

Guidelines for UK clinical psychology services in cystic fibrosis

July 2024

The UK Cystic Fibrosis Trust Clinical Psychologists Guidelines Working Group

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Acknowledgements

Thank you also to the following for valued input to these guidelines.

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Executive summary: The work of the UK CF clinical psychologist (CF CP)

UK Standards of Care require cystic fibrosis (CF) services to have CF clinical psychologists (CF CPs)* as core members of the CF multidisciplinary team (MDT).¹ Other guidance^{2,3} has outlined key roles to be provided by the CF CP, but the UK CF CP guidelines provide the first comprehensive recommendations for the work of clinical psychologists in UK CF care.

The roles of the CF CP within CF care are multiple, and their unique position in the team may be best understood as formulation driven (rather than diagnostic), within the biopsychosocial model. This approach and the wide range of services offered by the CF CP in both paediatric and adult services are described in fuller detail in the following guidelines. This short summary gives an overview of these multiple roles and may be particularly useful for MDTs and CF service leads. The core roles that it is recommended a CF CP should provide, if services are sufficiently staffed and integrated with the CF MDT, are listed throughout the main guidelines document. They are also listed separately in a self-assessment/audit tool for CF CPs to use, found in Appendix 1 of the guidelines.

Further information for people with CF about CF CP services is also available. There is also separate information about the qualifications and governance of clinical psychologists, and about employing clinical psychologists in CF teams, including recruitment, retention, and resources required to deliver a CF CP service.

The CF CP will take a holistic, preventative and proactive approach to helping people with CF achieve their best possible quality of life and health outcomes. CF CPs help people optimise their emotional and mental health, their physical health, and their management of CF in an individualised way. They will take into account the multiple 'internal' factors impacting on health and wellbeing (for example, personal strengths, relationship with CF, coping styles, health beliefs and other cognitions), but also the 'external' factors that significantly influence outcomes (for example, family systems, cultural factors, level of social support, health inequalities, trauma and other adversity).

CF CPs can work directly with people with CF and their caregivers, but can also work indirectly to improve outcomes. Through indirect work, the CF CP can support the work of the CF team in various ways. This is to ensure that psychological care is embedded within the CF service, and that CF care by the whole team is 'psychologically informed'. The CF CP can also lead on how psychological care is delivered in CF services, supporting service improvement work and the development of holistic CF care.

The key roles of the CF CP can be seen as being delivered in several different ways, and across a range of areas of health, both psychological and physical. The following charts provide a summary of both and may be particularly useful for CF MDTs, for example to help them evaluate if they are able to access the full range of services typically provided by CF CPs.

Key approaches and areas of work of the CF clinical psychologist



Annual assessment and other screening, for example through:

- Annual psychological screening covering a range of emotional health and CF management issues
- Additional screening/monitoring of people with CF at times of potential vulnerability or key changes in CF including, for example, at diagnosis, CFTR modulator introduction or cessation, transition to adult care, pregnancy, or transplantation.
- Monitoring the emotional wellbeing of caregivers in paediatric care.
- Monitoring of wellbeing of people with CF and caregivers indirectly, for example through attendance at MDT meetings and clinics.



Direct work with people with CF and caregivers – working in inpatient, clinic and other CF care settings, face to face or virtually, for:

- Engagement work
- Assessment
- Formulation
- Delivery of a wide range of therapeutic /psychological interventions.
- Evaluation of psychological interventions delivered.
- Assessing and helping to manage risk and safeguarding issues.



Indirect work – supporting psychologically informed care by the wider CF MDT, for example through:

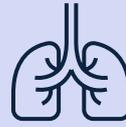
- Supporting the MDT to provide support to people with CF and their families.
- Joined-up working with CF MDT colleagues.
- Consultation and advice to CF team on psychological issues and care.
- Teaching and training on psychological issues.
- Supervision and reflective practice for MDT staff.
- Gatekeeping to, and liaison with, external services, for example specialist mental health provision.
- Delivery of integrated psychosocial care in conjunction with CF social workers.
- Helping the CF MDT practice good self-care.
- Psychological input to CF service improvements, quality initiatives, strategy and service planning.
- Research and audit.

Through a combination of screening, direct work and indirect work as above, the CF clinical psychologist can work with people with CF, caregivers and CF teams to provide psychological care and interventions in the following areas:



Optimising emotional wellbeing and mental health

- Taking a preventative and early intervention approach that recognises individual strengths and coping styles.
- Providing screening, assessment and psychological intervention for a wide range of emotional health issues.
- Recognising wider influences on emotional health, including diversity and health inequalities or other adversity.



Managing challenges with CF and CF care throughout the lifespan

Diagnosis at any age; issues around CFTR modulators; infection control; procedural distress; CF admissions; nutritional/eating/GI issues; CF diabetes; complications of CF; medical trauma; invasive treatments; difficult symptoms; additional needs and CF; lung transplantation; advanced CF and end-of-life care.



Managing change throughout the lifespan with CF

- CF and change in childhood, developing understanding of CF, school issues.
- Adolescence and transition to adult life and adult CF care.
- Adulthood and growing older with CF.



Managing adherence to CF treatment regimes and behaviour change

- Understanding psychological aspects of treatment adherence and behaviour change.
- Psychologically based interventions to address issues with adherence and behaviour change.
- Supporting the CF team to work effectively on adherence. issues.



CF and relationships

- Developing a healthy relationship with CF and self-identity.
- Relationships between the CF team and people with CF and caregivers.
- Parenting a child with CF.
- Relationships for children and young people with CF.
- Relationships for adults with CF, reproductive issues, and parenting as an adult with CF.

* It should be noted that certain other practitioner psychologists, for example counselling psychologists, may be employed by the NHS to provide CF psychology services if they meet the person specification for a CF psychologist post and can deliver the full range of roles required in the job description.

1. Foreword

1.1 Document development

These guidelines for cystic fibrosis clinical psychology care in the UK were written by the members of the Cystic Fibrosis Clinical Psychology Guidelines Working Group between 2019 and 2023. All Working Group authors were members of the UK Psychosocial Professionals in Cystic Fibrosis Group (UKPPCF). A further group of UKPPCF CF clinical psychology members have also contributed to the content of the guidelines and have reviewed drafts.

These guidelines have not been produced using a formal consensus methodology but reflect general agreement by the professionals involved.

Production of these guidelines was commissioned and supported by Cystic Fibrosis Trust, and the development process by the Trust has also included review by relevant CF experts from across the CF multidisciplinary team (MDT) professions (through their Clinical Advisory Group (CAG)), people with CF, and parents of those with CF.

The cystic fibrosis clinical psychology guidelines have been developed alongside cystic fibrosis social work (CF SW) guidelines, and together these documents constitute guidelines for specialist psychosocial care in CF.

1.2 How to use this document

This document is developed as a comprehensive support document for clinical psychologists working with infants, children, young people and adults with CF in England, Wales, Northern Ireland, and Scotland. **It should be useful for:**

- clinical psychologists in CF services, especially those starting work in the field of CF
- other multidisciplinary professionals in CF teams, including CF service leads.
- providers of CF services (NHS trusts)
- commissioners of NHS CF services
- people with CF, their families and carers, and the public.

This new detailed guidance may be used for: the induction of new clinical psychologists to CF posts; planning and benchmarking current care; addressing any gaps in current CF clinical psychology provision; and shaping future CF clinical psychology services. A summary of key CF CP roles and responsibilities is provided in section

3 of this document. Further summary information for commissioners of CF services and CF service directors is available separately and gives more information on employing a clinical psychologist, governance issues, and what infrastructure is required to carry out the CF CP roles (see Summary information for commissioners of CF services and CF clinical directors). That information should be used alongside these guidelines.

A summary of frequently asked questions about CF clinical psychology services for people with CF and their families is also available separately (see Information for people with CF and their families/carers). Both documents are available at cysticfibrosis.org.uk/consensusdocuments

There is a lack of peer-reviewed research in many of the areas covered by these guidelines, and so many of the recommendations for clinical practice are taken from the extensive and specialised experience of the authors and other named contributors. The Working Group consider this to provide best practice guidelines while retaining scope for variation in practice where appropriate, for example due to local service delivery arrangements and innovations in practice in particular services. The Working Group recognises that CF CP practice may differ in some details between different CF centres and between paediatric and adult services.

These guidelines should be used alongside other existing guidance, available in/relevant to some or all of the countries of the UK, including the following:

- UK standards of care for CF¹
- CF social worker guidelines for care in the UK (in production) standards of care and guidance for good practice by other disciplines in CF MDTs^{4,5,6,7}
- for those providing services in England: NHSE service specifications for CF care of children and adults^{8,9}
- any current guidance available from the UKPPCF. The UKPPCF committee is contactable at ukpp-cf-request@jiscmail.ac.uk. The UKPPCF comprises clinical psychologists and social workers from paediatric and adult centres, and can therefore provide consensus consultation
- ECFS standards of care, including: Framework for the Cystic Fibrosis Centre,² Best Practice Guidelines,¹⁰ Quality Management in Cystic Fibrosis,¹¹ Standards of care for CFTR variant-specific therapy (including modulators) for people with cystic fibrosis,¹² Standards for the

care of people with cystic fibrosis (CF): A timely and accurate diagnosis,¹³ Standards for the care of people with cystic fibrosis; establishing and maintaining health.¹⁴

- NICE guidelines for cystic fibrosis (NG 78) (for England, Wales and Northern Ireland) and other relevant NICE guidance³
- further CF-specific guidelines and consensus documents referred to throughout this document
- further guidelines and consensus documents related to the practice of UK clinical psychologists, including Health and Care Professions Council (HCPC) standards, and British Psychological Society (BPS) and Association of Clinical Psychologists (ACP) publications
- individual CF centre guidelines.

1.3 Review of the document

These guidelines will be reviewed every three years, or earlier if required. With the introduction of CFTR modulators for many – but not all – people with CF, and other changes in CF care, including impacts of the COVID-19 pandemic, psychosocial needs and service provision is evolving rapidly.

1.4 Audit of CF clinical psychology services

In the past, CF services were subject to regular peer review. As this does not currently take place, the emphasis is on self-review by CF services. It is therefore important for CF CPs to audit their own services.

A self-assessment tool for the CF CP, made up of the core roles that should be provided (if the CF clinical psychology service is sufficiently staffed and integrated into the CF MDT, and in line with local practices) is included in Appendix 1.

2. Introduction

2.1 Cystic fibrosis (CF) and CF care in the UK and worldwide

Cystic fibrosis (CF) is a genetic condition affecting more than 10,900 people in the UK, and diagnosis is made in approximately 1 in every 2,500 babies born. Nearly 58% of the UK population with CF is now over the age of 18 years. The median age of the UK population with CF is 21 and the median predicted survival age is 53.3 years. However, the median age at death in 2022 was 33 years.¹⁵

The nature of CF as a lifelong and progressive condition, including high treatment burden and impacts of cross-infection risks, can result in physical, emotional and social challenges for people living with CF and their families. For reviews of current issues in CF in general, including the need for more recognition of CF and equity of care in the global CF population, and likely future developments and priorities in CF care, see, for example, Bell et al. (2020), De Boeck (2020), Barry & Plant (2021).^{16, 17, 18}

Over recent decades, there has been a significant evolution of CF care, resulting in increased survival age for people with CF in the UK. This is largely attributed to improvements in care, including establishment of multidisciplinary centres, creation of large epidemiological datasets, and emphasis on early diagnosis, together with important new treatments produced as a result of pre-clinical and clinical research.

Following these developments, we are now in a new period of unprecedented change in the world of CF and CF care. In 2020, new CFTR modulators, also referred to as VSTs, including Kaftrio[®] became available to approximately 85–90% of those with CF over age 12 in the UK, with roll-out to the eligible 6–11 age group commencing in the UK in early 2022, and the 2–5 age group in late 2023. These developments are expected to have significant impacts on health and healthcare for the majority of people with CF. It is essential to remember the needs of the people with CF who do not yet have access to such potentially life-changing treatments. The current disparities in access to new treatments have an impact not only on the individuals and families involved but also on the CF community as a whole. In addition, for those who are taking new CFTR modulators, the long-term medical and psychological benefits and other outcomes are not yet known. Different cohorts of people with CF will benefit to different degrees, and some may need to stop taking CFTR modulators due to adverse effects or other issues.

Work is ongoing to develop other new treatments for the benefit of all people with CF.

Alongside the major developments in CF treatments available, the worldwide COVID-19 pandemic has taken place and also impacted significantly on people with CF and their families, and on delivery of CF services. The rapidly evolving nature of CF and CF care at the present time is reflected as much as possible in the current guidelines.

In the UK, people with CF should receive all or some of their care from a specialist CF centre. Some centres (usually paediatric ones) will also have network care arrangements between a specialist CF centre and an outreach clinic – a ‘shared care’ arrangement. In 2022 there were 56 specialist CF centres in the UK (30 paediatric and 26 adult) and 75 networked clinics. A specialist CF centre will have a minimum of 50 children or adult patients (but typically approximately 100) and should have a core multidisciplinary team (MDT) of trained and experienced CF specialist healthcare professionals, including CF clinical psychologists (CF CPs) and CF social workers (CF SWs).^{1, 8, 9, 15}

2.2 Clinical psychologists in UK CF services

Detailed guidelines for the work of UK CF CPs are new, but the specialised role, and the requirement for CF services to include CF CPs, are not. CF CPs have been embedded in CF MDTs in the UK for many years, with this role having been established three decades ago in some centres. The early integration of specialist psychological care into CF teams reflects the generally holistic model employed in UK CF services. This recommended model of closely integrated medical and psychological care (and the potential benefits to patient care), is now well-recognised across UK healthcare provision.^{19, 20, 21} See also [this discussion paper by the Psychological Professions Network](#).

The particular ‘added value’ to the CF MDT by a CF CP is described throughout the current guidance. The work of the CF CP is complex, with interventions at both direct and indirect levels. These depend on expert knowledge of CF and CF care, and close liaison with the rest of the CF MDT. The range of roles offered cannot therefore be provided in the same way by clinical psychology services or other psychological practitioners outside CF care.

Current UK CF standards of care require all CF centres to include clinical psychology provision as a core part of the CF MDT¹ and state that all those receiving CF care must have access to clinical psychology within the CF team. The need for specialist clinical psychology provision to be embedded within the CF team, and the key roles to be provided, are also outlined in NICE guidance for CF³ and in European standards of care for CF.^{2,10}

Clinical psychologists are part of a wider category of specialised, doctorate-level trained practitioner psychologists, as defined by the Health and Care Professions Council (HCPC). Specifying the profession of clinical psychologist for particular posts (as opposed to the wider category of practitioner psychologist) is subject to ongoing debate within the profession. It was not possible to reach a full consensus on this issue amongst the writers of the current guidelines. It should be noted that certain other practitioner psychologists, for example counselling psychologists, may be employed by the NHS to provide CF psychology services if they meet the person specification for a CF psychologist post and can deliver the full range of roles required in the job description. However, the title CF clinical psychologist (CF CP) will continue to be used throughout the current guidelines.

For more information about employing CPs in UK CF services, including staffing levels, recruitment, job planning, management, governance, CPD and the infrastructure required to deliver a CF clinical psychology service, see our summary information for commissioners of CF services and CF clinical directors.

The work of the specialist CF CP runs alongside that of the specialist CF SW. Both are required within UK CF services to provide joined-up psychological and social care, and the two together are viewed as the CF psychosocial team (see Section 6.5). There is now increasing recognition of the impacts of health disparities and health inequalities, with both physical and mental health outcomes greatly influenced by a range of wider determinants of health (including social, economic and environmental factors). NHS policy now requires action on health equalities in all services (see [Health disparities and health inequalities: applying All Our Health](#)). Within CF services, work must be done to identify the key issues and the actions needed. The CF psychosocial team are crucial to this work and consideration of wider determinants of health and health-related behaviour forms a key part of the work of the CF CP.

Psychological principles also inform much of the care of people with CF by other members of the CF team, for example through effective communication skills, and their supportive and collaborative holistic approach to those with CF and their families. Some areas covered by these guidelines, for example managing CF care and treatment, are also very much the remit of the rest of the CF MDT. The guidelines recognise these shared areas of work but focus specifically on the roles and recommendations for good practice for the CF CP in each of the areas described.

CF care in the UK is currently under some review, largely due to recent developments with CFTR modulators, increasing use of IT and remote monitoring, and the impacts of these on delivery of CF care. In a time of such change, the longer-term future psychological needs of people with CF remain to be seen. This is a time of great possibility but also uncertainty in CF care. The challenge for CF clinical psychology services of the future (as for CF services as a whole) will be to keep good practices that are still required, while adopting new ones in tandem. The current guidance draws on the experience of CF CPs, and the needs of those with CF, over the last three decades of CF CPs providing specialist services in CF care. However, recent and ongoing changes in CF care are reflected here, too, and future CF CP provision is also considered (see Section 10).

The essential criteria for a CF CP and key recommendations for service delivery are listed in Section 3. **Detailed information on all the key areas of work by the CF CP is then given in Sections 4–9, with recommendations in each section for:**

- The core roles that are recommended should be provided by the CF CP (if the CF clinical psychology service is sufficiently staffed and integrated into the CF MDT, and in line with local practices)
- additional good practice points and suggestions for CF CPs.

3. Core roles of the CF CP and delivery of the CF clinical psychology service

3.1 Roles of the CF CP: Background

- Roles of the CF CP have been outlined elsewhere^{2, 22, 23, 24} and are summarised in the CF Trust Standards of Care for CF (2011).¹ The multiple and evolving roles that should currently be provided by a CF CP in UK CF care are, however, described in more detail for the first time in this guidance.
- While the CF CP can contribute in multiple ways to the CF service, provision of all these roles will only be possible if sufficient CF CP staffing is in place. If not, some roles will need to be prioritised over others and priorities should be decided in conjunction with the CF service director at a local level.
- Job plans should be created for CF CP posts to determine the most appropriate mix of activities. Where there is a CF CP team, job planning will also help ensure that the time of differently banded CPs is used optimally. For more information on job planning, see the summary information for commissioners of CF services and CF service directors
- Roles of the CF CP may differ between paediatric and adult CF services. In the former, the CF CP may be more routinely embedded within CF clinics, seeing all families regularly (in some services), working with parents and other family members as well as children and young people with CF, and focusing on building skills and the prevention of problems through joint consultations with CF MDT colleagues. In adult services, there may be a model of referral to psychology, and more direct and individual psychotherapy work. But work alongside the CF clinic, ward, or team, indirect work, and the preventative approach are also very much part of the role.
- In adult services, work with parents or carers of people with CF is also approached differently. Family members may be involved in the work of the CF CP with an adult with CF, if this is the wish

of that adult. However, it is unlikely that the CF CP in adult services will provide any significant assessment or therapy/intervention role with individual caregivers, family members or partners of people with CF outside this scenario. These individuals may be signposted to services they can access for themselves as appropriate, and the adult CF SW and/or specialist nurses may also carry out some further supportive work with them if appropriate.

- See Appendix 1 for a complete list of the recommended core roles to be provided. Self-assessment by audit against these core roles can be completed.
- Further recommendations for good practice in all areas of CF CP work are also made in Sections 4–9.

3.2 Core roles of the CF CP

3.2.1 Essential criteria

- CF psychology services must be provided by a clinical psychologist,* and at larger centres by a consultant clinical psychologist.
- All people with CF in UK services must have access to a CF CP as part of the CF MDT.
- CF CPs must be registered with the Health and Care Professions Council (HCPC), and maintain CPD, supervision and registration appropriately, in line with HCPC standards.
- CF CPs should be members of the UK Psychosocial Professionals in CF Group (UKPPCF).
- CF CPs must maintain specialist knowledge about CF, for example through appropriate CPD, membership of the UKPPCF and attendance at UKPPCF and other CF training events.

*See Section 2.2 about the employment of other practitioner psychologists

3.2.2 Service delivery – recommendations:

The CF CP should:

- Respond to referral of inpatients within one week and outpatients within two weeks (using a triage system to assess the urgency of referrals).
- Use a preventative model of psychological care, with screening and other monitoring of both psychological health and health management, early intervention, and a proactive outreach approach to people with CF and their caregivers.
- Help ensure equitable and sufficient access to CF CP care within a centre and to those receiving network care.
- Advise CF commissioners, service directors and others as appropriate on development and provision of CF clinical psychology services, and help ensure appropriate: staffing and banding, recruitment and retention, job planning, professional management, CPD, governance and access to the facilities and systems required for effective service delivery. For more details on service delivery, see our summary information for commissioners of CF services and CF clinical directors.
- Ensure good self-care, including access to the required level of clinical supervision.
- Help ensure people with CF and their families are aware of the CF psychology service and provide information about how CF CP might help and how to access their service.
- Ensure adequate systems for clinical record-keeping, taking account of governance and needs for retaining confidentiality of some CF CP records.
- Collect data on psychological work and activity delivered in the CF service, as required locally or regionally (including for the NHS Specialised Services Quality Dashboard for CF in England).
- Carry out psychological research, quality improvement initiatives and audit in CF, and disseminate this work through publication and/or presentation, with awareness of the research priorities of the CF community.^{25,26}

- Collect and share feedback from people with CF and their caregivers about CF psychology services, involve them in service improvement work and research, and use their 'expert by experience' knowledge and skills where possible.
- Provide training placements for students, including doctoral practitioner psychologist trainees, and supervision of postgraduate research, as appropriate to the needs of the service.

See Sections 4–9 for the further recommended core roles of the CF CP (highlighted in yellow), and additional good practice points (highlighted in grey).

See Appendix 1 for a complete list of core roles of the CF CP and the self-audit tool.

4. Annual assessment and additional screening by the CF CP

4.1 Annual assessment and screening

Screening and monitoring of emotional health and health management in CF by the CF CP allows a preventative and early intervention model to be delivered in CF care. Existing standards of care require CF CPs to carry out psychological assessment or screening regularly, for example at annual review.^{1, 3, 10} A strategy for CF CP screening, surveillance, and monitoring of psychological health for people with CF (or for caregivers in paediatric care, especially in the early years) needs to be in place in UK CF centres, with at least annual screening delivered by the CF CP themselves. Methods, exact content and measures used in the CF CP annual screening process may differ from centre to centre depending on local practices.

International guidelines on screening for symptoms of anxiety and depression in people with CF aged 12 and over (and in caregivers of children with CF aged 0–17) are available from the [ECFS International Committee on Mental Health in Cystic Fibrosis \(ICMH\)](#).

ICMH guidance also provides pathways for assessment and psychological (and/or pharmacological) interventions for anxiety and depression. The screening measures recommended by the ICMH for people with CF are the PHQ9 and the GAD7. These widely used measures have the advantage of being free and available in other languages, but may require additional interpretation in people with CF, especially when they are unwell, due to the inclusion of several physical symptoms. Evaluation of the implementation of the ICMH guidelines has also highlighted some issues with reduced sensitivity of the PHQ9 and GAD7 measures in adolescents with CF and caregivers when compared with information from clinical interview.²⁷

A wider range of psychological issues, including other emotional health issues in addition to symptoms of anxiety and depression, and health management concerns, should be asked about at CF annual review for people with CF in the UK.³ The CF CP annual assessment should not be limited to measures of symptoms of anxiety and depression. In addition to concerns about anxiety or mood,

adults with CF have indicated a wish to discuss a range of issues including but not limited to life transitions, quality of life, life stress, adjustment to CF, existential issues, relationships, loss, work-life balance, treatment adherence, stigma/disclosure re CF, and self-esteem.²⁸ CF CP help may also be needed for other issues that are not easily identified by questionnaires pertaining only to anxiety and depression, for example difficulties with adjustment to CF, managing aspects of health and care, coping style, health beliefs etc. For information about quality-of-life measures, use of other patient-reported outcome measures (PROMs) and examples of further questionnaire measures used by CF CPs for assessment purposes, see Section 7.

Consensus guidance on the suggested content and format of CF CP annual reviews is available from the UKPPCF. See Appendix 2 for guidance in paediatric CF services, and Appendix 3 for guidance in adult CF services (due for review).

In large centres, annual assessment by the CF CP may need to be paper-based (or use an electronically delivered annual screening tool). In smaller and/or well-resourced centres, face-to-face assessment by the CF CP on an annual basis may be possible and preferable. In paediatric services, the CF CP annual assessment with children should be carried out via direct assessment rather than solely via questionnaires or other paper screenings.

Areas to be covered in the CF CP annual assessment will evolve as CF care and the needs of people with CF and their families change. For example, questions about the impacts of changes such as the COVID-19 pandemic, the availability of new CFTR modulators to many, and the resulting effects on emotional and physical health and the delivery of care can be added to annual screening protocols to identify needs in these areas. See Section 10 for further information about future care.

Psychological wellbeing data is not currently collected in the UK CF Registry. Work to identify potentially suitable data that could be collected in a standardised way across centres is underway and will further inform the content of the CF CP annual assessment.

4.2 Other screening/ surveillance by the CF CP

NICE guidance for CF³ recommends that monitoring, screening or proactive assessment by the CF CP may also be needed at times other than annual review, for example through the review of vulnerable groups and people at particularly complicated stages of cystic fibrosis, or at times of significant changes in health. Additional screening or monitoring may be required at specific times, for example when CF is newly diagnosed; during significant CF exacerbations; when there are secondary diagnoses; pre- and post-transition to adult services; during pregnancy and assisted conception; when there is gender dysphoria; for neurodivergent people, when a person with CF becomes a parent; at advanced stages of the condition or end-of-life-care; and at referral for transplant. (See Sections 7–9 for more information). Screening and monitoring of emotional health is now also recommended when new CFTR modulators are introduced.¹² Assessing and monitoring the emotional health of those who are not able to access such new treatments, or who need to stop those treatments for any reason, may also be particularly important.

