Mental Health in Cystic Fibrosis (CF):

The impact of CF on mental health and the impact of mental health conditions on CF

Learning Outcomes

At the end of this module, you should be able to:

- **Describe** the impact of CF on mental health and the prevalence of mental health conditions in CF populations
- **Explain** the impact of mental health conditions on self-care, general health, functioning and quality of life for people with CF
- **Differentiate** between the various psychosocial risk factors and challenges faced by people with CF at each stage of life
- **Explain** the mental health care needs of people with CF at each stage of life
- **Describe** the impact of CF on the mental health of carers.

Module Overview

There are four components of this module:

- **Part 1:** Improved longevity in CF and its implications for mental health
- **Part 2:** Prevalence of mental health conditions in people living with CF
- **Part 3:** Understanding and identifying mental health issues in people with CF at different life stages
- **Part 4:** Effects of mental health conditions in CF on carers

Expert steering committee

Dr. **Anna M. Georgiopoulos**, *Associate Professor of Psychiatry, Harvard Medical School and Consulting Psychiatrist, Massachusetts General Hospital Cystic Fibrosis Program*

Dr Georgiopoulos is an adult, child and adolescent, and consultation-liaison psychiatrist who has been working with the Massachusetts General Hospital CF Program (Boston, USA) since 2005 to promote psychiatric resilience and optimal medical outcomes. Dr. Georgiopoulos chaired workgroups for the CF Foundation/European CF Society consensus statements for screening and treating depression and anxiety and the CF Foundation models of palliative care delivery guidelines, and is co-principal investigator for clinical trials in CF mental health and palliative care. She leads the education and training subgroup of the CF Foundation Mental Health Advisory Committee, co-chairs the Junior Investigator Award for Clinical Research at the North American CF Conference, and serves on the CF Foundation Data Safety Monitoring Board and European CF Society Mental Health Working Group. In 2018, she received the CF Foundation's Mattingly Leadership in Mental Health Care Award. Dr. Georgiopoulos had the great pleasure of meeting members of the CF community and CF care teams in 6 Australian cities during the CF Australia Mental Health Roadshow.

Dr. **Lucy Holland** (PhD). *Adjunct Fellow, Queensland University of Technology. Accredited Social Worker, Professional Coach & Consultant, Simplify Consulting, Australia.*

Lucy is an accredited social work clinician, professional coach, consultant and academic who has worked in health and adolescent health for over 15 years. As a Senior Social Worker, Lucy has worked with individuals and families across tertiary and community health sectors, in acute, trauma and community settings. Lucy also works as a professional coach and adolescent health consultant through her business Simplify Consulting. Lucy has significant academic teaching and research experience in adolescent health, integrated care, professional education, mixed methods and health systems research and evaluation. Lucy has recently completed a PhD in Public Health that examined the optimisation of care for adolescents and young adults living with cystic fibrosis. Lucy is passionate about the working at the interface between practice and research to drive effective health service development, delivery and evaluation. She is committed to ensure meaningful, consumer-led, humanistic approaches to research and care that prioritise true consumer partnership.

Anthony Talbot, *Clinical psychologist, Melbourne, Victoria, Australia*

Anthony is a clinical psychologist who has worked in CF care at the Alfred for 10 years. He works with CF patients presenting with a range of psychological concerns, often in the context of significant illness, loss and understandable distress. His approach is to explore a person's own goals for achieving physical and mental wellbeing in the larger context of their life, relationships, values and strengths. Psychotherapy ideally supports adaptation, improved behavioural management of chronic illness, new coping skills and strengthened confidence to make changes. His approach integrates cognitive behavioural therapy, mindfulness, lifespan development and existential psychotherapy. He has a background in smoking cessation. Other interests include loss and grief, motivational interviewing, developmental transitions and sexual identity.

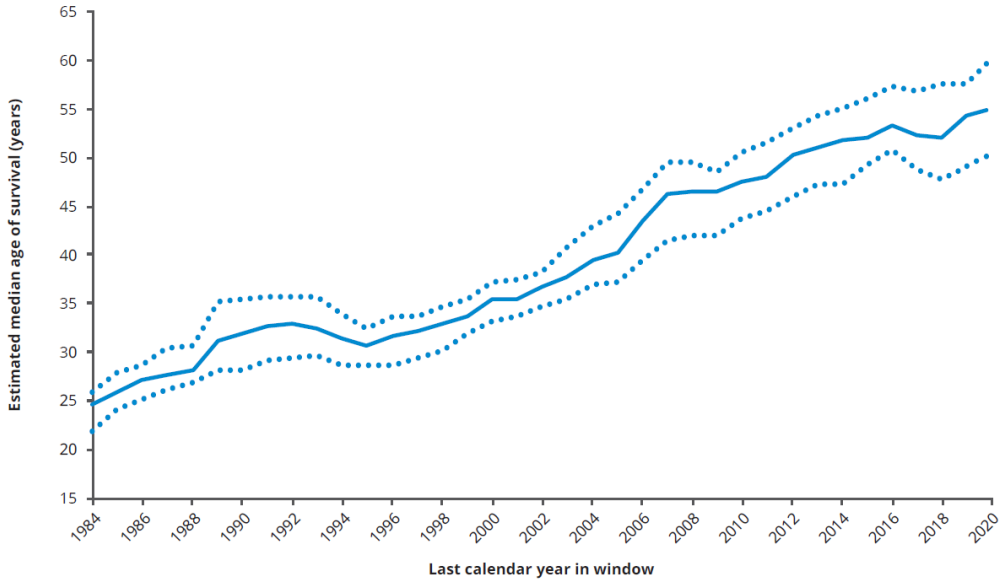
Improved longevity in CF and its implications for mental health

Part 1

The improvement of median age of survival[a] in CF is a major medical achievement

In Canada, **median age of survival surpassed 50 years of age for the first time in 2012 and has remained steady since.** The number of Canadian adults living with cystic fibrosis has also increased in the last few decades.

Estimated median age of survival for a moving 5-year window with 95% confidence intervals, 1984 to 2020.



**Adapted from The Canadian Cystic Fibrosis Registry Annual Data Report 2020.*

Accordingly, most Canadians with CF today will require more life years of care to be provided in adulthood.

CF IN CANADA: QUICK STATS

Median age:
23.8 years

Estimated median age of survival:
55.4 years

Proportion of people with CF who are adults:
62%

[a]The median age of survival is the age beyond which we expect 50% of babies with cystic fibrosis born today to live, under the assumption that recent age-specific mortality rates will hold for the rest of their lives. This measure is used in lieu of life expectancy because it is less influenced by extreme values.[1]

Cystic Fibrosis Canada. (2022). *The Canadian Cystic Fibrosis Registry 2020 Annual Data Report*. Retrieved from https://www.cysticfibrosis.ca/Registry/2020AnnualDataReport.pdf.

This improvement is principally driven by innovations in treatment and diagnosis

In addition to key scientific advances, the steady improvement in survival has also emerged from important improvements in care, resulting from careful clinical observation and a systematic approach to treating disease complications.[1]

These include:[1]

- Introduction of **intensive antibiotic regimens and pancreatic enzymes**
- Optimisation of **nutrition**, **effective airway clearance** and **lung transplantation**
- Optimisation of anti-inflammatory **drugs** and drugs that enhance sputum clearance, and more recently, introduction of CFTR modulators
- The advent of a **multifaceted approach to CF care**, delivered by a specialist multidisciplinary team
- **Improvements in diagnosis**, including routine newborn screening.

