



Whelan, Frieda (2018) *Cystic fibrosis and family relationships: adolescent, parent and health-professional perspectives*. D Clin Psy thesis.

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**Cystic fibrosis and family relationships: adolescent, parent and
health-professional perspectives**

And

Clinical Research Portfolio

Frieda Whelan, BSc Honours

Submitted in partial fulfilment of the requirements for the degree of
Doctorate in Clinical Psychology

Institute of Health and Wellbeing
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October 2018

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Acknowledgements

Firstly, thank you to all of the participants who shared their experiences with me. I am very grateful for your time and openness and could not have done the project without you.

Many thanks to Lesley Blaikie and Linda Cassidy (Clinical Nurse Specialists) for all their time and efforts in recruiting to the project.

Thank you to my academic supervisor Dr Alison Jackson for her knowledge, guidance and support throughout the project. Thank you to Dr Louise Phin for her support and enthusiasm in developing the project with me. My thanks also to my supervisors this year, Dr Ruth Hind and Dr Stephanie Bannon, for their help and encouragement. It was a pleasure to work with you all.

I would also like to thank my friends, including all my Highland friends who have made the last three years so fun and memorable. Thank you, Tracey and Claire, for sharing the journey! A special thanks to my family, particularly my mum for her continual love and support.

Finally, thank you to Greg for your constant support and belief in me throughout this journey. Thank you for being a husband, best friend, proof-reader, mock-interviewer and technician all in one!

Chapter 1: Systematic Review

Family system and parental functioning and the mental health of children and adolescents with cystic fibrosis

Prepared in accordance with the author guidelines for the Journal of Pediatric Psychology (see author guidelines in appendix 1.1 on page 86)

Chapter word count (including figures, tables, references):7473

Abstract

Objective: To explore the association between family functioning and mental health in children with cystic fibrosis.

Methods: A systematic search was conducted to identify papers reporting on an association between a child mental health variable and a measure of family/parental functioning. Search terms yielded 26 articles suitable for review. Studies were rated for quality and 11 “poor” quality articles excluded from quantitative synthesis. Participants included 1740 children and 1684 parent/caregivers (median number of families per study = 49.0).

Results: In all cross-sectional studies, at least one measure of family functioning was associated with a child mental health measure. Associations with parenting stress were mixed. In longitudinal designs, significant associations diminished by year three follow-up.

Conclusions: Less adaptive family functioning is associated with poorer child mental health in all cross-sectional studies except two. Clinically, results suggest the importance of the inclusion of family and parental functioning in assessments of emotional needs, to ascertain if individual or systemic intervention is warranted. An understanding of which family functioning factors are associated with child psychological wellbeing may also inform future systemic interventions for children with cystic fibrosis and their families.

Key words: systematic review; cystic fibrosis; mental health; children; family functioning

Introduction

Cystic fibrosis (CF) is a life-limiting genetic condition, affecting around 10,400 people in the UK and 100,000 worldwide (Cystic Fibrosis Trust, n.d). Abnormalities in salt transport around the body lead to a build-up of mucous, affecting the lungs predominantly but also other organs (Elborn, 2016). CF treatment involves a time-intensive daily management regime (Ziaian *et al.*, 2006), which can be burdensome for both the individual and family. Furthermore, it is recommended that individuals with CF are segregated for infection control reasons (Cystic Fibrosis Trust, 2004) and so family emotional support may be of increased importance due to a lack of face-to-face peer support. Those with CF and their caregivers experience increased rates of anxiety and depression, such that routine screening of these difficulties is recommended (Quittner *et al.*, 2016). The practical and psychological toll of CF, therefore, can be a source of pressure on the family system. As such, it is of importance that the emotional needs of those with CF and their families are understood and met.

Family Functioning in Paediatric Chronic Health Conditions

Studies exploring interactions between childhood chronic health conditions and family functioning have found mixed results. Herzer *et al.* (2010) analysed studies which used the Family Assessment Device in healthy children and children with CF, obesity, sickle cell disease, inflammatory bowel disease and epilepsy. For children with CF ($n = 59$), 22% of families were rated in the “unhealthy” range for overall family functioning, however, this was not significantly different to the functioning of other groups (*range* 23-32%). To examine the differences in family functioning between families of children with health conditions and those without, McClellan and Cohen (2007) reviewed six

studies featuring a group with CF. Two reported enhanced functioning, three reported poorer functioning and one found no significant differences. All studies had relatively small sample sizes (group with CF *mean n* = 29, *range* = 15-41), and all but two used different measures. In a broad review of studies on CF and the family, Berge and Patterson (2004) similarly found mixed results. Again, studies tended to have small sample sizes (*mean n* = 29, *range* = 23-41). Possibly, small sample sizes and varied measures of family functioning have contributed to inconsistencies. Indeed, Alderfer *et al.* (2008) suggest that family functioning in paediatric populations cannot necessarily be compared with healthy populations and that ostensibly problematic family interactions may be functional in the context of a chronic health condition. The authors suggest that, for example, circumscribed roles may promote treatment management. Inflexibility of roles and routine may facilitate consistent adherence to daily regimes of CF care, whereas in healthy populations role inflexibility may be considered problematic.

Family Functioning and Child Mental Health

Assessing the relationship of family functioning with clinically relevant variables, such as the child's psychological health, could help identify potentially modifiable targets for intervention. Drotar (1997) reviewed 50 studies examining family functioning in a wide range of chronic paediatric health conditions including CF, with 46 reporting on significant associations between family functioning and child psychological adjustment. Studies on CF were not reviewed in isolation, so it is unclear how family functioning interacts with CF specifically. Indeed, Drotar (1997) suggested that future research should explore whether family functioning interacts with adjustment differentially across health conditions. CF may be considered a particularly unique

condition in terms of its life-limiting and incurable nature, with demanding treatment regime.

Leeman *et al.* (2016) conducted a meta-analysis exploring the relationship between family functioning and child wellbeing across a wide range of chronic conditions. To facilitate comparability, articles were included if one of four specified family measures was used. Broadly, Leeman *et al.* (2016) concluded that family functioning was related to child psychological wellbeing, particularly cohesion and conflict. Only one study on CF was identified (Szyndler *et al.*, 2005). However, a range of measures of family functioning have been employed in studies of CF. Examples of measures used in CF studies include family support (Graetz, Shute & Sawyer, 2000) and parental coping (Wong & Heriot, 2008). Knafl *et al.* (2015) critique that existing reviews on paediatric health conditions and the family often do not specify which family factors they will include, and so they developed a framework for how variables can be conceptualised. Understanding how family functioning variables interact with child mental health can potentially inform future systemic interventions. This is especially important as there remains limited evidence-based psychological treatments for those with CF and their families, particularly systemic interventions (Goldbeck *et al.*, 2014).

Rationale

Previous reviews have included an aggregate group of paediatric health conditions, but this could potentially obscure the presence or absence of associations in CF specifically. Reviews by Drotar *et al.* (1997) and Leeman *et al.* (2016) have included children with a broad range of chronic health conditions, including diabetes, cancer, sickle cell disease, asthma, epilepsy and juvenile rheumatoid arthritis. However, CF is

a particularly unique chronic health condition in terms of its life-limiting nature, with particularly demanding daily treatment regime and lack of availability of face-to-face peer support. Although these reviews have been helpful in understanding the commonalities between various chronic health conditions, this could potentially obscure the presence or absence of associations in CF specifically. Indeed, the same family factors have been differentially related to different child mental health outcomes in diabetes and CF (Mullins *et al.*, 1995). Understanding which family functioning factors are related to child mental health outcomes and in which direction, can guide assessment and intervention in families with CF. As such, the purpose of the present article is to consider family functioning in a broad sense, so that a systemic review of CF alone can be conducted.

Objectives

1. To summarise the literature on the association of family functioning and mental health in children with CF.
2. To explore how family functioning has been measured.

Methods

Eligibility Criteria

Inclusion criteria:

- Quantitative studies published in English-language peer-reviewed journals
- Participants are children/adolescents (aged ≤ 18) with a diagnosis of CF and/or family member(s)
- A family functioning variable must be measured and reported, guided by criteria developed by Knafel *et al.* (2015; appendix 1.2, page 88)
- A child mental health variable must be measured and reported
- A statistical association is made between the variables

Exclusion criteria:

- Reviews, dissertations, single case studies and conference abstracts
- Qualitative methods
- Intervention studies

Information sources.

1. The following databases were searched in June 2018: Medline (1946 – 2018), PsychINFO (1967-2018), EMBASE (1947-2018), CINAHL (1971-2018).
2. Key journals were hand-searched. The Journal of Cystic Fibrosis was searched from its inception in 2002 until May 2018. All articles in the Journal of Pediatric Psychology containing the words “cystic fibrosis” were searched (1976 – June 2018).
3. References of included articles and other key studies were considered.

Search strategy.

A university librarian was consulted for assistance in refining search terms. Full search terms for all databases are shown in appendix 1.3 (page 90). Variations of the following terms were used:

1. Cystic fibrosis

AND

2. Parents or family or caregivers or mother or father or brother or sister

AND

3. Adjustment or adaptation or wellbeing or mental health or depression or anxiety or internalising or externalising or emotion or psychopathology or resilience

AND

4. Child or adolescent or paediatric or youth or young person/people or teen or infant

Study selection.

The process of study selection is outlined in Figure I. Search terms yielded 3356 studies. Studies were gathered in EndNote and duplicates ($n = 756$) removed. Guided by eligibility criteria, 2600 titles and abstracts were screened for relevance; resulting in the removal of 2534 records. 66 full-text articles were accessed to determine eligibility. 40 articles were then removed, most frequently because a child mental health variable was not reported ($n = 16$). The remaining 26 articles were considered suitable for review and were assessed for quality (see below). Eleven articles were rated as poor quality and excluded from the final review, leaving 15 articles for inclusion in the quantitative synthesis. Articles by Thompson *et al.* (1992, 1994, 1999)

shared a cohort, as did those by Ward *et al.* (2009) and Sheehan *et al.* (2012; 2014), so they are considered as two studies. The review included 11 separate studies.

Quality rating.

A quality rating tool was developed (appendix 1.4, page 92) to standardise the appraisal process. A tool was developed as existing tools did not cover the range of factors most relevant to the type of studies included. The following guidelines were consulted in the development of the tool: SIGN 50 guidelines (2015), CONSORT guidelines (Boutron *et al.*, 2008) and the STROBE Statement (von Elm *et al.*, 2007). The tool was piloted and refined to ensure it captured the important features of included studies. Points were awarded to studies for the abstract, rationale and objectives, description of participants, outcome measures, confounding variables, statistical analysis and discussion. A maximum of 36 points could be awarded, with total scores calculated as a percentage. Studies achieving fewer than 18 points were excluded from the review. A subset of four papers was co-rated by another researcher (trainee clinical psychologist), and any discrepancies in ratings discussed until agreement was reached. Studies were considered “good quality” if they achieved a rating of 80% or above, “moderate” if between 50% and 80% and “poor” if below 50%. The following 11 studies were rated below 50% and were therefore excluded: Cappelli *et al.* (1989), Cowen *et al.* (1985), Granvold, Woltman & Hocher(1990), Johnson *et al.* (1985), Kucia *et al.* (1979), Pumariega *et al.* (1993), Simmons *et al.* (1987), Stawski *et al.* (1997), Steinhausen & Schindler(1981), Steinhausen, Schindler & Stephan(1983) and Wilson *et al.* (1996). Poor-quality studies were published between 1979 and 1997, suggesting it would be unlikely that authors could be contacted for further information. Four authors of included studies (Sheehan *et al.*, 2012, Smith *et*

al., 2010, Szyndler *et al.*, 2005, Wong & Heriot, 2008) were contacted for further information on their reported results, for example, if correlations were available for subscales of measures. No responses were received, and this did not influence quality ratings. All included studies were rated as “moderate” quality; ratings are displayed in Table I.

Data collection process.

A data extraction tool (appendix 1.5, page 95) was developed to facilitate the process of identifying relevant information from articles.

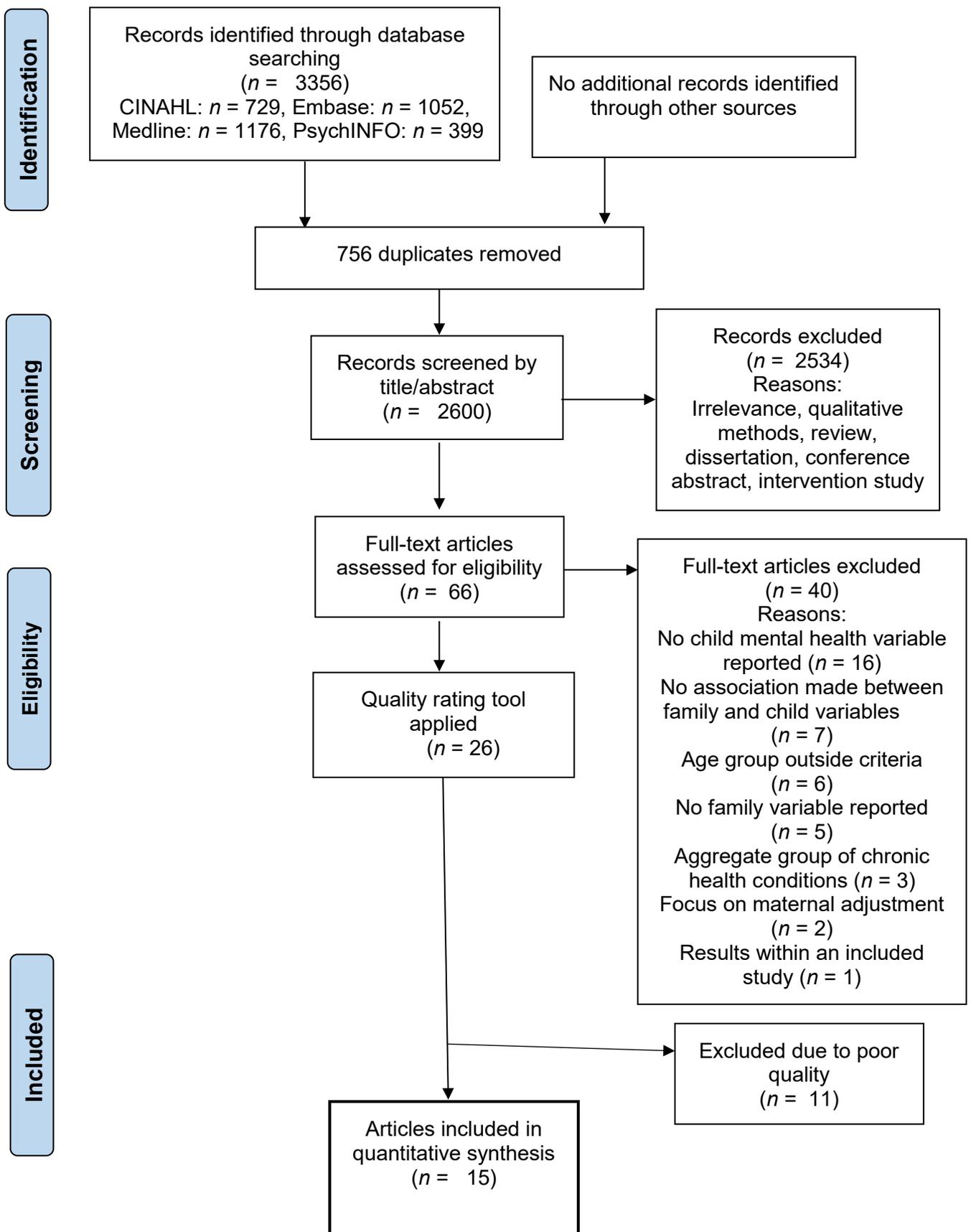


Figure I. PRISMA flow diagram (Moher *et al.*, 2009) of process for identification and inclusion of relevant studies

Results

Table I summarises included studies.

Study Characteristics

Sample.

Of the eleven studies with independent cohorts, 1740 children participated alongside 1684 family members (all parent/caregivers). Of these, four studies examined adolescents (Graetz *et al.*, 2000; Neri *et al.*, 2016; Quittner *et al.* 2014; Szyndler *et al.*, 2005), three examined a wide age-range (7-17; Cappelli *et al.*, 1988; Smith *et al.* 2010; Thompson *et al.*, 1992,1994,1999), and two reported on younger children (aged 5-12: Wong & Heriot, 2008, aged 7-13: Mullins *et al.*, 1995). The remaining studies followed up children from infancy (Goldberg *et al.*, 1997; Sheehan *et al.*, 2014).

All but two studies (Neri *et al.*, 2016, Quittner *et al.*, 2014) reported on gender of the children included in the relevant analyses ($n = 409$), with 53.3% males and 46.7% females. All studies except two included both the child and a family member (Graetz *et al.*, 2000, Szyndler *et al.*, 2005). For two studies, the gender split of the participating family members was not clear (Goldberg *et al.*, 1997, Quittner *et al.*, 2014). Three of the studies included solely mothers ($n = 112$; Cappelli *et al.*, 1988, Mullins *et al.*, 1995 and Thompson *et al.*, 1992,1994,1999). In the four remaining studies, including 404 family members; 72.8% were mothers, 27.0% were fathers and one person (0.002%) was a grandparent (Neri *et al.*, 2016, Sheehan *et al.*, 2014, Smith *et al.*, 2010 and Wong & Heriot, 2008).

Six studies reported on disease severity (Sheehan *et al.*, 2014, Smith *et al.*, 2010, Szyndler *et al.*, 2005, Thompspon *et al.*, 1992,1994,1999, Wong & Heriot, 2008). Four studies used objective medical measures such as mean forced expiratory volume in one second (Smith *et al.*, 2010, Szyndler *et al.*, 2005) and Schwachman-Kulczycki score (CF severity measure; Szyndler *et al.*, 2005, Thompson *et al.*, 1992,1994,1999). One reported on hospitalisations in the past year (Sheehan *et al.*, 2014). Two described subjective ratings of disease severity from medics (Mullins *et al.*, 1995) and family (Wong & Heriot, 2008).

One study (Quittner *et al.*, 2014) was conducted across eight European countries and the United States of America (USA). Neri *et al.* (2016) sampled Italian families in a multi-centre study. All other studies were conducted in circumscribed geographical areas in Australia (Sheehan *et al.*, 2014, Wong & Heriot, 2008, Szyndler *et al.*, 2005, Graetz *et al.*, 2000), USA (Smith *et al.*, 2010, Thompson *et al.*, 1999, Mullins *et al.*, 1995), and Canada (Goldberg *et al.*, 1997, Cappelli *et al.*, 1988).

Design of studies.

Studies followed observational, cross-sectional ($n = 7$) or prospective longitudinal designs ($n = 2$). Two studies reported on cross-sectional and longitudinal data. Recruitment was usually limited to one site, with small numbers of families sampled ($Mdn = 50.0$, $range = 24-1286$). An exception was the large international study by Quittner *et al.* (2014).

Table I: Summary of Included Articles

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
Neri et al. (2016) <ul style="list-style-type: none"> • Italy • 72% 	Observational cross-sectional survey. Explore impact of CF-related caregiving strain on occupational outcomes, and correlates of caregiver strain.	211 adolescents (aged 13-17) receiving treatment at one of 19 CF centres, and a family caregiver.	<u>Child-reported:</u> SF-12 Mental Component Summary (MCS) <u>Caregiver reported:</u> Proxy SF-12 MCS	<u>Caregiver-reported:</u> Caregiver Burden Scale	Spearman's correlation	Higher caregiver burden positively correlated with parent's perception of adolescent's mental health quality of life (QOL), but not with adolescent-reported QOL.
Quittner et al. (2014)* <ul style="list-style-type: none"> • Belgium, Germany, Italy, Spain, Sweden, Netherlands, Turkey, USA, UK • 53% 	Observational cross-sectional survey. Explore levels of anxiety and depression and their concordance across parent-adolescent dyads.	1286 adolescents (<i>mean</i> age 14.84, <i>SD</i> = 1.69) seen at one of 154 CF centres, including 1122 adolescent-dyads.	<u>Child-reported:</u> Hospital Anxiety and Depression Scale (HADS) And/or† Center for Epidemiol-	<u>Caregiver-reported:</u> HADS And/or† CES-D	Odds ratio	Adolescents 2.32 †† times more likely to report elevated symptoms of depression if at least one parent did; 2.22 times more likely for anxiety.