Additional monitoring by the CF CP is also likely to be needed when mental health problems are observed to have developed; in cases of substance misuse; and where there are considered to be significant risks. People at risk of developing emotional health difficulties (or of relapse to previous known difficulties) may also benefit from more informal CF CP reviews on a more frequent basis than annually (including regular screening or review after a period of intervention to check maintenance of improvement). This allows early intervention for emerging or recurring difficulties, and preventative work.

Especially in paediatric centres, monitoring is often delivered via the routine presence of the CF CP in MDT outpatient clinics or during CF admissions (as well as at annual review). This allows for more informal screening and monitoring of psychological wellbeing and health management issues. It also allows the CF CP to meet people with CF and their families within routine CF care, build relationships, provide brief assessments and interventions, offer consultation and advice to the team, have joint consultations with families and other team members, and signpost to other psychological and wellbeing services where needed. Similarly, in both paediatric and adult services, the CF CP is able to carry out further monitoring of psychological wellbeing, or identification of signs of emerging difficulties, through participation in MDT discussions about

patient care. CF CPs are also likely to work closely with other members of the CF MDT who regularly review the emotional health of people with CF and their caregivers in their day-to-day care. Close liaison with CF MDT colleagues can also aid early identification of those who may benefit from specialised CF CP assessment and intervention.

CF MDT awareness of signs of early difficulty in coping, and identification of those that may benefit from CF CP assessment and intervention, is essential to allow preventative work to be carried out. Following identification of emerging difficulties, CF teams should be encouraged to 'refer early' to the CF CP, before difficulties become entrenched, to minimise the negative impacts of poor psychological health in CF (see Section 7). Proactive identification of current or emerging emotional health difficulties is also likely to be needed, as people with CF may not identify such issues unless asked.¹⁰ CF teams may need guidance on what an appropriate referral to the CF CP looks like. Education and information provided to people with CF about recognising the need for CF CP intervention and how to access this (including self-referral where available) will also help achieve early access to the CF CP.

Following screening by the CF CP, detailed clinical assessment may then be indicated, for example for issues around emotional health, behaviour and health management, which will be arranged according to local protocols for access to work with the CF CP. For more detail on CF CP assessment and interventions, see Sections 5 and 7–9.

4.3 Annual assessment and other screening by the CF CP: Core roles and further good practice points

4.3.1 Annual assessment and other screening: Core roles

It is recommended that the CF CP should:

- Devise a robust local strategy for monitoring of psychological health for people with CF, with developmentally appropriate screening by the CF CP carried out at least annually, through paper/digital or face-to-face assessments at the CF annual review.
- Provide psychological screening at the CF annual review that allows early identification of a wide range of psychological difficulties

and an overview of both emotional wellbeing/mental health issues (including but not limited to anxiety and depression) and health management issues.

- In paediatric centres, screen/monitor the wellbeing of caregivers (in adult services, CF CP annual assessment will be limited to people with CF and their caregivers will not be routinely screened).
- Be aware of guidance from the International Committee on Mental Health in Cystic Fibrosis (ICMH) on annual screening of people with CF (and caregivers in paediatrics) for anxiety and depression, and apply as appropriate locally.
- Provide information as part of the annual assessment about how to access further assessment and intervention from the CF CP and an opportunity to ask for this if needed.
- Be aware of guidance on screening and monitoring of emotional health when people with CF are starting new CFTR modulators, and implement locally as appropriate
- Provide additional monitoring of the psychological health of people with CF (and of caregivers in paediatric services) through a variety of other means, for example through attendance at CF clinics and/or MDT meetings and close liaison with CF team colleagues.

4.3.2 Annual assessment and other screening: Further good practice points for the CF CP

- Provide annual review that is not solely problem-focused and that includes opportunity for people with CF and their caregivers to report, for example, good psychological function, coping strategies already used, personal strengths and sources of support.
- Share information from the CF CP annual assessment with the MDT as appropriate, (with consent of the person with CF/ caregiver).
- Provide additional proactive screening and monitoring to certain individuals or groups of individuals in the service or at key points in the CF experience (for example to parents of newly diagnosed children with CF, when there are significant changes to CFTR modulators, at transition to adult services, at adult diagnosis of CF, to patients planning or having families, at assessment for solid organ transplant, for coping with significant co-morbidities, to patients in the end-stage of the condition, to people with neurodivergence).
- Provide advice as required to the CF team, to help them also identify the psychological needs of people with CF and their families in routine CF care.

5. 'Direct work' by the CF CP

5.1 'Direct work': Background

A large part of the CF CP role will be to carry out 'direct work' with people with CF of all ages or their caregivers. This will include the processes of: engagement work, assessment and formulation of psychological issues, delivery of evidence-based interventions, and evaluation of the outcomes of interventions. Delivery of psychological therapy and interventions through direct work with people with CF and their caregivers is the foundation of the work of a clinical psychologist and may be the role most expected and understood by CF services. However, in CF care this is usually delivered in addition to substantial amounts of 'indirect work' by the CF CP (see Section 6).

5.2 'Direct work' for engagement, assessment, formulation and intervention by the CF CP

Through direct work with people with CF and their caregivers, CF CPs can carry out engagement work, readiness-for-change work and relationship-building (for example for those who are unsure about working with the CF CP) and can offer flexibility in service delivery to take account of CF and CF care issues. Locality-based psychological services may differ in some significant ways to CF clinical psychology services, may not be able to offer the same flexibility (for example fitting appointments around health and care needs) and will not generally have knowledge of CF.

When a likely need for direct work with the CF CP has been identified, the CF CP will meet with an individual or individuals to carry out an assessment of presenting issues. NICE guidance in CF³ reported a lack of validated tools for psychological assessment within CF services; assessment processes must therefore currently be based on the opinion and experience of CF CPs. Limited psychometric testing may also be carried out by some CF CPs, with referral for specialist assessment and services where required.

As part of the assessment, CF CPs will carry out formulation – an evolving, individualised and theory-based hypothesis about the particular experiences of a person, what is contributing to their presenting difficulties and what psychological interventions might help. CF CPs do not use a medical diagnostic model for assessment, but

a biopsychosocial model that recognises multiple factors impacting on an individual, including the role of trauma, and power and threat experiences and social adversity (see Section 7 for more information). Assessments should be 'trauma-informed' in approach – see, for example, the [Evidence-based guidelines for conducting trauma-informed talking therapy assessments](#).

Following assessment and formulation, a range of recognised therapeutic approaches are likely to be used in direct work by CF CPs, including: behavioural interventions, cognitive behavioural therapy (CBT) or CBT-informed interventions, trauma-based and trauma-informed interventions, 'third wave' CBT approaches (including metacognitive therapy, acceptance and commitment therapy (ACT/FACT), dialectical behaviour therapy (DBT)), mindfulness-based interventions and compassion-focused therapy (CFT). Interpersonal therapy (IPT), systemic therapies including narrative approaches, psychodynamic therapy, solution-focused approaches (SFT or SFBT) positive psychology approaches and specific grief-related work may also be offered by the CF CP. Health management work will also be carried out, using a variety of the above therapy approaches and also strategies such as motivational interviewing, use of health belief models and systemic models, and other psychological approaches to behaviour change and coping with long-term health issues (see Section 8).

Direct work with the CF CP will be carried out with informed consent and will be collaborative, to build skills that optimise emotional health, effective management of CF and quality of life. Any issues arising relating to risk or safeguarding will need to be managed. In paediatric care, the CF CP may offer direct work for CF-related issues to caregivers/family members especially in the early years, (as well as to children and young people with CF) but can also signpost or refer caregivers to local services, for example for mental health or other support.

Direct work may be carried out in person, meaning face-to-face in a range of settings, or virtually. Appropriate governance must be in place for virtual work by CF CPs, and the benefits and risks must be considered. The same requirements for confidential space in which to conduct therapy sessions will apply. For guidance for psychologists on virtual working, see the British Psychological Society's resources, [Psychological assessment undertaken remotely](#) and [Effective therapy via video: Top tips](#). The CF CP is likely to carry out direct work with

people with CF and families for a very wide range of difficulties, including emotional wellbeing and mental health as well as management of CF and CF care. For more details on the wide-ranging issues that may require direct work with the CF CP, see Sections 7–9.

5.3 Direct work by the CF CP: Core roles and further good practice points

5.3.1 'Direct work' with people with CF and their families: core roles

It is recommended that the CF CP should:

- Carry out engagement work with people with CF (and caregivers in paediatrics), for example when a person seems likely to benefit from CF CP input but feels unsure about the value of CF CP input or has barriers to this (for example perceived stigma).
- Offer direct CF CP assessment, formulation and intervention to people with CF (or caregivers where appropriate), for a wide range of issues including emotional wellbeing, mental health and management of CF.
- Evaluate interventions, including use of standardised outcome measures where appropriate.
- Assess and manage any safeguarding or risk issues arising in direct work, in conjunction with other members of the CF team (especially CF social workers), appropriate NHS Trust departments and other agencies as required.
- Offer direct work through 1) presence in the CF clinic where appropriate (especially in paediatric care) 2) CF CP outpatient appointments (arranged together with the CF clinic appointments where helpful) 3) during CF admissions as required.
- Offer a hybrid model of face-to-face and virtual working, with telemedicine/ video consultations and telephone work as appropriate, taking account of medical, psychological, social and governance issues that should influence decisions about how CF CP care is delivered.
- Ensure that all contact for face-to-face direct work with people with CF follows

infection control guidelines including any CF-specific guidance, and that any environment used is appropriate in this regard.

- Ensure that suitable therapeutic space is used for direct work with people with CF and their families.

5.3.2 'Direct work' with people with CF and families: Further good practice points for the CF CP

- Offer brief or ad hoc direct interventions within routine CF care, for example within CF clinics or CF admissions (especially in paediatric care).
- Carry out psychometric testing of cognitive function (for example using the WISC) and of neuropsychological issues as appropriate (expertise in this area of work and access to testing materials will vary) but refer on to specialist neuropsychology services where this is more appropriate.
- Consider offering group interventions using virtual platforms, for example to facilitate peer support, deliver psychoeducation or provide group therapy.
- Consider whether CF CP direct work should be delivered in person in homecare settings (as well as in outpatient and ward settings) depending on CF CP demand and provision, taking account of impacts of home visits on people with CF and their families, governance issues and local lone-working policies.
- Offer flexibility in appointments to allow access to CF clinical psychology services at times of poor health, adapting delivery of assessment and interventions to people who are acutely or seriously ill.
- Involve family members/other important people in direct work with people with CF as appropriate, i.e. with their consent and when clinically indicated.
- Work collaboratively with colleagues in the CF MDT to provide coordinated direct interventions, for example through joint sessions with people with CF or caregivers.
- Manage complex issues of confidentiality and appropriate information-sharing about direct work with people with CF or caregivers with the MDT (and with others as needed).

6. 'Indirect work' by the CF CP

6.1 'Indirect work' by the CF CP: Background

As well as providing the expected 'direct' psychological assessment and therapy services to people with CF and caregivers, CF CPs also have a significant role in helping the whole CF service ensure that care provided is 'psychologically informed'. Where a person with CF or their caregivers do not require direct intervention from the CF CP, or they decline it, indirect working is also an effective way for the CF CP to support other CF MDT professionals with psychological aspects of care.

The CF CP can help ensure that psychological principles and evidence are used throughout the CF service – in its direct patient care, quality improvement (QI) initiatives, all forms of communication, strategy, and service development and planning. For more information on QI in CF care, see also [Atul Gawande's article "The Bell Curve"](#) and the [Cystic Fibrosis Trust's pages on quality improvement](#).

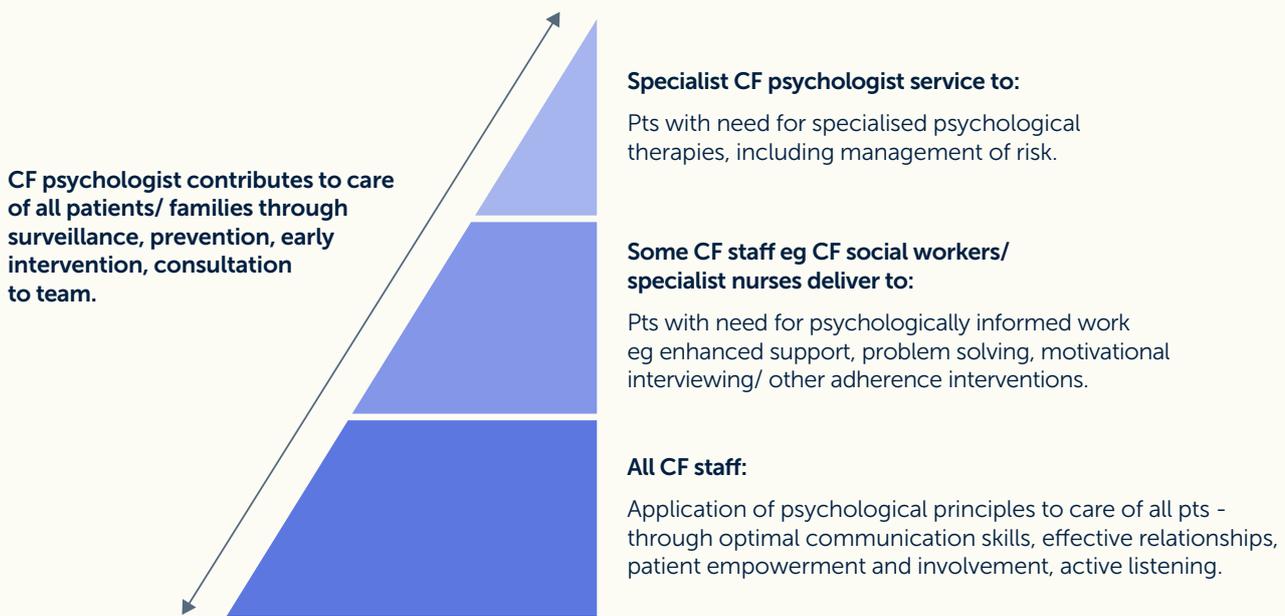
CF CP representation in CF team structures such as working groups and service lead/senior management meetings will allow psychologically informed care to be embedded within the CF service delivery as a whole. The CF CP will have specific training and skills in leadership which can be used by the CF MDT, as well as within the CF clinical psychology service itself. The CF CP can also support a 'stepped care' model, including provision of consultation and advice, training and supervision for staff, and close collaborative working with MDT colleagues. They can help the CF team to function healthily and to promote and support self-care. This 'indirect' work may be a substantial part of the work of the CF CP but is not likely to be available in the same way from external clinical psychology (or other therapy) services outside the CF service.

6.2 The 'stepped care' model of preventative psychological care in CF

Direct work by the CF CP can be resource-heavy and with limited CF CP resources available, allocation of this may need to be selective. For example, this may need to be reserved for those presenting with the highest levels of complexity, or only where there are psychological issues directly affecting CF management and outcomes including morbidity and prognosis.

Other CF team members also have a significant role to play in delivering psychologically informed care. They can recognise emotional health concerns or early difficulties in coping, and provide significant emotional support, depending on their skills, training and aptitude for this work. Some staff will also be able to deliver limited specific psychological interventions or support, where CF CP input is not indicated, but with access to the CF CP for advice as required. In this way, a 'stepped care' model of provision of psychological care can be implemented. This utilises the considerable existing skills of the CF MDT skills effectively, makes efficient use of a limited CF CP resource and ensures that psychologically informed care is available to everyone receiving CF care. See Figure 1 for a summary of the 'stepped care' model in CF, and Appendix 4 for content of a 2015 European CF Society conference poster outlining the long standing preventative approach of the UK CF CP, including provision of indirect care.

Figure 1: Psychological care in CF: model of stepped care and opportunities for preventative actions



Management of risk in the 'stepped care' model

As for all health professionals, CF MDT members have a duty to identify and manage a variety of non-medical risks; for example to safeguard children or vulnerable adults, and to manage suicide risk and risks to others. Training will be available to all NHS staff, delivered through their local trust, on risk assessment, risk management and safeguarding. CF CPs are experienced in the assessment and management of risk and can also support the CF team with advice and local protocols as required. For example, a CF team may benefit from a pathway, advice, or training on how to ask about and respond to signs of risk of suicide or other risk, particularly when the CF CP is not available for consultation on an urgent basis. Advice on when to refer on or 'step up' care, for example to specialist mental health services, may also be required to manage risk. The approach to risk management in paediatric care will differ in some ways to adult services.

6.3. CF CP 'consultation' to the CF MDT

In addition to supporting other CF MDT members to deliver some psychological support and care themselves, the CF CP can also, through attending CF MDT meetings, providing training, presenting at educational meetings or journal clubs, and other means:

- Provide access to a psychologically based framework to all staff, for the understanding and care of a person with CF or caregiver in the service. This may involve giving advice, or disseminating psychological knowledge, research and theory to the CF MDT. Sharing of CF CP formulations and hypotheses about presenting issues (with appropriate consent if specific people with CF or families are being discussed) can help teams develop a psychologically informed understanding of issues.
- Help the CF MDT understand and manage a wide range of CF-related issues, including coping with new diagnoses, managing medical procedures, treatment adherence issues, making decisions about treatments and providing care plans for psychological care at any stage of CF.
- Support the CF team with communication issues – for example having difficult conversations, breaking bad news, and healthy team functioning.

- Work closely with CF colleagues to ensure the whole service approach is psychologically informed around complex presenting issues, for example difficult gastrointestinal symptoms, chronic fatigue, fibromyalgia, medically unexplained or persistent physical symptoms, and chronic pain.
- Help CF teams understand the impact of key psychological processes such as attachment, lifespan developmental psychology, trauma and trauma-informed care, interpersonal development, and systemic influences, to aid understanding of people's behaviours and to help care to be psychologically informed.
- Support the CF team's understanding of systemic organisational processes, for example by helping the team understand team dynamics or the impact of systemic organisational factors, such as NHS/hospital trust policies and processes on staff's emotional health.

6.4 CF CP liaison outside the CF team

The CF CP has a specific role to play in providing expert liaison with services and agencies outside the CF team, using specialist skills and knowledge that other CF MDT members may not have, to optimise coordination of care and/or make referrals on behalf of the CF team. **Such services include:**

- Educational psychology services, neuropsychology services and services for people with learning disabilities or neurodiversity, including attention deficit hyperactivity disorder (ADHD) and autistic spectrum conditions.
- Community mental health services, including NHS Talking Therapies in England, children and young people's mental health services (CYPMHS), Community Mental Health Teams for adults (CMHTs) and specialised services, for example eating disorder services or Early Intervention in Psychosis teams.
- Hospital emergency psychiatry services, such as on-call psychiatry or mental health liaison teams.
- Community social services, in conjunction with the CF SW who is likely to take the lead role and be the first point of contact in this area. For example, this may be where there is a child in need, a vulnerable adult, or another safeguarding concern.

- Schools, colleges, universities and employers.
- Other hospital staff providing psychologically informed care, for example play specialists.
- Other services, for example transplant teams, pain management services, and hospices.

6.5 CF CP and CF SW roles: A cohesive CF psychosocial team

The CF CP will work collaboratively with all colleagues within the CF MDT and particularly closely with the CF SW service. The two professions create a CF psychosocial team, providing a holistic range of roles and much professional peer support. Such a psychosocial team can also coordinate their service improvement work, audit and research, and CF MDT education and support. CF CP and CF SW services will need to define and communicate about their respective roles and responsibilities locally, to minimise any unnecessary overlap and to provide clarity about roles for themselves and the CF team.

While the CF CP and CF SW will both work with people with CF and/or caregivers in many key areas – for example in supporting adherence to treatment, managing transition to adult services, or managing advanced CF and end-of-life care – specific roles should exist for each. For example, the CF CP will usually have lead responsibility for providing evidence-based psychological interventions and therapy to people with CF or caregivers, while the CF SW will take responsibility for work around the social factors affecting function, health and quality of life. Together, the psychosocial team is also able to offer a stance somewhat outside the medical model and can provide a different view of CF-related issues for the benefit for people with CF, their families, and the CF team.

CF specialist nurses also provide much psychosocial support to people with CF and families. Specific roles, expertise and responsibilities for the CF CP, the CF SW and the CF specialist nurse service can be defined and agreed at a local level, depending on the specific training, skills and availability of team members.

6.6 CF CP support for the CF team

CF can be a challenging (as well as rewarding) area of work due to multiple factors, including the length of relationships and frequency of contact with people with CF and their families, which results in strong relationships and even attachments. The adversity sometimes faced by those under the care of a CF team, and the need for provision of palliative and end-of-life-care (often at a younger age than in most specialties) can also make work in CF emotionally demanding. The current rapid rate of change in CF care may result in a need for much adaptation, and new challenges or stresses within CF teams. The COVID-19 pandemic also placed additional stresses upon many NHS staff.

CF teams need to be mindful of the need for good self-care. Appropriate structures and processes should be in place in the CF team management structure to ensure good professional support is always available if needed. The CF CP may also play a role in helping the CF team and team leaders ensure effective teamworking, by helping staff to optimise self-care and to access additional support where required. The CF CP should not, however, act as therapist to members of the CF team, who should access appropriate services as required – for example through their own trust’s wellbeing services. **NHS support to staff** increased as a result of the COVID-19 pandemic, with enhanced access to staff wellbeing services in hospital trusts.

CF CPs may offer opportunities for staff to reflect on challenging aspects of their work. This could be through group supervision or reflective practice sessions and consultation about clinical care at individual or staff group levels. The amount of time required to provide this role will be greater in larger teams. CF CPs can also help the CF team access existing supportive processes, such as Schwartz Rounds²⁹ and critical incident support sessions (both often offered by NHS trusts to employees). The CF CP can also participate in or lead facilitation of in-house CF team reflective sessions after patients’ deaths or other difficult events (see also Section 8.2.14). CF CPs do of course need the same self-care and access to support as any other member of the CF team.

6.7 Indirect work by the CF CP: Core roles and further good practice points

6.7.1 ‘Indirect work’: Core roles

It is recommended that the CF CP should:

- Provide a specialised psychological viewpoint to general CF service development, quality improvement initiatives, research, audit and strategy/ future planning.
- Develop a ‘stepped care’ model of psychological care, utilising the skills of the wider CF MDT to deliver psychologically informed and supportive care and interventions for people with CF and caregivers.
- Provide consultation, advice, teaching, training and supervision as required to the CF MDT, for example through attendance at MDT meetings, education sessions and formal or informal supervision arrangements.
- Work closely with CF MDT colleagues to provide ‘joined-up’ care and with the CF SW to form an integrated CF psychosocial team.
- Advise and support the CF team on management of risk issues when required, for example issues around suicidality, safeguarding children and vulnerable adults, or self- neglect.
- Provide expert liaison with multiple statutory and non-statutory services outside a CF centre, including shared care CF services, educational settings, social services and other health services and professionals.
- Provide expert gatekeeping and liaison and make onward referrals to mental health services, including liaison psychiatry and community services, and coordinate access to on-call child and adult psychiatry services when needed.
- Help the CF team develop good models of self-care, for example through advising on team processes and systems for preventing, monitoring and managing staff stress.

6.7.2 'Indirect work': Further good practice points for the CF CP

- Lead on development of psychological aspects of the CF service as a whole, for example through involvement in leadership teams, working groups and away days.
- Offer additional resources, for example psychosocial MDT meetings or reflective practice sessions, to help the CF team understand psychological aspects of presentations and care, best approaches to these, and impacts of CF care on the team.
- Present information about psychological aspects of CF care at external CF educational meetings and conferences as appropriate.
- Contribute a psychological perspective to written information provided by CF teams to people with CF and/or caregivers.
- Produce and use psychologically informed psychoeducational resources (locally or nationally) to help inform people with CF and/or caregivers about commonly encountered psychological and health management issues.

7. Emotional wellbeing and mental health in CF: Key issues and roles of the CF CP

7.1 Emotional wellbeing and mental health in CF: Background

Living with a long-term condition can be a significant life stressor. Thirty percent of people living with a long-term condition describe a co-existing mental health issue, and there is a two to three times increased risk of depression.¹⁹ Having co-morbid physical and mental health problems can delay recovery from both and is generally associated with poorer clinical outcomes, lower quality of life and increased healthcare costs.³⁰ Recent large-scale research also shows higher incidence of common and serious physical health problems and death, as well as mental health problems, in mothers of children with a life-limiting condition.³¹

Emotional health and wellbeing are the result of complex interactions between any individual's history, their current life stressors and the wider systems around them. What could be labelled or diagnosed as emotional health problems or mental illness symptoms within a particular individual can often be better seen as understandable responses to adverse circumstances. Such adversity includes trauma, threat, and power differences, and the meaning ascribed to these experiences. Responses to adversity are then understood as adaptive attempts to survive such trauma and threat (see BPS's **Power Threat Meaning Framework**).

While the medical model (and much of the literature on psychological health in CF) tends to measure and report psychological distress as 'problems' at an individual level, the CF CP will take a broader stance in understanding psychological wellbeing, including the impacts of social and health inequalities. CF CPs need to acknowledge and understand both approaches, to help with shared understanding and communication with CF colleagues.

Living with a long-term progressive condition such as CF can bring challenges, and these may impact upon emotional wellbeing or mental health. However, living with adversity can also result in great strengths, resilience and evidence of coping.

