PARENTS' UNDERSTANDING OF CURRENT AND FUTURE TREATMENTS FOR CYSTIC FIBROSIS AND THE IMPLICATIONS FOR THEIR CHILD'S FUTURE

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The candidate confirms that the work submitted is her own and that appropriate credit has been given where reference has been made to the work of others

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ABSTRACT

Current treatments for cystic fibrosis (CF) have largely focused on symptom management. Consequently little attention has been paid to understanding the genetics of CF, in particular gene mutations and mutation classes. Advances in treatments for CF, the most recent development being Ivacaftor, are starting to become mutation specific. It is becoming increasingly important to understand what patients with CF and parents know about the genetics of CF, in particular whether they are aware of gene mutations and classes. Until now, research exploring parents' knowledge of CF is almost absent.

30 parents of children with CF aged 5 or under participated in a survey that explored their knowledge about CF. They were asked to identify their child's CF gene mutations, mutation classes, and how much of an impact that this has on their child's health. The results of this survey found that most parents knew their child's gene mutation, though only a significant minority correctly identified mutation class. A further 7 parents took part in semi structured interviews that explored their understanding of CF in more depth, their views of current and future treatments, and future outlooks. An understanding of how much they were aware about Ivacaftor, other treatments in clinical trials, and what this means to them was also explored. Using thematic analysis, five core themes and subthemes were identified: the arrival of CF; adjusting to CF; management of treatments; approaches to thinking about the future; and what I know and feel about new developments.. The themes and implications for clinical practice are discussed.

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CHAPTER I

1.0 INTRODUCTION

1.1 Background

Cystic fibrosis (CF) is a chronic and life shortening genetic disorder (Jessup et al., 2010, Bobadilla et al., 2002) caused by a number of genetic mutations (Derichs, 2013), that is typically diagnosed in the first year of life, and with a current life expectancy of up to 33.4 years (e.g. Szyndler et al, 2005). What was once regarded as a terminal childhood disease (e.g. Carpenter et al, 2004, Jessup et al, 2010), has now, through early neonatal screening and major advances in treatments targeting the severity of symptoms, become essentially a life-limiting adult condition (e.g. Robinson, 2001, Rosenfeld, 2005). Furthermore, the advances in treatments have led to views that children with CF born in the year 2000 and onwards, may expect to live to 50 years of age or over (Dodge et al, 2007). Nonetheless, despite the treatments to slow down the symptoms, CF remains a progressive disease without a cure.

In recent years, a new generation of treatments for CF has been in development which offers the promise of massively increased effectiveness. These are qualitatively different from what has come before in that they target mutation class, and aim to prevent the condition of CF from deteriorating by disrupting the causal mechanism in CF rather than alleviating the symptoms. They will also potentially lead to a change in the way CF is viewed by teams, patients and parents. Until now treatment for CF has been broadly similar; though mutation class forms part of the medical history, it has limited implications for treatment. Because they target mutation class the new treatments will not be suitable for all patients with CF, and may potentially lead to CF being reconsidered as a cluster of related but different conditions rather than one. As ever with new treatments, and most recently seen in the early days of research into gene therapy, there may also be a mixture of beliefs held by patients and parents around the potential of this new class of treatments. At the very least there may be a realistic appraisal of the specificity of the new treatments as those patients who have a mutation class that is *not* targeted by the treatments recently brought on to the market will not benefit from them, but may need to invest hopes that development in this new line of treatment will produce a drug that does target their particular mutation class.

The development of a new approach to treatment for CF raises a need to better understand the beliefs of parents: what do they know about the genetics of CF, new treatments, and the interaction between them. The following chapter will review research that explores these issues. First, it will describe CF. Second, it will present an

overview of how CF impacts patients and parents. Third, it will examine current treatments for CF, and what we know of the beliefs about treatments of parents and patients. Fourth, it will describe advances in CF treatments and draw upon research that explores parents' knowledge of CF to speculate about how these advances may affect the way CF is viewed. Finally, it will provide a rationale for the current research and present the research aims.

1.2 Understanding Cystic Fibrosis

1.2.1 What is Cystic Fibrosis

CF is a life-limiting genetic condition that brings a wide range of physical, social, and emotional difficulties for individuals with CF and their families. Historically, it's likely that medieval accounts of 'cursed' infants' and children' whose foreheads tasted salty which predicted imminent death were cases of CF (Quinton, 1999). Other descriptions included additional symptoms of CF, for example; Busch (1990) shared Pieter Pauw's (1595) autopsy of a 'bewitched' child whose pancreas were 'hard', 'swollen' and 'gleaming white'. These descriptions, along with others over time, eventually fed the emerging diagnostic criteria, and most notably the sweat test for CF (Hull, 2003). Cystic fibrosis was first seen as a genetic disease in 1946 (Anderson et al, 1946), and the gene causing CF was officially identified in 1989 (Rommens et al, 1989).

1.2.2 The science of CF

CF is an inherited gene disorder present in children born to parents who are both carriers of a faulty gene. Individuals must inherit two copies of the faulty gene to develop CF (Thompson and Harris, 2008). Some individuals may inherit only one copy and are said to be 'carriers' of the CF gene although they do not go on to develop CF and associated symptoms. The CF gene is located on the arm of chromosome 7 and is responsible for the production of a protein known as the Cystic Fibrosis Transmembrane Conductance Regulator (CFTR). In healthy cells CFTR acts as a protein chloride channel and is responsible for the transport of sodium, chloride, and bicarbonate between cells (Conway, 2008). The presence of the faulty gene causes a disruption in how CFTR is manufactured and functions. CFTR is located on the surface of epithelial cells lining major organs including the lungs, sweat glands, pancreas, and digestive systems (e.g. Davies, 2013). Consequently, CF can be viewed as a multisystem condition causing a range of symptoms.

1.2.3 Symptoms of CF

Symptoms of CF vary between individuals however a common problem is the severe decline of salt and water movement, which in turn leads to the secretion of mucus. The loss of function in the CFTR affects organs in different ways. Two major systems that are significantly affected by CF are briefly described below:

<u>Lungs</u>

In the lungs the defective CFTR creates a loss of chloride movement and an increase in sodium reabsorption across the airway cells. The imbalance of chloride and sodium results in a build-up of mucus preventing the cilia hairs lined on the surface of the airways from continually pushing mucus up and out of the lungs. In turn, the airways become drier and stickier, creating a hospitable home for bacterial infections. Consequently CF individuals are prone to breathing difficulties, chronic chest infections, and bronchiectasis (scarring). The most common cause of death for an individual with CF is the respiratory infections that result in non-repairable damage to the lungs (Conway, 2008).

Gastrointestinal tract and nutrition

Pancreatic insufficiency, a common problem for CF individuals that often leaves them susceptible to developing infections (e.g. Glasscoe et al, 2008), is present in 90% of the CF population (Kriendler et al, 2010). CF individuals have an inability to produce enzymes involved in the digestion and absorption of food leading to poor growth, digestive symptoms, and malnutrition (Littlewood et al, 2006). Healthy lung functions are associated with early good nutritional status and growth (e.g. Peterson et al, 2003; Pedreira et al, 2005) and dietary management is therefore a vital part of care management for patients with CF.

1.2.4. CF-related co-morbidities

As CF patients move from childhood to adulthood, a greater number of CF related co-morbidities develops including diabetes, bone and joint disease, and kidney disease (Quon et al, 2012), with diabetes being the most prevalent co-morbidity (Cystic Fibrosis Foundation Patient Registry, 2010). CF-related diabetes is also associated with a number of other health complications including pancreatic insufficiency, liver disease, and more severe pulmonology disease (Marshall et al, 2005). Consequently, CF physicians additional challenges to manage not only CF itself but other related health complications as patients move into adulthood.

1.2.5. Mutation classes of CF

Since the identification of the CFTR gene, over 1500 mutations have been identified each of which affects CFTR functioning and the presentation and severity of CF in individuals (e.g Zielenski & Tsui, 1995). Zielenski & Tsue (1995) initially proposed five groups of mutations that result in the development of CF, however in more recent years, research suggests 6 classes of mutations (e.g. Pettit, 2012). The identification and understanding of mutation class and how they affect CFTR was hoped

to enable researchers to develop a range of treatments tailored to address both the symptoms of and underlying mutations causing CF. Table 1 is an overview of the six mutation classes in CF, detailing each class, the production and function of CFTR, and the prevalence within the CF population.

Table 1: Six mutation classes of CF

Mutation class	CFTR production and function	Gene mutation Examples	% of individuals with mutation class
I	Missing genetic information in process of making CFTR results in no CFTR gene made	R1162X G542X	10
II	CFTR folds incorrectly, and cannot move from inside of the cell to where it is needed	ΔF508 N1303K	80
III	CFTR is made and reaches the right part of the cell membrane, but the channel does not open properly (known as 'gating mutations').	G551D	4-6%
IV	CFTR reaches the cell membrane but the chloride channel CFTR is too narrow and prevents salt and water from passing through cells	R1178 R347P	2%
V	Mutations cause a reduced amount of functioning CFTR to reach the cell membrane	3489+10 kb C ⩾T	<1%
VI	CFTR reaches the cell membrane, but the mutations cause the protein to move away from the cell surface too quickly, creating a shortage of CFTR	4326delTC 4279insA	<1%

Information from Cystic Fibrosis Foundation (2010) and Pettit (2012)

The most common gene mutation is Delta F508, which accounts for 80% of CF patients in the UK (Pettit, 2012). The mutation classes can be further classified into two main groups. Within the first group are gene mutations that cause insufficient CFTR to be developed (mutation class I, II, and V), and in the second group (mutation class III, IV, and VI) are gene mutations where sufficient CFTR is produced and delivered to the right

part of the cell membrane, however the gene mutations prevent CFTR from functioning properly. To add to the complexity of understanding CF, both copies of the faulty CF gene can be caused by different gene mutations and consequently CF individuals may have two different gene mutations. Individuals who have one or both gene mutations from the second group are thought to have mild to moderate symptoms of CF as there is still some presence of residual CFTR (Cystic Fibrosis Foundation, 2010).

1.3 A review of the literature

The previous section presented an overview of CF and its manifestations, and the complexity underlying the condition of CF, with multiple gene mutations forming six classes of mutations. The following section draws upon research that illustrates the impact of CF on patients and parents. Although parents were the main focus in this research, previous research on patients with CF were included briefly to provide a context which enables the reader to grasp an understanding of what it is like to live with CF.

1.3.1. Adjustment to illness

Before examining the impacts of CF on individuals and their families, it would be helpful to briefly explore how people adjust and respond to illness.

The transactional stress and coping model (TSC) developed by Thompson and Gustafson (1994) has been drawn upon frequently in the chronic illness literature to explain how mothers adjust to their child's chronic condition. This model, situated within an ecological systems theory perspective, views chronic illness as a potential stressor which results in the individual and family systems seeking new ways to adapt. Two parameters have been proposed within this model that influence maternal adjustment to their child's chronic condition; the illness parameter and the demographic parameter. The former is the type and severity of illness while the latter is the age, gender, and socio-economic status. The two parameters, although are important, have been described to account for a relatively small amount of variance in psychological adjustment to a chronic condition. Guided by Lazarus and Folkman's (1984) cognitive model of stress and coping, the TSC model was amended by Thompson and Gustafson (1994) to include three psychosocial/mediating factors that influence psychological adjustment to a chronic condition. These were (a) cognitive appraisals of stress associated with completing daily tasks related to the chronic condition and expectations of efficacy in completing those chronic illness tasks (Bandura, 1977), (b) coping methods, and (c) family functioning (Moos and Moos, 1981). In the TSC, coping

methods have been categorised as either adaptive and problem focused or palliative, a combination of avoidance, wishful thinking and emotional focused coping. Both coping methods involve altering the self or the environment in response to and as part of adapting to the presence of stress, for example a chronic condition. The use of palliative coping methods had been found to be associated with poorer maternal adjustment to chronic illness (e.g. Felton et al, 1984). With regards to family functioning, the support given has been found to influence adjustment to the chronic illness, with lower levels of supportiveness being associated with poorer maternal adjustment to the child's chronic illness (e.g. Kronenberger et al, 1992, Thompson et al, 1992).

The TSC has been used widely to examine adjustment to chronic conditions including CF. Thompson et al (1993) explored 68 mothers' adjustment to their child or adolescent's CF (aged between 7-17 years). Based on the TSC model, the authors hypothesised that though all mothers might be similar in terms of the illness and demographic parameters, mothers with poor adjustment will report higher levels of perceived stress with daily and illness related tasks, lower levels of efficacy expectation, higher palliative coping and less family support than mothers with good adjustment. Mothers were asked to complete a number of self-report inventories including the Hassle Scale questionnaire (Kanner et al., 1981), Ways of Coping questionnaire (Folkman and Lazarus., 1980), The Family Environment Scale (Moos et al., 1981), and the Symptoms Checklist 90-Revised (Derogatis, 1983) for psychological adjustment. The authors found that twenty three mothers made either a good or poor adjustment to their child/adolescent's CF and two subgroups were thus formed; good adjustment and poor adjustment. The authors found no significant differences between both sub-groups in terms of illness and demographic parameters, supporting their hypothesis. Furthermore, the poor adjustment group were found to have higher levels of perceived stress and palliative coping, and lower efficacy levels of illness related tasks and family supportiveness. Interestingly, of the mediating processes, perceived stress of daily tasks was found to largely account for poor maternal adjustment. The authors suggested that routinely caring for a child with CF perhaps had made general daily hassles more stressful. Although this study is outdated, a key strength is that it utilised a number of measures to assess all three psychosocial/mediating factors included in the TSC and has provided an insight in how they can together or individually affect maternal adjustment to a child's chronic condition. The TSC model is a useful framework that highlights how individuals adapt to the presence of a chronic condition. In the context of children with cystic fibrosis, it is important to consider how the chronic condition impacts on parents' adjustment as this in turn significantly influences how the child will view and live with their condition.

1.3.2. Patients

The impact of CF in affected individuals include; physical, social, and emotional difficulties. These have been widely documented and appear to differ within different age groups. Early studies showed that depression and anxiety were found at higher rates in patients with CF aged between 5 and 19 (e.g. Turk et al, 1964; Lawler et al, 1966). At that time treatment outcomes were poor and early death was more common. Interestingly, as treatments have advanced over time, several later studies have also demonstrated that children and adolescent still experienced some difficulties, for example, with self-esteem (e.g. Sawyer et al, 1996) and mental health (e.g. Wennstrom et al, 2005). Quality of life has also been identified as being poor, particularly in younger children (e.g. Thomas et al, 2005). In addition, Bregnballe et al's (2007) study with 43 Danish children with CF aged between 7–14 years using the Beck Youth Inventory (BYI) of emotional and social impairment found high levels of anxiety in children aged 7-10 compared to healthy children. The authors suggested that children aged 10 -14 were less anxious because they had a greater understanding about CF from attending classes at their CF centre where they learn about CF and management of the condition.

In the CF literature, research has provided inconsistent findings on the prevalence of depression and anxiety, and furthermore adolescents and adults have tended to be the focus rather than children. A number of studies have highlighted that compared to the general population, rates of depression is significantly higher in adolescents and adults with CF ranging from 11 - 14.5% and 29 - 46% respectively (e.g. Birmaher et al, 1996; Burker et al, 2004). In contrast, other studies such Modi et al (2011) found that compared to normative data, there were lower rates of depressive symptoms in adolescents and adults with CF. With regards to anxiety, normal levels in adolescents and adults with CF have been identified (e.g. Bregnballe et al, 2007; Havermans et al, 2008), whilst other studies have reported higher levels (e.g. White et al, 2009; Modi et al, 2011). Research has also found that there are differences in rates of depression and anxiety between adolescents and adults with CF. The International Depression/anxiety Epidemiology Study UK (TIDES-UK) by Duff et al (2014) explored the prevalence of depression and anxiety in 2065 adolescents (aged 12-17) and adults (>18 years) with CF across 39 CF centres in Europe. Participants were asked to complete a HADS in routine outpatient appointments, and scores were then compared to normative scores. The authors found that adolescents with CF scored lower on depression and anxiety compared to adults with CF, although adults with CF had similar scores to the general population. The authors attributed increased scores of depression in adults with CF to a decline in health as age increased and resulting in unemployment.

Similarly, with anxiety, the authors suggested that as adult participants aged, their levels of anxiety increased, particularly among those who were not able to work due to poor health. In terms of the low rates of depression and anxiety seen in adolescents with CF, the authors suggested that this may be accounted for by increased clinical psychological care provision in CF centres that allows greater detection of depression and anxiety in patients and are given support. TIDES was the first study that explored depression and anxiety across a large sample of patients with CF from different CF centres in Europe, and has provided an updated understanding, although the picture regarding children remains unclear.

More current research has moved beyond examining the negative physical and emotional difficulties that result from CF, to exploring how the identity and perception of life of people with CF is constructed and whether this changes over time. Moola at al (2012) conducted qualitative research with two CF adolescents; the narrative of one of the participants described how as a child he was 'uninformed and apathetic' about CF, and obeyed the instructions related to his treatments issued by his parents without question. He also said that as a child he had no plans for his future, such as attending university, and instead imagined that he would continue to live with his parents for the duration of his life for fear of being exposed to health risks. This participant's perception of life when he was a child radically changed upon participating in a programme called CF Chatters[1] (See Moola et al, 2011 for a more detailed review) where he learned the importance of actively caring for his health, and developed a large social network (Moola et al, 2012). Moola et al (2012) found that the participant's description of himself had changed from a child who was 'passive' to a person with ambitions and plans for his future. Other research has demonstrated similarly positive developments linked with the current generation of CF patient's experiences in adulthood, finding that, in general, as CF patients grow older, they develop meaningful relationships, and enjoy a range of employment activities (e.g. Duncan-Skingle and Pankhurst, 2001).