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
		caregiver dyads.	ogical Studies-Depression scale (CES-D)			
Sheehan <i>et al.</i> (2014)** <ul style="list-style-type: none"> • Australia • 78% 	Observational cross-sectional survey. Investigate caregiver coping strategies and association with child internalising and externalising behaviours.	101 caregivers of children (aged 3-8), being seen at one of three hospitals.	<u>Caregiver-reported:</u> Child Behaviour Checklist (CBCL)	<u>Caregiver-reported:</u> Brief COPE	Adjusted logistic regression	Proactive coping not related to CBCL. For each unit increase in avoidant coping, odds of a child experiencing clinically significant internalising behaviours increased by 1.3. No relation to externalising.
Sheehan <i>et al.</i> (2012)** <ul style="list-style-type: none"> • Australia • 56% 	Prospective cohort. Follow-up of Ward <i>et al.</i> (2009)* Determine prevalence of problem	102 families of children (aged 3-8), being seen at one of three hospitals.	<u>Caregiver-reported:</u> CBCL ($n = 73$)	<u>Caregiver-reported:</u> Questions measuring parenting warmth, consistency, overprotection from Longitudinal	Adjusted logistic regression	5.5% of children had persistent externalising/internalising behaviour over time. Low parenting warmth (only significant predictor at baseline) not a significant predictor of

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
	behaviours over time and identify predictors.			Survey of Australian Children and National Longitudinal Survey of Children and Youth. Depression Anxiety Stress Scales		internalising/externalising at three-year follow-up.
Smith <i>et al.</i> (2010) <ul style="list-style-type: none"> • USA • 67% 	Observational, cross-sectional survey. Test hypotheses that families will report heightened depressive symptoms, be less adherent to lung clearance, and relationship insecurity will be associated with	39 children (aged 7-17) being seen at one CF centre.	<u>Child-reported:</u> Children's Depression Inventory (CDI)	<u>Child-reported:</u> Relatedness Questionnaire	Independent t-test	Children whose relationships with a parent were categorised as "insecure" had significantly higher levels of depressive symptoms than children in "secure" relationships.

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
	child depressive symptoms.					
Ward <i>et al.</i> (2009)** <ul style="list-style-type: none"> • Australia • 69% 	Observational cross-sectional survey. Determine prevalence of problem behaviours and association of child variables with parenting style.	117 families of children (aged 6 months – 5 years old), being seen in one of three hospitals.	<u>Caregiver-reported:</u> CBCL ($n = 84$)	As Sheehan <i>et al.</i> (2012)	Adjusted logistic regression	Child four times as likely to experience elevated internalising/externalising if experiencing low warmth parenting. No significant relationships between externalising/internalising and inconsistency/over-protection.
Wong & Heriot (2008) <ul style="list-style-type: none"> • Australia • 61% 	Observational, cross-sectional survey. Examine vicarious hope, vicarious despair and coping strategies, and association with child variables.	34 caregivers of 35 children (aged 5-12), attending a CF support group.	<u>Caregiver-reported:</u> Child Health Questionnaire: Parent Form 50 (mental health subscale)	<u>Caregiver-reported:</u> Vicarious Futurity Scale Brief COPE	Correlations Hierarchical regression	Venting moderately correlated with better child mental health, and behavioural disengagement and self-blame moderately negative correlated. Vicarious hope strongly positively correlated with better mental health and vicarious despair strongly negative correlated.

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
						High hope, low despair and lower self-blame all significant predictors of child mental health. For each 1 unit in (improved) mental health, change of 0.94 (hope), -0.82 (despair) and -4.31 (self-blame).
Szyndler <i>et al.</i> (2005) <ul style="list-style-type: none"> • Australia • 58% 	Observational cross-sectional survey. Examine interrelationship of family functioning and psychological factors.	52 adolescents (12-18), attending one CF clinic.	<u>Child-reported:</u> Cystic Fibrosis Questionnaire (CFQ) Symptom Checklist-90-Revised (SCL-90-R)	<u>Child-reported:</u> Family Environment Scale	Spearman's correlations	Significant negative correlations between psychopathology and family cohesiveness (moderate), expressiveness (low) and organisation (moderate). No correlations with CFQ emotional functioning subscale. Moderate positive correlation of better body image with family cohesion, and moderately correlated with family conflict.
Graetz, Shute & Sawyer (2000)	Observational cross-sectional survey.	35 adolescents (aged 11-18)	<u>Child-reported:</u>	<u>Child-reported:</u>	Multiple regression	Internalising scores: total support from family and non-supportive behaviours

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
<ul style="list-style-type: none"> • Australia • 53% 	Identify perception of supportive behaviours by family and friends and associations with psychological wellbeing.	seen at one clinic.	Youth Self Report Form	Chronic Disease Support Interview		significant predictors, accounting for 15% and 24% of variance. Externalising scores: positive association with non-supportive family behaviours, accounting for 26% of variance. Total scores: non-supportive behaviours a significant predictor (25% of variance).
Thomson <i>et al.</i> (1999)*** <ul style="list-style-type: none"> • USA • 53% 	Prospective longitudinal survey. Explore stability and predictors of child adjustment.	59 children (7-17), at one CF centre, and their mothers.	<u>Child-reported:</u> Child Assessment Schedule (CAS) Self-Perception Profile for Children <u>Mother-reported:</u>	<u>Mother-reported:</u> SCL-90-R – T1: anxiety and depression subscales; T2/T3: Global Severity Index	ANOVA Hierarchical multiple regression	T1, T2 and T3: no significant difference in T1 maternal distress between children with stable good adjustment and children with stable poor adjustment. T1: mother's anxiety added significant variance to child's MCBC internalising (10%), externalising (16%), CAS total score (6%) after demographic variables and self-worth entered.

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
			Missouri Children's Behavior Checklist (MCBC; children aged ≥14)			T2: T2 maternal distress added significant variance (6%) to CAS total score after T1 CAS score entered. T3: no significant contribution of T1 maternal distress.
Goldberg et al. (1997) <ul style="list-style-type: none"> • Canada • 58% 	Prospective longitudinal survey. To examine how early internalising problems can be predicted by health and psychosocial variables.	Caregivers of 47 children aged 1 at baseline, at one CF centre.	<u>Caregiver-reported:</u> CBCL at age 4.	<u>Caregiver-reported:</u> Parenting Stress Index (PSI)	Regression	PSI best and most consistent predictor of total CBCL (13% variance at year 1, 15% at year 2, not significant at year 3) and internalising scores (9% variance at year 1, not significant at years 2/3).
Mullins et al. (1995) <ul style="list-style-type: none"> • USA 	Observational cross-sectional survey.	24 children (aged 7-13) and their 24 mothers,	<u>Child-reported:</u> CDI	<u>Caregiver-reported:</u> Brief Symptom Inventory (BSI) –	Hierarchical multiple regression	BSI did not significantly predict child depressive symptoms/state anxiety. Maternal depression predicted significant

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
<ul style="list-style-type: none"> • 56% 	Investigate patterns of adaptation and relationship between child and maternal adaptation.	attending one hospital.	State-Trait Anxiety Inventory for Children (STAIC)	depression and anxiety subscales		variance in child trait anxiety after physical health and demographic variables entered (39%). Higher depression predicted lower child anxiety.
Cappelli et al. (1988) <ul style="list-style-type: none"> • Canada • 50% 	Observational cross-sectional survey. Exploration of interaction of disease with parental/family functioning on child's psychosocial functioning.	38 children (aged 7-16) and 29 mothers, attending one CF clinic.	<u>Child-reported:</u> STAIC (aged 7-13) or State-Trait Anxiety Inventory (STAI; aged 13+) CDI Self-Perception Profile for Children <u>Mother-rated:</u>	<u>Caregiver-reported:</u> STAI Parental Bonding Instrument (PBI) Coping Health Inventory for Parents Impact on Family Scale (IFS)	Block stepwise entry multiple regression	Mothers' trait anxiety a significant predictor of behavioural problems (43.1% of variance). Overprotective parenting accounted for further significant 8.2% of variance. Coping by understanding medical situation accounted for 14.4% of athletic self-esteem. IFS predicted 21.3% of variance for physical appearance self-esteem. No other significant associations.

Article <ul style="list-style-type: none"> • Location • Quality rating 	Design & aims	Participants	Measure of child mental health	Measure of family functioning	Data analysis	Results
			CBCL	Family Functioning Index		

† Measures varied across countries; 56% used both.

†† 2.32 is reported in the abstract and 2.39 in the results.

*Besier & Goldbeck (2011) was excluded as this study presents the German results within Quittner *et al.* (2014).

**Articles from the same cohort. Ward *et al.* (2009) features baseline data from 2007; Sheehan *et al.* (2012) report on follow-up data; Sheehan *et al.* (2014) report on cross-sectional data.

*** Includes articles from the same cohort (Thompson *et al.*, 1992, 1994).

Findings

Measures of family system and parental functioning.

As participating family members were always parent/caregivers, studies are divided into those utilising measures of family system functioning, and those measuring parental functioning. This is guided by criteria by Knafl *et al.* (2015) for conceptualising family variables.

Family system functioning and mental health of children with CF.

1. General family functioning.

Two studies reported on general family functioning. Szyndler *et al.* (2005) reported significant negative correlations between adolescent-rated cohesion, expressiveness and organisation on the Family Environment Scale with an overall measure of adolescent-reported psychopathology (Symptom Checklist 90-R; SCL-90-R). Additionally, positive body image was positively correlated with cohesion and negatively correlated with conflict. Cappelli *et al.* (1988) used two measures of general family functioning; Family Functioning Index and Impact on Family Scale (IFS). IFS contributed a significant 21.3% of variance to physical appearance self-esteem, but no other associations were found between general family functioning and child mental health.

2. Family processes: coping.

Three studies reported on family coping. In the aforementioned study, Cappelli *et al.* (1988) also administered the Coping Health Inventory for Parents (CHIP). Coping by understanding the medical situation contributed a significant 14.4% of variance to athletic self-esteem. However, no other coping styles (i.e. maintaining family

integration and social support) were associated with mental health outcomes. Sheehan *et al.* (2014) administered the Brief COPE to assess proactive and avoidant caregiver coping. Child mental health was measured with the Child Behaviour Checklist (CBCL); a caregiver-reported measure of internalising and externalising behaviours. Each unit increase in avoidant coping increased the chances of a child experiencing clinically significant internalising difficulties by 1.3 times, with no effects on externalising. Wong and Heriot (2008) utilised the 14 coping style subscales of the Brief COPE, alongside the Vicarious Futurity Scale; a self-report measure of the hope/despair a parent has for their child's future. Vicarious hope, vicarious despair and self-blame were significant predictors of a mental health subscale score. Hope predicted improved mental health, while despair and self-blame predicted higher distress.

3. Family relationship quality.

Two studies reported on the quality of family relationships. Smith *et al.* (2010) used the child-reported Relatedness Questionnaire to categorise parent-child relationships as either "secure" or "insecure". Children who were characterised as insecurely attached to either parent had higher depressive symptoms, scoring marginally under the threshold for clinically-significant depression. Graetz *et al.* (2000) found that adolescents' perceptions of family supportiveness significantly predicted internalising and externalising difficulties on the Youth Self Report Form. For internalising, supportiveness accounted for 15% of variance, and non-supportiveness 25% when combined support from family and friends was accounted for. Only non-supportiveness significantly predicted variance in externalising (27% of variance) and total scores (25% of variance).

Parental functioning and mental health of children with CF.

1. Parental mental health.

Five studies reported on parental mental health. Quittner *et al.* (2014) administered parent-adolescent dyads a combination of the Hospital Anxiety and Depression Scale and Center for Epidemiological Studies Depression Scale. Adolescents were over two times as likely to report symptoms indicative of depression and anxiety if their parent also did. Thompson *et al.* (1992, 1994, 1999) examined maternal distress using the SCL-90-R. At baseline anxiety was a significant predictor of child externalising (10% of variance), internalising (16%) and child-reported mental health (6%). One year later, time 2 maternal distress similarly predicted 6% of variance in child-reported symptoms, controlling for time 1. Time 1 maternal distress did not predict variance at follow-up. Cappelli *et al.* (1988) found that maternal anxiety was a significant predictor of CBCL scores, accounting for 43.1% of the variance. Mullins *et al.* (1995) measured maternal anxiety and depression using a shortened version of the SCL-90-R. Maternal mental health did not predict child depressive symptoms or state anxiety, however, depression was a predictor of *lower* trait anxiety in the child (39% of variance). In a longitudinal study, Sheehan *et al.* (2012) found that caregiver mental health at baseline did not predict persistent internalising/externalising difficulties in children three years later. However, this calculation was likely underpowered.

2. Parenting style.

Two studies reported on parenting style. Parenting style was examined in one study using questions from national surveys (see Table I) measuring warmth, consistency and overprotection (Sheehan *et al.*, 2012, Ward *et al.*, 2009). At baseline, children

were four times more likely to experience internalising or externalising problems if they experienced parenting characterised by low warmth. No significant relationships were found with other parenting styles (Ward *et al.*, 2009). Three years later 5.5% of children continued to experience internalising or externalising difficulties, with no parenting styles significantly predicting persistent problems (Sheehan *et al.*, 2012). However, it appears that the analysis may have suffered from low power due to only four children having persistent mental health problems (of $n = 73$ with CBCL data). Cappelli *et al.* (1988) utilised the Parental Bonding Instrument to assess overprotection and care. Overprotection was a significant predictor of CBCL scores, accounting for 8.2% of the variance, with higher levels of overprotection predicting higher distress.

3. Parenting stress.

Two studies reported on parenting stress, finding mixed results. In a longitudinal study, Goldberg *et al.* (1997) measured CBCL scores at age four as an outcome. Parenting Stress Index scores at ages one and two predicted 13% and 15% of the respective variance in internalising/externalising at age four. At three years it was not significant. Neri *et al.* (2016) used the Caregiver Burden Scale, finding higher caregiver burden was positively correlated with parent-perceived mental health quality of life, but not associated with adolescent-reported.

Discussion

The primary aims of this review were to examine whether family functioning is related to the mental health of children with CF and to explore how family functioning has been measured. Across all cross-sectional studies, significant findings were reported for at least one measure of family/parental functioning and child mental health, suggesting that less adaptive general family functioning, family coping, relationship quality, parental mental health and parenting style are related to poorer outcomes for children with CF. An exception is the study by Mullins *et al.* (1995) where maternal depression predicted *lower* child anxiety. Also, mixed associations were found for parenting stress, with Neri *et al.* (2016) finding a positive association between caregiver strain and parent-reported mental health quality of life in adolescents. In studies using regression methods, family functioning variables predicted levels of variance ranging from 6% (Thompson *et al.*, 1992) to 43% (Cappelli *et al.*, 1988). These variations may result, in part, from studies utilising a broad range of measures and methodologies. Overall findings are consistent with the meta-analysis of Leeman *et al.* (2016) who found that mental wellbeing was associated with family functioning across paediatric conditions. Similarly, literature reviews by Drotar (1997) and Berge & Patterson (2004) report consistent significant associations between family functioning and mental health in children across paediatric conditions, including CF.

Conversely, in longitudinal studies parental functioning at baseline had diminishing effects on child mental health, until no longer significant. This contrasts with longitudinal studies on other health conditions, for example, Thompson *et al.* (2003) found that baseline family conflict increased the odds of having elevated CBCL

scores at nine-year follow-up in children with sickle cell disease. Consistent with this review, a meta-analysis has found that the strength of the relationship between maternal and child mental health in non-paediatric populations decreases into adolescence (Connell & Goodman, 2002). This could be related to mothers having less of an influence on children as they become older.

Included studies which compared mental health difficulties in children with CF to control groups or normative samples found elevated levels of clinically-significant symptoms in the children with CF (Goldberg *et al.*, 1997, Quittner *et al.*, 2014, Smith *et al.*, 2010). One exception is the finding by Szyndler *et al.* (2005) that adolescents had lower levels of psychopathology than community samples. Similarly, Ward *et al.* (2009) found comparable levels of mental health difficulties with normative samples, but at three-year follow-up, internalising difficulties were significantly higher in the group with CF (Sheehan *et al.*, 2012). On balance, evidence suggests that clinically-significant mental health difficulties are more common in children with CF, and therefore emphasises the importance of assessing these difficulties and providing timely psychological interventions. Furthermore, there is evidence of elevated psychological distress in adolescent-caregiver pairs (Quittner *et al.*, 2014), suggesting the value of routine screening of both individuals with CF and caregivers (Quittner *et al.*, 2016). However, research is needed to ascertain the best tools to do this (NICE, 2017). It could be helpful to extend routine screening to include measures of family/parent functioning across a range of factors and not just psychological distress. This would highlight if systemic or individual intervention is most appropriate to achieve the best outcome. Currently, there are limited evidence-based treatments for people with CF and their families. CF-specific interventions have tended to focus on treatment

adherence, with no evidence-based systemic interventions, or interventions targeted to family-caregiver wellbeing (Goldbeck *et al.*, 2014). Evidence for interventions which target family/parent factors across paediatric conditions are in early stages, but there are promising results for treatments such as problem-solving therapy (Law *et al.*, 2014). Further evidence-based treatments for those with CF and their families are greatly needed to help improve quality of life, and to indirectly improve life expectancy by reducing factors associated with poorer treatment adherence, for example, child depressive symptoms and problematic family relationships (Smith *et al.*, 2010).

Limitations of Included Studies

Family functioning was measured using a wide range of tools, however, many of the widely-used and validated tools available for measuring family functioning in paediatric populations were not utilised, for example, Family Assessment Device, Family Assessment Measure-III and Family Adaptability and Cohesion Evaluation Scale-IV (Alderfer *et al.*, 2008). Ideally, family functioning would be captured by all members of the system, although this can be a practical challenge. Most often only one family member completed the family functioning measure, biasing results towards one viewpoint. Furthermore, most often the mother participated, with smaller subsets of fathers whose data sometimes had to be excluded due to low numbers. There are also problems associated with one family member completing all measures. Parents' own wellbeing can influence their ratings of child behaviour (Treutler and Epkins, 2003). Poorer mental health could lead a parent to perceive the family as less functional. The perspective of another family member would help to test this hypothesis. Studies varied in whether measures were completed by the child, relative or both, hindering comparison across studies.

All tools used were self-reported, which brings several limitations, including socially desirable responding (van de Mortel, 2008). Observational measures may help to strengthen research designs by offering an additional more objective perspective. Several observational tools have good reliability and validity in paediatric populations (Alderfer *et al.*, 2008). However, utilising these in clinic settings, where most studies were conducted, may prove difficult.

Limitations of Review

The scope of the review was necessarily broad to allow only CF-related articles to be included. A broad age range was reviewed (0-18), which represents a heterogeneous group and CF and family factors may interact differentially across ages and developmental stages. The number of studies was limited due to the exclusion of 11 low-quality articles. Most excluded articles reported significant associations between poorer family functioning and higher child distress with the exception of two (Kucia *et al.*, 1979 reported on significant associations between better adjustment and maladaptive patterns of family functioning, and Simmons *et al.*, 1987 found no significant associations for boys). Furthermore, generalisability has been limited by the nature of the studies; most had small samples which were recruited from one clinic. With the exception of Quittner *et al.* (2014) who recruited participants across nine countries, most studies were conducted solely in North America, Canada and Australia, with one in Italy. Potentially, the interaction of family functioning and child wellbeing may differ depending on the unique challenges of living in different countries (e.g. healthcare costs). This also limits generalisability to other countries not included in these studies, for example, the United Kingdom.

Conclusions

All cross-sectional studies examining the association between family functioning variables with the mental health of children with CF have reported significant associations. In longitudinal research, family functioning variables have diminishing effects, and results for parenting stress are inconclusive. Notwithstanding, these results suggest that family functioning variables remain clinically relevant factors to consider, across a range of indices of family and parental functioning. Future research using observational measures and within larger populations may help to strengthen the evidence base and elucidate which family factors are the most potent indicators of child mental health difficulties. This will help to inform assessment of children and families, and the development of individual and systemic interventions to target problematic interactions within families.

References

*Reviewed studies

**Excluded studies

Alderfer, M.A., Fiese, B.H., Gold, J.I., Cutuli, J.J., Holmbeck, G.N., Goldbeck, L., Chambers, C.T., Abad, M., Spetter, D. & Patterson, J. 2008. Evidence-based assessment in paediatric psychology: family measures. *Journal of Pediatric Psychology*, 33, 9, 1046-1061.

Berge, J.M. & Patterson, J.M. 2004. Cystic fibrosis and the family: a review and critique of the literature. *Families, Systems, & Health*, 22, 1, 74-100.

Besier, T. & Goldbeck, L. 2011. Anxiety and depression in adolescents with CF and their caregivers. *Journal of Cystic Fibrosis*, 10, 6, 435-442.

Boutron, I., Moher, D., Altman, D.G., Schulz, K.F. & Ravaud, P. 2008. Extending the CONSORT statement to randomised trials of nonpharmacological treatment. *Annals of Internal Medicine*, 148, 295-309.

*Cappelli, M., McGrath, P.J., MacDonald, N.E., Boland, M., Fried, P. & Katsanis, J. 1988. Parent, family, and disease factors as predictors of psychosocial functioning in children with cystic fibrosis. *Canadian Journal of Behavioural Science*, 20, 4, 413-423.

**Cappelli, M., McGrath, P.J., MacDonald, N.E., Katsanis, J. & Lascelles, M. 1989. Parental care and overprotection of children with cystic fibrosis. *British Journal of Medical Psychology*, 62, 281-289.

Connell, A.M. & Goodman, S.H. 2002. The association between psychopathology in fathers versus mothers and children's internalising and externalising behavior problems: a meta-analysis. *Psychological Bulletin*, 128, 5, 746-773.

Cystic Fibrosis Trust. N.d. *Cystic fibrosis FAQs*. [online]. Available at: <https://www.cysticfibrosis.org.uk/what-is-cystic-fibrosis/faqs#How%20common%20is%20cystic%20fibrosis?> [Accessed 29 June 2018].