7.2 Potential challenges for parents, children, young people and adults with CF

Challenges of living well with CF at any age or stage include:

- Coping with, and adapting to, the diagnosis of CF (see Section 8.2.1).
- Achieving accurate and evolving knowledge and understanding about CF over time, in changing times and at different developmental stages, including learning that CF is a life-limiting condition and adjusting to this.
- Developing healthy self-esteem and sense of identity, sometimes in the face of perceived 'difference'.
- Developing healthy body image, eating patterns and relationship with food (see Section 8.2.6).
- Building practical coping skills and emotional resilience when facing the challenges of CF and other life challenges not specific to CF, for example trauma or social deprivation and disadvantage.
- Managing change, including progression of CF (with its physical, emotional and social impacts), and tolerating uncertainty and unpredictability in CF through different developmental stages and in the current time of rapid development in CF care and treatments (see Section 8.1).
- Coping with CF-specific challenges and CF care throughout the lifespan (see Section 8.2).
- Coping with CF where there are additional needs such as neurodiversity, learning disability or sensory impairment (see Section 8.2.12).
- Successfully incorporating CF care and treatments into everyday life and balancing the demands of the two (see Section 8.3).
- Managing any impacts of CF on relationships (see Section 9).

7.3 Emotional wellbeing and mental health data in CF: Data from research and clinical experience

7.3.1 Existing data on mental health for people with CF and recommended strategies

Some data on the emotional health of people with CF over the last decade is available, particularly around symptoms of anxiety and depression. The ECFS has [a list of publications](#) in the field of psychological health and CF.

Symptoms of anxiety and depression are common in CF.³² The CF TIDES study³³ reported a rate two to three times higher in people with CF and parents of children with CF than in community samples (worldwide data). The UK data subset for adolescents and adults with CF showed some differences compared to worldwide data,³⁴ which have been explored.³⁵

Elevated depression scores in people with CF have been associated with worse adherence to treatments³⁶ and quality of life.^{37, 38} Mental health can also affect physical health outcomes for people with CF, with larger declines in lung function shown by adults with depression³⁹ and an increased risk of death for CF patients who screened positive for depression.⁴⁰

As a result of the TIDES data, consensus statements for screening, prevention and treatment of depression and anxiety have been produced.⁴¹ See also the [guidelines from the ECFS](#) for more information. This guidance also includes pathways for both medical and non-medical interventions for anxiety and depression, and information about managing suicidality. Guidelines also recommend routine screening for a primary caregiver of children with CF (aged 0–17 years) to allow monitoring of parental emotional wellbeing, early intervention and prevention of significant difficulties in parents.⁴²

Guidance on the promotion of emotional wellness (for example building resilience) in children with CF is also available,⁴³ as well as strategies for mental health assessment and intervention in this age group⁴⁴ and approaches to prevention of anxiety and depression in people with CF.⁴⁵ Strategies to address psychosocial risks, and to promote resilience to overcome psychosocial challenges for people with CF across their lifespan, have also been described.⁴⁶ For guidance on incorporating screening and treatments for anxiety and depression into routine CF care, see Smith et al (2016).⁴⁷ For a further summary of the management

of mental health in CF, including areas beyond anxiety and depression, and an overview of psychological approaches and psychiatric treatments that may be required, see Bathgate et al (2022).⁴⁸

Quality of life is also of key importance in CF – for a review, see Habib et al (2015).⁴⁹ For information about patient-reported outcome measures in CF (PROMS) in general, see Abbott (2018).⁵⁰ For a summary of quality-of-life measures relevant to CF care, see qol.thoracic.org/sections/specific-diseases/cystic-fibrosis.html. The importance of ‘thriving with CF’ through empowerment has also been recently highlighted.⁵¹ Work is currently underway to develop and update quality-of-life measures so that they accurately assess the areas that are important to people with CF.⁵²

7.3.2 Other clinically observed emotional health issues for people with CF and caregivers

Clinically, many other emotional health and behavioural issues have been seen by CF CPs working with people with CF and caregivers, in addition to the anxiety and depression which is the usual focus of CF research into emotional health. For further detail on issues impacting on emotional health that have historically been experienced by parents of children with CF, and children, adolescents and adults with CF, see Appendix 5. In the new era of CFTR modulator treatments for many people with CF, clinically observed emotional health issues may be expected to evolve over time.

Significant behavioural or emotional difficulties in combination with CF can create complex interactions between mood, self-esteem and self-efficacy, motivation, anxiety-driven behaviours and impacts on self-care and physical wellbeing, which create a vicious circle of decline in physical and emotional health, sometimes requiring urgent intervention. Unhelpful coping strategies may also develop, and a higher rate of excessive alcohol use has been reported in people with CF.⁵³

People with significant low mood, demoralisation or depression may have feelings of wishing to be dead, suicidal ideation (thoughts about suicide) and/or suicidal intent (actual plans to end one’s life). According to the Cystic Fibrosis Foundation Patient Registry, 4.4 % of (US) deaths were categorised as suicide or drug overdose in 2020. The UK CF Registry 2022 Annual Data Report (September 2023) shows fewer than five deaths due to suicide and/or trauma combined between 2020 and 2022.¹⁵ Suicidal ideation was expressed by 10.4% of patients sampled in a UK centre.⁵⁴ Rates of self-harm in people with CF are similarly not currently known, but UK rates in general are

increasing.⁵⁵ Helping manage risks of self-neglect, self-harm or suicide will be an important part of the work of the CF CP.

Significant mental health issues such as severe depression can potentially impact on capacity, resulting in complex issues in CF care. People with CF who are not depressed and who have full mental capacity may also sometimes express wishes to end their life with CF earlier than would occur naturally. CF CPs can help CF teams understand different presentations and encourage open discussion about concerns and wishes for the future when patients express ambivalence about survival.

Self-esteem and self-worth are central to emotional health and psychological wellbeing. There has been limited investigation of this important issue for people with CF, with one small-scale study finding equivalent self-esteem in people with CF compared with population norms.⁵⁶ However, low self-esteem in those with CF is often seen clinically by CF CPs. Self-esteem and self-worth may be negatively impacted by factors including growing up with a sense of differentness, or through other early adversity. Low self-worth and self-esteem will impact on emotional health but may also affect self-care, motivation, and confidence to manage CF, engagement with CF care and relationships. Proactive identification of issues around self-esteem will be important within CF care, and the CF CP has a central role in this and in providing psychological intervention where required.

Further work will be required to continue to identify the range and incidence of behavioural and emotional health concerns in people with CF and caregivers, especially in a changing era of CF care for many. However, CF CPs will continue to need to assess and intervene for a wide range of issues relating to emotional wellbeing and mental health, in addition to anxiety and depression.

7.3.3 Emerging data on impacts of new CFTR modulators and the COVID-19 pandemic on the emotional health of people with CF

Much of the existing data on the emotional wellbeing and mental health of those in CF services predates major developments and changes in CF care that have taken place over the last few years. These include the roll-out of new CFTR modulators to a majority of those with CF in the UK, and also the impacts of the worldwide COVID-19 pandemic on the lives of those with CF and their families and on the ways in which CF care is delivered. For a summary of some early evidence on the impacts on the CF community in both these areas, see

Appendix 6. For ECFS standards of care for CFTR variant-specific therapy (including modulators), including the need to prepare, advise and support people with CF and caregivers around any psychological impacts of these treatments, see Southern et al (2023).¹² For more detail on how CF clinical psychology services and knowledge may need to further develop in future, and in light of recent developments in CF, see Section 10.

7.4 Other factors impacting on emotional wellbeing for people with CF and their families

CF CPs will have knowledge and understanding of other factors outlined below, that may impact upon the emotional wellbeing and mental health of people with CF and their caregivers. They can work both directly and indirectly with those using CF services and use this knowledge to optimise emotional health for people with CF and caregivers.

7.4.1 Sexual orientation and gender identity

LGBTQI+ people in general in the UK are at higher risk of experiencing mental health issues, with half reporting experience of depression and three in five reporting anxiety. While there is limited data available in CF, rates of anxiety and depression have also been seen to be higher in gender diverse youths with CF than in their cisgender peers.⁵⁷ There is an absence of UK resources to support CF teams in providing appropriate care to LGBTQI+ individuals specifically. US-based resources are available and may be helpful to CF CPs and teams when considering these issues in an individualised way in UK care – see for example [Tips for CF teams – Caring for gender and sexual minority \(lesbian, gay, bisexual, transgender, queer, questioning, intersex; LGBTQI+\) people with CF](#).

Where specialised services are required, for example for gender dysphoria, the CF CP can also support the team in making appropriate referrals. See also [the BPS guidelines for psychologists working with gender, sexuality, and relationship diversity](#).

7.4.2 Social determinants of health and CF

Social deprivation is known to have widespread negative impacts on both physical and mental

health. The King's Fund has [more information on health inequalities](#) in general.

NHS policy now requires that all services understand and take action on the impacts of health inequalities.

Social deprivation has been shown to be associated with worse outcomes in people with CF, including effects on growth, lung function, healthcare use, employment chances and mortality.⁵⁸ For a review of demographic factors associated with poorer health outcomes in CF (and possible interventions for healthcare teams to implement to address these), see McGarry et al (2019).⁵⁹ Even in the new era of CFTR modulators for many, understanding social determinants of health will continue to be essential in providing CF healthcare.

The CF SW will play a lead role in helping CF services understand and mitigate impacts of, for example, poverty, adversity, discrimination, difficulty in accessing services and other social determinants of health and health inequalities. See [Guidelines for social work services in CF care](#) (in production). The CF CP will work closely with the CF SW in a joined-up psychosocial team approach to the emotional wellbeing of people with CF and their caregivers/families, and to understand and mitigate the impacts of health inequalities in CF.

7.4.3 Strengths, resilience, coping skills, adaptation and social support in people with CF and their caregivers

People with CF and parents of children with CF show considerable strengths, resilience and coping skills, and fostering resilience should be part of a preventative approach taken by the CF CP. Resilience may be considered as the capacity to adjust to changing circumstances, and flexibility, rather than reliance on a fixed set of coping strategies, is a crucial factor in maintaining resilience.⁶⁰ Resilience (when defined as 'personal competence and acceptance') and tolerance of uncertainty was seen to be elevated in one sample of adults with CF, and some aspects of this were positively associated with quality of life.⁶¹ Resilience in CF specifically has also been described as 'the ability to recover from negative events with an absence of lasting emotional disturbance', with a recommendation made to foster resilience in people with CF especially at key points in the CF 'journey'.¹⁰ Approaches and strategies for the promotion of such resilience have been described.⁴⁶ The role of self-compassion in emotional wellbeing for people with CF has also recently been investigated, with positive associations found, and recommendations made for mindfulness and self-compassion interventions as potentially beneficial for adults with CF.^{62, 63}

While the concept of resilience is important, and individual skills, strengths and commitment do impact on coping, these should not be overemphasised, as wider factors, for example an individual's family and other networks, environment, socioeconomic wellbeing, adversity, place in culture and society, and impacts of inequality and discrimination also affect outcomes in multiple ways. For further information, see Seligman & Csikszentmihalyi (2000), Edward (2013), Gouzman et al. (2015), Davis et al. (2009), and Fletcher & Sarkar (2013).^{64, 65, 66, 67, 68}

Coping style and adjustment are also of key importance in CF. 'Active coping' has been shown to be associated with better quality of life in some domains for adults with CF, compared with 'distraction coping'.⁶⁹ A systemic and developmental view of adjustment, for example of a family's adaptation to chronic illness, is also important in optimising the CF team's understanding, and assessment, formulation and intervention by the CF CP.⁷⁰ Other models relevant to coping and adaptation include the Moss-Morris multifaceted model of adjustment⁷¹ and the common-sense self-regulation model.⁷²

Clinically avoidant coping, minimisation and denial are sometimes observed in people with CF and parents or carers, and can be seen as understandable and sometimes very adaptive responses to threat. Understanding the role of such coping styles and strategies, as well as any unintended consequences, and helping people with CF and caregivers adapt their coping style and strategies when required, are important parts of a CF CP's role.

Social support is also likely to be key in developing and maintaining resilience and the ability to cope with adversity, as well as the improvement of emotional health difficulties. Understanding the role of (or lack of) such support in a person's overall emotional health is essential for the CF team and CF CP. Social support has been seen to be associated with better physical and emotional health outcomes in adults with CF, as in those with other chronic illnesses.⁷³

Emotional wellbeing for people with CF and caregivers results from a complex combination of factors, and at this time of great change in CF care, will require continuing evaluation. Good understanding of all these factors impacting upon the psychological and emotional wellbeing of people with CF and families is required to deliver appropriate care. The CF CP is uniquely placed in the CF team for this. The specific roles of the CF CP in assessing and intervening to optimise emotional wellbeing are considered below.

7.5 Emotional wellbeing and mental health: Screening, assessment, formulation and intervention by the CF CP

Much can be done to prevent, mitigate and improve difficulties with emotional wellbeing and mental health and to help those with CF thrive and achieve good quality of life. The CF CP has a key role in this through both direct work (see Section 5), for example, specialised psychological assessment, formulation and intervention with people with CF, and indirect work via the CF MDT (see Section 6). Complex interactions between emotional health and physical health mean that with expert knowledge of CF, the CF CP is able to assess and intervene with emotional health issues in a way that psychological therapists outside CF care may not be able to do. Psychological intervention from within the CF team also means that such work can be integrated with physical healthcare in a holistic way.

In paediatric care, the CF CP may also offer assessment and interventions for emotional health issues relating to CF to caregivers (as well as to children and young people with CF themselves) and can then signpost or refer on to local mental health or support services if required. In adult services, the CF CP would not usually offer intervention to carers or family members of people with CF in respect of their own emotional health, although important others may be involved in some therapeutic work between the CF CP and an adult with CF if indicated and requested by the person with CF.

CF CPs will carry out assessments around emotional wellbeing and mental health through a variety of methods, including clinical interview and use of standardised measure where appropriate. For examples of measures that may be helpful in aiding assessment of emotional health, see box below.

Examples of measures that may be helpful to CF CPs assessing emotional wellbeing/ mental health

- GAD (GAD7, GAD2) (for age 12+)
- PHQ (PHQ2, PHQ8, or most commonly PHQ9) (for age 12+)
- Work and social adjustment scale WSAS⁷⁴
- Distress thermometer scales, for example see Dhingra et al.⁷⁵

- Paediatric Index of Emotional Distress (PI-ED) (for children aged 8-16 years)⁷⁶
- Strengths and difficulties questionnaire (SDQ) (for age 3+)⁷⁷
- Revised Child anxiety and depression scale (RCADS) (for age 8+)^{78, 79}
- Distress in CF scale (DCFS)⁸⁰
- Eating Attitudes and Behaviour measure⁸¹
- Memorial symptom assessment scale (MSAS)⁸²
- Quality of life measures, for example CFQ-R,⁸³ CF QoL,⁸⁴ AWEScore CF⁸⁵
- Measures of resilience, for example Connor-Davidson Resilience Scale (CD-RISC),⁸⁶ Brief Resilience Scale⁸⁷

Following assessment and formulation, CF CPs will deliver evidence-based interventions for emotional health, using a wide range of therapeutic approaches (see Section 5.2). Much of CF CP work with younger children in paediatric care will be around positive parenting (see Section 9.3). Intervention for issues around feeding and eating, especially in the early years, may also be indicated (see Section 8.2.6.1). Both paediatric and adult CF CPs are likely to work with people with CF who struggle to manage procedures; from understandable dislike, for example of venepuncture, through to significant anxiety or phobias around such procedures. (see Section 8.2.4). CPs are not prescribers (unless they have undertaken additional specific training in this) and will therefore not make decisions about pharmacological treatments for psychological health. However, CPs may be involved in discussions which indirectly support people with CF in their decision-making regarding such medications.

Data about which psychological interventions have good outcomes in the area of CF and emotional health specifically is limited, and it has been concluded that 'the overall evidence on psychological interventions for individuals with CF and their families is insufficient'.⁸⁸ There is, however, no reason to believe that evidence-based psychological interventions in general are not applicable to people with CF and their caregivers. For details of psychological treatment modalities and screening tools used in CF care in Europe and beyond, there is more information on the ECFS website.

CF CPs will measure outcomes of interventions for emotional health in a number of ways. Selecting formal outcome measures to evaluate the progress of usefulness of psychological intervention can be complex. Some commonly used measures, for example of depression and anxiety, include physical health symptoms that can make it difficult to interpret results for people with CF. Others are not sensitive enough to change for them to be used repeatedly, or may not measure the aspects of emotional health or function that are most important to people with CF.

CF CP direct work on emotional health issues can range from short interventions for specific issues to longer-term therapy work for more complex issues. Brief interventions from a range of therapeutic options, (for example CBT-informed, ACT-based, and solution-focused approaches) may also be offered within CF clinic consultations or inpatient admissions. Therapeutic interventions for emotional health by the CF CP may be offered remotely (via video or telephone sessions) as well as face-to-face, and group interventions can also happen remotely. Feasibility and potential efficacy of ACT for anxiety and depression symptoms delivered via teletherapy to people with CF have been reported⁸⁹ and a CF-specific, internet-based CBT intervention has been shown to be acceptable.⁹⁰ Further evaluation of psychological interventions with people with CF for emotional wellbeing is however still required (see Section 10).

Whether direct work and interventions for identified issues with emotional wellbeing and mental health should be offered by the CF CP or by community services, for example CYPMHS or CMHTs (and NHS Talking Therapies services in England), will depend on a complex range of factors. These include CF CP provision and level of need in the CF service (with increasing numbers in many services), availability of appropriate community services, geography, patient preference, and the level of need for liaison or joint work with the CF MDT. CF CPs can work with a high level of complexity of mental health issues, including severe and enduring psychological issues, and this may be required in CF care, especially when community psychological therapy services are not easily accessible. However, referral to specialist services will be required in some cases and where there is severe mental illness, psychosis, high level of risk and need for access to crisis support or out-of-hours care. The CF CP has specialist knowledge to inform such decision-making about referral to specialist services in connection with mental health and can liaise with such services.

In addition to direct work for emotional health, the CF CP will also support the emotional health of those in CF services through their indirect work.

This may involve, for example, training for the CF MDT to recognise significant emotional health issues, assess risk, and provide immediate support to those in crisis. The CF CP will also support other members of the CF MDT who help people with CF and caregivers with emotional wellbeing, for example through consultation, education and training, and reflective practice or informal supervision with staff (see Section 6).

7.5.1 Emotional wellbeing and mental health: Core roles of the CF CP

It is recommended that the CF CP should:

- Take a preventative approach to mental health issues for people with CF (and caregivers in paediatric care), for example through help to build coping skills, empowerment and promotion of emotional wellbeing.
- Work directly with people with CF to provide screening, assessment, formulation and evidence-based interventions for emotional wellbeing and mental health (not limited to anxiety and depression).
- Be aware of the International Committee on Mental Health in Cystic Fibrosis (ICMH) guidance on screening, assessment, prevention and treatment for anxiety and depression in CF services and apply as appropriate locally.
- Be aware of guidance on the need for CF MDT familiarity with the potential wide range of psychological impacts of CFTR modulators, and offer CF CP intervention as indicated.
- Offer assessment and intervention for emotional health to caregivers as appropriate in paediatric care, or signpost to appropriate services as required.
- Include family members/caregivers in work with people with CF where appropriate, to provide systemic interventions for emotional health.
- Facilitate other members of the CF team to support and promote the emotional health of people with CF and their families through indirect work, for example through consultation, education and training, reflective practice and supervision, and service improvements.

7.5.2 Emotional wellbeing and mental health: Further good practice points for the CF CP

- Help the CF team educate and support people with CF and caregivers about any emotional health aspects of CFTR modulators, including support for complex decision-making, for example where there is uncertainty about the costs and benefits of such treatments.
- Help the CF team monitor emotional health impacts of taking CFTR modulators, including the use of mood screening questionnaires, especially for those with pre-existing mental health issues (for example via clinic reviews, CF CP annual review, or other methods of follow-up), with awareness of standards of care and guidance in this area.
- Work closely with CF MDT colleagues to help people with CF manage any negative impacts of CFTR modulators on emotional health, or when CFTR modulators need to be stopped for any reason.
- Help the CF team monitor the emotional health of people with CF and/or caregivers with respect to any ongoing impacts of the COVID-19 pandemic, offer assessment and intervention as required, and help CF teams ensure they are also managing stressors related to post pandemic care.
- Be aware of the sexual orientation and gender identity of people with CF, support with any impacts on emotional health, and help the CF team ensure CF care is appropriate for all those in the LGBTQ+ community.
- Be aware of the emotional health needs of people with CF with gender dysphoria, including screening and intervention as required, but ensure that individuals are signposted or referred to specialist support or services where required.
- Take account of social determinants of health and emotional health for people with CF in formulation and interventions.
- Refer to the CF SW where there are significant social or economic factors impacting physical and emotional health, including issues around education, work, future planning, benefits or other financial issues, and family functioning.

- Help the CF team understand and normalise the need for psychological adjustment to health changes (and at different developmental stages in children and adolescents), and to provide support for emotional health as required, with referral to the CF CP where further assessment, formulation, or intervention for emotional health is required.
- Support the CF team in assessing the level of risks associated with mental health when this presents to them, for example through education about initial risk assessment and management in CF clinics when the CF CP is not available.
- Support the CF team with complex issues, for example around mental capacity relating to emotional health and decision-making processes in situations of self-neglect or refusal of life-saving CF treatments, or in cases of child abuse, neglect or safeguarding issues relating to emotional health.
- Provide guidance to the CF team on when to refer to CF CP and when care should be 'stepped up' to external specialist services, for example child and adolescent mental health services (CAMHS), community mental health teams (CMHTs) or hospital mental health liaison teams, Early Intervention in Psychosis services, eating disorder services, gender dysphoria services and neurodiversity services, and liaise with all such services as needed.

8. Managing CF and CF care throughout the lifespan: Key issues and roles of the CF CP

Despite recent advances in treatment, managing CF throughout the lifespan from diagnosis onwards continues to pose various potential challenges for people with CF and their caregivers. The CF CP has a key role to play to help those with CF 'live well' with this condition – to adapt to significant CF-related changes at different life stages (for example in health status or to new treatments), to manage the challenges of CF and to achieve appropriate levels of adherence to CF treatments. This section outlines the roles of the CF CP in helping people with CF and caregivers to manage the combination of demands that CF and CF care brings. Much of the CF CP work in these areas will be carried out alongside the rest of the CF MDT and will be embedded within care delivered by the rest of the team, through joint sessions and close consultation.

8.1 Managing change and transitions in CF

8.1.1 Managing change and transitions in CF: Background and core roles for the CF CP

The CF CP can help people with CF and caregivers to become comfortable and confident in coping with change, and to develop positive coping strategies and resilience in the face of both anticipated transitions and unexpected ones (see also Section 7.4.3 for provisos around resilience). This psychological flexibility will help people with CF and caregivers to manage CF optimally and to see CF as part of themselves and their life, but not be defined by it. With experience of CF and expertise in developmental theory, CF CPs can often anticipate challenges or periods of greatest change for people with CF and their families and help them to manage these.

CF care and the outlook for people with CF have also seen change over recent decades, with whole cohorts of people with CF reaching stages of life they did not expect to reach. CF care is now at a further stage of unprecedented change due to the recent introduction of new CFTR modulators for many. While most welcome, this may also present some challenges, including increased disparities in

care in the CF community, need for adaptation to an unexpected future for some, uncertainty when the picture of the future is shifting, and sometimes regret for past decisions.

Managing change and transitions in CF: Core roles of the CF CP

The CF CP should:

- Provide specialised psychological assessment and interventions and support the work of the CF team as required, at times of change and transitions for people with CF, including in the following areas:
 - managing CF and change at different developmental stages in childhood
 - adolescence and transition to adult life and adult CF services
 - managing growing older with CF.

8.1.2 Managing CF and change in childhood

From the time of diagnosis onwards, people with CF or their parents have the challenge of adapting to new treatment regimes, managing any early changes in health and learning about CF and CF care. For information about psychological needs at diagnosis of CF, see Section 8.2 1. Subsequent medical change may also impact on families to a significant degree – for example the first growth of pseudomonas or the first admission to hospital – and support in managing such changes may be required.

Parents of a child with CF need to cope with the introduction of first treatments and to remain educated about treatments, including when there are complex decisions to be made about these. CF teams need strategies to educate and inform patients and families about CF in a systematic and ongoing way that is manageable over time and at different developmental stages. Understanding of the developing child's knowledge of illness, health and body function is needed for successful education. See [What's it all about?](#) for a resource to educate children about CF.

Knowledge about CF helps with shared decision-making and agreed treatment planning with patients and parents, which improves adherence.⁹¹ Ensuring routine and 'normalising' it as part of everyday living helps to take the spotlight off CF and encourage mastery and control over intrusive aspects of care. For more detail on adherence to CF treatments, see Section 8.3.

Starting in nursery or other childcare settings and then starting primary school are also key transitions for any child and family, including where there is a child with CF. Challenges may include managing feelings about separation, managing practicalities – for example carrying out CF treatments away from the home – and communication with care providers and schools. CF SWs and specialist nurses are likely to have the lead roles in supporting children with CF to start nursery or school and achieve good attendance. Educational Health and Care Plans (EHCP) in England (and equivalents in Wales, Scotland and Northern Ireland) may be required when there are further needs as well as CF, or when significant treatments need to be done in school or in early years settings. We also have [resources for parents of preschool- and primary-aged children with CF and information for primary schools about CF](#).