1.3.3. Parents

Cystic fibrosis' effects are not limited to individuals with CF, but extend to their families and, in particular, their parents. Parents of children with CF have been shown to

[1]CF Chatters is a 6 week physical activity programme, which was developed by Moola et al (2011) as a pilot programme to explore youth with CF's attitudes towards physical activity. It sought to educate participants about the importance of physical activity and appreciate the value of it. See Moola et al 2011 for a detailed review.

experience high levels of stress, and concerns which are related to the treatment regimens, the impact of CF on family relationships, and the difficulty acknowledging and accepting that CF is a terminal condition (Levers et al, 1996). High levels of anxiety, and insomnia (e.g. Sawyer et al, 2000), maternal depression, marital discord, and divorce rates (e.g. Carew, 2001) have also been identified. Furthermore, parents reported experiencing a high level of morbid thoughts (e.g. Ingersky et al, 2010).

In the words of Glasscoe et al (2008), 'the diagnosis of CF in a child is a major life event for parents'. Parents have described the diagnosis of CF as devastating, and that led to a complete change in their life (e.g. Carpenter et al, 2004). Although treatments have increased the longevity of their child's life, the diagnosis of CF remains a huge source of distress for parents (Carpenter and Narsavage, 2004). Parents' former identity often becomes compromised when they enter the role of caring for their child with a chronic condition, including CF. The aspects that previously may have helped to construct a parent's identity such as friendships, job, and time in a relationship becomes lost as they dedicate their time to their child's condition and ensuring treatments are followed (e.g. Hodgkinson et al, 2002). Other areas of research has highlighted that parents' perception of their child, and their relationship with them, can be affected by the presence of CF. For example; in their qualitative study Grob (2008) shared a mother's description of the time she had learned that her son had CF. She described that he was no longer a healthy baby, but 'a *sick child who needed to be treated'*, and this meant that she *'had to learn everything about him, how to treat him, and how to do physio'* (Grob., 2008, pg 1059).

Research is unequal in exploring fathers' views compared to mothers' views, in that fathers' perspectives have not been as well explored (e.g. Hayes et al, 2008). The existing research shows that fathers of chronically ill children describe more stressful events which include changing their identities, social and personal relationships, and experience a lower self-esteem than fathers of healthy children. They also report experiencing a range of emotions frequently including; anger, anxiety, and guilt (e.g. Katz et al, 1999; McGrath et al, 2004). In contrast, mothers are more frequently studied in the literature on CF. They have been found to experience a significant reduction in their interactions with others (e.g. Giess et al., 1992), and higher levels of stress, as they consistently battle with keeping life as normal as possible whilst managing their child's condition (e.g. Steward et al, 1994; D'Auria et al, 1997). Mothers may also experience denial in the face of their child with CF. For example; in an early study Tropauer et al (1970) found that mothers who were informed that their child had a prognosis of living up to a further 5 years believed they would make a complete recovery. Although some of these studies may not be directly relevant to the current study, they highlight the

importance of updating our current understanding about how parents cope with their child's CF, and in turn how they view their child's treatments, and their future.

1.4 Treatments for CF

The first part of this section will examine current developments in CF treatment and provide an overview of key treatments for patients with CF, before focusing on the beliefs about them. The second part will draw upon qualitative research that examines the further impacts of living with CF and associated treatments, and future outlooks.

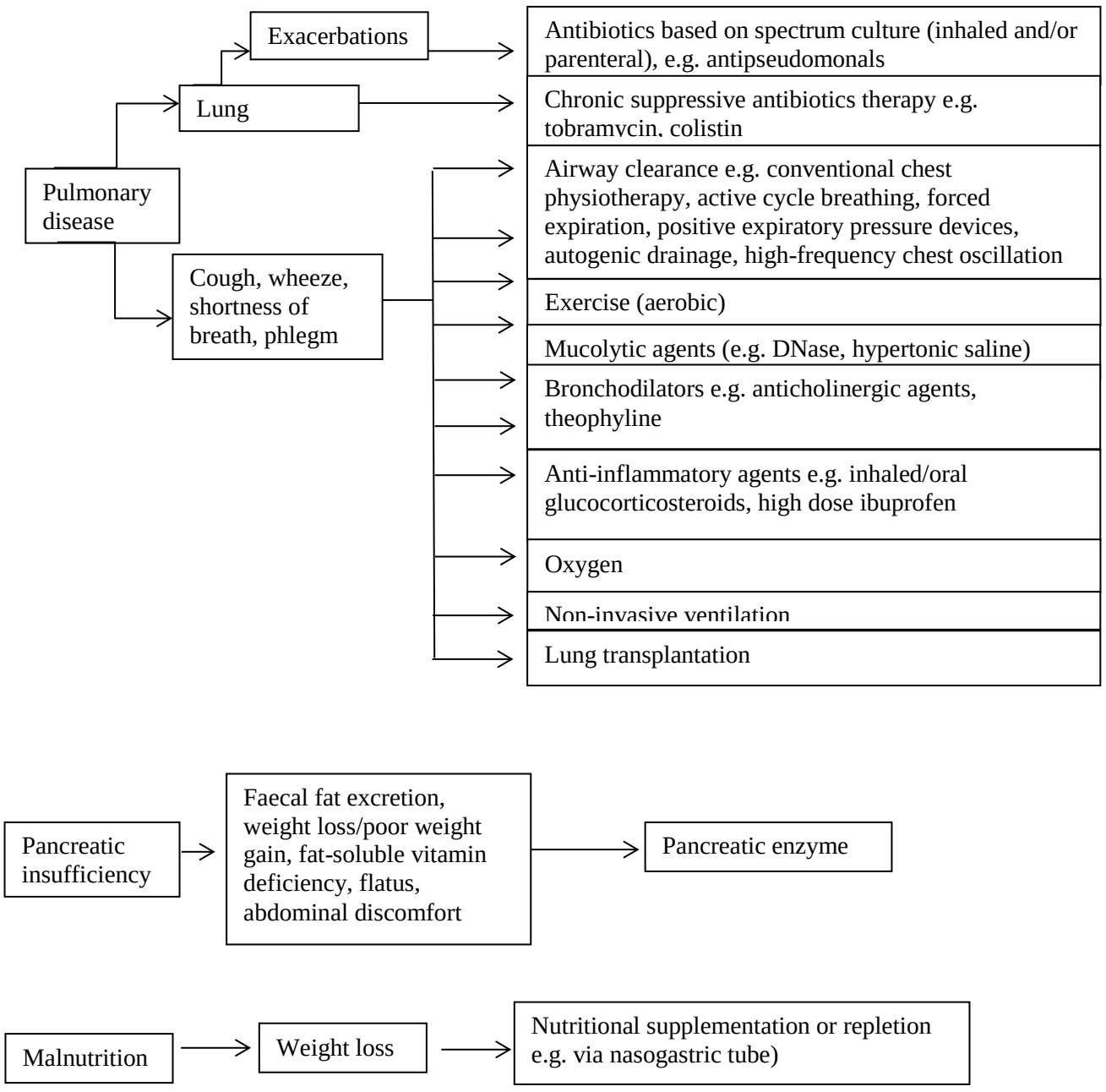
1.4.1 The complex set of current treatments for CF

Current modes of treatment target the secondary effects of CF. They have been remarkably successful, leading to greatly increased life expectancy (Sawicki et al, 2012), though CF remains an ultimately fatal condition. Such treatments involve infection control, mucus thinning agents, dietary management with replacement enzymes (e.g. Jones et al, 2002), oral antibiotics, and physiotherapy (e.g. Llorente et al, 2008). Physiotherapy typically consists of using airway clearance techniques (ACTs) such as Chest Physical Therapy (CPT) and Postural Drainage and Percussion (PD&P) where the chest is tapped continuously to dislodge and move mucus out of the lungs and in turn enable for easier breathing.

Sawicki et al (2012) provided an overview (Figure 1) which illustrates how CF affects adult individuals and lists current therapies to manage a range of symptoms. Although Figure 1 outlines CF therapies for adults, children also follow similar regimens (Sawicki et al, 2012). There are fewer CF related complications in childhood though these can often increase significantly with age. Some of the key treatments for children with CF involve ACTs, physical activities, inhaled medications, enzymes replacement therapy and ensuring nutritional needs are met. Treatments can be particularly demanding to parents of children with CF as they are mainly home based and responsibility falls onto them to ensure their child's adherence (e.g. Anthony et al, 1999).

Adherence to these treatments are hugely important to CF individuals maintaining a healthy lifestyle for the longer term. However, treatments to maintain health are becoming increasingly complex and burdensome. Indeed, CF chronic care guidelines recommend individuals to use between 3-5 respiratory therapies and 3-5 non respiratory therapies daily (Flume et al., 2009). Some research has explored how long it takes CF patients to complete their treatments which are discussed later on. Sawicki's (2012) review of treatments has indicated a need for better therapies where demands on individuals to engage in multiple treatments as part of caring for their health is reduced.

Figure 1 : An overview of symptoms of CF and treatments



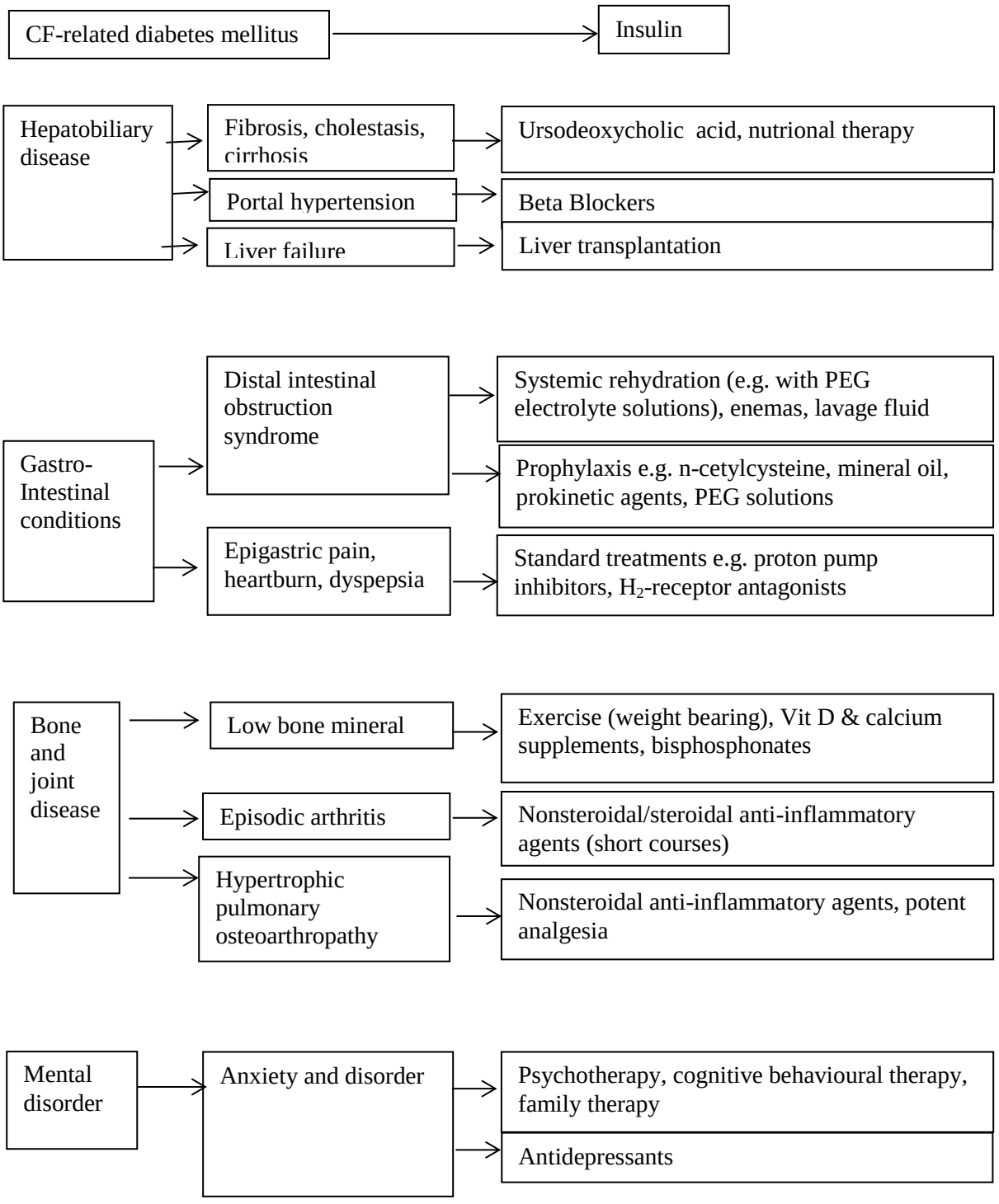


Figure 1: Extract taken from Sawicki et al (2012)

Although life expectancy is improved with current treatments, Foster at al (2001) have argued that parents now face the task of administering an increasing number of complex treatments daily to their child in order to help them live longer. This is explored further later on.

1.4.2 Beliefs about treatment and adherence

The current demands of adhering to the treatment regimen in CF is widely regarded as burdensome and demanding to the child and their family in the literature. Treatments now require regular hospital visits and substantial home care from the family (e.g. Eiser et al, 1995), and so the day to day treatment of CF has been labelled as the 'chronic burden of care' by Levers et al (1996). Glasscoe et al (2008) also quoted phrases such as 'time consuming' and 'labour-intensive' when describing current treatments, in their qualitative research with CF patients. Furthermore, some treatments, for example, chest physiotherapy, often leads to secondary and unpleasant effects such as; stomach pains, bloating, and bulky stools (Koocher et al, 1992). These effects, along with the time it takes to complete treatments can lead to a negative view of treatments by patients and their parents, and in turn can affect rates of adherence. Research has found that for children with CF, their treatment adherence rate is about 50% (Modi, 2006).

Little research on CF has focused on exploring patients and parents' beliefs about medications and treatments of chronic conditions, and as a result, understanding within this area of health is limited. Other health conditions however have been widely examined including asthma. Conn et al (2005) explored parental attitudes to asthmatic treatment (daily controller medications) and assessed the impact of this on their childrens' rate of adherence. The authors used the Beliefs about Medications Questionnaire (BMQ) among parents of 150 asthmatic children, which included two subscales that were of particular interest; necessity and concern. The authors found that although the majority of parents believed the medications were necessary for their children, a third were concerned about the medication itself, and further reported a relationship between the level of parents' concerns about the medication and their child's poor adherence. This may suggest that a child's ability to adhere to treatment could be dependent on their parents' perception of that treatment. Similarly previous studies with asthmatic children have shown that prescribed medications were discontinued by parents when they had worries about unknown possible side effects (e.g. Peterman-Sweeney et al, 2003; Handelman et al, 2004). Although Conn et al's (2005) study provides an attempt to understand the relationship between parental attitudes, concerns, and medication use by their children, CF was not featured in this research. The BMQ is becoming increasingly used in CF research to develop an understanding with

regards to how parents view current treatments for CF, and whether this impact on the child's adherence to treatments.

A small body of research that has examined CF treatments has highlighted issues around adherence in patients with CF and their families'. This research has tended to focus on adults' (with CF) perspectives on treatments and consequently little research exist that examines treatment issues from the perspectives of children with CF and their parents (e.g. O'Toole, 2012). Ziaian et al (2006) explored children and young adolescents with CF, diabetes, and asthma aged 10-16 and their parents' views of treatments. Data was collected at baseline (1 year from diagnosis), 1 year, and 2 year follow up assessments. A child and parent version of health related quality of life questionnaires were utilised as measures. Both children and parents were asked to describe all treatment tasks that the child engaged in as part of their chronic care management (i.e. time when tasks commenced and duration), and rate the level of hassle the child experienced over a period of 24 hours on three occasions per month. The authors found that a greater amount of time spent on completing treatment tasks was reported by children with CF (76 minutes) compared to 56 minutes and 6 minutes reported by children with diabetes and asthma respectively. Children with CF and diabetes also reported a higher level of hassle than those with asthma in completing treatment tasks. Ziaian et al's (2006) study has contributed to the small body of research growing on treatment burdens from the child's perspective. The authors further stated that there is need for future treatments to reduce demands on children's time. The usefulness of Ziaian et al's study could have been extended further if parents of children with CF had also rated the level of hassle in providing and/or assisting in treatment tasks to their child with CF, and this had been compared to the child's. Other research has shown that particular CF treatment tasks can take up a substantial amount of time which in turn can lead to increasing barriers to treatment adherence (e.g. White et al, 2007). For example, taking multiple medications for CF (e.g. Doring et al, 2000; Flume et al, 2007) and airway clearance techniques, which have been reported to take up to 29 minutes at a time and these are performed several times a day (e.g. Sawicki et al, 2009). CF treatments also include ensuring nutritional needs to promote growth and consequently healthy lung functions are met, which usually involve a high protein diet.

Parents of CF patients have rarely been asked for their views about CF treatments. As parents are implicated in effective treatment implementation, there is a need for research to explore this further. Previous research that has looked at this area has found that parents reported less confidence in managing nutrition-related CF issues than in respiratory therapies, and oral and inhaled medications (McDonald et al, 2009).

The next section examines further the impact of CF; particularly treatments on patients and parents using qualitative designs.