**Cowen, L., Corey, M., Keenan, N., Simmons, R., Arndt, E. & Levison, H. 1985. Family adaptation and psychosocial adjustment to cystic fibrosis in the preschool child. *Social Science & Medicine*, 20, 6, 553-560.

Cystic Fibrosis Trust 2004. *Pseudomonas aeruginosa infection in people with cystic fibrosis. Suggestions for prevention and infection control*. [online]. Available at: <https://www.cysticfibrosis.org.uk/~media/documents/the-work-we-do/care/consensus-docs-with-new-address/pseudomonas-aeruginosa.ashx?la=en> [Accessed 21 June 2018].

Drotar, D. 1997. Relating parent and family functioning to the psychological adjustment of children with chronic health conditions: what have we learned? What do we need to know? *Journal of Pediatric Psychology*, 22, 2, 149-165.

Elborn, J.S. 2016. Cystic fibrosis. *The Lancet*, 388, 2519-2531.

Goldbeck, L., Fidika, A., Herle, M. & Quittner, A.L. 2014. Psychological interventions for individuals with cystic fibrosis and their families. *Cochrane Database of Systematic Reviews*, issue 6, CD003148.

*Goldberg, S, Janus, M, Washington, J, Simmons, R.J., MacLusky, I. & Fowler, R.S. 1997. Prediction of preschool behavioral problems in healthy and pediatric samples. *Developmental and Behavioral Pediatrics*, 18, 5, 304-313.

*Graetz, B.W., Shute, R.H. & Sawyer, M.G. 2000. An Australian study of adolescents with cystic fibrosis: perceived supportive and nonsupportive behaviors from families and friends and psychological adjustment. *Journal of Adolescent Health*, 26, 64-69.

**Granvold, D.K., Woltman, K.A. & Hocher, M.J. 1990. Effects of parental involvement in treatment on the anxiety of children with cystic fibrosis. *Child and Adolescent Social Work*, 7, 1, 7-17.

Herzer, M., Godiwala, N., Hommel, K.A., Driscoll, K., Mitchell, M., Crosby, L.E., Piazza-Waggoner, C., Zeller, M.G. & Modi, A.C. 2010. Family functioning in the context of pediatric chronic conditions. *Journal of Developmental & Behavioral Pediatrics*, 31, 1, 26-34.

**Johnson, M.C., Muyskens, M., Bryce, M., Palmer, J. & Rodman, J. 1985. A comparison of family adaptations to having a child with cystic fibrosis. *Journal of Marital and Family Therapy*, 11, 3, 305-312.

Knafelz, K., Leeman, J., Havill, N., Crandell, J. & Sandelowski, M. 2015. Delimiting family in syntheses of research on childhood chronic conditions and family life. *Family Process*, 54, 173-184.

**Kucia, C., Drotar, D., Doershuk, C.F., Stern, R.C., Boat, T.F. & Matthews, L. 1979. Home observation of family interaction and childhood adjustment to cystic fibrosis. *Journal of Pediatric Psychology*, 4, 2, 189-195.

Law, E.F., Fisher, E., Fales, J., Noel, M. & Eccleston, C. 2014. Systematic review and meta-analysis of parent and family-based interventions for children and adolescents with chronic medical conditions. *Journal of Pediatric Psychology*, 39, 8, 866-886.

Leeman, J., Crandell, J.L., Lee, A., Bai, J., Sandelowski, M. & Knafelz, K. 2016. Family functioning and the well-being of children with chronic conditions: a meta-analysis. *Research in Nursing & Health*, 39, 229-243.

McClellan, C.B. & Cohen, L.L. 2007. Family functioning in children with chronic illness compared with healthy controls: a critical review. *The Journal of Pediatrics*, 150, 3, 221-223, 223.e1-2.

Moher, D., Liberati, A., Tetzlaff, J. & Altman, D.G. 2009. Preferred reporting items for systematic reviews and meta-analyses: the PRISMA statement. *PLoS Medicine*, 6, 7: e1000097.

*Mullins, L.L., Chaney, J.M., Hartman, V.L., Olson, R.A., Youll, L.K., Reyes, S. & Blackett, P. 1995. Child and maternal adaptation to cystic fibrosis and insulin-dependent diabetes mellitus: differential patterns across disease states. *Journal of Pediatric Psychology*, 20, 2, 173-186.

National Institute for Health and Care Excellence. 2017. *Cystic fibrosis: diagnosis and management*. NICE guidance (NG78).

*Neri, L., Lucidi, V., Catastini, P. & Colombo, C. 2016. Caregiver burden and vocational participation among parents of adolescents with CF. *Pediatric Pulmonology*, 51, 243-252.

**Pumariega, A.J., Pearson, D.A. & Seilheimer, D.K. 1993. Family and childhood adjustment in cystic fibrosis. *Journal of Child and Family Studies*, 2, 2, 109-118.

Quittner, A., L., Abbott, J., Georgiopoulos, A.M., Goldbeck, L., Smith, B., Hempstead, S.E., Marshall, B., Sabadosa, K.A., Elborn, S. 2016. International Committee on Mental Health in Cystic Fibrosis: Cystic Fibrosis Foundation and European Cystic Fibrosis Society consensus statements for screening and treating depression and anxiety. *Thorax*, 71, 26-34.

*Quittner, A. Goldbeck, L., Abbott, J., Duff, A., Lambrecht, P., Solé, A., Tibosch, M.M., Bergsten Brucefors, A., Yüksel, H., Catasini, P., Blackwell, L. & Barker, D. 2014. Prevalence of depression and anxiety in patients with cystic fibrosis and parent

caregivers: results of The International Depression Epidemiological Study across nine countries. *Thorax*, 69, 1090-1097.

Scottish Intercollegiate Guidelines Network (SIGN). 2015. *SIGN 50: a guideline developer's handbook*. Edinburgh: SIGN.

*Sheehan, J., Hiscock, H., Massie, J., Jaffe, A. & Hay, M. 2014. Caregiver coping, mental health and child problem behaviours in cystic fibrosis: a cross-sectional study. *International Study of Behavioral Medicine*, 21, 211-220.

*Sheehan, J., Massie, J., Hay, M., Jaffe, A., Glazner, J., Armstrong, D. & Hiscock, 2012. The natural history and predictors of persistent problem behaviours in cystic fibrosis: a multicentre, prospective study. *Archives of Disease in Childhood*, 97, 625-631.

**Simmons, R.J., Corey, M., Cowen, L., Keenan, N., Robertson, J. & Levison, H. 1987. Behavioral adjustment of latency age children with cystic fibrosis. *Psychosomatic Medicine*, 49, 3, 291-301.

*Smith, B.A., Modi, A.C., Quittner, A.L. & Wood, B.L. 2010. Depressive symptoms in children with cystic fibrosis and parents and its effects on adherence to airway clearance. *Pediatric Pulmonology*, 45, 756-763.

**Stawski, M., Auerbach, J.G., Barasch, M., Lerner, Y. & Zimin, R. 1997. Behavioural problems of children with chronic physical illness and their siblings. *European Child & Adolescent Psychiatry*, 6, 20-25.

**Steinhausen, H. & Schindler, H. 1981. Psychosocial adaptation in children and adolescents with cystic fibrosis. *Developmental and Behavioral Pediatrics*, 2, 3, 74-77.

**Steinhausen, H., Schindler, H. & Stephan, H. 1983. Correlates of psychopathology in sick children: an empirical model. *Journal of the American Academy of Child Psychiatry*, 22, 6, 559-564.

*Szyndler, J.E., Towns, S.J., van Asperen, P.P. & McKay, K.O. 2005. Psychological and family functioning and quality of life in adolescents with cystic fibrosis. *Journal of Cystic Fibrosis*, 4, 135-144.

*Thompson, R.J., Gustafson, K.E., Hamlett, K.W. & Spock, A. 1992. Psychological adjustment of children with cystic fibrosis: the role of child cognitive processes and maternal adjustment. *Journal of Pediatric Psychology*, 17, 6, 741-755.

*Thompson, R.J., Gustafson, K.E., George, L.K. & Spock, A. 1994. Change over a 12-month period in the psychological adjustment of children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology*, 19, 2, 189-203.

*Thompson, R.J., Gustafson, K.E., Gil, K.M., Kinney, T.R. & Spock, A. 1999. Change in the psychological adjustment of children with cystic fibrosis or sickle cell disease and their mothers. *Journal of Clinical Psychology in Medical Settings*, 6, 4, 373-392.

Thompson, R.J., Armstrong, D., Link, C.L., Pegelow, C.H., Moser, F. & Wang, W.C. 2003. A prospective study of the relationship over time of behaviour problems, intellectual functioning, and family functioning in children with sickle cell disease: a report from the Cooperative Study of Sickle Cell Disease. *The Journal of Pediatric Psychology*, 28, 1, 59-65.

Treutler, C.M. & Epkins, C.C. 2003. Are discrepancies among child, mother, and father reports on children's behaviour related to parents' psychological symptoms and aspects of parent-child relationships? *Journal of Abnormal Child Psychology*, 31, 1, 13-27.

Van de Mortel, T.F. 2008. Faking it: social desirability response bias in self-report research. *Australian Journal of Advanced Nursing*, 25, 4, 40-48.

von Elm, E., Altman, D.G., Egger, M., Pocock, S.J., Gøtzsche, P.C. & Vandenbroucke, J.P. 2007. The strengthening the reporting of observational studies in epidemiology (STROBE) statement: guidelines for reporting observational studies. *The Lancet*, 370, 1453-1457.

**Wilson, J., Fosson, A., Kanga, J.F. & D'Angelo, S.L. 1996. Homeostatic interactions: a longitudinal study of biological, psychosocial and family variables in children with cystic fibrosis. *Journal of Family Therapy*, 18, 123-139.

*Ward, C., Massie, J., Glazner, J., Sheehan, J., Canterford, L., Armstrong, D., Jaffe & Hiscock, H. 2009. Problem behaviours and parenting in preschool children with cystic fibrosis. *Archives of Diseases in Childhood*, 94, 341-347.

*Wong, M.G. & Heriot, S.A. 2008. Parents of children with cystic fibrosis: how they hope, cope and despair. *Child: care, health and development*, 34, 3, 344-354.

Ziaian, T., Sawyer, M.G., Reynolds, K.E., Carbone, J.A., Clark, J.J., Baghurst, P.A.,...French, D.J. 2006. Treatment burden and health-related quality of life of children with diabetes, cystic fibrosis and asthma. *Journal of Paediatrics and Child Health*, 42, 596-600.

Chapter 2: Major Research Project

Cystic fibrosis and family relationships: adolescent, parent and health-professional perspectives

Prepared in accordance with the author guidelines for the Journal of Pediatric Psychology (see author guidelines in appendix 1.1, page 86)

Word count (including quotes and references): 8528

Word count (excluding quotes): 6350

Plain English Summary

Title: Cystic fibrosis and family relationships: adolescent, parent and health-professional perspectives

Background

Cystic fibrosis (CF) is a chronic and life-limiting health condition affecting mainly the lungs but also several other organs in the body. Recent improvements in the treatment of CF have meant that the average life expectancy has increased, with approximately half of people with CF living to 47. However, people with CF and their caregivers experience greater levels of anxiety and depression. Therefore, it is important that people with CF and their caregivers are well supported. There are times when support may be more important; for example, during stressful times such as adolescence, a time when there are many changes and an expectation for increasing independence. However, there are few studies exploring experiences of family relationships during this time.

Aims

The aim of the present study was to explore the experiences of family relationships during adolescence from the perspective of adolescents with CF, family caregivers and health-professionals working in CF services.

Methods

Eligible participants were approached by specialist nurses in CF services in two Scottish NHS health-boards. Participants were included if they were an adolescent with CF (aged 13-18), a caregiver of an adolescent with CF or a health-professional working in a CF service. Participants were not eligible to take part if they met any of the following:

- they had a learning disability or communication difficulties
- were not fluent in English
- participation would cause excessive distress

Participants took part in an interview lasting approximately one hour. Interviews were audio-recorded, typed up and analysed for common themes in participants' experiences.

Main Findings and Conclusions

Three adolescents, two mothers and two health-professionals took part. Interviews with participants resulted in three themes, each with two sub-themes. The first theme was family coping; sub-themes explored a tendency for coping with the demands of CF by not talking about it in day-to-day life, and experiences of accessing support inside and outside the family. A second theme explored roles and boundaries in families and how these might change during adolescence. A final theme explored how adolescents and parents manage balancing the goals of the family with the teen's task of increasing independence during adolescence.

As a small number of participants took part, it is not possible to say if these themes would be found in a larger population of people. However, results suggest that it is important to understand each individual family's needs and offer extra support if required. This may be particularly important where families have little existing support to manage the challenges of CF and adolescence. It may also be helpful for teams to provide information and support on how families can have difficult conversations and manage relationship changes in adolescence.

Abstract

Objective

The aim of the present study was to explore the experiences of family relationships during adolescence from the perspective of adolescents with cystic fibrosis, family caregivers and health-professionals.

Methods

In-depth semi-structured interviews were conducted with three adolescents with cystic fibrosis (aged 15-18), two mothers and two health-professionals.

Results

Thematic analysis produced three themes, each with two sub-themes. Theme 1: family coping (using avoidant coping to manage the challenges of CF and the availability of support), theme 2: roles and boundaries (how these are adapted in families and the transition of these during adolescence), and theme 3: managing adolescence versus managing adolescents (adolescents balancing CF life with teen life and families balancing family and adolescent developmental tasks).

Conclusions

Although the sample was small, results suggest the importance that each individual family's psychological needs are understood and appropriate support offered if required. Support to initiate difficult conversations and interventions aimed at the whole family may help to manage the psychological distress that can occur during stressful times such as adolescence.

Key words: cystic fibrosis; family relationships; adolescents; caregivers; qualitative research

Introduction

Cystic fibrosis (CF) is a life-limiting and incurable genetic condition, affecting one in every 2500 births in the UK and 100,000 people worldwide (Cystic Fibrosis Trust, n.d.). Median life expectancy continues to rise and is currently 47 in the UK (Cystic Fibrosis Trust, 2017). Early detection through newborn screening, along with improvements in medical treatments have contributed to this. Additionally, future treatments are being investigated which target the genes implicated in CF and may hold promise in further improving life expectancy (Antoniou & Elston, 2016). Thus, there is growing need to meet the longer-term psychological needs of those with CF and their families.

CF treatment involves a demanding and time-consuming daily schedule (Ziaian *et al.*, 2006), which families have a role in supporting during childhood. CF can challenge the family system, with its uncertain nature and exacerbations which may require hospital admissions. Indeed, individuals with CF and their parents experience elevated levels of anxiety and depression (Quittner *et al.*, 2014). Models of family coping, such as the Family Adjustment and Adaptation Response (FAAR) Model (Patterson, 1988) have outlined the challenge of maintaining stability within the system when the family is faced with stressful situations. In this model, the family must continually adjust itself to maintain stability, and family crisis may occur where demands outstrip available resources. In chronic health conditions, demands could represent an accumulation of daily strains (e.g. treatment regimens) or an acutely stressful event such as hospitalisation. Resources are drawn from individual members (e.g. personality traits), the family, or external sources (e.g. professional support). Family resources include the adaptive functioning of the family unit; for example,

cohesion, clear family structure and clear communication (Patterson, 1988). McClellan and Cohen (2007) reviewed quantitative evidence, finding mixed results as to if and how the presence of CF affects family functioning. Despite this, the nature of family relationships is predictive of both medical and emotional outcomes. DeLambo *et al.* (2004) found that a more positive observed family environment predicted improved adherence to lung treatments as reported by both young people with CF and their parents, but not enzymes, nutrition or antibiotics. Additionally, ability to balance the needs of the family with those of the young person has been shown to predict improved lung function (Patterson *et al.*, 1993). Conversely, relationship strains between females and their parents predict poorer lung function (Patterson *et al.*, 2009). Children with CF whose caregivers have avoidant coping styles have higher chances of experiencing clinically significant emotional difficulties (Sheehan *et al.*, 2014).

For a child with CF, adolescence is a particularly complex time where they will gradually take on more of the management of their condition, while sharing responsibility with caregivers (Drotar & Ievers, 1994). Adolescence is a stressful period characterised by multiple transitions, developmental tasks such as seeking identity and autonomy, and numerous biopsychosocial developmental changes; many of which are especially difficult to negotiate while managing a chronic health condition (Williams, Holmbeck & Greenley, 2002). Research suggests a decline in CF-treatment adherence during adolescence (Bishay & Sawicki, 2016), which may place further pressure on the family. Despite adolescence being a particularly unique developmental stage, in a review of CF and the family by Berge and Patterson (2004), there were very few studies featuring a solely adolescent sample. None of these studies were qualitative, except Graetz Shute and Sawyer's (2000) mixed methods

study. Berge and Patterson (2004) argue for the addition of qualitative data to future research, and for the use of multiple perspectives when examining the complexity of family relationships. Cohen's (1999) review suggests the importance of understanding paediatric chronic health conditions within the context of the family system, with studies demonstrating that family variables are stronger predictors of psychological outcomes than disease-related factors.

To date, there has been a small pool of qualitative research exploring CF and the family. Phillips *et al.* (1985) explored the influence of CF on 43 families with a child aged up to twenty-one with a healthy sibling, from the parental perspective. They analysed responses to 62 potential problem areas in a semi-structured interview; with hospitalisation most frequently endorsed as a major problem. A third of mothers cited communication with the father as a significant difficulty. However, only 3% of fathers endorsed this. Using a grounded theory approach, Foster *et al.* (2001) explored themes within the topics of medical adherence and treatment of siblings, sampling eight children aged 10 to 18, eight siblings, eight mothers and a father. This multi-perspective study found that CF could challenge family relationships, with both children with CF and parents describing that siblings garner less family attention, leading to a perception of siblings as resentful. Furthermore, all parents described a continued role in medical management, even in late adolescence.

In a mixed methods study of 35 adolescents, Graetz *et al.* (2000) carried out a qualitative coding of adolescent-reported behaviours, finding that non-supportive behaviours from family members predicted poorer psychological adjustment.

However, they only asked one question about non-supportive behaviours and did not consider the mechanisms by which this might happen. Barker *et al.* (2011) extended this by offering qualitative descriptions of why 24 adolescents rated behaviours as unsupportive. However, Barker *et al.* (2011), Graetz *et al.* (2000) and Phillips *et al.* (1985) used qualitative methods but limited the responses participants could give. Qualitative methods which utilise in-depth semi-structured interviewing allow for a more detailed exploration of personal experiences and therefore are well suited to exploring the complexities of relational interactions (Ganong & Coleman, 2014).

Aims

The aim of the present study is to explore experiences of family relationships during adolescence from multiple perspectives; those of adolescents with CF, family caregivers, and health-professionals working alongside these families.

Methods

Design

In-depth interviewing was employed to capture rich accounts of participants' experiences. Thematic analysis was selected as the most appropriate method for analysing the dataset to provide a meaningful account of common themes across adolescents, mothers and health-professionals. Originally, it was planned that adolescents and family caregivers would be recruited, with a view to analysing data using Interpretative Phenomenological Analysis. Due to an unexpectedly high opt-out rate, a decision was made to recruit health-professionals also; details of which can be found in appendix 2.1 (page 97). As this increased heterogeneity of the sample, thematic analysis was used.

Thematic analysis is a flexible approach, not bounded by any one particular theory (Braun & Clarke, 2006). As such, this makes it a versatile approach, with Braun and Clarke (2006) suggesting that researchers should therefore specify the approach taken to the analysis. The aim of the present study was to explore participants' experiences and therefore an inductive approach was favoured to allow for themes to be identified from the data, independent of any pre-existing framework or theory. Generation of codes can be at both a semantic and latent level (Clarke & Braun, 2014), reflecting descriptive and more underlying meanings. This can help to gain an understanding of participants' experiences and the meaning attached to them.

Participants

Participants were adolescents (aged 13-18) attending CF services in either NHS Highland or NHS Greater Glasgow & Clyde (NHS GG&C), and/or a caregiver/parent, and health-professionals from these services. One member of the family could choose to participate without the other; if only a caregiver chose to participate, then it was ensured that the adolescent agreed to this. Type of caregiver was not restricted, as to allow for a variety of family structures. However, all caregivers recruited were mothers. Potential participants were deemed eligible for participation if they were able to give informed consent. To facilitate engagement in a spoken interview, only participants fluent in English were eligible. Exclusion criteria included a known learning disability, communication difficulties or acute psychological distress. In total, 23 of 25 families met eligibility criteria for participation.

Procedure

CF clinical nurse specialists provided all eligible families with a research pack including: a covering letter (appendix 2.2, page 100) and participant information sheets (PIS) (appendices 2.3 and 2.4, pages 101-106). Families sent back an opt-in form to consent to contact from the researcher or to opt-out of further contact. Health-professionals were approached to determine their interest in participating and provided with a PIS (appendix 2.5, page 107).