Managing change in childhood with CF: Further good practice points for the CF CP

- Help CF teams develop psychologically informed processes and practices to educate about CF throughout childhood and beyond, taking into account developmental stages, different learning styles, family preferences, and cultural and religious beliefs.
- Support the CF team in helping children learn to communicate about CF, to carry out usual activities of childhood alongside CF, and to gain independence appropriate to their developmental stage.
- Work directly with caregivers and children, for example within paediatric clinics, to address understanding of CF and adapting to new challenges or significant change in multiple areas of life.
- Help caregivers manage any anxiety or other psychological issues that may impede developing independence in their child by using parenting interventions as needed.
- Contribute to an EHCP (or equivalent) as needed where this is in place and liaise with educational settings when helpful.

- Liaise as required (with the CF team, educational settings, people with CF and families) around any identified cognitive needs, carry out assessments, for example neuropsychological testing, as appropriate to skills and competence in this area, and support the CF team to refer to specialist services where needed.
- Attend Team Around the Family (TAF) meetings as required and participate in any safeguarding discussions and meetings as appropriate.

8.1.3 Adolescence and transition to adult life and adult CF services

Moving through adolescence will bring many 'ordinary' life changes, as well as those specifically related to CF or CF care. Young people aged around 12–25 may also be navigating transitions to high school, college or university, first travel without parents, moving out of the family home, getting first jobs, and many more new experiences.

For those with CF, adolescence and transition to secondary education may bring new challenges including explaining CF and treatment to new peers, less parental involvement and control, and challenges in adopting self-management of health and adherence to CF treatments (see Section 8.3). Some features of adolescence, including increased risk-taking behaviour, may occur later than developmentally average for adolescents who have chronic health conditions,⁹² and it is important that this is recognised in both paediatric and adult CF services.

Parents may appreciate advice about supporting their children with CF during adolescence, for example in making the shift from a 'doing' to a supportive model. An example of this is Teen Triple P.⁹³ There may be challenges for parents in letting go of responsibility for CF care and treatment at this stage of life and development, and also for adolescents in taking responsibility for their CF care. CF can therefore become a conflict point in adolescence and this can impact on family relationships, in addition to the usual family changes during this period. Adolescents whose life experiences have been significantly impacted by CF may find it harder to achieve usual levels of young adult independence and may also need support from the CF team in doing so. Adolescents (and their families) may have unique needs which differ to those of either children or adults. Knowledge of adolescence will support all those working with this age group – see Blakemore & Choudhury (2006)⁹⁴ for more information about the adolescent brain. Adolescents with

additional needs (for example learning disability or neurodiversity) will likely have enhanced needs around the transition period, and care from paediatric and adult centres must be adapted as required to provide a good transition experience.

In England and Wales CF is usually introduced in schools in the Key Stage 3 curriculum as a way of illustrating genetics and inheritance.⁹⁵ There is a wide variation in how young people may feel about knowledge of CF being shared in their school environment. Some may not have shared their diagnosis with friends and peers, or may have limited knowledge about CF themselves. It is important to prepare young people and their families that information may be given that they were not aware of, or that may be outdated. We have [more information for parents of secondary age young people with CF, and their teachers](#).

Many learning difficulties may go unnoticed or undiagnosed until later childhood or adulthood⁹⁶ and CF teams should be aware of any emerging cognitive issues or neurodiversity as these may well impact on CF care and self-management.

An anticipated and important transition for children and adolescents with CF is the transfer of care from paediatric to adult services. Detailed recommendations for UK services about such transitions (across medical specialties) are available⁹⁷ and CF-specific recommendations are also made by NICE³ and the Cystic Fibrosis Trust.¹ For a framework for successful transition, see [GOT Transition](#). For suggestions to improve transition for young people with long-term conditions, see Colver et al (2020).⁹⁸ For reviews of transition challenges in CF, see Goralski et al (2017)⁹⁹ and Singh et al (2022).¹⁰⁰ UK transition tools are available for use across specialties including CF, for example [Ready Steady Go](#). Cystic Fibrosis Trust also has [resources about transition for CF teams, and young people with CF and their families specifically](#).

A small number of UK CF services have 'lifespan' psychology services, which means the same CF CP provides services to both paediatric and adult CF settings. However, in most areas in the UK, paediatric and adult centres have distinct CF clinical psychology services. Close liaison around transition will be essential, and CF CPs in any setting should be involved with facilitating successful transition as required, including in adult services.

Adolescence and transition to adult life and adult CF services: Further good practice points for the CF CP

- Contribute to teaching/training for CF MDTs in both paediatric and adult services around

working with young people and achieving successful transitions to adulthood.

- Contribute psychological opinion and expertise to any local shared transition processes and ensure that service-wide planning and improvements are psychologically informed.
- Help the CF team in paediatric care educate young people and caregivers about any differences between paediatric and adult CF services, both locally and generally, for example through transition events or joint paediatric and adult clinics.
- Share information about the psychological needs of individuals transferring to adult services with the MDT as appropriate, and also between CF CPs in paediatric and adult services.
- Ensure that transferring patients are aware of the adult CF psychology service and how to access this, and consider whether recently transferred people with CF would benefit from early screening or assessment of needs by the CF CP in the adult service.
- Support adult services in the continuing transition of young adults with CF after transfer to adult services (including their parents, if appropriate), for example in acquiring knowledge about CF, gaining autonomy and independence, and making plans for adult life, with individual or systemic assessment and interventions as required.

8.1.4 Growing older with CF

In 2022, 62.9% of people with CF in the UK were aged 16 or over, and the age of median survival has been steadily increasing,¹⁴ even before taking into account the impacts of the new CF treatments now available to many with CF. Adult CF services are likely to steadily grow in size in the future, with increasing numbers of people with CF in middle and older age (see Section 10).

Growing older with CF can result in some additional challenges, historically presenting adults with increasing burden of care, the eventual introduction of more visible or intrusive treatments, new complications or diagnoses, and increasing morbidity with the advancing stages of the condition (see Section 8.2 for more detail). A proactive approach to supporting people with CF with the recognition of likely stressor points (such as a new 'stage' of CF or increasing competing demands) is recommended.¹⁰

Achieving the usual goals of adult life alongside the demands of a condition such as CF can also be challenging and can result in a complex 'juggling act' between health and care demands, psychological wellbeing and the demands of 'real life'. Middle-aged and older adults also have other life-stage changes and stressors to manage, including physical and emotional aspects of normal ageing, such as menopause and age-related illnesses. They may also experience changing roles, including early retirement from work for health reasons, caring for or loss of ageing parents alongside parenting (and grandparenting) themselves, and loss of peers with or without CF.

While increasing challenges may be offset by greater life experience and coping skills, CF services and CF CPs need to be responsive to the needs of this cohort of people with CF. For some adults with CF, survival and life expectancy has far exceeded expectations, and while most welcome, this can trigger complex emotions for some. The introduction of new CFTR modulators for many may result in some similar emotions (see Section 7.3.3).

Growing older with CF: Further good practice points for the CF CP

- Help the CF team support people with CF with the challenges of growing older with CF, for example through consultation and psychologically informed service provision and planning for this age group.
- Offer individual assessment and intervention to adults with CF around issues specifically related to ageing with CF as required, with a proactive approach at potential stressor points.
- Work closely with CF SWs to help mitigate impacts of social, economic, and employment-related stressors or adversity during adulthood with CF.

8.2 Managing challenges in CF and CF care throughout the lifespan: Core roles of the CF CP

In addition to the need for adaptation during periods of change and transitions at each stage of life with CF, there are some specific challenges of CF and CF care that may arise across the lifespan. While many of the CF MDT will also have roles in the following areas, the CF CP is uniquely placed

to ensure CF care is psychologically informed and to provide psychological assessment and intervention where needed. Potential challenges in managing CF are outlined below, together with further recommendations for good practice by the CF CP in each area.

Managing challenges in CF and CF care throughout the lifespan: Core roles of the CF CP it is recommended that the CF CP should:

Provide specialised psychological assessment and interventions around managing the challenges of CF and CF care, and support the work of the CF team/provide joined-up care, including in the following areas:

- new diagnosis of CF at any age
- issues relating to CFTR modulators
- psychological aspects of infection control
- procedural distress
- CF admissions and outpatient care
- nutritional, eating and GI issues
- CF diabetes (CFD)
- secondary diagnoses and complications
- medical trauma
- intrusive treatments and environments
- medically unexplained or persistent physical symptoms
- neurodivergence, neurodevelopmental conditions and other additional needs and CF
- lung transplantation issues
- palliative care, advanced stages of the condition and end-of-life care.

For further detail and good practice points in each of the above areas, see Sections 8.2.1–8.2.14 below.

8.2.1. Managing the diagnosis of CF

Diagnosis of CF (at any age) is a key time to consider psychological wellbeing, and the CF CP has a significant role in helping CF services support parents, children, young people and adults who receive this life-changing diagnosis. CF standards of care recognise the need for emotional support and/or specialist psychological support at diagnosis^{3,10} and state that access to psychological support at the time of 'late diagnosis' (for example in adulthood) is essential.¹

For more information on diagnosis and screening in CF in general, including carrier and prenatal screening issues, see Bell (2020).¹⁵ For information on diagnosis of CF in childhood and psychological support that may be needed, see Havermans et al. (2015)¹⁰¹ or Bryon & Tluczek (2018).¹⁰² Or see [our resource for parents of children diagnosed with CF](#).

Most diagnoses of CF in the UK now occur as a result of newborn screening. However, 9.5% of adults with CF in the current UK CF Registry were diagnosed at age 16 or over, with a small number receiving diagnosis after age 50.¹⁵ See [our late diagnosis factsheet](#) for more information.

Adults diagnosed with CF will have specific needs for education and adjustment.¹⁰³ Psychological support should be available¹ and CF CP input or screening to identify any issues with adjustment may be required.

In addition to those diagnosed with unequivocal CF at any age, others will receive more uncertain diagnoses, including that of CFSPID (CF screen positive, inconclusive diagnosis). For guidance on management of children with CFSPID, see Barben et al. (2020).¹⁰⁴ Others may receive a diagnosis of, for example, CFTR-related disorder or CFTR-related metabolic syndrome (CRMS) with varied clinical consequences and unclear longer-term impacts. For further information on these issues, see for example Bell (2020),¹⁶ and Cystic Fibrosis Trust's [information on CFSPID](#).

Diagnosis of CF: Further good practice points for the CF CP

- Help ensure the CF team has a robust pathway for managing the psychological impacts (for caregivers especially) of diagnosis of CF in babies (after newborn screening) and children.
- Deliver more specialised interventions to caregivers around diagnosis when required, for example when the diagnosis is impacting on parental mental health or if made aware of a pre-existing mental health diagnosis in a parent.
- Help the CF team assess a child with CF's increasing level of understanding about diagnosis over time, to ensure they are included in information about the condition, and the requirements of daily treatments as appropriate to their developmental stage.
- Help ensure the CF team has a robust pathway for the care of newly diagnosed adults with CF that includes individualised

education, proactive awareness of psychological needs and support with adjustment to diagnosis, including access to screening and specialised intervention from the CF CP as required.

- Ensure that those receiving a new diagnosis of CF at any age are made aware of the CF psychology service, for example by giving written information about this and inviting them to self-refer if they have any concerns about emotional reactions to diagnosis or adjustment.
- Offer training, or support the work of other MDT staff involved in the diagnostic process (at any stage) as appropriate, to ensure that team support to families is psychologically informed, for example when sharing sad/bad news.
- Be aware of those with CFSPID in paediatric care and support the CF MDT in, for example, monitoring how parents are making sense of the diagnosis, managing any parental health anxiety about their child and ensuring the child is not over-medicalised.
- Be aware of those diagnosed with 'atypical CF' or CFTR disorder in adult services, assist the CF MDT in supporting them and offer specialised intervention if required (CF psychology services provided will depend partly on what services and level of follow-up is offered by the CF team as a whole to these individuals).

8.2.2 Issues relating to CFTR modulators

The availability of new CFTR modulators to most but not all people with CF has had an overwhelmingly positive impact on care overall. However, challenges may arise for the significant minority who are not currently able to access such treatment options and CF services must ensure they continue to meet all the needs of this group. Those who are able to access CFTR modulator treatments may also experience challenges. This includes people who report complex or negative impacts on emotional health, and those who have to stop such treatments for any reason. Complex and emotive decisions about when or even whether to take CFTR modulators may also be required by some people with CF and caregivers.

CF teams are now adapting to changing practice in service delivery and clinical care relating to CFTR modulator treatments, and managing such rapid change can also result in challenges for teams. CF CPs themselves are also developing their clinical

practice due to impacts of new treatments for CF and may need to develop knowledge and skills and access CPD in new areas of work as a result. See sections 7.3.3 and appendix 6 for more detail.

Issues relating to CFTR modulators: Further good practice points for the CF CP

- Be aware of emerging knowledge and guidance on any psychological issues relating to CFTR modulators, access CPD in this area, and adapt practices as required.
- Ensure that the psychological impacts of not being able to take modulator treatments are understood and addressed by the CF service as a whole and by the CF CP as required.
- Help CF teams support people with CF and/or caregivers with complex decision-making about taking modulator treatments, as required.
- Help teams to adapt services and to manage significant levels of change in delivery of CF services where required.

8.2.3 Managing psychological aspects of infection control

Separation (or 'segregation') from others with CF for the purposes of infection control is an essential part of CF care but results in unusual isolation from peers with the same condition and has been associated with negative psychological impacts in the past.¹⁰⁵ First growth of *Pseudomonas aeruginosa* may also be a significant event, and identification of a new microbiological status that might affect physical health outcomes, CF centre care, a new level of segregation from other family members with CF, or change in access to lung transplantation may all have psychological and emotional impacts too. See Cystic Fibrosis Trust's resources for [more information about cross-infection](#). The COVID-19 pandemic may have resulted in some normalisation of increased infection control practices, such as routine use of more personal protective equipment (PPE) in CF care, but the degree and impacts of such changes are not yet known.

Managing psychological aspects of infection control: Further good practice points for the CF CP

- Help CF teams minimise any negative psychological impacts of new

microbiological status or infection control issues, for example through awareness of possible impacts such as anxiety, stigmatisation and isolation.

- Support people with CF and families to connect with peers in the CF community, for example through social media (with awareness of both the benefits and potential downsides of the online CF world).
- Offer direct therapeutic intervention as appropriate to people experiencing significant negative psychological impacts due to new microbiological status or infection control issues.

8.2.4 Managing procedural distress

Up to 60% of young children in the general population report medical procedure-related distress¹⁰⁶ and a fear of needles has also been demonstrated in 20–50% of adolescents and 20–30% of young adults.¹⁰⁷ Procedural distress is therefore a widespread issue requiring normalisation by CF teams. A range of procedures are common in CF care, and psychologically informed processes need to be in place to manage procedural distress in routine care and to identify when specialist interventions are needed. The BPS has [guidelines for dealing with this](#).

Around 10% of adults and children have significant needle anxiety which will require specialised intervention, for example if anxiety goes beyond anticipatory anxiety, or if techniques such as distraction or play specialist intervention are no longer effective.¹⁰⁶ There is a developmental component to significant needle anxiety (which can often present in 4–7-year-olds), but this can present at any age either endogenously or related to one or repeated traumatic events. Medical procedures causing trauma in childhood may have lasting impacts, which can in turn affect mental wellbeing and adjustment through later years.¹⁰⁸ Procedural distress can also result in avoidance of CF care and treatments, and it is essential that CF services have robust processes in place to help people with CF manage this. Caregivers may also require help in how to manage their own anxiety and how to model coping. The CF CP can assist teams in ensuring the correct support to manage any type of medical procedure is available. This can, however, be a very time-sensitive activity requiring immediate availability, which can be a challenge in busy services. For more information on supporting with procedural anxiety in CF care, see www.cff.org/managing-cf/procedural-anxiety.

Procedural distress: Further good practice points for the CF CP

- Work closely with CF MDT colleagues, especially nursing and medical colleagues, to help them prevent and manage procedural distress in CF outpatient and ward settings.
- Help the CF team ensure that previous trauma is recognised, that trauma-informed care is provided, and that re-traumatisation is avoided during medical procedures.
- Help ensure the CF team has psychologically informed protocols in place for supporting people with CF and caregivers with managing procedures, including accessing appropriate services such as hospital-based play specialists and developing individual hospital passports where required.
- Offer specialised psychological interventions to people with CF and caregivers around procedural distress, for example needle phobia, when required, including individualised formulation of difficulties and evidence-based interventions and joint work with CF colleagues.

8.2.5 Managing CF admissions and CF outpatient care

Delivery of CF care is undergoing a period of change for many with the introduction of CFTR modulators. This is resulting in reduced CF admissions and clinic attendances. The COVID-19 pandemic also impacted on delivery of care, with a significant increase in remote monitoring and home-based care or remote consultations. However, children and adults with CF may continue to find aspects of inpatient and/or outpatient care challenging, disruptive or stressful. This may depend on factors including age, severity of CF and the level of care required, home and social circumstances, adaptation and adjustment to CF, and individual and family differences in coping with these challenges. Supporting people with CF and families during inpatient admissions is likely to be largely the remit of CF nursing teams and/or CF social workers. However, the CF CP may also have a specific role, for example in helping people with CF or their families manage significant anxiety about hospital admissions or psychological issues that impact on accessing CF care in general.

Managing CF admissions and outpatient care: Further good practice points for the CF CP

- Help the CF MDT understand the difficulties when people with CF or their caregivers are struggling to manage routine CF care, including outpatient attendance and inpatient admissions.
- Support MDTs with psychologically informed strategies, (e.g. including 'behavioural contracts' if required) where there are barriers to accessing CF care in the usual ways.
- Provide individual assessment, formulation and intervention where required, for example where fear around hospital admission or avoidance of appointments reduces access to care.

8.2.6 Psychological aspects of nutritional, eating and gastrointestinal issues

8.2.6.1 Nutrition, weight management, disordered eating and body image issues

Nutritional issues in CF care are at a time of significant change, with CFTR modulators further influencing weight and BMI for many. CF teams and CF CPs therefore now need new strategies to help people with CF in this area of health.

Previous research in this area has been carried out when difficulty in gaining and maintaining sufficient weight was the main issue for people with CF and CF teams. Parents of early-years children with CF have reported feeding issues, for example concern that the child is not eating enough or is eating a rigid range of food types, and advice for parents has been available.¹⁰⁹ Following specific nutritional advice and interventions may however impact on the relationship a person has with food and eating at any stage of life and may interfere with the development of healthy eating attitudes or behaviours.

Previous research has also indicated a slightly increased rate of disordered attitudes or behaviours related to eating in adolescents with CF compared to community norms¹¹⁰ and a CF-specific measure was developed to allow accurate assessment of eating attitudes and behaviours in people with CF.⁸¹

Data on body image in people with CF in the same period has suggested females with CF had better body image (in terms of weight) than males with CF, but that preference for low weight by females could compromise physical health.¹¹¹ Poor body

image (whether related to being underweight or overweight) may impact significantly on mental wellbeing, and the CF CP will have a major role to play in such instances. CF can also result in other visible differences in appearance – see [Cystic Fibrosis Trust’s website for more information](#).

Issues associated with being underweight may persist for some people with CF, and services must continue to meet the needs of these individuals and families while also developing new interventions for the new nutrition-related issues. There is now also an immediate need for understanding of the issues associated with unwanted weight gain and obesity in CF, and for delivery of psychologically informed interventions. For a review of changes in the field of nutrition in CF, including some psychological consequences of undesired weight gain and strategies for the CF team in this area, see Bailey et al. (2022).¹¹² While weight and BMI continue to be key clinical outcome measures, there is also an increasing emphasis on measures such as body composition, and psychologically informed goals with regards to nutritional status should focus on patient’s goals around functionality, strength, fitness and body shape in addition to weight. See Gabel et al. (2022)¹¹³ for more information on this and approaches to weight management work including motivational interviewing.

For those people with CF unhappy with weight gain for any reason, adherence to some treatments, for example PERT and/or insulin, may be reduced to achieve weight loss. This may then impact on GI symptoms, diabetes control or overall health. New weight-related body image concerns may become increasingly prevalent. People with CF who want to lose weight need to be supported to do so in safe and effective ways, with the line between dieting and disordered eating being carefully considered. It will be particularly important to look for early warning signs for relapse in those with a history of eating disordered behaviour. There are recent developments around achieving healthy nutrition, such as the use of mindful eating approaches, including a recent study with people with CF.¹¹⁴

Psychologically informed care is essential in the area of weight management. Difficulties in gaining or losing weight may be maintained by factors including mood disorders, anxiety, complex emotions such as shame, interpersonal dynamics, social factors and issues with readiness for change. CF CPs have important roles to play in helping people with CF and CF teams understand nutritional issues, advising on interventions, and liaising with specialist eating disorder services where appropriate. The CF CP may work on nutritional issues on an individual basis with people with CF. They may also work in close collaboration with CF MDT colleagues, including joint sessions,

and should be involved in service development and direct work on this issue from an early stage.

8.2.6.2 Gastrointestinal (GI) problems

Gastrointestinal (GI) difficulties are common in the CF population, with 65% of patients having significant GI symptoms,¹¹⁵ and are of high concern to the CF community, including professionals, patients and parents.²⁶ Symptoms in CF may overlap with those of other GI disorders, for example irritable bowel syndrome (IBS). Understanding the brain-gut relationship will be important for people with CF and CF teams in managing some common GI issues. There is good evidence that cognitive behavioural therapy (CBT) can reduce symptoms and improve quality of life in IBS.^{116, 117} The CF CP has a key role in supporting both people with CF and CF teams with psychologically informed management of GI difficulties where required.

Managing psychological aspects of nutritional and gastrointestinal issues through the lifespan: Further good practice points for the CF CP

- Help the CF team manage the way routine nutritional advice and interventions are delivered throughout the CF lifespan, to minimise any unintended negative psychological impacts of routine CF care in this psychologically complex area.
- Support the team in delivering psychologically informed interventions around behaviour change for weight management (both weight gain or difficulty achieving a healthy weight) through individual work where indicated, consultation and joint working with CF team colleagues.
- Offer psychologically informed formulation and interventions for complex issues around eating behaviours or disordered eating and issues with body image (including body dysmorphia), through both individual work where indicated and through consultation and joint working with CF team colleagues.
- Consult, liaise with, and make referrals to specialist eating disorder services on behalf of the CF team when more specialist assessment or intervention by these services is required.
- Help individual patients and CF teams understand the bidirectional gut-brain axis through education and consultation.

- Offer psychologically informed formulation and interventions for managing troublesome GI issues through both individual work where indicated, and consultation/joint working with CF team colleagues.

8.2.7 Managing cystic fibrosis diabetes (CFD)

Over 33% of people with CF aged 16 and over are now on treatment for cystic fibrosis diabetes (CFD).¹⁵ The diagnosis of this complication of CF can require significant changes to an individual's lifestyle, with the potential of a heavy treatment burden.¹¹⁸ All children with CF should have an annual diabetes screen from age 10 onwards,³ and there may be anxiety about required blood tests or the need for insulin if diabetes is diagnosed in children. CFD diagnosis frequently occurs around the age of 21, at a time of other important life transitions. For some, the diagnosis of CFD may represent a worsening of their condition and contribute to the development of health-related anxieties. Meta-analyses of the prevalence of psychological distress in type 1 and 2 diabetes indicate that rates of comorbid depression and anxiety are higher than the general population,^{119, 120} and psychological distress is elevated across adolescent and adult CF populations in general.³³ See Cystic Fibrosis Trust's [new guidance on the management of CFD](#) and a [factsheet on cystic fibrosis diabetes](#).

Various models of intervention, including cognitive, psychodynamic and systemic therapies, have been suggested to be effective for people with type 1 and type 2 diabetes^{121, 122} but are yet to be evaluated within CFD. Poor adherence to insulin treatment is prevalent in people with diabetes in general and is influenced by both pragmatic factors (managing diet and remembering insulin), and also complex psychosocial factors (such as experiential avoidance of unwanted cognitions).^{123, 124} Difficulties with diabetes management have been linked to reductions in cognitive functioning; and resultant difficulties with memory, retention of information, and planning can potentially further contribute to poor insulin management and adherence.^{125, 126, 127} Impaired cognition, including difficulties with executive function, has also been reported in people with CFD.¹²⁸

Managing cystic fibrosis diabetes (CFD): Further good practice points for the CF CP

- Support the CF MDT to provide psychologically informed care to help people with CF manage the diagnosis of

CFD (including normalising of difficult emotions while adjusting to this) and to deliver education about CFD (taking into account learning style and any factors affecting learning on this complex issue).

- Support the CF MDT to provide psychologically informed care to help people with CF manage treatments for CFD including venepuncture for screening or treatment, as required.
- Work directly with people with CF to deliver psychological interventions around issues relating to management of CFD, both individually and in joint sessions with CF MDT colleagues.
- Consider inclusion of screening for psychological distress in relation to CFD at CF annual review to help proactively identify any concerns.
- Assess for cognitive difficulties if required, or refer for specialist assessment if needed, and provide consultation to the wider CF MDT about how to adapt care planning accordingly.

8.2.8 Managing secondary diagnoses/ complications of CF

The UK Cystic Fibrosis Registry 2022 Annual Data Report, September 2023¹⁵ lists 42 secondary diagnoses/complications of CF, and throughout the lifespan people with CF may experience other diagnoses that may be related, directly or indirectly, or unrelated to their CF. These include CF liver disease, renal problems, GI issues, musculoskeletal problems, and cancers. The psychological impact of any significant new diagnoses should be considered by the CF team, and the CF CP may have a key role to play in supporting patients adjusting to these.

Managing psychological aspects of secondary diagnoses in CF: Further good practice points for the CF CP

- Help the team to recognise possible psychological impacts of secondary diagnoses, co-morbidities and complications for people with CF, and to monitor the wellbeing of people with CF who are experiencing these.
- Offer individual psychological assessment and intervention where diagnosis of additional health problems results in significant difficulties with coping or psychological difficulties.