1.4.3 Living with CF and associated treatments

The literature on CF has highlighted the emotional impacts that results from CF, however some of the qualitative studies have increased our understanding further about how CF and its' impacts are constructed by those living with the condition and their families. Although research has provided little insight into how individuals with CF and their families view the treatments themselves, a growing body of research has explored the costs of adhering to treatments and has generated key themes of compromised normalcy, and temporal losses (Moola and Faulkner, 2012). A number of key studies are described below in detail.

1.4.4 The concept of 'normal'

Normalcy is compromised in those who have CF, but research has highlighted that individuals with CF strive to live life akin to those without CF. For example; in his research with adults with CF, Admi (1996) found that they did not label themselves as having CF until the arrival and experience of associated symptoms led them to finally accept that they had it. Similarly Huyard's (2008) research with adult CF patients found that the absence of symptoms resulted in them thinking that the CF did not yet exist, and their experience of normalcy was not therefore threatened. It was only when symptoms surfaced that the concept of living a normal life became compromised.

Normalcy can be interpreted in different ways and research has shown that this can influence whether patients with CF chooses to engage with their treatment, or become non-adherent. For example; Moola and Faulkner's (2012) qualitative study used a narrative framework to understand what life was like living with CF and the impact that this had on physical activity from the perspectives of six young children, though they only discussed two of the participants' narratives in detail (one male and one female). Although their research focused on physical activity related to CF, Moola and Faulkner (2012) highlighted that for the female participant, when she compared herself to her sisters (who did not have CF) she felt that she was not a 'normal' person, which caused distress and ultimately led her to reject her treatments. In addition, this participant said that her discovery that CF was genetic and fatal further reinforced her decision to reject her treatments. In contrast, the male participant, upon learning about CF and how the treatments worked, chose to engage with actively taking care of his health and took sole responsibility of adhering to his treatments. For him, the treatments became a 'normal' part of his life.

Preserving a sense of normalcy is not restricted to those with CF, but rather extends to parents of children (young or old) with CF. Earlier research, particularly Bluebond-Langner's (1979) study with parents of children with CF, has highlighted their feelings that they consistently battled against CF from 'claiming their child's body' and banishing any sense of normalcy in their lives. When CF threatens an individuals' sense of normalcy, the striving to attain normalcy in their lives becomes significant, and is often fought for. Carpenter and Narsavage's (2004) qualitative study with families of children with CF identified a model of 'falling apart, pulling together, and moving beyond'. Within this model, participants felt that regaining a sense of normalcy was important in the process of adjusting to their child's CF. Normalcy was fought for by ensuring they learned everything about CF, became familiar with treatments, and planned routines consistently around treatments. Even then, there appears to be a conflict with how much normalcy can be allowed. For example, parents strive to create a normal life for their child with CF, and yet place social restrictions on them by not letting them play with other children to prevent exposure to germs that could result in fatal infections (e.g. Moola, 2012).

Other research has shown parents making efforts to construct treatments for CF as 'normal' and as part of the individual and family's life. The parents in Hodgkinson et al's (2002) study found that by integrating their children's treatments into their daily routine, normality could be resumed. Similarly, in their study, Glasscoe and Smith (2008) recruited a sample of four mothers of children with CF aged from 7-11 years old who consented to be interviewed in depth for a series of case studies (published in 2008 and 2011). The authors were interested in the mothers' caregiving experiences and explored their concepts of what constituted treatment burden using a qualitative approach. The transcripts were analysed using interpretative phenomenological analysis (IPA). From the case studies, one particular study that attracted the authors' attention was that of the participant ('Jacqui') who was a CF specialist nurse, and who later gave birth to a child called 'Daniel' who was diagnosed with CF. The authors felt that the unusual position of a woman who had both a specialist's knowledge of CF and a caregiver's experience warranted further exploration. Glasscoe and Smith (2011) explored some of the challenges that were involved in parenting a child with CF, and identified themes that related to the child's treatments. The authors described that because the nature of CF meant that treatments needed to be carried out relentlessly, for Jacqui and her son it became 'normal' in their lives. Jacqui further felt that because Daniel was born with CF, he had nothing to compare his life to, and as far as he knew and believed, his life and the treatments were 'normal' and part of 'his routine'.

1.4.5 The meaning of time and disruptions caused by treatments (temporal losses)

Research suggests that the demands of current treatments for CF has led to a loss of freedom, and for patients' and parents' concept of time to take on a different meaning. In their qualitative study, Jessup et al (2010) interviewed children, adolescents and adults with CF and their parents on the challenges of living with CF. Together, the participants discussed their perception that the condition demands their attention constantly, particularly with making sure the medications and treatments are taken religiously. They used the metaphor of a fight - CF needed to be fought against relentlessly through ensuring they took their treatments in a consistent and daily manner; this fight was not a *'Monday to Friday one, with weekends off'* (Jessup et al, 2010, pg. 357).

Moola's research (2012) provides further insight into the temporal losses associated with chronic conditions by examining the similarities and differences in care giving experiences of 16 parents of children with congenital heart disease (CHD), and 13 parents of children with CF. The children's ages ranged from 10-18 years. The parents participated in semi structured interviews that were then analysed using thematic analysis. Participants were included in the analysis and generation of themes, which in turn increased the validity of the findings reported. Three themes were identified; presence of stress, temporal losses (that resulted from engaging with time consuming treatments), and learning to put things into perspective. Of particular interest, Moola's (2012) research highlighted the impact of treatments for CF on 'time'. Participants of children with CF described that time - in particular the luxury to do as they please and live life spontaneously - was taken away by CF. Time was instead spent on ensuring their child completed a daily set of complex and time consuming treatments. Although treatments were viewed as important, they inevitably interfered with the time that could have been spent on family activities. Participants' description of treatments in the study conveyed feelings that treatments always come first, and demand to be prioritised over anything else. It further suggested that participants did not perceive time as fluid, but rather in strict segments where tasks related to their child's CF were required to be completed. This study has helped to consider beyond the initial and often the obvious impacts of CF (e.g. lung infections, physical differences) by exploring more broadly the difficulties that CF brings within a family.

Interestingly, Moola's (2012) study also found that parents of children with CF felt the situation was often made bearable when they compared their child's condition to other chronic conditions, such as childhood cancer. One parent described CF *'as the best fatal illness that you could have'* (Moola, 2012, pg. 220) because unlike cancer, for example, there was no chemotherapy, or radiation therapy. Although CF treatments were

viewed as demanding and needed to be a part of one's daily life in Moola's (2012) research, the parents still perceived them as conferring freedom in terms of completing the treatments in the privacy of their own home.

1.4.6 The impact on how children's futures are viewed

There is a small body of research that explores how the future is viewed by patients and parents of children and adults with CF. These studies have used qualitative methodologies with grounded theory or interpretative phenomenological analysis, as it enables for in-depth and rich narratives to be gained that are viewed as *'unfettered from researchers, clinicians and healthcare professionals'* (Glasscoe et al, 2008).

The Glasscoe and Smith (2008, 2011) case study series explored mothers' caregiving experiences, and in their first paper, Glasscoe and Smith (2008) examined how a mother viewed her child's future. They found that even when their child was well the mother always felt cautious about the future, knowing that CF is never gone, but *'is an ominous lurking presence'*. She further stated that she could not plan too far ahead because of the uncertainties that the condition brings. Other studies from fathers' perspectives have also presented similar themes, where fathers have described avoiding thinking about the future because they did not want to think about the illness deteriorating further (e.g. Peck et al, 2005). Glasscoe et al's (2008) study had presented a detailed insider perspective of what it is like to parent a child with CF, and has highlighted how restrictive the condition can be on a person's life. A particular strength of this research was that the researchers worked collaboratively with the mother in analysing the interview she gave about her son, adding 'truth' in the findings that this study presented.

In another qualitative study, Jessup et al (2010) explored the issues associated with living with CF from the perspectives of children, adolescents, young adults and parents. The transcripts were analysed using a phenomenological perspective by the researchers. A number of themes emerged including 'fear'. Parents described the fear of their child's death as 'loitering' in the background, with one father stating that CF will *'get you at some stage'*. The researchers' findings conveyed the uncertainty that parents felt about their child's future. Furthermore, a theme centred on 'future' emerged and the researcher shared the parents' descriptions, where they stated the future for their child was immediately lost at diagnosis, rather it became dependent on the treatments available and their child's responses to them. One of the interesting findings that surfaced in this study was that researchers observed a difference in adolescents' views of their futures, where they described their future ambitions and hopes, compared with earlier research, where adolescents had expressed an ambition to survive and become a

teenager (e.g. Tropauer et al., 1970). It is likely that the difference in these expressed views regarding their future may be related to the advances made in treatments that have improved life expectancy.

1.5 Advances in treatment

The previous section explored current treatments, views surrounding them and future outlooks. This next section briefly reviews research on gene therapy before describing a major new advance in CF treatment and explores its implications. Focusing on mutation class, which is crucial to the new treatment, this section goes on to examine what parents know about the genetics of CF.

1.5.1 Gene therapy

Since the identification of the defective CF gene, gene therapy was initially considered to be the most promising intervention to address the underlying cause of CF (Derichs, 2013). Gene therapy aims to replace the defective gene causing CF through using viral and non-viral delivery methods (Griesenbach et al, 2012). Both methods share a common goal to insert genetic material into a host cell with instructions to introduce a sequence coding for a normal CFTR (e.g. Derichs, 2013).

The promise is great, but there are several complications which explain the slow progress of gene therapy to date, including barriers to successful gene transfer (Derichs, 2013). CF patients have a thick layer of mucus on their airways and on the lung surface area making it difficult to transfer viral and non-viral vectors into the lungs (e.g. Lethem et al, 1990 and Kitsom et al, 1999). Although early clinical trials have demonstrated successful gene transfers using both delivery methods (e.g. Davies et al, 2010, Derichs, 2013), both were also found to have side effects. Davies at al (2010) provided a summary of clinical trials conducted so far and their outcomes. Some of the earlier outcomes included a patient who became unwell with fever and hypoxia following a high dose viral vector administration. Clinical trials with lower doses were later conducted but they were also found to be unsuccessful (Davies et al, 2010). This resulted in a further set of clinical trials that have attempted to administer low doses of viral vector repeatedly, but these too have had limited success for example, Harvey et al (1999) found that administering a viral vector with CFTR sequence code three times over nine months resulted in a diminished ability to produce copies of normal CFTR.

Other complications preventing successful gene therapy include identifying which cells are best to target CF in terms of ensuring the vector successfully attaches itself to the cell and results in maximum clinical benefit to patients (Davies et al, 2010), as well as ensuring that the gene expression once a viral and/or non-viral agent is

successfully attached to a cell works. Research has highlighted that the instruction to introduce a sequence coding for a normal CFTR (also known as gene expression) has generally not been long lasting enough to produce copies of a normal CFTR gene to work effectively (Davies et al, 2010).

At present, the UK CF consortium is supporting a gene therapy programme investigating whether a particular non-viral agent will be successful (e.g. Griesenbach et al., 2012). In general, however, the early promise of a relatively rapid and safe cure for CF that raised hopes of patients, parents, and treating teams has not as yet been met. In fact gene therapy research on CF has slowed, partly for the reasons mentioned above and partly because of the amount of time and expense needed to conduct clinical trials (Derichs, 2013). It likely that gene therapy still requires extensive development before it can be used successfully. Meanwhile, other types of interventions aimed at addressing the cause of CF rather than treating symptoms have been found to have more success, and the main one that is attracting a great deal of attention in the CF literature is described below.

1.5.2 Ivacaftor

A radical new approach to treatment has been developed that targets the malfunctions in cellular processes in patients with CF. The first of this new generation of drugs is known as Ivacaftor (Kalydeco in Europe), named VX-770 during development. Ivacaftor is an orally administered CFTR potentiator. It is designed primarily to target gene mutation G551D, a mutation class III which causes the CFTR protein channel to misfold when it reaches the surface of cell membranes and prevents the channel from opening properly to allow sufficient amount of salt and water to pass through cells (Davies, 2013). Once the CFTR protein channel reaches the cell membrane, Ivacaftor binds to the channel to induce a non-conventional mode of gating that allows chloride ions and fluid to pass cells through the channels. The effect of this is to prevent mucus building up, enable the individual to breathe unhindered, and become significantly less vulnerable to developing lung infections. It is estimated that 4-6% of the CF population has this type of gene mutation (Cystic Fibrosis Foundation, 2010) and a few will therefore benefit (though the largest concentration of CF patients with gene mutation G551D is in Ireland and the UK).

1.5.3 Effectiveness of Ivacaftor

There is a growing body of research on the effectiveness of Ivacaftor. Coghlan (2011) found that 161 CF individuals with mutation class III had improved lung function by 20% compared to those who were given placebo. A more recent and detailed study

by Davies et al (2013) examined the effectiveness of Ivacaftor on children between 6 and 11 years of age by assessing whether there were any changes in participants' forced expiratory volume (FEV_1[2]), body weight, sweat chloride concentration (a measure of CFTR function), and reported respiratory symptoms using the CFQ-R. The authors also evaluated the safety of patients using Ivacaftor. The study was restricted to participants who had CF, had G551D on at least one of the allele, had a body weight greater than or equal to 15kg and a FEV_1 of 40-105%. A total of 52 participants took part, with half the sample randomly assigned to self-administer Ivacaftor orally at a dose of 150g every 12 hours, and the remaining half to take a matched dose of placebo over a course of 48 weeks. All participants also continued to adhere to their pre-existing prescribed therapies. Compared to the placebo group, the Ivacaftor group were found to have a significantly improved FEV_1 and body weight, a significant reduction in sweat chloride concentration, and fewer participants' respiratory symptoms reported. The authors identified that the changes were evident in the early stages of taking Ivacaftor and remained stable throughout. Although the authors have highlighted the effectiveness of Ivacaftor in reducing symptoms of CF, and with the younger age group of the CF population, they also found side effects of Ivacaftor. These included headaches, pulmonary exacerbation, and diarrhoea. Davies et al (2013) concluded that although their research was conducted with a small sample of participants and for a limited time, further research is warranted and on a longer term to investigate the safety of Ivacaftor particularly if CF individuals are encouraged to self-administer Ivacaftor for the duration of their life.

Ivacaftor has nonetheless been approved in the United States and has recently been approved by NICE guidelines in the UK (in January 2013). The approval of Ivacaftor will potentially mean a complete change in the current treatments of CF. Ivacaftor is promising and offers the hope of a major advance in treatment for CF, highlighted by the fact that patients receiving the treatment subsequently fail the sweat test, the main diagnostic test of presence of CF. The knowledge that Ivacaftor will only target and work for a small number of the CF population, however, highlights the importance of exploring what parents know about the genetics of CF, and whether they are aware of different mutation classes, which is discussed below.

[2] FEV_1 is a measure of the ability to force air out of the lungs; in healthy people, the average FEV_1 is between 75-80%.

1.5.4 What is known about the genetics of CF?

Little research has focused on parents' knowledge and understanding about CF, in particular, their awareness of the potential importance of the different classes of mutations. Only two studies have explored parents' detailed knowledge about CF.

McCrae et al, (1973) interviewed 50 parents in Scotland and 50 in Northern Ireland with a view to ascertain the level of knowledge they had about CF. They found that 71 of the 100 parents showed a 'poor understanding', which was defined as knowing only that CF was an inherited disorder. The researchers had highlighted lower levels of intelligence and education, poor diagnosing by physicians, and psychological barriers as possible reasons for the parents' 'poor understanding'. In contrast, a later study by Nolan et al (1986) explored the knowledge of parents and patients with CF who were between 10 to 21 years old. The researchers developed and administered a questionnaire that assessed knowledge about disease pathophysiology, associated treatments, genetics, and reproduction. The researchers found that the patients and parents had a reasonably good knowledge about the disease pathophysiology and associated treatments. However, they demonstrated a 'poor' knowledge of the genetics of the condition and issues with reproduction.

These studies suggest detailed knowledge of CF might be limited in patients and parents, though both are outdated and arguably do not represent an accurate picture of current parents' knowledge. A clearer, and more contemporary understanding may be helpful to healthcare professionals in considering and anticipating how parents will respond to the prospect of a new generation of treatments, particularly when that parent's child may not be immediately eligible for them.

1.6 Summary and rationale for current study

CF is an inherited, life-limiting condition with a known genetic cause. Research has made us aware of the difficulties that individuals with CF encounter in managing the symptoms associated with the condition; for example, persistent coughing and malnutrition. There are also social and emotional impacts that result from having CF and these affect the families and, in particular, parents who enter the role of a caregiver, and need to find a way to cope with knowing that they have potentially limited time with their child.

The current treatments for CF work by reducing the physical symptoms caused by the condition, and these have been remarkably successful in increasing life expectancy. Nevertheless, children and young people still die each year from the condition, and life for those with CF, however much it is prolonged by recent

developments, remains limited. Similarly, the emotional and social impacts of CF continue for the child and their parents. Current treatments also come with a cost, as research has highlighted that treatments actually exacerbate some of the emotional and social impacts of the condition. For example, freedom, independence, and time taken away from other activities as patients and parents attempt to follow the complex daily treatment regimens. Given how treatments affect individuals and families, it is not surprising that adherence rates are generally low.

Ivacaftor has the potential to fundamentally change the lifestyle of an individual with CF, and their parents. It stands out from the current treatments as it is the first to target the underlying cause of CF, prevent the further decline of the condition, and (potentially) significantly increase life expectancy. Although not yet clear, there is also the prospect that by targeting the mechanism that causes secondary complications in CF, the need for other treatments (and the associated treatment burden) may in the future be significantly reduced and quality of life increased for both the individual and their family. On the other hand, adherence to this new treatment still requires (and in some ways increases the importance of) a strict regime, and as mentioned above it may not be without side effects.