The researcher contacted interested individuals by email or phone to discuss the study in more detail, and then the interview was arranged if appropriate. Two

participants were interviewed in a clinic room at their respective hospital; one of whom was an inpatient. Three participants chose to be interviewed at home. Health-professionals were interviewed in clinic or by phone. Before each interview participants were informed of the limits of confidentiality and voluntary nature of their participation. The researcher answered any questions and then written consent was obtained (example consent/assent forms in appendix 2.6, pages 110-113). Consent was sought to send a letter to inform the adolescents' general practitioner of their participation. Participants were also asked if they wished to receive a summary of the study's results.

Participants took part in a semi-structured interview lasting approximately an hour. Separate interview schedules were developed for adolescent, caregiver and health-professional participants (appendix 2.7, pages 114-120) which acted as a loose guide for the interviews. If a participant became distressed, they were offered a break and reminded that they could withdraw from the interview. Interviews were audio-recorded and transcribed verbatim. Identifying information, such as names and places were removed prior to analysis to protect participant anonymity, and pseudonyms were given. Due to the small population of individuals with CF, gender references were also removed. Specific job titles are not provided to protect the anonymity of health-professionals, however, both health-professionals were currently working with at least one of the adolescents and/or mothers.

Data analysis

The iterative process of analysing the interviews was guided by steps outlined by Braun and Clarke (2006). The first stage was to become familiar with the data by

engaging with it through transcription and repeatedly re-reading the interviews. Initial ideas were noted during this process. Following this, more detailed notes were made by reading the accounts line-by-line and assigning codes reflecting the meaning of small segments of data. All relevant ideas were coded for initially. Each transcript was analysed separately and then codes were interpreted together to identify broader patterns across participants. Provisional themes related to the research question were identified and then mapped and refined to ensure that each theme reflected a clear pattern in the data, based in participants' accounts. The full data-set was then re-read to ensure that themes made sense in relation to the accounts, leading to further refinement of themes.

Two transcripts were also analysed by two research supervisors with experience in qualitative methodology, to ensure that identified themes were similar. A third research supervisor read a draft of the results section to check that their interpretation of the themes was concordant with the researcher's identified themes.

Researcher reflexivity

Braun and Clarke's (2006) version of thematic analysis acknowledges that researchers will bring their own preconceived ideas to the analytic process and that themes do not simply "emerge" but are part of an active process between the researcher and the material. This fits with a critical realism stance which acknowledges that there is not an "accurate reality" to data (Clarke & Braun, 2014). Although themes will be driven by the content of the data within an inductive approach, it is important that the researcher is aware of their own biases which may be brought to the

interpretation of the data and that these are made transparent (Braun & Clarke, 2006). The researcher is a trainee clinical psychologist working within a paediatric clinical health psychology service, with an understanding of possible psychological effects of living with a chronic health condition, alongside knowledge of developmental, attachment and systemic models. Contact with the local CF service was avoided to minimise any bias from hearing about participants. Throughout analysis, the researcher remained aware of any assumptions and kept a reflective diary following interviews.

Ethics

The study was approved by the South East Scotland 01 Research Ethics Committee and sponsored by NHS Highland Research and Development department. Board approval was also granted by NHS GG&C (appendix 2.8, page 121-129). An amendment was also approved to recruit health-professionals (appendix 2.9, pages 130-135). For participants under the age of 16, consent was obtained from both the adolescent and parent/caregiver, in line with Scottish Children's Research Network guidance (ScotCRN, 2012).

Results

Seven participants took part. Of the 23 families approached, five participants agreed to take part from four families. Two health-professionals, two mothers and three adolescents (aged 15-18) took part; one mother and one adolescent were related. Participants are listed in table I.

Table I: *Participant Roles and Pseudonyms*

Role	Pseudonyms
Adolescent with CF	Ainsley Alex Jude
Mother of adolescent with CF	Isla Morag
Health-professional working in CF team	Jools Sam

Themes

Themes are outlined in Table II.

Table II: *Themes and Sub-Themes*

Themes	Sub-themes
<ul style="list-style-type: none">• Family coping	<ul style="list-style-type: none">○ Avoidant coping: <i>We just don't talk about it and it's fine</i>○ Sources of support: <i>Each jigsaw puzzle is different...it's about helping them to create a full picture</i>
<ul style="list-style-type: none">• Roles and boundaries	<ul style="list-style-type: none">○ Adapted roles and boundaries: <i>Have we let [them] do too much?</i>○ Transition of roles and boundaries in adolescence: <i>Some do well with it, other people I guess struggle to find where they fit in that situation.</i>
<ul style="list-style-type: none">• Managing adolescence versus managing adolescents	<ul style="list-style-type: none">○ Teen life versus CF life: <i>They just want to be normal</i>○ Family versus teen struggles: <i>You're trying to do it for their own sake...but you're trying to let them grow as well</i>

Theme 1: Family Coping.

There's not a day that goes by that CF is not...on your mind. (Isla, mother, p. 18)

Since early in life, families have had to face the reality of having a child with a serious and life-limiting illness. Participants described the challenges and uncertainty of the diagnosis experience and first few years of life, providing a stressful context for early development. This will lead to families establishing ways of coping with the challenges of CF, for example, the practical burden of treatment regimes, appointments and hospitalisations, alongside the emotional impact of a progressive and life-limiting illness. By adolescence, it could be expected that families may have developed a pattern of coping with these challenges.

Sub-theme 1: Avoidant coping

We just don't talk about it and it's fine (Isla, mother, p. 28)

Commonly, participants described a tendency for avoidant patterns of coping, potentially because this helps to minimise the impact of CF-related challenges on the family.

It never really comes up. (Morag, mother, p. 36)

There may be varying functions to this avoidance, for example, maintaining a sense of normality.

You just kinda plaster a smile on your face and kinda get on with it. (Alex, adolescent, p. 13)

A lot of people when you tell them like you have a condition they automatically like have sympathy or like get worried because they think you're ill but I don't

like that because I don't want people thinking that I'm like unwell or like ill. I just want them to know I'm a normal person like everyone else. (...) I just try to act like there's nothing wrong with me. (Ainsley, adolescent, p. 12)

Participants described conversations about CF as infrequent, and when it was discussed, these tended to focus on the practical management of the illness and hopeful topics, such as cures. This suggests that the more emotional side tends to be hidden from conversation.

Like there is sometimes we do talk about it but it's not very often and if we do it's like mostly just to do with like cures and all that, so it's not really to do with me like physically or anything. (Ainsley, adolescent, p. 32/33)

Holding on to the idea that the illness could one day be cured may help the family to maintain a sense of hope for the future. Participants also spoke of the distressing emotions that the presence of CF could elicit, for example, parental guilt at transmitting the CF gene, fear or sadness about shortened life expectancy and sibling resentment at the child with CF's needs being prioritised.

They [referring to parents] don't y'know, want to sort of tell their child that almost [referring to genetic implications of CF] (...) because it's as if they've failed them in some way. (Jools, health-professional, p. 10)

Avoidance of conversations may therefore act as a protective mechanism by defending family members from distressing emotions, with some participants describing this explicitly. Isla highlighted her ambivalent position of wishing to have sensitive discussions about CF but avoiding doing so for fear of the outcome. This was a particularly emotive reflection for Isla.

I've never ever said to [them] about dying with CF, I've never spoke to [them] about that. I just can't bring myself to. But like you kind of want to say like all this medication is to help you live longer, y'know, if you don't take the medication, you're going to damage your insides even more (...) but I-I can't bring myself to start that conversation with [them], it's far too...hurtful for me.

(Isla, mother, p. 15)

Alex describes their mother as having an avoidant coping style and how this has influenced their relationship by placing distance between them.

We've just never been close because of CF (...) she said that she can't handle it. (Alex, adolescent, p. 22)

Jude also links a pattern of avoidant coping by their mother with difficulties in their relationship, culminating in its eventual breakdown.

I think it all just got too much for her and she didn't really want to like cope with everything...herself anymore. 'Cause like she did do it for a...long if you know what I mean. (...) I know it's hard to like open up and talk to other people about it, but I wish she would do that. She's not really that type of person."

(Jude, adolescent, p. 33/34)

Other more subtle forms of avoidance were apparent, e.g. providing reassurance and downplaying the severity of the illness. Again these could serve as protective strategies for managing the distress associated with the illness, however, it may preclude open discussion about CF, leaving questions unanswered. In Morag's experience, reassurances may have comforted the adolescent in the short-term, but appears not to allay the teen's fears in the longer-term as this cycle of reassurance continues. This is illustrated in Morag's description:

That's been the only time really that the CF was an issue for [adolescent] when people were saying that [you're gonna die of CF soon] to [them] and [adolescent] was 'oh what if I die?' and I'm saying 'you're not going to die, [adolescent], you're healthy' y'know. I suppose the internet doesn't help either, y'know like [they'll] send me things people have gone through a double lung transplant and y'know you'll see the photos and, 'oh mum, what if I need that?'

(Morag, mother, p. 17)

Alongside parents using this coping style, adolescents may also avoid discussions to protect themselves and others.

The child's maybe frightened to ask the parents or frightened to bring it up in case they upset the parents (...) they know that that will upset their parents so they choose not to bring it up, it's a kind of protective thing for them.

(Jools, health-professional, p. 10)

When you show you're happy but you're actually sad. It's like you've got to kinda put a mask on. (Alex, adolescent, p. 12)

Sub-theme 2: Sources of support

Each jigsaw puzzle is different...it's about helping them [the family] to create a full picture (Sam, health-professional, p. 22)

Participants spoke about the availability of support for managing the challenges of CF. Over the course of life, there could be times when families require additional support to manage living with the challenges chronic illness can bring. This might be extra important during adolescence when there are extra challenges to navigate. For some

participants, family relationships allowed for the provision of this support. Ainsley reflected on their mother's adjustment to CF, which they believed had allowed her to take on an emotionally-supportive role.

She's quite...supportive of like me anyway, but I feel like she's came to terms with the CF as well. (Ainsley, adolescent, p. 23)

Ainsley described family as a positive source of emotional support, in terms of motivating them to complete treatment. For Ainsley, professional support felt less needed.

I think that's actually what does give me the positive attitude as well 'cause I'm not just doing it for like me, I'm doing it for my family as well. (Ainsley, adolescent, p. 25)

Similarly, although Jude's relationship with their mother had broken down, they felt well-supported by other family members. Jude tended to view practical support, for example, reminders about treatment, as positive.

...it doesn't annoy me, it just shows me that [sibling] actually cares and must worry about it. (Jude, adolescent, p. 16)

For others, external professional support was sought, with mixed experiences of accessing this. Morag reported a positive experience of accessing family and external support when needed.

We do have good family support round about, and likes of [CF team-member], if there's any issues I've just got to phone [them] and [they're] on the case.
(Morag, mother, p. 7)

However, Isla described a process of seeking professional support but finding that these requests were not met.

[CF team member 1] kind of said, asked me why I was getting upset. [Exhales] I ended up just having to – I lost it – y’know, started shouting at [them] ‘ why am I getting upset? Do you think this is what I want for my child?...blah, blah, blah, I’ve got no support.’ And I ended up, I just had to walk out. Em, [CF team member 2] came out looking for me and...there was still no support put in [laughs]. (Isla, mother, p. 16)

Isla’s laughter contrasts with her sense of exasperation at the situation. Her sense of being unsupported was likely magnified by Isla’s report of difficulties accessing informal support within her own networks also, with a description that others “don’t understand” (p. 9).

Participants commonly highlighted a lack of support for the wider family, namely, siblings. Unmet needs within the wider family was also echoed by those working in CF teams, with a description that teams are becoming more aware of these gaps in service provision.

I feel like there should be like...more something like not just for CF patients but more something for their like sisters and brothers and that. (Jude, adolescent, p. 24)

Support services are directed at the patient and at the families (...) the parents, but often the siblings are the bit of the jigsaw puzzle that’s forgotten about (...). I think as a CF service and teams, we’re becoming much more aware and wakening up to the fact that maybe we’ve got young people [referring to

siblings] who are struggling with the idea that (...) it's a life-limiting illness.

(Sam, health-professional, p. 7)

Considering the previous theme about the unspoken nature of CF-related topics within the family, siblings therefore are likely to be unheard both in the family and wider network, placing them in a potentially lonely position.

I think it impacts them too...like I dunno, they don't really want to say it to you though. (Jude, adolescent, p. 15)

Theme 2: Roles and Boundaries.

Sub-theme 1: Adapted roles and boundaries.

Have we let [them] do too much? (Isla, mother, p. 5)

Participants described how CF could influence the family unit by positioning family members into adapted roles. From diagnosis, family members take on an additional role as a carer, assisting the child with intensive treatment regimes, such as physiotherapy and nebulised medications. For Morag, taking on this role resulted in her neglecting her own emotional needs and adjustment, possibly out of necessity during the early days of the illness, until these finally “hit” her (p. 44). She described a strong role as a protector, explaining that CF contributed to her relationship with her child becoming “too close” (p.49).

[Child] was in and out of hospital and they finally diagnosed [them] and everybody collapsed, but I was the strong one because I had to learn about [their] medication, about [their] physio. And then, maybe a year down the line it hit me. (Morag, mother, p. 44)

Participants spoke of a tendency for mothers to take on the carer role, attending hospital appointments and managing treatments.

My mum was always here [hospital] with me, like here when I was sick (...) every time I'd be unwell like she would be here and like every time I had an operation she'd be there for me. (Ainsley, adolescent, p. 19)

It was never [father], it was always me. (Morag, mother, p. 59)

For Jude, a sibling stepped into this role after the breakdown of their relationship with their mother.

[They're] always the one like 'are you still taking it?' like your medicine. (...) I guess [they're] just trying to be the caring one. (Jude, adolescent, p. 16)

Participants spoke about the role other family members found themselves in as the needs of the young person with CF were necessarily prioritised at times. Frequently, siblings were spoken about as being “*pushed aside*” (Jools, health-professional, p. 14), possibly leading to a sense of their needs being subjugated in the family.

My [sibling] would have to sit and watch a movie or something like that because mum would be too busy doing my medicines. (Jude, adolescent, p. 23)

Siblings may find themselves in the position of adapting their needs in order to place less strain on the family unit, as Isla highlights below.

I get on great with [sibling], [they are] just...lovely (...) but I think it's because [sibling] is, [they don't] want to give us any problems because we've enough with [adolescent]. (Isla, mother, p. 20)

Although Isla views this positively, for others, this can be problematic. In Alex's experience, their sibling not conforming to "normal" role expectations was viewed as a difficulty for them.

It's like I kind of pave the way for the path myself, and a normal teenager like they kind of pave it for their younger [sibling]. (Alex, adolescent, p. 27)

Alternatively, this could lead to a build-up of anger and resentment in the sibling which may manifest in ways which are more disruptive to relationships.

I think the siblings as I say maybe show it in more negative ways (...). I definitely think siblings sometimes get angry and resentful of the child with CF for the attention they get (...). When you actually sometimes get y'know kind of get them [parent] to think about it, it's actually because the sibling's craving attention or just wanting acknowledgement. (Jools, health-professional, p. 15/16)

Morag reports that the closeness of her and the adolescent had resulted in distance with her partner.

I would always take [child's] side y'know, and I think [father] did feel pushed out by that. (Morag, mother, p. 49)

Mothers and health-professionals described a fluidity in boundaries for the adolescent with CF, driven by a sense of parental guilt about transmitting the CF-gene.

I felt sorry for [child], I think well [they've] got an illness, leave [them] alone.

(Morag, mother, p. 50)

I feel guilty and I think...[adolescent] knows [laughs]. [Adolescent] knows how to play you as well, y'know, I mean I don't spoil [adolescent] loads but

[adolescent] does get a lot more than what a normal teenager would get because I do feel guilty. (Isla, mother, p. 19)

Mothers expressed uncertainty about their style of parenting. Isla also wondered if fear for the future contributed to the flexibility of boundaries. In the following quote, Isla's distress is exemplified by her difficulty in labelling the unspoken content of her fears.

Y'know, is [adolescent] too spoilt? (...) are we giving [them] too much because I'm scared...is [adolescent] going to be here for –do you know what I mean?
(Isla, mother, p. 5/6)

Sub-theme 2: Transitioning of roles and boundaries in adolescence.

Maybe it is a loss of a role (...) some do well with it, other people I guess struggle to find where they fit in that situation. (Sam, health-professional, p. 4)

During adolescence it would be expected that roles and boundaries start to shift as adolescents begin to develop their own sense of individuation from the family. Health-professionals described the desired shift in boundaries; a gradual change, which they would attempt to model in clinic.

We'd be trying to get the mum to become much more of a supportive role as opposed to that carer role (...) where they're coming alongside the young person. (Sam, health-professional, p. 17)

For some participants, the movement of boundaries had been more successfully negotiated. Morag had described her relationship as "too close" with the

adolescent, with her partner feeling “*pushed out*” (p. 49), but this had improved as she gradually reduced her involvement over adolescence and became more able to enforce discipline.

I've backed off from [adolescent] and let [them] do [their] own thing so maybe that's improved their relationship too because I'm not all for [adolescent] (...) whereas now I'll give [child] a row because [adolescent's] older now (...) [father] probably feels well y'know we're more on the same page. (Morag, mother, p. 58)

For Isla, movement of roles and boundaries was less straight-forward, and was possibly inhibited by family avoidance and conflict.

If any of that's [referring to treatment]...mentioned, [they] get annoyed, em, and then I start saying you know 'this is for your own good we're telling you, blah, blah, blah' and then...it just escalates into a row. (Isla, mother, p. 28)

On the other hand, Jude was “*thrown in the deep end*” (p. 39) when their mother's role as a carer suddenly changed in adolescence, leading to them attending clinics on their own at an early age. Jude expressed this statement in a quiet voice, possibly reflecting a sense of loss, shame or guilt at the situation:

Em...I don't really know why I went to clinic myself but...I guess that was like to do with like my mum or whatever I think. It's...I guess like everybody's relationship's different, but I don't really have a strong relationship with my mum. (Jude, adolescent, p. 28)

Theme 3: Managing Adolescence Versus Managing Adolescents.

You've got to live your kid's life. (Alex, adolescent, p. 7)

Participants described the difficult task of managing family developmental tasks during adolescence (balancing the adolescent's involvement in the system with spending increasing time away from the family), alongside adolescents meeting their individual developmental needs, such as seeking their own sense of identity and individuation from the family. The management of these developmental tasks during adolescence can pose particular additional challenges within the context of a chronic health condition. Participants described a sense of internal struggle for adolescents as they balance CF life with teen life. This could then be reflected in the wider system as a struggle between family and individual developmental tasks.

Sub-theme 1: CF Life versus Teen Life.

They just want to be normal. (Jools, health-professional, p. 20)

Participants described a sense of an internal struggle within the adolescent as they negotiated CF life with maintaining a normal adolescence. Jude describes focusing on the present, which could be a feature of the adolescent mind-set which tends to be more here-and-now focused. It may also be a protective strategy to avoid the consideration of possible future outcomes. Adolescents may also have many other competing priorities to consider, such as school and extra-curricular activities.

I think that I'm healthy enough right now so...that's it really. I'll just come to it once it came to it if you know what I mean. (Jude, adolescent, p. 36)

I get more stressed about school than I do about my actual like CF. (Ainsley, adolescent, p. 7)

Alex described the ambivalent position of trying to live a “normal” teenage life (p. 8) alongside CF and having to “fight” for independence (p. 27). They highlight the frustration of this experience:

It's also a bit frustrating because you don't get to do stuff with your peers. As well, I'm not allowed to do the normal teenager things like going out drinking and all that, 'cause like mixing medication's not allowed and it's frustrating, especially when you're a teenager with CF because it's like you don't get to like test certain things out. (Alex, adolescent, p. 8)

Ainsley described how their family recognised the internal struggle CF could create, and so sought to minimise this in order for them to feel “normal”. This links back to the first theme, suggesting that a possible function of avoidance of discussions is to maintain normalcy.

Talking about it all the time that makes me feel like I'm not the same as everyone else, so I think they just kind of like try to treat me like just everyone else and just like not talk about it that much. (Ainsley, adolescent, p. 32)

Sub-theme 2: Family Versus Teen Struggles.

You're trying to do it for [their] own sake...but you're trying to let [them] grow as well. (Isla, mother, p. 28)

Adolescence is a normatively stressful time for young people as they navigate various developmental tasks in the context of biopsychosocial changes and life transitions, but

CF can pose additional challenges in terms of a “*battle*” (Jools, health-professional, p. 1) for control between parents and adolescents. This could arise from the different priorities of adolescents and parents, with parents keen to maintain consistent regimes to protect their health, but teens focusing on here-and-now priorities such as friends and school.

They're only thinking about the here and now, right now, yeah, whereas we're thinking about the long-term effects, if you stop doing these treatments, that'll cause damage, then that'll impact on your life expectancy. Whereas they're thinking 'oh not right now, I don't need to worry about that, that's a long way away.' (Sam, health-professional, p. 14)

Just kinda seeing that [treatment routine] kinda almost fall apart sometimes is just too much for the parents. They want to have that control because they know that that control is almost what's kept the child well for so long. (Jools, health-professional, p. 5)

Navigating the usual developmental tasks of the individual and family is complicated by the presence of a chronic illness. At a time when adolescents are seeking increased responsibility, they also retain a heightened sense of reliance on their family as compared with healthy peers, in terms of the support required for hospital appointments and treatment. Participants described a sense of fear about family support being reduced, even in those adolescents who considered themselves independent in the management of their condition.