Improved survival also means a higher treatment burden[1]

As an increasing number of treatments are prescribed, maintaining sustainable adherence to therapies is an ongoing and important issue – the time commitment associated with adequate adherence is close to 2 hours per day.[1]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

An ageing CF population imposes a new set of needs and challenges

Specialist adult CF units and appropriately trained health professionals

There is already a shortage, with strong reliance on support from paediatric colleagues.[1]

Close follow-up and monitoring for age-related medical complications

Increased incidence of CF-related diabetes, bone disease, arthropathy and cancer is expected.[1]

Support for mental health

Increasingly complex disease management makes patients and carers more vulnerable to psychological problems.[2]

1. Balfour-Lynn IM, King JA. Paediatr Respir Rev 2020 doi: 10.1016/j.prrv.2020.05.002. 2. Havermans T, Staab D. Thorax 2016;71(1):1-2.

Why does CF carry such a burden on mental health?

CF is a chronic, progressive and disabling disease that continues to be one of the most challenging to manage.[1,2]

It is a daily struggle that requires people with CF and their families to adopt multiple health-related behaviours, and continuously adapt to new CF-related challenges.[1-4] This can be overwhelming and interrupt many normal aspirations of fulfilment.[3]

Visit the following slides to discover some of the more challenging experiences of living with CF.

Biological

Psychosocial

Psychosocial impacts of CF on mental health will be explored in further detail later.

1. Quittner AL, et al. Thorax 2016;71(1):26-34. 2. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78. 3. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 4. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283.

Biological aspects of CF that affect mental health

- **Symptom burden:**
  People with CF face chronic lung infection and inflammation, gastrointestinal issues, respiratory symptoms, chronic sinusitis, nasal polyps, late onset puberty and infertility; sleep disturbance is also an issue and pain is common throughout the disease course.[1,2]
- **Treatment burden:**
  Treatment regimens are increasingly complex, time-consuming and usually involve multiple therapeutic modalities (e.g. airway clearance techniques, acute and chronic antibiotic treatment, enzyme replacements, vitamin supplements, high-calorie diet).[1,3-5] CF patients aged 6 years and over are prescribed a median number of 8 chronic medications.[5]
- **Disease progression:**
  Even though life expectancy has improved, patients must still cope with severe pulmonary exacerbations, comorbidities, transplantation, gradual deterioration in functional capabilities, and untimely death.[3,4]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283 2. Dancey DR, et al. Eur Respir J 2002;19(3):504-510. 3. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 4. Havermans T, Staab D. Thorax 2016;71(1):1-2. 5. Rouze H, et al. Patient Prefer Adherence 2019;13:1497-1510.

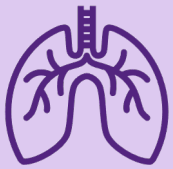
Psychosocial aspects of CF that affect mental health

- **Impact on social roles & relationships:**
  Deteriorating health can cause people with CF to withdraw from social life and/or have strong reliance on family for housing, finance or medical care; they also face concerns about finding a partner and managing parenthood.[1]
- **Occupational challenges:**
  CF can affect employment goals through activity limitations, needing to take time off for treatment, financial issues, risk of occupational health hazards, or concerns about disclosure of their disease.[1]
- **Infection control & social isolation:**
  Any in-person contact between individuals with CF is prohibited, limiting their ability to find peer support.[2]
- **Administrative challenges:**
  Managing medical paperwork, prescriptions, appointments, applying for benefits and health insurance can be complex.[1]
- **Impact on mental health**
  Going through the natural human life cycle can be difficult for a person with CF and affect their psychological wellbeing.[1] The prevalence of mental health conditions in the CF community will be explored in the next section.
- **Impact on families & carers**
  CF is one of the most challenging paediatric illnesses for families to manage.[3] Mental health in parents and caregivers of those with CF will be explored in the final section of this chapter.

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78. 3. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

Mental health matters: Link between mental and physical health is clear

The physical symptoms of CF can have an impact on mental health.[1] However, this relationship is bi-directional, with psychological distress and ill-health in both CF patients and their carers known to affect key health outcomes:[1,2]



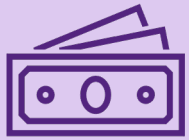
Decreased lung function

Lower BMI

Worse treatment adherence

Worse health-related quality of life

Increased exacerbations and hospitalisation



Increased healthcare costs

As patients living with CF age and the complexity of their disease increases, effective management of mental health issues becomes a critical component of care.

1. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78. 2. Quittner AL, et al. Thorax 2016;71(1):26-34.

However, mental health care delivery in CF remains inadequate

Despite much research and consistent interest in the psychological wellbeing of those with CF, support for patients and their families is still lacking.[1]

The burden of chronically addressing psychosocial challenges and mental health conditions remains primarily with individuals and their families.[2]

Findings from a survey of over 1,400 CF health professionals found:[3]

- Over 70% did not have any personal experience with mental health screening
- Many did not have a colleague trained in the skills to manage mental health issues
- Around 1 in 3 felt unable or unsure if they could refer to mental health clinicians in their hospital
- Almost half did not have an up-to-date list of mental health resources and referrals.

Training in mental health screening and access to mental health resources/ referrals were two of the most pressing needs identified, which this module aims to address.[3]

1. Havermans T, Staab D. Thorax 2016;71(1):1-2. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Abbott J, et al. J Cyst Fibros 2015;14(4):533-539.

Prevalence of mental health conditions in people living with CF

Part 2

People with CF are at higher risk for depression and anxiety

People with CF can be affected by depression and anxiety.

Depression

Depression "affects the way a person feels, thinks or behaves, which impairs social or occupational functioning".[1]

- Central to this is a depressed mood or loss of interest in most activities.[1]
- Risk for suicide is a core component.[1]

Anxiety

Anxiety is a "state of intense apprehension, uncertainty, and fear resulting from the anticipation of a threatening event or situation to the degree that normal physical and psychological functioning is disrupted".[1]

Procedural anxiety

Procedural anxiety is particularly relevant to CF – that is, an acute and excessive fear of a medical or surgical procedure that results in acute stress or avoidance, which can have negative health consequences.[1]

1. Quittner AL, et al. Thorax 2016;71(1):26-34.

Prevalence rates amongst those with CF are 2–3x higher vs community samples

While many individuals with CF and their families are tremendously resilient, an increased risk of anxiety and depression has been established.[1,2] Estimated prevalence rates are listed below.

