School of Psychology
Ysgol Seicoleg

Living into Adulthood with Cystic Fibrosis

Thesis submitted in partial fulfilment of the requirement for the degree of:

Doctorate of Clinical Psychology (DClinPsy)

South Wales Doctoral Programme in Clinical Psychology

Cardiff University

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October 2020

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Acknowledgements

First, I would like to thank all of the participants who expressed enthusiasm and volunteered their time to take part, for without their contribution, this thesis would not be possible. I would also like to thank the cystic fibrosis centres which kindly hosted this research.

I would like to say a big thank you to Dr Jennifer Moses and Dr Anna McCulloch, who have shared many helpful insights, invested their time and ultimately supported me in undertaking this research. A big acknowledgement is also necessary for the 2017 DClinPsy Cohort; you are all brilliant and inspiring people.

This thesis would not exist without the support of my family and friends. You have all kept me going throughout these past three years, and all deserve acknowledgement! Dave and Harriet, thank you for lending your expertise. Paul and Donnchadh, thank you for your proof-reading. Sam, thank you for your helpful perspectives. Hel, thank you for being the best distraction I could have asked for. James, thank you for always inspiring me to be a better person.

The final and most important acknowledgement goes to my mum. Your love, wisdom and support has been integral in getting me here today. Thank you.

Preface

This thesis is submitted in partial fulfilment of the requirement for the degree of Doctorate of Clinical Psychology (DClinPsy). It is a portfolio thesis, and consists of two separate papers. Both papers have been prepared in accordance with the author guidelines for submission to the British Journal of Health Psychology.

Cystic Fibrosis

Cystic fibrosis (CF) is an inherited (genetic) condition that results in excess mucus in all bodily organs. The largest effect is in the lungs, and the majority of people eventually experience lung failure as a result. In order to attenuate the rate of organ damage, people with CF (PwCF) can engage in self-administered health-management regimes (HMRs). These are time-intensive, invasive, and can have a detrimental impact on perceived well-being.

The last four decades have seen life expectance improve exponentially, and the current ageing cohort of PwCF (aged 40 years and over) were not expected to survive childhood. Advances in knowledge and understanding have resulted in a linear increase in life expectance with time, which has meant that the life expectance of the ageing cohort has increased as they have aged, resulting in frequent prognostic-shifts.

Paper 1: Systematic Meta-ethnography

Studies have observed that PwCF's engagement in HMRs is often inconsistent with the prescriber's expectation; that is, people typically do not routinely engage with all their treatments. Qualitative studies have attempted to understand the perspectives of PwCF when engaging with their HMRs. A range of methodological approaches have been utilised, resulting in a variance of perspective. Paper 1 sought to synthesise the available qualitative studies, to provide a meta-perspective of the current research in the area and provide new

theoretical insights. A meta-ethnographic approach was used to facilitate this objective. The synthesis of 11 papers produced a conceptual understanding of the psychological processes involved in health-related behaviours for adults with CF.

Paper 2: Empirical Paper

To-date, research has not explored the experiences of PwCF when growing older. Further, there is no understanding about how PwCF adapt to older-age or to frequent prognostic shifts. Paper 2 sought to develop an understanding of both areas using a qualitative semi-structured interview design, and analysed elicited data using a constructivist grounded theory approach.

A tentative psychological model was iteratively developed through the simultaneous collection and analysis of qualitative interview data. The model describes a set of psychological processes that offer insight into how the participants have adjusted to new health insults and/or prognostic shifts. These processes are contextualised for the ageing cohort of PwCF, who were born into a time where expectations were bleak, and where the concept of an uncertain mortality has always been present. The model offers an understanding and explanation for how the participant sample responded to such existential uncertainty.

Summary

Both papers offer tentative, novel theoretical insights which have implications for clinical practice, service development and future research. Although framed from a psychological perspective, these implications extend to any healthcare professional supporting adults with CF.

The Experience of Adults with Cystic Fibrosis Engaging in a Complex Health Management Regime: A Meta-ethnography

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Systematic Meta-ethnography Word Count: 7894

This meta-ethnography has been prepared for the British Journal of Health Psychology. The author guidelines can be found in Appendix A.

The South Wales DClinPsy programme have placed a word limit of 8000 on this paper due to the removal of paper 3.

ABSTRACT

Purpose

Cystic fibrosis (CF) requires complex, self-administered health management regimes (HMRs) in order to maintain health and function, and slow progressive organ damage. The demands of HMRs are often unpleasant, invade on daily life and impact perceived well-being, and health-related behaviours often do not match with the prescriber's expectation. Qualitative studies have sought to understand how people with CF experience their HMRs. This meta-ethnography sought to provide an up-to-date synthesis of the available qualitative studies examining the experience of adults with CF in engaging with their HMRs, and to provide new insight into engagement with HMRs.

Methods

PsycINFO, ASSIA, CINHL, EMBASE and Medline databases were systematically searched to identify studies reporting qualitative interview data from adults with CF regarding engagement with their HMRs. Articles were excluded if they did not consider adult perspectives (child and adolescent samples) or interactions of their experiences and engagement with HMRs. Studies were quality appraised using the Critical Appraisal Skills Programme. A meta-ethnographic approach was used for synthesising the studies, which involved translation and re-interpretation of the study findings.

Results

A conceptual model emerged from the synthesis, which provides new insight into the psychological processes underpinning engaging with HMRs, from the perspectives of adults with CF. Complex and multi-factorial inter- and intra-personal processes interact, and collectively influence engagement with HMRs.

Conclusions

The model provides an opportunity to inform clinical practice, service development and future research. Avenues in which to support adults with CF in engaging with their HMRs during clinical encounters are considered.

INTRODUCTION

Cystic fibrosis (CF) is a life-limiting autosomal-recessive inherited condition, caused by a mutation on the CF transmembrane conductance regulator (CFTR) protein, and affects approximately 1 in every 2,500 new-born babies (CF Trust, 2019). CF results in thick mucus secretions, typically leading to progressive failure in multiple bodily organs, such as the lungs, pancreas and digestive tract (CF Trust, 2019; Lobo & Noone, 2014). The most prevalent effect occurs in the lungs, with the excess mucus causing recurrent chest-infections, leading to lung damage and ultimately respiratory failure in over 80% of people living with CF (PwCF) (O'Sullivan & Freedman, 2009). Many PwCF also develop CF related diabetes (CFRD), which brings added self-management demands to an already challenging health condition (Alves, Della-Manna & Albuquerque, 2020).

In the UK, there are approximately 10,655 people currently living with CF (CF Trust, 2019). Historically, CF was considered a life-limiting condition of childhood (Dodge & Lewis, 2005); however, this has improved considerably over the past four decades (McIntyre, 2013). The current 'ageing' generation of PwCF are predicted to have a median life expectancy of 29 to 34 years (95% CI). Whilst there are no accurate figures regarding the upper range, there are multiple reports of PwCF living into their 40s and less commonly even into their 50s, 60s and 70s (CF Trust, 2019). This predicted median life expectancy for PwCF born today is 49.1 years (CF Trust, 2019).

Living with CF brings with it arduous health-management regimes (HMRs) that take several hours per day (Sawicki, Sellers & Robinson, 2009), and include self-administration of prescribed physiotherapy exercises to clear mucus from the chest, medication, careful management of nutritional needs and lifestyle factors such as regular exercise (Kerem, Conway, Elborn & Heijerman, 2005; Yankaskas, Marshall, Sufian, Simon & Rodman, 2004; Mogayzel et al., 2013). Usually, some medication is inhaled via a nebuliser, such as antibiotics to manage chronic and

acute infections, and mucolytics to help clear the airway (Sawicki & Tiddens, 2012). Despite the complex HMRs, the responsibility for this falls entirely to the PwCF which can subsequently impact their perceived well-being (Alves et al., 2020). Typically, PwCF only receive support from healthcare professionals during routine check-ups and periods of acute exacerbation (e.g. Flume et al., 2009).

Understandably, engaging in such a demanding HMR can be extremely difficult for PwCF, especially given treatments can be unpleasant and have adverse side effects (Chopra, Paul, Manickam, Aronow & Maguire, 2015). Some have gone as far to say that CF could be one of the most difficult to manage chronic health conditions (Sawicki et al., 2009). Non-engagement in HMRs can significantly increase the risk of morbidity, mortality and healthcare costs (Kymes et al, 2016; Khan & Socha-Dietrich K, 2018), and lead to reduced quality of life (Benjamin & Sacks, 1994; Makela, Backer, Hedegaard, & Larsson, 2013; Patterson, Goetz, Budd, & Warwick, 1993; Rapoff, 1999; Strausbaugh & Davis, 2007). The important and necessary role for clinical psychologists in supporting PwCF with the psychological challenges of living with CF and engaging with HMRs was first recognised nearly 20 years ago (CF Trust, 2001; Havermans & Staab, 2016), and since then regular guidance has been published and updated (Castellani et al., 2018; Cystic Fibrosis Trust, 2011; NICE, 2017)

Engagement in Health Management Regimes

Compliance, adherence and concordance are three terms which are frequently used to describe a person's engagement with HMRs, and are not intended to be synonymous. Compliance denotes the extent to which the prescriber's advice is followed by the PwCF (Haynes, Taylor & Sackett, 1979; Stimson, 1974). Adherence superseded compliance; it implies the person's right to engage or disengage with advice, and accounts for the extent to which a PwCFs behaviour is consistent with agreed upon actions with the prescriber (Horne et al.,

2005). Concordance, which has not been adopted by the CF field, supersedes compliance and adherence and reflects a working alliance between the prescriber and recipient whilst respecting their beliefs and wishes (Bell, Airaksinen, Lyles, Chen & Aslani, 2007; Horne et al., 2005). Essentially, these terminologies imply a differing degree of power/powerlessness in relation to medical professionals/HMRs by PwCF (Stimson, 1974). The researcher has chosen not to use the terms adherence and compliance in relation to the CF literature due to these implicit connotations, and will argue instead that improved understanding of the engagement process is necessary (e.g. Michie, van Stralen & West, 2012).

In order to improve the engagement process of PwCF in HMRs, recent innovations to support this have been developed. For example, while medications, such as mucolytics and antibiotics, are effective at reducing the rate of exacerbations (e.g. respiratory) and improving/preserving lung function (Eakin, Bilderback, Boyle, Mogayzel & Riekert, 2011; Ryan, Singh & Dwan, 2011; Smith, Rowbotham & Regan, 2018; Yang & Montgomery, 2018), new technologies have been developed to facilitate PwCF's engagement with this medication (Tibble et al., 2020). Despite this progress in supporting engagement in HMRs, this area of research and innovation is in its infancy. Important topics, such as PwCF's engagement in optimising lung function and health, or applying psychological models of regime engagement has not received similar levels of investment or research (Amico, Mugavero, Krousel-Wood, Bosworth & Merlin, 2018).

There have been varying reports on the extent to which PwCF engage with their HMRs. Understandably this varies across the range of treatments, and figures from another study indicate that PwCF engage with their respiratory medication 62% of the time, digestive 88%, physiotherapy 41% and nutritional supplements 59% (Llorente, Garcia & Martin, 2008). However, it is important to acknowledge that self-report accounts are vulnerable to a number of cognitive biases (Kimmel, Lewis, Jaskowiak, Kishel & Hennessey, 2003; Paulhus & Reid, 1991; Wagner & Miller,

2004), and are therefore considered to be inaccurate predictors of engagement (Daniels et al., 2011).

Several researchers have also used pharmacy dispensary data in order to understand rates of engagement (e.g. Burrows, Bunting, Masel & Bell, 2002; Elkins et al., 2006; Modi et al., 2006; Quinn et al., 2004; Quittner, Drotar & Ievers-Landis, 2004; Zindani, Streetman, Streetman & Nasr, 2006). They found that PwCF engaged with 67% of their oral antibiotics, 31 – 53% of their inhaled antibiotics, 53 – 79% of their mucolytic agents and 41 – 72% of their hypertonic saline (Burrows, Bunting, Masel & Bell, 2002; Elkins et al., 2006; Modi et al., 2006; Quinn et al., 2004; Quittner, Drotar & Ievers-Landis, 2004; Zindani, Streetman, Streetman & Nasr, 2006). Again, there is no guarantee that PwCF have used their dispensed medications, and therefore there are limitations associated with this research also.

It is difficult to determine accurate rates of engagement with HMRs due to the unreliability of self-report measures and dispensary data. To overcome these limitations, Daniels and colleagues (2011) collected data from modern nebulisers, which record frequency of use. They found that despite self-report engagement ratings of 80%, objective engagement behaviour was significantly lower, at only 36%. Further, some more recent studies have used objectively measured nebuliser use alongside qualitative interview data to examine the range of factors influencing individual engagement (Arden, Drabble, O'Cathain, Hutchings & Wildman, 2019). These findings state that interventions should consider a PwCF's beliefs about engagement with HMRs and facilitate the development of healthy routines, habits and problem-solving skills (e.g. Michie, Atikins & West, 2014).

Understanding Engagement in Health Management Regimes

Healthcare professionals caring for PwCF are interested in factors mediating engagement with HMRs. Numerous studies have investigated the association between various psychological constructs (e.g. optimism, hopefulness and

avoidance) and engagement in HMRs (e.g. Abbot, Dodd, Gee & Webb, 2001). Substantive theories have been developed to understand and enhance engagement with HMRs for people living with chronic health conditions (e.g. Munro, Lewin, Swart & Volmink, 2007).

Given the proliferation of psychological theories to facilitate understanding of health behaviours, the Theoretical Domains Framework (TDF) was developed, which synthesised 33 behaviour change theories (including their unique constructs and components) into 14 key domains (Cane, O'Connor & Michie, 2012). These 14 domains were then conceptualised within 3 constructs: Capability; Opportunity; and Motivation, which together are purported to be implicated in health-related Behaviours (COM-B) and as such, could be a useful framework with which to understand engagement in HMRs (Michie et al., 2012; Michie et al., 2014).

Despite many empirical studies examining the associations between psychological constructs (e.g. optimism, hopefulness and avoidance; Abbot, Dodd, Gee & Webb, 2001), and the development of behaviour change theories (such as those synthesised in the TDF), interventions specifically aimed at improving engagement with HMRs are shown to have mixed effectiveness (Nieuwlaat et al., 2014), with some demonstrating poor outcomes for PwCF (Goldbeck, Fidika, Herle & Quittner, 2014; Quinn et al., 2004).

This has prompted qualitative researchers to attempt to understand the experiences (e.g. Badlan, 2006; Arden et al., 2019), perceived facilitators and barriers (e.g. Arden et al., 2019; Barker, Moses & O'Leary, 2017; Eaton et al., 2020; George et al., 2010; Hogan, Bonney, Brien, Karamy & Aslani, 2015) and meanings developed in relation to engagement with HMRs (e.g. Chapman & Bilton, 2004; Drabble et al., 2019; Grossoehme et al., 2020; Grossoehme et al., 2012; Oddleifson & Sawicki, 2017). There is now a growing body of qualitative research in cystic fibrosis, which prompted Macdonald et al. (2016) to undertake a meta-synthesis of the available research. Their review synthesised 8 studies, totalling 22 findings

aggregated into 4 categories, arriving at 1 synthesised finding. This proposed that adults with CF need to strike a balance between 'adhering' to HMRs in order to avoid the psychological burden of 'non-adherence', whilst attempting to fulfil a 'normal' lifestyle and quality of life. However, the sparsity of available articles at that time resulted in the authors including a heterogeneous sample that possibly overlooked or minimised more nuanced influential factors (e.g. the relational aspects of engagement behaviours (Chiang, Gui, Amico, Atkins & Lester, 2018)).

Aim of the Meta-ethnography

Given that psychological research regarding cystic fibrosis is in its infancy, it is a quickly evolving field. The meta-synthesis by Macdonald and colleagues (2016) is the first in the field. However, since its publication, the TDF (Cane et al., 2012; Michie et al., 2014) has been applied in quantitative studies in CF (Arden et al., 2019) and a range of qualitative studies offer additional insights that have not been captured within their synthesis. With more qualitative studies in this area there is opportunity for application of refined inclusion/exclusion criteria which focus more explicitly on engagement with HMRs, from a psychological perspective. Some of the latter publications also examine the role of engagement with HMRs through the lens of well-evidenced theories (e.g. Leventhal's revised Self-Regulatory Model (Leventhal, Halm, Horowitz, Leventhal & Ozakinci, 2005); Theoretical Domains Framework (Cane et al., 2012; Michie et al., 2011; Michie et al., 2014)).

Furthermore, in the UK an innovative targeted treatment that acts on the defective CTFR gene, 'triple combination therapy', has been licensed and included within clinical guidance since 2020 (Iacobucci, 2020). This treatment is expected to be efficacious for three out of five PwCF (dependent on their type of CF genetic mutation) in markedly improving clinical outcomes (Gramegna et al., 2020). This is also likely to have implications for engagement in health care regimes for many

PwCF (Pedemonte, 2020). Given all of the above, it is an important time to re-visit the literature.

METHOD

Focus of the Meta-ethnography

This meta-ethnography aims to synthesise primary qualitative findings regarding engagement of PwCF with their HMRs. Given that Macdonald's (2016) review cited both adolescent and adult perspectives, this meta-ethnography will exclude studies considering an adolescent perspective and/or the period of transition from parental/care-giver support to independence.

Rationale for Using Meta-ethnography

To synthesise the qualitative studies meeting the eligibility criteria (Appendix B), Noblit and Hare's (1988) seven-stage meta-ethnographic approach was used. See Appendix C for description of how these stages were followed. This approach is both theory-based, and has the potential to generate new theory by achieving a higher-level conceptual understanding of the primary qualitative data (France et al., 2019). This can be achieved by firstly synthesising primary data from eligible studies by juxtaposing data and examining the connections between them (Harvey, 2007), then by focusing on the interpretation rather than aggregation of the data (Walsh & Downe, 2005).

Search Strategy

The STARLITE pneumonic (Booth, 2006) facilitated the development of the search strategy. PsycINFO, ASSIA, CINHL, EMBASE and Medline databases were used for a global electronic search, and were searched on 14th August 2020 by the primary researcher. Search terms were informed by two main groupings; (1)

engagement, adherence and compliance; and (2) subjective experiences, quality of life and wellbeing (Appendix D). Terms utilised those from Macdonald et al.'s (2016) meta-synthesis protocol, and were also added to in order to elicit additional studies that explored issues from a psychological perspective. The search strategy utilised a combination of keywords and subject headings/indexed terms, combined using Boolean operators, which were modified depending upon the database and available controlled vocabularies. A process of 'forward-chaining' and 'back-chaining' was followed whereby citation and reference lists were reviewed from the eligible studies to ensure the search criteria were sufficient and that no studies were missed, as detailed by France et al. (2015).

Given on-going research since Macdonald et al.'s (2016) review, the researcher excluded papers that relied predominantly on adolescent samples, examined the transitional period from parental/care-giver support to independence, and those specifically examining cystic-fibrosis related diabetes, acute exacerbations (e.g. pulmonary) or organ transplantation. By creating a homogeneous set of studies, a more rigorous synthesis of adult experiences was enabled.

Search Outcome

The search strategy identified 1,108 papers, which was reduced to 1,074 after duplicates were removed. The titles and abstracts were screened against the eligibility criteria (Appendix B) by the primary researcher, and 25% of the studies returned were screened by two independent researchers. Any disagreements were resolved through discussion and mutual consensus. This process returned 30 papers that required full-text screening against the review criteria, and a further 19 were rejected due either to use of a quantitative methodology or because they did not explore the interaction between experiences of treatment and engagement with HMRs (PRISMA diagram in Figure 1). Again, 25% of the final 30 requiring full-text

screening were screened by two independent researchers, and any disagreements were resolved through discussion and mutual consensus. The rationale for the rejection of full-text papers is provided in Appendix E.

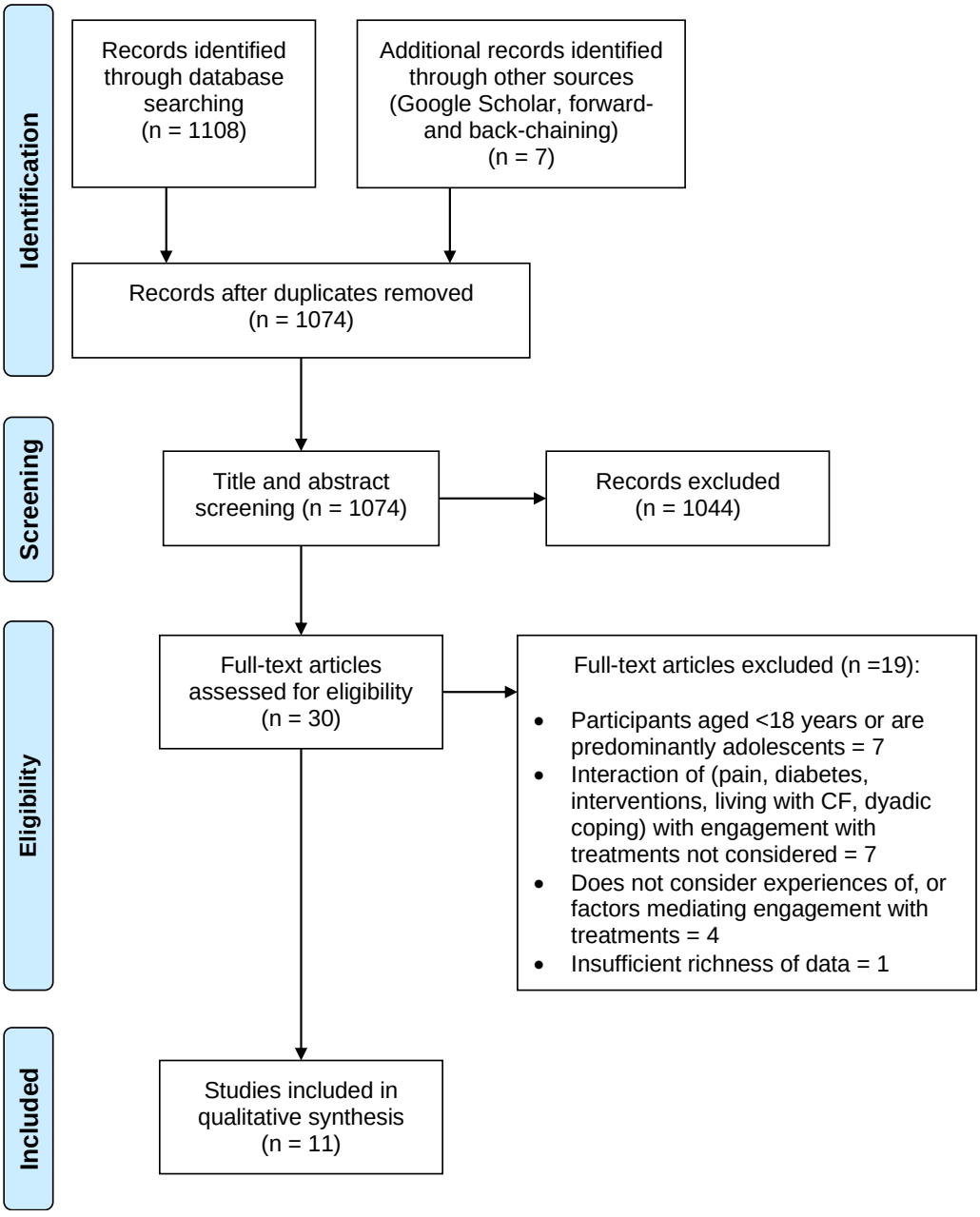

Figure 1. PRISMA diagram (Moher, Liberati, Tetzlaff & Altman, 2009).

Included studies covered a range of qualitative methodologies: framework analysis (n=1); framework *and* discursive analysis (n=1); thematic analysis (n=3);

grounded theory (n=2); hermeneutic phenomenology (n=1); ‘naturalistic enquiry’ (n=1); and interpretative phenomenological analysis (n=2). Data were collected from participants resident in: the UK (n=5); USA (n=5); and Australia (n=1). Full study characteristics can be found in Table 1 below.

Table 1. Characteristics of the included studies to be synthesised.

Source Paper (*n* = 11)	Country Setting	Sample (*n; age; sex)*	Ethnicity/Race/ Religion	Method of Data Collection	Method of Analysis	Aim
Chapman & Bilton (2004)	UK	*n* = 26 Aged 18 – 48 (M_{age} = 24) 'equally divided in terms of sex'	Not stated.	26 semi-structured interviews	Interpretative phenomenological analysis (Chapman & Smith, 2002)	To ascertain the extent to which patient thoughts about genetic determinism influences/interacts with their adherence behaviours.
Badlan (2006)	UK	**Overall:** *n* = 31 Aged 17 – 39 **Group:** *n* = 18 M_{age} = 28.4 9 male, 9 female **Individual:** *n* = 13 M_{age} = 26.2 8 male, 5 female	Not stated.	Seven group interviews (2 – 4 participants per interview) and 13 individual interviews. Structure of interviews not stated.	Hermeneutic phenomenology (Moustakas, 1994)	To explore the subjective perspectives of young people with cystic fibrosis so as to put the 'problem' of non-compliance into the context of their lives.
George, Rand-Giovannetti, Eakin, Borrelli, Zettler & Riekert (2010)	USA	*n* = 25 9 were in the 16 – 19 age category 10 were in the 20 – 29 age category 6 were in the 30 – 35 age category	Ethnicity not stated. *n* = 2 African-American *n* = 23 Caucasian	25 semi-structured interviews	'Naturalistic inquiry approach' (Lincoln & Guba, 1985)	To qualitatively explore the perceptions of barriers and facilitators of treatment adherence experienced by older adolescents and adults with CF.
Grossoehme, Ragsdale, Cotton, Meyers, Clancy, Seid & Joseph (2012)	USA	*n* = 12 M_{age} = 47 *n* = 10 female; *n* = 2 male	Ethnicity/Race not stated. *n* = 7 Protestant *n* = 2 Spiritual/'non-religious' *n* = 2 Non-religious *n* = 1 Unknown	12 semi-structured interviews	Grounded theory (Charmaz, 2006)	To explore whether adults diagnosed with CF after age 18 years would relate their spirituality with their CF.