Ivacaftor looks set to benefit any individual with mutation class III, however it might be argued that it offers the most promise to infants and children who have been diagnosed with CF (and have mutation class III), but where the onset of the symptoms has not been significant. Not only will it potentially prevent the emergence of symptoms that is characteristic of an older individual with CF, but some of the emotional and social problems that CF individuals eventually experience may also be avoided. Ivacaftor therefore means that we are on the cusp of a radical change in how CF is viewed and lived, particularly for the younger generation. At present Ivacaftor is the only treatment available that works in this manner, however it has raised the prospect of new treatments that target other mutations. Even so, it is not clear whether these new generations of treatments will work for other mutation classes, or whether or when they will be developed. As ever with new treatments, it becomes important to consider the wider impact on patients and families. Some will hopefully benefit directly from the new treatment, and others may have hopes and fears raised. One important aspect is that the new treatments target specific mutation classes, and therefore an understanding of the genetic basis of CF has become much more important than before. In particular, patients, parents and even the multi-disciplinary teams that treat CF have generally seen CF as one condition. The new treatments may potentially change that.

1.7 Introducing the current study

This current study aimed to contribute to research on CF by exploring parents' knowledge and understanding about CF (particularly the genetic characteristics influencing the presentation in their children), beliefs about current treatments and what they mean for their child's future, and also to explore the impact of knowledge of the new developments in treatments for CF including on adherence to current treatments. Although this research planned to focus on both parents' of children with CF, it recognised that mothers have been placed as their child's main caregiver within the literature and therefore it anticipated that much of what is understood about the genetics of CF and views of current treatments was likely to come from mainly mothers.

1.7.1. Research questions

The current study had three research questions:

1. What do parents know and understand about CF and in particular the genetic characteristics influencing the presentation in their children

2. What are parents' beliefs and views about current treatments and what do they perceive their children's' futures to be like

3. Does parents' knowledge of new developments in treatment for CF changes their beliefs about their child's CF and the importance of current and future treatment.

CHAPTER II

2.0 METHOD

2.1 Design

A mixed design was considered most suitable to understand what parents knew about CF, and to qualitatively explore their feelings towards current and new treatments. The design of this research involved a newly constructed questionnaire administered to a sample of parents of children with CF, and qualitative semi structured interviews with 7 parents. Questionnaires were analysed using descriptive statistical analysis. Qualitative data were transcribed verbatim and analysed using Braun and Clark's (2006) thematic analysis guidelines. The themes are presented in a table, with each theme explained.

2.2 Methodological Considerations

Both descriptive statistical analysis and thematic analysis were used for data analysis. This section will provide a summary of why a mixed design was used before describing the qualitative approach utilised and the rationale for its use.

2.2.1 Using a mixed design

Psychological research in CF has typically used either a quantitative design (e.g. Szyndler et al., 2005, Llorente et al., 2008) or qualitative design (e.g. Hummelinck et al., 2006, Grob., 2008), with very few employing mixed methods (e.g. Bywater., 1981). Quantitative and qualitative designs on their own provide different benefits. One of the greatest benefits from using a quantitative design is that the researcher is 'distanced' from participants and therefore any data collected can be said to be from an objective viewpoint. Furthermore, quantitative designs can allow for easy comparisons with other studies to be made (Kruger, 2003). They are not without limitations however as they can arguably provide a narrow and 'superficial' level of data.

Qualitative designs compensate for this as they can provide detailed narratives and/or generate theories about any phenomenon explored. It is argued that greater understanding about human experiences can be achieved from qualitative designs (Denzin and Lincoln, 2005) as they enable rich descriptions from participants to be generated (Smith, 2008), and allow researchers to explore how participants make sense of their experiences, for example living with a chronic illness (Willig, 2008). With qualitative designs, the researcher's personal attributes and skills in data collection and analysis heavily influence the quality of the data analysis. It becomes crucial that the

researcher conducts the analysis to a high standard. A lot of subjectivity is also placed in data analysis which can lead to generating rich descriptions of data. However, as there is no room for objectivity in data analysis, the findings are difficult to generalise.

Using both approaches can maximise the potential to generate rich and useful data. Both methods can compensate for shortcomings of the other, which is typically referred to as methodological triangulation (Jick, 1979). I felt that using a combination of a qualitative and quantitative method would be of benefit for several reasons. In the first instance, using questionnaires would enable the sample to be situated with regards to how much parents in a CF centre understood about the genetics of CF. The use of semi structured interviews would further provide an opportunity to explore parents' views in depth. Together, both methods would provide a well-rounded collection of information for analyses on the research topic. Furthermore, given the limited amount of research on CF using mixed approaches, I felt that this research would contribute to expanding that field.

2.2.2 Qualitative methods

Qualitative methods are very varied and complex (Holloway et al., 2003) and can be divided into two main clusters. The first cluster contains methods that are bound to an epistemological position such as conversational analysis (e.g. Hutchby et al., 1998), interpretative phenomenological analysis (e.g. Smith et al., 2003), and grounded theory (e.g. Glaser., 1992). These approaches follow a rigorous analytical procedure on data collected and essentially involve developing a theory that explains the phenomena explored. Within the second cluster are methods that can be independent of a theoretical position or development such as thematic analysis, and consequently can be viewed as being able to offer a greater degree of flexibility in how analysis can be conducted and interpreted (Braun and Clark 2006). In particular, the researcher is given the freedom to choose whether they wish to organise their data in a way that leads to theory development.

2.2.3 Choosing a qualitative method and rationale

Two approaches that were considered for this research were grounded theory (GT) and thematic analysis (TA). The method of analysis needed to be one that enabled the researcher to capture themes relevant to the research aims and produce a coherent account of those themes, as reported by the participants in the study. Both methods share a common feature in that they are flexible (Braun and Clark 2006), but have different requirements for how data is used, for example unlike GT, data analysis subjected to TA does not necessarily have to be organised to develop a theory. The themes from TA can

be presented as they are or organised into a theory that encapsulates the themes. After careful consideration TA was chosen as I felt that it would permit me the flexibility to explore and understand participants' stories without a demand to organise this into a general theory and potentially losing the distinctiveness of their views. TA and the process involved in using this approach are described next.

2.2.4 Thematic Analysis

There has been debate about whether (TA) is a qualitative 'method' in its own right, or whether it is a technique that can be used across a range of qualitative methods (e.g. Boyatzis, 1998; Ryan & Bernard, 2000). Furthermore, some have argued that TA has been used improperly in research (e.g. Antaki et al, 2002) because there is no clear definition or guidelines for implementation (e.g. Silverman, 1993; Tucket, 2005). Braun and Clarke (2006) in response presented a detailed overview of the approach, and step by step guidelines on using TA. The authors argued that TA is the most valued method for qualitative analysis that enables researchers to analyse and identify patterns and report themes (Braun and Clark, 2006). The authors insisted that TA should be the first method that researchers learn to use when undertaking qualitative analysis as it will enable them to acquire the skills needed to use other methods such as GT and discourse analysis. As well as reflecting core skills in qualitative analysis, they asserted that TA should be perceived as a defined method for qualitative analysis in its own right. Furthermore, the authors highlighted that a major strength of TA is that other than the inductive philosophy of science that underpins both TA and all qualitative research, it is a method that is largely independent of any particular epistemology. It involves generating themes, but this activity is not explicitly linked to a need to develop theory, though some researchers may choose to do so. TA is therefore flexible enough to be applied to all areas of qualitative enquiry.

Braun and Clark's (2006) overview was predominantly used in this research as guidance in the current research data analysis. It was also used to develop my knowledge of TA and enhance my ability to produce a good piece of TA.

2.2.5 What makes a good thematic analysis

There are a number of approaches possible within TA, such as using realist and constructionist methods. These have a different focus in terms of how a researcher performs the qualitative analysis and how they might interpret data. A good TA is one where the researcher makes it clear what method within TA they have chosen and why. Braun and Clark (2006) stated the importance for researchers to clarify what qualitative analysis has been performed, and the assumptions that informed their analysis. Without

these, it is difficult to evaluate and make comparisons with other research in similar areas (Attride-Stirling, 2001). Researchers have frequently omitted the 'how' part of their analysis (Lee et al., 1996) and consequently prevented readers from gaining a full understanding of how the researcher has come to identify the reported themes.

Braun and Clark (2006) also draw attention to recognising the role the researcher played in the process of analysing data. Previous qualitative research typically describe how themes 'emerged' and in doing so it denies the active role that the researcher engaged in pulling out and identifying themes, and which further suggests that themes 'reside in data' (Taylor et al., 2001). In reality, researchers using qualitative analysis do not simply provide a voice for their participants and report their views, but instead play an active role in selecting, editing and sharing narratives (Fine, 2002). Understanding our position as a researcher and our values is therefore of critical importance before and during our engagement in data analysis as these inform how analysis is conducted and what we might find in our data (Braun et al., 2006). The decisions that the researcher makes in this process are therefore active and conscious decisions that needs to be acknowledged.

2.2.6 Researcher reflexivity

Braun and Clark (2006) described that the researcher needs to engage in a process of constant reflexive dialogue whereby they consider the decisions that needs to be made regarding their personal and theoretical position as well as the level of data analysis being performed. Furthermore, the researcher is encouraged to consistently ask questions about the data during analysis. Throughout this research, I consistently reflected on my theoretical position, personal bias, and approach in analysis. These are described next:

2.2.7 Introducing the researcher

I am 27 years old, a British Indian female working towards her doctorate in clinical psychology. I have a particular interest in health psychology and client groups who experience chronic health difficulties. At the time of completing this research I was working with clients with cardiac and pain experiences. My theoretical orientation in clinical practice tends to be person centred, compassion focused, narrative, and systemic.

As a psychologist in clinical training (PICT), meeting with individuals who present with health and/or mental health difficulties, it is my role (and interest) to listen to their stories but also to support them in thinking about how they might be able to manage their difficulties. I was aware that entering the role of a researcher means that I

would need to refrain from listening to participants as a PICT and considering with them ways to problem solve. It was important to adopt the stance of a researcher

I acknowledged my position as a researcher who was new to qualitative analysis and specifically to TA. These issues were discussed in supervision during data collection and analysis. It was essential for that I developed a thorough understanding about TA and the decisions that needs to be made during the process which influence how analysis is performed.

On a more personal level, I anticipated that mainly mothers would participate in this research and I identified myself as someone who hopes to enter motherhood in the near future. I recognised that this may influence how I perceive and interpret mothers' views, and particularly when they describe their child's future. For example I might view a parent whose child has a potentially fatal condition as being unaccepting of their child's condition and/or distraught, as I might imagine myself to be feeling this. I acknowledged the need to be open, and of being alert to situations in which I may project my own views on to those of my participants. I further recognised the importance of reflecting on my feelings about the mothers that I interviewed and the situations they described both in and out of supervision.

I am a researcher who was diagnosed with a profound hearing loss. Although my disability is very different to CF, both are life-long conditions and can have a range of emotional impacts. I was aware that potential participants might express a variety of emotions such as anger, sadness, and/or fear particularly when recalling the time when their child was diagnosed with CF, and when thinking about their future. I was also mindful that potential participants may describe times of 'accommodating' to their child's medical needs. These feelings and experiences might evoke past memories of how my parents might have felt at the time of my own diagnosis, as well as how they might have had to accommodate to my needs. It felt important to be aware of my disability in terms of how I might respond to potential participants during the interviews and how I might interpret and analyse their views about their child's CF.

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2.2.8. Decisions made in thematic analysis

During the process of developing my understanding about TA, a number of key decisions were made that guided the approach used in my data analysis. These decisions have been shared to enable the reader to understand what approach in TA was utilised.

Realist, constructionist, or contextualist approach

TA can be a realist, constructionist, or contextualist method. In essence, the former involves reporting participants' experiences and meanings. A constructionist

method focuses on discourses within a society that may be influencing participants' realities, meanings, and experiences. A contextualist method explores how individuals make sense of their experiences within a wider social context. I had a particular interest to understand parents' experiences of dealing with their child's CF, views of current treatments, and hopes for the future. I recognised that I would be entering the parents' private worlds and I did not want to lose my focus on them. I viewed the realist method as best allowing me to stay with the parents and report their experiences.

Inductive versus deductive approach

An inductive (or 'bottom up') approach to analysis using TA involves exploring data and identifying themes with no specific, a priori questions in mind. Themes are not looked for and the intention is not necessarily to create a theoretical framework. As there is no specific question, the researcher is more likely to generate rich descriptions of the entire data sets. In comparison, a researcher using a deductive (or 'top down') approach has a specific interest in the data and the data analysis is driven by them to answer a specific question in mind. The outcome of using a deductive approach is that only parts of the data are fully explored (Braun and Clark, 2006). I was keen to understand as much as I could about parents' views and experiences about CF in general, in relation to their child, and their perceptions of their child's future. Although I was keen to answer my research questions, I wanted to approach the data analysis openly, and therefore I chose to employ an inductive approach.

Semantic versus latent approach

The level at which data is analysed can be at a semantic or latent level. A semantic approach involves looking at the form and meaning of data at a surface level (i.e. what has been articulated and what that means). A latent approach in contrast focuses on the underlying ideas that gave its meaning. In essence, it is interpretative and involves exploring how that person has come to say what they said through thinking about what might have been going on for them at that time. I chose to analyse data at both semantic and latent level, staying close to what participants reported and what meanings they gave, but also thinking about why and how they have come to have said particular things.

2.3 Ethical considerations

2.3.1 Ethics approval

This research gained ethical approval by the Leeds Bradford Committee in September 2013 (Appendix A).

2.3.2. Informed consent

Participants were given detailed information about the study and were asked to sign a consent form after they have read the information sheets, and had opportunities to ask questions. The right to withdraw at any time were made explicitly clear to participants and they were informed that this would not affect the standard of care that they received.

2.3.3 Confidentiality

Each participant was asked to provide an anonymous code on the questionnaire to protect their identity and enable me to remove their questionnaire from the data analysis if they decided that they no longer wanted to take part. Participants who took part in the interviews used the same code to enable me to correspond their interview data with the questionnaire data. Interviews were audio taped and transcribed by an independent transcriber who signed a confidentiality agreement. Transcripts were password protected on an encrypted memory stick. Identifiable information was removed from the transcripts and participants were assigned a pseudonym name for dissemination purposes.

2.3.4 Distress and false hopes

Ivacaftor is designed to address mutation class three and only 4% of the CF population is believed to have this mutation. It was anticipated that most potential participants' children would not be eligible for this treatment and this may invoke negative feelings. I was conscious that I would need to sensitively manage and contain parents' distress and/or anger. Furthermore, it was viewed that some participants may hold unrealistic hopes about their child's condition if they believe that they may be eligible for it. To minimise potential distress, participants were given two information sheets (for the questionnaire and for the interview) that provided some information about Ivacaftor, the purpose of the research, and what their involvement would be if they chose to participate. Contact numbers were provided on the information sheets for participants and they were encouraged to use them if they had any concerns, questions, and/or to talk to someone about how they have felt during the study. The CF team

(including psychologists) were available for any participant who may have experienced any negative emotions.

Participants who chose to take part in the interviews were also forewarned by the researcher about the topics that might be brought up regarding CF, particularly related to their child. Those who became visibly upset during the interviews were given the option to stop. At the end of the interview, I ensured that participants felt contained before terminating the interview session.

2.3.5. Seeing the researcher as being able to help their child

At the time of completing this research, Ivacaftor was being prescribed for some children in the CF centre and it was recognised that for some potential participants may have viewed the researcher as someone who could help their child receive Ivacaftor. It was important to make clear to potential participants that my role was a researcher only with no involvement in the CF team or other medical teams. Several participants asked about Ivacaftor after the interview to which a brief explanation was provided, followed by a strong encouragement for them to liaise with the CF team for more information.

2.4 Sampling

2.4.1 Participants

Participants were parents of young children with CF. It was expected that mothers would form all or most of this group because they are viewed by the CF team as being the main caregiver for their child, and in general are more frequent attenders at clinics than are fathers. Nonetheless, it was recognised that some fathers attend the clinics and may ask to participate. Fathers would have been accepted if they wanted to take part in this research.

2.4.2 Inclusion criteria

Participants needed to be parents of children aged 5 or younger with a diagnosis of CF only, and with no other chronic illness. Furthermore, the child must have received a diagnosis of CF at least six months previously. The rationale for using parent of children with only a diagnosis of CF was to avoid the potential risk of their views and beliefs about treatments for CF becoming influenced by treatments that their child may also be receiving for other chronic conditions**.** Parents of children who were recently diagnosed (less than six months) were excluded as they may be significantly more distressed and may not have had as much opportunity to develop their knowledge of CF.

2.5 Recruitment method

2.5.1 Recruitment of participants

Parents who attended the Leeds Regional Paediatric CF Centre were recruited. At the time of this research, the team at Leeds CF centre were actively encouraging parents to develop their understanding of the causes, mechanisms, and treatments of CF, in particular the role of different mutation classes, through a computer based education programme. It was hoped that these parents would develop a more accurate and potentially more positive understanding of their child's CF and future prospects, and in turn this would encourage their child to plan their future positively, and develop appropriate educational and occupational aspirations. Although this programme had not been evaluated in this research, it is hoped that the findings from the questionnaires will provide some indication about whether parents who attend the CF centre have a good awareness about CF. Furthermore, the Leeds CF Centre were also routinely informing parents about new developments in treatments for CF, including Ivacaftor. The team were aware, however, that parents would not necessarily have processed or remembered this information. With this in mind, it was anticipated that many participants would be aware of Ivacaftor but may not potentially have a good understanding about it.