8.2.9 Managing medical trauma

People with CF may experience stressful and sometimes significantly traumatic medical events associated with CF, including distal intestinal obstruction syndrome (DIOS), severe hypoglycaemic attacks, pneumothorax (sometimes requiring a chest drain), minor, moderate or massive haemoptysis (the expectoration of blood), and gastrointestinal bleeds. Medical trauma may result in psychological difficulties such as post-traumatic stress disorder (PTSD), requiring intervention from the CF CP. Traumatic medical events may also have a high risk of recurrence, and can be difficult to manage for people with CF, families and CF teams. For emerging themes and symptoms of medical trauma in CF specifically, see Cuneo et al (2023).¹²⁹

Managing psychological aspects of medical trauma: Further good practice points for the CF CP

- Help the CF team be aware of the possible psychological consequences of any traumatic medical events (for people with CF, their family members/caregivers and CF staff).
- Help ensure that people with CF who have experienced significant medical trauma are appropriately screened or monitored for post-trauma reactions or difficulties with adjustment, and provide psychoeducation to people with CF and caregivers about the symptoms and reactions that might be expected following a trauma (medical or non-medical), as required.
- Offer psychological assessment and intervention as appropriate when significant psychological impacts are identified, or where there is opportunity for prevention of such difficulties.
- Refer to specialist services if PTSD is identified and if the CF clinical psychology service is unable to provide specialised trauma-focused psychological interventions.¹³⁰

8.2.10 Managing invasive CF treatments and environments

Some people with CF will need to adjust to increasingly visible and intrusive CF treatments and interventions, for example the insertion of portacaths and gastrostomies, and the long-term need for supplemental oxygen and non-

invasive ventilation (NIV). Adjustment to these more intrusive treatments, and especially those that may be associated with progression of the condition, may be psychologically challenging. Some people with CF may also need to undergo care in intensive care settings (for example during periods of critical illness and post-transplant surgery). Such environments may be challenging, and management of trauma associated with invasive procedures may be needed. See Murray et al. (2020)¹³¹ for detail on therapeutic work after intensive care unit (ICU) care, NICE guidelines for guidance for PTSD¹³⁰ and Section 8.2.13 for more information about psychological care of people with CF peri- and post-transplant.

Managing invasive CF treatments: Further good practice points for the CF CP

- Help the CF team support people with CF with adjustment to more visible or intrusive treatments in routine care, for example through normalisation of difficult emotions and support during adaptation to such changes.
- Provide individualised assessment, formulation, and intervention where direct work is required, to help people with CF adapt to increasingly intrusive or emotionally challenging CF treatments.
- Be aware of local arrangements for supporting people with CF when they are admitted to intensive care environments and provide psychological intervention and support in that setting as appropriate, depending on local protocols.

8.2.11 Managing persistent physical symptoms (PPS) or medically unexplained symptoms (MUS)

PPS (or other symptoms of poorly understood syndromes) and MUS (i.e. physical symptoms that lack an identifiable organic cause) can include a very wide range of symptoms, including unexplained bodily pain, headache, functional neurological symptoms, GI symptoms, chronic fatigue, and breathlessness/dyspnoea. MUS and PPS are common in the healthcare system in general, accounting for up to 45% of all GP appointments, and with 50% of patients in secondary care having no clear diagnosis at three months.¹³² For information for health professionals on working with MUS, see [Guidance for health professionals on medically unexplained symptoms \(MUS\)](#).

While the prevalence and nature of MUS and PPS in people with CF is unknown, management of these issues in CF is likely to be especially complex, requiring careful understanding and management by the whole CF team. Management of multifactorial breathlessness in CF may be especially complex, and CF CPs will need to work closely with CF team colleagues including physiotherapists around issues such as breathing pattern disorder and associated difficulties. Communication about MUS and PPS with people with CF, and the management of investigations and interventions, requires high levels of MDT skill to ensure psychologically informed care.

MUS or PPS in children with CF would usually be reported to the CF team by caregivers rather than by the child directly. This gives an additional dimension, including the possibility of over-reporting of symptoms due to for example hypervigilance or anxiety, which may also need to be considered by the CF team. MUS and PPS in children require a thorough assessment of the child or young person's wellbeing, as well as parental emotional functioning, and liaison with the CF SW as required.

Managing medically unexplained symptoms (MUS) and persistent physical symptoms (PPS) in CF care: Further good practice points for the CF CP

- Help the CF MDT in understanding, communicating about and managing MUS and PPS collaboratively with people with CF and their caregivers, for example through education, training, consultation and joint working with the CF MDT as required.
- Provide individual assessment, formulation and intervention work to help people with CF manage MUS and PPS, for example if causing distress or negative impacts on function, taking account of the complexities that can arise in the context of CF care.
- Be aware of the potential for safeguarding issues in children when MUS are being investigated (depending on age of the child) and liaise closely with CF colleagues, especially the CF SW.

8.2.12 Managing CF with neurodivergence, neurodevelopmental conditions or other additional needs

The terms neurodivergence, neurodevelopmental disorders and additional needs are used here to encompass a range of possible co-morbid

diagnoses or difficulties experienced by some people with CF, including: learning disability, attention deficit hyperactivity disorder (ADHD), autistic spectrum conditions (ASC), dyslexia, dyspraxia, specific learning difficulties, physical disabilities, sensory difficulties including deafness, and communication difficulties.

Approximately 1.5 million people in the UK have a learning disability, and this is associated with health inequalities and poorer outcomes in both physical and mental health. See, for example, [information from Mencap on health inequalities](#).

Prevalence of neurodivergence or specific learning difficulties in the general UK population is difficult to state at present. Factors include much raised awareness in recent years, rapidly increasing rates of diagnosis in some areas, unmet need for specialist assessments, and possible under-recognition of issues in some sections of the population. For more information, see, for example, resources from [ADHD Aware](#) and the [National Autistic Society](#).

The prevalence and impact of learning disability, neurodivergence, neurodevelopmental disorders and other additional needs in people with CF specifically are as yet under-investigated. There is some data on ADHD, with elevated rates of possible symptoms in people with CF reported compared to the general population.^{133, 134} Clinical implications of this for people with CF and for CF services, and possible causal mechanisms, are outlined in Lemiere & Havermans (2018).¹³⁵ Rates of autistic spectrum conditions within CF specifically are not currently known, but challenges for children with CF and autism have been described.¹³⁶ More research is needed to identify prevalence of all forms of additional needs in people with CF, and to ensure UK CF services are aware of such needs and able to adapt services accordingly.

Clinical experience indicates that for people with CF, managing additional needs including neurodivergence alongside CF can present challenges, with possible impacts on: managing information about CF and healthcare, managing CF consultations, appointments and admissions, treatment adherence, emotional wellbeing and quality of life.

All healthcare organisations, including CF teams have a legal duty to adapt their care provision with reasonable adjustments to ensure that their services are accessible to people with physical or sensory disabilities, learning disabilities, or other long-term conditions such as dementia.¹³⁷ Reasonable adjustments are person-centred changes to usual pathways of care. They may include providing shorter or longer appointments, providing easy-read materials, the use of handheld

personal documents such as one-page profiles or hospital passports, and communication via an interpreter as required. [NHS England has more information.](#)

CF CPs may take a lead role in offering screening and making referrals for specialist assessment to people with CF presenting with possible undiagnosed neurodivergence (or other additional needs), may advise other members of the CF team in this area, or may signpost via the GP for referrals. Similarly, CF CPs may support the team in ensuring the needs of those who are neurodivergent or have other additional needs are taken into account in CF care generally. CF CPs may also need to use adapted measures in their own assessment and intervention with people with CF, including, for example: Glasgow Anxiety Scale for people with an Intellectual Disability (GAS-ID), Glasgow Depression Scale for people with an Intellectual Disability (GAD-ID), and adapted versions of the PHQ-9 and GAD-7 measures for people with learning difficulties or disabilities.

Managing CF with neurodivergence or additional needs: Further good practice points for the CF CP

- Be familiar with NHS/hospital trust policies and procedures relevant to supporting patients with additional needs in healthcare and hospital settings.
- Help ensure equitable access to CF services for patients with neurodivergence or additional needs, by supporting and advising the CF MDT in making reasonable adjustments to CF service delivery, involving the individual and caregivers as appropriate, and contributing to training in this area.
- Carry out limited neuropsychological testing in some cases, where there are the necessary competencies, for example administering the Weschler Intelligence scale for Children (WISC), with referral to specialist assessment services as required.
- Support the CF MDT with conducting capacity assessments and best-interest decision-making when indicated.
- Use screening tools as appropriate when referring for specialist assessment for neurodivergence, for example in adult services the [ASRS for ADHD](#)¹³⁸ and the [AQ10 for autistic spectrum conditions](#).¹³⁹
- Support the CF team in making referrals for specialist testing and diagnosis, for example for learning disability or neurodivergence when required, and with liaison with

specialist teams and other services in the community, where appropriate.

- Use measures for psychological assessment that have been adapted for use by those with intellectual disability, if required.
- Adapt individual therapeutic work with people with CF as needed to make psychological assessment and interventions equally accessible to all.

8.2.13 Managing lung transplantation

In 2019, in the UK, 49 people with CF aged 16 or over received bilateral lung transplants. Numbers of lung transplants were then much reduced (to 12) in 2020 (possibly largely due to the COVID-19 pandemic), and to less than 5 in both 2021 and 2022.^{15, 140} Future rates now remain to be seen. The introduction of CFTR modulators seems likely to result in a reduction in the number of referrals for lung transplantation and increased age at referral for those able to access CFTR modulators. However, lung transplantation will remain a necessary treatment option, with the CF CP as well as transplant services psychologists having key roles in supporting people with CF and families through this process.

For a summary of information relating to lung transplantation for people with CF, see Bell et al. (2020).¹⁶ For a summary of issues related to paediatric lung transplant (and end-of-life-care) in cystic fibrosis, see Dellon et al. (2017).¹⁴¹ For Cystic Fibrosis Foundation (CFF) consensus guidelines on lung transplant referral for people with CF, see Ramos et al. (2019a).¹⁴² For consensus statements for the care of CF lung transplant recipients including screening for mental health issues in both transplant recipients and caregivers, see Shah et al. (2021).¹⁴³ For further summaries of issues, including decision-making about transplant, delays in referral, and barriers to accepting referral for transplantation, see Daniels et al (2017),¹⁴⁴ Robertson & MacDonald (2017),¹⁴⁵ and Ramos et al. (2019b).¹⁴⁶

For information about lung transplant eligibility, assessment, risks and contraindications in general, see Leard et al. (2021).^{147, 148} Previous or current significant psychological or psychiatric difficulties can potentially impact an individual's ability to successfully cope with a transplant, so access to help with such difficulties is essential.

Helping people with CF maintain hope and countering demoralisation or exhaustion are important for those awaiting transplant, and interventions for managing this stressful situation should be part of pre-transplant care.¹⁰ The process

of lung transplant may involve traumatic elements for people with CF, including difficult experiences in ICU, unexpected complications, and the use of extracorporeal membrane oxygenation (ECMO) pre- or post-transplant. See also Section 8.2.10 for more detail about supporting people after ICU care.

While lung transplantation can offer people with CF significant improvements in quality of life, the process of adjustment may involve a range of emotional experiences, including feelings of guilt, increased anxiety, and difficulties adjusting to unfamiliarity.^{149, 150, 151} See also Rosenberger et al. (2012)¹⁵² for a review of psychosocial issues faced by lung transplant recipients. See [useful resources about transplantation for people with CF and their relatives](#) on the CF Trust website.

Provision, site and availability of CF-related and psychological care post-transplant currently varies in different areas of the UK. This can depend partly on proximity of the transplant centre to the patient's home and local agreements between the CF and transplant teams about levels of care offered by each. Ensuring that appropriate psychology input is available to people post-transplant should be a shared responsibility between transplant and CF centres, and any gaps in such care will need to be addressed.

The CF CP has key roles to play in supporting people with CF and families through the transplant process alongside MDT colleagues, including around decision-making, at assessment, during 'prehab' work, with coping while on a transplant waiting list, and, in some cases, post-transplant. Close liaison between CF and transplant psychologists is the expected norm, and a template for sharing information between these services at the assessment stage is available (see Appendix 7).

Lung transplantation for people with CF: Further good practice points for the CF CP

- Offer resources, including support, education and a reflective space for people with CF (and caregivers, where appropriate), to help with decision-making around transplant.
- Use a 'prehab' model along with CF and transplant team colleagues to help optimise health management and psychological readiness for transplant.
- Offer direct assessment and psychological intervention to people with CF and also consultation to the CF team as required, to help people manage the complex process of transplant at any stage.

- Offer the opportunity for advanced care planning while preparing for transplant (if this is not the specific role of others in the CF team). See 8.2.14 for more detail on end-of-life-care in CF.
- Make links with and liaise with transplant CPs at the appropriate transplant centre(s) to coordinate care pathways and share information relating to transplant assessment and psychological care pre-, peri- and post-transplant.
- Support the process of transition from one care provider to another through working collaboratively with CF and transplant team colleagues, for example by sharing information about needs, preferences and coping strategies used that would be useful post-transplant.
- Liaise closely with the transplant psychologist to agree the best pathways for psychological support and intervention during extracorporeal membrane oxygenation (ECMO) bridge to lung transplant treatment if this is available locally.

8.2.14 Palliative care, advanced stages of the condition, and end-of-life care

Palliative care is relevant at all stages of CF when understood as an approach to managing difficult symptoms.¹⁵³ Difficult symptoms, for example pain and fatigue, may be under-reported by those with CF and anecdotally there tends to be recognition of a high degree of stoicism and acceptance of symptoms in this population. Unmet needs for palliative care in people with CF have been reported,¹⁵⁴ and a screening and triage programme to identify palliative care needs has been described.⁷⁵ Consensus guidelines on delivering palliative care to individuals with CF are available.¹⁵⁵ Misunderstandings and concerns about the meaning of palliative care and resulting barriers to accessing this may need to be addressed, for example 'to dissuade patients and CF care providers of the notion that PC means abandonment of standard care'.¹⁵⁶

Life expectancy with CF has improved dramatically over recent decades, with a median predicted survival age of 56.1 years and median age of death of 33 years in 2022.¹⁵ With the availability of new CFTR modulators, the outlook for many with CF is now hoped to improve significantly further. However, how end-of-life-care may be impacted, for example by changing survival or cause of death for people with CF, is still unclear, and advanced stages of the condition and end-of-life care will

remain part of CF services, mostly in adult centres. The CF CP will continue to have a key role at this stage of care, including, for example, work on emotional adjustment, symptom management including breathlessness, life review work, and work with anticipatory grief.

Care at advanced stages of CF requires specific and sensitive issues to be raised by the CF team. CF has some specific challenges compared to end-of-life-care in some other specialties, for example related to: the long relationships formed between CF teams and people with CF and their families, the possibility of lung transplantation for some, and the natural course of advanced CF with severe exacerbations with subsequent recovery. Consensus guidelines for the care of individuals with advanced lung disease are available, including the need for screening emotional health at this stage of life with CF.¹⁵⁷

End-of-life care plans should be in place for all expected deaths within CF care as in other specialties. There is however a lack of training, knowledge and confidence in CF practitioners about both palliative and end-of-life care.^{158, 159, 160} A CF-specific palliative care training programme was shown to have increased staff 'comfort with core skills'.¹⁶¹ Wide variation in the offer of advance care planning (ACP) by CF teams at different centres has also been reported, with a recognition that barriers to this need to be overcome.¹⁶² See CF Trust [information on planning for end of life](#) for a CF-specific ACP document and guidance, and for resources for people with CF, families and teams about end-of-life-care in CF.

Great variation in how specialist palliative care services are integrated into CF teams has also been reported in the UK,¹⁶³ but an integrated palliative-care model has been shown to be beneficial to patients, CF teams and the palliative care team.¹⁶⁴ Training in breaking bad news and advanced communication training has been developed to enhance staff skills, for example.¹⁶⁵ Such continuing professional development (CPD) may be useful for CF teams in managing this challenging area of work.

Psychological and emotional support of patients at the end-of-life stage will be done by various members of the CF team. CF CPs can support the work of other team members at this stage of care, and will work closely with MDT colleagues, for example through consultation, joint working and case discussion, as well as working directly with people with CF.

Death of a person with CF will impact on a range of people, including parents, siblings, adult partners, children, peers with CF and also the CF team. Self-care within the CF team is important,

through appropriate management, and team and hospital trust structures and processes for staff support. Alongside other senior members of the MDT, CF CPs can help to facilitate reflective spaces as one way of optimising self-care for the CF team after a CF death. This also gives an opportunity to review end-of-life care, learn from events and further develop the centre's end-of-life care as a result. Analysis and discussion of patient deaths by a staff team has been developed in other specialties, for example paediatric intensive care. MDT review of practice, and support of the CF team following a patient death, are recommended in CF care.^{1, 10} Teams can find their own language and style for such sessions, supported by the CF CP as appropriate. Useful approaches could include narrative and/or systemic therapy, such as 're-membering conversations'.^{166, 167} However, decisions about who facilitates these sessions should be made on a case-by-case basis, and it may not be appropriate for an embedded CF psychologist to facilitate a session for a patient that they also worked closely with, to allow them to take part in the process in the same way as the rest of the CF team. Hospital trusts may also provide support with running such sessions, for example through staff emotional wellbeing services.

Palliative care, advanced stages of the condition and end-of-life-care for those with CF: Further good practice points for the CF CP

- Help the CF team to educate and provide information to people with CF about palliative and end of life care, and to ensure the needs of those within CF services are met in this respect.
- Develop shared pathways with the CF team for psychologically informed management of complex medical issues, such as pain and fatigue, where required.
- Work closely with the CF MDT, for example through consultation and teaching/training around psychological aspects of palliative and end-of-life care.
- Be aware of different roles in the CF MDT at the stage of end-of-life-care, with clear understanding of own roles, and help the team ensure that care provided by different members of the team is well-integrated.
- Provide individual psychological assessment and intervention as appropriate for people with CF with advanced stages of the condition, for example for anticipatory grief, managing distress, managing difficult symptoms, and preparing for the end of life.

- Liaise as appropriate with other agencies, such as hospital palliative care teams and community palliative care services including hospice services.
- Carry out ACP work with people with CF where appropriate (if this is not the specific role of others in the CF team).
- Establish who will support carers and families of those with CF at end of life, especially with respect to the children of those with CF.
- Take an appropriate role in any reflective process and meetings in the CF team following the death of people with CF and lead these where required.
- Help the CF team identify who in the CF MDT will provide any support to bereaved families (and other patients or parents in the service if required, for example when they know the patient who has died), including signposting to other services.
- Work with team leads and directors of services to ensure that staff education and support is in place for this potentially challenging area of care, and support the CF team and managers in building good team self-care strategies.
- Ensure own self-care is in place for this potentially challenging area of work.

8.3 Managing adherence to treatments in CF: Key issues and roles of the CF CP

8.3.1 Managing adherence to treatments in CF: Background

NB the term 'adherence' to CF treatments is used throughout these guidelines. This term describes 'the degree to which patient behaviour matches recommendations from the prescriber'.¹⁶⁸ Other terms are in use, including 'compliance', 'persistence', 'co-operation' and 'concordance'.⁴⁶ Different CF teams or team members may use a variety of terms, and this may or may not reflect any actual differences in philosophy or approach to supporting people with CF to manage CF treatments.

Steadily increasing survival for people with CF over recent decades has come with increased burden of treatments, in terms of number of

treatments, complexity and time required.¹⁶⁹ In 2018, simplifying treatment was identified as the top research priority by the international CF community and the sixth (out of 10) priority was 'what effective ways of motivation, support and technologies help people with CF improve and sustain adherence to treatment?'²⁵

More recently, the introduction of CFTR modulators for many, but not all people with CF, is bringing further significant change. Changes in health for some on new treatments may impact on feelings about treatment burden and decision-making about conventional treatments.¹⁷⁰ New data on adherence to both new and existing CF treatment regimes is needed, and work has begun to investigate whether some conventional treatments can be reduced for those on modulators – see Cystic Fibrosis Trust's [website](#). However, it seems likely that the issue of adherence to treatment will remain a key issue for CF care in the foreseeable future, for people with CF and families/caregivers, for CF teams, and for the CF CP, who has specific skills and roles to help people with CF take any treatments in an optimal way.

Adherence to health and lifestyle advice, including to prescribed treatments, is an unresolved challenge in healthcare generally, with the paradox that perfection (100% adherence) is desirable, but that non-adherence is to be expected.¹⁷¹ In developed countries, adherence to long-term therapies in the general population is thought to be around 50 per cent.¹⁷²

Within CF, describing a 'rate of adherence to treatment' is of limited value, with many different treatments involved, possibly all with different levels of adherence. Studies have shown widely varying rates of adherence to different treatments ranging from approximately 30–70%^{173, 174, 175, 176} with real-world, objectively measured adherence lower than that reported in clinical trials.¹⁷⁷ Adherence may also change continuously. Poor adherence to treatment in CF is linked to early deterioration in health¹⁷⁸ and high healthcare costs,¹⁷⁹ lower quality of life and poorer mental health,³⁶ with possible feelings of guilt or inadequacy as a further outcome.

Rates of adherence to new CFTR modulators outside clinical trials are not yet fully understood. Some early data suggested a very high level of prescription uptake of an early modulator treatment,¹⁸⁰ but more work is required. Evidence from other specialties, such as HIV, where there have been significant advances in treatments available,¹⁸¹ do suggest that adherence will remain an issue in CF care, requiring careful attention.

Accurate monitoring and measuring of adherence to CF treatments is an essential task for the CF

team but is complex. When monitoring adherence, a multimethod approach that includes both subjective and objective measures, preferably by several team members sharing information, is important, as each method has advantages and drawbacks.¹⁷⁵

The most effective approaches to enhancing adherence are not yet clear. A systematic review of interventions for enhancing medication adherence across conditions¹⁸² concluded that interventions studied were mostly complex and not very effective, with more research needed to identify effective strategies. Within CF, evidence for effective interventions, including those for adherence, was generally found to be insufficient,⁸⁸ and a problem-solving intervention with adolescents was not found to improve adherence.¹⁸³ A recent UK trial of an intervention to support adherence to nebuliser use in CF showed no reduction in exacerbation rates, but an increase in objectively measured adherence and lower perceived treatment burden.¹⁸⁴ A survey of US CF centres concluded that quality improvement projects to increase adherence promotion work would be useful in CF care.¹⁸⁵

The approach to adherence in CF services can differ somewhat in paediatric care compared to adult services. In the former, parents are usually in charge of CF treatments for many years and have a responsibility to deliver treatments as prescribed with the child patient having gradually increased agency in adherence and agreeing to treatment. In paediatrics, non-adherence to CF treatments by parents can be a child protection issue, but this is a psychologically and ethically complex issue. The illness representation model and ethical frameworks can be applied to inform clinician decision-making on how to manage cases where parents do not provide optimal CF care.¹⁸⁶ In addition, children's competence to make decisions about their medical treatments (**Gillick competence**) may also need to be considered where there is uncertainty or ambivalence about a treatment.

Adolescents may particularly struggle with adherence to CF treatments due to changes at this life stage, for example an increased desire to fit in with peers, increased risk-taking and experimentation behaviours, and changing relationships with parents and levels of autonomy. This may cause significant concern for families and CF teams, and the challenge is to maintain relationships and open communication while encouraging adherence to CF care, which can be a difficult balance.

In adult services, the right of an individual with capacity to make even apparently 'unwise choices' about CF treatments is paramount, and there will

be more of an approach of informed decision-making about treatments, including which to prioritise in terms of a balance with quality of life. Decisions to undertreat one's CF may occur frequently¹⁸⁷ and may provoke significant discomfort for some in CF teams and families, but there is a need to help individuals achieve autonomy and support well informed decision-making – even where this may reduce longevity. Rationalisation or cessation of some treatments will also have a specific place in the care of those with advanced CF or at end-of-life-care.

Alongside this 'intentional non-adherence', many caregivers or adults with CF simply struggle to manage all prescribed treatments, and this non-adherence is better viewed as non-intentional. In all settings, adherence to treatments is impacted by a complex mix of treatment, individual, systemic and healthcare characteristics and is best addressed with a team approach involving the people with CF, their parents and carers (where appropriate), and the various members of the CF MDT. It is important that patient reports of simply 'forgetting' to take their treatments are further discussed in light of all of these factors, so that obstacles to adherence can be more fully explored.¹⁸⁸ Further issues in adherence to CF treatments and the roles of the CF CP more specifically are outlined below.

8.3.2 Understanding adherence

Various frameworks and models can be used to help understand (and then intervene with) adherence issues:

- General frameworks for understanding (and intervening with) adherence include the WHO model¹⁷² with five dimensions to help understand wider social and systemic context around adherence, as well as individual factors. These dimensions are socioeconomic status and social support, healthcare team and system-related, patient and carer, condition, and therapy.
- For an approach to understanding adherence in terms of engagement, knowledge, measurement issues and regimen characteristics, see Duff & Oxley (2016).²³
- For an educational initiative to support healthcare professionals in measuring and addressing adherence challenges in CF, including tools and resources, see the **CF Care website**.
- General psychology and health psychology models, including the following, will also aid understanding of adherence behaviour in any condition and may be used by the CF CP:
 - the necessity/concerns framework¹⁸⁹
 - the theory of planned behaviour model¹⁹⁰

- self-regulation models, for example the common-sense model of self-regulation (CSM)⁷²
- the readiness for change model¹⁹¹
- the COM-B model¹⁹²
- systemic models, for example the Family Systems-Illness Model¹⁹³

8.3.3 CF CP approaches to adherence

There is no one evidence-based model of assessment and intervention for addressing suboptimal adherence. However, CF CP formulation of difficulties with adherence enables an integrative multi-model understanding of this, including identifiable barriers, which can guide selection of the best intervention approaches for an individual by the CF CP and the CF team. Barriers are likely to be missed if not actively explored. The CF CP can employ a mix of direct work with people with CF or caregivers (often conducted jointly with CF MDT colleagues), indirect work (for example formulation of adherence issues) with the CF team, and systemic work, depending on a person's needs.