Source Paper (*n* = 11)	Country Setting	Sample (*n; age; sex)*	Ethnicity/Race/ Religion	Method of Data Collection	Method of Analysis	Aim
Hogan, Nonney, Brien, Karamy & Aslani (2015)	New South Wales, Australia	*n* = 10 Aged 22 – 45 years *n* = 5 female; *n* = 5 male	Not stated.	10 semi-structured interviews	Thematic content analysis (Pope, Zeibland & Mays, 2000)	To explore experiences of adult patients with CF when taking nebulised medicines, identify factors that impact their adherences, and strategies they use to facilitate adherence to their nebulised medicine regimes.
Barker, Moses & O'Leary (2017)	UK	*n* = 9 *n* = 5 female; *n* = 4 male Aged 21 to 50	Not stated.	9 semi-structured interviews	Interpretative phenomenological analysis	To explore how people with CF experience being a parent, which includes the experiences of both mothers and fathers and the implications for healthcare professionals.
Oddleifson & Sawicki (2017)	USA	*n* = 14 *n* = 6 female; *n* = 8 male Aged 16 – 72 (median = 21)	Not stated.	26 unstructured interviews (including initial and follow-up interviews)	Thematic narrative analysis	To explore how recursive perception can interact with a person's daily decisions about self-care, medical treatments and lifestyle choices.
Arden, Drabble, O'Cathain, Hutchings & Wildman (2019)	UK	*n* = 18 *n* = 5 female; *n* = 13 male	Not stated.	18 semi-structured interviews	Framework analysis (Ritchie & Spencer, 1994)	To utilise the Theoretical Domains Framework (TDF) to assess the factors affecting nebuliser adherence in adults with CF to inform the development of an intervention.
Drabble, O'Cathain, Arden, Hutchings, Beever & Wildman (2019)	UK	*n* = 18 'mostly male' sample… distributed across age groups' Specific sex and age not stated.	Not stated.	18 semi-structured interviews	Framework analysis (Ritchie & Spencer, 1994) and further discursive analysis on extracts of data referring to 'forgetting'	To enable the functions that forgetting talk served to be made more visible, in particular, how speakers use forgetting talk to justify nonadherence including avoidance of accepting responsibility for their nonadherence.

Source Paper (*n* = 11)	Country Setting	Sample (*n; age; sex)*	Ethnicity/Race/ Religion	Method of Data Collection	Method of Analysis	Aim
Eaton, Beachy, McLean, Nicolais, Bernstein, Saez-Clarke, Quittner & Riekert (2020)	USA	*n* = 28 (*n* = 14 adolescents, *n* = 14 adults) **Adolescents** M_{age} = 15.89 *n* = 9 female; *n* = 5 male **Adults** M_{age} = 30.03 *n* = 9 female; *n* = 5 male	**Adolescents** *n* = 10 Caucasian *n* = 2 African-American *n* = 1 Hispanic/ Latino *n* = 1 Asian **Adults** *n* = 8 Caucasian *n* = 2 African-American *n* = 4 Hispanic/ Latino	28 semi-structured interviews	Thematic content analysis	To identify adherence barriers for which: (1) similar words were used to describe barriers, but people with CF and caregiver explanations of these barriers differed from healthcare providers; and (2) people with CF and caregivers perception of barriers that were different to healthcare providers.
Grossoehme, Cole, Lewis, Stamper, Teeters & Joseph (2020)	USA	*n* = 20 Aged 21 – 43 (M_{age} = 30) Sex not stated.	Ethnicity/Race not stated. *n* = 11 Religious **and** spiritual *n* = 3 Spiritual **but not** religious *n* = 3 Religious **but not** spiritual *n* = 1 Neither	20 semi-structured interviews	Grounded theory (Charmaz, 2006)	To: (1) qualitatively describe how adults diagnosed with CF as children use spirituality to construct meaning and to cope; and (2) to compare these qualitative results with how adults diagnosed with CF as adults described their use of spirituality in a prior study (Grossoehme et al., 2012).

Quality Appraisal

The 11 included papers were quality appraised using the Critical Appraisal Skills Programme tool (CASP, 2018). The CASP allows for a more standardised approach in quality assessing qualitative studies when interpreting their data for the purposes of a meta-synthesis (Dixon-Woods et al., 2007). It is comprised of 10 questions, the first two enable an additional level of screening for eligibility, and the further eight facilitate assessment of the study's credibility and relevance. Questions consider the research aims, design, recruitment strategy, data collection method, reflexivity of the researchers in interpreting the data, research ethics, rigour of data analysis, accessibility of the findings and utility of the findings.

To facilitate interpretation of the CASP, and therefore the extent to which findings of particular papers offer a credible perspective, a three-point (0, 0.5 or 1) scoring system was applied as developed by Feder, Hutson, Ramsay & Taket (2006), which has been used in many subsequent meta-syntheses (e.g. Duggleby et al., 2011; Elmir, Schmied, Wilkes & Jackson, 2010). To mitigate against any potential bias in the quality appraisal process, 25% of the eligible articles were also quality appraised by two independent researchers, and any disagreements were resolved through discussion and mutual consensus. For each article, the questions were calculated and totalled to produce a maximum possible score of 9 (the last item was not scored but appraised against three criteria, as illustrated in Table 2). Details of how each included paper was appraised and scored using the CASP is provided in Table 2 below.

Table 2. Scoring of the Critical Skills Appraisal Programme (CASP) for each of the included articles. Each article was assigned a score (in the final column) ranging from 0 to 9 to facilitate interpretation, as developed by Feder et al. (2006). A score of 0 was applied where there was little to no justification for a particular issue (e.g. the relationship between the researcher and participants was not considered), 0.5 if an issue was addressed but not sufficiently elaborated upon (e.g. the researcher made their views and potential biases explicit, but did not reflect on how this influenced the study or was mitigated against), and 1 if articles extensively justified and explained an issue (e.g. reflexive measures that were put in place to mitigate biases). A question mark is used to denote a 'can't tell' rating on the CASP.

Article	**CASP Question**										**Score**
	1	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	
Arden et al. (2019)	✓	✓	✓	✓	✓	✘	✓	✘	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✓	**7**
Badlan et al. (2006)	✓	✓	?	?	✓	✘	?	✓	✓	Contribution to the literature: ✓ Identified further areas for research: ✘ Considered generalisability: ✘	**6.5**
Barker et al. (2017)	✓	✓	✓	?	✓	?	?	✓	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✘	**7.5**

Article	CASP Question										Score
	1	2	3	4	5	6	7	8	9	10	
Chapman et al. (2004)	?	✓	?	?	✓	✗	?	?	✓	Contribution to the literature: ✓ Identified further areas for research: ✗ Considered generalisability: ✗	**5.5**
Drabble et al. (2019)	✓	✓	✓	✓	✓	✗	✓	?	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✓	**7.5**
Eaton et al. (2020)	✓	✓	✓	✓	✓	✗	✓	✓	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✓	**8**
George et al. (2010)	✓	✓	?	✓	✓	✗	?	✓	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✓	**7**

Article	CASP Question										Score
	1	2	3	4	5	6	7	8	9	10	
Grossoehme et al. (2012)	✓	✓	✓	?	✓	✗	?	✓	✓	Contribution to the literature: ✗ Identified further areas for research: ✓ Considered generalisability: ✓	**7**
Grossoehme et al. (2020)	✓	✓	✓	?	✗	✗	✓	✗	✓	Contribution to the literature: ✓ Identified further areas for research: ✓ Considered generalisability: ✓	**5.5**
Hogan et al. (2014)	✓	✓	✓	✓	✓	✗	?	?	✓	Contribution to the literature: ✓ Identified further areas for research: ✗ Considered generalisability: ✓	**7**
Oddleifson et al. (2017)	?	?	✓	✓	✓	?	✓	?	?	Contribution to the literature: ✗ Identified further areas for research: ✓ Considered generalisability: ✗	**6.5**

Data Abstraction and Synthesis

Britten et al. (2002) and France et al. (2019) discussed the notion of first-, second- and third-order constructs. First-order constructs represent the participant's views, accounts and interpretations of their experiences. Second-order constructs represent the views and interpretations of the participant's views by the authors of each included paper. Third-order constructs therefore represent the views and interpretations of the current researcher, of the second-order constructs. Thus, the aim of the meta-ethnographic approach is to develop a new, third-order conceptualisation and/or understanding of the first and second-order constructs.

Data abstraction and synthesis was guided by the principles and stages dictated by Noblit and Hare's (1988) meta-ethnographic approach (Appendix C). A process of 'reciprocal translation' ensued whereby the researcher utilised a constant comparative approach to juxtapose common themes and concepts from the included papers. This involved constantly reviewing the primary data in each paper, to ensure the commonality of themes and concepts. A process of 'refutational translation' followed, whereby the researcher searched for differences across the studies to ensure a level of rigour and that the nuanced aspects of each paper were not reduced or missed altogether. Although the included papers examined different aspects of engagement, few differences were identified amongst the papers, and the themes subsequently fed into a 'line of argument' synthesis. Contextual information, in particular the characteristics of the included studies (Table 1), was also considered at each stage when determining the relatedness of concepts.

Determining How the Studies were Related

Studies were carefully read and re-read in chronological order to identify the first and second order constructs. Key second order constructs were subsequently abstracted from each constituent paper (Table 3). To maintain the original context and relatedness of constructs, conceptual maps were drawn to note their

interactions (e.g. Britten et al., 2002; France et al., 2019). Analytic 'memos' were kept throughout this process, which influenced the development of subsequent third order constructs.

Table 3. Key (second order) themes, concepts and metaphors identified in each constituent paper.

Paper	(Second Order) Themes, Concepts and Metaphors
1. Arden et al. (2019)	Support of being in hospital; social support; planned reward; routine, cues and prompts; positive (praise) and negative (told off, negative outcomes) reinforcement; competing demands of life goals (e.g. holidays, travel); impact of stressful events (e.g. Christmas); impact of side effects; ability to self-monitor (objective nebuliser readings); 'intentional forgetting'
2. Badlan et al. (2006)	Visible versus hidden identities; 'normal' versus 'different' versus 'imposter'; interaction of health management versus life goals; biographical disruption; pressured into 'patient' roles
3. Barker et al. (2017)	Being a parent on compressed time; competing priorities (own versus child's needs); competing life demands (e.g. goals to have children versus maximise health); perceived importance of treatments; intentional non-adherence
4. Chapman et al. (2004)	Role of genotype versus phenotype; understanding of impact of environment; influence on locus of control; role of healthcare professionals in supporting understanding for people with cystic fibrosis
5. Drabble et al. (2019)	Perceiving self as more 'normal' by not engaging in treatments; role of routine and reminders; moral dilemma of (not) engaging in treatments; influence of normative expectations of treatment engagement; discourses to manage power dynamic between healthcare professionals and people with cystic fibrosis; role of 'forgetting' discourse
6. Eaton et al. (2020)	Miscommunication between people with cystic fibrosis and healthcare professionals; gaps in knowledge; locus of control; role of routine; impact of unexpected events; competing life demands (e.g. work, treatments, travel); facilitative role of attuned relationship between people with cystic fibrosis and healthcare professionals
7. George et al. (2010)	Influence of perceived treatment efficacy; role of stigma and embarrassment; role of social support; role of reminders, habit and routine; planned reward (e.g. later non-adherence, night out); perceived ease of treatment; role of the cystic fibrosis clinic; role of complementary/alternative treatments; role of 'forgetting' discourse
8. Grossoehme et al. (2012)	Role of spirituality in guiding treatments; re-framing illness identity; locus of control (accountability to God versus God-given condition); collaborative partnership with healthcare professionals (and God)

Paper	(Second Order) Themes, Concepts and Metaphors
9. Grossoehme et al. (2020)	Role of spirituality in guiding treatments; re-framing illness identity; locus of control (accountability to God versus God-given condition); role of perceived value and importance of treatments; role of social support (e.g. from congregation)
10. Hogan et al. (2015)	Role of perceived health benefits/treatment efficacy; influence of mood and embarrassment; influence of perceived responsibility (in order to maintain health and function); role of external and social support; role of routine and structure; importance of time management and organisational skills; competing demands (treatments, work, social life); role of (healthcare professionals in supporting) understanding of adaptability of treatments to meet competing demands; role of relationship with healthcare professionals
11. Oddleifson et al. (2017)	Influence of perceptions of self (and perceptions of how others perceive self); striving to be 'normal' ('identity performances'); influence of perceived treatment benefits/treatment efficacy; competing demands and priorities; time intensiveness of treatments; role of forgetfulness; influence of recursive perception with healthcare professionals

Translating Studies into One Another

This step necessitated that the researcher considered the relationships between the included studies and their exemplified constructs (as in Table 3). Following Britten and colleagues' (2002) approach to cross-tabulating the studies and their common constructs, the researcher constructed a Microsoft Excel spreadsheet (Appendix F), which went through several iterations. Studies were carefully read and re-read by the researcher using a 'constant comparative' approach to ensure that the concepts were grounded in the primary data. Eventually recurrent constructs were populated, and organised by the emergent third order constructs (the first column). The overall second order interpretations made by each study were also included to aid this process and preserve the validity of the constructs. An overview of the constructs is provided in Table 4. A detailed illustrative example of how a third order construct 'Conceptualisation of Health' emerged from first and second order constructs can be found in Appendix G.

To remain grounded in and preserve the integrity of the constructs, terminology was preserved wherever possible. For some third order constructs, terminology was borrowed from a constituent paper, such as the 'illness representation' construct that was used in multiple constituent papers (e.g. Chapman et al., 2004). Another example is 'locus of control', which was discussed to differing extents in some constituent papers (e.g. Grossoehme et al., 2020).

Table 4. Overview of constructs. Numbers associated with endorsed papers are carried over from Table 3.

Super-ordinate Third Order Construct	Sub-ordinate Third Order Construct	Second Order Construct(s)	Papers Endorsing Construct
Conceptualisation of Health	**Illness Representation**	• Degree of understanding of the role of the environment (i.e. treatments) on genes (phenotype). • Degree to which illness identity is accommodated into overall identity ('biographical disruption') and influence on visible versus hidden identities • Positive re-framing (e.g. in context of spiritual beliefs)	1, 2, 3, 4, 6, 10, 11
	Locus of Control	• Internal (phenotype, responsibility to self and others) versus external (genotype, God-given condition) • Perceived efficacy of treatments and influence on perceived control	1, 4, 7, 8, 9, 10, 11
Management of Health	**Behavioural Regulation Strategies**	• Structure and routine • Organisational and self-management skills • Availability of positive and negative reinforcement	1, 3, 5, 6, 7, 10
	Planned Non-adherence	• Reward for period of engagement with treatment • Engagement with life (e.g. social) events • Competing priorities (e.g. childcare, work)	1, 3, 5, 6, 7, 10, 11
	Alternative Coping Strategies	• Beliefs about complementary treatments • The function of 'forgetting' discourses	1, 2, 5, 7
External Influences	**-**	• Role of the cystic fibrosis clinic (e.g. problem solving, re-framing, motivation/reinforcement) • Relationship and communication between healthcare professionals and people with cystic fibrosis • Dynamic between healthcare professionals and people with cystic fibrosis (e.g. 'patient' roles) • Availability of social support	1, 2, 4, 5, 6, 7, 8, 10, 11
Practical Influences	**-**	• Perceived 'ease' of treatments • Balancing competing demands (e.g. treatments, childcare, work) • Understanding of how to adapt treatments	1, 3, 6, 7, 10, 11

Synthesising Translations

By reading all the constructs from the cross-tabulation, alongside analytic 'memos' and concept maps, the researcher was able to ascertain associations and relationships between the included studies. It was apparent that the study findings were not refutations of each other, and this remained true even when constructs were not elucidated in a particular paper (represented by empty cells in the cross-tabulation, Appendix F). Rather, the constructs appeared to examine engagement behaviours from a range of perspectives and appeared reciprocal (Thorne, Jensen, Kearney, Noblit, & Sandelowski, 2004), subsequently leading to a line of argument that provided further insight into patterns of (non)engagement with HMRs. As per Britten et al. (2002) and France et al. (2019), the line of argument consisted of a synthesis leading to third-order constructs, developed from the first- and second-order constructs of the included papers.

RESULTS

Quality Appraisal

The translation of constructs into a line of argument synthesis resulted in four over-arching constructs which were well endorsed: 'Conceptualisation of Health'; 'Management of Health'; 'Practical Issues'; and 'External Influences'. Comments are made throughout regarding the endorsement of each individual construct. Although some of the synthesised studies are of lower quality (as illustrated in Table 2 above), they typically share a conceptual space with studies of higher quality. It could therefore be argued that the studies which were rated as lower quality did not adversely affect the overall synthesis. Where constructs are commented upon as being well-endorsed, and no comment is made regarding the quality appraisal, the reader should assume that these constructs are well-endorsed by sufficient high-rigour studies.

There were ‘subordinate-constructs’ that were less-well endorsed, such as the ‘Alternative Coping Strategies’ and aspects of the ‘Behavioural Regulation Strategies’ construct (regarding positive and negative reinforcement), whereby the quality appraisal of the synthesised studies is more paramount. Where constructs were endorsed by few papers and where those papers are of mixed quality, they were not raised to stand alone constructs within the model. For example, the above constructs were translated into the super-ordinate construct of ‘Management of Health’ because individually they were endorsed by a small number of studies of mixed-quality. Such super-ordinate constructs should be regarded more tentatively.

Main Results

First- and second-order constructs that were exemplified within the included papers were translated into one another (as described above), which led to the emergence of several third-order constructs. Appendix G provides an illustrative example of how a third-order construct ‘Conceptualisation of Health’ was borne out of the translation of first- and second-order constructs from the included papers. The emergent third-order constructs were then synthesised into a line of argument, arriving at two over-arching constructs, which resulted in the model depicted in Figure 2 below. As above, this was achieved by scrutinising the cross-tabulation of constructs, analytic memos and concept maps. This required a constant-comparative approach, reading and re-reading of the included articles, to ensure that the interpretations being made by the researcher were valid and grounded in the original first- and second-order constructs. The extent to which each construct was endorsed by the synthesised papers is commented upon throughout the results, and a critical discussion about the quality appraisal process and weighting of various constructs within the model is offered in the discussion section. Although components of the model are discussed separately below, the model’s constructs interact and represent psychological processes.

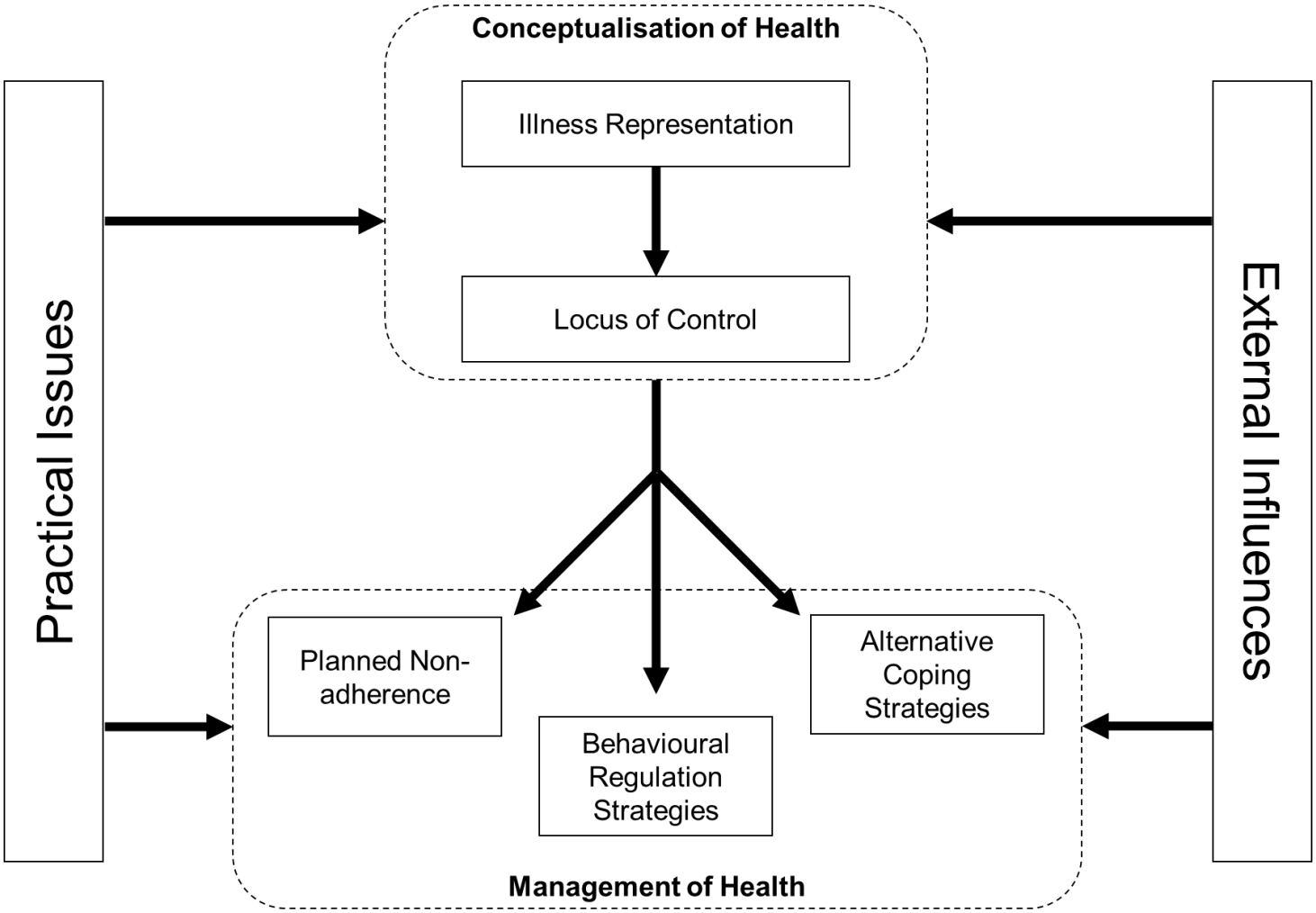

Figure 2. Model developed from a line of argument synthesis of the included papers.

Conceptualisation of Health

The synthesis of constructs resulted in an over-arching construct 'Conceptualisation of Health'. This construct concerns the internal conceptualisation, and subsequent identities and meanings that PwCF construct of their CF (illness representation). This was often based on previous experiences of their condition (and illness severity and/or instability) and heavily inter-related with that is how ready, able and willing they feel (either consciously or unconsciously) to be in control of their CF (locus of control). This construct is also impacted upon by the perceived 'external influences' and 'practical issues' constructs, discussed below. The two constructs that constituted this process were the most heavily endorsed by the included papers. How CF is internalised (e.g. genetic condition given by God; appreciation of the role of environment on genes (phenotype)) is proposed to have a direct influence on the perception of control over the condition (e.g. external/'spiritual' locus of control; perceived responsibility and accountability to self and others).

As described above, illness representation is a construct that was discussed in multiple constituent papers (Arden et al., 2019; Badlan et al., 2006; Chapman et al., 2004; Eaton et al., 2020; Hogan et al., 2015; Oddleifson et al., 2017). Authors that interpreted participant accounts within this concept referenced Leventhal's Self-Regulatory Model (Leventhal, Leventhal & Contrada, 1998), wherein illness representation makes up one component. Consistent with this model, data from the included studies mostly endorsed the 'identity' aspects of illness representation. Many papers discussed the trade-off between identifying in a 'patient' or 'sick' role and the dissonance this creates with wanting to fulfil a 'normal' societal role (e.g. being a parent) (Arden et al., 2019; Badlan et al., 2006; Barker et al., 2017; Eaton et al., 2020; Hogan et al., 2015; Oddleifson et al., 2017) . Psychologically, this reflects a tension between values in which PwCF can live by and express, such as

maintaining health and avoiding illness versus maintaining meaningful roles which are fulfilling.

> *"If I was in a social situation, out to dinner and a movie, people over, party off with friends spending the night, I didn't want to feel inadequate; I didn't want to feel lesser than they did. And for some reason in my head I was thinking that because I have this illness that I was not as good as they were so I didn't feel like I should be taking medication in front of them." (Hogan et al., 2015, pp. 90).*

This dissonance was conceptualised by one of the studies as the degree of 'biographical disruption' (Badlan, 2006) that PwCF experience, which could manifest as a sense of being an 'imposter in a normal world'. Although coined by Badlan, this concept has resonance across all the cited studies to some extent. This was considered to be a dynamic process, illustrated by one participant's account: *"It's unusual, it's a bit odd isn't it really 'cos one minute you feel like normal… then you don't feel normal, it's like swings and roundabouts"* (Badlan, 2006, pp. 267).

Further, identifying towards the 'normal' end of the spectrum, at least for some, 'sharpened' their focus and appeared to be facilitative of treatment engagement (*"He keeps me focused to stay alive basically to stay healthy as I can to see him grow up and not get into any trouble.",* Barker et al., 2017, pp. 748). However, when more aligned with an 'imposter' or 'abnormal' identity, this appeared to lead to embarrassment, perceived stigma and/or have an impact on mood, which in turn had a negative effect on engagement with treatments (Badlan et al., 2006; Drabble et al., 2019; George et al., 2010; Hogan et al., 2015; Oddleifson et al., 2017). It appeared that this reflected a process of tolerating (their) difference, for example if intolerant of difference then engagement in treatments was likely to be reduced if it made their CF visible. PwCF frequently disclosed that they would strive to be more normal by intentionally not engaging with treatments, which was influenced by contextual factors ('external influences').

> *"It's frustrating. All my friends can kind of do whatever they want. They don't have to maintain their health in the same way I do. I want to be like that. But I can't. And they don't fully understand." (Oddleifson et al., 2017, pp. 73)*

> *"When I was married I wouldn't do my breathing treatments around my wife…like my nebuliser, after I use it, I push it under my bed. I guess maybe I am hiding it…It is more my concern how they are going to react seeing their sick friend, sick husband, sick boyfriend." (George et al., 2010, pp. 427).*

> *"I work full-time and I work probably 50 hours a week right now so wanting to have a 'normal' life, where you go out to get drinks with friends, go to spend time with friends, or go for a bike ride, or a run or do something to try to maintain a normal life. The most difficult part is fitting the treatments into that." (George et al., 2010, pp. 427).*

PwCF's beliefs about the cause of their condition interacted with the construct of illness identity and influenced perceptions of (locus of) control. Across the papers, CF was seen as having a genetic cause, but how this was interpreted and understood by PwCF appeared to have a consistent and significant influence on their engagement with treatments (e.g. *"The genes are already there aren't they? You can't do nothing about it at present",* Chapman et al., 2004, pp. 378) (Chapman et al., 2004; Grossoehme et al., 2012; Grossoehme et al., 2020). Perhaps because some comorbidities of CF are easier or more difficult to control (e.g. excess mucus managed with physiotherapy or an airway clearance vest), and that their control may have limited immediate perceived benefits (e.g. Arden et al., 2019; George et al., 2010) subsequent perceptions of their efficacy as treatments (e.g. Oddleifson et al., 2017; Grossoehme et al., 2012; Grossoehme et al., 2020) may be impacted and consequently perceptions of their relevance to maintaining health destabilised.