A total number of 46 parents who attended the CF centre met the inclusion criteria in that they had a child aged 5 or under with CF. A non-purposeful sampling strategy was used initially for most parents, by which parents were approached to complete a questionnaire. From there, participants who expressed a willingness to take part in an interview were recruited on a *'first come first served basis'* until enough participants were recruited. Before I describe the procedure, the measures used are outlined next.

2.6 Measures

This section describes two methods of data collections, a questionnaire and an interview schedule respectively and how they were developed.

2.6.1 Questionnaire and its' development

This research aimed to explore participants' knowledge and understanding about the genetics of CF and new treatments and it intended to achieve this through using a questionnaire. At present, there are a number of questionnaires about CF including The Cystic Fibrosis Questionnaire (CFQ), and Quality of Life questionnaire (QoL). These measures assess but are not limited to knowledge about CF, and include other areas

related to CF such as; dietary management, physical exercise, and health related quality of life. The research needed a questionnaire that specifically focused on parents' knowledge about the genetics of CF. As there were no measures designed for this purpose a questionnaire was developed by the researcher, two clinical psychologists and a consultant paediatrician. Several versions were designed and amended before a final version was agreed on. The questionnaire asked participants to identify their child's mutation gene and mutation class, whether it had clinical implications on what treatments their child had to take, and whether they were aware of new developments in treatments for CF. The questionnaire consisted of seven items and took an average of 10 minutes to complete (appendix B).

2.6.2 Semi Structured Interviews and its' development

The interview schedule was developed by the researcher, two clinical psychologists, and a consultant paediatrician. The semi structured interviews was to enable the researcher to further explore parents' knowledge about CF, views about current treatments, and the implications that these had on their outlook for their child's future. It was also an opportunity for the researcher to determine whether the participants were aware about the new approach to treatment, what they knew about it, and whether this changed how they viewed their child's future. Therefore, it was important that the interview schedule was developed with these topics in mind. The interview schedule was divided into six sections starting with exploring knowledge and ending with what they knew about the new treatment and how they perceived the possible implications (appendix C). Particular questions for each section were included to guide the interview, taking care to use open ended questions, for example;

> *'How well would you say you understand your child's cystic fibrosis?'... Can you tell me more about that…'*

2.6.3 Pilot of questionnaire and interview

A patient care advisor, who also has CF and previously attended the Leeds CF centre, was contacted through the CF trust website. She agreed to share her views about the questionnaire and interview. The patient care advisor also invited a mother of a child with CF to provide feedback. Between them, their feedback was invaluable and helped to further refine both.

The patient care advisor and the mother stated that the questionnaire should only ask questions that are factual; they felt that questions that invite participants' to express their views and feelings should only be asked in the interview to minimize feelings of

distress. This particular feedback helped to separate which questions should be asked on the questionnaire and which in the interview only.

Both the patient care advisor and the mother felt that the initial interview schedule needed to have a better ordering of the questions. For example, the original version of the interview schedule started by asking questions about what parents knew about CF and moved on to current treatments, before exploring symptoms that their child experiences. The mother and the patient care advisor felt that asking about symptoms before moving on to treatments might flow easier. The mother also identified some questions that needed to be rephrased in order to elicit specific information to address the research questions, in particular to gather information about the medical aspects of CF. An example of a question that was rephrased was *'Can you tell me what you know about how CF is caused and what the fault actually is..'* as opposed to '*Can you tell me what life is like generally with CF'* which is more of a general question and one that does not invite participants to talk about their medical knowledge of CF.

2.7 Procedure

2.7.1 Questionnaires

The clinical team at the Leeds CF centre distributed information sheet (appendix D) about the research during routine outpatient consultations and ensured ample opportunities to ask questions or share their concerns were given. Participants who verbally consented to take part in the survey were given the questionnaire to complete at the centre and they were then returned to the clinical team. The CF consultant was also available throughout to provide any support the participants may have needed. The researcher regularly attended the CF centre to collect the questionnaires for analysis.

2.7.2 Semi structured interviews

Participants who took part in the survey were immediately invited to take part in a semi structured interview (SSI) with a researcher. Interested participants were given an information sheet (appendix E) by the CF clinical team and were asked to provide their email address as a way for the researcher to contact them. The researcher contacted those potential participants and arranged to meet at their home at a suitable time. On arrival, the researcher explained the nature of the study, answered their questions, and asked them to sign a consent form (appendix F) if they were happy to take part.

Participants were contacted on a *first come first served basis* (i.e. each participant left their details, I contacted them straight after). It was initially difficult to contact potential participants, as it became apparent that most were not responding (n=10). The first four mothers who responded to the researcher's email reported time constraints in checking their email account and further stated their preference to have been contacted on their mobile telephone. This issue was raised with the research supervisors and the ethics committee that had previously approved this research were consulted. A minor amendment was applied for and granted that enabled potential participants to be asked to also provide their mobile numbers as another way for the researcher to contact them.

2.7.3 Changes to the inclusion criteria and procedure: contextual issues

There were two main issues that surfaced and resulted in changes made to the inclusion criteria and procedure. The first issue was that the sample of parents to be interviewed was shaping up to be predominantly mothers of children with the most common gene mutation (DeltaF508). A non-purposeful strategy was initially used as the researcher was keen to interview participants of children with different gene mutations including the one targeted by Ivacaftor to add variability in the sample. During the recruitment stage however, the researcher liaised with the CF team and became aware that a) most of the children who attended the centre had DeltaF508 gene mutation and b) there were only three parents of children with the gene mutation G551D and who were taking Ivacaftor. These children were also aged between 6 and 10 years old. The researcher recognised that the original inclusion criteria meant that those parents of children on Ivacaftor could not take part. As the current study aimed to explore parents' views of current and new treatments including Ivacaftor, the researcher was keen that at least one mother of a child with a G551D gene mutation was included in the sample. Therefore a substantial amendment form was submitted and approved by Ethics for the inclusion criteria to include a mother of a child aged 10 or under to take part in the interview. The CF team purposefully informed those three parents about the research and followed the procedure as outlined in sections 2.7.1 and 2.7.2 if they were interested to take part. The interview schedule was slightly amended during the interview for the participant purposefully recruited as her child was already taking Ivacaftor and I was keen to explore more about Ivacaftor itself rather than current treatments. Questions about the effectiveness of Ivacaftor for her child, side effects, and hopes for the future were more of a focus in this participant's interview. Appendix G shows the revised interview schedule (the future treatments section).

The second issue that surfaced was in the fourth interview with a mother which took place at her home on an evening. The father was also present and unexpectedly asked to take part. He reported that his work commitments meant that he had not been able to attend the CF centre as often as the mother did. Nonetheless, he was interested in the research after his partner informed him about it. The researcher had maintained from the start that fathers were welcome to participate if they wished to. The interview procedure remained the same for both the mother and father. He was given an information sheet and asked to sign a consent sheet. The researcher was careful to listen to both parents' views and had changed the interview schedule where appropriate to fit the demands of their contexts. The researcher had also planned to treat both parents' data separately in the analysis however the father was absent for most part of the interview as he needed to attend to their baby. He came back in the room occasionally and 'joined' in the interview. On no occasion did the mother's views appear to be influenced or changed by the father's views. There was insufficient data from the father to be treated separately and therefore it was treated with the mother's data in analysis.

2.7.4 Conducting the interviews

Each interview took place in the participant's home. Data collection, transcription, and analysis were conducted simultaneously. The interview schedule was used as a guide to initiate a discussion with participants around topics relevant to the research. It was intended to be utilised in a flexible manner to provide some structure to the interviews but allowing participants to move beyond initial questions asked and share their narratives. Particular questions on the interview schedule were omitted and/or adapted to fit the demands of the participants' contexts. I adopted a stance of allowing participants space in which to share their stories, and offered frequent summaries and reflections to maintain the flow of the discussions. However, from reading through the first transcript, I observed that I had a tendency to assume what participants were feeling without checking this out with them. An example is provided from the first transcript. The participant was describing examples of children with CF in the media who had done well and I shared my thoughts of how the participant viewed this or felt.

Participant:… *they've done really well and I'm really pleased for them*
Researcher: *Really inspiring*
Participant: *Yes, inspiring. Yes.*

My tendency to make unchecked assumptions about participants' experiences was discussed in supervision which led to increased self-awareness. Consequently, I became more cautious in ensuring that my interpretations were not shared in future interviews.

Transcripts from previous interviews were read before the next interview which helped to hone the interview schedule and for me to potentially inquire about emerging categories identified in previous transcripts. I also recorded my reflections of the participants and their stories. These reflections included my impressions of them and how they presented themselves. This was viewed as an important part of the data collection process, keeping the memory of each participant and their interview alive in mind when undertaking data analysis.

The following two chapters describe quantitative (chapter III) and qualitative (chapter IV) data analysis before presenting the results. They are divided into two chapters in order to enable the researcher to understand the findings.

CHAPTER III

3.0 QUANTITATIVE RESULTS

3.1 Survey

Short questionnaires were administered to parents of children with CF aged 5 or under at the Leeds CF Centre. The questionnaires were used to situate the sample and provide an understanding of how much parents knew about CF, the genetic aspect underlying symptom severity, and eligibility for the new treatment. It was also used to provide a context for the interviews which will be explored in the next chapter.

3.2 Participants

46 parents who were eligible to take part in the study were invited to take part. 30 participants consented to complete the questionnaire and all were females.

3.3 Results

Each participant's data was entered into SPSS and descriptive statistics were performed. The 7 questions asked on the questionnaire are listed below with respondents' answers provided.

1. *Knowledge of child's mutation type*

26 participants stated that they knew their child's gene mutation; 22 said Delta F508, 1 G551D, 3 other gene mutations (G542x, TRP1282X, and A89+1). 4 participants stated that they were not sure what their child's gene mutation was.

2. *Knowledge of child's mutation class*

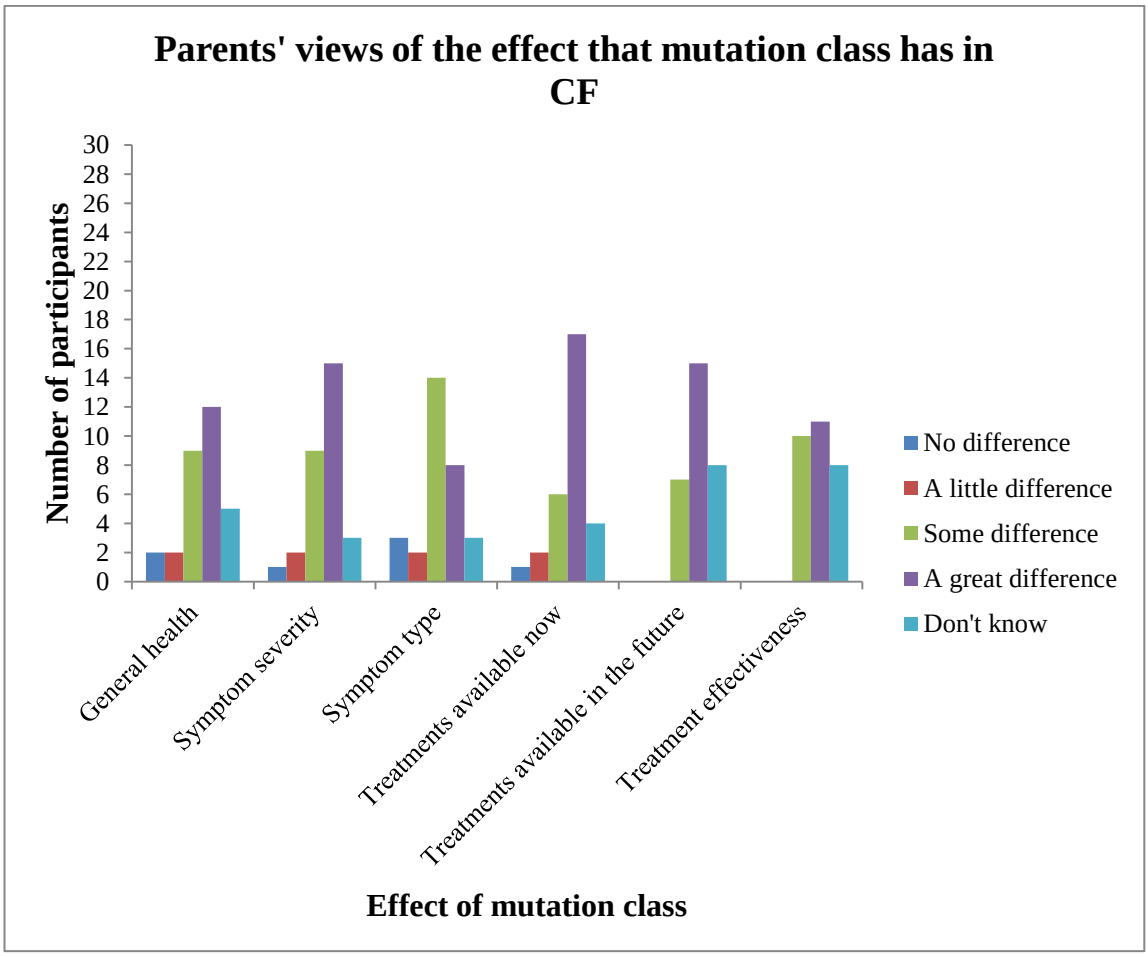
12 participants stated their child's mutation class (2 reported mutation class 1, 9 class 2, and 1 class 4). 18 participants were not sure what mutation class their child's gene mutation fell into. In terms of the accuracy of those that indicated a mutation class, 9 ticked the correct mutation class and 3 were incorrect.

3. *Effect of mutation class in CF*

Figure 2 shows how participants viewed the effect of mutation class in CF. Most participants were aware that mutation class is important, particularly for symptom severity and treatment efectiveness, though a significant minority were unsure. The majority felt that the type of mutation class did have an impact on general health, with

some believing it made a great difference (40%), and others some difference (30%). Half the sample believed mutation class also affected the severity of the symptoms (50%), although most believed that it had less impact on the type of symptoms developed (47%). The type of treatments available now and in the future for their child were viewed to be influenced greatly by mutation class (57% and 50% respectively). There was also a slightly less than equal split between participants' views on how likely treatments would be effective, with some stating that the type of mutation class made some difference (33%), and others reporting a great difference (37%). A smaller group reported that they were unsure (27%) (see Figure 2).

Figure 2: Participants' views of the effect that mutation class in CF has



4. Severity of own child's mutation class compared to other children with CF

10 participants rated the severity of their child's mutation class as moderate (33%), 9 reported mild (30%), 8 reported severe (27%). Only 3 participants did not know (10%).

5. Effect of mutation class on symptoms of CF

Table 2 show that most participants believed that their child's gene mutation affected their health in a number of ways; however two key areas were viewed to be significantly affected. These were the lungs (90%) and pancreas (83%). This view is accurate as these are the organs most seriously affected in CF. One participant was unsure of how their child's gene mutation affected their health (Table 2).

Table 2: Participants' views on what areas of their child's health their mutation class affects

	Yes	No	Not sure
Lungs	27 (90%)	2 (6.7%)	1 (3.3%)
Kidneys	10 (33.3%)	19 (63.3%)	1 (3.3%)
Bladder	7 (23.3%)	22 (73.3%)	1 (3.3%)
Pancreas	25 (83.3%)	4 (13.3%)	1 (3.3%)
Intestines	12 (40%)	17 (56.7%)	1 (3.3%)

6. Inheritance of CF

All 30 respondents knew that both parents are carriers of the gene that causes CF

7. Awareness of new approach to treatments for CF

20 participants (66.7%) reported that they were aware of new developments while 10 participants (33.3%) stated they did not.

3.4 Summary of findings

The survey was given out to parents of children with CF, aged 5 or under at the Leeds Children CF unit in order to capture an understanding of how much parents knew about the genetics of CF. In particular, it aimed to explore whether they had an awareness of their child's gene mutation, mutation class, and whether this made any difference with regards to their health, symptoms developed and severity, and the type of treatments available to them now and in the future.

Over half of the sample that was eligible to take part in the survey completed the questionnaire (n=30). All participants were mothers and therefore the survey only provides an understanding of what mothers know. All mothers knew that CF was an inherited condition that came from both themselves and the father of their child.

At the time of the survey, twenty-six mothers (87%) reported that they knew their child's gene mutation, with most stating Delta F508. Each patient actually has two

copies of the CF gene, and it is not clear from participants' responses whether they know that their child has two copies of gene mutation, or whether parents named the one they knew. Three mothers named both gene mutations.

Only nine mothers (30%) were correct in identifying what mutation class their child's gene mutation fell into. In the literature on CF, the only comparable research are McCrae et al's (1973) and Nolan et al's (1986) both of which were conducted 40 years ago. McCrae et al's research in the early 1970's demonstrated that parents had a 'poor' understanding of CF and only knew that it was an inherited condition. It seemed parents' understanding of CF had improved slightly by the time Nolan et al (1986) carried out their research as the parents in his study showed that they had a reasonably good understanding about the symptoms and treatments of CF. However, those same parents' knowledge about the genetics of the condition remained undeveloped. It is perhaps surprising given the publicity attached to Ivacaftor that parents in the current study – though clearly better informed than those in previous research were often unsure or incorrect about their child's mutations.

The gene mutation Delta F508 was the most frequently cited by mothers (n=22) in terms of what their child's mutation was and this particular gene is classed as being within the severe group of CF (e.g. Cystic Fibrosis Foundation; 2012). However, the survey found that only eight mothers viewed their child's CF as being severe compared to other children with CF in general. It is unclear how the remaining fourteen mothers have come to view their child's gene mutation DF508 as less than severe.