Approaches to working with adherence issues may vary somewhat between adult and paediatric care, for example due to any differences in general approach to adherence, how services are delivered, and the developmental stage of the person with CF. In paediatric care, work may mostly be via parents, with advice given on strategies to try, and how to help their children take treatments optimally. There will be gradual change in the responsibility for treatment becoming held by a young person as they progress into adulthood. This process will also be influenced by wider familial and psychosocial support systems. In adult services, a more direct talking therapy model may be offered, with both emotional health and health psychology models utilised in individual work – for example on beliefs and emotions as well as behaviour change strategies.

CF CP interventions for adherence in paediatric CF care may be with children, young people or caregivers and may be done within routine CF clinics and in joint consultations with CF MDT colleagues. Liaison with schools may also be required to support any treatment-taking there. Children's understanding of CF and treatments depends on their stage of development and this should also be considered in understanding and intervening for adherence to treatments.¹⁷¹

Adults with CF may also lack knowledge or understanding about treatments, depending on the level of education received in paediatric care,

and this should be reassessed upon reaching adult services. Interventions for adherence by the CF CP with adults are likely to be formulation-based, encompassing both individual and wider systemic influences on health behaviour.

Mental health issues at any age may also impact on treatment adherence and thus clinical outcomes. Detailed assessment and formulation of such factors will be essential in understanding and intervening for adherence difficulties in such cases. For example, generalised loss of drive and motivation, high hopelessness in the context of very low mood, or low self-esteem or self-efficacy will all be likely to make optimal treatment adherence very much harder. Where non-adherence to CF treatments constitutes self-neglect, for example due to mental health difficulties rather than informed non-adherence, care may be complex with CF CP involvement especially key.

Assessment tools to enhance understanding of the reasons for non-adherence or partial adherence may be used by the CF CP or the CF team. These may be general measures, for example the beliefs about medicines questionnaire¹⁹⁴ or CF-specific, for example the barriers to adherence in CF assessment.¹⁹⁵

Following assessment and formulation of any issues with adherence, at any age, the CF CP may use a combination of the following approaches in working with caregivers or people with CF.

- Psychoeducation, for example in joint sessions with CF pharmacists to aid knowledge and understanding of CF treatments and approaches to improving adherence.
- Use of behavioural strategies to positively reinforce desired behaviours.
- Practical approaches, including problem-solving and SMART goal setting (ensuring goals are Specific, Measurable, Attainable, Relevant and Time-based).
- Work to identify values, to ensure that when setting goals around CF treatments they are meaningful to the person, and not only the result of wider familial, professional or cultural expectations and pressures.
- Exploration of health beliefs, their role in determining adherence, and the scope to change adherence behaviour by changing certain beliefs.
- Understanding of motivation, factors affecting motivation and interventions designed to optimise this, for example motivational interviewing.^{196, 197}

- Understanding that even when motivation is high, behaviour change may not occur due to other barriers. For example, the COM-B model is a way of understanding behaviour change in the context of capability and opportunity as well as motivation.¹⁹²
- Work on habit formation to make adherence to treatments less effortful.¹⁹⁸
- Psychotherapeutic work, for example on emotional and mental health barriers affecting adherence to treatments, including low mood, anxiety, low self-esteem, low self-efficacy.
- Understanding of cognitive factors and neurodiversity that might impact adherence, for example learning disabilities, ADHD, autistic spectrum conditions.
- Work on any systemic factors affecting adherence, including relationships between a person with CF and caregivers or family and with the CF team.
- Understanding of the role of social factors in adherence, including adversity, poverty and lack of social support (with referral to the CF SW as required).

8.3.4 Supporting CF team interventions for adherence

Much of the rest of the CF MDT will also be engaged in work on adherence with people with CF and caregivers. See for example [Section 13 of the Cystic Fibrosis Trust's Standards of Care of Good Clinical Practice for the Physiotherapy Management of Cystic Fibrosis](#). The CF CP can work alongside their MDT colleagues to help ensure that approaches to adherence are psychologically informed.

Empathy, curiosity and patience should underpin all conversations exploring adherence, by anyone in the CF team, with the aim of trust, openness and honesty. General principles²⁴ include:

- accept that partial adherence is normal
- establish an empathic team approach
- avoid blame or criticism
- provide written copies of treatment plans
- prioritise and simplify regimes, setting realistic goals.

CF teams need to demonstrate empathic understanding of the challenges of CF treatment adherence, balanced with the need to challenge beliefs and behaviours, where appropriate. An understanding of other factors that may affect

adherence, for example neurodiversity, trauma, adversity and socioeconomic factors, is also essential.

For a detailed summary on adherence in CF, including information on strategies to help CF teams (and CF CPs) with adherence, see Duff et al. (2020).¹⁹⁹

In summary, work by the CF CP in the area of adherence may involve a combination of behavioural, cognitive, interpersonal and systemic level understanding and intervention. The CF CP has specific skills and knowledge of evidence-based psychological models of difficulties with adherence to CF treatments, to add to the expert knowledge of the CF team in this area. This can inform the work of the CF CP directly with people with CF and caregivers, and also the work of the rest of the CF MDT.

8.3.5 Managing adherence to CF treatments: Core roles of the CF CP and further good practice points

8.3.5.1 Adherence to CF treatments: Core roles of the CF CP

It is recommended that the CF CP should provide specialised psychological input around adherence to CF treatments through:

- Direct work with people with CF or their caregivers on adherence issues, using a variety of evidence-based psychological models, theories and approaches.
- Supporting the work of the CF MDT around adherence issues, for example through joint work, consultation, teaching/training, and service improvement.

8.3.5.2 Managing adherence to treatment in CF: Further good practice points for the CF CP

- Help the CF MDT understand and formulate adherence issues (including for people with CF/families they are not seeing directly), using a biopsychosocial model and psychologically informed understanding of adherence from different theoretical perspectives.
- Help the CF team understand adherence difficulties in the context of any neurodiversity, other neurodevelopmental issues and/or learning disability, and advise on adaptations to care as needed.

- Help the CF team assess, monitor and address issues around adherence to all treatments including CFTR modulators, through participation in CF clinics (especially in paediatric care), joint sessions with CF MDT colleagues, and consultation.
- Contribute to quality improvements, audit and research in the area of adherence to CF treatments.
- Understand the complex issues of informed non-adherence versus child protection issues in paediatric care (and work closely with the CF team, including the CF SW, in such complex cases).
- Support teams to consider capacity issues when thinking through informed non-adherence versus self-neglect in adults.

9. CF and relationships: Key issues and roles of the CF CP

9.1 CF and relationships: Background

Relationships and their quality are central to psychological health and wellbeing, and to the quality of CF care. Recent events, including the COVID-19 pandemic and the introduction of new CFTR modulators for some people with CF, may also impact on relationships. For example, shielding during the pandemic led to loss of support and isolation for some; treatment advances are resulting in reduced contact with the CF team for many people with CF; and there is expected to be a significant increase in the number of children born to people with CF. The CF CP has a key role to play in helping people with CF, their caregivers and the CF team manage relationships in an optimal way.

9.1.1 CF and relationships: Core roles of the CF CP

It is recommended that the CF CP should:

Provide specialised psychological assessment and interventions, and support the work of the CF team as required, including in the following areas.

- The CF team and relationships.
- Parenting children with CF.
- Relationships for children and young people with CF.
- Relationships for adults with CF including:
 - sexual and reproductive health
 - parenting as an adult with CF
 - relationships with partners, peers and parents.

See the following sections for further good practice points for CF CPs.

9.2 The CF team and relationships: Key issues and roles of the CF CP

Healthy relationships within the CF team, and between the CF team and people with CF and their caregivers and families, are key to good care. Relationships between services, for example in shared CF care, between paediatric and adult CF services, and with other voluntary or statutory organisations involved in care, will also be important in determining quality of care. NHS England has more information on optimising [team interactions](#) and [staff wellbeing](#).

The relationships that a CF team develops with people with CF, and with their caregivers, is a complex dynamic developed over years. The initial relationship – usually between the CF team and parents – must be established in a positive way from the outset. The CF CP has an important role to play in promoting this, for example by introducing themselves to parents and offering the opportunity to share reflections about their previous relationship to healthcare services, family and others.²⁰⁰ The relationship is then developed with the child/young person with CF as they grow older.

Long relationships between CF teams and people with CF and their families mean that CF work can be particularly emotive, as teams offer support through major health events and transitions. Consistency of care and clear boundaries are important to fostering realistic expectations and maintaining good therapeutic alliances. The need for advanced communication skills in CF teams has been identified, and strategies to optimise these in CF services have been described.²⁰¹

Involvement of people with CF and their caregivers in the design and improvement of CF services, with expert patient initiatives and public and patient involvement (PPI) work, will also help foster good working relationships and enable the ‘voice’ of people with CF to be heard in services.

Using their systemic perspective – recognising the influence and impact of multiple systems and influences around the team, patient and family, and using formulation to understand how these shape experiences and drive behaviour, the CF CP can help optimise those relationships that are key to quality of life, emotional health and effective management of CF. See also Section 6 on indirect care by the CF CP.

9.2.1 The CF team and relationships: Further good practice points for the CF CP

- Help the CF team achieve healthy team functioning, for example through facilitating reflection, consultation, or supervision around any team difficulties where appropriate, and encouraging high levels of support and compassionate leadership within different parts of the CF team.
- Promote healthy relationships between people with CF/caregivers and the CF team through work with individuals, families and the team as required.
- Help the team ensure that reasonable adjustments are made for people with CF with additional needs that may impact on communication or relationship-building with the CF team.
- Help the CF team understand complex patient presentations, including perceived difficult behaviours, and any impacts on the staff team, relationships, or care.
- Offer and contribute to specific forums for the CF team, such as reflective practice sessions, team meetings, critical incident support and discussions after patient deaths or other difficult events.
- Offer consultation and training in areas such as communication skills, trauma-informed care, having difficult conversations, and understanding the dynamics of healthcare professional/patient relationships.
- Foster effective working between the CF team and other services, for example during shared care, transition to other services and liaison with community and statutory services.
- Facilitate involvement of people with CF and caregivers in CF service developments and improvements.

9.3 Parenting a child with CF: Key issues and roles of the CF CP

Parenting a child with a long-term, life-limiting condition can bring challenges (see also appendix 5). Research indicates a high subjective care burden perceived by parents, increasing with the child's age, infection with *Pseudomonas aeruginosa* and/or other raised morbidity.²⁰² Thresholds have also been described, when the 'chronic sorrow'

experienced by parents of children diagnosed with CF is heightened, including at diagnosis of CF, acquisition of pseudomonas, first exacerbation and increase in treatment burden.^{203, 204} A primary task for parents is adapting to the diagnosis of CF in their child at whatever age this occurs (see Section 8.2.1) and this event may also impact on parental relationships or family dynamics. Cultural influences may also determine how parents incorporate CF into family life.

Understanding the impact of a condition such as CF on parenting and vice versa is essential, and CF CPs have a key role to play in this. Support for parents around their relationships with family and friends in the context of having a child with CF, for example by helping family members understand the illness and what it means for the person with CF and family life, may be needed. Parents also need to help their child with CF (and siblings) develop their own understanding of their condition, to adjust to and communicate about CF and to develop a relationship with CF that incorporates it into their life, but is not all-consuming. Much of the work needed to support parents of children with CF will be around positive parenting – for detailed guidance including tips for parents, see Bryon & Titman (2019).¹⁰⁹ CPs may also signpost parents to many other parenting books they might find useful, for example Siegel & Bryson (2012).²⁰⁵

9.3.1 Parenting a child with CF: Further good practice points for the CF CP

- Support the team in helping parents develop optimal relationships with their child with CF, for example through presence in CF clinics, recognition of any difficulties in this area, and ensuring early intervention and increased support by the team and the CF CP where required.
- Help parents optimise their own relationships and support them with communication with family and friends about CF, and any cultural aspects of CF and CF management.
- Offer a reflective space to parents to consider the impacts of diagnosis of CF and parenting a child with a long-term condition (as well as signposting to any other services required).
- Promote positive parenting, through advice and education to parents in routine CF consultations, to help optimise their relationships with their children with CF and their siblings.

- Monitor the emotional wellbeing of parents of patients from the point of diagnosis of CF, and promote healthy parental emotional wellbeing.
- Be aware of guidance from the International Committee on Mental Health in Cystic Fibrosis (ICMH) on screening and assessment of caregivers of children with CF, and apply as appropriate locally.
- Offer individualised assessment and intervention to families around parenting issues as required.
- Support parents with the interface between statutory and non-statutory services offering input to them and their children.

9.4 Relationships for children and young people with CF: Key issues and roles of the CF CP

As a child with CF develops, some key tasks are: to negotiate ordinary relationships; to adjust to a growing understanding of CF; to develop a healthy relationship with CF and self-identity; and to acknowledge that CF is a significant part, but not all of this. Parents and other adults can model a 'coping' relationship with CF to promote the child's own relationship with CF. For summaries on promoting emotional wellness in children with CF, and assessment and intervention strategies, see Prieur et al. (2021)⁴³ and Georgiopoulos et al. (2021).⁴⁴

At adolescence, a person with CF may experience change in relationships with parents and CF care teams. Understanding developmental stages and any other factors impacting an adolescent's relationships in CF is essential, for example any additional communication/intellectual difficulties or concurrent social and contextual challenges. The CF CP can help the team in understanding relationships for people with CF at this life stage.

Relationships with siblings are key for people with CF and may be impacted in different ways.^{206, 207} The CF CP can have a role in supporting development of healthy relationships with siblings with and without CF. For the perspective of a person with CF and their experience of losing a sibling with CF, see [youtube.com/watch?v=Dkffpibi-Dc](https://www.youtube.com/watch?v=Dkffpibi-Dc)

Good relationships with peers are also important for children and young people with CF. Helping people manage these relationships may include work on communication about CF, and useful interventions may include roleplay work. For resources on helping children manage a visible difference, see the [Changing Faces website](#). Potential to build friendships (and more) with peers with CF is limited by infection control practices (and there was significant loss of contact for many older people with CF when practices changed). See also Section 8.2.3.

Social media and internet use is now a given in the lives of people with CF and parents of children with CF. This can provide contact with peers and be used to access information about CF and wellbeing and to maintain social communication, for example when children or young people with CF are in hospital. CF teams may, however, also need to support children and young people with CF to manage any negative impacts of social media and the CF CP can play a part in this.

9.4.1 Relationships for children with CF: Further good practice points for the CF CP

- Help CF teams understand the nature of relationships for children and young people with CF at different stages of development, and the multiple factors that can impact on these relationships.
- Help children and young people with CF develop good relationships with their condition, parents, siblings, peers, CF care teams and other important influences in their lives, such as educational settings, for example through MDT work, clinic-based interventions and support, liaison work and individual work with children and families if required.
- Help children and young people with CF to explore their feelings about CF and to communicate about CF as they would like.
- Help parents to foster healthy relationships within the family, for example between siblings with and without CF.
- Help the CF team promote healthy and safe use of social media, including accessing information about CF.

9.5 Relationships for adults with CF: Key issues and roles of the CF CP

As people with CF pass through adolescence and reach adulthood, the importance of family, partner and social relationships continues, and it is essential that the CF team understands the impact of CF on these relationships. With improvements in life expectancy due to advances in treatments, many adults with CF can now expect to have similar choices and opportunities in adulthood to people without CF, and this includes parenting. The introduction of new CFTR modulators for many adults with CF may also impact on relationships in various ways. It is expected that many more people with CF will have children in future, and CF team support for education, planning and having children for people with CF will need to be rapidly developed. CF CPs play an important role in supporting adults with CF to enjoy successful relationships of any kind, working both directly with people with CF and indirectly through work with the CF MDT. The following areas should be considered.

9.5.1 Sexual health and reproduction for people with CF

In CF services, discussion about sex and relationships needs to take place in both paediatric and adult services. Unsafe sexual activity is one of several health risk behaviours recognised during adolescence. For a framework for education of young people in general about sexuality, see the World Health Organisation Standards for Sexuality Education in Europe 2010.²⁰⁸

Education about sexual and reproductive health in CF may require special attention by CF teams, and an existing lack of knowledge about this has been demonstrated.^{209, 210, 211} For a review of sexual and reproductive health in CF, see Frayman et al. (2020).²¹² Unmet needs have been identified in this area of CF care,^{213, 214} with suggestions made for improvements to services, including a more standardised provision in both paediatric and adult services (albeit with room for individual preferences about discussion of these topics) and refreshers in adulthood.²¹⁵ For a summary of a wide range of sexual and reproductive health issues for people with CF and a guide to assessing needs in areas including gender diversity, body image and menopause, see West et al. (2022).²¹⁶ This is a fast-moving area of CF care and further development of approaches taken by CF teams is likely, with CF CPs having a role to play in ensuring that education and care are psychologically informed.

Common sexual difficulties have been described by people with CF, including erectile dysfunction²¹⁷ and female sexual dysfunction and impacts of CF on sex.²¹³ CF may impact on sexual wellbeing, through both physical (for example breathlessness, fatigue, pain) treatment-related (for example need for oxygen) and psychological factors (for example body image concerns, low self-esteem, low mood, and anxiety). A correlation between measures of quality of life and sexual satisfaction has been noted in people with CF.²¹⁸ There may be avoidance of asking about sexual wellbeing and function in CF services (possibly exacerbated by the long and familiar relationships that develop over time) so effort must be made to ensure this is not neglected in CF reviews.

CF has a significant impact on fertility for many people with CF, with almost all men with CF considered to be infertile.²¹⁹ Difficulties in accessing useful information about fertility have been reported,²²⁰ with resulting gaps in education about male infertility²¹¹ and possible misconceptions.²²¹ Some adult males receive their diagnosis of CF as a result of fertility investigations, so may be dealing with adjustment to both infertility and a significant new medical diagnosis. A lack of standardised approaches to men's health has been highlighted, with a wish for more in-depth discussion. Poor quality information about sexual and reproductive health may have mental health ramifications for men involved.^{222, 223}

In the past, 35–50% of women with CF have struggled to conceive naturally.²¹² However, the number of women with CF having children is already increasing with the introduction of CFTR modulators, and increased provision of information and support about fertility for females with CF is a key part of CF service development.

Previous research has highlighted complexity in decision-making about having children for people with CF.^{220, 224, 225} Guidelines for pregnancy, including psychosocial aspects are available,²²⁶ but this is a fast-changing area of CF services and new UK guidance is needed. Managing pregnancy with CF generally involves an increase in health and care demands, and this may be an additional stress for some mothers-to-be with CF. For a recent review of issues and recommendations around pregnancy in CF, including monitoring of emotional health, see Jain et al. (2022).²²⁷ For a review of issues around family-building and parenting considerations for people with CF in the modulator era, see Kazmerski et al. (2022).²²⁸

Where pregnancy is considered particularly high-risk, difficult decisions may be involved, including the need for termination of pregnancy on medical grounds for some. Pre-implantation diagnosis may be one part of the potential stresses of fertility

interventions for others. Having a child through other routes, for example surrogacy or adoption, may also be considered by some people with CF but may not be straightforward. Decisions about CF treatments during pregnancy and breastfeeding can also be complex and can cause stress where there are competing priorities. The CF CP may need to be involved with individuals facing any of these particularly complex situations and decisions.

9.5.2 Parenting with CF

Becoming a parent is a huge life adjustment for any person, bringing both benefits and challenges for people with CF,²²⁹ and is expected to be experienced by many more people with CF in the future. Benefits described include enhanced motivation and adherence to treatment, and a new sense of 'normality' and purpose.²²⁴ Challenges reported include balancing the competing demands of being a parent and managing CF, worrying about the impact of health decline and mortality on children's wellbeing, and navigating how to communicate with children about CF.^{224, 230} Dilemmas about parenting well while adhering to CF treatment regimes, mixed emotions, concerns with communicating about CF and the future with children, and issues managing CF care have also been reported. Fathers with CF may be at risk of significant health decline in the two years after birth of a child.²³¹

In the general UK population, 10–20% of mothers experience mental health problems including during pregnancy and in the first year following birth.²³² There is a lack of data on rates of perinatal mental health issues for women with CF specifically, but there may be additional stressors to deal with as above. Monitoring of wellbeing and health management after the birth of a child, as well as liaison with community resources and services is likely to be an essential part of care by the CF MDT.

The CF team also may need to offer support and advice to parents with CF about communicating with their own children about CF, and the CF CP can help ensure this is done in a developmentally appropriate way. **Resources are also available to help with this.**

9.5.3 Relationships with partners, peers and parents

Adults with CF will be subject to the same issues in relationships as adults without CF. However, disclosure of CF-related information to romantic partners may be an additional concern for some.^{233, 234, 235} CF teams and CF CPs can support people with CF to manage this process,

including level of comfort in doing treatments in front of partners and decision-making about balancing adherence to treatments with disclosure about CF.

CF CPs may have specific skills to offer some intervention around relationship difficulties but will also refer to specialist services where more appropriate, for example **Relate**. They will also need to be aware of any domestic violence/abuse within relationships and follow appropriate guidance where safeguarding action is required, with support from the CF SW. Partners (or sometimes other family members) may sometimes be involved in a systemic intervention by the CF CP for an adult with CF. However, intervention, for example for partners of adults with CF for their own psychological wellbeing, will not usually be offered, although this is sometimes requested.

Adults with CF may also have health-related issues to navigate in social relationships, including disclosure to others such as new friends, fellow students or work colleagues. Relationships with peers with CF has been influenced by infection control measures, but some adults with CF do have relationships and friendships predating this or that have developed online. CF peer support has many advantages, but CF teams may also need to support with any difficulties arising, for example through loss of peers to CF or exposure to deteriorating health in a way that can be difficult, especially if it is a repeated experience.

Changing relationships with parents during the transition to adulthood can be a challenging time for some, and CF may add complexity. Relationships with parents continue to evolve during adulthood and when health declines, people with CF's relationships with parents (and others) may change again, especially if more practical help and support is required, and parents become more involved than would be usual in adulthood. Alternatively, a loss of care and support may be experienced by people with CF as their parents age. Becoming a carer for elderly parents is becoming increasingly common for people with CF and is likely to become even more so.

9.5.4 Adults with CF and relationships: Further good practice points for the CF CP

- Help CF teams deliver psychologically informed care in the areas of sexual and reproductive health (SRH) and wellbeing, including around education and decision-making about reproduction issues, for example through consultation, other indirect work and service development.

- Help CF teams support people with CF to manage sexual wellbeing or difficulties related to CF and offer direct interventions where appropriate, but with referral to specialised services where required.
- Help teams and people with CF to manage particularly complex and difficult decisions and situations around SRH issues through direct and indirect work as required.
- Help the CF MDT support people with CF through pregnancy, for example through awareness and monitoring of emotional wellbeing, direct therapeutic intervention where this is required, and support in managing the additional health and care demands CF places on pregnancy if needed.
- Consider routine screening for emotional health issues perinatally, for both mothers and fathers with CF (or support the team to do this).
- Help people with CF to manage parenting with CF (including providing resources and signposting to external agencies where required, for example to young carers' organisations).
- Support people with CF to develop healthy relationships with partners and peers (especially where interpersonal issues or low self-worth impact upon relationships or difficulties in developing these).
- Support the CF MDT in awareness of the impact of any significant relationship problems or domestic abuse on the wellbeing of people with CF, including the need for safeguarding procedures and referral to specialist services as required and particularly in the case of domestic abuse.
- Help people with CF to manage changing relationships and any significant difficulties with their parents.

10. Looking forwards: The changing landscape of CF and implications for CF CP care

At such a time of rapid change in CF care, new guidelines for care are in danger of being out of date as quickly as they are written. While many aspects of care by a UK CF CP may remain the same going forwards, others may change. This section therefore outlines where the work of the CF CP may evolve in the future, and where guidance may need to be developed or amended over time as a result.

Awareness of the mental health needs of the UK population as a whole has arguably never been greater, with emerging data suggesting significant increases in need across many parts of the community, including children.²³⁶ The financial crisis of 2022 onwards is also expected to have significant impacts upon people with CF and their families as well as on the general population. Very significant changes in CF care and how it is delivered have also occurred, and the CF CP's role in helping those with CF manage their health is also therefore undergoing change. This section outlines how CF CP practice may best continue to develop in the future, in light of the multiple factors now influencing psychological wellbeing and CF care.

10.1 Recent changes in CF, and future CF care

The roll-out of CFTR modulators to a majority of, but not all, people with CF in the UK is significantly impacting those with CF and their families, and CF care. Over a similar time period, the COVID-19 pandemic also affected people with CF and delivery of care in CF services. For information on some early data on the impacts of these developments on emotional health in CF see Appendix 6.

Delivery of CF services is also changing in several ways, with reduced contact between CF teams and many people with CF and their families due to improved health on CFTR modulators, reduced hospital admissions for many, the introduction of 'hybrid' models of both face-to-face and virtual consultations, increasing remote monitoring and home-based care for people with CF, as well as some ongoing remote working by CF staff. Evaluation of these changes is needed by CF teams, to ensure sufficient and equitable access to high-quality CF services continues. Participation by people with CF and caregivers in the design of future services will be essential.