> *"I can tell the difference when I do my Vest and when I don't…When I don't do my enzymes, I can definitely tell the difference. My stomach hurts." (George et al., 2010, pp. 428).*

> *"So it's like what's the point in taking all this medication if 6 months down the line let's say, it turns out that it's really done nothing, it's been a waste of time" (Arden et al., 2019, pp. 366)*

A mediation process was revealed in how PwCF believed the cause of their CF and their day-to-day symptomology (e.g. coughing, chest infections) was genetic

but felt the efficacy of various treatments (e.g. nebulised antibiotics) influenced their sense of reduced (locus of) control. Some second-order constructs examined this through the lens of the nature/nurture debate (Chapman et al., 2004), whereas others through a spiritual lens (Grossoehme et al., 2012; Grossoehme et al., 2020). Those PwCF who evidenced beliefs that biology (phenotype) was not deterministic and could be modified with treatment or environmental adaptation were more likely to have a hopeful outlook regarding their treatment efficacy. These PwCF tended to report a sense of responsibility to either themselves or others (e.g. God, healthcare professionals).

Management of Health

This concept reflects how PwCF self-regulate their response to CF, using behavioural regulation strategies, alternative coping strategies and/or planned non-adherence. In the emergent model, these regulatory responses are proposed to be directly influenced by the Conceptualisation of Health concept, as well as both external influences and practical issues (discussed below). Individually, the constructs underpinning this construct were endorsed by at least half of the included papers, where some were of mixed quality (Table 2). However, collectively, 'Management of Health' as an over-arching third-order construct is heavily endorsed by all included papers (Arden et al., 2019; Badlan et al., 2015; Barker et al., 2017; Chapman et al., 2004; Drabble et al., 2019; Eaton et al., 2020; George et al., 2010; Grossoehme et al., 2012; Grossoehme et al., 2020; Hogan et al., 2015; Oddleifson et al., 2017), and without refutational arguments, fed into a clear line of argument.

Behavioural Regulation Strategies

This construct, although not evident in all papers, received endorsement from some of the highest quality synthesised studies. This construct refers to the strategies that are directly within the person's control. An almost unanimous report was that if treatment regimens were embedded within a robust structure and routine this had a significant influence on engagement with treatments, *"it comes down to good time management"* (Hogan et al., 2015, pp. 90) (Arden et al., 2019; Barker et al., 2017; Drabble et al., 2019; Eaton et al., 2020; George et al., 2010; Hogan et al., 2015). This appeared even more important for PwCF balancing multiple roles (e.g. a patient with CF, striving for 'normality', being a parent), and required a degree of organisational ability and self-management (Arden et al., 2019; Barker et al., 2017; Hogan et al., 2015). This finding also converges with the common experience of some 'practical issues' such as staying away from home, long travel, holidays or stressful occurrences subsequently breaking that routine and having an adverse effect on treatments (Arden et al., 2019; Eaton et al., 2020; Hogan et al., 2015).

> *"It's like being on a diet. For a few weeks, you are like hardcore into it and you are totally serious and you do it every single time. And then [after] several weeks you fall off the wagon. And that's it, you're done. Nothing for weeks. Then you feel guilty enough to get back on the wagon. Or you get sick or something happens." (Oddleifson et al., 2017, pp. 72).*
>
> *"It definitely is a routine mindset . . . I don't even think about it in the morning particularly, if I'm going to work" (Arden et al., 2019, pp. 368).*
>
> *"On the whole I'd say it's alright like majority of the time I feel like I manage it quite well but I think that sometimes I don't know I think more recently I've noticed it that like if I get out of a routine then it becomes difficult to manage" (Arden et al., 2019, pp. 370)*

Less consistently reported (and subsequently less well endorsed) was the notion of embedding positive and negative reward systems into their routines (Arden et al., 2019; George et al., 2010). This aspect of the construct should therefore be interpreted more tentatively. For example, the delaying of something pleasant (e.g. accessing social media) until after treatments were completed, whereas some

reported that they engaged with treatment regimes to mitigate the experience of guilt for not doing treatments.

> *"I love Facebook. I made this deal that I couldn't go on Facebook until after I had done my therapies for the day"* (George et al., 2010, pp. 428-9).

Planned Non-adherence

This construct received good endorsement from studies of mixed quality. Given that there were no refutational constructs/data, this was not perceived to adversely affect the synthesis of this construct. The construct reflects an intentional choice that PwCF make to disengage with their treatment regime (Arden et al., 2019; Barker et al., 2017; Drabble et al., 2019; Eaton et al., 2020; George et al., 2010; Hogan et al., 2015; Oddleifson et al., 2017). Closely connected with behavioural strategies, some PwCF planned a period of non-adherence as a reward for their earlier engagement in treatments, e.g. to have a night off from treatments (Arden et al., 2019; George et al., 2010). Similarly, if there were particular events planned (e.g. a social event) or competing priorities (e.g. caring for child) then participants often made an active decision to not engage with their treatments at these times (Arden et al., 2019; Eaton et al., 2020; Oddleifson et al., 2017). Understandably, this also closely relates to the construct 'Conceptualisation of Health', whereby PwCF often wished to feel more 'normal' within social contexts. For some, intentional non-adherence was a dichotomy whereby either they met their own needs (engage with treatments) or met their other responsibilities (e.g. caring for their child, work commitments) (Arden et al., 2019; Barker et al., 2017; Oddleifson et al., 2017).

> *"and I was supposed to take it 4 times a day. Even, when I were proper on it I'd only take it 3 times a day" (Arden et al., 2019; pp. 372)*

> *"Due to the seasonality of my job, you could say I neglect myself. I try and put more effort into my work and my social life than I do my physical state and over the past 2 years in the same job I don't feel it has sinned me." (Badlan et al., 2006, pp. 267).*

> *"Not wanting to let CF run the show all the time…you feel better when you are not doing your treatments." (George et al., 2010, pp. 430).*

Alternative Coping Strategies

Alternative coping was less well explored in the included papers, however the researcher felt that it contributed usefully to the emergent model. Similarly to 'Planned Non-adherence' above, although less well endorsed, at least half of the papers were of good quality, and there were no refutational data, meaning that the lower quality papers did not adversely influence the synthesis of either aspect of this construct. The alternative coping construct consisted of two elements: engagement in pro-health behaviours other than medical treatments (Arden et al., 2019; Badlan et al., 2006; George et al., 2010); and coping with the emotional experience associated with (non)engagement with medical treatments (Drabble et al., 2019; George et al., 2010).

Some participants experienced a strong belief that engagement in 'complementary' therapies, such as hiking, swimming, running or going to the gym, were important aspects of maintaining their health in relation to CF, and substituted these for clinically required CF treatment regimes (e.g. "*Riding that bike, in my mind replaces the Vest"* (George et al., 2010, pp. 430); *"I keep myself active and get more benefit from doing those activities than I get from physio" (*Badlan et al., 2006, pp. 267)). It is possible that opting to engage in alternative coping strategies re-locates the (locus of) control within the PwCF, as opposed to this being located externally between them and their clinician. What is not clear from the synthesised data, is whether there is perceived equity between regimes and if PwCF 'forget' to engage with their alternative coping strategies (as discussed below). It is also possible that there are differing beliefs about the efficacy of these alternative therapies as opposed to their clinically required treatments. Both of which are likely to inform their illness representation (Conceptualisation of Health).

The other facet of alternative coping relates to management of the emotional experience of (non)engagement with treatments (Drabble et al., 2019; George et al., 2010). The perceptions of PwCF regarding 'adherence', although not explicitly voiced, were consistently interpreted as a normative behaviour, aligned with an assumption that one should engage with 100% of one's treatments. This belief appeared to be formed from the interpersonal context (e.g. interactions with healthcare professionals). Whilst some PwCF would acknowledge they were unable to achieve 100% engagement (*"It's just life. It's never going to be absolutely hundred percent is it?",* Arden et al., 2019, pp.370) others were hopeful that they could achieve 100% (Arden et al., 2019). However, as established and exemplified, this outcome from any CF treatment regime is unlikely, due to the significant demands of the regime in the context of trying to live a 'normal' life. Therefore, a common finding across these papers was that PwCF engage in a 'forgetting' discourse, which has been interpreted as a mechanism used to manage the emotional consequences of not meeting self-imposed treatment standards (Drabble et al., 2019; George et al., 2010). A forgetting discourse created a more 'morally' and 'socially acceptable' rationale for not engaging with all treatments. This process implies that some PwCF are locating the (locus of) control in the interaction with the 'other', e.g. health care professionals.

> *"I think the consequences of that [missing your nebuliser] is you know sometimes it can lead to sort of forgetting a bit more often but I think you know the times that I forgot recently, I've sort of known about it and I've been more conscious about it" (Drabble et al., 2019, pp. 2126).*

External Influences

This construct represents a fundamental aspect of the model, synthesised from the included papers (Arden et al., 2019; Badlan et al., 2006; Chapman et al., 2004; Drabble et al., 2019; Eaton et al., 2020; George et al., 2010; Grossoehme et al., 2012; Hogan et al., 2015; Oddleifson et al., 2017). It is well endorsed by first-

and second-order constructs, and despite being heterogeneous in its nature, all sub-ordinate constructs evidently had an influence on both super-ordinate third order constructs (Conceptualisation of Health and Management of Health).

Consistently reported was the role of the CF clinic in supporting engagement with HMRs (Arden et al., 2019; Eaton et al., 2020; George et al., 2010; Grossoehme et al., 2012; Hogan et al., 2015). Often, PwCF visit the same clinic, and the same healthcare professionals, for many years. This has the potential to be facilitative of engagement with HMRs, as there is opportunity to develop long-term relationships where professionals can adopt active-listening skills, experience empathy for the day-to-day struggles of the PwCF, and facilitating re-framing, collaborative experimentation and active problem solving. Further, visiting the CF clinic is often the 'push' that PwCF require, whereby they can receive feedback on their health metrics (e.g. lung function) and experience positive (e.g. praise) and/or negative (e.g. disapproval) reinforcement from professionals (Arden et al., 2019; George et al., 2010). How PwCF prepared for clinic visits was not explored in the papers, however the papers suggest that the response of the clinician has a strong influence on a person's subsequent engagement in treatments.

> *"And that's when it really kicked in when I saw my lung deteriorating and it were like I can do a lot more than what I have been doing. That's what really did it. Ever since then I would say I have been between 80-90%" (Arden et al., 2019, pp. 372)*

> *"If I go to the doctor and my PFTS are lower than usual…it does make it more likely to take my medicine and be on a tighter regimen." (George et al., 2010, pp. 427).*

There can often be relational and communication-related issues between PwCF and professionals that have an adverse effect on engagement with treatments (Chapman et al., 2004; Badlan et al., 2006; George et al., 2010; Hogan et al., 2014; Oddleifson et al., 2017). Multiple PwCF reported the struggle of interpreting 'doctor speak', and it would often transpire that although professionals

and PwCF believe they are 'on the same page', their perception of the agreed upon treatment regime differs (*"sometimes doctors use these big words and it's like you don't know what it all means"* (Chapman et al., 2004, pp. 377)). This suggests a limited opportunity, perhaps due to limited external resources (e.g. time constraints on clinical sessions), for a strong attunement process.

Communication between professionals and PwCF also appeared to lack nuance. For example, PwCF derived a black and white understanding of messages and perceived there to be a 'right' and 'wrong' way to use treatments (e.g. Badlan et al., 2006; Hogan et al., 2015). This resulted in a perceived lack of flexibility and adaptability of treatments if, for example, competing demands (e.g. travel, work, parenting) arose. Understandably, this will affect a person's beliefs about treatment efficacy, and their perceived ability to engage with treatments (illness representation). PwCF may report forgetting or intend to be non-adherent (as above) when, had there been mutual understanding regarding their ability to engage with treatments more flexibly around other commitments, their health may ultimately have benefitted.

Lastly, in relation to the CF clinic, it was often reported that PwCF felt somewhat forced into a 'patient' role (Badlan et al., 2006; Barker et al., 2017; Drabble et al., 2019; Eaton et al., 2020; George et al., 2010). It was often the perceived expectation that professionals only viewed PwCF within their 'patient' or 'sick' roles, despite fulfilling many other roles and identities (e.g. parent, employee) outside of their CF (linked to their illness representation). This implies a constrained set of perceived needs by healthcare professionals regarding the management of CF, perhaps with an under appreciation of what maintains the PwCF's well-being (not reliably reflected in administration of standard psychometric measures, e.g. Gorbatenko-Roth, Levin, Altamaier & Doebbeling, 2001; Pinto, Fumincelli, Mazzo, Caldeira & Martins, 2017). At times, this appeared to push PwCF into the 'white

coat compliance' phenomenon and was reported to link with the 'forgetting' discourse discussed above.

An additional external influence was the degree to which PwCF reported they had social support (Arden et al., 2019; George et al., 2010; Grossoehme et al., 2020; Hogan et al., 2015). The type of social support varied between studies, from a friend asking after their treatments and wellbeing, to a partner (e.g. wife or husband). Although not reflected in detailed elaboration, the consensus was that this appeared to be a strong facilitative factor that was associated with engagement with treatment regimes. However, the mechanism for this influence was unclear (e.g. was it indirectly that emotional support from peers/partners enhanced capacity to subsequently engage in treatments?).

Practical Issues

This final construct 'Practical Issues' was well endorsed by the high-quality studies and translated from several first- and second-order constructs. Translation of this construct resulted in an emerging continuum, whereby the ease (and time commitment) of a particular treatment influenced the likelihood of the treatment being completed (i.e. the quicker and/or easier the treatment, the more likely it was to be completed) (Arden et al., 2019; Eaton et al., 2020; George et al., 2010; Hogan et al., 2015; Oddleifson et al., 2017). The perceived 'ease' of treatments was multi-factorial. One aspect concerned the presence (or not) of side-effects (Arden et al., 2019; Hogan et al., 2015), whereas another concerned the degree to which the PwCF had to balance competing roles/demands (e.g. providing care to child, balancing work demands, navigating unexpected events/stressors) (Arden et al., 2019; Eaton et al., 2020; Hogan et al., 2015; Barker et al., 2017; Oddleifson et al., 2017).

"If it's a choice between going away for work and taking my machine or taking meds that I think might get in the way, I'm probably going to choose work." (George et al., 2010, pp. 427).

"I think the most difficult thing is travel. When I'm traveling for work, I lose control of my schedule because I'm not necessarily in charge." (Eaton et al., 2020, pp. 1590).

DISCUSSION

This meta-ethnography provides a conceptualisation of the psychological research regarding the experiences of PwCF's engagement with HMRs, given the recent licensing of triple combination therapy (Gramegna et al., 2020; Iacobucci, 2020) and the subsequent consequences this will have for many PwCF in minimising their daily treatment regimens (Pedemonte, 2020). By utilising a meta-ethnographic approach (Noblit & Hare, 1988), this facilitated the synthesis and argument for a tentative conceptual model of psychological processes underlying engagement. The synthesised papers drew on a range of successively dominant theoretical models when designing their studies and this subsequently framed their data collection and analysis. Thus, it would be anticipated that the current model's derivation has recognisable theoretical alignment with continually revised understandings of the processes of engagement with HMRs for people living with chronic health conditions. The researcher argues that the model developed from this synthesis elaborates on these pre-existing theoretical models in application to the CF population, subsequently providing new insight and new avenues for clinical practice and research at a moment in time when triple combination therapy is becoming more widely adopted.

Summary of the Model

The model synthesised first and second-order constructs, many of which were explicitly elicited in relation to pre-existing theoretical models (e.g. the TDF and Self-Regulation Model; Cane et al., 2012; Michie et al., 2014; Leventhal et al.,

2005), and others more implicitly. There are four super-ordinate third order constructs, two of which represent more 'central' processes, and two 'peripheral' processes (Table 4). The central two concern how PwCF construct and internalise an understanding of their condition ('Conceptualisation of Health') using the relational space with others (e.g. healthcare professionals) and individually using cognitive behavioural processes. This is hypothesised to directly influence how they behave and manage their condition ('Management of Health'). Both processes are then hypothesised to receive influence from the two peripheral constructs: 'Practical Issues'; and 'External Influences'. Thus, the model has synthesised both internal and external influences on how PwCF make sense of (the 'treatability' of) their condition and the observable responses this influences in terms of their engagement with treatments.

Fit with Other Models

When considering more nuanced elements, there are several areas of conceptual overlap with pre-existing models. The most pertinent overlap is the adoption of the 'illness representation' label in the present model, which is a key component in Leventhal and colleagues' (2005) Self-Regulatory Model (SRM). The SRM has been widely applied in the field of health psychology (e.g. Leventhal et al., 2005; Benyamini & Karademas, 2019) and considers a person's illness identity and locus of control (amongst other elements), internal and external influences on these constructs, and the direct impact they have on illness-related behaviours. The 'Conceptualisation of Health' construct in this current model closely mirrors this well-regarded theoretical construct (Leventhal et al., 2005). The SRM also purports that illness-related behaviours (e.g. engagement in treatments; appraisals of treatment efficacy) has a subsequent impact on their illness representation (e.g. controllability), similarly to the current model. However, where the two models diverge is in appreciating the congenital nature of CF, arduous HMRs, high (internal

and external) ‘adherence’ expectations and the challenge of maintaining perceived well-being.

Strengths and Limitations of the Meta-ethnography

By utilising a systematic search strategy (based upon a well-endorsed framework (Booth, 2006; France et al., 2019)) and a well-established systematic approach for synthesising the identified first and second order constructs (France et al., 2019; Noblit & Hare, 1988) to develop new theoretical insight, this helped to enhance the reliability of the review. Further, steps were adopted to mitigate undue bias on the part of the researcher, such as involving independent researchers during the screening and quality appraisal phases of the review. For the reader to further assess the review’s credibility, it is important to consider the epistemological heuristics of the researcher, and the influence that this could have had on the synthesis. As described (Appendix C), bracketing processes were necessary to attenuate biases from the researcher’s own preconceptions when building the themes and key constructs. One un-mitigating bias is the researcher’s position as a Trainee Clinical Psychologist, and interpretation of research through a psychological lens (versus a medical lens, discussed below). The validity of the review is further strengthened by the clear resonance with existing theoretical models (e.g. the SRM), whilst also recognising how the current model diverges given the unique context for PwCF.

The current meta-ethnography is however limited by the epistemological foundations of the synthesised papers. It is apparent that all the included papers framed the problem as ‘adherence’ or ‘compliance’, and commonly placed this problem within the PwCF. On a superficial level, PwCF hold the *responsibility* for engaging in their HMRs. However, given that the collection of data in the included studies occurred through the lens of ‘(non)adherence’, this immediately limits the scope of elicited participant responses. Quality appraisal using the CASP does not

explore the theoretical underpinnings of a study, however it is apparent that only two of the included studies (Arden et al., 2019; Chapman et al., 2004) stated these explicitly. Therefore, it is difficult to ascertain the context of some of the studies and whether sufficient bracketing practices (e.g. Ahern, 1999) were adopted by the authors of the included studies. The second order interpretations of 'sick', 'patient' and 'normal' roles, for example, further implies that the problem is located within the PwCF, whereas the third order interpretation in the current synthesis was one of psychological dissonance resulting from a conflict in values. The labelling of such roles further perpetuates a disconnect between PwCF and healthcare professionals, which is problematic given the current model indicates that the development of an attuned relationship with healthcare professionals can be facilitative of engagement (*concordance*) with HMRs.

Further, some constituent papers (Arden et al., 2019; Drabble et al., 2019) arbitrarily categorised participants by level of adherence (e.g. 'low-' and 'high-adherers'), and pursued their analyses using these crude categories. It is therefore possible that this adversely influenced the development of the model. To mitigate against this, the researcher ensured that concepts were well-endorsed by a range of papers, including at least some papers of higher quality.

Clinical and Theoretical Implications

The model indicates that clinical interventions to improve engagement need to be sophisticated and multifactorial. For example, interventions that have targeted 'forgetting' in isolation have reliably low effectiveness (Choudhry et al., 2017; Kahwati et al.,2016). Of course, this could be due to people managing with the emotional burden of choosing not to engage in treatments, as discussed in the model. The model would indicate that interventions would both need to target internal factors (e.g. the person's illness representation and sense of (locus of) control), and the external factors influencing this process (i.e. practical issues and

external influences such as the relationship and discourses with healthcare professionals). When considering the current model in the context of other health psychology models, it is also apparent that there are gaps in the research relating to the CF population, with recommendations needed to further substantiate the current model. For full consideration of the clinical and research implications, refer to Table 5.

Table 5. Implications for clinical practice and research based upon the findings of this meta-ethnography and emergent theoretical model.

Clinical and Research Implications
• Future research should seek to make sense of PwCF's perceptions of the relational space through dyadic qualitative research, to elaborate on (dyadic) perceptions of the 'relational adherence space' (e.g. clinical interactions, interactions with significant others). Currently only one paper (Werner, Hochman, Rosenne & Kurtz, 2020) considers the role of dyadic coping more generally when living with CF, but not in relation to treatments.
• Research concerning engagement with CF treatment regimes is framed from an 'adherence' perspective and largely grounded in a medical model (that the barriers/facilitators of 'adherence' exist within the person). It would be useful for the CF field to consider the adoption of 'concordance' when engaging with and supporting PwCF with their engagement with treatments.
• The model indicates the influential role that an attuned relationship between a PwCF and their healthcare professional can have on supporting their engagement with treatments. Given the noted barriers to this, the model would suggest that organisations adopting a more compassionate leadership style (West & Chowla, 2017) could be facilitative of engagement behaviours, as has been indicated in other areas of chronic illness (e.g. Trzeciak, Roberts & Mazzarelli, 2017).
• There is a need for careful consideration and exploration of 'forgetting' discourses, as although they may be authentic, they may also serve to protect PwCF from experiencing guilt and/or shame regarding their treatments. Thus, application of compassion theory (as applied for other conditions, e.g. Munro et al., 2007) could help to normalise the impracticability of engaging with 100% of treatments, whilst not letting PwCF 'off the hook'.
• There may be a need for preparatory work with PwCF to explore their illness identity (representation) (e.g. their understanding of CF, how CF has been incorporated into their identity or how this clashes with their sense of self), given the influence this is hypothesised to have in health management behaviours.
• A role for clinical supervision and team formulation approaches (Johnstone & Dallos, 2014). If clinicians become complacent and/or collude with the PwCF's limited engagement with health management regimes, then PwCF may not get the necessary positive and/or negative reinforcement they need to support them in developing sufficient motivation to engage with treatments.
• Interventions targeting individual elements of this model are unlikely to be effective in supporting PwCF with engagement with their health management regimes. Interventions need to be multi-factorial, and consider both inter- and intra-personal processes.

As the developed model indicates, influences on PwCF's engagement with treatments are wide-ranging and involve many internal *and* external factors. As

illustrated above, the current research is limited based on its epistemological foundations. Thus, the researcher advocates the adoption of updated terminology in the field of CF, which may also be facilitated by organisations that embrace a compassionate leadership stance (e.g. West et al., 2017). Other fields of clinical health psychology have adopted the use of *concordance* to describe the complex processes involved in the interpersonal negotiation of engagement with their HMRs (Bell et al., 2007; Horne et al., 2005). A plausible hypothesis based on the model would be that focusing solely on constructs within the 'Management of Health' concept (e.g. 'adherence' behaviours) would be ineffective to support PwCF in managing their complex health condition, and shifting towards 'concordance' would facilitate approaches that target more global processes.

Conclusion

The current meta-ethnography offers a theoretical update regarding how PwCF engage with their HMRs, given the now wide availability of new treatments that will alter management regimes for future generations. The tentative theoretical model can facilitate the development of clinical interventions to support PwCF with their engagement with HMRs by applying a psychological understanding, and offers opportunities for positive organisational change. The model also provides scope for future research to further substantiate the model and test the psychological processes that have been hypothesised here to underpin engagement behaviours.

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Ageing with Cystic Fibrosis: How do Older Adults with Cystic Fibrosis Adapt to Change

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Empirical Paper Word Count: 7870

This empirical paper has been prepared for the British Journal of Health Psychology. The author guidelines can be found in Appendix A.

The South Wales DClinPsy programme have placed a word limit of 8000 on this paper due to the removal of paper 3.

ABSTRACT

Objectives

'Older' adults (>= 40 years) living with cystic fibrosis (CF) today were not expected to survive childhood. Life expectance has improved thanks to advances in knowledge and understanding, leading to this cohort experiencing frequent prognostic shifts. To date, there are no published studies examining the experience of adults with CF when growing older and negotiating these prognostic shifts. Further, existing psychological theories do not appreciate the nuances of growing older with CF. This study sought to capture the experiences of people with CF about growing older, and to understand how they re-adjust in-light of prognostic shifts.

Design

Data were elicited from 10 people with CF aged 40 years and over, using semi-structured interviews. The interview schedule was iteratively developed from simultaneous collection and analysis of interview data, according to theoretical sampling practices of grounded theory.

Methods

Qualitative data were analysed using a constructivist grounded theory approach. This included initial line-by-line, then later focused and theoretical coding practices. Memo writing enabled theoretical links to be made more explicit. A constant-comparison approach was used to facilitate data analysis, and remain grounded in the data. Reflexive practices were adopted throughout.

Results

A tentative theoretical model was borne out of the qualitative data analysis. This describes the psychological processes involved when people with CF re-adjust

to new prognostic information, and places these within the unique contextual factors influencing this ageing cohort.

Conclusions

This model has implications for clinical practice, current theoretical understanding and indicates avenues for future research.

INTRODUCTION

Cystic fibrosis (CF) is a life-limiting inherited condition, caused by a mutation on the CF transmembrane conductance regulator (CFTR) gene, and affects approximately 1 in every 2,500 new-born babies (CF Trust, 2019). CF results in thick mucus secretions, typically leading to progressive failure in multiple bodily organs, such as the lungs, pancreas and digestive tract (Lobo & Noone, 2014; CF Registry Report, 2019). The most prevalent effect is in the lungs, with the excess mucus causing recurrent chest-infections, leading to lung damage and ultimately respiratory failure in over 80% of people living with CF (PwCF) (O'Sullivan & Freedman, 2009). Many PwCF also develop CF related diabetes (CFRD), which brings added self-management demands to an already challenging health condition (Alves, Della-Manna & Albuquerque, 2020).