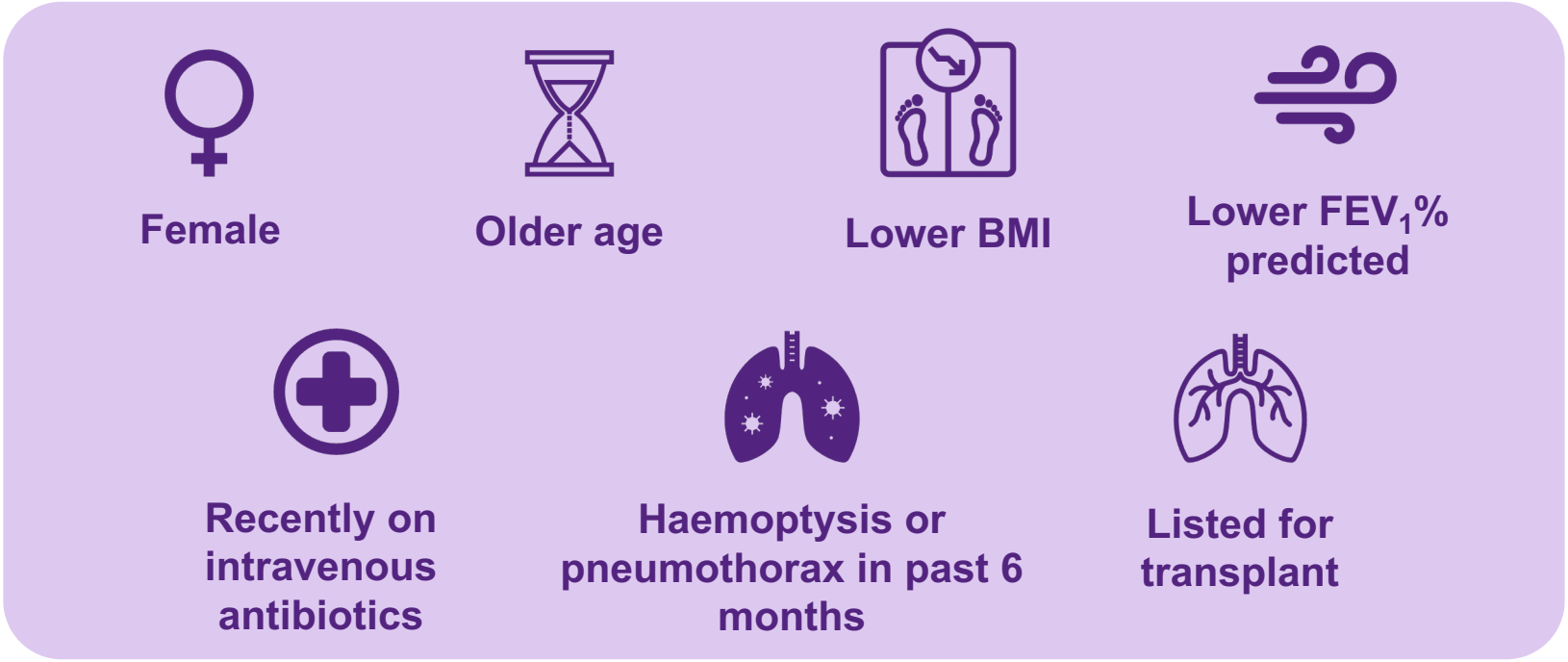
Comorbidity of anxious and depressive symptoms is also high amongst people living with CF.[1]

Children (8-11yo)	Adolescents (12-18yo)	Adults	Overall prevalence
15% of 8-11 year olds have major depression[3]	**10%** with elevated depression[1]	**19%** with elevated depression[1]	**17%** with depression[1] *2x higher vs community*
45% of 8-11 year olds have an anxiety disorder[3]	**22%** with elevated anxiety[1]	**32%** with elevated anxiety[1]	**30%** with anxiety[1] *2-3x higher vs community*
	6% with elevations in both[1]	**14%** with elevations in both[1]	
	15x increased risk of comorbid depression in those with anxiety[1]	**16x increased risk** of comorbid depression in those with anxiety[1]	

1. Quittner AL, et al. Thorax 2014;69(12):1090-1097. 2. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 3. Georgiopoulos AM, et al. Pediatr Pulmonol 2020; doi: 10.1002/ppul.24977. Online ahead of print.

Biological risk factors include older age, being female and recent changes in health status

Predictors of elevated depression and anxiety in CF populations:[1]



1. Quittner AL, et al. Thorax 2014;69(12):1090-1097.

There is a clear correlation between older age and poorer quality of life

Quality of life assessment amongst people with CF (aged 16-42 years) showed statistically significant negative correlations between age and:[1]

- **Physical Functioning** (limitations in daily activities)
- **Physical Roles** (difficulty with work/daily activities due to physical health)
- **Bodily Pain** (presence of and limitations due to pain)
- **Vitality** (loss of energy or presence of fatigue)
- **Mental Health** (presence of depressive feelings or nervousness)
- **Social Functioning** (limitations in social activities).

Age-associated disease progression may be one of many contributing factors to the decreased scores in both physical and mental functioning – functional status affects emotional status, and vice versa.[1,2]

1. Uchmanowicz I, et al. Adv Clin Exp Med 2015;24(1):147-52. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 4. Harness-Brumley CL, et al. J Womens Health(Larchmt) 2014;23(12):1012-1020.

Are women more affected by the impacts of CF?

On average, women assess their quality of life more negatively in all subscales compared to men.[1]

Girls experience earlier pulmonary function decline and higher mortality rates.[3,4]

While the reasons remain unclear, it is potentially driven by unfavourable effects of oestrogen on mucosal fluid dynamics, bacterial virulence and the inflammatory response, or by chromosomal differences vs males.[4]

1. Uchmanowicz I, et al. Adv Clin Exp Med 2015;24(1):147-52. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 4. Harness-Brumley CL, et al. J Womens Health(Larchmt) 2014;23(12):1012-1020.

Prevalence rates for depression and anxiety are likely to be underestimated

- Those experiencing the most severe symptoms of depression and anxiety are less likely to attend regular clinic visits or give informed consent for participation in research.[1]
- Many are likely to have subclinical manifestations.
  - Feelings of worry or sadness are common when dealing with a chronic illness.[2]
  - Disorders can develop slowly and have a lifelong course of relapse and remission.[2]

Prevention strategies and promotion of wellbeing are just as critical as screening and treatment.[2]

1. Quittner AL, et al. Thorax 2014;69(12):1090-1097. 2. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78.

Although causality isn't always clear, CF may contribute towards other mental health conditions too

With a comorbidity of CF, the impacts of an underlying mental health disorder can be substantially more significant on both physical health and wellbeing.[1-6] They can also impact CF care and will likely require additional therapeutic strategies.[7]

It is important to continue to improve understanding about other mental health conditions in the context of CF. Full psychosocial screening for those with CF may also be prudent.

Visit the following slides to learn the CF-specific implications of the following disorders.

- **Substance misuse**
- **Sleep disturbances**
- **Eating disorders / Disordered eating**
- **Attention-deficit / hyperactivity disorder (ADHD)**

1. Cystic Fibrosis Foundation. Substance misuse [Internet]. Available at: https://www.cff.org/Life-With-CF/Daily-Life/Emotional-Wellness/Substance-Misuse/ [Accessed July 2022]. 2. Lowery EM, et al. J Cyst Fibros 2020;19(1):84-90. 3. Tomaszek L, et al. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub 2018;162(3):212-218. 4. Shakkottai A. et al. Sleep Med Rev 2018;42:100-110. 5. Linkson L, et al. Paediatr Respir Rev 2017 Mar 23. https://doi.org/10.1016/j.prrv.2017.03.002. 6. Spitzer N, et al. Cureus 2018;10(7):e3048. 7. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78.