There was some uncertainty around on the effects of mutation class in CF. The majority felt that the type of mutation class their child fell into had a significant impact on their general health and the severity of symptoms, with less influence on the type of symptoms developed. Most participants were aware of how their child's CF affected their health, with most believing it significantly affected the lungs, closely followed by the pancreas. There were a smaller number of mothers who reported that CF affected the kidneys, bladder and intestines and these are much less affected by CF.

With regards to mothers' awareness about new approaches to treatment, two thirds reported that they were aware.

3.5 Conclusion

The survey highlighted that although there were some areas where the participants' knowledge about CF was good, there were other aspects of CF that were much less well understood. In particular, knowledge of mutation class, the effects that mutation class has in CF, and the severity of their child's gene mutation.

Though the sample size was relatively small and only mothers took part, the survey is the only recent attempt to explore parental knowledge of the genetic aspects of CF. It raises important questions about what information about CF is given to parents at the time of their child's diagnosis, how it is presented to them, and what parents are using to judge the severity of their child's condition. These questions were touched upon in the qualitative interviews with parents which were conducted to generate further understanding of their knowledge about CF, their views about treatments, and how they perceive their child's future.

CHAPTER IV

4.0 QUALITATIVE ANALYSIS

4.1 Participants

Semi structured interviews with 7 participants were conducted to further explore parental knowledge of CF, views about current and future treatments, and their thoughts about their child's future. This chapter is divided into two sections. The first gives an outline of the application of TA and an example of how it was used with a transcript. The second section presents the findings of the thematic analysis carried out on transcripts from the seven interviews.

4.2 Using thematic analysis

As described earlier, data collection and data analysis ran concurrently. Once the transcripts were completed, I listened back to the audio recordings to check the transcripts for any inaccuracies and make necessary adjustments. I also listened to familiarise myself to participants' stories, how they spoke about various aspects of CF, and how I had felt listening to them. Transcripts were analysed individually and compared to existing transcripts before being analysed altogether. I followed Braun and Clark's (2006) guidelines as described earlier however I also held in mind my research questions to help me contain my analysis. As a reminder, the research questions were; what does this participant know about CF? How do they view current treatments? How does this participant see their child's future? What do they know about new treatments?

A description of each phrase of TA is outlined together with an example of how the researcher conducted each phrase with the third interview transcript 'Louisa' to illustrate the entire process. The third transcript was chosen as I had started to feel confident both interviewing participants and using TA.

Phrase one: Familiarising yourself with your data

The first phrase of thematic analysis is where the researcher immerses themselves in the data to become familiar and develop an understanding of the data. Braun and Clark (2006) argue the researcher should actively search for meanings and patterns, and begin to make note of initial ideas or codes from the data that they find interesting and relevant to their research question. Though often skipped, Braun et al

(2006) advised that this stage should not be rushed, but conducted with time and patience as it will provide a foundation for how the rest of the analysis is developed

I read Louisa's transcript twice and listened to her audio recording once to gain an overview of her interview and to make sense of what she had shared with me. I was keen to understand who she was and how she talked about and viewed CF. During this process of active reading, I deliberated what Louisa said, and considered both how and why she said what she had. I also labelled each section of the transcript with what I felt Louisa was talking about (for example, 'diagnosis', 'knowledge about CF' and 'current treatments'). This helped me to break up the transcript into manageable sections to analyse and also made it easier to look back at the transcript and locate relevant sections when comparing transcripts and doing a group analysis.

Phrase 2: Generating initial codes

Once the researcher has become familiar with their data and has produced an initial list of ideas for coding, they can begin the process of formal coding in which they go through the entire data set, and identify aspects of each data item that are of interest, and which may start to develop into themes. The researcher is advised give each data item full attention and items that do not appear to match previously generated codes should not be ignored, but taken into account and coded.

I began to do a line by line coding of the entire transcript, and generated codes for things that I found interesting and relevant to my research questions. An example of how I coded is shown below

Table 3: Example of the coding process

Sentence	**Code given**	**What might be going on**
"For me it's not actually that important. I like to keep up to date with it but at the same time, as far as Abi goes, I just like to deal with what is specific to her as it comes along.	Not important to know everything Keep up to date generally Limits self to knowing what's specific Know things as it comes along	Sense of just dealing with what I can at that time Sense of knowing what's relevant to me
I don't like to… and I have been like this since Lauren was diagnosed, I just want to know the things that are important at	Want to focus on the now Don't want to look into the future Don't want to think about what	Sense of feeling afraid to look too far ahead What's happening now is enough for me to know

that time rather than looking into the future too much and seeing what might happen to them and extra medication and conditions that might affect them.	might happen Don't want to think about health deteriorating	Implies negative things will happen to them
Although I'll read a bit about it and might be aware, I think it scares me a bit too much to think oh well this could…because it might never happen, and so there's no point.	Reads a little about CF Scary to know too much Thinking about the what ifs No point thinking about it	Sense of limiting engagement on knowing much about CF Sense of denial, protecting self from knowing too much

In addition to staying close to each data item and making codes that captured what was said, I also considered more broadly what might have been going on for Louisa or how she liked to deal with CF, and included this in the column 'What might going on?' This was not a phrase that was suggested by Braun and Clark (2006), but I found it helpful in enabling me to think more about the participant and gain an understanding about what they were doing. I was also conscious that I did not want to lose the individual when it came to conducting the group analysis.

Phrase 3: Searching for themes

The third phrase involves re-focusing the analysis to identify themes by sorting out the codes generated and arranging all the relevant coded data extracts into potential themes. Here, the relationship between different codes are considered and questioned whether they can be grouped to form an overarching theme. Relationships between different themes and at different levels (e.g. overarching themes and sub themes) are also considered.

A total number of 1,325 open codes were developed across all 7 transcripts and the relationships between the codes were considered. I began to group codes and relevant extracts into categories in order of what they represented, for example, codes such as 'want to know as much I can about CF', and 'want to hear about CF kids doing well' were grouped together and labelled 'seeking information'. Similarly codes such as 'I don't want to know about kids not doing well', and 'I don't want to know too much about CF' were grouped together as 'avoiding information'. The codes that I

additionally made about what might be going on for participants (see table 3 column: 'what's going on') were also included in this process.

A total number of 170 categories were generated and the relationships between them were examined in order to identify themes. This process led to a total of 31 subcategories developed. In the process of beginning to group categories, I started to notice a pattern of 'opposites' for example, 'seeking versus avoiding information', and 'knowing everything vs knowing what's relevant'. It felt more natural to work in this way rather than to develop a series of single categories and given an overarching theme name. Once I had completed an initial draft of grouping categories, I began to consider what they shared in common in order to identify a theme name, for example, subcategories such as 'seeking versus avoiding information', and 'knowing what's relevant versus knowing everything' felt to me epitomised how participants dealt with and processed information about CF. I gave the overarching theme name of 'Processing information on CF'.

Phrase 4: Reviewing themes

Once potential themes have been identified, the researcher begins a process of refining those themes and considering whether they are separate, combined with other themes, or broken down into discrete themes. Two levels of reviewing and refining are involved in this process (Braun and Clark, 2006).

During this process, I considered how the themes and subthemes related to each other (level 1), and overall (level 2). I also reviewed the potential themes and subthemes by going back to the transcripts and examining the coded extracts. I considered whether the extracts matched a coherent description that I had produced for each subtheme and main theme. Where they did not, I reconsidered and moved categories around to form new main themes and subthemes only stopping once I was satisfied that a description matched a theme.

Phrase 5: Defining and naming themes

In this phrase, the researcher begins a process of further defining and refining the themes that will be presented in their analysis. The themes should each 'tell a story of what the theme is about', highlight which data represents that theme, and how they fit into the overall story of the data and in relation to the research question(s).

The themes and subthemes were reviewed in terms of ensuring they were clearly refined and labelled in a way that that described what they represented. I considered how each theme told a story on its own, and altogether. My research questions guided me throughout the whole process of TA as I thought about how these categories, subthemes,

and themes answer my research question. There were other themes that did not directly relate to my research questions however I found them interesting and did not want to lose them. These themes were included as they added richness to the story that I was starting to see emerge about participants' understanding and way of dealing with CF.

Phrase 6: Producing a report

The final phrase of TA involves writing a detailed report that tells a story of the data. Each theme and sub theme must be articulated and accompanied with supporting evidence. As guided by Braun and Clark (2006), I considered how I wanted to tell a story of my themes and ensured that my discussion did not simply provide a description of the themes but that they had a 'narrative' feel to it.

4.3. A change in the analysis

All transcripts were analysed individually in the procedure outlined above and were then analysed together except for one transcript, ('Jade'). Jade was the mother of a child taking Ivacaftor and she was purposefully recruited to share what could be a different perspective compared to the other participants. There were several differences between her and the participants including that her child was older (9 years old). Another difference was that she had lived with her son's diagnosis for 7 years – longer than other participants, some of whom had only received the diagnosis 10 months previously to being interviewed, and therefore unlike other participants she had a much longer time to adjust to and learn about CF. Her child had also gone through some of the stages that participants voiced their worries about - such as attending school - and so developmentally there were many differences between Jade's child and other participants' children. For these reasons, I considered and discussed in supervision both options of including Jade's transcript in the group analysis or keeping it separate in supervision. I also discussed this with a clinical psychologist who had no involvement in the research who I considered an independent expert. I had initially planned to merge several themes that I saw in Jade's transcript with the themes that were developed from the group analysis as there were many similarities, and to present other themes from Jade's transcript separately (such as her views of Ivacaftor) as I appreciated that she had different experiences from the other participants. In the end, after much deliberation I felt that it was best to completely exclude Jade's transcript from the group analysis and present it individually. I believed that she may have had a different perspective and to embed her story with the rest of the participants would lose that perspective, which would defeat the whole purpose of recruiting her. Furthermore, there was a natural shift in the focus of the interview schedule as there was more emphasis on discussing her

views of Ivacaftor and her child's future and as such I did not follow the interview schedule in the same way as I had with the other participants, though I asked some of questions that I had asked the other participants. I analysed Jade's story separately to the other participants using the same TA procedure and also carefully considered the similarities and differences between her story and the other participants' stories.

5.0 Quality checks

It is important when undertaking qualitative data analysis that it is performed with particular rigor to enhance the quality of findings generated. A number of procedures were utilized by to achieve this.

5.1 Supervision

Supervision with my academic supervisor occurred monthly and was a particularly important source of support and advice from the start of data collection to finalising the themes. Supervision was utilized in a number of ways. First, I shared my reflection of each participant and gave an overview of their interview. This helped me to develop an understanding about each participant, their story, and to recognize when and how the interview schedule needed to be amended to generate further understanding in later interviews. Second, supervision allowed the supervisor to comment on the emerging ideas and interpretations of transcripts, and often gave me a different perspective on analyzing the transcripts. Finally, I reflected on my own feelings towards the participants to prevent forming any potential bias or developing unchecked assumptions about participants' experiences.

5.1.2 Qualitative research peer support group

I attended several sessions of the qualitative peer group facilitated by an academic with extensive knowledge and experience of using qualitative research. This occurred bi-monthly and I used this time to check in with other researchers employing qualitative methods including TA. This was a useful format that enabled me to reflect on the research process and seek advice where appropriate.

5.1.3 Participants' validation

Participant validation has been viewed by some researchers as a strong quality check (e.g. Lincoln et al., 1985; Smith, 2008) enabling the researcher to ascertain whether their interpretation of data accurately represents the participants' reported experiences. Each participant received a summary report that described my interpretations and identified themes that emerged across the 7 interviews. They were

asked to consider whether this was consistent with their own views. Three participants replied in time to be included in the thesis, and they all found it interesting:

> *"It was interesting to read, one of the most interesting elements was that as I don't get to speak to many other parents who have children with CF a lot of us have the same worries. It brought up a lot of emotions as now CF is part of everyday life, I tend to forget all the bad elements but that reminds me how serious it is". (Sophie)*

> *"It was interesting to read them; I can identify with many of the comments made by the other parents" (Carole)*

> *"I read it all and was hooked! It's very interesting to read the views of other parents of children with CF" (Lorraine).*

6.0 Qualitative results

6.1 Participants

A total of 21 mothers expressed their willingness to participate in the semi structured interviews by leaving their contact details behind at the CF centre. Of these, 10 responded to my email, 3 declined to take part leading to a total of 7 mothers were interviewed; on one occasion a father was also present and took part in the interview with the mother and they were both viewed as 1 participant. Of the 7 participants interviewed, 6 had a child who was not eligible for Ivacaftor; one had a child who was eligible and taking Ivacaftor. The interviews lasted between 55 minutes and 1 hour 30 minutes. Table 4 provides basic demographic details of 7 the participants who took part in the interview:

Table 4: Demographic details of participants

Participant	*Age*	*Occupational status*	*Child's name and age*	*Mutation gene*
Carole	38	Homemaker	Harry, 3	DeltaF508
Lorraine	35	Paid part time	Jake, 3	DeltaF508
Louisa	40	Homemaker	Abi, 3 ½	DeltaF508
April	21	University student	Oscar, 10 months	DeltaF508
Chris	21	Paid full time		
Sophie	30	Paid full time	Ruby, 11 months	DeltaF508
Fran	40	Homemaker	Max, 3	DeltaF508
Jade	28	Homemaker	Luke, 9	G551D

6.2 Pen portraits

Before presenting the themes, pen portraits of the first six participants are given which include their backgrounds and my impressions of them. The pen portrait and themes for the final participant are presented separately.

Carole

Carole is 38 years of age. She is married to Mark and they have a three years old son, Harry, who was diagnosed with CF at birth. Carole identified herself as having some understanding about CF but described that at the time of her son's diagnosis, her knowledge about CF was outdated. She described that this was upsetting for her as she did not fully know what having CF meant for her son. Carole added that Mark previously knew someone who had CF and so he was also aware about the condition. However, like Carole, Mark did not have an updated knowledge about CF. At the time of the interview Carole was also expecting another child and had opted for genetic screening to test for whether the child has CF. Carole talked a lot about focusing on the now and did not want to think about the future. As well as focusing on the now, she was

particularly keen to know about positive news in the CF world for example, hearing about kids who are doing well as opposed to finding out about CF itself, what might happen and thinking ahead. I gained the impression that she actively avoids knowing anything about CF that would cause her distress.

Carole was warm and engaging but at the same time she did not offer a lot of information. She seemed initially nervous about taking part in the interview and I sensed that she tried to stay on safe topics for example; she did not often share her feelings about her son's diagnosis and how it felt to think about the future. Instead she talked about what her son's treatments involved and what they were for. After the interview, she said that she was nervous of what psychological questions she might be asked and thought she would have to share her emotions which she said she did not feel comfortable to do. The conversation flowed well, but there were times where I felt it stalled and I needed to rely on the interview schedule to keep it going.

Lorraine

Lorraine is 35 years old and was recently divorced from her husband. She works part time and has two children, Jake and Hannah. Jake is three years old and he was diagnosed with CF at birth. Hannah is 1 year old and does not have CF. Prior to Jake's arrival, she described her knowledge of CF as non-existent. She also said that the first four months of Jake's life was emotionally difficult before she was able to eventually adjust to his CF.

The conversation with Lorraine flowed easy and the interview lasted 52 minutes. There were several things that I noticed about Lorraine such as that she tended to say 'you' as opposed to 'I' when describing things for example, when she talked about her child's diagnosis she said 'you just cry for days and weeks'. I felt she was distancing herself from talking about her own emotions and I wondered whether this was a coping strategy. I also gained the impression that she was particularly adamant that she would not let CF take over and stop her son from living his life. It seemed to be important to her to make life as normal as it could be for her son. Lorraine came across as someone who really wants to help research on CF in any way that she can. I was struck by her positivity that having CF is not the end of the world and that one day, CF will be something that is 'not a big deal'.

During the interview, Jake frequently entered the room and wanted his mother's attention. He was a very active, playful, and happy young child and I saw the close bond that he has with his mother. She delighted in playing with him and chasing after him round the living room.

Louisa

Louisa is 40 years old and she is married to Jeremy. She identified herself as a full time mother to her three children Joe, Lauren, and Abi. Joe is aged 8 and does not have CF. Lauren and Abi are aged 6 and 3 ½ respectively, and they were both diagnosed with CF at birth. Prior to Lauren's diagnosis, both Louisa and Jeremy had no knowledge of CF. She said that she and Jeremy know what gene mutation Jeremy has, but they did not know what gene mutation she has. They have been unable to find this information out and she described that she wanted to know even more now upon hearing about the new treatments.

Louisa came across as a very bright and engaging woman who was keen to talk about her children's CF. Our conversation lasted 1 hour and 5 minutes. She said that she liked to talk about CF when she can as she does not get many opportunities to do this. I also noticed that Louisa sometimes appeared to try to limit how much she knows about CF and from thinking about the future. I felt like there was a deep sadness within Louisa. There were frequent times where she was visibly upset, and other times when she laughed but it was a sad laughter, as though she was brushing off painful memories and emotions. I wondered several times whether she had accepted her children's diagnoses, as she tended to say things that suggested otherwise. I sensed that her children's diagnoses had left a powerful mark within her. She added that '*cystic fibrosis is close to my heart'*.

April and Chris

April lives with her boyfriend, Chris, and they have a 10 months old son, Oscar, who was diagnosed with CF two weeks after birth. April attends university and Chris works in a full time job. Both are 21 years old and have their own home. Chris took part in the interview as he was keen to be involved. The couple described a difficult beginning, in that Oscar needed surgery because he had Meconium ileus; a sticky and blocked bowel which is one of the first signs of CF. The couple said that as soon as they heard about CF being a possibility, rather than waiting for the genetic test results to come back as confirmation, in their minds they had already accepted and knew Oscar had CF.