In the UK, there are approximately 10,655 people currently living with CF (CF Trust, 2019). When CF was first reported in 1938, it was poorly understood and there were limited medical treatments available (Davis, 2006), which resulted in the majority of children dying within their first year of life (Andersen, 1938; Simmonds, 2013). Over the past four decades there have been important advances in our understanding of CF, such as the identification of the CFTR gene in 1989 (e.g. Rommens et al., 1989), the development of novel treatments for CF symptoms such as the development of acid-resistant digestive enzymes to improve diet (Edwards, Clarke & Greenop, 2013) and triple combination therapy, a targeted treatment addressing some of the causal factors underpinning CF, which has recently been approved through clinical trials (Gramegna et al., 2020).

These advances in understanding and treatment have significantly altered the CF prognosis, and life expectance has increased significantly over the past two decades (McIntyre, 2013). The current 'ageing' generation of PwCF are predicted to have a median life expectancy of 29 – 34 years (95% CI). This is in stark contrast to those who are born today with CF, who have a projected median life expectancy of

49.1 years (CF Trust, 2019). This picture is also gendered, with males born today likely to live between 49 – 54.6 years, as opposed to 42.6 – 49.2 years for females (CF Trust, 2019).

The older PwCF living today will have been born prior to the identification of the CFTR gene and are likely to have experienced several different treatment regimens and been provided with revised prognoses over the years. Understandably, this will have influenced significant life choices, such as opting to start a family or establishing a career (e.g. Higham, Ahmed & Ahmed, 2013; Lomas & Fowler, 2010). Qualitative research examining the views of PwCF who have become parents has provided insight into the perceived time pressures of being a parent both on a limited life trajectory and with arduous health management regimes (Barker, Moses & O'Leary, 2017; Jacob, Journiac, Fischer, Astrologo & Flahault, 2020). Considering these findings, it is possible that those who decide not to make such significant life decisions in-light of earlier, outdated prognoses, may experience a degree of psychological distress (Simmonds, 2013).

Although medical knowledge has advanced significantly for PwCF (e.g. the advent of triple combination therapy (Gramegna et al., 2020)), research in the psychological domain has not always kept pace (e.g. Sherman et al., 2020). Some authors have speculated about the likelihood of increased psychological distress when growing older with CF (e.g. Simmonds, 2013), in the context of ever-changing external factors (e.g. medical advancements, increasing prognostic-shifts or increased comorbidities). Despite this speculation, large-scale UK-based epidemiological studies (*n* = 2,065 participants) have indicated that PwCF had an estimated prevalence of anxiety and depression that was similar to the general population (Duff et al., 2014). This is consistent with other large-scale studies outside of the UK (e.g. Goldbeck et al., 2010) but inconsistent with findings for other chronic health conditions such as chronic obstructive pulmonary disease, which

show significantly higher prevalence rates of depression and anxiety (Matte et al., 2016; Sampaio et al., 2019; von Siemens et al., 2019).

There are data from studies utilising psychometric measures exploratively that suggest PwCF not only are able to tolerate uncertainty to a comparable level relative to the general population, they also scored higher than the general population on measures of resilience (Mitmansgruber et al., 2016). However, there is no current psychological understanding as to why this is the case, or how older PwCF have managed with the complexities of growing older with CF, as described above. The role of clinical psychologists in the care of PwCF has been long reported (e.g. WHO, 1999), and has been part of UK standards since 2001 (CF Trust, 2001). Clinical psychologists have now been embedded in CF multi-disciplinary teams and it is possible that this model of care has given PwCF an opportunity to process their emotional responses to CF in a very live and timely way (NICE, 2017).

There have been numerous psychological theories developed, such as the Self-Regulatory Model (Leventhal, Halm, Horowitz, Leventhal & Ozakinci, 2005) and the Health Belief Model (Rosenstock, Strecher, & Becker, 1988) which are designed to inform understanding of how people living with chronic health conditions understand, manage and live with their condition. The researcher argues that there is perhaps no theoretical understanding that can account for the idiosyncratic nature of CF, given its genetic (and life-long) origins, and particularly in relation to the now 'older' generation that have experienced significant contextual changes.

However, one theory that helps to begin understanding these processes is hope theory (Snyder, 2002; Snyder et al., 1991) which has been applied to the field of health psychology when considering (typically acquired) chronic and/or terminal health conditions, such as spinal cord injury and cancer (e.g. Hill & Feudtner, 2018). In such settings, hope has been operationalised as a broader concept, which can be considered as: (a) a tangible hope (linked to a particular outcome); (b) hopeful

patterns of thinking (that are amenable to situation-dependent change); and (c) a general optimism that things will work out (Feudtner, 2014). The theory offers an explanation for how people living with chronic health conditions strike a balance between striving for one desired outcome (e.g. cure-seeking goals) and targets/goals that some may deem more adaptive (e.g. goals to achieve greater quality of life in the context of such circumstances, or engagement in palliative care) (Gum & Synder, 2002).

Although hope theory has not been applied to PwCF, it could inform hypothesis generation as to how PwCF adapt to constant change (e.g. new medical knowledge, new treatments or a deterioration in their health/new prognostic information). Hill et al. (2018) proposed that people living with chronic health conditions may need to 're-goal' in response to such changes, for example shifting from cure-seeking goals to engagement in palliative care. However, what is unclear is how this would apply for the ageing cohort of PwCF, particularly those diagnosed at or soon after birth. A unique experience for the current ageing cohort of PwCF is that having survived childhood, their predicted life expectancy has been shifting as they have matured, and so the concept of an approaching mortality has always been present. Although hope theory has some utility in understanding hopefulness in the ageing CF population, it does not provide a comprehensive theoretical understanding of the additional nuances that accompany the notion of prognostic shift. For example, despite rapid prognostic shifts, how do people maintain a sense of agency and hope? Is hope one of the processes that fosters resilience in the CF population?

Aim

The aim of the present study was to understand, from the perspective of 'older' PwCF, both the experience of growing older alongside such prognostic-shifts and how these shifts have been processed and managed from a psychological

perspective. Given the identification of the CFTR gene in 1989, and current median life-expectancy data, 'older' PwCF were defined as 40 years and over, consistent with other fairly recent studies (Rodman et al., 2005; Hodson et al., 2008; Simmonds, Cullinan & Hodson, 2009). Only people who were diagnosed with CF during childhood (which represents 82% of the CF population (CF Trust, 2019)) were recruited to ensure that they have lived through and experienced these medical and prognostic changes.

METHOD

A constructivist grounded theory approach was used to analyse qualitative interview data elicited from semi-structured interviews, as described by Charmaz (2006; 2014). This method of analysis was selected for the following reasons: (1) it is a hypothesis-generating approach that can be used to study a phenomenon whereby no current conceptual understanding exists; (2) it emphasises how a theoretical understanding can be systematically developed (constructed) by the researcher through an interactional process with the data; and (3) previous grounded theory approaches, such as Glaser and Strauss' (1967), suggest that theoretical understandings are 'discovered' and 'emerge' from the data, with less emphasis on the interactive construction of a conceptual understanding between the researcher and the data.

Researcher Positionality

Given the use of a constructivist approach, it was important for the researcher to recognise their position within the research and maintain (reflexive) bracketing practices (e.g. Ahern, 1999). It has been recommended that qualitative researchers maintain a process of reflexivity throughout the entirety of the research process (e.g. Pidgeon & Henwood, 1997). This started with the researcher recognising their own position within the research. At the time of undertaking the

study, the researcher (first author) was a Trainee Clinical Psychologist in his final year of doctoral training. Although the primary researcher had a general pre-existing awareness of CF, he did not have a sophisticated level of insight concerning matters directly related to CF. However, the researcher had pre-existing knowledge and experience of clinical (health) psychology, and theoretical health psychology models (e.g. the Health Belief Model (Rosenstock et al., 1988) and Self-regulatory Model (Leventhal et al., 2005)), and so did not approach the research from an entirely 'naïve' perspective.

The researcher also held a dual role; working as a clinician in children's services (not with PwCF) and a researcher working with adult PwCF, and therefore had to be reflexive in their stance within research interviews (e.g. Brinkmann & Kvale, 2005; Thompson & Russo, 2012). This required careful reflection to delineate the two roles appropriately, and to clearly express to participants the purpose of the *research* interview.

Rigour

Reflexive Bracketing

A criticism of qualitative research is the opportunity for subjectivity and bias, projected by the researcher(s) onto all stages of the study (epistemological foundations, construction of the interview schedule, data analysis/coding, expressing the results, critiquing the study; Elliot, 1999). Unlike previous (classical) versions of grounded theory (e.g. Glaser & Strauss, 1967), a constructivist approach allows the researcher to undertake the literature review prior to or during data collection and analysis (Charmaz, 2006; Thornberg, 2012) to facilitate analysis through multiple lenses, prevent blind spots in data collection and to prevent duplication of existing theory (Thornberg, 2012; Thornberg & Charmaz, 2014). It has also been acknowledged elsewhere that qualitative researchers cannot be

entirely objective (Porter, 1993). Given that the researcher was cognisant of pre-existing health psychology theories, and a constructivist approach was being adopted, the steps described by Ahern (1999) were followed in order to engage in reflexive bracketing to mitigate the influence of biases and heuristics (Crotty, 1996).

This included keeping a reflexive journal throughout the research process. The researcher initially wrote a reflexive entry regarding their pre-conceived ideas about the application of health psychology theories to PwCF, attending particularly to the process of growing older, his personal experiences of encountering people living with chronic ill health conditions, and attitudes towards health more generally. This was important to recognise the researcher's epistemological influences and areas that could indicate a lack of neutrality. When constructing the interview schedule, writing entries in the reflexive journal facilitated reflection on the questions to determine whether they were too heavily informed by the researcher's pre-existing theoretical knowledge or heuristics, and perhaps whether they limited the opportunity to elicit more open-ended data. The researcher would also write a reflexive entry both after an interview and writing memos (discussed below), as these are key analytical tools where analytical connections between data begin to be constructed.

Reliability and Validity

In order to enhance the credibility and inter-rater reliability of the data analysis, i.e. to assess the 'fit' between the primary data and the researcher's representation of those data in the form of codes (Tobin & Begley, 2004), segments of data were coded by two independent researchers at each stage of coding (initial line by line, focused and theoretical – discussed below). Both researchers had experience of engaging in qualitative research and constructivist grounded theory approaches. Although codes are very idiosyncratic dependent upon the researcher (Charmaz, 2014), this facilitated discussion whereby the researchers were able to

reach consensus and mitigate against undue biases when coding the data (Jonson & Jehn, 2009; Elliot, 1999).

An additional credibility check involved the assessment of respondent validity, through 'member checking' (Birt, Scott, Cavers, Campbell & Walter, 2016). The researcher invited back all participants to comment on the findings, and two responded (18% response rate). The process involved the researcher presenting the outcome of the analysis to participants, and allowed them to respond as to whether they reflected their lived experiences. Participants involved reported a close resonance with the findings.

Procedure

The study received ethical approval by NHS Wales (REC 6; IRAS ID = 257144; Appendix H).

Recruitment of Participants

Initial eligibility criteria (Appendix I) required participants to be aged 40 years and over, and to have been diagnosed with CF during childhood. As above, this was to purposefully sample participants who will have lived through a period of significant medical advancement and experienced several prognostic-shifts (Patton, 2014). Following approval, the researcher collaborated with the CF Trust to run a pilot interview with a participant meeting the study criteria. This was to ensure the appropriateness of the interview schedule and the process surrounding the interview (e.g. length, need for breaks) by receiving feedback from the pilot participant. The participant observed the interview schedule and process were fit for purpose. Data from the pilot interview were included in the final analysis.

All subsequent participants meeting the initial eligibility criteria were identified by a clinical psychologist working in two regional CF centres based in Wales and England, UK. An 'opt-in' recruitment strategy was used, whereby a 'Participant

Information Sheet' (Appendix J) and 'Consent to Contact Form' (Appendix K) were posted to eligible participants in batches. A total of 38 were sent, and 14 responses received. This allowed the researcher to contact respective participants to discuss the study and ascertain their wish to participate. Of the 14 initial responses received, 11 were contacted, and 10 subsequently provided written informed consent, as one person opted not to take part (recruitment rate = 26%). Consent form in Appendix L.

Participants

Ten adults aged 40 years and older (M_{age} = 43.5 years; range = 40 – 47 years, 4 male, 6 female) participated in the study. In order to situate the sample (Elliot, 1999), metrics were collected on respective participant lung function (%), blood sugar (HbA1c), whether participants had received a lung transplant and the number of hospital admissions in the past year. These data can be seen in Table 1 below.

Table 1. Data captured in order to situate the sample. Gaps in the data are indicated using a hyphen. HbA1c of below 42 is considered 'healthy'.

Participant	Age	Sex	Lung Function (%)	Blood Sugar (HbA1c)	Lung Transplant (Yes/No)	Hospital Admissions (12 month period)
1	46	M	89	-	No	0
2	40	F	75	46	No	0
3	43	F	57	47	No	2
4	41	F	17	78	Yes	1
5	46	M	65.3	37	No	1
6	40	F	44.9	39	No	0
7	47	M	74.1	52	No	1
8	41	F	26.2	41	No	3
9	44	F	50	39	No	2
10	47	M	42	62	No	3

Interview Procedure

Five interviews were completed face-to-face (*n* = 2 at the participant's home; *n* = 3 at the respective CF unit – pre-pandemic) and a latter five via video call using Skype for Business (post-pandemic onset). Interviews ranged from 45 to 80 minutes, were audio recorded and subsequently transcribed verbatim. Nine interviews were completed in one sitting, and one took place over two sessions due to the participant's fatigue levels. Interviews were guided by the interview schedule which was updated as data were collected and analysed to theoretically sample data, as discussed below ('theoretical sampling').

Qualitative Data Analysis

Qualitative interview data were analysed by the procedures dictated by a constructivist grounded theory approach (Charmaz, 2014). NVivo 12.6 software was used for analysis.

Coding

During all stages of coding, a constant-comparative approach was used, whereby the researcher continuously re-visited and compared the primary data, codes and memos (described below) to one another (Charmaz, 2006; Glaser & Strauss, 1967). This process also helped to ensure that all stages of coding were appropriately grounded in the data.

As described by Charmaz (2014), the researcher engaged in initial, line-by-line coding of the interviews by scrutinising the interview data in small batches (see 'theoretical sampling' below) as they were completed. Line-by-line coding was utilised to remain grounded in the data, and to begin defining what was happening in the data (Charmaz, 2014). To facilitate the definition/detection of processes, the researcher coded predominantly using gerunds, as suggested by Glaser (1978).

To progress the data analysis, and begin making conceptual connections between the data, codes and memos, the researcher engaged in focused coding (Charmaz, 2014; Glaser, 1978). This form of coding facilitated the identification of similarities, connections and differences in the most salient initial line-by-line codes, and the subsequent synthesis of larger segments of data. To conceptualise the relationships between focused codes, the researcher engaged in theoretical coding. This final stage facilitated the formulation of the nascent theoretical model, underpinned by hypothesised interactions between focused codes (Glaser, 1978; Glaser, 2005; Charmaz, 2014). See Appendices M, N and O for illustrative examples of how line by line codes were raised to focused codes, and subsequently to theoretical codes.

Memo writing

The memo functionality within NVivo was used throughout all stages of data collection and analysis (post-interview, line by line coding, focused coding, theoretical coding) to capture the researcher's analytical ideas, facilitate a constant comparative approach and allow an additional layer of reflexivity (Charmaz, 2006). They were used to capture the researcher's thoughts about salient codes that kept re-emerging, provided a place to develop early (tentative) hypotheses about particular codes and how they related to one another, and to recognise gaps in the data (for example, where too much was being assumed). This latter point also allowed the researcher to engage in an additional measure of reflexive bracketing; that is, if too much was being assumed, then *what* was being assumed and *why*? Example memos can be seen in Appendices P and Q.

Theoretical Sampling - Interview Schedule Development

The original interview schedule consisted of eight stem questions (and an additional two closing questions), with each stem question having its own prompts (if needed). Interview questions aimed to explore how PwCF: had been impacted by medical advances; experienced and managed uncertainty; experienced and managed setbacks; set goals in the context of their terminal condition; managed their condition day-to-day; and constructed meaning about growing older. Intentional effort was also made to keep them open and accessible, particularly given that older PwCF may have had a disrupted education due to their health needs (Gathercole, 2019). To enhance the rigour in this area, the researcher bracketed their pre-conceived ideas (as above), and consulted with a group of experts by experience (adult PwCF) via the CF Trust, which led to amendments to the interview schedule (see Appendix R for the original and updated interview schedule, and Appendix S for an example feedback form used to inform the initial interview schedule development).

As described above, the researcher engaged in line-by-line and focused coding in small batches. Four interviews were completed initially (including the pilot interview), and then these were coded before engaging in further data collection. Early focused codes begun to emerge, such as '*maintaining low expectations'* and *'avoidance of uncertainty'*. During the memo writing process, a link between these was beginning to emerge, however it was still unclear how they interacted (e.g. the impact of uncertainty on maintaining expectations and/or goals). Up until this point, all participants talked about experiencing setbacks (e.g. a medical crisis, subjective well-being), but it was unclear at this stage of coding *how* participants responded to/recovered from the experience of setbacks.

In-line with a grounded theory approach, emerging gaps in the initial data (such as the above example) facilitated the development of the interview schedule, specifically the stem-question prompts (the stem questions remained unchanged) (Charmaz, 2014). This updated interview schedule (Appendix R) guided the data collection of a further four interviews. The same line-by-line and focused coding procedures were then followed as before. No new superordinate codes/themes emerged, and two further interviews (without amendments to the interview schedule) were completed and coded to ensure theoretical sufficiency (Dey, 1999).

RESULTS

This study sought to understand how PwCF have experienced growing older amongst prognostic shifts, and how they have processed and managed with these shifts. The iterative data collection and analysis process, informed by grounded theory (Charmaz, 2014), led to the emergence of a tentative model which captures the processes involved for older PwCF in adapting to new health/prognostic information (see Figure 1 below). These processes are lifelong and involve an on-going set of adjustment response(s) that ultimately lead to the development of a 'new normal'. The process of re-appraisal was underpinned by a transitory grief

response (to the loss of previous self) and this is proposed to be the catalyst for re-appraisal. Recognition of an (un)certain mortality (implied by living with a terminal, fluctuating condition) and low expectations (that have been internalised from a young age) permeate participant's narratives. The model proposes that a life-long process of adapting and re-adapting to a 'new normal' directly influences how participants responded to such uncertainty, and lived their day-to-day lives (e.g. goal setting and planning, and operationalising their expectations of 'normal').

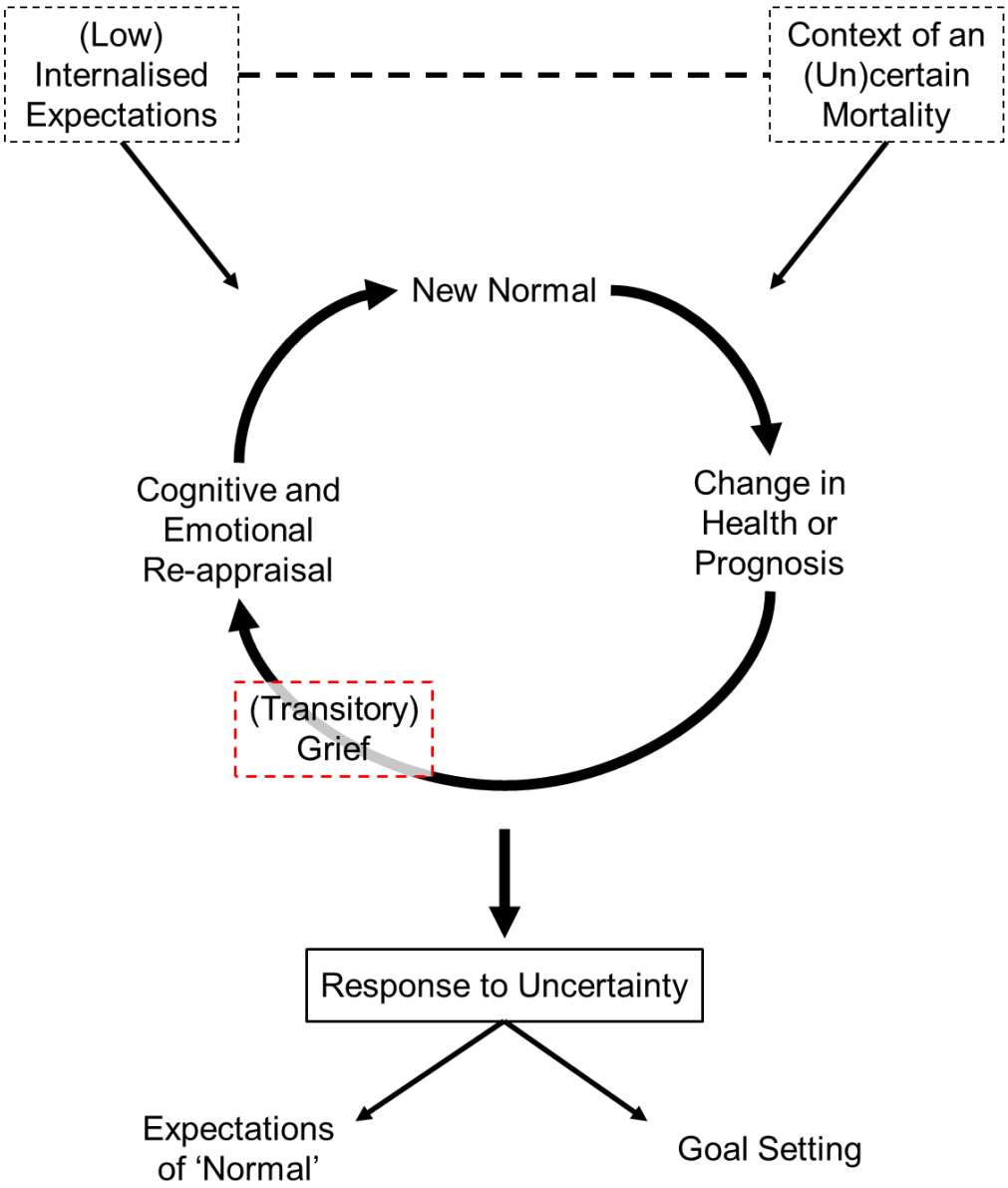

Figure 1. Tentative conceptual model developed from the analysis of qualitative interview data.

Living with an (Un)Certain Mortality

The construct of an (un)certain mortality has existential elements which older PwCF share with the oldest old (Agren, 1998; Gasiorek, Fowler & Giles, 2019; Nilssin, Sarvimaki & Ekman, 2003). However, there are many other uncertainties associated with living with CF, illustrated within other aspects of the model. The lives of PwCF are in the context of a certain mortality, as can be said for everyone. However, for PwCF, this certain mortality can be very uncertain and sudden, and can feel like *"you've always got that dark cloud hanging over you" (Participant 3).* The (un)certain nature of mortality for PwCF is articulated in the following quote:

> *"Um, CF is, you know, the uncertainty of CF is I expect to die before my time. I expect it to be a slow-, a slow debilitating process of being well, not as well, ill, ill, ill... My, my main hope in life is that when I'm, say, 62, I'll be killed by a 'Mind that bus', 'What bus?' Splat. Because that is far more easy and better to deal with than the complete what is expected of the, the-, you know, this shallow curve and then maybe a slight step off a cliff, um, of my CF death." Participant 5*

Several participants could vividly recollect learning about their own mortality, mostly during their childhood or adolescent years, although one participant reported only learning about their limited life expectancy in their early twenties. The same participant, however, went on to reflect about how they had "buried" their awareness of their limited life expectancy, and recounted learning about it in primary school. Many participants also reflected on how, throughout their lives, they have been living on the edge of mortality, "*I always seemed to be as they kind of pushed it, the age, up, I seemed to be matching it."* (Participant 7). For the generation of participants interviewed, this pattern continued from their childhood years, all the way through their lives and appeared to be imminent according to the statistics at the time.

Managing 'Existential' Uncertainty

Participants disclosed responding to the existential uncertainty by avoiding thinking about it. For some, that was an active avoidance of thinking about it altogether, some conceptualised it as 'forgetting' or 'displacement', and for others it was a process of making decisions in life to minimise having to think about it (e.g. not having a pension, life insurance or savings). For example, some would respond by acknowledging the certainty of an early death, and state that *"there is no dealing with it"*. One participant reflected on how they consciously try to forget about their (un)certain mortality and some of the ways in which this continues to permeate into their day-to-day life:

> *"Just accept it and-, yeah, been told I deal with it well, but I think it's just that-, but then I get upset about, you know, a little thing, you know, like the house is messy or-,*
>
> ***Yeah.***
>
> *Something silly, really. And I think that's probably displacement or something.*
>
> ***Yeah. Yeah.***
>
> *I can't really think about what, what's happening most of the time. I just try and forget about it and focus on something else. Yeah." Participant 8*

As articulated by one of the participants, the presence of an '(un)certain mortality' can be considered a super-ordinate theme, like a *"dark cloud hanging over you"*. This metaphor fits with the emerging model, with the concept of an (un)certain mortality manifesting in many ways and setting the context for all other aspects of the model.

Maintaining Low Expectations

In addition to the context of an (un)certain mortality, another strong contextual factor in the model is that of internalised low expectations, with participants reflecting on how this permeates their sense of self. For example, one participant identified as a *"pessimist defensive",* and kept their bar (of expectations) extremely low to the point where they *"expect to be tripped up by that low bar*" (Participant 5). Some reflected on the interaction with their low expectations and new medical treatments, "*I don't instantly think it's going to be a game changer… I've got quite a neutral expectation*" (Participant 7). Others reflected on their perspective on life more broadly.

> *"I've got a different viewpoint, definitely, than lots of other people, non-CF people. I'm sure that's true with all, all people with long-term illnesses-,*
>
> ***Yeah.***
>
> *Have got quite a different viewpoint on life. I generally don't expect things to happen, but then I'm very pleasantly surprised when nice things do happen.*
>
> ***Okay.***
>
> *I just kind of roll with it a lot more than a lot of other people. A lot of other people worry about what's going to happen, whereas I'm just like, 'It'll happen or it won't,' and just kind of-, I'm a lot more steady, I think, generally."*
>
> *Participant 6*

Several participants expressed how they believed this was almost a habitual process that was internalised from a young age, influenced by the pessimistic medical prognoses. For many, the medical advice for parents was that "*oh she won't live that long*" and simply to "*take me home and enjoy me"* (Participant 3). Thus, these low expectations were initially set for the parents of PwCF whilst the

participants were young and appear to have been inherited and internalised by older PwCF.