And [they'll] be 'oh what happens if I have to get IVs [intravenous anti-biotics] you won't be able to stay in the hospital with me' that's [their] main thing.

(Morag, mother, p. 55)

I know that my mum's not there to do everything anymore and I think that makes me more nervous. (Ainsley, adolescent, p. 30)

For Isla, there is a conflict between allowing the adolescent to become more independent and the provision of the support that she feels the young person needs. Isla describes this as a losing battle: “*you can't win*” (p. 14).

[They're] wanting to be more adult about it but [they're] not mature enough, in my opinion. (Isla, mother, p. 14)

A level of rebellion and struggling against the family would be considered normative during the adolescent period. Again, chronic illness could complicate this process, with non-adherence to treatments becoming a vehicle for rebellion.

Sometimes the young person just feels the parent is constantly nagging them or constantly on their case to do things and it is just a bit of rebellion y'know and sometimes it's the thing they know they can rebel with is their treatment because it sometimes is the thing that will upset their parents the most.

(Jools, health-professional, p. 4)

For other families, this struggle may be minimised. Where participants reported positive family relationships, reciprocation of responsibility between parent and adolescent, or a routine for illness-management; conflict tended to be minimised. Referring to treatment management, Sam gives an example of what works well:

It's just part of the, a natural kind of occurrence as opposed to it being a big event (...) it becomes part of the routine. (Sam, health-professional, p. 25)

My mum is literally like one of my best friends. (Ainsley, adolescent, p. 19)

Linking back to the first theme, avoidant coping and unspoken emotions may also intensify the struggles of adolescence.

You've kind of got an end-stage disease idea in your head but that's way out there but when your young person starts challenging and not wanting to do their treatments you worry that that time is going to come in and be shorter (...) parents can really struggle with that and something what's their fear then becomes anger and it leads to conflict. (Sam, health-professional, p. 12)

Sometimes you'll bottle them up, and sometimes you'll just kinda get on with it. But sometimes you'll have a big lash out and a screaming match. (Alex, adolescent, p. 9)

Discussion

The present study sought to understand experiences of CF and family relationships during adolescence from the perspective of adolescents, mothers and health-professionals. The interplay of family factors with chronic health conditions has been extensively studied, however, quantitative research has been limited in exploring the more complex nature of how family relationships are experienced and how this may interact with CF at different developmental stages (Berge & Patterson, 2004). The present study adds to the literature by exploring the relational experiences of adolescents with CF, mothers, and health-professionals working closely with families.

The FAAR model (Patterson, 1988) suggests that families are in a constant dynamic of trying to maintain stability in the face of challenges to the family system, including the demands of chronic illness. Furthermore, managing chronic illness during adolescence places further pressures on the family (Rolland, 1987). Participants commonly described a tendency not to discuss CF. During adolescence, it would be expected that distance between family members increases as the adolescent seeks individuation (Combrinck-Graham, 1985) and this could explain why conversations may be less frequent. However, in the descriptions given by participants, it appears that the avoidance described serves a protective function. Not discussing CF regularly may allow the adolescent and family to create a sense of normalcy and echoes previous qualitative research on CF which has found “normalisation” to be a prominent coping strategy for families during the transition period (Dupuis, Duhamel & Gendron, 2011). Avoidance of discussions appeared to be a two-way process, and findings are consistent with a small qualitative exploratory study which found that most young

adults found out about life expectancy from their doctor rather than their family, and many wished to shield their parents from the emotional distress that such discussions could cause (Farber *et al.*, 2018). Although protective, at other times this strategy may preclude open discussion around important topics, leaving questions unanswered for adolescents and family.

Dealing with the stress of a chronic illness meant that family members were required to adapt their normal roles, for example, siblings were described as missing out. In their grounded theory study, Foster *et al.* (2001) similarly reported on a perception that siblings experience resentment at the needs of children with CF appearing prioritised. In the current study, participants described a tendency for mothers to take on a carer role, for example, in accompanying the child to appointments and administering treatments. Indeed, Hodginson & Lester (2002) found that all of seventeen mothers interviewed about their experiences of caring for a younger child with CF experienced their role as stressful, including the heightened sense of responsibility that they held. Berge and Holm (2007) posit that for families of children with a chronic condition, the uncertainty and challenges this poses can lead to “boundary ambiguity” where there is uncertainty around roles within the family; for example, uncertain expectations for the child’s behaviour. Ambiguity may place extra strain on navigating role and boundary changes during adolescence. Where caregivers experience uncertainty and stress, this can reduce personal and family resources to manage the challenges of stressful times. Also, if a parent is experiencing distress, then it is more likely the adolescent will too (Quittner *et al.*, 2014).

Participants described the challenges of adolescence, in terms of negotiating the balance between adolescents completing their individual developmental tasks, alongside family developmental tasks. Transition of CF care is a complex process influenced by many factors, including parental, child and family factors (Leeman *et al.*, 2015). Difficulties might arise where there is a mismatch between the family conferring responsibility to the adolescent, and the teen's ability or willingness to take on greater responsibility. For some families, where this balance is negotiated, additional difficulties may be minimised. However, for others this could increase the burden of CF, causing a struggle between parent and adolescent. Parent-teen interactions have been listed by adolescents with CF and their caregivers as one of their top three most problematic situations in a structured interview (DiGirolamo *et al.*, 1997). It is important that conflict in relationships with caregivers is minimised as far as possible. Family difficulties, for example relationship strains, can have negative implications for treatment adherence and physical health (DeLambo *et al.*, 2004, Patterson *et al.*, 1993, 2009). As such, it is of great importance that family needs are monitored, and appropriate support offered to proactively reduce stress, prevent ruptures in the family and indirectly influence physical health outcomes. Assessing family needs could be challenging at a time when adolescents are encouraged to attend clinics on their own and suggests that it may be important for parents and teens to have time alone to discuss any difficulties. Indeed, this is particularly important considering results that adolescents and parents may tend not to discuss CF, sometimes due to the distress it may cause to themselves or others.

Strengths and Limitations

A strength of this study was the attempt to gather multiple perspectives regarding family experiences. However, a major limitation of this study was the small sample size, despite efforts to recruit a larger sample. Furthermore, participants were a heterogeneous group, with varying roles, family backgrounds and structures. As such, participants are therefore not necessarily representative of the wider CF population. With a small sample size, it is possible that data saturation was not reached and that the themes may not be replicated in a larger sample. However, participants shared very emotive experiences on topics such as family breakdown and the life-limiting nature of CF which provided in-depth accounts.

Clinical Implications

Adolescence is a particularly complex time, especially in the context of a chronic and life-limiting health condition. Although the results are limited in their generalisability, they suggest that supporting the family during this time is of great importance, where needed. Some participants spoke of a difficulty in initiating CF-related conversations. Support in clinic could be augmented with the provision of resources for parents and adolescents to consider how they might approach difficult conversations. Furthermore, this highlights the importance of both adolescents, their carers and siblings having a safe space to explore emotions. Currently, there is a lack of evidence-based interventions to provide support for people with CF and their families (Goldbeck *et al.*, 2014) and so new treatments are greatly needed. It may be particularly important for families to access professional support if resources are lacking within their existing networks; potential service development could include the provision of support groups

for caregivers and siblings; or information on maintaining relationships during adolescence. NICE (2017) guidelines advise that the emotional needs of the individual and family should be assessed at critical time points such as transition. The use of screening tools may help to quickly identify families who are experiencing difficulties at an early stage.

Future Research

Due to the small sample size, future research could replicate the study with a larger sample size to ascertain if the same themes are present in a more representative group. As recruitment of participants was difficult, it would be important to consider how to engage families in such research. Furthermore, carrying out interviews with other family members, for example, fathers and siblings, could provide more comprehensive data on experiences of relationships within a system.

Conclusion

The present study sought to explore experiences of CF and family relationships during adolescence from multiple perspectives. It is important that each individual's and their family's psychological needs are understood, and appropriate support offered, to promote well-being and reduce psychological distress. This may then positively influence treatment adherence and ultimately improve physical health.

References

- Antoniou, S. & Elston, C. 2016. Cystic fibrosis. *Medicine*, 44, 5, 321-325.
- Barker, D.H., Driscoll, K.A., Modi, A.C., Light, M.J. & Quittner, A.L. (2011). Supporting cystic fibrosis disease management during adolescence: the role of family and friends. *Child: care, health and development*, 38, 4, 497-504.
- Berge, J.M. & Holm, K.E. 2007. Boundary ambiguity in parents with chronically ill children: integrating theory and research. *Family Relations*, 56, 123-134.
- Berge, J.M. & Patterson, J.M. 2004. Cystic fibrosis and the family: a review and critique of the literature. *Families, Systems & Health*, 22,1, 74-100.
- Bishay, L.C. & Sawicki, G.S. 2016. Strategies to optimize treatment adherence in adolescent patients with cystic fibrosis. *Adolescent Health, Medicine and Therapeutics*, 7, 117-124.
- Braun, V. & Clarke, V. 2006. Using thematic analysis in psychology. *Qualitative Research*, 3, 2, 77-101.

Clarke, V. & Braun, V. 2014. 'Thematic analysis', in Teo, T. (ed.), *Encyclopedia of Critical Psychology*. New York: Springer, pp. 1947-1952.

Combrinck-Graham, L. 1985. A developmental model for family systems. *Family Process*, 24, 139-150.

Cohen, M.S. 1999. Families coping with childhood chronic illness: a research review. *Families, Systems & Health*, 17, 149-164.

Cystic Fibrosis Trust. N.d. *Cystic fibrosis FAQs*. [online] Available at: <https://www.cysticfibrosis.org.uk/what-is-cystic-fibrosis/faqs#How%20common%20is%20cystic%20fibrosis?> [Accessed 1 July 2018].

Cystic Fibrosis Trust. 2017. *Cystic fibrosis: strength in numbers. UK Cystic Fibrosis Registry 2016 Annual Data Report*. [online]. Available at: https://www.cysticfibrosis.org.uk/~/_media/documents/the-work-we-do/uk-cf-registry/uk-cf-registry-annual-data-report-2016.ashx?la=en [Accessed 1 July 2018].

DeLambo, K.E., Ievers-Landis, C.E., Drotar, D. & Quittner, A.L. 2004. Association of observed family relationship quality and problem-solving skills with treatment

adherence in older children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology, 29, 5, 343-353.*

DiGirolamo, A.M., Quittner, A.L., Ackerman, V. & Stevens, J. 1997. Identification and assessment of ongoing stressors in adolescents with a chronic illness: an application of the behavior-analytic model. *Journal of Clinical Child Psychology, 26, 1, 53-66.*

Drotar, D. & levers, C. 1994. Age differences in parent and child responsibilities for management of cystic fibrosis and insulin-dependent diabetes mellitus. *Developmental and Behavioral Pediatrics, 15, 4, 265-272.*

Dupuis, F., Duhamel, F. & Gendron, S. 2011. Transitioning care of an adolescent with cystic fibrosis: development of systemic hypothesis between parents, adolescents and health care professionals. *Journal of Family Nursing, 17, 3, 291-311.*

Farber, J.G., Prieur, M.G., Roach, C., Shay, R., Walter, M., Borowitz, D. & Dellon, E.P. 2018. Difficult conversations: discussing prognosis with children with cystic fibrosis. *Pediatric Pulmonology, 53, 592-598.*

Foster, C., Eiser, C., Oades, P., Sheldon, C., Tripp, J., Goldman, P., Rice, S. & Trott. 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.

Ganong, L. & Coleman, M. 2014. Qualitative research on family relationships. *Journal of Social and Personal Relationships*, 31, 4, 451-459.

Goldbeck, L., Fidika, A., Herle, M. & Quittner, A.L. 2014. Psychological interventions for individuals with cystic fibrosis and their families. *Cochrane Database of Systematic Reviews*, issue 6, CD003148.

Graetz, B., Shute, R.H. & Sawyer, M.G. 2000. An Australian study of adolescents with cystic fibrosis: perceived supportive and nonsupportive behaviors from families and friends and psychological adjustment. *Journal of Adolescent Health*, 26, 1, 64-69.

Hodgkinson, R. & Lester, H. 2002. Stresses and coping strategies of mothers living with a child with cystic fibrosis: implications for nursing professionals. *Journal of Advanced Nursing*, 39, 4, 377-383.

Leeman, J., Sandelowski, M., Havill, N.L. & Knaf, K. 2015. Parent-to-child transition in managing cystic fibrosis: a research synthesis. *Journal of Family Theory & Review*, 7, 167-183.

McClellan, C. B. & Cohen, L. L. 2007. Family functioning in children with chronic illness compared with healthy controls: a critical review. *Journal of Pediatrics*, 150, 221-223.

National Institute for Health and Care Excellence. 2017. *Cystic fibrosis: diagnosis and management*. NICE guidance (NG78).

Patterson, J. M. 1988. Families experiencing stress. I. The Family Adjustment and Adaptation Response Model. II. Applying the FAAR model to health-related issues for intervention and research. *Family Systems Medicine*, 6, 2, 202-237.

Patterson, J.M., Goetz, G., Budd, J. & Warwick, W.J. 1993. Family correlates of a 10-year pulmonary health trend in cystic fibrosis. *Pediatrics*, 91, 383-389.

Patterson, J. M., Wall, M., Berge, J. & Milla, C. 2009. Associations of psychosocial factors with health outcomes among youth with cystic fibrosis. *Pediatric Pulmonology*, 44, 46-53.

Phillips, S., Bohannon, W. E., Gayton, W. F. & Friedman, S. B. 1985. Parent interview findings regarding the impact of cystic fibrosis on families. *Journal of Developmental and Behavioral Pediatrics*, 6, 122-7.

Quittner, A. Goldbeck, L., Abbott, J., Duff, A., Lambrecht, P., Solé, A., Tibosch, M.M., Bergsten Brucefors, A., Yüksel, H., Catasini, P., Blackwell, L. & Barker, D. 2014.

Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers: results of The International Depression Epidemiological Study across nine countries. *Thorax*, 69, 1090-1097.

Rolland, J.S. 1987. Chronic illness and the life cycle: a conceptual framework. *Family Process*, 26, 203-221.

ScotCRN, 2012. *Obtaining informed consent for clinical research in children and young people under 16 in Scotland*. [online]. Available at: http://www.scotcrn.org/documents/ScotCRN_Guidance_Documents/ScotCRN_informed_consent_7.08.12_V1.pdf. [Accessed: 2 June 2018].

Sheehan, J., Hiscock, H., Massie, J., Jaffe, A. & Hay, M. 2014. Caregiver coping, mental health and child problem behaviours in cystic fibrosis: a cross-sectional study. *International Study of Behavioral Medicine*, 21, 211-220.

Williams, P. G., Holmbeck, G. N. & Greenley, R. N. 2002. Adolescent health psychology. *Journal of Consulting and Clinical Psychology*, 70, 828-42.

Ziaian, T., Sawyer, M. G., Reynolds, K. E., Carbone, J. A., Clark, J. J., Baghurst, P. A., Couper, J. J., Kennedy, D., Martin, A. J., Staugas, R. E. & French, D. J. 2006.

Treatment burden and health-related quality of life of children with diabetes, cystic fibrosis and asthma. *Journal of Paediatrics and Child Health*, 42, 596-600.

Appendices: Chapter 1

Appendix 1.1: Author guidelines for the Journal of Pediatric Psychology

Organization of manuscripts

Manuscript Central will guide authors through the submission steps, including: Abstract, Keyword selection, and the Manuscript. The manuscript must contain an Introduction, Methods, Results, Discussion, Acknowledgements and Reference List.

Length of manuscript: Original research articles should not exceed 25 pages, in total, including title page, references, figures, tables, etc. In the case of papers that report on multiple studies or those with methodologies that necessitate detailed explanation, the authors should justify longer manuscript length to the Editor in the cover letter. Case reports should not exceed 20 pages. Review articles should not exceed 30 pages. Invited commentaries should be discussed with the Editor. The *Journal of Pediatric Psychology* no longer accepts brief reports but will accept manuscripts that are shorter in length than the 25 page manuscript.

Manuscripts (text, references, tables, figures, etc.) should be prepared in detailed accord with the Publication Manual of the American Psychological Association (6th ed.). There are two exceptions:

The academic degrees of authors should be placed on the title page following their names, and a structured abstract of not more than 250 words should be included.

The abstract should include the following parts:

1. Objective (brief statement of the purpose of the study);
2. Methods (summary of the participants, design, measures, procedure);
3. Results (the primary findings of this work); and
4. Conclusions (statement of implications of these data).

Key words should be included, consistent with APA style. Submissions should be double-spaced throughout, with margins of at least 1 inch and font size of 12 points (or 26 lines per page, 12-15 characters per inch). Authors should remove all

identifying information from the body of the manuscript so that peer reviewers will be unable to recognize the authors and their affiliations.

Informed consent and ethical treatment of study participants: Authors should indicate in the Method section of relevant manuscripts how informed consent was obtained and report the approval of the study by the appropriate Institutional Review Board(s). Authors will also be asked to sign a statement, provided by the Editor that they have complied with the American Psychological Association Ethical Principles with regard to the treatment of their sample.

Clinical relevance of the research should be incorporated into the manuscripts. There is no special section on clinical implications, but authors should integrate implications for practice, as appropriate, into papers.

Terminology should be sensitive to the individual who has a disease or disability. The Editors endorse the concept of "people first, not their disability." Terminology should reflect the "person with a disability" (e.g., children with diabetes, persons with HIV infection, families of children with cancer) rather than the condition as an adjective (e.g., diabetic children, HIV patients, cancer families). Nonsexist language should be used.

Special Instructions for Types of Manuscripts

- **Systematic reviews**

(b) Systematic reviews: Systematic reviews should not exceed 30 pages. Authors are required to attach the PRISMA checklist and flow diagram as supplementary material for each submission. Authors can find the PRISMA checklist and flow diagram in downloadable templates that can be re-used [here](#). Authors of systematic reviews that do not include a meta-analysis must provide a clear statement in the manuscript explaining why such an analysis is not included for all or relevant portions of the report.

