
  - Parental relationship stability (communication and coping styles)
  - Patient and parent/caregiver adjustment to diagnosis
  - Patient's self-esteem and self-image
  - Adjustment to parenthood, parenting coping styles
  - Other relevant psychosocial stressors (needs of other family members, during hospital admission, extraneous circumstances)
  - Financial stressors (employment flexibility, benefit payments, returning to work)
  - Family's health literacy, comprehension of information provided and capacity to administer treatments
  - Patient's social wellbeing (school, extracurricular activities)

### 3.2.2 Provide referrals education and Information resources

- a. Referral to external support services (e.g. respite services, counselling)
- b. Age appropriate referral for patients to online peer support groups
- c. Make a Wish or Starlight Foundation
- d. Education support services
- c. Financial assistance and income support applications (information, support and referral)

### 3.2.3 Assist with practical and logistical issues

- a. Patient travel claims
- b. Transport to and from hospital

### 3.2.4 Provide psychosocial and emotional support regarding:

- a. Patient and parental / caregiver adjustment to school
- b. Patient and parental / caregiver grief and loss
- c. Developing and maintaining new school and CF routine
- d. Parenting / caregiver style and communication
- e. Coping styles and strategies
- f. Parental/ child attachment

### 3.3 References

Branch-Smith, C, Shaw, T, Lin, A, Runions, K, Payne, D, Nguyen, R, Hugo, H, Cross, D 2017, 'Developing an innovative online intervention to support schooling for children and young people with cystic fibrosis', *Journal of Cystic Fibrosis*, vol.16, pg. 39, DOI: 10.1016/S1569-1993(17)30286-2.

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Quittner, A, Saez-Flores, E & Barton, J 2016b, 'The psychological burden of cystic fibrosis', *Current Opinion in Pulmonary Medicine*, vol. 22, no. 2, pp. 187-191, DOI: 10.1097/MCP.000000000000244.

# 4. Adolescence: late primary school-young adulthood

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Perform annual psychosocial assessment, with patients and parents/caregivers.
- Standard 2** Identify any need for and provide relevant referrals, education and information resources.
- Standard 3** Encourage and support independence in self-care and private consultations.
- Standard 4** Provide ongoing psychosocial/ emotional support during this period based on individual needs.

### 4.1 Overview

#### Patient self-care and CF management

It is well known that adolescence is a developmental stage which presents new challenges for young people and their families. For young people with CF there are unique dimensions to this transition period. Many patients experience a loss of freedom and opportunities because of the health impacts and treatment burden of CF. Tensions may emerge between parents/ caregivers and patients as the responsibility for treatments begins to shift from parent to child.

To encourage autonomy and empower patients it is important that clinicians begin

to speak directly young people and listen to their needs. Patients report feeling

ignored and devalued when clinicians speak only to their parents/caregivers and use complex medical terminology (Jamieson et al 2014). Ideally, patients should begin to attend some or part of their appointments alone.

The invasive nature of some CF treatments causes some patients to feel a loss of control over their body (Jamieson et al 2014). Fostering independence and self-management with young people is

essential to create feelings of normality and self-confidence.

#### Risk taking

Risk taking behaviours such as drug, tobacco and excessive alcohol use have additional health risks for young people with CF. These behaviours and poor adherence to CF treatment become common during this time (Keyte, Mantzios & Jackson, 2017). Often patients will avoid speaking to their care team about high risk activities in front of parents/caregivers, leading to high risk behaviours, defiance and hiding issues with adherence (Jamieson et al 2014).

#### Identity, self-esteem, social wellbeing and sexual health

Adolescent patients described relationships with other people with CF as particularly important for their wellbeing (Jamieson et al 2014). Online forums for this age group have been shown to have a positive effect on feelings of acceptance and social support (Kirk & Milnes 2015). Studies show that optimism may act as a protective factor against the stigma of chronic illness (Oliver et al. 2014).

Body image and self-esteem can become issues due to the short stature, weakness and susceptibility to infections experienced by patients. Young men can feel particularly concerned about not being able to achieve a typically 'masculine' physique (Jamieson et al. 2014).

Sexual and reproductive health (SRH) is profoundly impacted by CF. Adolescents with CF reach the same developmental milestones as their healthy peers and at this stage, SRH should be discussed with their medical consultants (Frayman & Sawyer 2015). Sexuality, consent and healthy relationships should be discussed and social workers should actively make space for patients to discuss these issues during outpatient or inpatient contact (Frayman & Sawyer 2015).