An Ever-changing Normality

In addition to the notion of an (un)certain mortality, PwCF live with many day-to-day uncertainties in terms of their ever-changing health and prognostic status. This model, which emerged from the interview data, suggests that there is a 'live' dynamic process that is always on-going, whereby PwCF are constantly monitoring changes in their health, acknowledging the impact of this on their subjective well-being, going through a process of adaptation and grieving for what they have lost, and eventually establishing a new normal. The model proposes that the process of adaptation is pervasive, whilst the experience of a grief response is more transitory and is the catalyst for adaptation. One participant used the metaphor whereby each cycle of this process represents an additional 'dent', and over time, repeated cycles can result in an accumulation of dents that take longer to 'pop out' and return to 'normal':

> *"...it's just every little knock gets you a little bit… dents you a little bit*
>
> ***And do those dents ever pop back out?***
>
> *Yeah… it just takes longer each time I think… you know for instance that one I didn't see it coming so it was even harder to cope with" Participant 4*

As will be considered later, this cyclical process of establishing a 'new normal' is proposed to have a direct influence on how PwCF respond to uncertainty and live their day-to-day lives.

Experiencing a Change in Health or Prognosis

The model postulates that the process of adapting to a 'new normal' commences when someone experiences a change in their health or receives new prognostic information. In the context of an (un)certain mortality, and attempted avoidance of this unfortunate reality, experiencing a change in health later in life for PwCF was often reported as coming as a shock. Participants described how a sudden, uncertain change in health made it impossible for them to avoid thinking about their (un)certain mortality. Many wondered whether the current health decline could ultimately result in their mortality, questioning "*was this going to be the end of my life"* (Participant 4). Whilst for some, the sudden uncertain change hit them *"like a tonne of bricks"* (Participant 7), others experienced familiar smaller fluctuations, describing that they "*pick up colds so easy"* and that they "*can go from one to another to another and it's restrictive" (Participant 2)*. The change in physical health status or prognostic information was not the only important factor, but the interactional process of these and the subsequent impact upon their quality of life.

> *"Last year I went from being really well to being really ill in about, like, three weeks or something and that was a real shock, because I went from, like, literally running for, you know, about an hour or something, to being really out of breath on the flat and being really, really ill and it was-, that was a really big shock, because I didn't really-, because all the other illnesses I've had before have been quite a slow decline really, probably-,*
>
> ***Okay.***
>
> *Um, so that was quite a mental blow again, because it was like, 'Oh, actually you might go from being-, having a great life to suddenly that's it, this is the new normal.'" Participant 6*

With an increased life expectancy, participants acknowledged additional consequences of growing older for PwCF such as other age-related health conditions. But, given that their life expectancy was uncertain, the relative risk and consequences of acquiring such conditions was minimised. PwCF appeared not to have given much, if any thought to the potential co-morbidities of growing older, such as developing cancer. Minimisation strategies applied to their inevitable CF-related health decline were also extended to other chronic health conditions of old age.

> *"so who would imagine I've got cystic fibrosis… I've had a liver transplant… heart and lung transplant… kidney transplant… diabetes… all the complications of transplantation and I always knew the worries and the complications to come with all of that and then suddenly you're faced with breast cancer and you're like 'where did that come from?!' it was like as if it was a steam train had just hit me from the side… erm… and it just… it bowed me over and I found it really difficult to cope with" Participant 4*

(Transitory) Grief

The model postulates that a transient grieving process is necessary to drive the process of re-appraisal, ultimately leading to the establishment of a new normal. The cognitive and emotional processes which allow adjustment to loss are as pervasive and diverse as grief processes (low mood, anger, regret, guilt, 'depression' – Participant 3) (e.g. the Dual Process Model; Stroebe & Schut, 2010). PwCF acknowledged that the concept of death is always present (as exemplified in the '(un)certain mortality' theme above), yet despite this appeared to experience grief for the present loss of past self/identity/health, rather than experience an anticipatory grief for inevitable future losses (as with lots of other chronic health conditions).

"I kind of have to have like a little bit of a 'grieving period' [laughs] I call it… and then time for… to…kind of you have to accept it and then proves it and move on… didn't even think of that when I was 17… you just did it… I just did it…" Participant 4

The degree to which a person's health is affected, or the degree to which a change in health brings about psychological distress, appeared to be idiosyncratic and unique for everyone. One participant reflected upon how the biggest part of the process for them was time to allow them to "*adjust to whatever new norm that is"* and *"adjust your expectations"* (Participant 6). Another participant described this same process, where they became *"really depressed",* and how they were able to positively reframe their circumstances and think "*it could be worse I could be dead... at least I've got a future to plan at the moment"* (Participant 3).

Re-appraising their changed health was considered a pivotal point in the process of re-adjusting. A shift from the psychological experience of distress (e.g. grief, low mood, anger), where motivation is possibly low, to a mindset of acceptance over their present circumstances (e.g. loss of health, function and/or wellbeing). This process suggests that PwCF may balance oscillations between the avoidance of, and sitting with, the psychological distress (e.g. grief) by integrating their past self with their current self whose opportunities are limited by their ill health.

"It's just all about looking after yourself and realising your potential… what you can do and what you can't do… you've got to realise that from a young age… you can't do what other people do… sometimes not all the time… and you gotta take a step back and think oh well I can't do that unfortunately" Participant 3

Establishing a 'New Normal'

Acceptance of the new normal enables PwCF to engage in the necessary measures to keep their CF under control. For example, one participant articulated how they had to accept a (temporary) new normal to manage a period of being unwell by receiving regular intravenous antibiotics, in order to prevent their health deteriorating further and prevent the need for hospital admission. In addition to maintaining their physical health, the adjustment process of grieving and acceptance enables PwCF to adopt a more psychologically healthy attitude, and by embracing their circumstances they can establish meaningful roles grounded by their values.

> *"Oh like my arts and crafts I suppose I get involved in that… and that focuses my mind on something else then… and like when I'm in hospital… cause I was in for three weeks I brought them with me… so instead of sitting in my room and watching TV and feeling bored and then the boredom gets to you which is the worst… do something that you enjoy even if it means bringing in three extra bags of beads and glitter and glues and… but then I was glad I did it because erm… I made fairy dolls out of silk and pipe cleaners and stuff and then I made little crystal and beaded angel charms so the staff all had one each cause I was making them for them… so when I really look back on it that was really my strategy when I was in hospital of coping… " Participant 3*

Following a process of grieving for the old, and accepting a new (permanent or temporary) normal regarding their health status, and the associated loss that incurs, a strong narrative of a 'fight' with CF emerged, *"it's like CF punches you one… and you're like 'right I'm gonna get back up now'"* (Participant 4). Using the previous metaphor offered by a participant, this process can be likened to popping out the 'dents' that can be incurred from an uncertain change in health. Often, this

process of engaging in a ‘fight’ with CF involves the appraisal of external influences. For example, learning about innovative medical approaches to transplant from the TV, establishing a good working relationship with a doctor/surgeon, or simply having good social support from family or a partner.

> *“I’m quite feisty and I suspect that’s because-, I always say to [husband] I’m a bit of a fighter and I’m sure that’s obviously come from CF, because, you know, if I fall over I’m going to get my-, get up and dust myself off. I might not do it well, but I’ll keep getting up.” Participant 6*

Others describe this same process as taking control of their CF, for example by establishing a sense of normal through routine and engagement with treatments and other pro-health behaviours (eating healthy, exercising, etc.), leading to the feeling that *“I have some control over it”* (Participant 8). What was apparent is that even when establishing a ‘new normal’, and keeping up that fight with CF whilst also living a life beyond managing CF, PwCF remain vigilant, for example having the thought that *“something is going to go seriously wrong”* (Participant 5).

Engaging in this fight with CF, whilst remaining vigilant for health crises, proved difficult for some PwCF. Some participants acknowledged the difficulty of maintaining a sense of ‘normal’ and finding meaningful roles, always questioning *“am I going to be well next week?”* (Participant 5). Whereas others, who had experienced less uncertainty and had relatively healthy lung function, were able to attain a sense of normality, e.g. *“I just view it as a cold or the flu”* (Participant 2) with more confidence. This suggested, given the progressive nature of CF, their sense of normality and confidence could be challenged were their health to begin to significantly deteriorate. Embracing a ‘new normal’, for some, included maintaining and/or re-establishing some of their ‘old normal’ and previous roles/routines, such as associating going back to work with *“back to normality”* (Participant 4).

Response to Uncertainty

This aspect of the model is intended to reflect the different responses that PwCF demonstrate, which is hypothesised to be influenced by the significant uncertainties that they face (as illustrated above).

Setting Goals

In the context of internalising and subsequently maintaining low expectations, amongst an uncertain and constant re-appraisal cycle, the participants also reflected upon their process of goal setting. Some considered how they set small goals for themselves, so that they are achievable despite the uncertainty, "*I probably limit the goals that I make, so that I can achieve them*" (Participant 6). Whilst others disclosed that they had not set themselves any goals at all, *"I don't make plans"* (Participant 3). It is possible that this is the approach taken to avoid disappointment or avoid not managing to achieve goals due to the uncertain nature of living with CF.

> *"You try and tell a 12-year-old boy to set life goals when he fully expects to die […] so I've always had not people telling me you're going to die, but somehow I was always aware of what is probably expected." Participant 5*

The day-to-day impact for some was being unable to maintain pre-determined plans, with one participant sharing how they reframed plans to catch-up with a friend (which had to be cancelled three times due to their health taking an unexpected decline), realising that *"we haven't seen each other for a year"*, and managing that uncertainty *"with my psychology of I don't expect much, it's not really that uncertain"* (Participant 5).

The participants recognised that the uncertainty associated with living with CF can be overwhelming, and so they tended to explicitly adopt a lifestyle choice of *"taking one day as it comes"* (Participant 3) and *"trying to keep putting one foot in front of the other"* (Participant 6). This is one way in which the participants could avoid/not attend to the significant levels of uncertainty. The participants reasoned that if they did not live this way, then they may 'dwell' on the uncertainty, for example their own mortality, how their ill health interferes with planning or goal setting and precludes engagement with roles defined as those of a 'normal person'.

> *"Take one day as it comes… never… I don't look too far into the future now cause it… like I said you don't know what is round the corner… in a normal person anyway… nobody knows what's gonna happen one day to the next… but when you've got a serious illness… if it's CF cancer… any serious illness… you don't know what's gonna happen so you just take one day at a time… today now I feel great… tomorrow I could be down in the dumps crying all day long… so it's just… each day as it comes…" Participant 3*

Expectations of 'Normal'

In the context of an always-changing sense of 'normal', and a constant process of re-adjusting their concepts of 'normal', some PwCF appear to manage this by establishing roles that are perceived as normative within their particular societal context. For some this related to educational experiences such as going to university, volunteering within their community, living independently, parenting or holding down employment.

> *"I know that more people are working with CF… and it's not all about the working but it's achieving normal things I think… you know things that people*

like you or my husband are able to do… you know why can't we do those things… why can't we go to university and live alone…" Participant 4

For others who were too unwell to fulfil a perceived normative role within their societal context, there were negative consequences for their psychological wellbeing, *"I thought I was unemployable really. Err, that had a massive effect on me"* (Participant 8). It appears therefore that a negative interaction can occur between the internalised low expectations for this generation of ageing PwCF and the fulfilment of normative roles within society.

DISCUSSION

This study aimed to explore both the experience of PwCF when growing older alongside medical advancements and prognostic-shifts, and to understand how these shifts have been processed and managed from a psychological perspective. The researcher utilised a grounded theory approach (Charmaz, 2014), which involved the iterative development of a theoretical model from the simultaneous collection and analysis of semi-structured interview data.

Summarising the Model

The emergent model illustrates an integrated set of processes that allow older PwCF to respond to health insults and/or prognostic shifts, both of which carry significant uncertainty. The model postulates that older PwCF live with a dynamic sense of 'normal', which is constantly evolving in response to external factors such as health insults or new prognostic information (e.g. new scientific findings, longer life expectancy statistics, availability of new treatments). These encounters are hypothesised to precipitate a transitory grief response, which is the catalyst that drives PwCF to adjust and re-appraise their new circumstances, ultimately developing a 'new normal'.

There are two influential factors that affect the cycle of re-appraisal, which provide insight into the unique circumstances for older PwCF. Whereas the re-appraisal cycle considers the *intra*-personal processes, the internalisation of low expectations and living in the context of an (un)certain mortality are considered more *inter*-personal processes. They are deemed inter-personal as they are socially constructed understandings that older PwCF have internalised, based on their and their parent/care-giver's interactions with members of society (e.g. each other, teachers, medical professionals, the media). Thus, this is the lens through which older PwCF perceive the world and where they position themselves in the world, and set the context for the re-appraisal cycle.

The model also illustrates how older PwCF may respond to living in the context of such uncertainty. For many, this involved strategies to minimise the uncertainty by living in the current moment, "*taking one day as it comes"* and *"keep putting one foot in front of the other",* which appears to be an approach that participants have intuitively adopted throughout their lives. It is hypothesised that some PwCF, where this approach does not come naturally, may experience higher levels of psychological distress. Although some participants talked about aspirations they had (e.g. to travel to certain holiday destinations), the majority of participants indicated that they do not set or commit themselves to concrete goals due to the uncertainty of their condition (e.g. booking last minute holidays).

The other way participants responded to uncertainty ('Expectations of 'Normal') was in their pursuit of fulfilling roles, many of which were perceived as normative within their societal context. There was a strong desire to achieve 'normal' things, whether that was maintaining a job, having children, or simply being well enough to buy some milk from the local shop. Understandably, there was diversity in the unique construction of what 'normal' was for this cohort (as would be expected for individuals without CF). The cohort of PwCF that were interviewed were born at least 40 years ago, where the inter-personal processes described

above were hypothesised to have led people to internalise a low set of expectations for themselves. This represents a point in time, or a cross-sectional expectation of 'normal' for older PwCF, born into that time. However, living in the current day, society is likely to have changed its perceptions of CF (e.g. longer life expectancies, 'scientific age', availability of new drugs such as triple combination therapy). The older PwCF living today are the exemplars for society of what is 'normal' for PwCF, yet society's expectation is mismatched with the reality of *current* older age for PwCF. This will have implications for clinicians when interacting with PwCF, and will require them to be reflexive about their assumptions and expectations relating to this cohort.

Fit with Other Models

One aspect of the current model that aligns with pre-existing models is the construction of a 'new normal', which is comparable to the 'illness representation' construct within Leventhal and colleagues' (2005) self-regulation model of illness. For example, the development of the identity aspect of Leventhal et al.'s illness representation construct is conceptually similar to the 'new normal' construct in this model, with both models highlighting their dynamic nature. The two models diverge when appreciating the unique contextual factors that influence older PwCF's development of a 'new normal', particularly the interpersonal constructs ('Internalised Low Expectations' and the 'Context of an (Un)certain Mortality'). Although there are other health conditions where there is overlap of such factors (e.g. HIV), there are nuanced differences for older PwCF that are unaccounted for by current theoretical models (e.g. the multiple layers of uncertainty, the internalisation of beliefs about the self and mortality).

An additional area of overlap with existing theory is the 'Cognitive and Emotional Re-Appraisal' element. An existing theory that offers insight into re-appraisal and adjustment processes is Snyder and colleagues' hope theory (Hill et

al., 2018; Snyder et al., 1991; Synder, 2002). Their theory concerns acquired rather than congenital conditions, and explains how people 're-goal' following a health insult/prognostic shift. Despite their different applications, the models overlap when concerning the cognitive and emotional elements of the re-appraisal process. Hope theory has proposed that it is necessary for individuals to experience some level of negative affect to precipitate consideration of new goals (e.g. shifting from life-curing treatments towards palliative care). Although on a superficial level, 'negative affect' could be synonymous with '(Transitory) Grief', Hill and colleagues do not deconstruct this element of their model.

It is possible to elaborate on these areas of overlap by applying Stroebe and Schut's (2010) dual process model, which has conceptual similarities with the '(Transitory) Grief' element in the current model, and may also allow insight into the 'negative affect' process in hope theory. One of the defining elements of theoretical models of grief is the attachment/separation distress in response to a loss or change (e.g. Parkes, 2006). In application to this model, this is hypothesised to be distress associated with the consequences of a loss of identity; that is, who PwCF saw themselves as before and after a health/prognostic shift. The Dual Process Model purports that people process their grief through oscillating between their experience of grief ('loss-orientated') and readjustment ('restoration-orientated'). This is similar to the current model, which proposes that people process their (transient) grief response through oscillations of the psychological distress (avoiding or sitting with distress) and re-appraisal of their coping skills and resources (e.g. social support, healthcare professionals). This model postulates that grieving enables people to re-appraise their circumstances, and therefore if they are unable to 'sit with' their grief, the model would propose that this would interfere with the re-appraisal process.

Critiquing the Model

Glaser's (1978) criteria of fit, work, relevance and modifiability have been used broadly in order to consider how grounded theories frame their data and to critique the quality of grounded theories (Charmaz, 2014; Lomborg & Kirkevold, 2003). The 'fit' criterion has been considered more elusive, as the determination of this depends upon the epistemological and ontological positions taken (Lomborg et al., 2003). For example, this study is ontologically underpinned by a social constructivist approach, and thus it could be argued that assessment of the 'fit' criterion is invalid, with an assumption that the data and subsequently the model is constructed between the researcher and the participants, based on the participants' own social constructions. As discussed earlier, this required a great degree of reflexivity on the part of the researcher to prevent the skewing of any aspect of the research processes based on his own epistemological heuristics (e.g. Ahern, 1999).

The researcher argues that the model fulfils Glaser's additional criteria of work, relevance and modifiability. The iterative development of a tentative theoretical model provides the basis for understanding the psychological phenomenon in question, and subsequently providing opportunity for clinical application. It is important to acknowledge that the developed model applies only to the cohort that were interviewed as part of this study (PwCF aged 40 years and over, who were diagnosed with CF during childhood), given the influence of the context-specific interpersonal processes. Although these context-specific inter-personal processes may only relate to this specific cohort, it is possible that the model could be modified and future iterations could consider the broader applicability of these (e.g. for younger generations of PwCF living in an era with the availability of triple combination therapy) (Glaser & Strauss, 1967). The model also presents opportunities for further research, discussed below.

Limitations

The researcher implemented steps discussed by Elliot et al. (1999) to enhance rigour (as discussed above). Although measures were used to enhance rigour and reduce threats to internal validity, such as having independent researchers code segments of data, rigour could have been further improved by having entire transcripts coded by multiple researchers. To mitigate against this, the researcher engaged in frequent in-depth discussions with the independent researchers concerning matters related to coding, and maintained a high level of reflexivity (e.g. by engaging in a reflexive diary (Ahern et al., 1999), as above). Measures were also implemented to enhance external validity, for example by presenting research findings to participants and gaining their feedback regarding how the model resonated with their lived experiences. Additionally, independent experts in the field of CF clinical psychology were consulted with regarding the development and subsequent interpretation of the model.

By their very nature, qualitative methodologies do not aim to produce generalisable findings (Charmaz, 2014). However, the iterative development of a conceptual model as part of a constructivist grounded theory approach can help to enhance the generalisability of the findings (Charmaz, 2014). Of course, the model is based upon interviews with just 10 participants from two UK nations, and although it provides hypotheses for further exploration and testing (discussed below), it still needs to be interpreted cautiously. Due to the availability of participants for such a study, and the ethical issues involved with interviewing participants that are too unhealthy, only one participant had undergone a lung transplant. Other health metrics of the included participants in terms of their blood sugar levels and lung function were more heterogeneous, enabling a greater diversity of perspective. It is possible that the model would have demonstrated nuanced differences if more people were to have experienced a lung transplant. However, the researcher hypothesises that the same cycle could have ensued, and the experience of an

organ transplant would have simply provided a more apparent exemplar of the cycle.

Clinical Utility

The developed model could be applied as a tentative theoretical foundation for which to base a psychological formulation of the person's 'presenting problem' (DCP, 2011; Johnstone & Dallos, 2013). PwCF could theoretically become 'stuck' at any stage of this cycle, and depending on the nature of the clinical formulation, clinical interventions could target some (or all) of the processes within the model.

For example, a person's avoidance of the grief associated with the loss of previous self (the consequence of a health deterioration), could be preventing their readjustment to a 'new normal'. Thus, a clinical intervention could facilitate the processing of their experience of grief, to support the person to accept their incurred loss of self and re-adjust (and re-frame) their current circumstances. Alternatively, if someone was hypothetically to become stuck in a grief response that was preventing them from accepting, re-appraising and adjusting in the context of a 'new normal', then a clinical intervention could focus on facilitating the person with the acceptance and re-appraisal processes within the model, for example by using a values-based approach (e.g. Acceptance and Commitment Therapy (Brassington et al., 2016)). Such an approach could also be facilitative at supporting PwCF whom do not intuitively manage to develop adaptive responses to uncertainty, such as living in the moment or achieving fulfilling 'normal' roles, and instead experience a high degree of distress relating to the uncertainty they face.

Further Research

Given that grounded theory is a hypothesis generating methodology, there are multiple avenues for future research (Charmaz, 2014). The model makes several predictions about the mechanism for appraisal and re-appraisal having

emotional and cognitive interacting elements. Further research would seek to examine the processes more specifically to gather evidence to reject the null hypothesis that re-appraisal does not occur. Following this, research would need to identify clinical interventions which might be suited to modifying the mechanisms and promoting healthier processes (such as those suggested above to facilitate emotional regulation, problem solving, acceptance, committed action), and examine their effectiveness. It would be a plausible hypothesis that the COM-B (influences of capability, opportunity, and motivation on behaviour) model borne out of the Theoretical Domains Framework (Michie et al., 2011) could also underpin such interventions, to facilitate adaptation to the cumulative 'dents' that older PwCF experience as they age.

This model applies specifically to older PwCF, given their unique set of context-specific circumstances. However, it is possible that these context-specific constructs may still be relevant to other generations of PwCF. Future research could consider the applicability of this re-appraisal cycle for younger generations of PwCF in a context that may, for example, involve less uncertainty.

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Appendix A. ***Author guidelines for submission to the British Journal of Health Psychology.***

Author Guidelines

The aim of the British Journal of Health Psychology is to provide a forum for high quality research relating to health and illness. The scope of the journal includes all areas of health psychology as outlined in the Journal **Overview**.

The types of paper invited are:

• papers reporting original empirical investigations, using either quantitative or qualitative methods, including reports of interventions in clinical and non-clinical populations;

• theoretical papers which report analyses on established theories in health psychology;

• we particularly welcome review papers, which should aim to provide systematic overviews, evaluations and interpretations of research in a given field of health psychology; and

• methodological papers dealing with methodological issues of particular relevance to health psychology.

Authors who are interested in submitting papers that do not fit into these categories are advised to contact the editors who would be very happy to discuss the potential submission.

All papers published in The British Journal of Health Psychology are eligible for Panel A: Psychology, Psychiatry and Neuroscience in the Research Excellence Framework (REF).

1. Circulation

The circulation of the Journal is worldwide. Papers are invited and encouraged from authors throughout the world.

2. Length

Papers describing quantitative research (including reviews with quantitative analyses) should be no more than 5000 words (excluding the abstract, reference list, tables and figures). Papers describing qualitative research (including reviews with qualitative analyses) should be no more than 6000 words (including quotes, whether in the text or in tables, but excluding the abstract, tables, figures and references). The Editors retain discretion to publish papers beyond this length in cases where the clear and concise expression of the scientific content requires greater length.

3. Editorial policy

The Journal receives a large volume of papers to review each year, and in order to make the process as efficient as possible for authors and editors alike, all papers are initially examined by the Editors to ascertain whether the article is suitable for full peer review. In order to qualify for full review, papers must meet the following criteria:

• the content of the paper falls within the scope of the Journal

• the methods and/or sample size are appropriate for the questions being addressed

• research with student populations is appropriately justified

• the word count is within the stated limit for the Journal (i.e. 5000 words, or 6,000 words for qualitative papers)

4. Submission and reviewing

All manuscripts must be submitted via **Editorial Manager**. The Journal operates a policy of anonymous (double blind) peer review. We also operate a triage process in which submissions that are out of scope or otherwise inappropriate will be rejected by the editors without external peer review to avoid unnecessary delays. Before submitting, please read the **terms and conditions of submission** and the **declaration of competing interests**. You may also like to use the **Submission Checklist** to help your prepare your paper.

5. Manuscript requirements

• Contributions must be typed in double spacing with wide margins. All sheets must be numbered.

• Manuscripts should be preceded by a title page which includes a full list of authors and their affiliations, as well as the corresponding author's contact details. You may like to use **this** template. When entering the author names into Editorial Manager, the corresponding author will be asked to provide a CRediT contributor role to classify the role that each author played in creating the manuscript. Please see the **Project CRediT** website for a list of roles.

• For articles containing original scientific research, a structured abstract of up to 250 words should be included with the headings: Objectives, Design, Methods, Results, Conclusions. Review articles should use these headings: Purpose, Methods, Results, Conclusions. As the abstract is often the most widely visible part of your paper, it is important that it conveys succinctly all the most important features of your study. You can save words by writing short, direct sentences. Helpful hints about writing the conclusions to abstracts can be found **here**.

• Statement of Contribution: All authors are required to provide a clear summary of ‘what is already known on this subject?’ and ‘what does this study add?’. Authors should identify existing research knowledge relating to the specific research question and give a summary of the new knowledge added by your study. Under each of these headings, please provide 2-3 (maximum) clear outcome statements (not process statements of what the paper does); the statements for 'what does this study add?' should be presented as bullet points of no more than 100 characters each. The Statement of Contribution should be a separate file.

• Conflict of interest statement: We are now including a brief conflict of interest statement at the end of each accepted manuscript. You will be asked to provide information to generate this statement during the submission process.

• The main document must be anonymous. Please do not mention the authors’ names or affiliations (including in the Method section) and always refer to any previous work in the third person.

• Tables should be typed in double spacing, each on a separate page with a self-explanatory title. Tables should be comprehensible without reference to the text. They should be placed at the end of the manuscript but they must be mentioned in the text.

• Figures can be included at the end of the document or attached as separate files, carefully labelled in initial capital/lower case lettering with symbols in a form consistent with text use. Unnecessary background patterns, lines and shading should be avoided. Captions should be listed on a separate sheet. The resolution of digital images must be at least 300 dpi. All figures must be mentioned in the text.

• For reference citations, please use APA style. Particular care should be taken to ensure that references are accurate and complete. Give all journal titles in full and provide doi numbers where possible for journal articles. For example:

Author, A., Author, B., & Author, C. (1995). *Title of book.* City, Country: Publisher.
Author, A. (2013). Title of journal article. *Name of journal, 1,* 1-16. doi: 10.1111/bjep.12031

• SI units must be used for all measurements, rounded off to practical values if appropriate, with the imperial equivalent in parentheses.