Substance misuse

- At least as common in the CF population compared to the general population.[1,2]
- May be a response to anxiety/depression, chronic pain, or self-medication of undiagnosed comorbid conditions.[1,2]
- Young age of initiation of alcohol use is the strongest risk factor.[1,2]
- Increases non-adherence, risk of negative medical outcomes and risk of dangerous interactions with CF medications.[1]
- Doubles severity of anxiety and depression compared to those who don't misuse substances.[1]
- May affect transplant eligibility.[1]
- Young, unmarried males with lower education levels are more susceptible.[2]

Binge drinking occurs in 17% of adolescents and 49% of adults with CF who drink alcohol.[2,3]

1. Cystic Fibrosis Foundation. Substance misuse [Internet]. Available at: https://www.cff.org/Life-With-CF/Daily-Life/Emotional-Wellness/Substance-Misuse/ [Accessed July 2022]. 2. Lowery EM, et al. J Cyst Fibros 2020;19(1):84-90. 3. Weitzman ER, et al. Pediatrics 2015;136(3):450-457.

Sleep disturbances

- Children, adolescents and adults with CF experience chronically worse quality sleep than the general population.[1-3]
- Causes include nocturnal hypoxemia, nocturnal hypercapnia, nocturnal cough, nocturnal enteral feeding, GI problems or medication.[2,3]
- Significantly impairs lung health, pulmonary hypertension, neurocognitive ability, mood, pain, and quality of life.[1-3]
- Daytime drowsiness is the most typical consequence.[1]
- Significantly higher prevalence of anxiety and depression in those with poor sleep.[1,2]
- The risk of insomnia is increased 4.3x in the presence of anxiety symptoms and 5.0x with depressive symptoms.[1]

Approx. 40% of people with CF suffer from disrupted sleep (including frequent awakenings, trouble breathing).[1]

1. Tomaszek L, et al. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub 2018;162(3):212-218. 2. Shakkottai A. et al. Sleep Med Rev 2018;42:100-110. 3. Dancey DR, et al. Eur Respir J 2002;19(3):504-510

Eating disorders/disordered eating

- While formal diagnoses are not more prevalent vs general population, there may be an increased risk of abnormal eating patterns amongst CF adolescents.[1-3]
- Risk factors directly related to CF include preoccupation with disease and dietary management, delayed growth/puberty, body image concerns, low BMI, comorbidities and reduced life expectancy.[1,4]
- Females with CF-related diabetes may be more susceptible due to increased concerns around eating and body image.[1]
- CFTR modulator therapies can result in rapid weight gain once initiated; women with CF may need support to manage body image concerns and changed ideas about eating.[5]
- Extremely low BMI has a far more significant physical health impact on an individual with CF and management can be complicated.[1]
- Eating disorders may affect transplant eligibility.[1]

Subclinical levels of eating disturbances occur in 53% of people with CF vs 40-47% in the general population.[2]

BMI: body mass index.

1. Linkson L, et al. Paediatr Respir Rev 2017 Mar 23. https://doi.org/10.1016/j.prrv.2017.03.002. 2. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 3. Mussche I, et al. Tijdschrift voor Geneeskunde 2014;70(18):1030-1039. 4. Helms SW, et al. J Pediatr Psychol 2017;42(9):1040-1050. 5. Cystic Fibrosis Reproductive and Sexual Health Collaborative. Body Image [Internet]. Available at https://cfreshc.org/SRH-Guide/body-image/ [Accessed July 2022].

ADHD

- ADHD is the most common behavioural disorder in the general paediatric population, characterised by persistent patterns of inattention and/or hyperactivity.[1]
- Diagnosis may not be recognised immediately if behavioural changes are attributed to disease symptoms or complications from treatments.[1]
- Underlying changes in executive functioning and behaviour can impair self-care skills and create challenges for managing and sustaining complicated health regimens, disrupting treatment adherence and negatively affecting prognosis.[1-4]
- Fatigue or sleep disturbances associated with CF, as well as depression and anxiety, can worsen the attentional symptoms of ADHD.[3]

The prevalence of ADHD in CF populations is estimated between 8-10% – at least equal to the general population, but studies suggest prevalence may be as high as 20%.[1-4]

ADHD: attention-deficit/hyperactivity disorder

1. Spitzer N, et al. Cureus 2018;10(7):e3048 2. Georgiopoulos AM, et al. Pediatr Pulmonol 2020; doi: 10.1002/ppul.24977. Online ahead of print. 3. Georgiopoulos AM, et al. J Cyst Fibros 2018;17(2):276-280. 4. Cohen-Cymberknoh M, et al. J Cyst Fibros 2018;17(2):281-285.

Understanding and identifying mental health issues in people with CF at different life stages

Part 3

The factors contributing to mental health challenges in CF are many and varied

As we have discussed briefly, in CF, both physical and mental health are influenced by numerous biological and psychosocial factors as the disease exerts its profound influence on daily life.[1-3]

An individual's emotional wellbeing fluctuates over time, impacted by the severity of their CF, their personality, their social role, their coping style and their medical and social support systems.[1] This can make identifying and treating mental ill-health complex, and emphasises the need for enhanced awareness by the CF care team of the factors affecting psychosocial functioning.[2,3]

To better understand the mental health care needs of the individual with CF, we will now explore some of the psychosocial impacts and challenges that CF poses throughout a patient's lifespan.

We will then complete this module by summarising the impacts of CF on family and caregivers.

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68

Common psychosocial risk factors can impact the health and wellbeing of people living with CF

However, these risk factors are modifiable, so it is important for CF care teams to understand these factors and opportunities for intervention to promote wellbeing and quality of life for people with CF.[1-3]

Visit the following slides to learn about the below risk factors.

- **Poor family functioning**
- **Maladaptive coping**
- **Poor social support / social isolation**
- **Low self-efficacy and self-esteem**
- **Lack of engagement in education, vocation and employment**

Other risk factors to look out for:

- Unstable home environment
- Financial instability
- Substance use
- Poor access to care and resources
- History of mental illness

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68.

Discordant family functioning

- CF is a “family diagnosis” where a large burden of treatment is placed on parents/carers.[1]
- Families may feel the need to be more structured in their interactions, at the expense of more emotional and social aspects of family functioning.[2]
- **The childhood context influences their attitudes towards life and their illness.3 There is a well-established association between a poor family environment and lower treatment adherence.**[2]
- Families with lower socioeconomic status are more at risk of lower levels of family functioning.[2]
- Although the risk for long-term health and wellbeing outcomes decreases in adulthood, CF can continue to disturb family dynamics in both the family of origin and present family.[2,3]

Intervention opportunity: Early family-centred care is important, including identification of significant psychosocial and mental health needs, aiming to enhance family cohesion and resilience over time.