Numbers of adults with CF are expected to rise in the future, with increasing numbers with mild forms of the condition and a significant cohort with more severe CF experiencing increasing co-morbidities and medical complexity as they age. The needs of those for whom new CFTR modulators are not available must be understood. Analysis of genotype data has identified that people from black, Asian and minority ethnic backgrounds are significantly less likely to be eligible for such treatments²³⁷ and this needs to be addressed. Those not able to access CFTR modulators will require the same level of care as before, as well as understanding around the impact of not being able to access high-profile new treatments that are available to others. Their needs must not be overlooked. Reviews of likely future issues in CF care in general are emerging that will help inform the work of CF services.^{16, 17, 170, 238, 239, 240, 241, 242, 243}

New research, registry data and quality improvement work are all required to ensure continuing high-quality CF care for all in the future. CF services throughout the UK are reviewing some of their current practices and planning new ones. In England, an NHSE 'toolkit' to identify new priorities and changing areas of CF MDT care is being produced to aid teams in local service delivery, while revised UK Standards of Care for CF are also expected.

10.2 The future of CF CP care

CF care in general is at a time when good practice and innovation from the recent past needs to be retained, while CF care and services must also evolve rapidly in a changing era of CF. For CF CPs the task is similar – to build on experience gained and good practice carried out over almost three decades of specialist CF clinical psychology provision in the UK, while building new skills, strategies and processes for future psychological care in CF.

Areas likely to be of key importance in psychological care in CF in the new era of modulator therapies have been described.²⁴⁴ Mental health implications of increased longevity and 'survivorship' (as in the cancer literature) with CF have also recently been explored²⁴³ with recommendations for future practice including areas such as recognising the impact of trauma

(both post-traumatic stress and growth), changing self-image/identity, and the impacts of having children. For UK CF CPs, development may be required in the following areas (see below): psychological wellbeing and health management for those not able to access or benefit from CFTR modulators; psychological impacts of new CFTR modulators for the majority of those with CF, including an expected increased longevity for many; changes in delivery of CF services, including increased remote monitoring and home-based care; service development and service improvement work by CF CPs to ensure equitable access to high-quality CF psychology services for all, including recognition of the social determinants of health for people with CF and the impacts of social inequalities; new research and data in many emerging areas of CF psychological care .

10.2.1 Psychological wellbeing and health management for those not able to access recent advances in CF treatments

CF CPs need to continue to meet the psychological and emotional health needs of those who are not able to benefit from recent advances in CF treatments, with acknowledgement of the new division in the CF community in terms of access to innovative treatments at present. There is a continuing need for monitoring and promotion of emotional wellbeing and quality of life in this group, as well as effective health management, including coping with the continuing high burden of treatment for many.

10.2.2 Psychological impacts of CFTR modulators and increased longevity for many with CF

CF CPs will also need to develop their practice and services to meet some changing needs of those taking CFTR modulators. This will include having strategies in place for monitoring the emotional health of those taking modulators (especially those with pre-existing psychological difficulties), educating about possible indirect emotional impacts of improved health, helping with management of any difficult apparent side effects, aiding complex decision-making about whether to take CFTR modulators or not, dealing with cessation of CFTR modulators for any reason, and achieving good adjustment to changing health. For standards of care for CFTR modulators, see Southern et al. (2022).¹²

Uncertainty about the long-term benefits of new treatments and emotional responses to any return of symptoms need to be taken into account. Emerging research data on the mental health impacts of the new CFTR modulators needs to be understood and built upon, and CF team and CF CP practices developed accordingly. Changes in the ways people with CF on CFTR modulators are accessing CF care, for example reduced admissions to hospital or fewer CF clinic visits, may result in reduced opportunities for the CF CP to monitor, assess and offer interventions themselves, and alternative ways of working, for example in more home-based and virtual care need to be considered and developed (see 10.2.3). Those who do require inpatient admissions may have complex health needs and CF clinical psychology services must continue to offer ward-based services in addition to those delivered elsewhere.

New needs are emerging, including the need for evidence-based interventions for managing unwanted weight gain, and CF CPs will need to develop practices and services accordingly in conjunction with MDT colleagues. The psychological needs of increasing numbers of parents with CF must be considered and met. As those with CF grow older, the psychological consequences of increased complexity of care, including managing menopause and increased co-morbidities such as cancers, will need to be addressed, with suitable resources available for support and intervention as required, as well as likely need for liaison with an increasing range of specialist services outside CF. Psychologically informed palliative and end-of-life-care will continue to be required for all with CF, albeit hopefully at an older age for many with CF in the future.

Good adherence to CFTR modulators should not be assumed, and this will continue to need monitoring alongside adherence to conventional treatments. While data about the appropriateness of reducing some conventional treatments for those on CFTR modulators is awaited (see the **CF STORM Trial**), some with CF already have many questions about this. It remains paramount to understand any reasons for reduced adherence, tackle identified barriers, which can be complex and multifactorial, and maintain open, honest, good-quality relationships between CF teams and people with CF. The CF CP will continue to help in this work by CF MDTs, for example in work to develop and evaluate new evidence and psychologically informed interventions to be delivered by the CF team alongside more specific individualised CF CP interventions.

10.2.3 Impacts of changes to the delivery of care in CF services

Delivery of CF team and CF CP care has changed in several respects, due to access to new CFTR modulators for many, and also through developments in working practices developed during the COVID-19 pandemic. Increased remote monitoring of the health and wellbeing of people with CF, and delivery of psychological interventions remotely, is likely to continue. For CF teams and CF CPs, knowledge of both the risks and benefits of this need to be understood. The mitigation of any identified risks should be considered and integrated into good practice. There is the potential for increased anxiety about health, or overfocus/hypervigilance that is encouraged by access to devices to measure health parameters at frequent intervals. Some may also find it feels intrusive to pay so much attention to CF outside the CF clinic or may be less engaged in consultations than when in CF clinic. Others may not wish to take more responsibility for self-monitoring, may lack knowledge or confidence, or may have other social, environmental, psychological or cognitive barriers to this. Benefits of home-monitoring need to be weighed against any unintended negative consequences at the level of a person with CF or caregiver.

A reduction in the quality of care or in relationships or therapeutic alliances must also be avoided when delivering remote monitoring or interventions, and good access to the space, privacy and means to undertake more care at home must be ensured. A hybrid model of care is likely to be the norm in future and face-to-face contact may be particularly important at certain times, for example when issues with engagement, emotional health, social circumstances or developmental stage suggest this. CF CPs may be able to help CF teams take individual differences of people with CF into account when designing the best form of care for them. CF CPs (in addition to MDT colleagues) may also continue to deliver care through some remote working themselves (from home), and adequate technical support and equipment must be available for this, alongside strategies to ensure continued confidentiality of care, effective communication and good teamworking.

For remote monitoring and delivery of interventions by the CF CP specifically, the same considerations will apply. Annual assessment by the CF CP may need to be conducted remotely, screening tools delivered electronically, and therapy sessions delivered by video or even telephone. Group sessions (for example psychoeducational or therapeutic) may now be delivered, thanks to the use of remote technology. Adequate access to quiet, private spaces with the

appropriate equipment must be available to CF CPs and the correct governance ensured, while facilities must also continue to be available for face-to-face interventions to be provided as required.

10.2.4 CF CP future service development and further service improvements

CF CPs need to develop robust data collection for the UK CF Registry, for example around the emotional health and wellbeing of those with CF nationally. Costs of the psychological aspects of CF care need to be considered and appropriate future funding ensured. Ways of reporting CF CPs activity against standards, for example through the [NHS England quality dashboard](#) for CF, also need optimising. Gaps in CF CP staffing need to be monitored and acted upon, to ensure access to CF CP for all people with CF, including in shared care services. Challenges in recruitment of CPs need to be considered and retention of skilled CF CP staff optimised. Skill mix in CF CP teams and possibly in addition to CF CPs may need to be considered, especially as some units increase rapidly in size. As more care is delivered at home, the inclusion of CF CPs in homecare services will be essential but may have resource implications. As their services continue to evolve, CF CPs will need to evaluate the quality of their own provision through self-assessment if formal peer review is not available in the future.

The impact of social determinants of health needs to be fully recognised, particularly with the evolving cost of living crisis, and responded to in CF care generally. While CF SWs may take the lead in this area, CF CPs will also have a significant part to play. Any wider factors which impact on community mental health needs and resources will also impact on CF clinical psychology services, as CF CP teams are often more accessible to patients than local services but may not always be the most appropriate point of care. For example, the mental health needs of adolescents may be increasing²⁴⁵ and increased demand may be seen for CF CP input to this age group. Furthermore, the demand for assessment for neurodivergence, and the need for reasonable adjustments for those with additional needs may also be growing within CF services and this needs to be reflected in service improvement work.

CF CPs will also need to continue to develop their psychological screening programmes to ensure early identification and intervention for a wide range of emotional wellbeing and health management issues in a growing population. The minimum content of psychological annual assessments may need to be further defined. The

preventative and 'early intervention' approach of CF CPs needs to continue, but new strategies for the promotion of physical and emotional health in people with CF may be needed for an increasing CF population who are expected to live longer. However, with increasing numbers of people with CF (in adult services particularly), the precise remit of the CF CP may need to be reviewed. For example, delivery of psychological therapy by the CF CP may increasingly need to be reserved for people whose CF is the central issue and specialist knowledge of CF or close co-ordination with the CF team care is essential, with increased signposting to existing community-based services including NHS talking therapies in any other cases.

Emerging evidence for the effectiveness of virtual reality technology,²⁴⁶ for example for managing medical procedures, should be taken into account and may be a useful tool for CF CPs and CF teams. New therapeutic protocols and pathways are also being developed, including more 'teletherapy', growing application of the ACT approach in CF and new group interventions, and these will all need to be evaluated.

10.2.5 Future research priorities for psychological care in CF

New research is needed in many areas pertaining to the work of the CF CP. Current and longer-term psychological wellbeing for those not receiving CFTR modulators as well as the longer-term and real-life impacts of the newest CFTR modulators on emotional health and quality of life of those taking them all need to be demonstrated. New data on some psychological and cognitive impacts of CFTR modulators is emerging but further research is required. Work is also needed to further understand what helps people with CF manage CF treatments and to identify effective psychologically informed interventions in this area in the modulator era. The research priorities of the CF community also need to continue to be identified and work to address these to be carried out.²⁶ New approaches and interventions by the CF CP for both emotional health and CF health management will need to be evaluated. The longer-term impacts of the Covid-19 pandemic upon emotional health and quality of life of the CF community will also need to be reviewed, perhaps especially for those who were at important developmental stages during this.

In summary, at the time of production of these new guidelines for CF clinical psychology services, much remains to be seen in terms of changes in CF and CF care. In paediatric care, for those taking CFTR modulators, contact with CF teams will likely be less frequent than for previous cohorts, and most children with CF will be more well physically, and hopefully psychologically. Many adults with CF are hoped to live 'longer and stronger', also requiring less – or different – contact with their CF centre and with changing psychological needs, including those relating to an unexpected and significant increase in life expectancy for some. Many people with CF and their caregivers may be expected to continue to experience significant emotional health and/or health management challenges, requiring the input of the CF CP, and psychologically informed care by the CF team will continue to be required by all.

Keeping pace with change can be difficult for busy professionals and whole CF MDTs will need to adapt quickly to changes in the way CF care is delivered. Decisions about what will be needed more and what might be needed less will need to be made. These are both exciting and demanding times for CF services. The wellbeing and good function of CF teams of the future must also be ensured, and CF CPs may be able to help teams achieve this aim. CF CPs, alongside their CF MDT colleagues, will need to be responsive, innovative, and flexible to continue to deliver the highly specialised CF CP care in the manner that has been established in the UK, with the aim of optimising the health, quality of life and wellbeing of people with CF and their families now and in the long term.

Appendices

Appendix 1: Self-audit tool for CF Clinical Psychologists

Number of patients in this centre	Current CF CP staffing in this centre	Recommended staffing (calculated according to UK Standards of Care ¹)	Comments

Essential criteria

Are the following criteria met? (Section 3)	Y/N	Comments
<p>CF psychology services must be provided by a clinical psychologist*, and at larger centres by a consultant clinical psychologist</p> <p>*see Section 2.2 about the employment of other practitioner psychologists</p>		
<p>All people with CF in UK services must have access to a CF CP as part of the CF MDT.</p>		
<p>CF CPs must be registered with the Health and Care Professions Council (HCPC), and maintain CPD, supervision and registration appropriately, in line with HCPC standards</p>		
<p>CF CPs should be members of the UK Psychosocial Professionals in CF Group (UKPPCF)</p>		
<p>CF CPs must maintain specialist knowledge about CF, for example through appropriate CPD, membership of the UKPPCF and attendance at UKPPCF and other CF training events.</p>		

Is the CF psychology service sufficiently staffed and integrated with the CF MDT to do the following?

	Y/N/partial/NA	Comments
Service delivery (Section 3)		
<ul style="list-style-type: none"> Respond to referral of inpatients within one week and outpatients within two weeks (using a triage system to assess the urgency of referrals) 		
<ul style="list-style-type: none"> Use a preventative model of psychological care, with screening and other monitoring of both psychological health and health management, early intervention, and a proactive outreach approach to people with CF and their caregivers. 		
<ul style="list-style-type: none"> Help ensure equitable and sufficient access to CF CP care within a centre and to those receiving network care. 		
<ul style="list-style-type: none"> Advise CF commissioners, service directors and others as appropriate on development and provision of CF clinical psychology services, and help ensure appropriate: staffing and banding, recruitment and retention, job planning, professional management, CPD, governance and access to the facilities and systems required for effective service delivery. For more details on service delivery, see our summary information for commissioners of CF services and CF clinical directors. 		
<ul style="list-style-type: none"> Ensure good self-care, including access to the required level of clinical supervision. 		
<ul style="list-style-type: none"> Help ensure people with CF and their families are aware of the CF psychology service and provide information about how CF CP might help and how to access their service. 		
<ul style="list-style-type: none"> Ensure adequate systems for clinical record-keeping, taking account of governance and needs for retaining confidentiality of some CF CP records. 		
<ul style="list-style-type: none"> Collect data on psychological work and activity delivered in the CF service, as required locally or regionally (including for the NHS Specialised Services Quality Dashboard for CF in England). 		
<ul style="list-style-type: none"> Carry out psychological research, quality improvement initiatives and audit in CF, and disseminate this work through publication and/or presentation, with awareness of the research priorities of the CF community.^{25, 26} 		

<ul style="list-style-type: none"> • Collect and share feedback from people with CF and their caregivers about CF psychology services, involve them in service improvement work and research, and use their 'expert by experience' knowledge and skills where possible. 		
<ul style="list-style-type: none"> • Provide training placements for students, including doctoral practitioner psychologist trainees, and supervision of postgraduate research, as appropriate to the needs of the service. 		
<p>Annual assessment and additional screening (Section 4)</p>		
<ul style="list-style-type: none"> • Devise a robust local strategy for monitoring of psychological health for people with CF, with developmentally appropriate screening by the CF CP carried out at least annually, through paper/digital or face-to-face assessments at the CF annual review. 		
<ul style="list-style-type: none"> • Provide psychological screening at the CF annual review that allows early identification of a wide range of psychological difficulties and an overview of both emotional wellbeing/ mental health issues (including but not limited to anxiety and depression) and health management issues. 		
<ul style="list-style-type: none"> • In paediatric centres, screen/monitor the wellbeing of caregivers (in adult services, CF CP annual assessment will be limited to people with CF and their caregivers will not be routinely screened). 		
<ul style="list-style-type: none"> • Be aware of guidance from the International Committee on Mental Health in Cystic Fibrosis (ICMH) on annual screening of people with CF (and caregivers in paediatrics) for anxiety and depression, and apply as appropriate locally. 		
<ul style="list-style-type: none"> • Provide information as part of the annual assessment about how to access further assessment and intervention from the CF CP and an opportunity to ask for this if needed. 		
<ul style="list-style-type: none"> • Be aware of guidance on screening and monitoring of emotional health when people with CF are starting new CFTR modulators, and implement locally as appropriate 		
<ul style="list-style-type: none"> • Provide additional monitoring of the psychological health of people with CF (and of caregivers in paediatric services) through a variety of other means, for example through attendance at CF clinics and/or MDT meetings and close liaison with CF team colleagues. 		

<p>‘Direct work’ with people with CF and families (Section 5)</p>		
<ul style="list-style-type: none"> Carry out engagement work with people with CF (and caregivers in paediatrics), for example when a person seems likely to benefit from CF CP input but feels unsure about the value of CF CP input or has barriers to this (for example perceived stigma). 		
<ul style="list-style-type: none"> Offer direct CF CP assessment, formulation and intervention to people with CF (or caregivers where appropriate), for a wide range of issues including emotional wellbeing, mental health and management of CF. 		
<ul style="list-style-type: none"> Evaluate interventions, including use of standardised outcome measures where appropriate. 		
<ul style="list-style-type: none"> Assess and manage any safeguarding or risk issues arising in direct work, in conjunction with other members of the CF team (especially CF social workers), appropriate NHS Trust departments and other agencies as required. 		
<ul style="list-style-type: none"> Offer direct work through 1) presence in the CF clinic where appropriate (especially in paediatric care) 2) CF CP outpatient appointments (arranged together with the CF clinic appointments where helpful) 3) during CF admissions as required. 		
<ul style="list-style-type: none"> Offer a hybrid model of face-to-face and virtual working, with telemedicine/ video consultations and telephone work as appropriate, taking account of medical, psychological, social and governance issues that should influence decisions about how CF CP care is delivered. 		
<ul style="list-style-type: none"> Ensure that all contact for face-to-face direct work with people with CF follows infection control guidelines including any CF-specific guidance, and that any environment used is appropriate in this regard. 		
<ul style="list-style-type: none"> Ensure that suitable therapeutic space is used for direct work with people with CF and their families. 		
<p>Indirect work with/via the CF MDT (Section 6)</p>		
<ul style="list-style-type: none"> Provide a specialised psychological viewpoint to general CF service development, quality improvement initiatives, research, audit and strategy/future planning. 		

<ul style="list-style-type: none"> • Develop a 'stepped care' model of psychological care, utilising the skills of the wider CF MDT to deliver psychologically informed and supportive care and interventions for people with CF and caregivers. 		
<ul style="list-style-type: none"> • Provide consultation, advice, teaching, training and supervision as required to the CF MDT, for example through attendance at MDT meetings, education sessions and formal or informal supervision arrangements. 		
<ul style="list-style-type: none"> • Work closely with CF MDT colleagues to provide 'joined-up' care and with the CF SW to form an integrated CF psychosocial team. 		
<ul style="list-style-type: none"> • Advise and support the CF team on management of risk issues when required, for example issues around suicidality, safeguarding children and vulnerable adults, or self- neglect. 		
<ul style="list-style-type: none"> • Provide expert liaison with multiple statutory and non-statutory services outside a CF centre, including shared care CF services, educational settings, social services and other health services and professionals. 		
<ul style="list-style-type: none"> • Provide expert gatekeeping and liaison and make onward referrals to mental health services, including liaison psychiatry and community services, and coordinate access to on-call child and adult psychiatry services when needed. 		
<ul style="list-style-type: none"> • Help the CF team develop good models of self-care, for example through advising on team processes and systems for preventing, monitoring and managing staff stress. 		
<p>Emotional wellbeing and mental health (Section 7)</p>		
<ul style="list-style-type: none"> • Take a preventative approach to mental health issues for people with CF (and caregivers in paediatric care), for example through help to build coping skills, empowerment and promotion of emotional wellbeing. 		
<ul style="list-style-type: none"> • Work directly with people with CF to provide screening, assessment, formulation and evidence-based interventions for emotional wellbeing and mental health (not limited to anxiety and depression). 		

<ul style="list-style-type: none"> • Be aware of the International Committee on Mental Health in Cystic Fibrosis (ICMH) guidance on screening, assessment, prevention and treatment for anxiety and depression in CF services and apply as appropriate locally. 		
<ul style="list-style-type: none"> • Be aware of guidance on the need for CF MDT familiarity with the potential wide range of psychological impacts of CFTR modulators, and offer CF CP intervention as indicated. 		
<ul style="list-style-type: none"> • Offer assessment and intervention for emotional health to caregivers as appropriate in paediatric care, or signpost to appropriate services as required. 		
<ul style="list-style-type: none"> • Include family members/caregivers in work with people with CF where appropriate, to provide systemic interventions for emotional health. 		
<ul style="list-style-type: none"> • Facilitate other members of the CF team to support and promote the emotional health of people with CF and their families through indirect work, for example through consultation, education and training, reflective practice and supervision, and service improvements. 		
<p>Managing CF and CF care throughout the lifespan (Sections 8.1 and 8.2)</p>		
<ul style="list-style-type: none"> • Provide specialised psychological assessment and interventions and support the work of the CF team as required, at times of change and transitions for people with CF, including in the following areas: 		
<ul style="list-style-type: none"> - managing CF and change at different developmental stages in childhood 		
<ul style="list-style-type: none"> - adolescence and transition to adult life and adult CF services 		
<ul style="list-style-type: none"> - managing growing older with CF. 		
<ul style="list-style-type: none"> • Provide specialised psychological assessment and interventions around managing the challenges of CF and CF care, and support the work of the CF team/provide joined-up care, including in the following areas: 		
<ul style="list-style-type: none"> - new diagnosis of CF at any age 		
<ul style="list-style-type: none"> - issues relating to CFTR modulators 		
<ul style="list-style-type: none"> - psychological aspects of infection control 		
<ul style="list-style-type: none"> - procedural distress 		
<ul style="list-style-type: none"> - CF admissions and outpatient care 		

- nutritional, eating and GI issues		
- CF diabetes (CFD)		
- secondary diagnoses and complications		
- medical trauma		
- intrusive treatments and environments		
- medically unexplained or persistent physical symptoms		
- neurodivergence, neurodevelopmental conditions and other additional needs and CF.		
- lung transplantation issues		
- palliative care, advanced stages of the condition, and end-of-life care.		
Adherence to CF treatments (Section 8.3)		
<ul style="list-style-type: none"> The CF CP should provide specialised psychological input around adherence to CF treatments through: <ul style="list-style-type: none"> - Direct work with people with CF or their caregivers on adherence issues, using a variety of evidence-based psychological models, theories and approaches. - Supporting the work of the CF MDT around adherence issues, for example through joint work, consultation, teaching/training, and service improvement. 		
Healthy relationships in CF (Section 9)		
<ul style="list-style-type: none"> Provide specialised psychological assessment and interventions, and support the work of the CF team as required, including in the following areas: <ul style="list-style-type: none"> - The CF team and relationships. - Parenting children with CF. - Relationships for children and adolescents with CF. - Relationships for adults with CF including: <ul style="list-style-type: none"> - sexual and reproductive health - parenting as an adult with CF - relationships with partners, peers and parents. 		

See further good practice points in Sections 4–9 of guidelines if needed.

Appendix 2: Guidance on format and content of the CF CP annual assessment in paediatric care

The psychological component of the annual review assessment in Paediatric Cystic Fibrosis Centres in the UK

UKPPCF Consensus Document

September 2022

The main aim of the psychology annual review is to assess psychological wellbeing and adjustment to CF, family-, educational-, friendship-functioning, understanding of CF and management of treatments; the review should be structured in a developmentally appropriate way that evolves over time in complexity as the patient gets older.

Example:

'As part of your annual review, as your psychologist, we also want to find out a bit more about how you are feeling, how your year has been, and how you are managing CF. So we have a few questions that either you can answer or your grown-up can help you with.'

Consensus

1. Assessment of understanding of CF as developmentally appropriate. The rationale for this question is to ensure that children and young people's understanding of CF is age-appropriate and that they have a rationale for treatment adherence. It will also offer opportunities with parents and children to model age-appropriate discussions about CF and give children ways to answer questions from peers. It enables an insight into the parental adjustment to the diagnosis and the identification of any areas that parents might need support to talk about with their child. Example questions are given below to be adapted to your way of working.

- In the first annual review of an infant, ask parents what they plan to say about CF to their child as they grow up and what they share with wider family and friends etc.
- Ask younger children – why do you come to see us at the hospital/come to clinic appointments? Do they know the name of the clinic? What does CF stand for?
- As they get older, do you understand/ know how CF affects your body? Do you know

which treatments help which parts of your body/the purpose of treatments? Is there anything you feel you don't understand or would like to know more about in relation to your health?

2. Family: Social and other support for the family. The rationale for this question is to gain insight into the family structure, who helps with treatments, and who is available for emotional and practical support in the family.

It may be helpful in the first annual review to do a genogram and in subsequent annual reviews ask about any changes to family structure and social support.

3. Attendances and absences from playgroups/nursery/pre-/school. This question offers another opportunity to assess adjustment to diagnosis and an insight into systemic expectations about illness.

- This question may provide an opportunity for education about infection risks and balance with social development.
- As the child gets older, this question will give insight into a wellness or illness model within the family and aspirations for the future.
- When children start pre-school/school think with parents and children how they might answer their peers' questions about what Creon is? Or why are they taking a tablet?
- At transition from primary to secondary school ask if they have thought about how they will talk to new people about CF (role play with them in clinic if this feels helpful).
- Prepare parents and children that they may learn about CF in their Genetics lesson in year 8/9.

4. Friendships/relationships. This question enables you to assess how they are managing with friendships and if CF impacting on these relationships.

- Assessment of social support, being able to talk about CF or treatment that needs to be done in front of others and support with 'feeling different'.
- Ask older children if/what have they have thought of how to talk about CF to someone who feels more special than other friends as well as what they might say to other peers.