• In normal circumstances, effect size should be incorporated.

• Authors are requested to avoid the use of sexist language.

• Authors are responsible for acquiring written permission to publish lengthy quotations, illustrations, etc. for which they do not own copyright. For guidelines on editorial style, please consult the **APA Publication Manual** published by the American Psychological Association.

• Manuscripts describing clinical trials are encouraged to submit in accordance with the **CONSORT statement** on reporting randomised controlled trials.

• Manuscripts reporting systematic reviews and meta-analyses are encouraged to submit in accordance with the **PRISMA statement**.

• Manuscripts reporting interventions are encouraged to describe them in accordance with the **TIDieR checklist**.

If you need more information about submitting your manuscript for publication, please email Hannah Wakley, Managing Editor (**bjhp@wiley.com**) or phone +44 (0) 116 252 9504.

6. Supporting information

We strongly encourage submission of protocol papers or trial registration documents, where these are in the public domain, to allow reviewers to assess deviations from these protocols. This will result in reviewers being unblinded to author identity.

Supporting Information can be a useful way for an author to include important but ancillary information with the online version of an article. Examples of Supporting Information include appendices, additional tables, data sets, figures, movie files, audio clips, and other related nonessential multimedia files. Supporting Information should be cited within the article text, and a descriptive legend should be included. Please indicate clearly on submission which material is for online only publication. It is published as supplied by the author, and a proof is not made available prior to publication; for these reasons, authors should provide any Supporting Information in the desired final format.

For further information on recommended file types and requirements for submission, please visit the **Supporting Information page** on Author Services.

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10. Colour illustrations

Colour illustrations can be accepted for publication online. These would be reproduced in greyscale in the print version. If authors would like these figures to be reproduced in colour in print at their expense they should request this by completing a **Colour Work Agreement** form upon acceptance of the paper.

11. Pre-submission English-language editing

Authors for whom English is a second language may choose to have their manuscript professionally edited before submission to improve the English. A list of independent suppliers of editing services can be found in **Author Services**. All services are paid for and arranged by the author, and use of one of these services does not guarantee acceptance or preference for publication.

12. The Later Stages

The corresponding author will receive an email alert containing a link to a web site. The proof can be downloaded as a PDF (portable document format) file from this site. Acrobat Reader will be required in order to read this file. This software can be downloaded (free of charge) from **Adobe's web site**. This will enable the file to be opened, read on screen and annotated direct in the PDF. Corrections can also be supplied by hard copy if preferred. Further instructions will be sent with the proof. Excessive changes made by the author in the proofs, excluding typesetting errors, will be charged separately.

13. Early View

British Journal of Health Psychology is covered by the Early View service on Wiley Online Library. Early View articles are complete full-text articles published online in advance of their publication in a printed issue. Articles are therefore available as soon as they are ready, rather than having to wait for the next scheduled print issue. Early View articles are complete and final. They have been fully reviewed, revised and edited for publication, and the authors' final corrections have been incorporated. Because they are in final form, no changes can be made after online publication. The nature of Early View articles means that they do not yet have volume, issue or page numbers, so they cannot be cited in the traditional way. They are cited using their Digital Object Identifier (DOI) with no volume and issue or pagination information. Eg Jones, A.B. (2010). Human rights Issues. *Journal of Human Rights.* Advance online publication. doi:10.1111/j.1467-9299.2010.00300.x

Appendix B. ***Eligibility criteria used for screening of the retrieved papers for meta-ethnography.***

Inclusion	Exclusion
Studies considering: • Factors influencing engagement in ('adherence' to) health-management regimes • Qualitative investigations of direct patient experiences (not parents/carers/healthcare professionals) • Adults (>=18 years)	• Children (<18) and participants identified as 'adolescents' • Studies specifically examining transitional period (this period already reviewed, finite period to independence/agency) • Studies specifically examining cystic fibrosis related diabetes (CFRD) • Studies specifically examining acute exacerbations (e.g. pulmonary). • Studies specifically examining organ transplantation (e.g. lung, kidney). • Studies not based on empirical findings • Studies that do not provide full explanation of methods, analysis or findings • Dissertations • Non-English language publications

Appendix C. ***Description of how Noblit and Hare's (1988) seven stage meta-ethnographic approach was followed.***

Noblit and Hare's (1988) Seven Step Meta-ethnographic Approach	Description of the Researcher's Application of Each Step
1. Getting started	A previous review (Macdonad et al., 2016) included a heterogeneous sample examining experiences of patients in adhering to cystic fibrosis treatment regimes. Since this publication, there have been numerous publications of qualitative studies investigating this phenomenon from more of a psychological perspective. Given more readily available papers, and the recent introduction and availability of triple combination therapy (and a consequential impact on treatment regimes for people prescribed this treatment), it was prudent to re-visit this area. A meta-ethnographic approach benefits from being more homogeneous in its nature, in order to mitigate the risk of making over-generalisations and reducing the meanings of first and second-order constructs (Noblit & Hare, 1988). Given this, the scope was narrowed from that of Macdonald's review, and excluded adolescent samples whom were experiencing a transitional period from parental/care-giver support to independence, and potential additional complications of cystic fibrosis (acute exacerbations, organ transplantation, cystic fibrosis related diabetes (CFRD)).
2. Deciding what is relevant to the initial interest	The search strategy was developed using the STARLITE pneumonic (Booth, 2006), and resulted in a refined inclusion and exclusion criteria (Appendix B) compared to Macdonald et al. (2016). Search terms were adopted from Macdonald and colleagues' review, whilst several additions to the subject headings and keywords used (Appendix D) to facilitate a broader search and identification of more psychologically informed papers. Systematic database searching was complemented with Google Scholar searches and use of forward- and back-chaining (Downe et al., 2009; France et al., 2015). Titles and abstracts were initially screened for the identified papers (against the inclusion and exclusion criteria, Appendix B), and the remaining papers were screened by considering the full-text. See PRISMA diagram (Figure 1) for figures. To enhance rigour, a sample (25%) of full-texts were also screened by two independent researchers. Any disagreements were discussed until mutual consensus was reached. Following consensus, a rationale was provided for the exclusion of full-text papers (Appendix E). For an overview, see PRISMA diagram in Figure 1. Each included study was then quality appraised using the Critical Appraisal Skills Programme (CASP) tool (2018). To enhance rigour, a sample (25%) of the included papers were quality appraised again by two independent researchers. Any disagreements

Noblit and Hare's (1988) Seven Step Meta-ethnographic Approach	Description of the Researcher's Application of Each Step
	were discussed until a mutual consensus was reached. Each CASP was then allocated a score to facilitate later interpretation, as demonstrated by several authors (Duggleby et al., 2011; Elmir, Schmied, Wilkes & Jackson, 2010; Feder et al., 2006). See Table 2 for an overview of the quality appraisal outcomes.
3. Reading the studies	The researcher carefully read and re-read each paper, on multiple occasions, which facilitated the identification of and familiarisation with the main first and second order concepts. The characteristics of each study were noted (summarised in Table 1) including the context for each study, and also their relevance to the synthesis. The researcher noted the epistemological stance of each paper, and noted whether the theoretical foundations were made explicit. These factors were all also considered in the context of the CASP quality appraisal.
4. Determining how the studies are related	Studies continued to be carefully read and re-read in chronological order by the main researcher. Key second order concepts were extracted from each paper (summarised in Table 3). The researcher developed conceptual maps to preserve the original interactional nature of many of these concepts, portrayed by the original authors (e.g. Britten et al., 2002; France et al., 2019). From this stage, the researcher kept analytic memos to capture initial conceptual ideas that later fed into the emerging third order concepts. A constant comparison approach was used throughout, comparing first and second order concepts, considering original conceptual interactions between concepts, the researcher's analytic memos, considering the context of the study characteristics and the quality appraisal.
5. Translating the studies into one another	The main researcher independently led on the translation of studies, which considered the interactional nature of each of the concepts (as exemplified in Table 3). To facilitate this process, the researcher adopted an approach exemplified by Britten et al. (2002), which involved cross-tabulating the concepts, relative to each of the original papers and the emerging third order concepts (Appendix F). This required studies to again be read and re-read, and the continued use of a constant comparative approach to ensure that emerging concepts were not too conceptual and that they were appropriately grounded in the first order concepts also. An illustrative example of how meanings were translated from multiple studies can be seen in Appendix J, and an overview of the concepts in Table 4.

Noblit and Hare's (1988) Seven Step Meta-ethnographic Approach	Description of the Researcher's Application of Each Step
6. Synthesising translations	By utilising the developed cross-tabulation, and including that within the researcher's constant comparative approach (with other analytic tools noted above), the researcher ascertained relationships/interactions between the concepts enabling the synthesis of concepts and generation of third order concepts. The researcher reciprocally translated the concepts (given an lack of scope for refutational translation) and this fed into a line of argument synthesis (see Figure 2 and associated supporting information in the body of the text). I.e. although studies often examined various phenomenon from a different perspective, no refutational 'cases' arose throughout this process (e.g. Thorne, Jensen, Kearney, Noblit, & Sandelowski, 2004), leading to the development of a line of argument.
7. Expressing the synthesis	This aspect of the meta-ethnography is expressed in the conceptual model and accompanying discussion. The synthesis was interpreted in the context of pre-existing theoretical understandings in the area of engagement with (adherence to) treatments when living with a chronic health condition. This included a critique of the emergent model, for example limitations in the context of the epistemologies of the included papers and their respective quality. The model was also situated in terms of the characteristics of the synthesised studies, and the availability of studies. The role of the researcher in interpreting and synthesising the included contexts was considered (i.e. reflexivity) as to how this could have influenced the outcome. Finally, implications for clinical research and practice were expressed based on the emergent synthesis.

Appendix D. ***Search terms used for meta-ethnography systematic database search, including Boolean operators.***

Population	Engagement with (adherence to) treatments	Personal experiences
Subject heading: • Cystic Fibrosis	Subject heading: • Treatment compliance/ Medication compliance/ Compliance • Drug therapy/ Drug/ Treatment • Self-management/ Self-care skills/ Self-care/ Self-efficacy/ Self-concept • Behavioural activation system	Subject heading: • Personal experience • Quality of life • Wellbeing • Life satisfaction • Subjectivity
Keywords (ti,ab,id. and .ti,ab,kf. and.ti,ab.kw.)		
Cystic fibrosis Mucoviscidosis	Adherence Compliance Concordance Management Activation Treatment Drug* Medication Self-care Self-efficacy Confidence	Experience* Subjective Well-being Quality of life
OR	**OR**	**OR**
Then…		
AND		

Appendix E. ***Excluded full-text articles and rationale for exclusion.***

Allgood, S. J., Kozachik, S., Alexander, K. A., Thaxton, A., Vera, M., & Lechtzin, N. (2018). Descriptions of the pain experience in adults and adolescents with cystic fibrosis. ***Pain Management Nursing, 19*****(4), 340-347. doi:http://dx.doi.org/10.1016/j.pmn.2017.11.011**

- The impact of pain on managing (adhering) to a health-management regime was not considered.

Barker, D. H., Driscoll, K. A., Modi, A. C., Light, M. J., & Quittner, A. L. (2012). Supporting cystic fibrosis disease management during adolescence: The role of family and friends. ***Child: Care, Health and Development, 38*****(4), 497-504. doi:http://dx.doi.org/10.1111/j.1365-2214.2011.01286.x**

- Study participants were adolescents (defined as <=18 years) and therefore the wrong participant group were studied for this review.

Collins, S., & Reynolds, F. (2008). How do adults with cystic fibrosis cope following a diagnosis of diabetes? ***Journal of Advanced Nursing, 64*****(5), 478-487. doi:http://dx.doi.org/10.1111/j.1365-2648.2008.04797.x**

- The interaction of adapting to cystic fibrosis related diabetes (CFRD) and managing an already demanding cystic fibrosis health-management regime is not explored.

Cronly, J., & Savage, E. (2019). Developing agency in the transition to self-management of cystic fibrosis in young people. ***Journal of Adolescence, 75*****, 130-137. doi:http://dx.doi.org/10.1016/j.adolescence.2019.07.006**

- Specifically explores transitional period to self-management (and includes predominantly adolescents).

Foster, C., Eisner, C., Oades, P., Sheldon, C., Tripp, J., Goldman, P., . . . Trott, J. (2001). Treatment demands and differential treatment of patients with cystic fibrosis and their siblings: Patient, parent and sibling accounts. ***Child: Care, Health and Development, 27*****(4), 349-364. doi:http://dx.doi.org/10.1046/j.1365-2214.2001.00196.x**

- Study participants ('patients') were aged <=18 years and therefore the wrong participant group were studied for this review.

Greenop, D., & Glenn, S. (2014). Self-care at the margins of healthcare: 'Malingering' and 'self-neglecting' cystic fibrosis patients. ***Qualitative Social Work: Research and Practice, 13*****(3), 389-405. doi:http://dx.doi.org/10.1177/1473325013479392**

- Study adopts a narrative approach, and does not explicitly explore/investigate factors mediating engagement with (adherence to) a cystic fibrosis health-management regime.

Keyte, R., Egan, H., Nash, E. F., Regan, A., Jackson, C., & Mantzios, M. (2019). An exploration into experiences and attitudes regarding risky health behaviours in an adult cystic fibrosis population. *Psychology, Health & Medicine*, 1-7. doi:http://dx.doi.org/10.1080/13548506.2019.1706750

- Study examines engagement in 'health risk behaviours', which have implications for physical health, however does not examine the interaction of these with engaging with (adhering to) a cystic fibrosis health-management regime.

Knudsen, K. B., Boisen, K. A., Katzenstein, T. L., Mortensen, L. H., Pressler, T., Skov, M., & Jarden, M. (2018). Living with cystic fibrosis - A qualitative study of a life coaching intervention. *Patient Preference and Adherence, 12*, 585-594. doi:http://dx.doi.org/10.2147/PPA.S159306

- Examines perceived effectiveness of a specific intervention rather than investigating factors mediating engagement with (adherence to) a health-management regime.

Myers, L. B., & Horn, S. A. (2006). Adherence to Chest Physiotherapy in Adults with Cystic Fibrosis. *Journal of Health Psychology, 11*(6), 915-926. doi:http://dx.doi.org/10.1177/1359105306069093

- Uses questionnaire methodology and not direct interviews. Qualitative content analysis of qualitative questions. Insufficient richness and rigour of data.

Nicolais, C. J., Bernstein, R., Saez-Flores, E., McLean, K. A., Riekert, K. A., & Quittner, A. L. (2019). Identifying factors that facilitate treatment adherence in cystic fibrosis: Qualitative analyses of interviews with parents and adolescents. *Journal of Clinical Psychology in Medical Settings*, No-Specified. doi:http://dx.doi.org/10.1007/s10880-018-9598-z

- Study participants were identified as 'adolescents' (aged between 11 – 20 years) and therefore the wrong participant group were studied for this review.

O'Toole, D. P. H., Latchford, G. J., Duff, A. J. A., Ball, R., McCormack, P., McNamara, P. S., . . . Southern, K. W. (2019). Adherence to aerosol therapy in young people with cystic fibrosis: Patient and parent perspectives following electronic data capture. *Qualitative Health Research, 29*(6), 846-856. doi:http://dx.doi.org/10.1177/1049732318805754

- Study participants ('patients') were aged <=18 years and therefore the wrong participant group were studied for this review.

Rodriguez Hortal, M. C., Hedborg, A., Biguet, G., & Nygren-Bonnier, M. (2018). Experience of using non-invasive ventilation as an adjunct to airway clearance techniques in adults with cystic fibrosis-A qualitative study. *Physiotherapy theory and practice, 34*(4), 264-275. doi:http://dx.doi.org/10.1080/09593985.2017.1400137

- Study does not explicitly explore/investigate factors mediating engagement with (adherence to) to a cystic fibrosis health-management regime.

Sawicki, G. S., Heller, K. S., Demars, N., & Robinson, W. M. (2015). Motivating adherence among adolescents with cystic fibrosis: Youth and parent perspectives. *Pediatric Pulmonology, 50*(2), 127-136. doi:http://dx.doi.org/10.1002/ppul.23017

- Study participants were identified as 'adolescents' (aged between 16 – 21 years) and therefore the wrong participant group were studied for this review.

Schmid-Mohler, G., Caress, A.-L., Spirig, R., Benden, C., & Yorke, J. (2019). "Thrust out of normality"-How adults living with cystic fibrosis experience pulmonary exacerbations: A qualitative study. *Journal of Clinical Nursing, 28*(1-2), 190-200. doi:http://dx.doi.org/10.1111/jocn.14646

- Study investigates experiences of pulmonary exacerbations specifically, not experiences of engagement with (adherence to) a cystic fibrosis health management regime.

Sylvain, C., Lamothe, L., Berthiaume, Y., & Rabasa-Lhoret, R. (2016). How patients' representations of cystic fibrosis-related diabetes inform their health behaviours. *Psychology & Health, 31*(10), 1129-1144. doi:http://dx.doi.org/10.1080/08870446.2016.1183008

- The interaction of living with cystic fibrosis related diabetes (CFRD) and managing an already demanding cystic fibrosis health-management regime is not explored.

Tierney, S., Deaton, C., Webb, K., Jones, A., Dodd, M., McKenna, D., & Rowe, R. (2008). Isolation, motivation and balance: Living with type 1 or cystic fibrosis-related diabetes. *Journal of Clinical Nursing, 17*(7b), 235-243. doi:http://dx.doi.org/10.1111/j.1365-2702.2008.02331.x

- The interaction of living with cystic fibrosis related diabetes (CFRD) and managing an already demanding cystic fibrosis health-management regime is not explored.

Varilek, B. M., & Isaacson, M. J. (2020). The dance of cystic fibrosis: Experiences of living with cystic fibrosis as an adult. *Journal of Clinical Nursing*, No-Specified. doi:http://dx.doi.org/10.1111/jocn.15397

- Study does not examine the interaction between experiences of living with cystic fibrosis and engaging with (adhering to) a cystic fibrosis health-management regime.

Werner, S., Hochman, Y., Rosenne, H., & Kurtz, S. (2020). Cooperation or tension? Dyadic coping in cystic fibrosis. *Family Process*, No-Specified. doi:http://dx.doi.org/10.1111/famp.12538

- Study does not examine the interaction between dyadic coping and engagement in (adherence to) a cystic fibrosis health management regime.

Willis, E., Miller, R., & Wyn, J. (2001). Gendered embodiment and survival for young people with cystic fibrosis. *Social Science & Medicine, 53*(9), 1163-1174. doi:http://dx.doi.org/10.1016/S0277-9536%2800%2900416-0

- Study examines participants that are transitioning to adulthood (i.e. from parental support to independence) and is therefore not eligible to include.

Appendix F. ***Cross-tabulation used to facilitate translation and synthesis phases, produced in Microsoft Excel.***

Concept	Chapman et al. (2004)	Badlan et al. (2006)	George et al. (2010)	Grossoehme et al. (2012)
Illness representation	Role of environment (phenotype) in taking control vs viewed as a genetic cause	Normal' vs. 'different'; 'biographical disruption'; 'imposter'	Influence of perceived health benefits Stigma/embarrassment	Spirituality guides care of body and treatment adherence Making meaning through cognitively re-framing
Locus of control	Internal (phenotype) vs external (genotype)	-	Internal - influenced by perceived immediate health benefit	Internal (accountability to God) vs. external (God gave me this condition)
Behavioural regulation strategies	-	-	Seeking social support; Reminders; Habit & routine; Reward	-
Practical issues	-	-	Ease of treatment (speed); Attendance at CF Clinic	-
Planned non-adherence	-	-	Planned non-adherence as a reward after a period of adherence (e.g. skipping treatments, a night out)	-
Alternative coping strategies	-	-	Substitution of medical treatments for hikes, swimming, gym "Forgetting"	-
External influences	Struggling with understanding of "doctor speak" and seeking medical information from alternative sources	Expectations from healthcare professionals that people are always in a "patient" role, whereas living with CF requires a balance of additional life demands Perception of how others view self	Anticipation and receipt of PFT results; Communications with healthcare professionals; empathy for challenges with engagement and encouragement to persevere; accountability to CF team ("white coat compliance"); need for adequate provision of knowledge about treatment purpose and expectations	Collaborative partnership between self, physician and God.

Concept	Hogan et al. (2014)	Barker et al. (2017)	Oddleifson et al. (2017)
Illness representation	Influence of perceived health benefits Mood/embarrassment	Dual role of parent and patient. Often de-prioritised self and prioritised child Guilt/anxious/frustrated/loss when one role was not prioritised above the other	Competing demands Time intensiveness of treatments Competing priorities
Locus of control	Internal - responsibility of maintaining 'normal' function	Internal - "sharpened focus" - own health impacts foetus - Responsibility	-
Behavioural regulation strategies	External/social support; Routine; Structure; Time management; Organisation	Development and adjustment of routines to meet two demanding roles	Forgetfulness Minimising/Avoiding CF
Practical issues	Competing demands of work and social life Fatigue	"I've got to prioritise" - balance of time constraints as a parent	Need for engagement of people with CF to support their communication about CF with others, thus mitigating a hidden identity. Potential for recursive perception with healthcare professionals
Planned non-adherence	-	Deprioritising own needs to meet child's needs	-
Alternative coping strategies	-	-	-
External influences	"Absence of additional support/knowledge from healthcare professionals about how to adapt nebuliser treatments in order to fit in with competing demands (e.g. tiredness, work/social demands, travel) Support from healthcare professionals through long-term relationship development"	Communication with medical professionals – need for appreciation of the balance of priorities for PwCF	"Need for engagement of people with CF to support their communication about CF with others, thus mitigating a hidden identity. Potential for recursive perception with healthcare professionals"

Concept	Drabble et al. (2019)	Arden et al. (2019)	Eaton et al. (2020)
Illness representation	Perceiving self as more "normal" by not engaging with treatment	Influence of perceived importance of treatments and health benefits Battle of having a "normal" life incl. life goals of family, career vs. engagement with treatments	-
Locus of control	-	Internal (holding realistic expectations of ability to be adherent) vs. external (idealistic expectations of ability to be adherent)	External (mis-communication/gaps in knowledge)
Behavioural regulation strategies	Routine; Reminders	Support of being in hospital; Social support; Planned reward; Routine, cues, prompts; Positive (praise) and negative (told off, negative outcomes) reinforcement	Routine
Practical issues	Tired/Apathetic	Competing demands of life goals. Stressful events e.g. Christmas. Side effects (feel worse rather than better); Tiredness. Ability to self-monitor treatment (nebuliser readings, lung function readings).	Having gaps in knowledge (assumptions by medical team). Unexpected events occur. Competing demands and balancing work. Tired/Fatigued/Burnout.
Planned non-adherence	Moral struggle for patients with low engagement to "admit" and instead use "forgetting"; those with high engagement disclose intentional non-adherence whilst acknowledging it is socially undesirable	Holidays/Travel	Travel (e.g. plan not to take equipment)
Alternative coping strategies	Forgetting to avoid CF and normalise non-engagement with treatment (more socially acceptable)	"intentional forgetting"	-
External influences	Using "forgetting" narrative to mask non-adherence in presence of power dynamic with medical professionals.	-	Professionals' use of active listening skills to understand barriers from patient perspective facilitative of coproduced care and potentially therefore engagement with treatments

Concept	Grossoehme et al. (2020)
Illness representation	Cognitively reframing CF as part of the Divine plan Perceived value & importance of treatments
Locus of control	Spiritual locus of control: Viewing body as a temple (accountability to God & responsibility)
Behavioural regulation strategies	Social support
Practical issues	-
Planned non-adherence	-
Alternative coping strategies	Support from the congregation
External influences	-

Appendix G. ***Illustrative example of translation for the emergent 'Conceptualisation of Health' third order concept.***

		Chapman et al. (2004)	Badlan et al. (2006)	George et al. (2010)	Grossoehme et al. (2012)
Conceptualisation of illness, development of an illness identity and their influence of engagement in (adherence to) treatments	**First-order concepts**	"The genes are already there aren't they? You can't do nothing about it at present". "It was something that I was born with and it can't be changed". "Genes – that's something you don't have a lot of control over". "So my knowledge goes to things I can physically do – treatments such as physio, nebulizers, taking antibiotics, doing a bit of exercise, eating properly, eating the right fatty foods, taking vitamins, doing all that is what I can do". "So the genes, I can't physically stop them producing CF so I leave that to the scientists".	"Not so many of my friends know I have CF I Don't really want them to feel sorry for me. I try to be as normal as I can really, hope to marry my girlfriend, go holidays maybe, if I can have a child". "I've never liked to think of myself as any different from anyone else and I am well determined to stay healthy really". "How can I explain trying to be normal, having a balance. It's unusual, it's a bit odd isn't it really 'cos one minute you feel like normal… then you don't feel normal, it's like swings and roundabouts".	"When I was married I wouldn't do my breathing treatments around my wife… like my nebulizer, after I use it, I push it under my bed. I guess maybe I am hiding it… it is more my concern how they are going to react seeing their sick friend, sick husband, sick boyfriend". "I have 2 roommates… I wasn't going to do it in front of him. I did it while they were off at work. I do that in my room". "When I am out with friends, I don't carry them (enzymes)… because I would feel embarrassed".	"It's nice that CF is genetic. God doesn't make mistakes, so it's a good thing… believing that God doesn't make mistakes is really helpful instead of feeling hurt, and asking, 'Why me?'". "God made my body. He gave me one, and I know He doesn't want me to abuse it… He wants me to take good care of it; after all, he sent nine months making it" "God helps you with staying healthy if you do what you need to do. You have to do your treatments and your medicine. Why should He give you health if you're not going to take care of it?".
	Second-order concepts	Dwelling on the genetic root cause might influence the perception that health maintenance efforts are of little use. Appreciation of an interactional process between root genetic cause (genotype) and influence of treatments/environmental factors (phenotype) could influence treatment approaches.	A sense of ambiguity emerged about 'normality' which reflected a degree of 'biographical disruption'. This depended on the extent to which a person fell into one of two states: "when you are well and when you are not well". This manifested as a sense of being an 'imposter' in a normal world.	Performing treatment regimes in public or in front of people can lead to stigma and embarrassment. "Absence of perceived health benefits". "Failure to achieve symptom relief prompted non-adherence."	Adults coped with their CF diagnosis by making meaning influenced by their spirituality. Spirituality guided the care of the body and treatment adherence. Some hold a belief that God's intervention was conditional on their engagement with treatments and other pro-social behaviours.