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

Lack of engagement in education, vocation and employment

- Health-related quality of life is associated with work status.[1]
- **People with CF who are working experience better health perception, role perception, physical functioning, social functioning and disease mastery, and lower depression scores.**[1]
- However, reconciling work with CF can be difficult; patients may delay or avoid treatments rather than disrupt professional duties.[1]
- Employment is also closely linked to financial issues – sick leave or the need to work part-time hours can negatively affect income and make financial independence difficult.[1]
- Poor educational attainment also impacts on the ability of people with CF to secure employment prospects independent of their disease severity.[1]

Intervention opportunity: Educational and vocational counselling should be developed in both pediatric and adult CF centres, as well as vocational rehabilitation guidance for those failing to maintain employment.[1]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

Low self-efficacy and self-esteem

- Self-efficacy is an individual's belief in their own ability to accomplish a task – relevant to CF considering the rigorous, daily and time-consuming aspects of CF care.[1]
  - This is true for both patients and carers.
- Self-efficacy is highly correlated with self-management and the performance of tasks required to maintain health in CF, as well as better physical, social and emotional quality of life.[1,2]
- **Low self-efficacy is linked directly to poor adherence and health behaviours, and ineffective pain management and chronic disease management.**[1]

Intervention opportunity: Promote confidence, motivation and problem-solving to address barriers to help maintain or improve self-efficacy and thus self-management.[1]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

Poor social support/social isolation

- Friendships and social relationships significantly contribute to wellbeing and help people with CF feel they are leading a normal life.[1]
- **Greater levels of social support are associated with fewer depressive symptoms, higher self-efficacy, greater adherence and better overall quality of life.**[2]
- Social support, connectedness and validation of experience is known to be beneficial across many illness populations, but this is severely reduced in CF due to infection control guidelines that limit in-person support and connection with others with the disease.[2]
- Newer technologies, such as internet-based forums and conferencing, apps and social media, could prove very effective in overcoming this.[2]

Intervention opportunity: Encourage appropriate methods to engage social support from friends, or others with a perceived ability to understand their experience.[2]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

Maladaptive coping

- Coping includes a range of emotional regulating strategies, thought processes and behaviours aimed at reducing disease-related distress and maintaining subjective wellbeing.[1]
- **Passive or avoidant coping strategies (e.g. disengagement, self-distraction, substance use) increase the risk of mental health symptoms, lower quality of life and are associated with worse treatment compliance.**[1-4]
- In contrast, active coping styles (e.g. problem solving, acceptance, seeking social support) predict better quality of life and improved self-management.[1-4]
- Active coping styles are associated with hope, optimism and ability to relieve the risk for depression and anxiety.[1-3]

Intervention opportunity: Support development of effective coping skills appropriate to the developmental age.[1,2]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 4. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68.

Developmental risk factors arise from unique psychosocial challenges at each life stage

As a person with CF progresses through life, the intersection of developmental processes, psychosocial variables and health outcomes evolves, as the focus on self-management and adherence increases.[1]

In the following slides, we will consider five broad life stages that mark key changes in psychosocial needs and challenges.

1. **Perinatal**
2. **Early childhood**
3. **Primary school age**
4. **Adolescence**
5. **Adulthood**

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283.

Perinatal: A critical time for the mental health of parents and carers

DEVELOPMENTAL CONTEXT

CF is increasingly being diagnosed prenatally and during the newborn phase, with most diagnosed by 2yo.[1,2] The parent-child relationship is established during this stage.[1] Quality of attachment predicts emotional and behavioural regulation, social skills and coping abilities.[1]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Parental distress throughout the diagnostic process

- **Parental distress throughout the diagnostic process** can make them vulnerable to depressive symptoms and have negative implications for infant/parent attachment.[1,2]
- Poor infant/parent attachment can result in significantly poorer nutritional status and lower BMI.[1]
  - However, CF itself does not increase the risk of maladaptive infant/parent attachment.[1]

Family interactions

- **Family interactions** may start being influenced by the burden of CF.[1]
  - Compared to healthy dyads, children 12-24 months with CF exhibit more whining and less responsivity to parents, while parents display more controlling, serious and less encouraging behaviours.[1]

The effects on parents and carers will be explored further in the next section.

BMI: body mass index

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92

Early childhood: Increased burden on family dynamics can impact mental health

DEVELOPMENTAL CONTEXT

Development of language skills results in a better understanding of the child's cognitive and emotional experience.[1] Many children understand they were born with their illness but are still developing understanding of what can and cannot be controlled.[1]

Parents must meanwhile promote normal childhood development in the context of CF.[1,2]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Behavioural feeding challenges

Medical milestones

Increased opportunities for parent/child conflict

- **Behavioural feeding challenges** are salient – focus on nutritional health can worsen the usual challenges of childhood eating.[1,2]
  - Children with CF display higher frequency of behaviours interfering with eating, while parents display increased coercion, commands, physical prompts and actual feeding.[1]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Behavioural feeding challenges

Medical milestones

Increased opportunities for parent/child conflict

- **Medical milestones** (e.g., initiation of new therapies, initial onset of CF-related infection, first hospitalizations) can be stressful.[2]
  - Procedural anxiety is experienced by 40-60% of children and can have longer-term behavioural and psychological impacts.[3]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Cystic Fibrosis Western Australia. Fact Sheet: Procedural Anxiety [Internet]. Available from: https://www.cfwa.org.au/wp-content/uploads/2018/02/CF-Fact-Procedural-Anxiety.pdf [accessed July 2022]

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Parents must meanwhile promote normal childhood development in the context of CF.[1,2]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Behavioural feeding challenges

Medical milestones

Increased opportunities for parent/child conflict

- **Increased opportunities for parent/child conflict** can lead to more authoritative or permissive parenting strategies, which negatively impact mental health and family functioning.[1]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

Primary school age: Experiencing the 'second diagnosis' and its impact on mental health

DEVELOPMENTAL CONTEXT

Cognitive and communication skills are evolving and there is increased emphasis on peer relationships as children develop their sense of identity and competence.[1]

Treatment burden can start to increase, interfering with school and peer-related demands.[1] The child's sense of ownership and control should be encouraged to develop self-efficacy.[1]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Comparison with peers

Increased feelings of loneliness

Self-perception of vulnerability

- **Comparison with peers** leads to a realisation of their disease on a deeper lever, as significant concerns about 'being different' emerge.[1]
  - Coping with negative peer reactions to the visible characteristics of CF (e.g. coughing, taking enzymes, fatigue) is considered one of the most stressful daily events.[1]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- **Comparison with peers**
- **Increased feelings of loneliness**
- **Self-perception of vulnerability**

- **Increased feelings of loneliness** can develop due to frequent disease-related absences that may make it more difficult to establish friendships.[2]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

Comparison with peers

Increased feelings of loneliness

Self-perception of vulnerability

- **Self-perception of vulnerability** may arise through parental behaviour coupled with illness uncertainty.[1]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92.

Adolescence: A time of immense change, complexity and increased risk of mental health issues

DEVELOPMENTAL CONTEXT

This is a period of rapid social, cognitive and physiological changes, with increasing autonomy and separation from parents as peers become more influential.[1-3]

It is also a time of disease progression, with more frequent symptoms and exacerbations.[1-3] Increased treatment burden coupled with the turmoil of adolescence makes treatment adherence more challenging.[1-3]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- Distress about body image
- Social Isolation
- Concerns about the future
- Family tension
- Risk taking behaviours

- **Distress about body image** given pubertal delay, GI symptoms, and other physical symptoms can feel isolating; sexual exploration adds another layer of complexity.[1,3]

GI: gastro-intestinal

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Withers AL. Pulm Med 2012;2012:134132.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- Distress about body image
- Social Isolation
- Concerns about the future
- Family tension
- Risk taking behaviours

- **Social isolation** is common, with those with CF having fewer friends and less likely to develop romantic relationships.[1-3]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Withers AL. Pulm Med 2012;2012:134132.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- **Distress about body image**
- **Social isolation**
- **Concerns about the future**
- **Family tension**
- **Risk taking behaviours**

- **Concerns about the future** (e.g., finishing school, having a job, sexual relationships, starting a family) may increase with a heightened sense of mortality.[1,3]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Withers AL. Pulm Med 2012;2012:134132.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- Distress about body image
- Social isolation
- Concerns about the future
- Family tension
- Risk taking behaviours

- **Family tension** is characteristic in adolescence; unsupportive family behaviours are the strongest predictor for poor psychological adjustment.[1-3]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Withers AL. Pulm Med 2012;2012:134132.