I was struck by how young the couple were to be parents of a child with CF, and more importantly how they dealt with it all. They both were very open about their experiences and how they deal with CF on a daily basis. Chris said it was very important to him to know everything about his child's CF so that he could answer any questions that his son might ask. He also said that he knows Oscar might not be around as long as he hopes and talked about the importance of being in the present and ensuring he makes the most of every day with him. April had a different way of dealing with Oscar's CF.

She came across as being worried and apprehensive about the future, particularly about whether Oscar's health will deteriorate. She also said that she preferred not to know too much about CF, or think about the future. They both came across as supportive to one another, and helping each other when they are at their lowest. The conversation flowed well and the interview lasted longer than I had expected; 1 hour and 11 minutes.

Sophie

Sophie is 30 years old and married to Michael. They have one daughter between them, Ruby, aged 11 months. Sophie described a strange and rather difficult beginning with Ruby. There were early signs or clues of CF during her pregnancy for example, she had a scan that showed her baby's belly was big but it was dismissed until when she gave birth, which in itself was traumatic and involved an emergency C section. Her baby was then taken away from her and transferred to Leeds. It wasn't until the early morning that she was finally allowed to see her for five minutes. There was a sense of an initial separation between Sophie and Ruby that I imagined she found very difficult. Ruby was diagnosed 6 days after her birth.

I gained the impression that Sophie was superstitious as she often touched her table made of wood and crossed her fingers when she talked about Ruby to avoid 'jinxing' things. Sophie described her fear of looking ahead and thinking about the things that are yet to come, as though there is an impending doom. She told me of times when she tried to be courageous and look into CF by going onto the internet or pick up a book, but then she would put it down straight away because there were too many 'horror stories'. I sensed that she often feels torn between trying to know everything about CF in order to prepare herself and not knowing everything because it makes her panic and worry. She talked about being guided by her emotions and moods. For example, on days where she feels stronger she would try and look up more about CF, and on days where she feels less strong she would avoid knowing things.

Sophie came across as very warm and bubbly in her nature, but underneath there was a sadness which rose to the surface quite quickly several times during the interview. I could see how much she really appreciates Ruby being here and she reiterated how lucky she was to have her.

Fran

Fran is Irish, 40 years old, a full time homemaker and has been married to Charlie for 16 years. Between them, they have 4 boys. Only the youngest - Max, aged 4, has CF. Max was diagnosed when he was 1 year old. Fran's story of how Max came to be diagnosed with CF was different to the other participants' stories in that Max initially had a false

negative test at birth. For a long year Fran described her son as being ‘always poorly’ with recurrent chest infections. Fran frequently sought medical help and sought a cause, however she described the healthcare professionals in her local area as having little knowledge of CF and so it took a year to finally receive a diagnosis. Her reaction at diagnosis was different in that rather than being distressed by the CF, she was concerned that she had missed a year’s worth of treatment and was worried about the impact of this.

Fran came across as being very articulate and as very matter of fact, but equally very warm. I sensed her attitude towards life was to make the best of any situation and deal with it as well as she could. She described herself as being someone who doesn’t dwell on the ‘what-ifs’ as it doesn’t make any difference to think about that. Rather, I gained the impression that she was a woman who focuses on the task in hand and what she can improve. I was surprised by her knowledge about the genetics of CF- for example, she knew about mutation classes, which classes her son’s genes fell into, and whether the genes were rare or common. She was also aware of new drugs that were in the ‘pipeline’ and could further identify the drugs’ names, which mutation classes that these drugs are designed to target, and whether these are going to help her child. Equally, I was also surprised by her approach to thinking about the future. She did not come across as someone who was afraid to find out about CF; instead she actively keeps abreast of developments in CF and makes enquiries about it. She also seems to think about the future and has thought about how she might manage future worries, in particular teaching her son the importance of adherence.

Fran surprised me further when she became emotional at the end of the interview as she had been so composed during it. She said that she herself was surprised at this and recognized that the pain was still within her. She acknowledged that the interview had aroused some of those painful feelings that she thought had either resolved or was buried deep within her. The interview lasted 1 hour and 20 minutes.

6.3 Core themes

Six core themes were developed from the thematic (see table 5). Each of the themes and subthemes will be now described, using illustrative quotations from participants.

Table 5: Core themes and subthemes

Core themes	Subthemes
1.0 An unknown territory	1.1 My initial reaction and the aftermaths of it 1.2 Feeling clueless vs recalling what I knew and trying to comprehend 1.3 Mentally preparing for the worst
2.0 Struggling with paradoxes	2.1 Forgetting CF exists vs worrying about CF 2.2 Refusing to let CF take over vs not restricting what my child can do 2.3 Making the best of it and feeling supported
3.0 Information overload	3.1 Information overload 3.2 Seeking information vs avoiding information 3.3 Knowing what's relevant vs knowing everything
4.0 Taking control but feeling uncertain	4.1 The importance of adherence 4.2 Process of administering treatments 4.3 Feeling ambivalent 4.4 Worry about whether it works and trusting the experts
5.0 Dealing with uncertain future	5.1 Avoidance 5.2 Hidden worries 5.3 Comforting myself
6.0 Quiet hopes and holding back	6.1 A vague awareness of new developments 6.2 Feeling pleased vs worry about funding 6.3 A healthy cynicism vs hopes for the new treatment 6.4 Focusing on the now vs feeling hopeful for the future

Core theme one: An unknown territory

This theme describes the arrival of CF at the very beginning when participants received their child's diagnosis. The theme encompasses three subthemes: my initial reaction; feeling clueless vs recalling what I know and trying to comprehend; and mentally preparing for the worst.

1.1 My initial reaction and the aftermaths of it

All participants experienced strong emotions when they first discovered that their child had CF and what this meant, with most stating that they felt shocked and distressed (6/6):

"It was quite a shock…to find out about…what the implications of it were and what it meant. It's a lifelong condition.' (Louisa)

"It was quite upsetting! Really upsetting…getting that diagnosis.' (Carole)

There was still variability within this reaction however: two participants who were both parents to the same child with CF described how they reacted differently to each other:

> *"When he first got the diagnosis... I was a lot more upbeat… not happy about it but I was... I could get on with it a lot easier than..." (April)… "You got on with it and I were right depressed… and we switched round didn't we… we went right bad and I were saying the same stuff to you…" (Chris)… "Then when Chris got his head around it… I just fell apart then and then I couldn't deal with it." (April)*

One participant recalled thinking it was a hoax and felt angry and confused:

> *"When I got the phone call to say that he had CF, I just thought it was like a hoax call or something. I really did. I was like who are you? Ringing me up…What are you talking about? She was like, my name's (NAME) I'm from the clinic and I'm coming to visit you in 2 hours." (Lorraine)*

Another participant initially blamed herself and experienced regret:

> *"When Lauren first got diagnosed, I remember going through a stage thinking…why…why didn't I check for this while I was pregnant, why didn't I… Why wasn't there a check that tests for this?" (Louisa)*

When it came to her second child's diagnosis, this participant recalled feeling more prepared to receive the diagnosis. But even then getting the diagnosis still shocked her:

> *'So even with Abi, we knew there was a risk that she might have CF when she was… when I was carrying her, we didn't know it was a girl…but we knew there was a risk that our baby would have CF. We were sort of more prepared… and then we knew at birth that she had CF because she… we already had an older child with CF and then Abi was born with a blocked bowel and so it was fairly obvious that she had CF…but it was still quite shocking." (Louisa).*

Prior to receiving a diagnosis of CF, some participants (2/6) also described a sense of feeling dismissed by healthcare professionals (who were not part of the Leeds CF team) when they were concerned about their baby, which with hindsight could be seen as early signs of CF:

> *"We were told at 39 weeks that she had a big belly on her scan, three people told us it but they didn't do anything about it, erm, and left it and said 'Oh you might just need a wee." (Sophie)*

> *"It was the admissions nurse I was talking to…I remember saying to her 'he could poo up to 8 times a day, and it was orange-y'. And she just went: 'oh right', and kind of laughed it off, now that's a sure sign of CF and as soon as I mentioned that to Doctor (*NAME at Leeds CF unit*) he was almost like: 'I don't need to know anymore, that's all I need to know, let do some tests' kind of thing." (Fran)*

Participants (4/6) recalled that as they began to move past the feeling of shock, it was a difficult beginning. Feelings of distress, loss, depression, and struggling to be with people were described:

> *"At the time I was just in an absolute daze. Because it's horrible. You just cry for days and weeks and months…I did cry for about four months." (Lorraine)*

Some participants (2/6) described struggling to deal with not being the parents of a child that they had expected, and almost grieving for that loss:

> *"We'd prepared to be parents of a nice healthy child…and obviously when someone turns around and says it's not going to be like that it's…just…yeah." (Chris)*

Several participants (3/6) initially isolated themselves from others as they felt unable to be around them:

> *"The first month we didn't go out, we didn't speak to people, we couldn't face the big bad world because we didn't want people to see (pause) the pain really, I didn't want to see people." (Fran)*

> *"We were quite down and depressed at the beginning, we did shut out quite a lot of them* (family). *And we just kind of wanted to be left alone and…that become an issue with family because they wanted to help and we wanted to be alone…" (Chris)*

Participants (2/6) described feeling almost unequipped in the beginning

> *"It was really quite tough when he was a baby, I think reflecting back, I don't think I realised how hard it was compared to somebody that didn't have a child with CF… It was abit overwhelming really, the amount of things that we had to do…it felt like it…but I think a part of it was…it was a new child. I didn't know how to deal with a baby for a start let alone a baby that had special needs…" (Carole)*

One participant said that she needed to seek support to deal with it all:

> *"…after we had Abi and she was poorly at birth and she was our second child with CF…I found it…well I went through a very difficult stage myself because I think after the stress of her birth, getting over that and then extra things to deal with her…I found it very hard…and I got myself some help." (Louisa)*

1.2 Feeling clueless; recalling what I knew to try and comprehend

Participants' understanding of what CF was at the time of their child's diagnosis varied from knowing nothing (3/6) to desperately trying to understand by recalling anything that they knew about it (4/6):

> *"I knew, I knew nothing before Ruby was born." (Sophie)*

> *"When I first found out, with Lauren my elder child, and they came and said that there's a possibility that she had CF, I didn't know anything about it. I didn't…we didn't…didn't…I'd hardly heard of it…We had no previous knowledge of it in our families even though we… we didn't even know we were carriers, my husband and I didn't know that we were carriers of the CF gene or anything like that." (Louisa)*

Some participants (4/6) tried to make sense of their child's diagnosis by reflecting on what they had heard about CF before and any past dealings with it that they might have had:

> *"All I knew is… All I knew is that he was going to die when he was 30 and it was something to do with his lungs because that's just what I knew in my life anyway about CF, that's the only thing that I'd heard of cystic fibrosis for." (Lorraine)*

> *"I had a very very tiny bit of background because my cousin had CF. But I was only young at the time so I didn't know the ins and outs of it at that stage, I just knew that he was poorly and things went on in the hospital…" (April)*

1.3 Mentally preparing for the worst

All but one participant (5/6) recalled thinking the worst when they first received the diagnosis. The belief that their child would die played on participants' minds significantly:

> *"I thought: 'oh my...' I thought he was just going to probably be ok for 20 years and then just deteriorate and die. That's what I thought. So that's what I thought in my head when someone said your little boy's got CF. I was like oh my god, oh my god, horrible…" (Lorraine)*

Another participant was told by the healthcare team to expect the worst when her child was born:

> *"They told that she might not make it on the first day she was born, to expect the worst."(Sophie)*

In contrast, the male participant Chris thought differently:

> *"One thing I remember you (April) saying is…every Christmas you're going to be thinking, that's another Christmas less didn't you? But for me, my attitude is…it's a Christmas gained." (Chris)*

Participants (2/6) thought about things other than the possibility of death including their child's capabilities and the kind of life they would live:

> *"There was a girl with Cerebral Palsy when Ruby was diagnosed and I thought CF was like Cerebral Palsy, so I was panicking thinking: 'Oh no, this girl has a lot of things she can't do' (emotional) I was thinking: 'it's awful, there's going to be so many problems'." (Sophie)*

> *"I thought it was going to be… I thought I wasn't going to be able to work and I wasn't… and we were going to have to look at him every night to check that he wasn't dying." (Lorraine)*

For one participant, she recalled that after she got over the initial shock of her child's CF, she felt both hopeful and determined to beat the odds of her child dying at a young age:

> *"I needed peace of mind right from day one that things were going to be okay, and obviously they couldn't reassure me that things weren't going to be tough, but there was hope and that's all I needed, that hope that somebody has survived to age seventy, that's my benchmark. Max's going to live beyond the*

age of seventy because somebody has done that already, so that's what we are going to aim for." (Fran)

As several participants (4/6) started to learn about CF, they began to feel better:

"And then we realised that CF was something totally different to cerebral palsy! So it was better when we understood." (Sophie)

"It was about a million times better that I understood what CF was." (Lorraine)

Core theme two: struggling with paradoxes

Core theme two described the struggles that participants experienced daily; forgetting CF exist vs worrying about it, and refusing to let CF take over vs restricting what their child can do because of it.

2.1 Forgetting CF exists vs worrying about CF

Participants described that they generally don't worry about CF and tend to forget that it's a part of their lives until something associated with CF surfaces:

"I actually don't…I don't worry about it as much at all. I forget about it, it's only when he has a bit of a cough or maybe the results come back and they're not quite as we would want it, it suddenly…it hits me again. Otherwise, normally on a day to day basis…I don't even give it a second thought." (Carole)

At the same time, participants (6/6) said that they worried about many things, suggesting that CF is actually very much alive in their minds:

"I worry how to protect him from the hidden bugs…. His chest and weight gain is our main concern, something that we keep an eye on…making sure when he eats something, it's very calorific…we monitor his progress by monitoring his poo every day…" (Fran)

One particular participant described worrying about her child being in situations where she could be exposed to germs and described the lengths that she goes to ensure that she doesn't:

"Like we had a christening the other week and I emailed the girl in the morning and said 'do you know of anyone going with a cold? Sounds ridiculous because you can't pre-empt anything but…' and she just said 'I don't think so…' so we

went there for an hour and then some people came and their little kids had a cold so I said 'we'll get off now' and we left straight away." (Sophie)

2.2 Refusing to let CF take over vs restricting what my child can do

As part of adjusting to CF, most participants (4/6) were quite adamant that they did not allow CF to hold their child back:

"I really believe that I don't…I do not want his life to be any different because he has got cystic fibrosis do you know what I mean. He's not going to be 10 and go, 'oh I couldn't do that because I had CF…' I'm like, no…I want him to do as much as he can, just have the same life." (Lorraine)

For one participant, her fear of her son growing up to resent her plays a role in ensuring that she doesn't let CF dominate his childhood:

"I don't ever want to say 'no you can't play with your friends…go on the paddling boat', I don't want him to ever turn against me and turn against the disease." (April)

Even then, there seemed to be a contradiction for some participants (3/6) as they discouraged their children from doing something that they know isn't a good idea:

"She'd like to be a nurse and we might have had a… we might…I think we had a little discussion about the fact that maybe, maybe being a nurse wouldn't be a very good idea for her because, because she'd be exposed to things that she shouldn't really be exposed to." (Louisa)

2.3 Making the best of it and feeling supported

This subtheme describes how participants tried to deal with the unfamiliar presence of CF and their struggles with paradoxes. They described how they tried to make the best of it and the invaluable support they received from the CF team. Making the best of CF involved for some participants (4/6) appreciating that CF happened to their first child:

"I think, uh, I am grateful in a way that she was our first child with CF and not our fourth child with CF…If I had a baby without CF and then a baby with CF, it'd be like (tired sigh) oh my god, so much work." (Sophie)

Participants (5/6) tended to normalise CF by viewing their day to day as being normal and seeing CF as a condition that does not have a significant impact on them:

"What we actually do all day, every day is not effected at all. I work the same as I would have if he didn't have CF. (FATHER'S NAME) works, he goes to school, everything is normal. It's good." (Lorraine)

"I think it will be more abnormal for us to have a baby without CF than a baby with CF." (Sophie)

For most (5/6), it was also about putting CF into perspective by comparing it other conditions that are worse to live with and appreciating that it is not an obvious disability:

"Compared to what other people have to deal with, you know he's mentally sharp, physically sharp, you know it's- it's maybe not the worst thing in the world that he could have, I would find other things a lot more difficult to deal with, a physical disability for example, would be more difficult to deal with." (Fran)

"Nobody knows that he's got CF…I love it. If he was in a wheelchair, everyone would know it. And everybody would talk about it…not in a bad way but you can't help but say 'oh the little boy in the wheelchair.' Do you know what I mean? Which…I'm really really really really pleased it's not a physical disability…" (Lorraine)

The male participant thought about CF in a different way in that he did not compare CF to other conditions, but instead tried to put it into perspective by thinking that we could all die at any time, whether we have CF or not:

"I've always got the attitude that I understand it's life limiting but I think…anything could happen to any of us…and unfortunately, I could go tomorrow…or you hear of things on the news where such young children develop something or get run over or anything like that…"…"But for me, my attitude is…yes it's a Christmas gained with Oscar every year, not…but also it's all of us…it's a Christmas less for all us realistically!" (Chris)

Even then, some participants (2/6) described still feeling saddened that their child has CF and wishing that they didn't:

"It can be hard, every now and then you have a day where you just want to cry all day and you think…why did it happen to us?" (April)

"You just think it's a little unfair sometimes, I think that's what it is, you think 'Aw why my Ruby'." (Sophie)