		Hogan et al. (2014)	Barker et al. (2017)	Oddleifson et al. (2017)	Arden et al. (2019)
Conceptualisation of illness, development of an illness identity and their influence of engagement in (adherence to) treatments	**First-order concepts**	"If I was in a social situation, out to dinner and a movie, people over, party off with friends spending the night, I didn't want to feel inadequate; I didn't want to feel lesser than they did. And for some reason in my head I was thinking that because I have this illness that I was not as good as they were so I didn't feel like I should be taking medication in front of them".	"When you're pregnant all of a sudden if you are ever lax with doing medication or anything like that you suddenly realise the importance of it because you want your children to be as healthy as possible". "If I had to weigh up do a bottle or do my nebulisers, it had to be the bottle". "If I did have to go in [to hospital] it would have to be childcare that would have to come first". "Not only does it [hospital] take you away from your family and it's not the normal thing… families without cystic fibrosis don't have to contend with that… it can get on your nerves to say it politely".	"So I went to college and acted like every other college freshman and did tons of stupid things [getting drunk] and um yeah when you have CF it doesn't work the same". "We all want to blend in to a certain extent. We all want to be in control of how other people see us". "It's frustrating. All my friends can kind of do whatever they want. They don't have to maintain their health in the same way I do. I want to be like that. But I can't. And they don't fully understand".	"… because that [nebuliser] is helping my chest, this keep certain infections at bay, but every now and then one will creep through". "I was told that [the bacteria] actually become acclimatised to it, and it would lose its effectiveness". "I do definitely think that all the nebulizer stuff is a really good a really good drug to to have and a necessary one I think" "And you can't turn stubborn off, you can't, if you're a stubborn person you're a stubborn person for life".
	Second-order concepts	Medication use was influenced by emotional state, specifically mood and embarrassment.	"It sharpened my focus": Development of a new perspective on optimising health by engaging with treatments. Need to de-prioritise own needs to attend to being the child's primary caregiver. Putting child's needs first at expense of engaging with treatments and maintaining own health.	An individual curates how they perceive others to see them by engaging in 'identity performances'. This can often mean not engaging in treatments to maintain health. Identity performances may also serve to convince the self of a particular identity and in-turn function as a coping mechanism.	Participants' perceived benefits and importance of treatments appeared to influenced their engagement with them. 'Social identities' and personal qualities can influence adherence. Life goals can conflict with adherence: wanting to be normal/have a normal life, family, or child-related goals.

		Drabble et al. (2019)	Grossoehme et al. (2020)	Emerging Third-Order Concept
Conceptualisation of illness, development of an illness identity and their influence of engagement in (adherence to) treatments	**First-order concepts**	"There is just going to be times when you just can't, like it might be someone's birthday, and you might have forgot to do it. And you are not just going to come home from someone's birthday to do your medication". "At the minute it's just like out the window. Like I'll say 'ah I'll do that in the morning.' Get up in the morning, and I'll find something else to do, and it wont be till later on in the day and I'm like 'oh I should have done that, oh I'll do it later when I get in then.' Get home later and it will completely slip my mind and I just forget to do it altogether" "I think it's more like a blocking and like I put out my mind. Like I say if I block it, it isn't there sort of thing… because like, if I don't, if I don't block it, I don't think about it so it's not there sort of thing I feel. So I can, I don't know how to say, not be normal, but more of someone my age sort of thing".	"God or Higher Power must have some purpose for me if I'm here for this long when by all the science and statistics I should have died a long time ago". "He gave it [body] to me and it's [body's] His property to a degree and I have to take care of it [body] for Him". "He gave it to me, and He only gives me what I can handle, so I don't let it control me, I control CF" "He gives me the tools to do it [engage in healthy behaviours/treatments] and it's my choice to follow it or not".	**Illness Representation – patients develop a construction of their illness, and an identity that either incorporates that illness identity or not (potentially resulting in 'biographical disruption'). The degree of dissonance between these identities appears to influence engagement in CF treatments.**
	Second-order concepts	Participants will use forgetting as avoidance of their CF. Using a dialogue of 'forgetting' leads others to believe that non-engagement with treatments is not a choice but a series of smaller decisions that add up to forgetting, and is therefore not intentional. Adherence is presented as a barrier to normality that can subsequently justify non-adherence.	Adults coped with CF by positively re-framing it as part of the Divine plan, and part of that plan is their capability to "control CF".	

Appendix H. *NHS ethical approval confirmation*

Ymchwil Iechyd a Gofal Cymru
Health and Care Research Wales

Gwasanaeth Moeseg Ymchwil
Research Ethics Service

Ariennir gan Lywodraeth Cymru
Funded by Welsh Government

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Dr Jenny Moses
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Cardiff and Vale University Health Board
11th Floor, Tower Building
70 Park Place
Cardiff
CF10 3AT

25 June 2019

Dear Dr Moses

Study title: **Maintaining hope when ageing with cystic fibrosis.**
REC reference: **19/WA/0185**
Protocol number: **SPON 1727-19**
IRAS project ID: **257144**

The Research Ethics Committee reviewed the above application at the meeting held on 19 June 2019. Thank you for attending to discuss the application.

The members of the Committee present gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below. .

Conditions of the favourable opinion

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

Number	Condition
1	The PIS should be amended under the heading 'Who has approved the study?' to include the Wales REC 6. It was not necessary to include dates and telephone numbers in this section. The sentence regarding approval to take place in the NHS by the HRA and HCRW should also be omitted as this is covered by including the Wales REC 6.
2	The charity PIS and consent form version numbers and dates should be linked and consistent.
3	The PIS page 2, third paragraph should mention that participants may be contacted again by *Skype* by the researcher.
4	The Protocol should omit reference to gift cards as these were not being offered.
5	The PIS should include information about direct quotes being used and the consent form should ask for permission for direct quotes to be used.
6	The PIS introductory paragraph should inform participants that the study was being carried out as part of an educational qualification.
7	The NHS consent form made reference to possible use of verbatim quotations and asked how the researcher would ensure the identification of the participant was protected if using verbatim quotes in the transcription

You should notify the REC once all conditions have been met (except for site approvals from host organisations) and provide copies of any revised documentation with updated version

numbers. Revised documents should be submitted to the REC electronically from IRAS. The REC will acknowledge receipt and provide a final list of the approved documentation for the study, which you can make available to host organisations to facilitate their permission for the study. Failure to provide the final versions to the REC may cause delay in obtaining permissions.

Confirmation of Capacity and Capability (in England, Northern Ireland and Wales) or NHS management permission (in Scotland) must be obtained from each host organisation prior to the start of the study at the site concerned.

Confirmation of Capacity and Capability (in England, Northern Ireland and Wales) or NHS management permission (in Scotland) should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).

Guidance on applying for HRA and HCRW Approval (England and Wales)/ NHS permission for research is available in the Integrated Research Application System.

For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.

Sponsors are not required to notify the Committee of management permissions from host organisations.

Registration of Clinical Trials

It is a condition of the REC favourable opinion that all clinical trials are registered on a publicly accessible database. For this purpose, clinical trials are defined as the first four project categories in IRAS project filter question 2. For clinical trials of investigational medicinal products (CTIMPs), other than adult phase I trials, registration is a legal requirement.

Registration should take place as early as possible and within six weeks of recruiting the first research participant at the latest. Failure to register is a breach of these approval conditions, unless a deferral has been agreed by or on behalf of the Research Ethics Committee (see here for more information on requesting a deferral: https://www.hra.nhs.uk/planning-and-improving-research/research-planning/research-registration-research-project-identifiers/

As set out in the UK Policy Framework, research sponsors are responsible for making information about research publicly available before it starts e.g. by registering the research project on a publicly accessible register. Further guidance on registration is available at: https://www.hra.nhs.uk/planning-and-improving-research/research-planning/transparency-responsibilities/

You should notify the REC of the registration details. We routinely audit applications for compliance with these conditions.

Publication of Your Research Summary

We will publish your research summary for the above study on the research summaries section of our website, together with your contact details, no earlier than three months from the date of this favourable opinion letter. Should you wish to provide a substitute contact point, make a request to defer, or require further information, please visit: https://www.hra.nhs.uk/planning-and-improving-research/application-summaries/research-summaries/

It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).

After ethical review: Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study, including early termination of the study
- Final report

The latest guidance on these topics can be found at https://www.hra.nhs.uk/approvals-amendments/managing-your-approval/.

Ethical review of research sites

NHS/HSC Sites

The favourable opinion applies to all NHS/HSC sites taking part in the study taking part in the study, subject to confirmation of Capacity and Capability (in England, Northern Ireland and Wales) or NHS management permission (in Scotland)being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

Non-NHS/HSC sites

I am pleased to confirm that the favourable opinion applies to any non NHS/HSC sites listed in the application, subject to site management permission being obtained prior to the start of the study at the site.

The documents reviewed and approved at the meeting were:

Document	*Version*	*Date*
Copies of advertisement materials for research participants [Sample Poster Advertisement]	1	18 February 2019
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Cardiff University Insurance Certificate]		20 February 2019
HRA Schedule of Events		20 June 2019
HRA Statement of Activities		20 June 2019
Interview schedules or topic guides for participants	1.2	06 February 2019
IRAS Application Form [IRAS_Form_20052019]		20 May 2019
IRAS Checklist XML [Checklist_20052019]		20 May 2019
Letter from sponsor		20 February 2019
Letters of invitation to participant [Participant follow-up letter]	1.0	18 February 2019
Participant consent form [NHS Consent Form]	1.2	15 February 2019
Participant consent form [Non-NHS Consent Form]	1.0	18 February 2019
Participant consent form [Consent to Contact Form]	1.0	15 February 2019
Participant consent form [Member Validation Consent Form]	1.0	11 February 2019
Participant information sheet (PIS) [NHS PIS]	1.3	15 February 2019
Participant information sheet (PIS) [Non-NHS PIS]	1.3	15 February 2019
Referee's report or other scientific critique report [Evidence of peer review]		02 October 2018
Referee's report or other scientific critique report [Evidence of peer review]		10 August 2018
Referee's report or other scientific critique report [Evidence of peer review]		02 October 2018
Referee's report or other scientific critique report [Evidence of peer review]		10 August 2018
Research protocol or project proposal	1.5	05 February 2019
Summary CV for Chief Investigator (CI) [CV for Dr Jenny Moses]		18 April 2019
Summary CV for Student [Steven Stirk]		29 May 2019

Validated questionnaire [Trait Hope Scale]		
Validated questionnaire [Resilience Scale]		
Validated questionnaire [Emotion Thermometer]		

Membership of the Committee

The members of the Ethics Committee who were present at the meeting are listed on the attached sheet.

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

User Feedback

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/

HRA Learning

We are pleased to welcome researchers and research staff to our HRA Learning Events and online learning opportunities– see details at: https://www.hra.nhs.uk/planning-and-improving-research/learning/

19/WA/0185	**Please quote this number on all correspondence**

With the Committee's best wishes for the success of this project.

Yours sincerely

pp. Dr Matthew Lawrence
Chair

E-mail: Wales.REC6@wales.nhs.uk

Enclosures: List of names and professions of members who were present at the meeting and those who submitted written comments

"After ethical review – guidance for researchers"

Copy to: Helen Falconer

Ymchwil Iechyd a Gofal Cymru
Health and Care Research Wales

Gwasanaeth Moeseg Ymchwil
Research Ethics Service

Ariennir gan Lywodraeth Cymru
Funded by Welsh Government

Wales REC 6
c/o Public Health Wales
Building 1
Jobswell Road
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SA31 3HB

Telephone : 01267 61 1164
E-mail : sue.byng@wales.nhs.uk
Website : www.hra.nhs.uk

Dr Jenny Moses
Academic Director & Consultant Clinical Psychologist
Cardiff and Vale University Health Board
11th Floor, Tower Building
70 Park Place
Cardiff
CF10 3AT

1 July 2019

Dear Dr Moses

Study title: **Maintaining hope when ageing with cystic fibrosis.**
REC reference: **19/WA/0185**
Protocol number: **SPON 1727-19**
IRAS project ID: **257144**

Thank you for your email of 1 July 2019. I can confirm the REC has received the documents listed below and that these comply with the approval conditions detailed in our letter dated 25 June 2019

Documents received

The documents received were as follows:

Document	*Version*	*Date*
Participant consent form [non NHS]	1.4	01 July 2019
Participant consent form	1.4	01 July 2019
Participant information sheet (PIS) [non NHS]	1.4	01 July 2019
Participant information sheet (PIS)	1.4	01 July 2019
Protocol	1.6	01 July 2019
Response to Additional Conditions Met [Response conditions table]		01 July 2019

Approved documents

The final list of approved documentation for the study is therefore as follows:

Document	*Version*	*Date*
Copies of advertisement materials for research participants [Sample Poster Advertisement]	1.0	18 February 2019
Evidence of Sponsor insurance or indemnity (non NHS Sponsors only) [Cardiff University Insurance Certificate]		20 February 2019
HRA Schedule of Events		20 June 2019

HRA Statement of Activities		20 June 2019
Interview schedules or topic guides for participants [Interview Schedule]	1.2	06 February 2019
IRAS Application Form [IRAS_Form_20052019]		20 May 2019
IRAS Checklist XML [Checklist_20052019]		20 May 2019
Letter from sponsor		20 February 2019
Letters of invitation to participant [Participant follow-up letter]	1.0	18 February 2019
Participant consent form [Consent to Contact Form]	1.0	15 February 2019
Participant consent form [Member Validation Consent Form]	1.0	11 February 2019
Participant consent form [non NHS]	1.4	01 July 2019
Participant consent form	1.4	01 July 2019
Participant information sheet (PIS) [non NHS]	1.4	01 July 2019
Participant information sheet (PIS)	1.4	01 July 2019
Referee's report or other scientific critique report [Evidence of peer review]		02 October 2018
Referee's report or other scientific critique report [Evidence of peer review]		10 August 2018
Referee's report or other scientific critique report [Evidence of peer review]		02 October 2018
Referee's report or other scientific critique report [Evidence of peer review]		10 August 2018
Research protocol or project proposal	1.6	01 July 2019
Response to Additional Conditions Met [Response table]		01 July 2019
Summary CV for Chief Investigator (CI) Dr Jenny Moses]		18 April 2019
Summary CV for student [Steven Stirk]		29 May 2019
Validated questionnaire [Trait Hope Scale]		
Validated questionnaire [Resilience Scale]		
Validated questionnaire [Emotion Thermometer]		

You should ensure that the sponsor has a copy of the final documentation for the study. It is the sponsor's responsibility to ensure that the documentation is made available to R&D offices at all participating sites.

19/WA/0185	**Please quote this number on all correspondence**

Yours sincerely

Sue Byng

Sue Byng
REC Manager

E-mail: Wales.REC6@wales.nhs.uk

Copy to: *Mr Steven Stirk*
Ms Jane Jones, Cardiff And Vale University Health Board

Appendix I. ***Inclusion and exclusion criteria for the empirical study.***

Inclusion Criteria	Exclusion Criteria
• Participant perceives themselves to have been diagnosed with cystic fibrosis during childhood. • Currently aged 40 years or older. • Sufficiently fluent in English to read and understand the information sheets, give informed consent and to participate in the interviews. • Sufficiently well clinically to take part in an interview lasting 1-1.5hours (with breaks as necessary).	• Diagnosed with cystic fibrosis in adulthood. • Currently aged below 40 years. • Currently presenting with high risk of harm to self (current self-harm, suicide attempt within the last six months). • Death is expected within several weeks (to minimise intrusion). • Acutely unwell/currently experiencing an acute exacerbation.

Appendix J. *Participant Information Sheet*

NHS WALES GIG CYMRU

School of Psychology
Ysgol Seicoleg

South Wales Doctoral Programme in Clinical Psychology
De Cymru Rhaglen Doethuriaeth mewn Seicoleg Glinigol

CARDIFF UNIVERSITY
PRIFYSGOL CAERDYDD

Cardiff University
Tower Building
70 Park Place
Cardiff CF10 3AT
Wales UK

www.psych.cf.ac.uk

Prifysgol Caerdydd
Adeilad y Tŵr
70 Plas y Parc
Caerdydd CF10 3AT
Cymru Y Deyrnas Unedig

PARTICIPANT INFORMATION SHEET

Study: Maintaining hope when ageing with cystic fibrosis.

IRAS ID: 257144

Do you have cystic fibrosis that was diagnosed during childhood?

Are you currently aged 40 years or older?

If **YES** to **BOTH** of the above, you are invited to take part in a Cardiff University research study that aims to find out how people growing older with cystic fibrosis manage with the concept of growing older with a 'childhood condition'. This information sheet will provide further information about the study – please take the time to read it carefully and discuss it with others if you wish. If you have any questions, please feel free to contact any of the researchers (details below).

Thank you for reading this!

Why is the study taking place?
This study is taking place as part of a doctorate qualification in clinical psychology (DClinPsy). Thanks to rapid medical advances people with cystic fibrosis (CF) are living longer than before. Given how quickly medical advances have progressed for people with CF, people have had to live with changing prognoses, and predicted life expectancies. This understandably leads to uncertainty and requires adjustment. It has meant that people living with CF have had lots of new opportunities, for instance, to have a family, work outside the home and engage with leisure opportunities. The current research aims to explore how people with cystic fibrosis have managed to adjust to their changing prognoses and futures.

How will the research help people in the future?
Understanding how people living with cystic fibrosis remain hopeful and resilient may inform the care given by health care professionals working with future generations of people growing older with cystic fibrosis.

What will happen if I decide to take part?
Participation in the study is optional. If you choose to participate, you will be invited by the researcher to share your experiences and views during an interview which might last between 60 – 90 minutes. The interview can take place at your local cystic fibrosis centre (Cardiff, Bristol or Oxford), via telephone/skype, or at your home. Before the interview, the researcher will go through this information sheet with you and ask you to complete a consent form and you will be given the opportunity to ask any questions you may have.
With your permission, the researcher will also ask your local cystic fibrosis centre to provide some basic details about your current health status, including clinical information such as your blood sugar levels, lung function and recent hospital admissions. This information will be kept strictly confidential and will be stored anonymously at Cardiff University for the purposes of the study only.

When the interview is finished, you will also be asked to complete some brief questionnaires which will ask you more about staying hopeful.

Following your interview after the researcher has analysed data from all the interviews, you may also be contacted again by the researcher to ask if you would like to meet so that the researcher can talk to you about the overall findings of the study. This meeting would take place via Skype. This will be an informal meeting, rather than an additional research interview. Participation in any future meeting is also voluntary and you do not have to take part if you don't want to.

Why am I being invited?
You are being invited to take part because you are aged 40 or over and the clinicians responsible for your care at your local cystic fibrosis centre believe you meet the inclusion criteria for the study.

What are the possible disadvantages of taking part?
It is possible that talking about your experience may lead you to consider things from a different perspective, which could lead you to experience uncomfortable emotions. The researcher will make sure there is time at the end of the interview to provide support and advice if this is needed.

What will happen if the researchers are concerned about my welfare following the interview?
By consenting to participate in this study, you will also be consenting for the researcher to contact the clinical psychologist in your local cystic fibrosis health service if they are concerned about your welfare. This referral will be discussed directly with you before contacting the clinical psychologist except in the case of an emergency.

What will happen with my information?
All of your information will remain confidential and be anonymised (unidentifiable). The interview will be audio recorded to allow the researchers to type up (transcribe) the interview, and the transcription will then be held securely within Cardiff University and analysed by the researchers. Direct quotes may be used for academic submissions and research publications, and given this all attempts to remove identifiable information will be made. Please see the additional information below regarding how your information will be handled, under the new GDPR legislation:

Cardiff University is the Sponsor for this study based in the United Kingdom. We will be using information from you in order to undertake this study and will act as the Data Controller for this study. This means that we are responsible for looking after your information and using it properly. Cardiff University will keep identifiable information about you for 15 years after the study has finished. The legal basis we will rely upon to collect and store your information is public task.

Your rights to access, change or move your information are limited, as we need to manage your information in specific ways in order for the research to be reliable and accurate. If you withdraw from the study, we will keep the information about you that we have already obtained. To safeguard your rights, we will use the minimum personally-identifiable information possible.

You can find out more about how we use your information at https://www.cardiff.ac.uk/public-information/policies-and-procedures/data-protection.
The University's Data Protection Officer can be contacted at: inforequest@cardiff.ac.uk.

If you consent to take part in the study, the NHS will collect information from you and your medical records for this research study in accordance with our instructions.

The NHS will use your name and contact details to contact you about the research study, and make sure that relevant information about the study is recorded for your care, and to oversee the quality of the study. Individuals from Cardiff University and regulatory organisations may look at your medical and research records to check the accuracy of the research study. The NHS will pass these details to Cardiff University along with the information collected from you and/or your medical records. The only people in Cardiff University who will have access to information that identifies you will be people who need to contact you about the study or to audit the data collection process. The people who analyse the information will not be able to identify you and will not be able to find out your name or contact details.

The NHS will keep identifiable information about you from this study for one year after the study has finished.

If you change your mind, you are free to withdraw from the research at any time and it will not affect your standard of care or treatment. If you choose to leave the study, we will need to keep any data you have provided up until the point you chose to leave the study and it may be included in the final analysis. As is the case for all participants, you will not be identified in any publications or presentations about the study. However, some quotations from your interview might be included in the published findings.

What if I choose not to take part?
Participation in the study is completely optional. If you decide not to take part, this will not have any influence on the care or treatment you receive.

Who has approved the study?
The study has been approved by an NHS Research Ethics Committee 'Wales REC 6'.

What will happen to the results of the study?
The study results will be written up as part of my doctoral thesis and may also be published in academic journals and presented at conferences. All results will remain anonymous and no one will be able to identify you from the results.

I want to take part – what do I have to do?
If after reading this Information Sheet you would be interested in taking part in the study, or if you would like further information, please return the 'Consent to be Contacted' reply form to the researcher using the enclosed pre-stamped envelope. The researcher will then contact you to arrange an interview.

If you want to find out more or ask any questions before you return the Consent to be Contacted form, please feel free to contact the **Lead Researcher** using the details below.

What if there is a problem?
If at any point during the research, you wish to make a complaint or wish to speak to someone independent of the study, then you can contact: Victoria Samuel (Senior Research Tutor) on 02920 870582.

What will happen next?
After the researchers have received your signed 'Consent to be Contact' form, they will contact you to arrange a time, date and venue that is convenient for you in order to complete the research interview. Upon meeting you/prior to starting the research interview, the researchers will go through this information again with you to check whether you have any questions and that you are still happy to take part and will then ask you to complete a brief Research Consent Form.

Thank you for taking the time to read this information sheet!

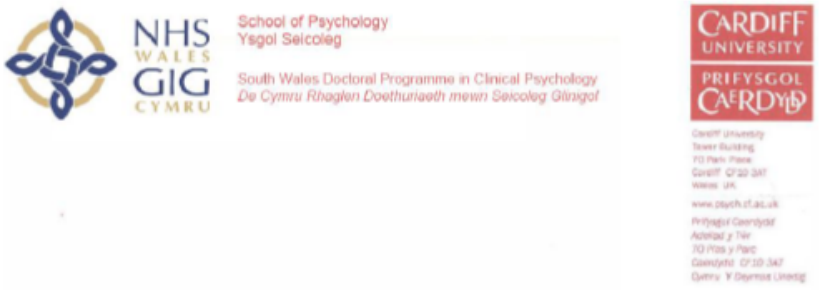
Contact details of the researchers:

Lead Researcher:
Steven Stirk, Trainee Clinical Psychologist & Postgraduate Research Student
11th Floor, Tower Building
70 Park Place
Cardiff
CF10 3AT
Email: stirks@cardiff.ac.uk
Tel: 02920 870582

Chief Investigator:
Jenny Moses, Consultant Clinical Psychologist
11th Floor, Tower Building
70 Park Place
Cardiff
CF10 3AT
Email: Jenny.moses@wales.nhs.uk
Tel: 02920 870582

Principal Investigator:
Anna McCulloch, Clinical Psychologist
All Wales Adult Cystic Fibrosis Centre
University Hospital Llandough
Penlan Road
Penarth
CF64 2XX
Email: Anna.Mcculloch@wales.nhs.uk
Tel: 02920 715937

Appendix K. *Consent to Contact form*



Participant ID: _____

CONSENT TO CONTACT FORM

Study: Maintaining hope when ageing with cystic fibrosis.
IRAS ID: 257144
Researcher: Steven Stirk

If you have read the enclosed Participant Information Sheet and you would be interested in taking part in the study, please complete your details below and return this form (using the enclosed pre-stamped envelope) to:

Steven Stirk
11th Floor, Tower Building
Cardiff University
70 Park Place
Cardiff
CF10 3AT

Please initial the following statement if you agree:

1. I confirm that I have read the information sheet dated 1st July 2019 (version 1.4) for the above study. I have had the opportunity to consider the information and would like to be contacted by the Cardiff University researcher using the contact details provided below. ☐

Please provide your contact details below:
Name:
Address:

Phone:
Email:

Please sign and date:

_____	_____	_____
Name (PLEASE PRINT)	Date	Signature

Appendix L. *Consent form*

School of Psychology
Ysgol Seicoleg

South Wales Doctoral Programme in Clinical Psychology
De Cymru Rhaglen Doethuriaeth mewn Seicoleg Glinigol

Cardiff University
Tower Building
70 Park Place
Cardiff CF10 3AT
Wales UK
www.psych.cf.ac.uk
Prifysgol Caerdydd
Adeilad y Tŵr
70 Plas y Parc
Caerdydd CF10 3AT
Cymru Y Deyrnas Unedig

Participant ID: _____

NHS CONSENT FORM

Study: Maintaining hope when ageing with cystic fibrosis.