Appendix 1.2: Conceptualisation of Family Functioning

Knafl *et al.* (2015) provide a framework for conceptualising family variables in research on paediatric health conditions within families. The authors distinguish between family system functioning variables (defined as family relationships, structure, processes, characteristics and resources) and family member functioning (defined as wellbeing, experiences and ability to carry out role, e.g. parenting style and stress). For the purposes of this review, family and parental functioning was separated according to these criteria:

Study	Family variable measured	Knafl <i>et al.</i> (2015) category
Neri <i>et al.</i> (2016)	Caregiver burden	Parental functioning (performance of parenting role)
Quittner <i>et al.</i> (2014)	Parental depression and anxiety	Parental functioning (wellbeing)
Sheehan <i>et al.</i> (2014)	Coping	Family system functioning (family processes, e.g. coping)
Sheehan <i>et al.</i> (2012)	Parenting style	Parental functioning (performance of parenting role and wellbeing)
Smith <i>et al.</i> (2010)	Security of relationships	Family system functioning (family relationships)
Ward <i>et al.</i> (2009)	Parenting style	Parental functioning (performance of parenting role)

Wong & Heriot (2008)	Coping	Family system functioning (family processes, e.g. coping)
Szyndler <i>et al.</i> (2005)	Family environment	Family system functioning (family system characteristics)
Graetz <i>et al.</i> (2000)	Perceived supportiveness	Family system functioning (family resources)
Thompson <i>et al.</i> (1992, 1994, 1999)	Maternal mental health	Parental functioning (wellbeing)
Goldberg <i>et al.</i> (1997)	Parenting stress	Parental functioning (performance of parenting role)
Mullins <i>et al.</i> (1995)	Maternal mental health	Parental functioning (wellbeing)
Cappelli <i>et al.</i> (1988)	Maternal anxiety	Parental functioning (wellbeing)
	Relationship quality Coping Impact on family	Family system functioning (family relationships, processes and resources)

Appendix 1.3: Search strategy

Database	Search terms used
CINAHL Medline PsychoINFO	<p>Major subject heading (MM) cystic fibrosis OR cystic fibrosis or CF</p> <p>AND</p> <p>MM relating to parents, family, caregivers OR mother* or father* or brother* or sister* or carer* of caregiver* or care giver* or parent* or family</p> <p>AND</p> <p>Subjects (DE): adjustment or DE emotional adjustment OR MM relating to emotions OR adjustment or adaptation or well#being or mental health or depression or anxiety or internali?ing or externali?ing or emotion* or psychopathology or resilience OR MM relating to psychopathology/adolescent psychopathology/child psychopathology</p> <p>AND</p> <p>Mm relating to child* or adolesc* or p#ediatric or youth or young person* or young people or teen* or infant*</p>
EMBASE	<p>Exp cystic fibrosis OR (cystic fibrosis or CF).mp</p> <p>AND</p> <p>Exp parent or exp family or EXP caregiver or (mother* or father* or</p>

	<p>brother* or sister* or carer* or caregiver* or care giver* or parent* or famil*).mp</p> <p>AND</p> <p>Exp psychological adjustment or exp emotion or exp psychological resilience or mental health or (adjustment or adaptation or well?being or mental health or depression or anxiety or internal#ing or external#ing or emotion* or psychopathology or resilience).mp</p> <p>AND</p> <p>Exp juvenile or (child* or adolesc* or p?ediatric or youth or young person* or young people or teen* or infant*).mp</p>
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Appendix 1.4: Quality rating tool

Abstract	The abstract provides a summary of the study including methods and results.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
Introduction	Provides scientific background and justification for study.	2- Well covered and justified 1 – Adequately covered and justified 0 – Not/poorly covered and justified
	Provides objectives/hypotheses.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
Methods • Design	Clearly states study design.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
• Participants	Justification for sample size.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
	Inclusion/exclusion criteria stated.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered

	Settings and locations and dates for data collection outlined.	2 – Well covered 1 – Adequately/partly covered 0 – Poorly/not covered
	Participant characteristics clearly described and representative of target population.	2 – Well covered and considered representative 1 – Covered but not considered representative/partly covered 0 – Not covered
• Outcomes	All of the outcomes are clearly described.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
	Measure of child/adolescent mental health is appropriate.	2- Standardised tools with reported validity/reliability 1 – Standardised tools but no report of validity/reliability or low validity/reliability 0 – Non-standardised assessment
	Measure of family functioning is appropriate.	2- Standardised tools with reported validity/reliability 1 – Standardised tools but no report of validity/reliability or low validity/reliability 0 – Non-standardised assessment
	Confounding variables have been reported and taken into account.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
Statistical analysis	Numbers given for each group analysed and non-participation described.	2 – Well covered 1 – Adequately covered

		0 – Poorly/not covered
	Statistical methods are appropriate.	2- Appropriate 1 – Adequate 0 – Not appropriate
	Findings are clearly described.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
	Where appropriate confidence intervals are reported.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
• Discussion	Limitations of study are clearly described, taking into account potential sources of bias.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered
	Generalisability of study is clearly described.	2 – Well covered 1 – Adequately covered 0 – Poorly/not covered

Score out of 36 =

% =

Appendix 1.5: Data extraction tool

Study authors:
Study characteristics
Study: Type of study: When conducted: Country: Setting:
Study aims
Primary aims: Secondary aims:
Participant characteristics
Parent participants/child participants/both <u>Child participants</u> Inclusion criteria: Sample size: Age range: Mean age: Gender: Disease severity: Length of diagnosis/age of diagnosis: Other: <u>Parent participants</u> Inclusion criteria: Sample size: Age range: Mean age: Gender: Other:

Child measure
Family measure
Statistical analysis
Study results
Notes

Appendices: Chapter 2

Appendix 2.10: Change in design and difficulties with recruitment

Change in planned design

In the original proposal (see appendix 2.10, p. 139), the planned project had been to recruit family dyads (an adolescent and a parent/caregiver from each participating family) and for accounts to be analysed using Interpretative Phenomenological Analysis (IPA). Due to low uptake by families (see below), a decision was made to make an amendment to recruit health-professionals from the CF services involved in recruitment. IPA relies on gaining rich accounts of small, homogenous groups (Smith, Flowers & Larkin, 2009). Multi-perspective IPA studies are few in number but growing, for example, Rostill-Brookes *et al.* (2011) interviewed foster children, foster carers and social workers on their experiences of foster placement breakdown, providing three shared themes. However, the limited word count of a professional doctoral thesis precluded a more detailed presentation of the analysis, and so data was re-analysed using thematic analysis, which is a more versatile approach with less focus on rich accounts.

Difficulties with recruitment

Recruitment began in January 2018. CF clinical nurse specialists (CNS) issued research packs to all eligible families in NHS Highland and NHS Greater Glasgow & Clyde ($n = 23$). These were most frequently provided to potential participants at clinic appointments, with some being posted to families. There was a slow response from families, with the initial few respondents opting-out of further contact from the researcher. Although the researcher had planned to contact any families who had not opted-out within six weeks of receiving the research pack, it was decided that the CNS

would make further contact. This was deemed appropriate due to the longstanding relationship that the CNS have with the families. All families were approached by nurses on at least one further occasion to gauge their interest in the research. By May 2018, all families had either participated or opted-out.

Amendment

As the group of potential participants had been exhausted an amendment was submitted in May 2018 to include the views of health-professionals within the research. Due to the slow uptake rate for families, it was thought that approaching another health-board would present the same issues.

Reflections on recruitment

On reflection, it may have been helpful to start the recruitment process earlier. This may have identified recruitment difficulties at an earlier stage and therefore allowed other plans to be put in place.

From discussions with the CF CNS prior to recruitment, it was thought that the minimum numbers would be achieved and so the uptake rate was unexpectedly low. This was discussed with the CF CNS who felt that the timing of the research had been problematic as it coincided with exam periods in winter and spring 2018. One CNS described that adolescents had a general lack of interest in research, and that it had been difficult to recruit to a patient wellbeing survey ongoing on at the same time. Those in good health were thought to be especially focused on their exams, while those with poorer health tended to have difficulties attending appointments. Another CNS was surprised by the low uptake rate, and suggested that this may have been due to the sensitive nature of the research, or that there were fewer perceived benefits from taking part compared with other research, i.e. drug trials.

References

Rostill-Brookes, H., Larkin, M. Toms, A. & Churchman, C. (2011). A shared experience of fragmentation: making sense of foster placement breakdown. *Clinical Child Psychology and Psychiatry*, 16, 1, 103-127.

Smith, J.A, Flowers, P. & Larkin, M. 2009. *Interpretative Phenomenological Analysis: Theory, Method and Research*. London: SAGE Publications Ltd.

Appendix 2.2: Invitation letter for potential participants (V3, 27/10/17)



Experiences of family relationships and managing cystic fibrosis in adolescence

Dear <name>

I am writing to let you know about some research that is being carried out by a final year Trainee Clinical Psychologist, Frieda Whelan. Frieda is completing the research as part of her doctoral degree at the University of Glasgow.

Frieda is interested in the experiences of young people (aged 13-16) and their parents/caregivers as they grow older with cystic fibrosis. Please find enclosed participant information sheets (one for teenagers and one for caregivers) which describes the study and what will happen should you decide to take part. Please take your time to read through this and feel free to discuss with friends and family, or to get in touch with the research team. There are contact details for the team at the end of the participant information sheet.

If you decide you would like to take part, please return the enclosed opt-in form to Frieda in the envelope provided. She will then contact you to discuss it in more detail.

Please note that I am independent of the research team. If you decide not to take part it will have no impact on the service that you receive from the Cystic Fibrosis team.

Thank you for taking the time to read this letter and to consider the research project.

Yours sincerely,

[CF nurse]
Cystic Fibrosis Service

Enclosed:
Participant information sheet for teenagers
Participant information sheet for caregivers
Opt-in form
Envelope to return opt-in form

Appendix 2.3: Participant information sheet for caregivers (V3, 20/10/17)



Experiences of family relationships and managing cystic fibrosis in adolescence

Researcher: Frieda Whelan

Participant information sheet for parents/caregivers

This information sheet describes a study which is being carried out about teenagers and their experiences of growing up with cystic fibrosis (CF) and their relationships with family members. We are also interested in the views of the parents/caregivers.

We would like to invite you to take part in the study, but first we would like to let you know about why the study is being done and what it would involve if you took part. Please read the information sheet carefully and you may wish to discuss it with someone else. If you have any questions please contact the researcher, Frieda Whelan, whose details are at the end. Thank you for reading this and for considering taking part in this study.

1. Why is the study being done?

The study aims to find out about the experiences of teenagers with CF. The aim is find out how teenagers find growing up with CF and about how their relationships with family affects this and vice versa. We are also interested in the views of the parent/caregiver. We know that growing up can be a difficult time, particularly with a health condition. Therefore, we are interested in how people might use family support at this time.

2. Why have I been invited to take part?

You are being asked to take part because you are the parent or caregiver of a teenager with CF being seen by the CF team in either NHS Highland or NHS Greater Glasgow & Clyde. The study hopes to recruit up to 10 people.

3. Do we have to take part?

No, it is up to you. If you decide to take part we will ask you to keep the information sheet and to sign a consent form. If you take part, you can stop taking part at any time. You will not need to give a reason and it will not affect the medical care of the teenager taking part. It will also not affect his/her medical care should you not wish to take part.

If you would like to take part, your teenager can decide whether they would like to take part also. However, if you would like to take part but the teenager does not, we will check with him/her that they agree to your participation.

4. What happens if we/I decide to take part?

If you decide to take part you will be asked to take part in one interview with the researcher (Frieda Whelan, who is a Trainee Clinical Psychologist). You will be interviewed alone, even if your family member also takes part. The interview will last up to an hour and will take place at a local hospital. The interview will be audio recorded and typed up and any details which could identify you will be removed. The interview will ask questions about having a child with CF. It will also ask about family relationships and how these might have changed as the teenager has gotten older.

5. Will the study help me/us?

There are no direct benefits to taking part. However, it could help the CF teams to understand more about how best to offer support to young people and their families.

6. Are there any disadvantages about taking part?

You might talk about experiences that cause you to feel some distress. You can stop the interview at any time and you can have a break whenever you wish. The researcher will check how you are feeling during the interview. You can stop the interview at any time and this will have no impact on your teenager's medical care.

Unfortunately we are not able to provide you with any expenses for travelling to the interview.

7. Will my information be kept safe?

Yes, all information will be kept safe and secure. Once the audio recording of the interview is typed up it will be destroyed. Any information which could identify you (for example, names or place names) will be removed as it is typed up. The typed-up interview will be given a study number so that your name is not on it. Some quotes might be used from your interview. If quotes are used, a pseudonym will be used and not yours.

All information will be kept confidential. However, there may be times when information needs to be discussed with other professionals, for example, if the researcher is concerned about any risks to yourself or others. Frieda will try to discuss this with you if it is necessary.

8. What will happen to the results of the study?

Once the research is finished, the researcher (Frieda Whelan) will write up the project and submit it to the University of Glasgow as part of the Doctorate of Clinical Psychology degree. A report might also be submitted for publication in a journal.

9. Further information and contact details

If you have any questions or would like more information please contact one of the research team below:

Name	Role	Contact details
Frieda Whelan	Trainee Clinical Psychologist, Chief Investigator	Phoenix Centre, Raigmore Hospital, Old Perth Road, Inverness IV2 3UJ 01463 705597 f.Whelan@research.gla.ac.uk
Dr Alison Jackson	Academic Supervisor	Institute of Health and Wellbeing, University of Glasgow, 1st floor, Administration Building, Gartnavel Royal Hospital, 1055 Great Western Road, Glasgow G12 0XH 0141 211 3917

10. Complaints

If you have a complaint about any aspect of the research, please contact your health board below:

NHS Highland	The Feedback Team, NHS Highland, PO Box 5713, Inverness IV1 9AQ nhshighland.feedback@nhs.net 01463 705997
NHS Greater Glasgow & Clyde	Complaints Department, West Glasgow Ambulatory Care Hospital, Dalnair Street, Glasgow G3 8SJ complaints@ggc.scot.nhs.uk 0141 201 4500

Appendix 2.4: Participant information sheet for teenagers (V1, 20/10/17)



Experiences of family relationships and managing cystic fibrosis in adolescence

Researcher: Frieda Whelan

Participant information sheet for teenagers

This information sheet describes a study which is being carried out about teenagers and their experiences of growing up with cystic fibrosis (CF) and their relationships with family members. We are also interested in the views of parents/caregivers.

We would like to invite you to take part in the study, but first we would like to let you know about why the study is being done and what it would involve if you took part.

Please read this information sheet carefully and you might want to talk about it with someone you know. If you have any questions please contact the researcher, Frieda Whelan, using the contact details at the end.

Thank you for reading this and for thinking about taking part in the study.

1. Why is the study being done?

The study aims to find out about the experiences of teenagers with CF. We hope to find out about how teenagers find growing up with CF and about how their relationships with family affects this and vice versa. We are also interested in the views of your family, so we may ask your parent or caregiver to take part too. We know that growing up can be a difficult time, particularly with a health condition. Therefore, we are interested in how people might use family support at this time.

2. Why have I been invited to take part?

You are being asked to take part because you are aged 13-18, have CF and see the CF service in either NHS Highland or NHS Greater Glasgow & Clyde.

We hope that up to ten people will take part in the study.

3. Do I have to take part?

No, it is up to you. If you decide to take part, we will ask you to keep the information sheet and to sign a consent form. If you take part, you can stop taking part at any time. You will not need to give a reason and it will not affect your medical care. Also, it will not affect your medical care if you choose not to take part.

We might also ask your parent/caregiver to take part. If they would like to take part but you do not want to, we will check with you that you are happy for your family member to take part.

4. What happens if we/I decide to take part?

If you decide to take part you will be asked to take part in one interview with Frieda, who is a Trainee Clinical Psychologist. You will be interviewed alone, even if your family member takes part too. The interview will last up to an hour and will take place at a local hospital. The interview will be audio recorded and typed up and any details which could identify you will be removed, for example your name. The interview will ask questions about growing up with CF. It will also ask about family relationships and how these might have changed with getting older.

5. Will the study help me/us?

There are no direct benefits to taking part. However, it could help the CF teams to understand more about how best to offer support to young people and their families.

6. Are there any disadvantages about taking part?

You might talk about experiences that make you feel upset. You can stop the interview at any time and you can have a break whenever you wish. The researcher will often check that you are feeling okay to continue. It might also be helpful for your parent/caregiver to wait in the waiting room while you take part.

Unfortunately we are not able to provide you with any expenses for travelling to the interview.

7. Will my information be kept safe?

Yes, all information will be kept safe and secure. The audio recording of the interview will be deleted after it is typed up. Any information which could identify you (for example, names or place names) will be removed while it is typed up. The typed-up interview will be given a study number so that your name is not on it. Some quotes might be used from your interview. If quotes are used, a pretend name will be used and not yours.

Frieda will keep your information confidential and private. If something you say during the interview makes Frieda worried about you or someone else, this may need to be passed on to someone else who cares for you, such as your doctor. If this happens, Frieda will try to talk to you about it first before telling anyone else.

If you agree, Frieda will also write to your GP to let them know you are taking part.

8. What will happen to the results of the study?

Once the research is finished, Frieda will write up the project and submit it to the University of Glasgow as part of the Doctorate of Clinical Psychology degree. A report might also be submitted to a journal.

9. Further information and contact details

If you have any questions or would like more information, please contact one of the research team below:

Name	Role	Contact details
Frieda Whelan	Trainee Clinical Psychologist, Chief Investigator	Phoenix Centre, Raigmore Hospital, Old Perth Road, Inverness IV2 3UJ 01463 705597 f.Whelan@research.gla.ac.uk
Dr Alison Jackson	Academic Supervisor	Institute of Health and Wellbeing, University of Glasgow, 1st floor, Administration Building, Gartnavel Royal Hospital, 1055 Great Western Road, Glasgow G12 0XH 0141 211 3917

10. Complaints

If you have a complaint about the research, please contact your health board below:

NHS Highland	The Feedback Team, NHS Highland, PO Box 5713, Inverness IV1 9AQ nhshighland.feedback@nhs.net 01463 705997
NHS Greater Glasgow & Clyde	Complaints Department, West Glasgow Ambulatory Care Hospital, Dalnair Street, Glasgow G3 8SJ complaints@ggc.scot.nhs.uk 0141 201 4500

**Appendix 2.5: Participant information sheet for health-professionals (V2,
12/6/18)**



**Experiences of family relationships and managing cystic fibrosis in
adolescence**

Researcher: Frieda Whelan

Participant information sheet for health professionals

This information sheet describes a study which is being carried out about teenagers and their experiences of growing up with cystic fibrosis (CF) and their relationships with family members. We are also interested in the views of the parents/caregivers and health professionals.

We would like to invite you to take part in the study, but first we would like to let you know about why the study is being done and what it would involve if you took part. Please read the information sheet carefully. If you have any questions please contact the researcher, Frieda Whelan, whose details are at the end. Thank you for reading this and for considering taking part in this study.

1. Why is the study being done?

The study aims to find out about the experiences of teenagers with CF. The aim is find out how teenagers find growing up with CF and about how their relationships with family affects this and vice versa. We are also interested in the views of the parent/caregiver and health professionals. We know that growing up can be a difficult time, particularly with a health condition. Therefore, we are interested in how people might use family support at this time.

2. Why have I been invited to take part?

You are being asked to take part because you are a health professional working within a CF service in either NHS Highland or NHS Greater Glasgow & Clyde. The study hopes to recruit up to 10 people in total.

3. Do I have to take part?

No, it is up to you. If you decide to take part we will ask you to keep the information sheet and to sign a consent form. If you take part, you can stop taking part at any time, with giving a reason.

4. What happens if we/I decide to take part?

If you decide to take part you will be asked to take part in one interview with the researcher (Frieda Whelan, who is a Trainee Clinical Psychologist). The interview will last up to an hour and will take place at the hospital or by phone. The interview will be audio recorded and typed up and any details which could identify you or others will be removed. The interview will ask questions about your experience of working with teenagers with CF and their families.

5. Will the study help me?

There are no direct benefits to taking part. However, the research could help teams working with young people with CF and their families to understand more about how best to offer support during adolescence.

6. Are there any disadvantages about taking part?

It is not expected that the interview will cause any distress or difficulties. However, you are free to withdraw your participation at any time without giving a reason.

7. Will my information be kept safe?

Yes, all information will be kept safe and secure, and only the research team will have access to it. Once the audio recording of the interview is typed up it will be destroyed. Any information which could identify you (for example, names or place names) or others will be removed as it is typed up. The typed-up interview will be given a study number so that your name is not on it. Some quotes might be used from your interview. If quotes are used, a pseudonym will be used and not yours.

8. What will happen to the results of the study?

Once the research is finished, the researcher (Frieda Whelan) will write up the project and submit it to the University of Glasgow as part of the Doctorate of Clinical Psychology degree. A report might also be submitted for publication in a journal.

9. Further information and contact details

If you have any questions or would like more information please contact one of the research team below:

Name	Role	Contact details
Frieda Whelan	Trainee Clinical Psychologist, Chief Investigator	Phoenix Centre, Raigmore Hospital, Old Perth Road, Inverness IV2 3UJ 01463 705597 f.Whelan@research.gla.ac.uk
Dr Alison Jackson	Academic Supervisor	Institute of Health and Wellbeing, University of Glasgow, 1st floor, Administration Building, Gartnavel Royal Hospital, 1055 Great Western Road, Glasgow

		G12 0XH 0141 211 3917
Professor Andrew Jahoda	Independent research advisor	Institute of Health and Wellbeing, University of Glasgow, 1st floor, Administration Building, Gartnavel Royal Hospital, 1055 Great Western Road, Glasgow G12 0XH 0141 2110282

10. Complaints

If you have a complaint about any aspect of the research, please contact your healthboard below:

NHS Highland	The Feedback Team, NHS Highland, PO Box 5713, Inverness IV1 9AQ nhshighland.feedback@nhs.net 01463 705997
NHS Greater Glasgow & Clyde	Complaints Department, West Glasgow Ambulatory Care Hospital, Dalnair Street, Glasgow G3 8SJ complaints@ggc.scot.nhs.uk 0141 201 4500
Argyll and Bute Health and Social Care Partnership	The Feedback Team, Argyll and Bute Health and Social Care Partnership, Main Building, Victoria Integrated Care Centre, 93 East King Street, Helensburgh G84 7BU Argyllandbutehscp.feedback@nhs.net 01436 635155

11. NHS Highland GDPR information

NHS Highland 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. NHS Highland will keep identifiable information about you for seven years after the study has finished.

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 <http://www.nhshighland.scot.nhs.uk/Pages/YourRights.aspx> or by contacting Donald Peterkin (Interim Data Protection Officer, NHS Highland) on 01463 704000

Appendix 2.6: Example consent/assent forms

Separate consent forms were developed for: teenagers under 16, teenagers 16+, caregivers and health-professionals. Assent forms were developed for teenagers to consent to caregiver participation if the teenager was not participating, and for caregivers to agree to the participation of a teenager under 16. Examples of a consent and an assent form are given below.



Experiences of family relationships and managing cystic fibrosis in adolescence

Researcher: Frieda Whelan

Consent form (for under 16s)

Thank you for your interest in the study. This form is to make sure you are happy to take part in the study and know exactly what it involves.

Please read the following sentences carefully. If you agree with them please sign your initials in the boxes. Please sign and put the date at the bottom of this sheet if you agree to taking part in the study.

1. I have had a chance to ask questions about the study.

Please initial

2. I am happy with the answers I have been given to my questions.

3. I have been told enough information about the study.

4. I agree to the interview being audio recorded. I also agree to the researcher typing out the interview, word by word, and removing any information about people and places so that no one can tell the interview is about me.
5. I agree to the research team looking at my interview, and some parts of the interview being used in a report so long as I cannot be identified in it.
6. I understand that the information I give will be kept safely. Only the research team will be able to see it.
7. I understand that it is my choice to take part in the study. I can stop taking part at any time. I do not have to give a reason and it will not affect my medical care.
8. I agree that the research team can tell my GP I am taking part.
9. I agree to take part in the above study.