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- **Distress about body image**
- **Social isolation**
- **Concerns about the future**
- **Family tension**
- **Risk-taking behaviours**

- **Risk-taking behaviours** can occur in a bid to gain acceptance from peers, minimise differences and as a distraction strategy.[3]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Withers AL. Pulm Med 2012;2012:134132.

Early Adulthood and Beyond: Fostering meaningful independence and nurturing mental health

DEVELOPMENTAL CONTEXT

Adult milestones such as educational/vocational achievement and family planning can be fulfilling and meaningful.[1] However, they can also add to the psychosocial burden of CF, as disease acceleration and end-of-life decision making may also be faced.[1]

COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- Increased frustration and discomfort
- Feelings of powerlessness
- Distress related to employment
- Financial concerns
- Parenthood

- **Increased frustration and discomfort** at their medical and social situation is common; self-esteem can also decline.[2]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- **Increased frustration and discomfort**
- **Feelings of powerlessness**
- **Distress related to employment**
- **Financial concerns**
- **Parenthood**

- **Feelings of powerlessness** can occur due to significant disease acceleration.[2]
  - CF-related diabetes can begin in late adolescence and the burden of an additional chronic disease can be overwhelming.[2]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283.

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- **Increased frustration and discomfort**
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- **Distress related to employment**
- **Financial concerns**
- **Parenthood**

- People with CF may experience **distress related to employment,** such as limited opportunities and fear about disclosing their disease to potential employers.[2,3]
  - Those with CF are less likely to be employed and more likely to remain in the parental home in adulthood (especially men).[2,3]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Withers AL. Pulm Med 2012;2012:134132.

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COMMON MENTAL HEALTH PSYCHOSOCIAL CHALLENGES

- Increased frustration and discomfort
- Feelings of powerlessness
- Distress related to employment
- Financial concerns
- Parenthood

- **Financial concerns** can be an issue, stemming from employment concerns.[2,3]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Withers AL. Pulm Med 2012;2012:134132.

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- **Increased frustration and discomfort**
- **Feelings of powerlessness**
- **Distress related to employment**
- **Financial concerns**
- **Parenthood**

- **Parenthood** is pursued by an increasing number of people with CF, but comes with practical, emotional and ethical considerations that can be stressful.[2,3,4]
  - One study in adults with CF in the US found that 68.6% of participants would like to have children in the future.[4]
  - Key concerns are the hereditary nature of CF and the high possibility of illness or death during their offspring's childhood.[2,3]

1. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Withers AL. Pulm Med 2012;2012:134132. 4. Kushary S, et al. Reprod Biomed Soc Online 2021;13:37-45

Other important themes related to psychosocial wellbeing

Growing older with CF means coping with new diagnoses, more medications and other healthcare-related issues, with associated psychosocial implications.[1]

Some of they key themes that become relevant as individuals with CF move from adolescents into adulthood are addressed on the following screens.

1. **Treatment adherence, difficulty sustaining daily care**
2. **Reproductive and sexual health**
3. **Lung transplantation**
4. **Relationships with healthcare providers**

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

Balancing normal life with adherence to increasing treatments is challenging

Adherence to treatment fluctuates over time, with periods when treatment is managed easily and others when it is more difficult.[1]

Important factors affecting adherence[5]

- Personality
- Age
- Gender
- Illness severity
- Knowledge
- Perceptions
- Emotions
- Attitudes & beliefs
- Socioeconomic issues

Facilitators of treatment adherence[5]

- Social support
- Community support
- Organisational strategies
- Intrinsic characteristics
- Combining treatments with pleasurable activity
- Flexibility
- Easier or faster treatments
- Prioritising treatments
- Negative effects of non-adherence.

While some level of non-adherence is normal at all life stages, adolescence is the period of lowest treatment adherence.[2-4]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 3. Withers AL. Pulm Med 2012;2012:134132.
4. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 5. Nicolais CJ, et al. J Clin Psychol Med Settings 2019;26(4):530-540.

Why is adolescence associated with the period of lowest treatment adherence?

Up to 50% of those CF have shown significant non-adherence with at least one aspect of management, usually during the adolescent period.[1-3]

Adolescence is a time of great change when adolescents are developing their identity and beliefs and pursuing independence from parents and caregivers, often with substantial influence from their peers and social environments.[4] These normal developmental milestones may contribute to or be perceived as psychosocial resistance that can impact negatively on treatment adherence in adolescents with CF.

Reasons for non-adherence:[1-3]

Inadequate knowledge	Educated non-adherence	Procedural anxiety
Underestimate severity of disease, poor knowledge about treatment, misinterpretation of treatment details	Difficulty with time management, desire to balance demands of CF treatment with goals for normal life (work, relationships, family)	Less parental supervision, family discord, oppositional behaviours, wish to conform to peers, rebellion, feelings of futility, avoidance coping, disagreeing with the physician

1.Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Withers AL. Pulm Med 2012;2012:134132. 3. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 4. Patton GC, et al. Lancet 2016; 387(10036):2423-2478.

Difficulty sustaining daily care can be attributed to multiple reasons[1-4]

Since adherence to treatment is closely related to prognosis, identifying strategies to address difficulties is important.

- Reasons for non-adherence are mostly treatment-specific, so tailoring treatments to lifestyle can help.[3,4]
- Drawing attention to the immediate and obvious benefits of treatment is also useful, particularly for adolescents.[3,4]
- Interventions that focus on an individual's strengths, build on prior success and/or use models successfully used by others are more likely to be efficacious.[5]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 3. Withers AL. Pulm Med 2012;2012:134132.
4. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 5. Nicolais CJ, et al. J Clin Psychol Med Settings 2019;26(4):530-540.

Reproductive and sexual health is a notable unmet care need for adolescents and adults with CF

Reproductive health includes providing advice about contraception, pubertal delay, fertility status, avoiding sexually transmitted infections, genetic counselling and the effects of pregnancy and parenting on health.[1]

Early knowledge of reproductive and sexual health is important because it may affect reproductive decision making and can help prevent CF-specific health risks resulting from unsafe sexual activity.[1,2]

However, knowledge is often poor in adolescents and education provided is inadequate,[1] which may impact on overall psychological wellbeing.

<50% of adult men learnt of their **probable infertility** from their preferred source, and substantially later than preferred.[2]

Only 50% of **parents** know about male infertility and only 30% feel confident discussing it.[1]

20% confused infertility with **impotence.**[1,2]

Girls can be confused about **potential for pregnancy.**[1,2]

Genetic and psychological counselling about parenthood should be offered to both men and women.[1,2]

1. Withers AL. Pulm Med 2012;2012:134132. 2. Frayman KB, Sawyer SM. Lancet Respir Med 2015;3(1):70-86

The prospect of lung transplantation can be highly distressing

Lung transplantation (LTx) is the most aggressive treatment for end-stage lung disease.[1]

Despite improving post-transplant outcomes, individuals with CF may consider lung transplantation as 'a death sentence' or have other negative connotations.[2,3]

- Between 2005 and 2016, the 1-, 3-, and 5-year survival rates of patients who underwent lung transplantation for CF in Canada were 90.5%, 79.9%, and 69.7%, respectively.[4]
- Discussions regarding LTx should be initiated when FEV1 is <50% predicted.[2]

Routine discussion of lung transplantation can help to normalise it and reduce fear and anxiety.[2]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Ramos KJ, et al. J Cyst Fibrosis 2019;18(3):321-333. 3. Withers AL. Pulm Med 2012;2012:134132. 4. Stephenson A, et al. J Heart Lung Tranplant 2021 Mar;40(3):201-209

The prospect of lung transplantation can be highly distressing (cont.)