#### Career planning

Young people with CF must consider several disease related factors when considering their future career choices including workplace risks like air quality and work place flexibility. CF can impact a persons' employment options. Not only due to the requirements of air quality, flexible hours, and fatigue but also due to educational attainment and proximity to treatment (Targett et al. 2013).

#### Psychosocial challenges:

- Shifting roles in CF management between patient and parents/caregivers
- Patient adjustment to diagnosis
- Identity development and self-esteem
- Social wellbeing
- Sexual and reproductive health
- Career / study planning

## 4.2 Guidelines

### 4.2.1 Annual psychosocial assessment

- a. Assess patient's willingness to engage in social work review and focus on outcomes desired by the patient.

A psychosocial assessment should be performed annually to identify risks to and protective factors for good CF management and wellbeing, including:

- Impact of CF on patient's life (including co-morbidities)
- Screen for anxiety and depression
- Patient understanding of their health status
- Patient's level of adherence to CF treatment
- Self-esteem/ body image
- Skills for independent living
- Level of education/future vocational aspirations
- Peer and family relationships; extended support systems
- Connections to wider community/social connectedness
- History of hospital admissions (e.g. first admission, regular admissions impact of, procedural anxiety/trauma, institutionalised outlook)
- SRH – relationship status, sexuality, sexual identity/ fluidity, gender identity / fluidity, sexual health (risk factors/ contraception and protection)

- Risk taking behaviours, drug, tobacco and alcohol consumption

### 4.2.2 Provide referrals, education and information resources

- a. Sexual and reproductive health resources
- b. Healthy relationships
- c. Risk taking behaviours and harm minimisation
- d. Discussion/ information regarding transplant
- e. Mental health services or ongoing counselling
- f. Online peer support or other peer support/ mentoring program

### 4.2.3 Assist with practical and logistical issues

- a. Liaise with vocational assistance in secondary and tertiary settings
- b. Explore income support options

### 4.2.4 Encourage private consultations

- a. Patients and parents/caregivers should be encouraged to allow for private consultations/ time alone with all members of the MDT

### 4.3 References

Jamieson, N, Fitzgerald, D, Singh-Grewal, D, Hanson, C, Craig, J & Tong, A 2014, 'Children's Experiences of Cystic Fibrosis: A Systematic Review of Qualitative Studies', *Pediatrics*, vol.133, no. 6, DOI: 10.1542/peds.2014-0009.

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Keyte, R, Egan, H, Mantzios, M & Jackson, C 2017, 'Paediatric cystic fibrosis patients' attitudes and beliefs regarding risky health behaviours', *Journal of Cystic Fibrosis*, vol. 16, pp. 39, DOI: 10.1016/S1569-1993(17)30287-4.

Kirk, S. & Milnes, L 2015, 'An exploration of how young people and parents use online support in the context of living with cystic fibrosis', *Health Expectations*, vol. 19, no. 2, pp.309-321, DOI: 10.1111/hex.12352.

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Targett, K, Bourke, S, Nash, E, Murphy, E, Ayres, J & Devereux, G 2013, 'Employment in adults with cystic fibrosis', *Occupational medicine*, vol. 64, no.2, pp.87-94, DOI: 10.1093/occmed/kqt140.

Quittner, A, Abbott, J, Georgiopoulos, A, Goldbeck, L, Smith, B, Hempstead, S, Marshall, B, Sabadosa, K, Elborn, S, Boyle, M & Crossan, A 2016, 'International committee on mental health in cystic fibrosis: Cystic fibrosis foundation and European cystic fibrosis society consensus statements for screening and treating depression and anxiety'. *Thorax*, vol. 71, no. 1, pp.26-34, DOI: 10.1136/thoraxjnl-2015-207488.

# 5. Transfer of Care

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Discuss transfer of care plans and readiness early and regularly with patients and parents/caregivers.
- Standard 2** Work collaboratively with paediatric / adult care teams to ensure successful transfer of care process.
- Standard 3** Provide appropriate education and information resources to patients, focused on addressing gaps in knowledge and empowering patients to self-manage their health care.