IRAS ID: 257144

Researcher: Steven Stirk

Please initial each statement if you agree:

1. I confirm that I have read the information sheet dated 1st July 2019 (version 1.4) for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐
2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected. ☐
3. I consent to use of audio-taping, with possible use of verbatim quotation being included within academic submissions and research publications. I understand that attempts will be made to protect my anonymity, by allocating me a pseudonym (fictitious name) and removing any personal information that could be identifiable. ☐
4. I understand that the information I provide during the interview will be fully anonymised and held securely within Cardiff University. ☐
5. I give permission for relevant information regarding my medical condition (obtained from my medical records) to be made available to the Cardiff University researchers for the purposes of the research. I understand that this information will be treated confidentially by the Cardiff University researchers and used only for the purposes of the study. ☐

6. I consent to the clinical psychologist within my local cystic fibrosis service (Cardiff, Bristol or Oxford) being contacted in circumstances whereby the researchers are concerned about my welfare. ☐

7. I agree to take part in the above study. ☐

Please sign and date:

Name of Participant (PLEASE PRINT)	Date	Signature
Name of Researcher	Date	Signature

Appendix M. ***Illustrative example of grounded theory coding progression***

Abstract from transcribed participant interview data	Line by Line Codes	Focused Codes	Theoretical Codes
Throughout your life you will have had to live with lots of messages about CF being life limited or a childhood illness… how do you think these messages have influenced goals that you have set yourself?			
To be honest… like I said I was born donkeys years ago…	Highlighting sense of old age		
and it was told take her home she's not gonna live long… then it	Describing early life expectancy		
was like oh CFs don't live past their 31st birthday and things like	Describing growing but limited life expectancy	Planning amongst	
that… I didn't make plans cause I thought 'well I'm gonna be dead	Avoiding goals due to limited life expectancy	uncertainty	
by the time I'm 31 anyway' and that's what… it… they… err how	Searching for words		
can I explain… it was a lot of negativity and like 'oh you wouldn't	Remembering pessimistic life expectancy		
live very old and you wouldn't do this…' and I thought 'well what's	Avoiding goals due to limited life expectancy		**Context of uncertainty: Minimising goals, planning & aspirations**
the point in making plans if I'm gonna be dead by the time I'm 31'…			
so I didn't… or you're in hospitals all the time and you did this… I…	Identifying barriers to goal fulfilment		
I think I tried my best at school but I'm not academic anyway… but I	Working hard at school	Planning amongst	
think I could've done a lot more looking back with my life instead of	Identifying missed opportunities	uncertainty	
thinking 'oh what's the point… they've told me I'm not gonna live	Avoiding goals due to limited life expectancy		
long so there's no point' so I think now… it's a lot… you know…	Noticing shift in life expectancy		
lifelong… they give you better prospects for when you're older…	Describing increasing prospects		**Internalising low expectations from others**
and people are living into their 60s and I'm thinking we weren't	Recalling low life expectations		
given that option years ago… I'm talking like I was born in the stone			
age isn't it… no you were just told so you think 'oh… just… just do	Joking about change in perspective		
what you can now and enjoy it' and the future… you've done what	Inheriting messages of low expectation	Developing	
you can… but err.. no I was a… like I said now… there's more	Recognising day-to-day accomplishments	expectations	
encouragement to do stuff because there's better longer lifetime so	Contrasting difference in hope		
I wish I knew that then and I could've perhaps done more…	Regretting missed opportunities		
perhaps I'd have still been who I am today… you just wish there	Contemplating own identity	Forming identity	
wasn't so much negativity back then you know… yeah… and said	Wishing for greater hope when young		
well… I suppose they didn't have the knowledge that they do now…	Recognising increasing knowledge		
and the facilities and the technology and stuff I suppose… you can't	Recognising medical advancements		
change it so… you can't go back in time and change… turn back	Accepting life		
the clock so…			

Appendix N. ***Illustrative example of grounded theory coding progression***

Abstract from transcribed participant interview data	Line by Line Codes	Focused Codes	Theoretical Codes
erm… but when I was an inpatient I used to get my mates to come and take me out for the night…. So I was known as the wanderer [laughs]… so even within the confines of a hospital I'm sure I broke all the protocols but they're… I think y'know they're smart enough to know that your mental state is as important as your physical…. Well… not at the… if you're very ill it's not but…. Most of the time…. So I've always found ways to push what might be termed as kind of normal…. And don't take what people say…. Don't take what people tell you as… y'know… challenge…	Defining coping strategy Identifying as 'wanderer' Identifying as rule breaker Recognising views on mental stage Noticing shifting priorities Finding ways to exemplify 'normal' Noticing importance of challenge	Forming identity	
And what is normal?			
well exactly… I… normal is… is what you currently know isn't it…. So you can… you can shift that.	Describing 'normal' Defining normal as transient	Establishing a 'new normal'	**An ever-changing normality**
I guess in those circumstances…. What would be seen as normal do you think… in that example you gave of being in hospital			
well being in hospital generally you lie there placid waiting for people of of huge intellectual and medical experience to come around and tell you what's gonna happen next…. Erm… and actually in hospital I think loss of power is so…. Erm… it's kinda draining isn't it…. You… your entire rhythm of life works on shift pattern… regardless of what you want… so…. I found that just… y'know that would send me round the twist… erm…. I was really lucky that I didn't have to endure it but erm… very much…. But you have to find little ways to fight against it… so most people I think… certainly my sister would say… there's sort an underlying cantankerousness so she would she would always wear heavy jewellery when she knew she was being weight y'know [laughs] little… little thing I'm gonna… I'm gonna not pulling… you can't really pull the wool over other peoples eyes… but it's sort of a game that everyone plays	Describing helplessness in hospital Conveying experience of powerful medic Acknowledging loss of power Describing emotional impact of powerlessness Fitting in with hospital routine Describing lack of choice Sharing emotional impact of no control Expressing gratitude for health Describing coping strategy Expressing view of others Describing emotional undertone Fighting the system Recognising intentions Accepting mutual roles in hospital	Fighting with consequences of CF Planning amongst uncertainty	**Responding to uncertainty**

Appendix O. ***Illustrative example of grounded theory coding progression***

Abstract from transcribed participant interview data	Line by Line Codes	Focused Codes	Theoretical Codes
Hmmmm…. I didn't at first it was just like… I don't know… you know you know you're gonna get worse as you get older… you've always got that cloud dark cloud hanging over you but… I just didn't know what to do but luckily you've got people like [psychologist] to talk to and things like that so… but… I don't know… you don't want to be told or given negative stuff throughout your illness because you know it's gonna happen anyway so you don't wanna be force fed stuff that you think well it may happen it may not and just in case… I hate that saying… but err… there needs to be… not more understand how can I say… I don't know how to word it… but like a say I was blazey about it until it happened so I didn't want to think about it… but then it does happen then and you… it hits you hard… so I think you… I don't know I can't explain it… I'm not very good at putting things into words anyway… I know how it sounds in my head but trying to say it out loud d'y'know what I mean [laughs]	Struggling with uncertainty Acknowledging progressive nature of CF Identifying 'dark cloud' Feeling aimless Identifying support structures Avoiding 'negative' information on health Justifying avoidance of CF Minimising severity Observing professionals minimising impact Managing uncertainty with avoidance Avoiding own mortality Recognising consequences of avoidance Searching for words	Making sense of uncertainty Avoiding uncertain mortality Living on the cusp of mortality	**Living with (un)certain mortality**
You're doing a great job			
But erm… no yeah… I don't know if those people should talk about… instead of sugar coating everything… which not everybody does do some people are quite brutal and a bit too honest but erm… perhaps it needs to be outlined a bit more that if these things happen they will happen slowly or you know gradually… quickly… everybody's… oh I suppose everybody's different you can't really explain… I don't know how to word it…	Wondering about change Wanting accurate information Considering need for balance Striving for more certainty Realising idiosyncratic nature of CF Searching for words		

Appendix P. ***Example memo***

A DYNAMIC NORMALITY

The sense of normality did not appear to be static, but rather a constantly evolving dynamic process (in the context of such uncertainty). PwCF constantly experience setbacks with their health, whether that's a minor setback (e.g. a 'cold') or a more major setback ('being hit by a train', e.g. being diagnosed with diabetes or cancer). Both have ramifications on quality of life ('you can go from one to another to another and it's restrictive'). Given the constant uncertainty, PwCF also seem to experience an anticipatory fear that their health will suddenly an unexpectedly deteriorate 'like falling off a cliff'.

Conceptualising Normality

"I think so because I just view it as a cold or the flu… in the same way Joe Bloggs down the road would have a cold or flu… I don't need to be hospitalised for it erm… I definitely would have antibiotics a lot quicker than a lot of people would but… I wouldn't contact the team here or anything over it… just make them aware of it if they ask next time kinda thing… erm… yeah but that's probably the level of it…" Participant 2

"up until my 30s and stuff, when I got married and I started, I think I was married and within 2 months that was my first lot of IVs, that was a big shock y'know, having to come in, then, it was once a year, I could cope with that, it was just coming for an mot type of think y'know, just to top up every year, then it started to get a bit more and a bit more" Participant 10

Normality is Dynamic/Fluctuating Normality

"well exactly… I… normal is… is what you currently know isn't it…. So you can… you can shift that... being in hospital generally you lie there placid waiting for people of of huge intellectual and medical experience to come around and tell you what's gonna happen next…. Erm… and actually in hospital I think loss of power is so…. Erm… it's kinda draining isn't it…. You… your entire rhythm of life works on shift pattern… regardless of what you want… so…. I found that just… y'know that would send me round the twist" Participant 1

I've really struggled since I hit my 40s, I was quite well before that and then I hit my 40s and rather than having a gradual decline I've had a massive dip, sort of like, right down" Participant 10

"I kept having quite a lot of exacerbations, would have an exacerbation and my lung function would drop again and then I would gradually make my-, but never get back to where I was, I would get better, bit worse, bit better, bit worse again" Participant 10

"I'd say yes because I pick up colds so easy… you can go from one to another to another and it's restrictive as in anyone who would get a cold… I appreciate that but I seem to get more than one so yeah it's restrictive erm… not terribly restrictive but you know like if you've got things you wanna do over several weeks and then you're nearly housebound because you're not well…. It is restrictive… it's restrictive cause you don't get to see the people you want to see and you don't want to share bugs and things like that so

yeah in that way I suppose I hadn't really thought of it but… yeah it does in general restrict things doesn't it…" Participant 2

"Um, I never would be coughing all the time, but I would have the typical CF cough. I'd have wheeze and I'd always bring up phlegm. I don't any more." Participant 5

When well, PwCF appeared to manage with the uncertainty surrounding their health in certain ways, as discussed above. What also transpired, is how PwCF managed with the regular fluctuations to their sense of normality. Some participants expressed their need to have a sense of purpose. One participant talked about one of her recent experiences of being admitted in hospital:

"Oh like my arts and crafts I suppose I get involved in that… and that focuses my mind on something else then… and like when I'm in hospital… cause I was in for three weeks I brought them with me… so instead of sitting in my room and watching TV and feeling bored and then the boredom gets to you which is the worst… do something that you enjoy even if it means bringing in three extra bags of beads and glitter and glues and… but then I was glad I did it because erm… I made fairy dolls out of silk and pipe cleaners and stuff and then I made little crystal and beaded angel charms so the staff all had one each cause I was making them for them… so when I really look back on it that was really my strategy when I was in hospital of coping… " Participant 3

ADAPTATION - ADJUSTING to ill health/ageing

Bouncing Back/Popping the Dents Out

"I mean, I have since bounced back, but that made me think I need to mentally prepare for that a bit more, because I hadn't thought about that, so I need to kind of-, but in some ways it's been useful, because it gives me time to process it a bit and think.
Yeah.
Now if it does happen I'll be like, 'Oh, well, that happened before. Maybe I'll get better, maybe I won't-,'
Yeah.
But it just made me think that when I get to the point where I don't get better again I'm going to need stuff to do round the house that's going to stop me going doolally-,"

"I've got this inner strength that sometimes I think I haven't got… that you think 'oh god I don't know if I'm gonna get over this one now'.. erm… like I said earlier when I was younger I could just feel as if I could conquer the world… and then I got through all of those and every kind of ill health that I've had since… it does push you down a little bit further and you think 'have I got the strength in me to get over this one now?' and… for some reason I manage to pull it out the bag… and I don't know how… so I've been given… I'm fortunate enough to be given this inner strength that I can rely on… maybe not straight away cause you kind of have to process.. grieve for the bit that you're not gonna be able to do again… and then move on and then you get the fight mode in and then I'm away… I'm like a dog with a bone then I just can't you know release it" Participant 4

GRIEVING

> *"I kind of have to have like a little bit of a 'grieving period' [laughs] I call it… and then time for… to…kind of you have to accept it and then proves it and move on… didn't even think of that when I was 17… you just did it… I just did it…" Participant 4*

Another participant described this same process, where they became 'really depressed', and how this shifted their mindset and reframed their circumstances ('it could be worse...'):

> *"It was hard at first… like that's when I got really really depressed… but then I just think 'y'know'…. I said it the other day and he [husband] said don't say stuff like that… I said well it could be worse I could be dead… at least I've got a future to plan at the moment… you know some people haven't… they're given like a three month you know… time… you know you've got three months left to live… so I think… even though I get my bad days and I feel like rubbish I think I'm a lot better off than a lot of other people so I just… I try and think of it like that and not feel so sorry for myself" Participant 3*

> *"Life is more precious isn't it… and… erm… you know your life might be deteriorating a little bit every year and so these things get to you a little bit more…" Participant 4*
> *"Oh it's a constant struggle of re-adapting all the time… so for instance before I had the breast cancer I was doing really well… so… I'll go back a little bit now then so I qualified as a doctor… so you know as a junior doctor you have to work these hideous hours… I was working sometimes 72… 80 hours a week and I was doing really well so I was flying and I was doing exactly what I wanted… my health was really good… and then it's almost as if a juggernaut comes and stops you in your tracks… erm… and so that… you kind of have to ok… readjust now… to the new kind of expectations of health…. And that's what it is with CF I think… you get so many kind of different illnesses and knocks that you… it's a constant readjustment and realigning your expectations for the future… and that's not easy… that's not easy… but you have to do it… you have to move on… and think well okay I can't do it this way so I have to do it another way… and that's what it is… basically being… not flexible… but readjusting is the word I think… it's not easy…." Participant 4*

Describing process of adjustment to the new norm, which takes time. During the process, when facing real adversity, finding small things in the day that bring happiness as a way to cope and get through those times.

> *"Um, er, I don't know. Just, just time and waiting for things to get better and slowly then over time just keeping-, just keeping doing the things you can do. Actually you just get used to it and then you adjust to whatever new norm that is. That's probably true when I've been ill and stuff as well. It's just keeping doing what you can do and then actually slowly you're kind of bringing-, cope, learned how to cope with it, um, and adjust your expectations. That's not very helpful advice to anyone who's going through it, but it, it-, for me, I don't know whether it's-, whether other (TC: 00:50:00) people have better coping strategies. Probably. But time is the thing for me that just seems to make the difference. It's not very easy when you're in the middle of something, like when I was ill last year, that was-, that was difficult*

to deal with. How did I deal with that then? Um, trying to be happy, trying to find some happiness during the day, whatever it was, because I was too tired and too ill to do very much. But okay, I did-, I enjoyed doing a puzzle. So I enjoyed that. Ooh, I listened to a nice radio programme or I went for a five-minute walk round the block or-,
Yeah.
Trying to find something in the day to make me happy, um, and then that day I could like, 'Tick, I've done that day. Going onto the next day.' And then tick. And then soon you've got-, banked up a group of days where you've been largely happy-,
Yeah.
Even if it's not where you want to be and you're not achieving any of the things or doing anything and you're feeling ill-,
Yeah.
If I can feel that I've felt-, if I've had a happy day then I can manage it. If I-, I mean, obviously I'm not happy all the time. Some days I'm just miserable as anything. But then again I-, even when I'm feeling miserable, I just try and find something that gives me some sort of sense of slight enjoyment or is less bad than everything else." Participant 6

Accepting Loss of Health and Function

Some participants spoke about their process of adapting to periods of ill health, whereby when they struggled for motivation they adopted a mindset of acceptance over what they can and can no longer do:

"By getting on as best I can so… it's not like you can maybe achieve everything that you want or need to achieve but you try your best don't you like anyone… but then if it's… yeah you just adapt really and just do your best… put things off I suppose where they can be put off… it's like everyone that's ill isn't it… like you get a mother who's ill she's still gotta look after her children and it's exactly the same isn't it… whereas it'll kill a man [laughs] but yeah…" Participant 2

"It's just all about looking after yourself and realising your potential… what you can do and what you can't do… you've got to realise that from a young age… you can't do what other people do… sometimes not all the time… and you gotta take a step back and think oh well I can't do that unfortunately… but it's just… that's about it really…" Participant 3

Interaction between setting small goals, finding a sense of achievement, and coping with adversity.

"Um, and then, yeah, um, I think that's-, for me that's probably the biggest thing, is just keep it small. If it's all-, if it's all too difficult, just really narrow it all down to something I can do. I mean, I love doing puzzles. I do puzzles all the time, you know, possibly because there's a start and an end and, ooh, I've achieved it." Participant 6

Finding Ways to Exemplify 'Normal'

In the context of an always-changing sense of 'normal', and a constant process of re-adjusting their concepts of 'normal', PwCF appear to manage this by establishing roles that are perceived as normative within their particular societal context.

"I know that more people are working with CF… and it's not all about the working but it's achieving normal things I Think… you know things that people like you or my husband are able to do… you know why can't we do those things… why can't we go to university and live alone…" Participant 4

"When I'm in hospital they always used to say to me 'you're a stubborn little sod' it's always like, if I've got to go to an X-ray, I'll struggle to X-ray y'know, I will not go in a wheelchair, I don't want to be in a wheelchair, that's always been my philosophy, so y'know they really have to struggle if I'm poorly" Participant 10

"I, I, I did want to, um-, I did just want to be like everyone else, really, and get, get-, yeah, get a degree, get-, I, I didn't know if I'd ever be employed full-time, because of the amount I had to do, and I thought-, I, I really didn't know about that. I thought I-, yeah, I thought I was unemployable really. Er, that had a massive effect on me." Participant 8

"Life is very busy as an adult… and it is just day to day routine isn't it… and normality and… erm… I've never had been one to stop and think about my health I suppose… erm cause I've always worked… always been at school and then worked… you just get on with life don't you" Participant 2

For one participant, this was important for them to re-establish their sense of normality, particularly as during a period of ill health in hospital, there was an internal struggle with being perceived as the 'patient with CF' and not '[*name]* the doctor' which is their work identity:

"when I had erm the breast cancer two years ago everyone said to me 'oh just have the year off'… I was back in work after 6 weeks because it's the sense of going back and being normal again for me… you know getting up… just the whole routine of going to work and being valued by my patients and my colleagues… erm that's really important to me… and being seen as maybe 'X the doctor' rather than 'X with CF'" Participant 4

"...going back to the whole work thing… it gives me a sense of worth and self-esteem and confidence to be that kind of… back to normality type of thing so it's really important for me… like I said it's not for everyone but it's really important for me anyway… and so as soon as I'm well I do go back to work because it helps me psychologically to get back to a normal space in my head if you know what I mean" Participant 4

Another participant talked about the importance of their experience of adjusting to a new role/normal whilst waiting for a kidney transplant:

"I couldn't go back to university until the September… so that's when I did all my charity work then cause I thought 'well I've got to do something'… and erm… so that's when I got really involved and I loved it…"

Whilst another participant reflected on the impact of the uncertain nature of CF on maintain a sense of normality:

"The uncertainty of whether I could or couldn't turn up to work, then I thought, 'Well, that's no good to man nor beast.' I honestly cannot tell you, you know-, am I going to be well next week? I do not know." Participant 5

Appendix Q. ***Example memo***

Developing Expectations

This code reflects two aspects of the development of low expectations. Initially, it reflects the significantly different medical expectations of the time, when these participants were born. It was not expected that they would reach adulthood, and so there was little hope. Despite the significant prognostic shift, participants maintained this approach to managing expectations. It appeared protective to continue to maintain low expectations, so that one cannot easily fail. Living in this context, with the maintenance of low expectations, links with memo attached to the 'Planning Amongst Uncertainty' code.

The PwCF interviewed within the study have all developed a process of living amongst an (un)certain mortality, whereby they developed exceptionally low expectations for themselves.

> *"You try and tell a 12-year-old boy to set life goals when he fully expects to die. Now, I apologise that you might need counselling after this session, by the way, you, you know. So I've always had not people telling me you're going to die, but somehow I was always aware of what is probably expected." Participant 5*
>
> *"So my psychology, I sort of self-taught to-, and I believe it was [clinician] that told me it's defensive, a pessimist defensive, so you raise the bar-, have the bar very low in life.*
> ***Yeah.***
> *You almost expect to be tripped up by that low bar. So then you're not upset when you do trip up, because that's what you expected, but if you get over it, it's like, 'Wow. Yay. Brilliant.'" Participant 5*
>
> *"I didn't go to uni because of being poorly, I just didn't contemplate it… if I was well, I would definitely have gone to university" Participant 9*
>
> *"Well I try and do everything that I can that year, just in case I'm not well the next year or not here, so I've sort of lived for the day kind of thing, I'm not really good at saving money I'll just spend it… I live for today rather than the future*
> ***Is that something you've always done?***
> *Yeah, my whole life I've done that" Participant 9*

Several participants expressed how they believed this was almost a habitual process that was internalised from a young age, influenced by the pessimistic medical expectations at that time:

> *"Well… they were low… err so my parents joined CF Haven and we knew when they would get phone calls from parents in the group that had just lost their children… so y…. you kind of know that cause they…. So you know that growing up… it's not… it's not weekly but y'know it was regular enough to know that that erm… people were succumbing to what we had" Participant 1*
> *"I think they probably took the view that while we've got them we'll make the best of it." Participant 1*

"my parents were told when I was born just to take me home and enjoy me" Participant 3

"Yeah cause like when I was born they were told like in the 70s then oh she won't live long" Participant 3

"I think I could've done a lot more looking back with my life instead of thinking 'oh what's the point… they've told me I'm not gonna live long so there's no point' so I think now… it's a lot… you know… lifelong… they give you better prospects for when you're older… and people are living into their 60s and I'm thinking we weren't given that option years ago… I'm talking like I was born in the stone age isn't it… no you were just told so you think 'oh… just… just do what you can now and enjoy it'" Participant 3

"No… we were always encouraged in those days not to work… so things have changed a lot now… I'm going back 20 years ago… it was always encouraged for us all not to work and to live off benefits…" Participant 4

The consequence of this for many participants was how they approached planning and goal pursuit, as informed by their internalised low expectations:

"So my psychology, I sort of self-taught to-, and I believe it was Jackie that told me it's defensive, a pessimist defensive, so you raise the bar-, have the bar very low in life.
Yeah.
You almost expect to be tripped up by that low bar. So then you're not upset when you do trip up, because that's what you expected, but if you get over it, it's like, 'Wow. Yay. Brilliant.'" Participant 5

"So linking it all, you could sort of almost come to the conclusion I don't hope, I don't wish and I don't have setbacks, because my defensive pessimistic way leads me to deduce what is a SMART target?" Participant 5

"I've got a different viewpoint, definitely, than lots of other people, non-CF people. I'm sure that's true with all, all people with long-term illnesses-,
Yeah.
Have got quite a different viewpoint on life. I generally don't expect things to happen, but then I'm very pleasantly surprised when nice things do happen.
Okay.
I just kind of roll with it a lot more than a lot of other people. A lot of other people worry about what's going to happen, whereas I'm just like, 'It'll happen or it won't,' and just kind of-, I'm a lot more steady, I think, generally." Participant 6

"But I definitely do expect things to go worse so that when things are good I'm like, 'Yay,' which is generally my default. But, but equally I don't think I'm pessimistic, because I think-, you know, I think I'm quite an optimist, so I don't quite know how that all sits. But, but yeah, it's definitely expecting things to go wrong so that when they don't I can feel quite happy about it-," Participant 6

"You know, when I hear something new's coming, you know, I don't instantly think it's going to be a gamechanger. My initial thing is, er, will I be able to tolerate it first.
Yeah. Okay, yeah.

You know. Um, yeah, I don't have an expectation on it, sort of thing, necessarily, um, because I know sometimes I can't tolerate some of the things that have come out, so-,
Yeah. So you've kind of-,
Yeah, so I don't-, yeah, I've quite a neutral expectation, I think, about stuff."
Participant 7

Appendix R. ***Theoretical sampling through interview schedule development***

Stem question	Example prompts	Example theoretical sampling prompts
1. During your lifetime there have been lots of medical advances which have impacted on people living with cystic fibrosis. To what extent have these had a personal impact on you?	• How did your expectations change as you grew older? • What were the expectations of your parents/caregivers, and did these differ from your own?	
2. If I understand right, living with cystic fibrosis often involves lots of uncertainty/not knowing what's around the corner. If this is right, what are some of the 'unknowns' you have experienced and how did you get through these times?	• How has your perception of these uncertainties changed over time? • What helps you manage day-to-day with the smaller ups and downs? • Have there been times where you have really struggled for motivation?	• How did you manage with the uncertainty? • How has the uncertainty shaped your personality? (*e.g. 'stubbornness', 'head strong')*
3. Throughout your life I guess you will have had to live with messages about CF being 'life limiting'. How has this influenced the goals you have set yourself?	• How might these have changed in response to an updated medical opinion about your health?	• How did the uncertainty impact on your goals? • How has this shaped your expectations?
4. Cystic fibrosis has been described as 'one of the hardest health conditions to manage'. What helps you to keep hopeful day-to-day?		
5. Lots of people with cystic fibrosis have difficult times when it is hard to see how you will achieve your goals. Can you tell me about how you get through such times?		• How have you (re)adjusted following a set back? • What helped you to adjust?
6. What have been the consequences for you of having survived into your 'older age' (40s, 50s, 60s) with CF?	• Prompt re. range of opportunities, fears, health/wellbeing etc.	

7. As I understand, some people living with cystic fibrosis are doing things that previously weren't possible, such as becoming parents. If you had your time again are there decisions that you might have made differently?	• What helped you to make the decision at the time? • What kind of feelings does this bring up for you now?	How do you manage now, with the decision that you made then?
8. What makes you proud about being an older person with cystic fibrosis?		

Appendix S. ***Feedback form used for interview schedule consultation process***

Cystic Fibrosis advancing care with clinical trials

Lay Reviewers Score Sheet for Patient Information Sheet

Study title	**Maintaining hope when ageing with cystic fibrosis.**		
Date for return of review feedback	2nd September 2019		
What were your initial or immediate thoughts on seeing the Interview Schedule?			
Lay Reviewers initials		Date of review	

These questions may help guide your review. Please add comments where you can – these provide the most useful feedback

KEY: 1=Poor; 2=Requires Improvement; 3=Adequate; 4=Good; 5=Excellent

Presentation of questions:-	**Rating 1-5**	**Please comment on your rating score?**
Clarity of initial instructions		
Clarity of the questions (is it clear what the questions are asking?)		
Use of plain English throughout		
Sensitivity of the questions (do you feel that the wording is appropriate?)		
Scope for Lay Reviewers	**Rating 1-5**	**Please comment on your rating score?**
Outline of review requirements or experience (how well did you know what was expected of you as a reviewer?)		
Timescales allowed		
Any other comments		

Thank you for your feedback