Numerous psychosocial and mental health concerns can emerge:

- Some patients may perceive LTx as a failure of their own adherence behaviours and subsequently feel shame or stigma – this is particularly true for those with a mindset of "beating" CF through vigilant adherence behaviours.[2]
- They may be concerned about increased complexity of care, potential decentralization of care and worsening QoL.[2]
- They may also feel concerns about the impact on family planning, relationships and their education/career.[2]
- Some patients may feel distress that someone else has had to die for them to live.[3,4]
- Referral for LTx doesn't always mean they will receive a transplant:
  - 25% of CF patients will die while on the transplant waiting list.[1,3]
  - Significant distress can also occur if they are found to be **ineligible for a transplant**.[4]

It is important to identify and address negative self-directed emotions and provide psychosocial support throughout the LTx process.[2]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Ramos KJ, et al. J Cyst Fibrosis 2019;18(3):321-333. 3. Withers AL. Pulm Med 2012;2012:134132. 4. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68.

Psychosocial and mental wellbeing are key components of transplant eligibility

In a 2019 set of consensus guidelines, the Cystic Fibrosis Foundation identified that the following can be barriers to lung transplantation:

- Substance use (as this has been associated with worse post-transplant outcomes)
- Non-adherence (many transplant centres consider non-adherence to medical care a relative, and sometimes absolute, contraindication to transplant)
- Anxiety and depression can impair recovery from transplant (these issues may negatively impact transplant candidacy)
- Other psychosocial barriers such as: health literacy, finances, (lack of) caregiver support

These barriers are all modifiable – it may take years of work in order to optimise a candidate for transplantation, so it is critical to identify and address any issues pre-emptively.

Be aware that uncertainty about eligibility may serve to increase ambivalence and ultimately lead to avoidance of transplant-related knowledge or health decisions.

Ramos KJ, et al. J Cyst Fibrosis 2019;18(3):321-333.

The interactions between healthcare team members and the person with CF strongly influence their wellbeing

The therapeutic relationship between individuals with CF and members of their healthcare team is an important part of the CF treatment process; both sides are involved in actively thinking about, discussing and participating in the process – or by contrast, neglecting it.

Healthcare teams need to be ready to adapt to the way the person with CF views their care plan. Some patients may need time to accept and adapt to changes in health status, or feel uncertain and anxious about new treatments.

Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015.

The interactions between healthcare team members and the person with CF strongly influence their wellbeing

Healthcare teams should also be aware that **the way they prioritise and define the aims of interventions will influence how the person perceives their care.**[1] Below are some examples of this, relating to the key themes we just explored:

Non-adherence can result when members of the healthcare team don't appear to be responsive to the patient's beliefs or concerns.[2]

An empathetic, non-critical approach that involves listening and some compromise is helpful.[2,3]

Some health professionals can find it embarrassing or have trouble finding time to discuss reproductive or sexual health, but many patients would welcome a physician-initiated discussion.[3,4]

The onus should be on the health professional to create these opportunities for patients, especially during adolescence.[3,4]

A patient's feelings towards transplantation are strongly influenced by the ways it is discussed by their healthcare team.[5]

Patients benefit from discussing lung transplantation early, and having it framed it as an option for extending life for end-stage CF, rather than the 'beginning of the end.'[5]

1. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 2. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 3. Withers AL. Pulm Med 2012;2012:134132.
4. Frayman KB, Sawyer SM. Lancet Respir Med 2015;3(1):70-86. 5. Ramos KJ, et al. J Cyst Fibrosis 2019;18(3):321-333.

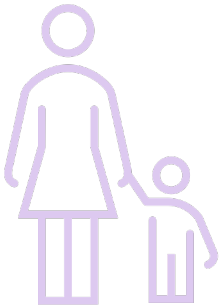
Effects of mental health conditions in CF on carers

Part 4

CF Parent caregivers experience higher psychological distress

Caregivers are central in the care of individuals with CF throughout the lifespan.[1] **While caregiving brings countless rewards, it also bears substantial emotional and existential tolls**.[1]

Concerns can include worries about their loved one(s) with CF, feeling overwhelmed, distress about medical decision making, inability to attend to their own needs and other responsibilities, and strained relationships.[1] Disease progression can also amplify caregiver burdens and needs.[1]



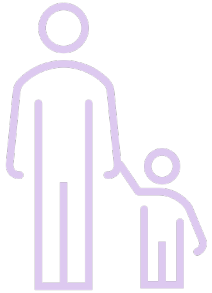
MOTHERS

37% with elevated depression[2]

48% with elevated anxiety[2]

31% with elevations in both[2]

16x increased risk of comorbid depression in those with anxiety[2]



FATHERS

31% with elevated depression[2]

36% with elevated anxiety[2]

21% with elevations in both[2]

9x increased risk of comorbid depression in those with anxiety[2]

Depression is 3x higher and **anxiety is 2-3x higher** in CF caregivers compared with community samples.[2]

1. Kavalieratos D, et al. J Palliat Care 2020 doi: 10.1089/jpm.2020.0311. 2. Quittner AL, et al. Thorax 2014;69(12):1090-1097

There are also significant associations between parental and adolescent mental health issues

Analyses of parent-adolescent dyads showed **adolescents were 2x more likely to be above the cut-off for depression or anxiety if at least one of their parents had elevated symptoms.**[1]

Parental psychological distress can be a cue for screening adolescents with CF for mental health disorders.

Quittner AL, et al. Thorax 2014;69(12):1090-1097

1 in 4 CF parent caregivers suffer from post-traumatic stress symptoms (PTSS)

- **Parents of children with CF are exposed to various potentially traumatic events**, such as:
  - Witnessing their child undergo painful procedures and recurrent hospitalisation
  - Lung transplantation and intensive post-transplant medical care.
- **PTSS is associated with functional impairment and can compromise the caregiver role.**
  - Hypervigilance, re-experience and arousal are the most frequent symptoms in CF caregivers.
  - Mothers may be more at risk than fathers.
  - PTSS is not related to objective disease severity – it is driven by the subjective perception of risk.

While parents acknowledge their problems and need for treatment, they are underdiagnosed and undertreated.
May believe it is a normal reaction to their situation, or prioritise their child's health over their own.

Cabizuca M, et al. Health Psychol 2009;28(3):379-388.

Parental depression and anxiety are linked to poorer adherence and health outcomes for the child[1,2]

With family functioning being a psychosocial risk factor for CF, there is a **clear rationale to ensure timely identification and treatment of psychological distress in parent caregivers.**

Guidelines recommend:[3]

- Preventative psychoeducational support
- Annual screening in at least one caregiver of children <18yo
- Clinical assessment/diagnosis following a positive screen
- Evidence-based psychological and pharmacological intervention when indicated (undertaken in primary care or mental health services following referral from the CF team).

1. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Quittner AL, et al. Thorax 2016;71(1):26-34.

What are the specific challenges and triggers for parents at each life stage?

Caregiving roles change over time, with both the degree and types of physical and emotional support dependent on the changing health status of their child with CF.

Distress begins during newborn screening and diagnosis[1,2]

Balancing the needs of the whole family can be challenging[1-3]

Transition from paediatric to adult care can be difficult for parents too[2,4]

Support is indicated during end-of-life care and upon death of their loved one[5-7]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 4. Leeman J, et al. J Fam Theory Rev 2015;7(2):167-183. 5. Withers AL. Pulm Med 2012;2012:134132. 6. Kavalieratos D, et al. J Palliat Med 2021;24(1):18-30. 7. Dellon EP, et al. J Cyst Fibros 2018;17(3):416-421.

Distress begins during newborn screening and diagnosis

Distress is particularly high while waiting for confirmatory sweat testing following positive screening results.[1,2]

- Depressive symptoms, anxious symptoms, hypervigilance and guilt are common.[1]
- Poor knowledge of the newborn screening process and CF in general leads to worse coping.[1]

Distress can also be amplified by healthcare providers.[1]

- Impersonal communication of results can be upsetting.[1]
- Primary care paediatricians may have low confidence and a lack of CF-specific training to appropriately manage consultations.[2]

Current quality improvement focuses on reducing parental and provider anxiety, by streamlining referrals to CF centres and minimising time from screening to sweat test.[2]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92

Balancing the needs of the whole family can be challenging

Families with a child with CF can have poorer adjustment, particularly in areas of family communication, emotional expression, interpersonal involvement and appropriate behavioural expectations.[1]

- Parents of pre-schoolers with CF report greater marital strain or breakdown, decreased home recreation time and elevated levels of depression, anxiety and stress.[1]
- Worsening health status (e.g. first hospital admission) can cause anxiety and raise unresolved grief about the diagnosis.[2]

Siblings can also be negatively impacted as CF dominates daily life[1,3]

- Differential treatment between siblings is common, with mothers spending more time with the sibling with CF.[1]
- Anger, resentment and other negative feelings can affect the quality of the sibling relationships, with long-term social implications for both children.[1,3]

Promoting effective parenting strategies (e.g. coping, social support) can help redistribute resources to self-care, marital health and maximising quality of time spent with all children.[1]

1. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68.

Transition from paediatric to adult care can be difficult for parents too

Parents can perceive the transfer of responsibility for care to their child as positive.[1]

- Transfer promotes a more mature identity as well as social and financial independence for their child.[1]
- Parents appreciate the respite from the demands, intensity and stress of the parental role in enforcing therapy.[1]

However, conflict and tension can arise over their child's self-management.[1,2]

- The child's greater independence in deciding whether to perform treatments generally leads to lower adherence.[1]
- Parents may find it difficult to 'let go' and change their role from primary carer to advocate.[2]

Information and preparation of caregivers is also important to promote successful transition to adult healthcare.[3]

1. Leeman J, et al. J Fam Theory Rev 2015;7(2):167-183. 2. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 3. Coyne I, et al. J Child Health Care 2018;22(4):646-657.

Support is indicated during end-of-life care and after death of a loved one

Although life expectancy for people with CF has increased, it is still a serious terminal condition.[1]

Care and comfort for a patient's family is an important part of end-of-life care.[1-3]

On the transplant list

End-of-life care

Death of a loved one

For carers whose loved one with CF is waiting for a transplant:

- Some candidates consider the waiting period to be the most difficult part of the transplantation process.[4]
  - Prior to being called for transplant, it is perfectly normal for candidates to experience fear, anxiety, and uncertainty.[4]
  - Throughout the transplantation process, help is available to assist candidates in coping with stress, managing pain, and other concerns.[4]
- It is impossible to know when a candidate will receive the call for transplant surgery, so candidates must always be ready, and a support person must always know how to reach the candidate.[4]

1. Withers AL. Pulm Med 2012;2012:134132. 2. Kavalieratos D, et al. J Palliat Med 2021;24(1):18-30. 3. Dellon EP, et al. J Cyst Fibros 2018;17(3):416-421. 4. Cystic Fibrosis Canada. Lung Transplantation and Cystic Fibrosis. Available at: https://www.cysticfibrosis.ca/uploads/intro%20to%20treatment/Lung_Transplants_WEB_Compressed.pdf [Accessed July 2022].

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End-of-life care

Death of a loved one

The Cystic Fibrosis Foundation palliative care delivery guidelines note that as individuals with CF approach the final stages of their lives, the CF care team remains critically important in providing comfort-focused care to alleviate suffering for patients and families.[2] Recommendations include:

- CF care teams take a collaborative approach in offering comprehensive, timely, and compassionate end-of-life care.[2]
- CF care teams should consider early engagement of experts in collaborative management of end-of-life concerns, including palliative care specialists, chaplains, and social workers.[2]

1. Withers AL. Pulm Med 2012;2012:134132. 2. Kavalieratos D, et al. J Palliat Med 2021;24(1):18-30. 3. Dellon EP, et al. J Cyst Fibros 2018;17(3):416-421.

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End-of-life care

Death of a loved one

The death of a loved one is a distressing event for parents, siblings and extended family, as well as for other families with a child with CF.[1]

Following death, ongoing contact by the CF team can be helpful to ensure specific needs of the family are being met and to facilitate the provision of ongoing bereavement support.[2,3]

1. Withers AL. Pulm Med 2012;2012:134132. 2. Kavalieratos D, et al. J Palliat Med 2021;24(1):18-30. 3. Dellon EP, et al. J Cyst Fibros 2018;17(3):416-421.

In summary

- An increased life expectancy is accompanied by increasingly complex disease management for people with CF, contributing to an increased vulnerability to mental health issues than in the past.[1-3]
- Psychological symptoms have an established negative effect on health outcomes and prognosis.[1,4]
- Individuals with CF, as well as their caregivers, are at a higher risk of anxiety and depression, likely associated with the chronic and challenging nature of the disease.[4,5]
- A range of biopsychosocial factors and challenges affect individuals with CF, as well as their caregivers, across life stages – care needs to adapt accordingly.[6-10]
- Older age correlates with poorer QoL, with those with CF experiencing increasingly difficult health and psychosocial issues as they progress through adulthood[6-11]

QoL: quality of life

1. Smith BA, et al. Pediatr Pulmonol 2016;51(S44):S71-S78. 2. Havermans T, Staab D. Thorax 2016;71(1):1-2. 3. Balfour-Lynn IM, King JA. Paediatr Respir Rev 2020 doi: 10.1016/j.prrv.2020.05.002. 4. Quittner AL, et al. Thorax 2016;71(1):26-34. 5. Quittner AL, et al. Thorax 2014;69(12):1090-1097. 6. Hubert D, Simmonds N. Living Longer with Cystic Fibrosis. Denmark: European Cystic Fibrosis Society; 2015. 7. Muther EF, et al. Pediatr Pulmonol 2018;53(S3):S86-S92. 8. Ernst MM, et al. Child Adolesc Psychiatr Clin N Am 2010;19(2):263-283. 9. Pfeffer PE, et al. J Cyst Fibros 2003;2(2):61-68. 10. Withers AL. Pulm Med 2012;2012:134132. 11. Uchmanowicz I, et al. Adv Clin Exp Med 2015;24(1):147-52.