_____ Date _____ Signature _____

Participant

_____ Date _____ Signature _____

Researcher

1 copy for participant, 1 copy for researcher

Would you like a summary of the results of the research once the project is complete (approximately August 2018)? *Please tick.*

Yes, by email. My email address is _____

Yes, by post. My home address is _____

No thank you



Experiences of family relationships and managing cystic fibrosis in adolescence

Researcher: Frieda Whelan

Assent form for parent/caregiver (for under 16s)

This form is to make sure you are happy for your child to take part in the study and know exactly what it involves.

Please read the following sentences carefully. If you agree with them please sign your initials in the boxes. Please sign and put the date at the bottom of this sheet if you agree to your child taking part in the study.

Please initial

- 1. I have had a chance to ask questions about the study that my child will take part in and I am happy with the answers I have been given.
- 2. I agree to the interview being audio recorded. I also agree to the researcher typing out the interview, word by word, and anonymising the information so my child cannot be identified. I agree that anonymous quotes can be used in reports/publications.
- 3. I understand that my child's participation is voluntary and they can stop taking part at any time. They do not have to give a reason and it will not affect their medical care.
- 4. I agree to my child's GP being informed of their participation in the study.
- 5. I agree to my child taking part in the above study.

Child's name _____

_____ Date _____ Signature _____

Parent/caregiver

_____ Date _____ Signature _____

Researcher

1 copy for parent/caregiver, 1 copy for researcher

Would you like a summary of the results of the research once the project is complete (approximately August 2018)? *Please tick.*

Yes, by email. My email address is _____

Yes, by post. My home address is _____

No thank you

Appendix 2.7: Interview guides

Interview schedule for teenagers (V1, 23/8/17)

Thank you for meeting with me today, my name is Frieda Whelan and I am a trainee Clinical Psychologist working for NHS Highland, undertaking a research project as part of my training. I am working alongside the CF team for my research project and am supervised by a psychologist within the team.

As you will be aware, the study is aimed at finding out more about the experiences of young people with cystic fibrosis. In particular it aims to explore relationships as a young person gets older and how this might affect managing CF. I will be recording today's interview so I can refer back to the information. Everything you say will be confidential and the information recorded will be stored securely. I may pass on information if there is anything which would make me concerned about the safety of yourself or someone else or if I feel you may benefit from more support from the team. I may pass this information on to the CF team, however, I would try to discuss this with you first. The interviews are anonymised, including any information which is used in a report of the study. You can withdraw from the interview at any time without having to give a reason, and this will not affect your clinical care. The interview will take up to an hour – please tell me if you would like a break at any point during the interview.

Young person's life with CF

- Can you tell me about what it is like to live with CF, at the moment?
 - Prompt: how does CF affect your day to day to life at the moment?
- What is going well/more difficult?

Exploring relationships at home

- Can you tell me about the people around you? (use a genogram/mapping exercise to plot who helps and in what way)
 - Prompt: who is in your family? Who else is around for you?
- Can you describe what closeness looks like in your family?

- Can you describe what gets in the way of closeness at home/what does distance look like?
- In your mind, what do you think your (main caregivers) think of CF?
- In what way, if any, does this influence closeness/distance?

Experiences of CF with transition into adolescence

- Has there been any change in how your CF affects day to day life as you have gotten older?
 - Prompt: how are things different now that you are in high school compared with primary school?
- Can you describe what these changes feel like for you?
- Has there been change in how confident you feel in managing your condition since you've gotten older? Can you tell me more about it?
 - Prompt: what does that feel like?
- How do you feel about the level of support you have to manage the condition?
 - Prompt: would you prefer more or less? Why?
 - Prompt: would you want the support to be different? In what way?

Exploring relationships and CF management

- As you've gotten older, how have your conversations with your family about CF changed?
 - Prompt: if no changes, why do you think this is? If there have been changes, how do you feel about this?
- Since becoming older, have you found that your relationships have changed?
- In what way have they changed?
- What has worked well in the relationship between you and (caregiver) in managing your CF?
- Is there anything about your relationship which makes it more difficult to manage your CF?

Effect of rurality/urban

- Have you experienced any challenges in attending appointments?
- Does living in a more rural area influence life with CF? Can you tell me more?

Interview schedule for caregivers (V1, 23/8/17)

Thank you for meeting with me today, my name is Frieda Whelan and I am a trainee Clinical Psychologist working for NHS Highland, undertaking a research project as part of my training. I am working alongside the CF team for my research project and am supervised by a psychologist within the team.

As you will be aware, the study is aimed at finding out more about the experiences of caregivers of young people with cystic fibrosis. In particular it aims to explore relationships as a young person gets older and how this might affect managing their condition. I will be recording today's interview so I can refer back to the information. Everything you say will be confidential and the information recorded will be stored securely. I may pass on information if there is anything which would make me concerned about the safety of yourself or someone else or if I feel you may benefit from more support from the team. I may pass this information on to the CF team, however, I would try to discuss this with you first. The interviews are anonymised, including any information which is used in a report of the study. You can withdraw from the interview at any time without having to give a reason, and this will not affect your clinical care. The interview will take up to an hour - please tell me if you would like a break at any point during the interview.

Demographic information

- What is your relationship to [young person with CF]?

General opening question

- Can you tell me about what it is like to be the [parent etc.] of (adolescent's name)?
- Can you describe what it is like to parent a young person with CF, at the moment?
 - Prompt: how does them having CF affect day to day to life at the moment?
 - Prompt: what is going well/more difficult?
 - Prompt: how do you feel about the support you have to manage (x's) condition?

Exploring relationships at home

- Can you tell me about who is around you and (x) at home?
 - Prompt: who is in your family? Who else is around you and (x)?
- Can you describe what closeness looks like in your family?
- Can you describe what gets in the way of closeness at home/what does distance look like?
- In your mind, what do you think (x's name) thinks of CF?
- In what way, if any, does this influence closeness/distance?

Experiences of CF with transition into adolescence

- Has there been any change in how CF affects (x's) life day to day as they've gotten older?
 - Prompt: how are things different now they are in high school compared with primary school?
- When did you notice things changing?
- Have you noticed a change in how confident (x) is in managing their CF as she/he has gotten older? Can you tell me more about it?
 - Prompt: how do you feel about these changes?
- How do you feel about the support (family and professional) (x) has received in doing this?
 - Prompt: would you prefer them to have more or less support? Why?
 - Would you want the support to be different? In what way?

Exploring relationships and CF management

- How have your conversations changed about CF as (x) has gotten older?
 - Prompt: if not, why do you think this is?
 - Prompt: how do you feel about the level of support/guidance you have received for these types of conversations?
- What way, if any, has your relationship changed as (x) grows older?
- In what way has it changed?
- What has worked well in the relationship between you and (x) in managing their CF?
- Is there anything about your relationship which makes it more difficult for you and (x) to manage CF?

Effect of rurality/urban

- Have you experienced any challenges in attending appointments?
- Does living in a more rural area influence life with a young person with CF?

Can you tell me more?

Interview schedule for health-professionals (V1, 11/5/18)

Thank you for meeting with me today, my name is Frieda Whelan and I am a trainee Clinical Psychologist working for NHS Highland, undertaking a research project as part of my training. As you will be aware I am working alongside the CF team for my research project and am supervised by a psychologist within the team.

The study is aimed at finding out more about the experiences of young people with cystic fibrosis. In particular it aims to explore relationships as a young person gets older and how this might affect managing CF. I am also interested in this from the perspective of health professionals working closely with young people with CF and families.

I will be recording today's interview so I can refer back to the information. Everything you say will be confidential and the information recorded will be stored securely. The interviews are anonymised, including any information which is used in a report of the study. You can withdraw from the interview at any time without having to give a reason. The interview will take up to an hour – please tell me if you would like a break at any point during the interview.

General questions about working with families

- Can you tell me about what it's like to work with young people with CF and their families?
- What are the families like that you work with?

Questions about working with families during adolescence

- Can you tell me about what it's like to work with the families of adolescents with CF?
 - Prompt: what works well (during adolescence in particular)?
 - Prompt: are there any challenges (during adolescence in particular)?
What are they?
 - Prompt: how does adolescence differ from other ages/stages?

- Have you noticed changes in changes in family relationships over adolescence? Can you describe these?

Questions about the interaction of CF with family relationships

- Do you think CF affects family relationships? If yes, in what way? If no, why?
- Do you think family relationships can affect how a young person manages CF?

Questions about family support

- Can you tell me about your views on the role of family for teenagers with CF?
 - Prompt: how do families help teenagers with CF?
 - Prompt: how do families hinder teenagers with CF?
- What do you think works well in terms of family support?
- What do you think works less well in terms of family support?
- What kind of support would be offered/suggested to families who are experiencing difficulties?
- **Question about recruitment to project**
- Why do you think it's been difficult to recruit families for a research project looking at experiences of CF and family relationships in adolescence?

Appendix 2.8: Approval from Research Ethics Committee and Research and Development

Lothian NHS Board

**South East Scotland Research
Ethics Committee 01**



Waverley Gate
2-4 Waterloo Place
Edinburgh
EH1 3EG
Telephone 0131 536 9000

www.nhsllothian.scot.nhs.uk

Date 06 December 2017

10 November 2017

(re-issued 06 Dec 2017 with corrections)

Mrs Frieda Whelan
Trainee Clinical Psychologist
NHS Highland
New Craigs Hospital
6-16 Leachkin Road
Inverness
IV3 8NP

Enquiries to: Sandra Wyllie
Extension: 35473
Direct Line: 0131 465 5473
Email: Sandra.Wyllie@nhsllothian.scot.nhs.uk

Dear Mrs Whelan

Study title: Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis
REC reference: 17/SS/0138
IRAS project ID: 228143

Thank you for your letter of 03 November 2017, responding to the Committee's request for further information and for submitting revised documentation.

The further information has been considered on behalf of the Committee by the Chair.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this opinion letter. Should you wish to provide a substitute contact point, require further information, or wish to make a request to postpone publication, please contact hra.studyregistration@nhs.net outlining the reasons for your request.

Confirmation of ethical opinion

On behalf of the Committee, I am pleased to confirm a favourable ethical opinion for the above research on the basis described in the application form, protocol and supporting documentation as revised, 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.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

Management permission 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 NHS permission for research is available in the Integrated Research Application System, www.hra.nhs.uk or at <http://www.rdforum.nhs.uk>.

Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.

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

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database within 6 weeks of recruitment of the

first participant (for medical device studies, within the timeline determined by the current registration and publication trees).

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact hra.studyregistration@nhs.net. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

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).

Ethical review of research sites

NHS sites

The favourable opinion applies to all NHS sites taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion" below).

Approved documents

The final list of documents reviewed and approved by the Committee is as follows:

<i>Document</i>	<i>Version</i>	<i>Date</i>
Copies of advertisement materials for research participants [Poster advertising study]	2	23 August 2017
Covering letter on headed paper [Cover letter - changes]	1	03 November 2017
GP/consultant information sheets or letters [GP letter for adolescent participants]	1	20 October 2017
Interview schedules or topic guides for participants [Interview schedule for caregiver participants]	3	23 August 2017
Interview schedules or topic guides for participants [Interview schedule for adolescent participants]	3	23 August 2017
Letters of invitation to participant [Letter of invitation - Glasgow]	3	27 October 2017

Letters of invitation to participant [Letter of invitation - Highland]	2	27 October 2017
Other [Opt in form]	3	03 November 2017
Participant consent form [Consent form for participants aged 16+]	3	03 November 2017
Participant consent form [Consent form for participants aged under 16]	3	20 October 2017
Participant consent form [Assent form for parents/caregivers if participant under 16]	3	20 October 2017
Participant consent form [Teen assent form is parent but not teen participating]	1	20 October 2017
Participant consent form [Consent form for caregivers]	1	03 November 2017
Participant information sheet (PIS) [Participant information sheet for caregivers]	3	20 October 2017
Participant information sheet (PIS) [Participant information sheet for adolescents]	1	20 October 2017
REC Application Form [REC_Form_29082017]		29 August 2017
Research protocol or project proposal [Research protocol]	2	20 October 2017
Summary CV for Chief Investigator (CI) [CV for chief investigator]	1	23 August 2017
Summary CV for supervisor (student research) [CV for academic supervisor]	1	23 August 2017

Statement of compliance

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.

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

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

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 Training

We are pleased to welcome researchers and R&D staff at our training days – see details at <http://www.hra.nhs.uk/hra-training/>

17/SS/0138	Please quote this number on all correspondence
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With the Committee's best wishes for the success of this project.

Yours sincerely



Mrs Christine Beadle Chair

Email: sandra.wyllie@nhslothian.scot.nhs.uk

Enclosures: "After ethical review – guidance for researchers"

Copy to: Ms Frances Hines, NHS Highland

Professor Angus Watson
Research, Development & Innovation Director
Research, Development & Innovation Division
NHS Highland
Centre for Health Science
Old Perth Road
Inverness
IV2 3JH

Tel: 01463 255822
Fax: 01463 255838
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05 December 2017

NHS Highland R&D ID: 1329
NRSPCC ID: NRS17/228143

Dr Stephanie Bannon
Clinical Psychologist
NHS Highland
Phoenix Centre
Old Perth Road
Inverness
IV2 3JH

Dear Dr Bannon,

Management Approval for Non-Commercial Research

I am pleased to tell you that you now have Management Approval for the research project entitled: **'Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis. [Protocol V2.0 20/10/2017].** I acknowledge that:

- The project is sponsored by the NHS Highland.
- The project has no external funding.
- Research Ethics approval for the project has been obtained from the South East Scotland Research Ethics Committee (Reference Number: 17/SS/0138).
- The project has been Site-Specific Assessment exempt.

The following conditions apply:

- The responsibility for monitoring and auditing this project lies with the NHS Highland.



Headquarters:
NHS Highland, Assynt House, Beechwood Park, Inverness, IV2 3HG

Chairman: David Alston
Chief Executive: Elaine Mead

- This study will be subject to ongoing monitoring for Research Governance purposes and may be audited to ensure compliance with the Research Governance Framework for Health and Community Care in Scotland (2006, 2nd Edition), however prior written notice of audit will be given.
- Any researchers coming into NHS Highland for the purposes of carrying out research with patients will require the submission of a Research Passport, Occupational Health approval and Letter of Access before starting the study at this site. Please contact Anna McIver (anna.mciver@nhs.net) for further assistance, if this is required
- You are reminded that all amendments (minor or substantial) to the protocol and associated study documents or to the REC application should be copied to the NHS Highland Research and Development Office together with a copy of the corresponding approval letter. Guidance can be found at <https://www.nhsresearchscotland.org.uk/services/permissions-co-ordinating-centre/permissions>
- The paperwork concerning all incidents, adverse events and serious adverse events, if thought to be attributable to participant's involvement in this project, and appropriate to the study, should be copied to the NHS Highland R&D Office. Please email documents to Anna McIver, RD&I Facilitator (anna.mciver@nhs.net).
- If applicable, monthly recruitment rates should be notified to the NHS Highland Research and Development Office, detailing date of recruitment and the participant trial ID number. This should be done by e-mail on the first week of the following month, to Debbie McDonald, RD&I Data Manager (debbie.mcdonald@nhs.net).
- Please report any other changes in resources used, or staff involved in the project, to the NHS Highland Research and Development Manager, Frances Hines (01463 255822, frances.hines@nhs.net).

Please quote your RD&I Highland reference number (Highland 1329).

Yours sincerely,



Frances Hines
RD&I Manager

cc Frances Hines, R&D Manager, NHS Highland Research, Development & Innovation Division, Phase 3, The Centre for Health Science, Old Perth Road, Inverness, IV2 3JH

Mrs Frieda Whelan, Trainee Clinical Psychologist, NHS Highland, New Craigs Hospital, 6 – 16 Leachkin Road, Inverness, IV3 8NP

Administrator: Sophie Bagnall

Telephone Number: 0141 232 1826

E-Mail: sophie.bagnall@ggc.scot.nhs.uk

Website: www.nhsggc.org.uk/r&d

**R&D
Management
Office
West Glasgow
ACH
Dalnair Street
Glasgow G3
8SW**

6 December 2017

Dr Ruth Hind
Clinical Psychology
NHS Greater Glasgow & Clyde
Department of Paediatric Clinical Psychology
Royal Hospital for Children
Glasgow
G51 4TF

NHS GG&C Board Approval

Dear Dr Hind,

Study Title:	Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis
Principal Investigator:	Dr Ruth Hind
GG&C HB site	Royal Hospital for Children
Sponsor	NHS Highlands
R&D reference:	GN17RM427
REC reference:	17/SS/0138
Protocol no:	v2 20.10.17

I am pleased to confirm that Greater Glasgow & Clyde Health Board is now able to grant **Approval** for the above study.

Conditions of Approval

1. **For Clinical Trials** as defined by the Medicines for Human Use Clinical

Trial Regulations, 2004 a. During the life span of the study GGHB

requires the following information relating to this site

- i. Notification of any potential serious breaches.
- ii. Notification of any regulatory inspections.

It is your responsibility to ensure that all staff involved in the study at this site have the appropriate GCP training according to the GGHB GCP policy (www.nhsggc.org.uk/content/default.asp?page=s1411), evidence of such training to be filed in the site file

2. **For all studies** the following information is required during their lifespan.

- a. Recruitment Numbers on a monthly basis
- b. Any change of staff named on the original SSI form
- c. Any amendments – Substantial or Non Substantial
- d. Notification of Trial/study end including final recruitment figures
- e. Final Report & Copies of Publications/Abstracts

Please add this approval to your study file as this letter may be subject to audit and monitoring.

Your personal information will be held on a secure national web-based NHS database. I wish you every success with this research study

Yours sincerely,



Sophie Bagnall Senior Research Administrator

Appendix 2.9: Approval from Research Ethics Committee and Research and Development for amendment

Lothian NHS Board

**South East Scotland Research
Ethics Committee 01**



Waverley Gate
2-4 Waterloo Place
Edinburgh
EH1 3EG
Telephone 0131 536 9000

www.nhslothian.scot.nhs.uk

Enquiries to: Sandra Wyllie
Extension: 35473
Direct Line: 0131 465 5473
Email: Sandra.Wyllie@nhslothian.scot.nhs.uk

13 June 2018

Mrs Frieda Whelan

Dear Mrs Whelan

Study title: Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis
REC reference: 17/SS/0138
Amendment number: 17/SS/0138/AM01
Amendment date: 11 May 2018
IRAS project ID: 228143

The above amendment was reviewed the Sub-Committee in correspondence.

Ethical opinion

The members of the Committee taking part in the review gave a favourable ethical opinion of the amendment on the basis described in the notice of amendment form and supporting documentation.

The Sub-Committee had no ethical concerns regarding the amendment however as a result of their discussion the Sub-Committee raised the following points and queries:

The Sub-Committee suggested that if the researcher was introducing the views of health professionals then this should be stated this in the participant/caregiver PIS. In response, the researcher has added into the participant/caregiver PIS that the views of healthcare professionals are included.

The Sub-Committee noted that there appears to be no independent contacts in any of the PIS and requested that this information has to be added to the PIS. In response, the researcher has added an independent contact from the University of Glasgow to the PIS.

The Sub-Committee noted that there were no GDPR wording added to the documents and queried whether the researcher plans to add this information at a later stage. In response, the researcher has added GDPR working to the PIS.

The Sub-Committee reviewed and approved the updated versions of the PIS for staff, caregivers and adolescents dated 12th June 2018. No further queries were raised.

Approved documents

The documents reviewed and approved at the meeting were:

Document	Version	Date
Letters of invitation to participant [Clyde/Argyll]	1	11 May 2018
Letters of invitation to participant [Glasgow adult service]	1	11 May 2018
Non-validated questionnaire [Staff interview schedule]	1	11 May 2018
Notice of Substantial Amendment (non-CTIMP)		11 May 2018
Participant consent form [staff participants]	1	11 May 2018
Participant information sheet (PIS) [adolescents]	3	12 June 2018
Participant information sheet (PIS) [caregivers]	5	12 June 2018
Participant information sheet (PIS) [staff]	2	12 June 2018
Research protocol or project proposal	3	11 May 2018

Membership of the Committee

The members of the Committee who took part in the review are listed on the attached sheet.

Working with NHS Care Organisations

Sponsors should ensure that they notify the R&D office for the relevant NHS care organisation of this amendment in line with the terms detailed in the categorisation email issued by the lead nation for the study.

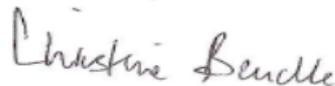
Statement of compliance

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.