### 5.1 Overview

#### Defining transfer of care

‘Adolescent transition’ and ‘transfer of care’ are often used synonymously. It is important to separate the period of adolescence from the transfer of care between CF teams. Transition occurs across the course of the lifespan and transfer of care is a discrete period when patients move from the paediatric care team to the adult care team. Conversations regarding transfer of care/transition in developmental stages should be discussed with patients and parents/caregivers across the lifespan.

#### Transfer challenges

The advances in CF treatment and care have extended life expectancy and improved the quality of life for CF patients.

This extension in life expectancy means that a patient will transfer from paediatric to adult care teams for the continued management of their CF. However, this transition/transfer presents several practical challenges in changing to a new team of health professionals, a new CF care centre and the shifting of treatment management from parents or caregivers to the new adult patient.

Challenges during transition/transfer to adult care can emerge due to the developmental and life stage in which it occurs. Issues that are part of ‘growing up’ like work and study choices, relationships, reproductive health, family planning, drug/alcohol/ tobacco use and identity and autonomy, all interact with CF treatment

transition. Abstract thinking is still developing at this young adult stage making it difficult for patients to plan and prepare for medical needs. Consequences of avoiding care seem remote and the pleasure of risk taking behaviours outweighs potential negative health outcomes. Due to these age and stage factors, assessing a patient's readiness to transition/transfer in relation to their self-care readiness, treatment adherence and mental and emotional wellbeing are critical (Okumura et al. 2014).

#### Health outcomes

These challenges can lead to a decline in adherence to therapies and treatments and can cause reduced health such as a loss in pulmonary function, decrease in body mass index and increased hospitalisations (Okumura et al 2014; Tierney et al. 2013). This phenomenon is not unique to CF. Young adults with other chronic conditions experience a deterioration in health that reflect difficulties in self-care and adherence to treatments (Okumura et al 2014). Care providers in both the paediatric and adult care teams can face challenges in providing care for young adults and can struggle to meet the psychosocial needs of patients during transition (Okumura et al. 2014).

#### Transfer processes

While there can be significant challenges for transition/transfer, the evidence suggests that young people with CF fare

better through transition/transfer than young people with other chronic conditions due to the benefits of dedicated CF care systems (Megumi, Okumura & Kleinhenz 2016). Each CF centre or care team will have different transition/transfer procedures and resources. Thus, the following guidelines are designed to address the common risks and best practices during transition/transfer. Structured transition/transfer processes are demonstrated to produce better health outcomes and patient experiences (Chaudhry, Keaton & Nasr 2013).

Transition/transfer practices can be categorised in the following way: assessing, planning, evaluating, providing information, promoting independence and following up (Al-Yateem 2013; Schwartz et al. 2014). Social workers are well placed to facilitate good transition due to social work competencies – developmental frameworks, resource facilitation, and patient/provider relationships. Akre and Suris' (2014) research highlights the importance of fostering shared management in early adolescence.

#### **Psychosocial challenges:**

- Adjustment to new care team
- Increase in patient's control and responsibility for CF management and health
- Continuity of care and risk of reduced health status

## 5.2 Guidelines

### **5.2.1 Discussing and planning readiness to transition/transfer**

- a. Clinicians should begin discussing transition/transfer with patients and parent/caregivers as early as appropriate
- b. Encourage and support young people to develop self-care skills
- c. Plan and discuss with patient and carers their readiness to transition
- d. Discuss school/education plans, housing and entitlements
- e. Assess psychosocial readiness for transition/transfer

### **5.2.2 Collaboration between paediatric and adult care teams**

- a. Successful transition/transfer requires collaboration between paediatric and adult care teams
- b. Clinicians from both care teams should maintain regular communication prior to, during and for a period after patient transitions/transfer to ensure continuity of care.

### **5.2.3 Provide education and information resources to patients, focused on addressing gaps in knowledge and empowering patients to self-manage their health care**

- a. Following assessment of a patient's readiness to transition/transfer any gaps should be addressed with the provision of appropriate information and education resources.
- b. Parents/ caregivers should be provided with appropriate context specific information regarding the transition/transfer process and their changing role in the patient's care
- c. Discussion/ information regarding transplant

### 5.3 References

Al-Yateem, N 2013, 'Guidelines for the transition from child to adult cystic fibrosis care: Evidence suggests that nurses require improved guidance on how to help transfer young patients with this condition to adult healthcare services' *Nursing children and young people*, vol. 25, no. 5, pp.29-34, DOI: 10.7748/ncyp2013.06.25.5.29.e175.