5. 'Wholeness of person.' What do you enjoy? What has gone well over this last year?

6. How do you manage your treatment/how manageable are your CF treatments? This question will change across development but it's to understand and capture whether children

know what treatments they are taking and what they do. Which treatments are easier to do, and which require more motivation to do. This may also be a springboard for co-working with CF colleagues.

- Young children – what supports the family in adhering to the child’s daily treatment routine?
- Older children/young people – how do they manage potentially competing demands of school work, socialising, leisure time and looking after their health? Have young people developed strengths of skills which assist them?

7. How do you feel about having medical procedures (for example blood tests, having a canular, long line inserted, X-ray, ultrasound) done/coming to clinic?

8. What has been more challenging over the last year (unless this has already been raised in earlier questions)? Has anything been getting you down, stressing you out or causing you to worry?

- Ask about risk if appropriate, eating, sleeping, mood and anxiety in young people.

9. What are your plans and hopes for the next year? You may want to ask both children/young people and their parents.

Appendix 3: Guidance on format and content of the CF CP annual assessment in adult services

The psychological component of the annual review assessment in Adult Cystic Fibrosis Centres in the UK

UKPPCF Consensus Document

Written 2016 (due for review)

Medical and psychological assessments at annual review are part of standard CF care in the UK (CF Trust, 2011).¹ This paper describes current best practice for psychologists across adult CF centres in the UK, enabling some standardisation across the centres.

A group of over forty psychologists working in adult CF centres hold annual meetings to develop best practice in CF psychology care in the UK, including working over a five-year period to iteratively develop the psychological component of the annual review assessment. The aim was to develop an annual screen concurrent with the medical annual review that best captured psychological wellbeing and health-related behaviour.

Through extensive debate and scrutiny of currently available psychometric measures, informal patient involvement, and clinical knowledge, a consensus was reached on areas that most accurately captured people’s quality of life and mental health over the previous calendar year. The annual review covers a wide range of disparate areas, including low mood, feeling anxious/worried, social support, sleep and anger difficulties, family relationship functioning, managing CF care, and procedural anxiety. These consensus areas are the recommended minimum content of the psychological annual review. The recent mental health guidelines developed by Quittner² and colleagues make recommendations for additional screening for anxiety and depression which may be useful additions to this guidance.

The annual review can be either a face-to-face or questionnaire-based format and there is no recommendation for specific timing of the screen. The Working Group felt it was important to be flexible enough to allow for regional differences given the complexity of living with Cystic Fibrosis, the variation of staffing levels and differences in procedures for completing the medical component of the annual review in different CF centres across the UK. Instead, the group emphasises the importance of professional judgement in delivering

psychological annual reviews in a manner appropriate to the person with CF and the service context of each centre.

Function of psychology reviews

The main function of psychology reviews in the UK is to screen people for a wide range of emotional and health-related behaviour concerns, and to identify those people in need of further assessment and/or psychological intervention. **There are a number of important secondary functions:**

- Informing people with CF (and their families) of the psychology service available at the CF centre for CF-related concerns
- Building working relationships with patients/families so they are more likely to access psychology at times of distress
- Psychoeducation: introducing people to psychological ideas, addressing misconceptions and reducing barriers to seeking help should problems arise in future
- Building up a picture of the person with CF over time. This provides important contextual information (preferred coping styles, strengths, sources of support etc.) to the psychologist and/or CF team should the person face challenges or seek help in the future
- Monitoring of certain groups of people with CF, who may have additional stresses/issues for example, those diagnosed in adulthood; those with learning disabilities or other complex needs; those making decisions about having a family; those with deteriorating health, advanced stages of the condition, and/or requiring transplant assessment etc.

Format of psychology reviews

Wherever possible, people should be offered face-to-face review with the appropriate psychological professional. We recognise that service constraints may make this difficult or impossible. Where face-to-face reviews cannot be offered to all patients in a given centre, a questionnaire-based review should be offered to all patients, or a mixed model adopted where patients are allocated either to paper-based screening or face-to-face review depending on certain characteristics (for example severity of CF, history of psychological problems, known social problems or patient choice). Where paper-based reviews are offered, adaptations need to be made for people who have difficulty completing questionnaires, in line with good practice and equality legislation.³

Informal psychological assessment is an ongoing process by all members of the MDT and part of routine care. MDT members can make a referral

to the team psychologist at any time of the year and may recommend a face-to-face psychological assessment if thought to be needed.

Content of psychology reviews

Having reviewed the currently available options for screening tools, the group believes that there are no available psychometric tools which could adequately provide an overall screen or review that capture the breadth and depth of CF-related issues. Examples of such issues that psychologists typically work with include supporting individuals and their families with psychological adjustment (for example, to diagnosis, deteriorating health or end-of-life issues), managing developmental challenges or transitions (for example, to adult services, related to employment or work, leaving home or preparing for pregnancy and/or parenthood) and supporting adherence and behaviour change. Given the diversity of this list, tools which are available all have limitations in terms of psychometric robustness, availability of appropriate cut-offs and/or limitations in scope compared to the role of psychological professionals in CF teams.

In spite of these reservations, the group agrees that available psychometric tools may form a useful part of psychological annual review, which would be in line with mental health guidelines, which highlight the importance of screening for anxiety and depression in people with CF. Centres may choose to adopt elements of these guidelines, specifically to enhance screening for these mental health difficulties. Their use should constitute part of the psychological review process and care should be taken to interpret any scores obtained in relationship with the medical review, specific health changes and other contextual information. The UK consensus document for annual review focuses on more than mental health diagnosis of anxiety and depression, focusing instead on formulation of psychological and health-related behaviour difficulties and how they interact with CF with an emphasis on prevention as well as treating identified difficulties. This highlights the benefits of embedded psychologists in CF teams who are able to assess wide areas of functioning.

As a **minimum**, the group agrees that the following areas should be covered.

- Information to allow informed consent to the procedure and information on data use and storage (including how any results will be shared with the wider CF team).
- Patient evaluation of their current emotional wellbeing and coping.
- Patient evaluation of their current support from family and friends and from CF professionals.

- Screening for patient perceived psychological difficulties/concerns commonly seen in adults with CF including mood problems (anxiety and low mood), stress, procedural anxiety, low self-esteem, perceived lack of support, anger, eating difficulties, pain, decision-making (both medical and non-medical), relationship difficulties, treatment adherence, trauma, sleep problems, concerns about changes in health. This broad range of areas is selected to reflect the broad scope of activity with which UK Clinical Health Psychologists should be concerned.⁴
- Opportunity to describe positive factors contributing to successful coping.
- Opportunity to describe any concerns not covered by a standardised framework.
- Opportunity to request or decline feedback.
- Opportunity to request a face-to-face meeting with the clinical psychologist, if the review is not conducted in person.
- Opportunity to opt into or out of contact with the CF psychologist even if problems are disclosed.

Summary and recommendations for the future

Psychologists working in UK adult CF centres see annual psychological review as an essential part of CF care, and consensus is that this should cover the wide range of psychological issues that may impact on health and quality of life and allow early identification of any emerging difficulties. The authors recognise that the use of the UK consensus for psychology annual reviews needs to be evaluated to assess the efficacy of the review process for patients and centres. This consensus guidance will be revised in future in light of any new information or developments.

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Appendix 4: European CF Society Conference poster 2015 – The preventative approach of the UK CF CP

A Preventative Psychosocial Care Approach (PPCA) for promoting psychological wellbeing for people with CF and their caregivers: the UK model

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On behalf of the UK Psychosocial Professionals Group (UKPPCF)

Introduction

The UKPPCF model of embedded psychological care

- People with chronic health problems are at risk for psychological distress¹ and physical health outcomes are improved if psychological distress is prevented².
- Service evaluation research indicates that psychologists embedded in the MDT can prevent mental health symptoms from developing into intractable mental health diagnoses³.
- The team clinical psychologist has expert knowledge of CF, and is therefore aware of any likely impacts of CF/ treatments on emotional functioning and the key triggers for potential distress.
- Anxiety, depression and other significant emotional health difficulties are important to consider. However, other psychological factors are also central to adherence and other aspects of physical health. A wide range of psychological issues may arise for people with CF and their caregivers see table 1.
- Consideration of psychological impacts, needs and possible interventions should be delivered alongside physical health care at the same time. Anticipated risk = opportunity for preventative intervention see table 2.

Table 1- Potential issues arising for PWCF and caregivers, requiring early identification, prevention or psychological intervention

Adjustment to diagnosis (occurring at any age)
Issues with feeding/eating/other behavioural issues
Managing demands of CF at key developmental stages/transitions/stress points
Managing transition to adult life/services.
Need for psychological resilience and positive coping strategies
Managing adherence to treatment, treatment burden, engagement in CF care and medical procedures required in CF e.g. venepuncture
Decision making re reproduction options
Dealing with challenging/traumatic medical events
Impact of CF on family functioning
Emotional health issues including: anxiety, demoralisation, depression, low self esteem/confidence, anger, relationship issues
Adjustment to declining health, additional diagnoses and challenges of growing older with CF
Emotional wellbeing at end stage disease and end of life care

Table 2- Psychological care delivered concurrently with physical health care

Cystic Fibrosis presenting problem	Examples of Physical management/ interventions	Examples of corresponding psychological work
Weight loss	PERT review, OGTT	Mealtime management, eating disorder/mood assessment
Reduced FEV1 due to Pa growth	Nebulised/Intravenous antibiotics	Work on: adherence to nebulisers, help for invasive procedure phobia
Secondary diagnosis eg CF related diabetes	Endocrinology review, insulin, dietary review	Assessment of trauma/stress/anxiety/ impact on adherence to all treatment
Bronchiectasis, breathlessness	Progression through regular IVABs, introduction of oxygen, transplant referral	Review of adjustment/mood/quality of life. Deliver family/individual intervention as required

Method

The UKPPCF has developed a preventative approach, refined over two decades, and based on detailed standards of care for CF^{4,5}. This model promotes psychological wellbeing, and is responsive to the psychological issues occurring in those with chronic physical health issues such as CF.

Table 3- Cost effectiveness of Psychosocial Preventative Care Approach⁶

Co-morbid mental health problems are associated with a 45-75% increase in service costs for long term physical health conditions (LTC) (after adjusting for physical disease severity).
Delivering effective care and support to people with LTC therefore involves: promoting good mental health, prevention of mental health problems, integrating mental health into chronic disease management programmes, collaborative and stepped care delivery and effective support of self management skills
“There is evidence that costs of including psychological initiatives within disease management programmes can be more than outweighed by the savings arising....”

Results

Development of psychological care in CF services in the UK has resulted in a PPCA approach that includes several key elements, summarised below:

Roles of the CF Psychologist

The overall role of the CF psychologist is to promote physical health outcomes, QoL and emotional wellbeing through the use of expert psychological knowledge and principles. Roles include:

- Formulation (not diagnosis) of presenting difficulties.
- Consultation to the CF team.
- Training, supervision and support to CF team.
- Surveillance of PWCF/caregivers (offering early identification of needs and preventative interventions).
- Promotion of positive coping strategies, development of resilience and harnessing existing strengths of PWCF/caregivers.
- Provision of expert assessment and delivery of evidence based psychological therapies.
- Interventions to optimise health management e.g. work on adherence, managing procedures, optimising engagement with healthcare team.
- Liaison and gatekeeping to agencies including: community mental health services, social services, liaison psychiatry services.
- Close liaison/co-working with the CF social worker to complement services offered by them, and optimise delivery of complete psychosocial care service to PWCF/caregivers.
- Development of model of psychological care in CF unit (through service improvement, audit, research).

PPCA

Surveillance of PWCF/ caregivers by the CF Psychologist

Surveillance aims to identify early signs of emotional/ other emerging difficulties and opportunities for early intervention. This can be achieved through:

- Attendance at MDT discussion of patients.
- Presence in CF clinics and CF wards and participation in consultations.
- Close liaison with other CF team members to facilitate early referral for specialist psychological help.
- Participation in the annual assessment of PWCF with a more formalised assessment of psychological wellbeing and areas of need.

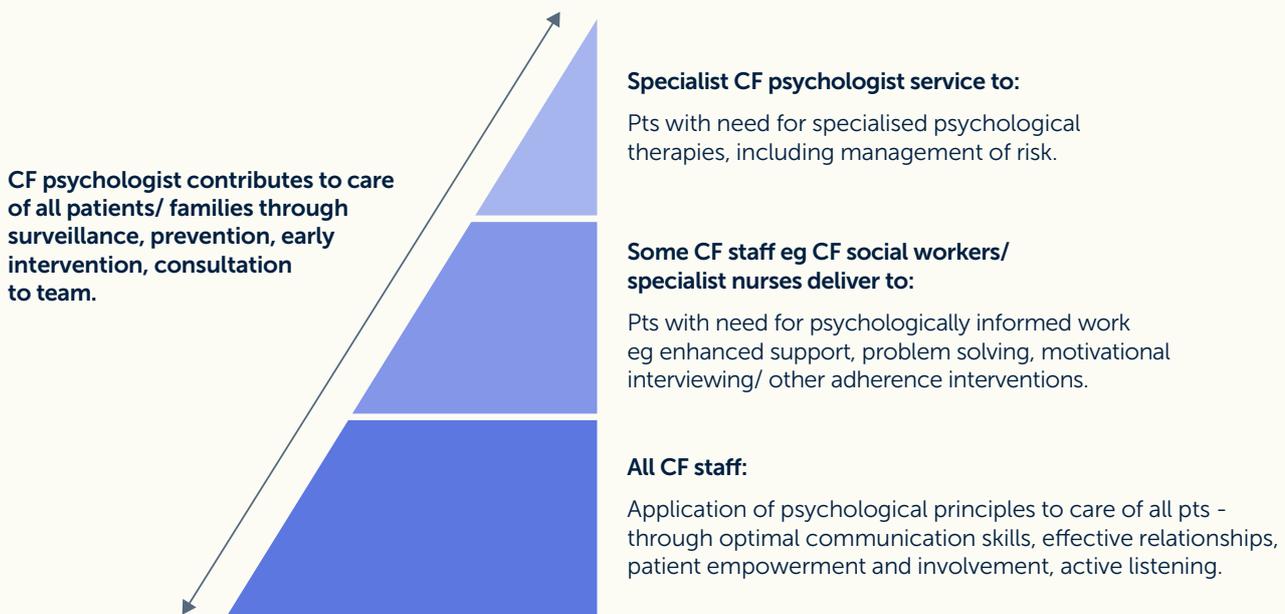
Annual screening assessment by the CF Psychologist

The CF Psychologist carries out an annual assessment: minimum data collected may include:

- Checklist of wide range of potential psychosocial issues experienced by PWCF.
- Subjective numerical ratings of emotional health/quality of life/current coping with CF.
- Subjective report of current level of support.
- Standardised measures of emotional health and quality of life.
- Opportunity to report positive coping skills used.
- Opportunity to request early psychological assessment/ review.
- Opportunity to report any other concerns not covered elsewhere.
- Provision of information about the CF psychology service and "normalisation" of this as part of the integrated CF service.

Annual screening methods will also need to include: protocols for gaining consent for assessment and appropriate information sharing, managing any risks identified, effective data collection and refining assessment on basis of service user feedback.

Psychological care in CF: model of stepped care and opportunities for preventative actions



Discussion and conclusions

- The PPCA outlined above describes a model of care that supports delivery of the UK standards of care for CF. Evidence suggests that this approach (preventative and integrated psychological care) is also a cost effective way of delivering care - see table 3.
- A PPCA aims to prevent mental health conditions developing in people with physical health conditions, particularly long term conditions for which there is no cure. It also aims to optimise physical health outcomes through a “joined up” and co-ordinated approach within MDT care.
- The UKPPCF supports the development of equitable psychological care throughout CF services in the UK, through the well established peer review process. The PPCA provides a model by which to deliver optimal psychological care for people with this lifelong and lifelimiting condition.

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Appendix 5: Issues historically experienced by (and seen in clinical practice with) children and their parents, young people and adults with CF

Further issues that may be experienced by parents of children with CF include:^{200, 247} (see also Bryon & Tluczek 2018, and Elderton et al.)^{102, 248}

- Adjusting to a diagnosis of CF, learning that this is a life-limiting condition and dealing with emotions, including uncertainty, shock and disbelief, loss and grief (for the child that they thought they had), and fear of the future.
- Possible disruption to the parent-infant bonding attachment following diagnosis of CF.
- Confusion around diagnosis in an apparently healthy child: Is my sick child healthy or is my healthy child sick?
- Navigating the new 'world of CF', explaining complex information to family and friends, and absorbing their reactions to this information.
- Establishing effective routines that incorporate everyday activities and CF care/treatments.
- Difficulties establishing positive feeding behaviours in young children.
- Coping with children's fears about CF treatment, hospital admissions, or invasive procedures, including being present and managing own distress as a parent.
- Concerns about children starting nursery, preschool or school and handing over responsibility for CF care to another adult.
- Talking about and explaining CF to children; providing accurate and age-appropriate information (and managing children's use of online information about CF).
- Maintaining effective parenting approaches in the face of additional stressors related to CF.
- Balancing the needs of other children and family members alongside a child with CF.
- Challenges of negotiating the care of a child with a physical health condition with statutory services (healthcare, education, etc).
- Future family planning once CF informs decision-making.
- Additional financial concerns as a result of CF.
- Ongoing parental adjustment and adaptation

to CF and associated parental wellbeing. Possible feelings of incompetence and lack of confidence, blame and guilt, fear, loneliness and isolation, anger, sadness.

Further issues that may be experienced by children with CF aged 0–12 years include (see also Elderton et al. (2018)):²⁴⁸

- Fears or concerns about CF treatment, clinic appointments, or hospital admissions.
- Anxiety and distress about invasive procedures such as cannulas, blood tests, throat swabs and swallowing tablets.
- Challenges with feeding behaviour/nutrition.
- Sleep problems if sleep hygiene skills have not developed, for example due to discomfort, parental anxiety/super surveillance, or a child's own anxieties.
- Managing increased separation from parents as children start nursery, preschool and school.
- Learning about CF at different developmental stages, including a possible 'second diagnostic period' at age 8–12.²⁴⁹
- Developing a positive sense of self that includes CF.
- Developing a sense of ownership/control over health and self-efficacy in CF-related care, and being actively involved in effective daily treatment routines.
- Adherence to treatment – children often accept many routine treatments started at diagnosis but co-operation with new treatments can be challenging (for example accepting a nebuliser for the first time), and can involve increasing recognition of the permanence of treatments.
- Peer relationship challenges, including managing disclosure about CF to friends and responding to peer questions, such as 'What is that medicine for?' or 'Why do you go to hospital sometimes?'
- Increased awareness of other people's emotion may mean children do not want to voice their own worries and concerns for fear of upsetting others.

Further issues that may be experienced by adolescents with CF include (see also Blakemore & Choudhury (2006) and Castellani et al. (2012)):^{94, 250}

- Difficulty in regulating own emotions, difficulties with impulse control and inability to delay gratification.
- Increased risk-taking and less future-oriented decision-making compared with adults.
- Increased importance of peer and intimate

partner relationships and challenges managing questions or social situations in respect of CF.

- Difficulties with adhering to treatment, for example due to lack of motivation, unhelpful personal or social circumstances, or challenges in managing the complex organisational aspects of CF healthcare.
- Increased independence and autonomy and continued identity development alongside the demands of a long-term condition, parental involvement and healthcare.
- CF may affect physical development, delaying puberty and growth, and contributing to a sense of feeling different to peers.
- Adolescents may have an adult understanding of death and dying, but without having the emotional maturity to make sense of this.

Further issues that may be experienced by adults with CF (in addition to reported anxiety or depression) include:²²

- Continuing negative impacts of adverse childhood events (ACEs), early trauma or attachment issues.²⁵¹
- Disrupted eating behaviours, relationship with food, and poor body image.
- Low self-esteem and low self-worth.
- Difficulties with adapting and adjusting to CF, and negative feelings, for example guilt or shame about the condition.
- Demoralisation – distinct from depression and characterised by feelings of hopelessness and powerlessness.
- Disruptive feelings of anger, or anger-management difficulties.
- Self-harming behaviours, for example cutting oneself to manage difficult emotions.
- Sleep disturbance.
- Managing disruptive physical symptoms such as pain and fatigue.
- Disordered eating or relationships with food.
- Disrupted personal or family relationships.
- Grief or bereavement reactions (including loss of others with CF).
- Feelings of isolation or lack of support.
- Psychological impacts of socioeconomic disadvantage or adversity.
- Substance misuse.

Appendix 6: Early data on impacts of new CFTR modulators and the COVID-19 pandemic on emotional wellbeing/mental health for people with CF and their families

At the time of writing, there is some existing and further emerging data on how new CFTR modulators and the COVID-19 pandemic have impacted the emotional wellbeing and mental health of people with CF. This is likely to be a rapidly evolving area over the coming years.

Early data on impacts of new CFTR modulators for people with CF

CFTR modulators are now becoming available to the majority of people with CF in the UK aged six and over. New data is therefore needed about the emotional wellbeing of people with CF in this new era of care, about the psychological and emotional impacts of this major development in treatment for many, and about the emotional wellbeing of those for whom new treatments are not available.

People with CF taking CFTR modulators such as Kaftrio® may require adaptation to changes in symptoms, treatments and contact with CF services. Known effects of Kaftrio®, including weight gain and change in respiratory symptoms, may require new types of input from the CF team, including the CF CP. Families, caregivers and partners of people with CF can also be impacted by new treatments and changes in health of a person with CF. (See [Kaftrio – Complex and individual experiences.](#))

Positive psychological benefits of CFTR modulators have been reported²⁵² and several themes relating to subjective experiences of CFTR modulators have been described.^{253, 254} The importance of recognising the individual and varied nature of psychological responses to CFTR modulators has been emphasised, for example depending on perceptions of illness, self-identity and outcomes.²⁵⁵ For those reporting mixed or negative impacts of CFTR modulators on emotional wellbeing, causation may be hard to unpick, especially as much of the roll-out of Kaftrio® in Europe took place during the pandemic. Clinical experience and limited case reports suggest that a number of psychological/psychiatric issues, apparently associated with CFTR modulators, have been experienced by some people with CF, including sleep disturbance,

anxiety, mood changes, suicidality and cognitive effects often described as 'brain fog'. There are several possible/theoretical mechanisms for such experiences, including drug-drug interactions and direct effects of modulators on CFTR function in the brain, as well as indirect effects. Some trial and later real-life data on the psychological impacts of CFTR modulators, both positive and negative, is available^{256, 257, 258, 259, 260, 261, 262} but more is required. This includes information from any formal reporting of possible side effects, for example via the MRHA 'yellow card' system in the UK. See Spoletini et al. (2022)²⁶³ for an approach to dose adjustment of CFTR modulators when mental health impacts are reported.

As the new CFTR modulators take effect and health improves, for some patients the burden of existing treatments may in fact feel greater and some may wish to reduce conventional treatments. Research in this area is ongoing (see the [CF STORM Trial](#)) but the CF CP may need to be involved, along with other members of the CF MDT, to support continued adherence (see also Section 8.3).

Data on psychological impacts of the COVID-19 pandemic

The full and long-term impacts of the COVID-19 pandemic on the emotional health of the UK population as a whole are not yet fully known, but early data suggested decreased life satisfaction and happiness and increased anxiety.²⁶⁴ Mental health impacts (as well as social and financial ones) have not been evenly distributed, with a reported widening of already existing mental health inequalities and with those affected by socioeconomic inequalities more likely to experience negative mental health impacts.²⁶⁵ The mental health and wellbeing of those with long-term conditions who were required to shield during the early part of the pandemic 'was hit particularly hard'.²⁶⁶

Early psychological impacts of the COVID-19 pandemic on people with CF were described, suggesting 'evident psychological impact'²⁶⁷ and fear, worry, isolation and loneliness, with a need for individualised and specific advice and support.²⁶⁸ Other studies have suggested similar high levels of distress in people with CF compared to the general population, and a lack of correlation between lung disease severity and distress.²⁶⁹ People with CF may have been more

rehearsed than others in the coping strategies needed, for example in avoiding infection and managing emotions about this.²⁷⁰ A reduction in screening for mental health difficulties but also a benefit of increased access to psychological support during the COVID-19 pandemic has been reported, including the helpful development of new telehealth interventions.²⁷¹ For articles relating to the impact of the COVID-19 pandemic on people with CF and CF care overall see Sabadosa et al. (eds) 2021.²⁷² Further work will however be required to identify the longer-term impacts of the COVID-19 pandemic on psychological wellbeing for children, young people and adults with CF and their families.

Appendix 7: Proforma for information-sharing between CF and transplant psychologists.²⁷³

The CF psychologist, in collaboration with the person with CF, will usually provide information to the transplant psychologist on the following areas. This may be via direct liaison, letter/report or use of this UKPPCF proforma designed for this purpose.

- Past and present psychological or psychiatric difficulties (including past trauma).
- Past and present adherence difficulties (medicines, nebulisers, physiotherapy, exercise, nutrition).
- Past and present nutritional management difficulties (tube feeding, supplements, eating disorders).
- Past and present substance misuse difficulties (including smoking, alcohol, illicit drugs, vaping and nicotine replacement therapy).
- Any family history of CF that might impact the person's transplant journey (for example, death of a sibling with CF).
- Other significant life events that might impact the person's transplant journey (pregnancy, relationship difficulties, history of abuse).
- Information on coping styles.

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Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all.

We won't stop until everyone can live without the limits of cystic fibrosis.

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We welcome your feedback on our resources. You can also ask for this resource in large print or as a text file.

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