We are pleased to welcome researchers and R & D staff at our Research Ethics Committee members' training days – see details at <http://www.hra.nhs.uk/hra-training/>

17/SS/0138:	Please quote this number on all correspondence
--------------------	---

Yours sincerely



Mrs Christine Beadle
Chair

South East Scotland REC 01

Attendance at Sub-Committee of the REC meeting

Committee Members:

<i>Name</i>	<i>Profession</i>	<i>Present</i>	<i>Notes</i>
Mrs Christine Beadle	Research Nurse	Yes	In the Chair
Dr Derek Santos	Senior Lecturer - Faculty Of Health Sciences	Yes	
Mrs Amy Shepherd	Senior Research Nurse - Regional Infectious Diseases Unit	Yes	

Also in attendance:

<i>Name</i>	<i>Position (or reason for attending)</i>
Mrs Sandra Wylie	REC Manager

Document: AM Approval letter Frances Hines
 27.02.17
 R&D Ref No: 1329
 REC Ref No: 17/SS/0138
 NRS Ref No: NRS17/228143
 EudraCT Ref No: NA MHRA
 Ref No: NA
 Today's Date: 18/06/2018

Research, Development &
 Innovation Manager
 NHS Highland
 Research, Development &
 Innovation Department
 Centre for Health Science
 Old Perth Road
 Inverness IV2 3JH
 Tel: 01463 255821
 E-mail:
frances.hines@nhs.net



Dr Stephanie Bannon
 Clinical Psychologist
 NHS Highland
 Phoenix Centre
 Old Perth Road
 Inverness IV2 3JH
 By email: s.bannon@nhs.net

Dear Dr Bannon,

LETTER OF APPROVAL OF YOUR RESEARCH PROJECT AMENDMENT

PROJECT TITLE: Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis

REC: 17/SS/0138

NHS Highland R&D Ref Number: 1329

Amendment Type:	Substantial (SA)	√
	Modified SA	
	Non-substantial (NSA)	
Amendment No:	AM 01	
Amendment Date:	11.05.18	

Current Protocol Version No:	V3 Dated 11.05.18

We have been notified of the above amendment to your research project and have received the following documents:

- Notification of Substantial Amendment form.
- Amended documents corresponding with those approved in the REC amendment approval letter (South East Scotland Research Ethics Committee 01) dated 13.06.18.

The **RD&I Division**, NHS Highland, is happy to **approve** this amendment as it is within the scope of the original Management Approval Letter (05.12.17).

Yours sincerely,



Frances Hines

NHS Highland Research, Development & Innovation Manager

Dear Dr Hind,

R&D Ref: GN17RM427 **Ethics Ref:** 17/SS/0138

Investigator: Dr Ruth Hind

Project Title: Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis

Protocol Number: v3 11.05.18

Amendment: SA01 11/05/18 and NSA02

Sponsor: NHS Highlands

I am pleased to inform you that R&D have reviewed the above study's Amendments SA01 11/05/18 and NSA02 and can confirm that Management Approval is still valid for this study.

I wish you every success with this research project.

Reviewed Documents:	Version	Dated
Ethics approval		13/06/18
Notice of substantial amendment		11/05/18
Letter of invitation to participant (Clyde/Argyll)	1	11/05/18
Letter of invitation to participant (Glasgow adult service)	1	11/05/18
Staff interview schedule	1	11/05/18
Consent form (staff participants)	1	11/05/18
Information sheet (adolescents)	3	12/06/18
Information sheet (caregivers)	5	12/06/18
Information sheet (staff)	2	12/06/18
Protocol	3	11/05/18
Notice of minor amendment (clarification)		29/06/18

Yours sincerely,

Research and Development Department
NHS Greater Glasgow and Clyde

Appendix 2.10: Research proposal

Experiences of family relationships and managing cystic fibrosis in adolescence using Interpretative Phenomenological Analysis

Abstract

Background

Early detection and improved treatments have meant that the life expectancy of those with cystic fibrosis (CF) has improved. However, those with CF and their caregivers can experience elevated levels of anxiety and depression. Accordingly, there is a need to provide for the psychological needs of people with CF and their families. Families may need support during significant life transitions such as from childhood into adolescence. However, there are few qualitative studies on how family relationships and support interacts with CF during adolescence; a time when increasing responsibility for treatment may be expected.

Aims

The aim of the study is to explore adolescent and caregiver experiences of the transition into adolescence with CF and how family factors interact with this. This will be explored using Interpretative Phenomenological Analysis (IPA).

Methods

Four to ten interviews will be carried out with adolescents with CF (aged 13-18) and caregivers, who will be purposively recruited from two CF services in NHS Highland and NHS Greater Glasgow & Clyde. Individuals will be interviewed on their experiences and the interviews analysed with IPA to generate common themes.

Applications

The results could help services to understand more about how individuals experience CF into adolescence, and what factors in the family may help or hinder the management of the condition, allowing for assessment and intervention to be adapted.

Introduction

Cystic fibrosis (CF) is a life-limiting genetic condition, affecting 10, 583 people in the UK, including 970 in Scotland, during 2014 (Cystic Fibrosis Trust, 2015). Median life expectancy has increased by 10 years in recent times and is currently 47 in the UK (Cystic Fibrosis Trust, 2016). Early detection through newborn screening, along with improvements in medical treatments has contributed to this. Additionally, future treatments are being investigated which target the genes implicated in CF and may hold promise in further improving life expectancy (Antoniou & Elston, 2016). Thus, there is growing need to meet the longer-term psychological needs of those with CF and their families.

The chronic and life-limiting nature of CF, along with the demands of managing the condition, can pose challenges to the family. Indeed, individuals with CF and their caregivers experience elevated levels of anxiety and depression, which in turn can affect treatment adherence and use of health services (Quittner *et al.*, 2016). A systematic review of 43 qualitative studies exploring children and adolescents' experiences of CF highlights the unique challenges they can face growing up with CF, along with resilience factors (Jamieson *et al.*, 2014). Themes included developing resilience, recognising restrictions, challenges of chronic treatment, effect of time limitations, emotional difficulties and thoughts around transplants. Altogether, the

studies included at least 729 children, however, just 12 of them included only adolescents (per the World Health Organisation definition of adolescence as spanning ages 10 to 19, WHO, n.d.).

Adolescence is a complex time, particularly with CF when children will gradually take on more responsibility for the management of their condition, whilst continuing to share responsibility with their parents (Drotar & Ievers, 1994). In early adolescence, individuals will transition from primary to secondary school, which will bring with it increased independence which may impact on the balance of responsibility for the management of their condition. Berge and Patterson (2004) review studies regarding family functioning and CF and highlight the complexity of the issue of how much responsibility to confer to the adolescent, and how this remains unresolved in the literature. Only six of the 54 studies were qualitative and none of the six studies considered a solely adolescent sample. A qualitative approach could be particularly useful because it affords a deeper understanding of the complexity of family relationships.

The nature of family relationships can be associated with treatment adherence and emotional distress. In one study, a more positive family environment was associated with increased self-reported adherence to lung treatments (DeLambo, Ievers-Landis, Drotar, and Quittner, 2004). Family relationships have also been related to emotional coping, for example, parent-reported attachment and communication have been shown to be related to self-reported internalising problems and emotional symptoms in young people with CF (Tluczek *et al.*, 2014). Graetz, Shute and Sawyer (2000) explored behaviours that adolescents perceive as supportive and non-supportive.

They found that perceived non-supportive behaviours from family members was a strong predictor of psychological maladjustment, however, they only asked one question around non-supportive behaviours and did not consider the mechanisms by which this might happen. Support from family members is likely to be of increasing importance since it is recommended that individuals with CF are segregated as an infection control measure (Cystic Fibrosis Trust, 2004).

Understanding the interplay of a chronic health condition with family relationships may be well-suited to a qualitative approach, for example Interpretative Phenomenological Analysis (IPA), which can help to explore how individuals make sense of significant life experiences through interpretation of rich and detailed data (Smith & Eatough, 2007).

Aims

There is a paucity of qualitative research on the relationship between growing up with CF and family relationships in adolescence. The aims of the present study are to explore the experience of adolescents' transition from late childhood to adolescence with a chronic health condition, and how family factors interact with this. Particularly, the research aims to ask whether there are changes in relationships, what works well in terms of family support, and what is problematic.

Plan of investigation

- Participants

Participants aged 13-16 will be purposively recruited through the paediatric service at the Royal Hospital for Children, NHS Greater Glasgow and Clyde (NHS GG&C). As of

October 2016, 26 of individuals currently under review were aged 12-16. In NHS GG&C, adolescents independently attend transition clinics between the ages of 14 and 16, and move into adult services at age 17.

Participants aged 13-18 will be recruited through NHS Highland. Five of the fourteen families being seen by the CF service will be eligible to take part during the recruitment period. The CF nurse for NHS Highland, who will act as a gatekeeper for recruitment, knows the families well and believes that there would be interest in taking part in research. There is no psychologist attached to the team but input from the paediatric health psychology service is available on a referral basis. In NHS Highland, the clinical team and hospital at which adolescents are seen remains unchanged into transition. The medical consultant changes, however, the CF nurse covers both paediatric and adult populations. At age 14 adolescents are introduced to the adult consultant and transition occurs between the ages of 16 and 18 (dependent on leaving school).

Participants will be aged between 13 and 16/18. This will allow for a relatively homogenous population to be recruited. Adolescents of this age will have left primary school, meaning they have experienced a significant life event (i.e. transition to high school), which inevitably affects independence. This could also affect responsibility for health management. Caregivers of adolescents meeting the above criteria will also be recruited. To ensure the study is open to adolescents from different family backgrounds, the caregiver could be a mother, father or other relative who is a primary caregiver for the participant. Details of the relationship of the caregiver will be collected. Obtaining more than one perspective will afford a richer understanding of family relationships during adolescence (Smith, Flowers & Larkin, 2009, p. 52).

Caregivers and adolescents of each family will both be invited to participate; however, it will not be necessary for both to participate. If a caregiver is willing to participate but not the adolescent, it will be ensured that the adolescent agrees to the caregiver's participation and to sharing of information which pertains to them.

- **Inclusion and exclusion criteria**

Inclusion criteria:

- CF diagnosis/are the primary caregiver of an adolescent with a CF diagnosis
- Attendance at CF service at the Royal Hospital for Children, Glasgow or Raigmore Hospital, Inverness
- Fluent in English
- Aged 13-18/a caregiver
- Consent to participate

Exclusion criteria:

- Known learning disability
- Lack of capacity to give consent
- Communication difficulties which preclude engagement in an interview
- Acute psychological distress and/ or the clinical team deems that participation would be an additional burden for the individual

- **Recruitment procedures**

Clinical teams will be asked to provide a research pack to individuals who would be eligible to participate. This will be posted to all eligible families (n = 31). If the

adolescent has a clinic appointment scheduled during recruitment period (approximately October 2017 – May 2018) then the clinician can discuss the study directly with them if there has been no response to the letter. The research pack will include information about the study and an opt-in slip, giving the researcher permission to contact them. Alternatively, they can reply by email or call the researcher. An opt-out slip will also be provided for those who do not wish to be contacted. The researcher will then contact interested participants by phone to discuss the study and to obtain consent. Those who have not responded to the letter and have not opted out might also be contacted by the researcher to check if they are interested. Also, posters advertising the study might be placed in waiting rooms and interested participants can contact the researcher directly. There is also a newsletter which is distributed to families in Highland and a parents' group and team website in Glasgow where information about the research could be presented. After consent is obtained, a suitable time will then be agreed with the participants for the interview.

- **Interview**

A semi-structured, in-depth interview will be carried out on a one-to-one basis, lasting up to an hour. An interview schedule will enable discussion around the aims of the study. Draft interview schedules, which have been reviewed by an expert in the field (Dr Louise Phin), are included in appendices 2 and 3.

Creative methods might be employed to facilitate discussion. For example, Veale (2005) discusses methods which can be employed to engage children in qualitative research, such as a “social mapping” exercise. This involves “mapping” out who helps a child and in what way. In a grounded theory study, Christian and D’Auria (1997) used

more concrete methods such as enquiring about earliest memories, asking adolescents to give advice to a younger child with CF and a timeline of critical events. Punch (2002) outlines methods for engaging 13-14 year olds in interviews; such as spider diagrams for eliciting information about problems and social support charts.

- **Design**

The study will employ a qualitative design using semi-structured interviews. IPA is an approach to qualitative data analysis which seeks to understand how people make sense of significant experiences through the researcher's interpretation of the person's narrative account (Smith *et al.*, 2009). Transition into adolescence, particularly with a chronic illness which may require increased self-management, could be regarded as an important life experience, and as such IPA is favoured over other designs.

- **Research procedures**

Using the interview schedule as a guide (see appendices 2 and 3), the researcher will initiate discussion to explore topics associated with the aims of the study. Participants will be interviewed alone to ensure there is no bias in their answers from having their caregiver/adolescent present. A waiting area will be available for anyone waiting for a participant. The researcher will remain sensitive to how the interviewee is feeling, and adapt the interview accordingly. The interview schedule will be piloted in the first two interviews and feedback sought from these participants. Emerging themes will also be considered and the interview schedule may be adapted. Interviews will be recorded, transcribed, and identifying information removed prior to analysis.

- **Data analysis**

Data will be analysed using IPA, a method used to understand how people perceive significant life experiences (Smith *et al.*, 2009). This involves thorough and repeated reading of the data, noting of initial ideas, identifying possible themes and mapping out connections between themes. This is repeated for each participant, prior to looking for patterns across participants (Smith *et al.*, 2009). An independent researcher will analyse a subset of transcripts to ensure the themes which have emerged are similar.

- **Justification of sample size**

Smith *et al.* (2009) advocate between 4 and 10 interviews for professional doctorate theses. As both adolescents and caregivers are being recruited then the number of interviews is likely to be within the higher range of this estimate.

- **Settings and equipment**

Participants will be interviewed within a private room at their respective hospital (Royal Hospital for Children/Raigmore). Where possible, this will be on the researcher's study days (Friday, and also Thursday as of April 2018). At the Royal Hospital for Children rooms can be booked in the multi-disciplinary team (MDT) hub. Local infection control procedures will be followed and the researcher will be trained by a clinician in infection control procedures when working with people with CF. In NHS Highland, rooms are bookable at the Phoenix Centre, Raigmore, where paediatric health psychology is based. If participants live far from Inverness, rooms are bookable at other satellite clinics. Another option is to conduct interviews by phone. By offering an option to those who may not be able to attend a clinic, for example due to difficulties accessing transport, childcare etc., a more representative sample can be included. There is

evidence to suggest that telephone interviews can yield a similar amount and depth of information as face-to-face interviews (Sturges & Hanrahan, 2004). Home visits may also be considered for participants who may be unable/unwilling to be interviewed in a clinic or by phone. Interviews will be recorded on a digital voice recorder. Access to transcribing software and an encrypted laptop will be required for analysis of the data.

Health and safety issues

- Researcher & participant safety issues

One-to-one interviews will be conducted in clinic rooms during working hours, and usual safe/lone working practices will be followed. A Health and Safety for Researchers form is included in appendix 4, detailing the measures which will be taken to minimise potential risks to the researcher and participants. Home visits will only be conducted in cases where the criteria in section 10 (iii) have been met.

Ethical issues

Sponsorship will be provided by NHS Highland. An application will be submitted to NHS Highland and NHS GG&C Research and Development departments and to the Research Ethics Committee. Concurrently, an application will be submitted to NHS Research Scotland Permissions Co-ordinating Centre (NRSPCC) for multi-site approval. Participants will be asked to provide written consent; if under 16, this will be sought from both the adolescent and caregiver/parent (ScotCRN, 2012). If aged 16 or above, only the participant will be required to provide consent. Identifiers will be removed from the data to allow anonymity and the Data Protection Act 1998 will be adhered to. Audio recordings will be held on an encrypted laptop and retained until no longer required for the purposes of the project. Prompting individuals to talk about CF

could initiate a reflective process about sensitive issues, such as the life-limiting nature of their condition. As such, participants may become distressed. The researcher will remain aware of the sensitive nature of the discussion and follow the procedure in appendix 4, Section 10 (ii) for managing distress.

Financial issues

Equipment to be borrowed from the University of Glasgow includes a digital voice recorder and encrypted laptop. Costs are outlined in appendix 5. As travelling expenses are not available from NHS Highland, where possible the researcher will plan interviews around travel to Glasgow for teaching.

Timetable

MRP proposal	30 th January 2017
Final approved MRP proposal and paperwork	15 th May 2017
Apply for ethical approval	June-August 2017
Begin recruitment	October 2017
Analysis	February-June 2018
Write up	May - July 2018
Submission of thesis	July 2018
Viva	September 2018

Practical applications

Understanding the experience of individuals with CF and their families will contribute to a greater recognition of how services can best support and meet the needs of

families. Understanding how the stress of managing CF impacts on family communication and relationships, in both positive and less helpful ways, can help teams to understand how best to maintain supportive relationships with adolescents and families during the transition period. This could lead to potential service development, such as the provision of new information leaflets; and inform clinical practice by allowing clinicians to guide their questioning and tailor intervention.

References

Antoniou, S. & Elston, C. (2016). Cystic fibrosis. *Medicine*, 44, 5, 321-325.

Berge, J.M. & Patterson, J.M. (2004). Cystic fibrosis and the family: a review and critique of the literature. *Families, Systems & Health*, 22, 1, 74-100.

Christian, B.J. & D'Auria, J.P. (1997). The child's eye: memories of growing up with cystic fibrosis. *Journal of Pediatric Nursing*, 12, 1, 3-12.

Cystic Fibrosis Trust (2004). *Pseudomonas aeruginosa infection in people with cystic fibrosis. Suggestions for prevention and infection control* [PDF file]. Retrieved from: <https://www.cysticfibrosis.org.uk/~media/documents/the-work-we-do/care/consensus-docs-with-new-address/pseudomonas-aeruginosa.ashx?la=en> (Accessed 21 January 2017).

Cystic Fibrosis Trust (2015). *Cystic Fibrosis: strength in numbers. UK Cystic Fibrosis Registry 2014 Annual Data Report – Scotland* [PDF file]. Retrieved from:

<https://www.cysticfibrosis.org.uk/~media/documents/the-work-we-do/uk-cf-registry/scottish-registry-report-2014.ashx?la=en> (Accessed: 21 January 2017).

Cystic Fibrosis Trust (2016). *Cystic Fibrosis: strength in numbers. UK Cystic Fibrosis Registry 2015 Annual Data Report* [PDF file]. Retrieved from: <https://www.cysticfibrosis.org.uk/~media/documents/the-work-we-do/uk-cf-registry/full-registry-report-2015.ashx?la=en> (Accessed: 21 January 2017).

Data Protection Act (1998). Retrieved from: <http://www.legislation.gov.uk/ukpga/1998/29> (Accessed: 4 December 2016).

DeLambo, K.E., levers-Landis, C.E., Drotar, D. & Quittner, A.L. (2004). Association of observed family relationship quality and problem-solving skills with treatment adherence in older children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology*, 29, 5, 343-353.

Drotar, D. & levers, C. (1994). Age differences in parent and child responsibilities for management of cystic fibrosis and insulin-dependent diabetes mellitus. *Developmental and Behavioral Pediatrics*, 15, 4, 265-272.

Graetz, B., Shute, R.H. & Sawyer, M.G. (2000). An Australian study of adolescents with cystic fibrosis: perceived supportive and nonsupportive behaviors from families and friends and psychological adjustment. *Journal of Adolescent Health*, 26, 1, 64-69.

Jamieson, N., Fitzgerald, D., Singh-Grewal, D., Hanson, C.S., Craig, J.C. & Tong, A. (2014). Children's experiences of cystic fibrosis: a systematic review of qualitative studies. *Pediatrics*, 133, e1683-e1697.

Punch, S. (2002). Interviewing strategies with young people: the "secret box", stimulus material and task-based activities. *Children & Society*, 16, 45-56.

Quittner, A.L., Saez-Flores, E. & Barton, J.D. (2016). The psychological burden of cystic fibrosis. *Current Opinion in Pulmonary Medicine*, 22, 187-191.

ScotCRN (2012). *Obtaining informed consent for clinical research in children and young people under 16 in Scotland* [PDF file]. Retrieved from: [http://www.scotcrn.org/documents/ScotCRN Guidance Documents/ScotCRN informed consent 7.08.12 V1.pdf](http://www.scotcrn.org/documents/ScotCRN_Guidance_Documents/ScotCRN_informed_consent_7.08.12_V1.pdf). (Accessed: 4 December 2016).

Smith, J.A. & Eatough, V. (2007). Interpretative phenomenological analysis. In E. Lyons & A. Coyle (Eds.), *Analysing qualitative data in psychology* (pp. 35-50). London: SAGE.

Smith, J.A, Flowers, P. & Larkin, M. (2009). *Interpretative Phenomenological Analysis: Theory, Method and Research*. London: SAGE Publications Ltd.

Sturges, J.E. & Hanrahan, K.J. (2004). Comparing telephone and face-to-face qualitative interviewing: a research note. *Qualitative Research*, 4, 1, 107-118.

Tluczek, A., Laxova, A., Grieve, A., Heun, A., Brown, R.L., Rock, M.J.,...Farrell, P.M. (2014). Long-term follow-up of cystic fibrosis newborn screening: psychosocial functioning of adolescents and young adults. *Journal of Cystic Fibrosis*, 13, 2, 227-234.

Veale, A. (2005). Creative methodologies in participatory research with children. In S. Greene & D. Hogan (Eds.), *Researching children's experiences: methods and approaches*, (pp. 253-273). London: SAGE.

World Health Organisation (n.d.). *Adolescent Health*. Retrieved from: http://www.who.int/topics/adolescent_health/en/ (Accessed: 27 November 2016).