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Okumura, M, Ong, T, Dawson, D, Nielson, D, Lewis, N, Richards, M, Brindis, C & Kleinhenz, M 2014, 'Improving transition from paediatric to adult cystic fibrosis care: programme implementation and evaluation' *BMJ Quality & Safety*, vol.23, pp. 64-72, DOI: 10.1136/bmjqs-2013-002364.

Schwartz, L, Daniel, L, Brumley, L, Barakat, L, Wesley, K & Tuchman, L, 2014, 'Measures of readiness to transition to adult health care for youth with chronic physical health conditions: a systematic review and recommendations for measurement testing and development', *Journal of Pediatric Psychology*, vol. 39, no. 6), pp.588-601, DOI: 10.1093/jpepsy/jsu028.

Tierney, S, Deaton, C, Jones, A, Oxley, H, Biesty, J. & Kirk, S 2013, 'Liminality and transfer to adult services: a qualitative investigation involving young people with cystic fibrosis', *International Journal of Nursing Studies*, vol. 50, no. 6, pp.738-746, DOI: 10.1016/j.ijnurstu.2012.04.

# 6. Adulthood

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Perform annual psychosocial assessment with patients.
- Standard 2** Identify any need for and provide relevant referrals, education and information resources.
- Standard 3** Provide ongoing psychosocial/ emotional support during this period based on individual needs.

### 6.1 Overview

#### Wellbeing and mental health

In addition to fatigue caused by the high level of care, people with CF may experience chronic pain and challenges with sexuality, relationships and autonomy (Pakhale et al. 2015). The interaction between symptoms, care and treatment burden and perceived reactions to CF can result in people with CF feeling stigmatised, experiencing psychological distress, negative emotions, low self-esteem, helplessness and depressive symptoms (Pakhale et al. 2015; Platten 2013; Quittner et al. 2016). The largest international study measuring depression and anxiety in people with CF found that 19% of adults experienced elevated symptoms of depression and 32% of adults experienced elevated symptoms of anxiety (Quittner et al. 2014).

Younger and older adults with CF may experience a range of concerns that may contribute to low mood, anxiety or depression. These include intimate and familial relationships, grief and loss, life transitions, decline/ perception of quality of life, pain management, worries, body image, self-esteem, life stress, life/death/existentialism, neurocognitive difficulties, CF adherence, changing work/life balance, stigma/ disclosure, traumatic experiences and adjustment to CF (Pakhale et al. 2015).

#### Psychosocial challenges:

- CF management fatigue
- Adjustment to changing health status
- Intimate and family relationships
- Family planning
- Managing CF and work
- Possibility of organ transplants

## 6.2 Guidelines

### 6.2.1 Annual psychosocial assessment

- a. A psychosocial assessment should be performed annually to identify risks to and protective factors for good CF management and wellbeing, including:
  - Family planning and pregnancy
  - Screen for anxiety and depression
  - Adjustment to health status
  - Employment, retirement and income support
  - Family planning

### 6.2.2 Provide relevant referrals, education and information resources

- a. Consultant referral to genetic counselling/ IVF services

- b. Referral to Advance Care Planning/ Respecting Patient Choices services
- c. Provision of information and referral for transplantation services
- d. External counselling
- e. Referrals for families- counselling support
- f. Discussion/ information regarding transplant

### 6.2.3 Provide psychosocial and emotional support regarding:

- a. Adjustment to health status
- b. Grief and loss
- c. Family planning
- d. Coping styles and strategies

## 6.3 References

Pakhale, S, Baron, J, Armstrong, M, Tasca, G, Gaudet, E, Aaron, S, Cameron, W. & Balfour, L 2015, 'A cross-sectional study of the psychological needs of adults living with cystic fibrosis', *PLOS ONE*, vol. 10, no.6, pp. 1-13, DOI: 10.1371/journal.pone.0127944.

Platten, M, Newman, E & Quayle, E 2013, 'Self-esteem and its relationship to mental health and quality of life in adults with cystic fibrosis', *Journal of clinical Psychology in Medical Settings*, vol. 20, no. 3, pp.392-399, DOI: 10.1007/s10880-012-9346-8.

Quittner, A, Goldbeck, L, Abott, J, Duff, A, Lambrecht, P, Sole, A, et al. 2014, 'Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers: Results of the International Depression Epidemiology Study (TIDES) across Nine Countries' *Thorax*, vol. 69, pp. 1090–1097, DOI: 10.1136/thoraxjnl-2014-205983.

Quittner, L, Abbott, J, Georgiopoulos, A, Goldbeck, L, Smith, B, Hempstead, S, Marshall, B, Sabadosa, K, Elborn, S, Boyle, M & Crossan, A 2016, 'International committee on mental health in cystic fibrosis: Cystic fibrosis foundation and European cystic fibrosis society consensus statements for screening and treating depression and anxiety', *Thorax*, vol. 71, no. 1, pp.26-34, DOI: 10.1136/thoraxjnl-2015-207488.

# 7. Transplantation

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

- Standard 1** Provide ongoing psychosocial support throughout the transplant process with regular patient contact.
- Standard 2** Prepare psychosocial assessment for transplant team.
- Standard 3** Identify any need for and provide relevant referrals, education and information resources.

### 7.1 Overview

#### Lung transplant

Lung transplantation is usually discussed before a patient's predicted life expectancy is less than two years. This will occur at different stages for everyone. Not all patients will desire or be eligible for a transplant. Current survival rates post lung transplantation in Australia and New Zealand is around 87% (one-year), 70% (three-year) and 60% (five -year) (ANZCOTR 2015). Unfortunately, organ availability will mean that some patients will die waiting for a transplant. Not all patients will want to be assessed for a lung transplant.

Being listed for a lung transplant can be an alarming signal for patients that their health has reached a critical stage (Jamieson et al 2014). Some people may feel unprepared and shocked. The time people wait on the transplant list is

characterised by 'uncertainty, anxiety and disappointing false alarms' (Jamieson et al.

2014). Reframing transplant as an opportunity for improved HRQOL may be a useful tool to overcome this. Transplant can also be a source of great hope and optimism for patients with the potential of improved health related quality of life.

There is limited research available on the psychosocial needs of patients throughout the transplantation process. Jamieson et al. (2014). described a lack of communication from the CF team while on the waiting list as a cause of distress.

#### Post-transplant care

David et al. (2015) recommended ongoing psychosocial support post-transplant, to help patients adjust to their new health status and particularly regarding fertility

and pregnancy (that is considered high risk post-transplant for women).

### Multi organ transplant

It is important to be aware that, while less common, some CF patients may require a liver or multi-organ transplant. For further information regarding this refer to the local state liver and lung transplant services.

## 7.2 Guidelines

### 7.2.1 Ongoing psychosocial support throughout the transplant process

- a. Education and information about the transplant process
- b. Refer to/ liaise with transplant service if patient is transferring to another service
- c. Explore income support options
- d. Regular contact while on waitlist
- e. Support adjusting post-transplant to new health status
- f. Focus on positive framing and improvement in HRQOL

### Psychosocial challenges:

- Distress and anxiety while on the transplant waiting list
- Adjustment to changing health status pre-and post-transplant

### 7.2.2 Prepare psychosocial assessment for transplant team

### 7.2.3 Identify any need for and provide relevant referrals, education and information resources

- a. External counselling
- b. Referrals for families- counselling support

### 7.3 References

Australia and New Zealand Cardiothoracic Organ Transplant Registry (ANZCOTR) 2015, 2015 Report, <<http://anzcotr.org.au/v3custom/anzcotr-2015.pdf>>

David, V, Feldman, D, Danner-Boucher, I, Le, A, Guyomarch, B, Ravilly, S & Marchand, C 2015, 'Identifying the Educational Needs of Lung Transplant Recipients with Cystic Fibrosis', *Progress in transplantation*, vol. 25, no. 1, pp. 18-25, DOI: 10.7182/pit2015526.

Jamieson, N, Fitzgerald, D, Singh-Grewal, D, Hanson, C, Craig, J & Tong, A 2014, 'Children's Experiences of Cystic Fibrosis: A Systematic Review of Qualitative Studies', *Pediatrics*, vol.133, no. 6, DOI: 10.1542/peds.2014-0009.

# 8. End of life care

## RECOMMENDED STANDARDS FOR SOCIAL WORK CARE:

**Standard 1** Provide ongoing psychosocial / emotional support during this period based on individual needs assessment.

**Standard 2** Assist with practical and logistical issues.

**Standard 3** Identify any need for and provide relevant referrals, education and information resources.

### 8.1 Overview

While advances in the treatment and management of CF have extended life expectancy into middle adulthood, CF remains a life limiting disease. End of life is a period when the health status of a person has deteriorated and death is likely to occur within a limited timeframe, typically within two years. During this time, there can be challenges for the treating team in delivering active treatment while providing adequate symptom control, like pain relief. This is especially difficult for patients on the lung transplant waiting list.

During this time several alarming, confronting and invasive treatments may take place. Patients and families require ongoing access to emotional psychosocial support.

#### **Psychosocial challenges:**

- Adjustment to prognosis
- Grief and loss
- Mental and emotional wellbeing – depression and anxiety
- Familial/parental/ partner emotion and mental wellbeing and grief and loss

## 8.2 Guidelines

### **8.2.1 Provide ongoing psychosocial/ emotional support**

- a. Ongoing grief and loss support for the patient and their family/carers/partners
- b. Adjustment to prognosis and health status
- c. Short term ongoing bereavement support for family/carers/ partners post patient death

### **8.2.2 Assist with practical and logistical issues**

- a. Travel and accommodation arrangements for families/ carers/ partners during palliative care

### **8.2.3 Identify any need for and provide relevant referrals, education and information resources**

- a. Referral to palliative care
- b. Referral for family/ carers/ partners to ongoing bereavement support and counselling

# APPENDIX 1

## Useful Online Resourc

Nutrition Education Materials Online cystic fibrosis resources

[https://www.health.qld.gov.au/nutrition/neo\\_cysticfibrosis](https://www.health.qld.gov.au/nutrition/neo_cysticfibrosis)

CFSmart information booklets: include everything you need to know about CF and school

<https://cfsmart.org/>

Online CF Sexual & Reproductive Health resource for young people with CF

<http://www.spill.org.au/>

Healthy cooking for people with Cystic Fibrosis

<https://www.cfcooking.org/1-healthy-fats>

Cystic Fibrosis Carrier screening- Information Resources Kit

<http://www.cysticfibrosis.org.au/vic/info-resources-kit>

Australian Indigenous HealthinfoNet- Cystic fibrosis

<http://www.healthinfonet.ecu.edu.au/key-resources/organisations?oid=1729>

Raising children Network- Cystic Fibrosis [http://raisingchildren.net.au/articles/cystic\\_fibrosis\\_d.html](http://raisingchildren.net.au/articles/cystic_fibrosis_d.html)

What they don't tell you- CF SRH

[http://www.rch.org.au/uploadedFiles/Main/Content/cah/What\\_they\\_dont\\_tell\\_you.pdf](http://www.rch.org.au/uploadedFiles/Main/Content/cah/What_they_dont_tell_you.pdf)

Cystic Fibrosis community care

<http://www.cysticfibrosis.org.au/vic/>

EDMed Professional development for teachers

<https://learningprogram.rmhc.org.au/about/edmed-professional-development.php>

Cystic Fibrosis Foundation

<https://www.cff.org/>

European Cystic Fibrosis Society

<https://www.ecfs.eu/>

Cystic Fibrosis Trust United Kingdom

<https://www.cysticfibrosis.org.uk/>

Cystic Fibrosis Australia

<http://www.cysticfibrosis.org.au/>

Cystic Fibrosis New Zealand

<http://cfnz.org.nz/>

Staying connected with friends

<https://learningprogram.rmhc.org.au/>

Make A Wish Foundation

<http://www.makeawish.org.au/>  
<https://www.makeawish.org.nz/>

Starlight Children's Foundation

<https://starlight.org.au/>

Carers Australia

<http://www.carersaustralia.com.au/>

CF Buzz- Parents & Carers- Learn how to manage cystic fibrosis for your children

[http://www.cfbuzz.org/parents-and-carers/?gclid=EAlaIQobChMI243L2NKg1QIVhgYqCh2YsQAbEAAAYAiAAEgKZ2PD\\_BwE](http://www.cfbuzz.org/parents-and-carers/?gclid=EAlaIQobChMI243L2NKg1QIVhgYqCh2YsQAbEAAAYAiAAEgKZ2PD_BwE)

Starship Child Health- Child and Youth Cystic Fibrosis Clinical Network

<https://www.starship.org.nz/for-health-professionals/new-zealand-child-and-youth-clinical-networks/child-and-youth-cystic->