All participants (6/6) particularly valued the support that they receive from the Leeds CF team which helped them to make the best of the situation:

> *"The care at Leeds is fantastic, couldn't fault it, everybody is amazing. And I don't think if it wasn't for that support I'd feel as comfortable as I do 'cos I know I can talk to them if I got problems… I feel like we've got a lot, a lot of support from them." (Sophie)*

Others (2/6) extended their appreciation to researchers and scientists:

> *"They're doing their absolute best, the absolute best that scientists can understand…" (Lorraine)*

Core theme three: Information overload

The core theme 'information overload' describes how participants managed the amount of information that came with the receiving the diagnosis of CF and since then:

3.1 Information overload

Participants (5/6) identified feeling overwhelmed by how much there was to know about their child's CF, and CF in general. Several participants described having to attend the clinic at the CF centre day after getting her child's diagnosis to learn more about CF:

> *"Then you have to go and spend the whole day at clinic with information overload. It's good but it's like you're so tired and you're emotional anyway." (Lorraine*)

> *"There was just so much information…just too much information all at once." (Chris)*

Participants (3/6) described wanting to know about the latest research on CF but time constraints makes it difficult to do so:

> *"I am interested in the latest research, things like the gene therapy and things and how that's coming along but I wouldn't…I just skim read a few basic points on that but I wouldn't read it in depth to see how it's going and…pilot studies and all of this. It just time really...I suppose it just having the time to read all the evidence..." (Carole)*

3.2 Seeking information vs avoiding information

At the beginning, most participants (5/6) initially sought out information that would help them to understand CF:

> *"When she was first diagnosed I said: 'give me as much information as possible to research it…" (Sophie)*

> *"At the beginning, at the first clinic appointment, I was asking all the questions about how it would affect him, what's the life expectancy, and what is it that kills them in the end." (Fran)*

As participants began to learn about the implications of their child having CF, they described feeling overwhelmed by the amount of information they were exposed to and they started to distinguish between information that they were keen to hear about and information that they preferred to avoid hearing about:

> "*I love reading good news stories about children who have done really well and that are fit and healthy, and they're top gymnasts or…doing well…or there were some children singing on X factor, two siblings that were really…both had CF and they were doing really well and…I just thought…I want to hear about those stories!" (Carole)*

Others liked to know about the latest developments in CF generally:

> *"I would always be happy to be given information about the latest on CF or about new treatments and stuff…" (Louisa)*

At the same time, most participants (5/6) said that they tended to avoid finding about CF because of it made them fearful and anxious about what could happen to their child, and also question their ability to look after them:

> *"Knowing everything it's a lot more scarier because you're aware of how many dangers there is in the actual world that might affect him. It can be scary to know everything because then we…you get worried thinking…can we do this?" (April)*

3.3 Knowing what's relevant vs knowing everything

Both fear and time constraints have led all participants (6/6) to develop strategies for managing information by which they only seek information that is directly relevant to them at that time:

> *"For me it's not actually that important. I like to keep up to date with it but at the same time as far as Abi goes, I just like to deal with what is specific to her as*

it comes along. I don't like to…and I've been like this since Lauren was diagnosed, I just want to know the things that are important at that time rather than looking into the future too much and seeing what might happen to them and extra medications and conditions that might affect them." (Louisa)

"I think you've kind of, because there's so much going on, you've got to filter the information, so you've got to know what you need to know, and you're aware of the other stuff but don't dwell on it too much because it doesn't make much difference to you." (Fran)

In contrast, the male participant felt that knowing everything was important to help him prepare for the future and to answer questions:

"For me it prepares you though doesn't it? Prepares you for what can be around the corner…"… "For me it's…the hard thing about it is people not understanding what it is and I want to be the person who basically is able to answer any questions that anybody's got about it. And him himself, so for me to actually learn the most about it as possible, is obviously…any questions he has then…I should have the answers…" (Chris)

Core theme four: Taking control but feeling uncertain

Core theme four describes how participants manage current treatments, and the emotional and practical burden of this for them. It contained four subthemes: the importance of adherence; the process of administering treatments; feeling ambivalent; worry about whether it works and trusting the experts. Participants described a sense of taking back power from CF by adhering to treatments yet at the same time they described feeling uncertain about their necessity and efficacy:

4.1 The importance of adherence

All participants (6/6) described their child's adherence to treatments as very important and they worked hard to ensure that their child did not miss any:

"We're so strict with ourselves. He does not miss his medication, on a day-to-day basis, you know it's just not an option, it's wherever we are, whatever we're doing, and it's not an option that he misses his medication." (Fran)

"And like when we're staying over at grandparents' houses or friends' houses, you just have to go…you just have to do it for 20 minutes no matter what. You just do it. 10 minutes running and then 20 minutes of chest physio…" (Lorraine)

In contrast, although one participant agreed that adherence was very important, she also said that they still had some 'treatment days off':

> *"But sometimes if we don't do it…like we do it every day…but say if we were at a wedding or a…Christmas day or something like that…we wouldn't do it, we'd just have a day off." (Lorraine)*
>
> *Interviewer: "How often does that happen?"*
>
> *"I would say about…10 times a year…something like that." (Lorraine)*

Another participant felt that her and her partner's families did not understand how important adhering to treatments were, which made it difficult for them to trust that their families would look after their son properly in their absence

> *"They say… like, oh just one time won't hurt him. But to us we know that one time this…whatever he has or what he's not meant to be doing may have one of these bacterias and that one time could lead him to then get it…" (April)*

Some participants (3/6) also reported that there were times when they 'missed' or were not able to do a treatment and that would make them feel guilty:

> *"I feel guilty if for some reason, I haven't managed to do their physio, something stopped me from doing their physio or anything like that. I will have a guilty feeling about thinking…I've missed their physio…or there might be times when I've missed out a medication or some reason, forgotten to do it or something like that…" (Louisa)*

For another participant, she described not doing a treatment if her child was too tired:

> *"We might be out at a friend's house and we might be driving home in the car and then we might be like…oh right we'll get home and then put them straight to bed because it will be half past seven. We're not going to do his treatment because he'll be absolutely knackered and crying because it's too late." (Lorraine)*

4.2 The process of administering treatments

This subtheme is a broad theme that describes how participants initially felt about administering treatments compared to now, what helps make giving treatments easier, and how long it takes them to administer treatments:

Several participants (3/6) recalled that in the early days following the diagnosis they had many concerns about treatments, particularly with ensuring that they administered them properly:

> *"The medicines, I used to have a chart in the, uh – I used to tick everything off and write everything down and I did that for about seven months before I would stop writing things down to make sure I was covering everything." (Sophie)*

> *"We were worried that we were doing it right because we were responsible for administering the nebulisers and sterilising all the equipment and ordering prescriptions, making sure they got them in the chemist…" (Carole)*

As participants (6/6) began to get into a routine of administering treatments, treatments became normal to them:

> *"We give him medicines…6…well 7, 8 syringes a day now it's just…it's normal now..."… "It becomes routine for when he's on his knee and you're watching TV just to kind of…give him a tap..." (Chris)*

> *"I find them second nature, I find them quite easy to do." (Fran)*

One participant described some of the treatments as being no different to what people without CF should or would be doing anyway:

> *"It's just like us though…if we go for a run, it's good for our lungs, it's even better for him…" (Lorraine)*

Participants (5/6) also stated that their children had grown up with treatments and saw them as normal, with some already taking an active part in administering them

> *"She just watches Mickey when we do physio, you wouldn't even know we're doing it. I think she thinks it's normal." (Sophie)*

> *"We still give medicine in a syringe…sometimes I've left them on the table where I've been out of the room and I've come back in and he's taken the syringes himself…they're empty when I come back!" (Carole)*

The process of administering treatments is easier when their child responds well to treatments most of the time:

"It's just second nature to him. It's just second nature. He has to take 1, 2, 3, 4, 5 syringefuls of medicine everyday which he takes just like that. You just approach him and he takes it." (Lorraine)

Almost all participants (5/6), however, said that treatments could be difficult to do at times:

"The only time it's not very…like it's easy, it's not enjoyable, but it easy. Sometimes if he's really tired…he just wants to be playing with his toys. You have to spend a few minutes like coaxing him, come on…because I don't want to make him have to do it, kicking and screaming." (Lorraine)

For one participant, it was about making sure the right amount of medication was given:

"It just getting that balance that he gets, the right Creon for the right amount of fat that he'd had for that particular meal and then spacing it out… um, so he has, he has one scoop of Creon for every 2.5grams of fat, we have to work out how much milk he's had on his breakfast, how much fat is that, and how many Creon scoops is that…" (Fran)

Despite participants' insistence that treatments are normal and routine in their lives, they also talked about the time consuming and constant nature of completing treatments every day:

"…It can be quite wearing because you can't ever switch off so…every morning you have to do their physio, you get up and give them their medication and do their physio and…they have their last medication at 10 o'clock at night, while they're asleep and so it's only really after that that I can switch off until…the next morning!" (Louisa)

"You have your bad days where you think: 'Oh this is such a chore…'" (Fran)

Participants (3/6) described the relief of not doing treatments on occasions:

"You do think, oh my god, it's quite nice not to have to do it…" (Lorraine)

"…Sometimes me friend…if she's round here she'll say: 'Oh I'll do the physio tonight if you want', y'know if me and (partner name) are having tea then she'll do it then and I think: 'Oh, god, that's nice'." (Sophie)

One participant described a way that she coped with these demands – recognising that even though treatments are demanding and difficult, it is important to balance this by remembering why they were doing them:

> *"But in the grander scheme of things, if it means that he's healthy and it maintains his, balance if you know, it's a small price to pay really." (Fran)*

There seemed to be differing views on how long treatments take to administer. Some participants (2/6) felt treatments did not take long and had minimal impact on their day to day life:

> *"Now with medicines, it takes a matter of a minute to give him his medicine…" (Chris)*

> "*We're only affected, every day we're affected for half an hour because we do 10 minutes of extra things in the morning, we do 20 minutes of extra treatments and exercises in the evening…" (Lorraine)*

In contrast, other participants (3/6) had different views:

> *"It depends, I suppose, on whether you see his breakfast as a treatment. I do because that's when he gets his Creon, his milk, his Urso, that takes half-an-hour, lunch can take easily half-an-hour, teatime easily half-an-hour…" (Fran)*

4.3 Feeling ambivalent

This subtheme describes participant's overall feeling of ambivalence about current treatments. Participants identified treatments that they generally found ok, treatments that they disliked, and treatments that they found uncomfortable:

Most participants were happy with the physiotherapy (mainly exercise) and Creon administration (it is taken with food) aspects of treatment:

> *"I like just running around with, just doing all the stuff that we do, so we sort of have a 10 minute football kicking session after his tea…" (Carole)*

> *"his best form of treatment is, uh, running around…when his brothers come home from school they torment him, by taking something from him, and running around just to make him round around…and he loves it, he loves the attention, he loves the chase, he loves winning 'cos they let him win sometimes… the whole thing works you know? It gives him his physio, gives him abit of enjoyment and interacting with his brothers." (Fran)*

"I mean his Creon, the enzymes… we…they're great aren't they…he's always putting his weight on… very happy with them." (April)

Treatments that were seen as 'invasive' and caused their child distress or to feel uncomfortable were disliked by participants (4/6):

"His nebulisers. We don't really like those, but I think it's because it's difficult to administer…when he was a baby, it was quite stressful so we'd come down, get him to sleep and then we'd wait for an hour or so and we'd make sure he was really fully asleep and then we'd nominate one of us to go up and do it. Sneak in and put the mask on him. Try not to wake him up…we had to draw straws on who did it each night!" (Carole)

"Sometimes…we have to take blood, that's horrible, but it'll be fine in years to come…it's a horrible age at the moment because you can't really explain it to him. But he just thinks…why is she hurting me?" (Lorraine)

Antibiotics were not just difficult to administer as several participants (3/6) also recognised that they felt uncomfortable that their child was permanently on them:

"It's…the times it upsets me is when…like I was on the bus going to the clinic, one of the women on the bus was…is he having calpol and…I just said yes because I didn't want to go into any details…" (April)

"It is always a bit strange to think that you're giving your children antibiotics everyday of their life for the… I must admit I was under the understanding that you should only really give antibiotics if you really needed it so it was a bit strange to be giving them the way they do give them so many antibiotics…antibiotics everyday any away and as soon as the first sign of cough or a cold, extra antibiotics and it does…should we really be giving them all of these antibiotics? Is it a good thing?" (Louisa)

4.4 Worry about whether it works and trusting the experts

Most participants (5/6) expressed concerns and doubts over whether treatments (mainly physiotherapy and antibiotics) actually made a difference. Worries about whether the treatments work tends to be addressed ultimately by a decision to trust the advice of the experts – their child's CF team:

"It hard to say whether it's making any difference because she wasn't actually seeming to be poorly in the first place…" (Louisa)

"Physio's hard to say because you don't ever see what it actually does…so that's quite hard because obviously we're trying to battle to do something which we've never seen the impact of…" (Chris)

The majority of participants said that the only way they would know if treatments works is by not doing them, but added that this is something that they would never do to their child:

"How do I know…I'll never stop him will I, if I stopped it all for a month then I would really find out but I'm not going to take the risk…So if he didn't have the drugs and the physio, he might be exactly the same but I'll never know." (Lorraine)

Two participants had focused on the yearly annual visit at the CF centre where the team reviews their child's health status as a chance to get feedback on whether treatment is helping, but though this can potentially be reassuring, it is also a great source of worry (and sometimes guilt):

"If his chest looks really nice and clear then obviously we'll be really happy but if it doesn't…that's when I'm going to get really worried…" "I think if it's a negative one we have this year, with this first one, I think that will be really hard to…I think that will be really difficult to…because you'll feel like you haven't done enough…" (April)

April's partner, the male participant, also expressed worry:

"It's like doomsday isn't it…it's like after a year's worth of work…we're finding out if everything…if we're doing everything right." (Chris)

One participant described the importance of trying to be positive and hopeful that treatments were effective:

"I think it much better to be positive about the current treatments working and them not being poorly…" (Louisa)

Another was confident that some treatments at least made a huge difference:

"The Creon is like a miracle drug, I have to say. He (pause) overnight started to put weight on once he started on the Creon and it is, it's fantastic, that is like a little bottle of miracle really." (Fran)

All participants (6/6) said that they felt that in the end they just had to trust that the experts know what they are doing with the treatments:

"I just trust the experts to be doing what they need to." (Carole)

Core theme five: Dealing with an uncertain future

Core theme five describes the ways in which participants approached thinking about the future, which was a source of uncertainty for them. Three subthemes are included: avoidance; hidden worries; and comforting myself.

5.1 Avoidance

Most participants (4/6) avoided thinking about the future because it's 'upsetting' and 'scary' for them to think about things that might happen to their child:

"I tend to try not think about the future because I don't want to get myself upset when he is here, he's healthy at the moment so just enjoy…what's going on now, do you know, rather than look ahead…" (April)

"I probably put myself in a little bubble about it sometimes because I just…that's just the way I like to deal with it." (Louisa)

One participant said that although she avoided thinking about the future, there was a part of her that knew that she needed to think about it:

"But at the same time you do have to think about…think about the future I guess and what might happen but at the end of the day, it's just better to try and focus on the present I think." (Louisa)

In contrast, the male participant viewed time as precious and genuinely wanted to make the most of now as opposed to wondering about the future:

"It's obviously something that I think about. For me I just think it's important to think about today and next week and stuff like that…instead of thinking of years in advance…concentrate on the time that we do have with him…because quite frankly we don't know what's round the corner." (Chris)

His partner also recalled him saying something that sunk in:

"he said a comment to me and I've always remembered it and he said…do not grieve for the condition now, grieve for when he's gone…if he did die early then we think we'd get upset about it afterwards than worry…spend our time worrying now…" (April)

5.2 Hidden worries

Most participants (5/6) tended to avoid thinking about the future, yet they identified a number of worries that they had about what might happen suggesting that perhaps the avoidance is not always successful. The worries that participants (6/6) described were broad.

Participants (4/6) worried that their child's health will deteriorate as they get older:

> *"I think it's deterioration in his condition and development of other conditions on top of his CF…I know maybe he's going to be susceptible to things like diabetes and things. I do worry that he's going to need more and more different sorts of treatments and more investigations as he gets older…" (Carole)*

The possibility of having a lung transplant in the future seemed to play on most participants' minds:

> *"You see so many young children having, being put on the transplant list in their teens, so that is obviously one of the biggest worries…" (April)*

Another major worry for participants (5/6) was about whether their child will accept their condition and adhere to treatments as they get older:

> *"…there will be a stage where he refuses his treatment and there will be a stage where he will ask why and why he's different…I think it will become a bigger deal." (Chris)*

> *"I'm hoping when he becomes a teenager…whatever he rebels against, it's his homework or whatever else, the one thing he doesn't rebel against is his CF treatments…" (Fran)*

For one mother, the worry about her child's future adherence to treatments was even more significant because she knew a young teenager with CF who died after becoming non-adherent:

> *"She rebelled against her CF and she rebelled against her treatments, and unfortunately she (pause)…she paid the ultimate price for that rebellious nature." (Fran)*

She described the importance of educating her son about the treatments and adherence and has thought about using what has happened to the girl who died as an example:

"…if we can get the message across to him…maybe it sounds harsh, but something good could come out of her death, because it'll teach him and it'll teach others that this is what happen if you rebel and you rebel too much, it's, you're not going to beat them, beat this argument, beat CF-it'll beat you. It will always win the argument… 'Yes, you want to beat CF, but beat it in the right way, by taking your medicine, by doing your physio and not letting it get the better of you by being positive, not negative'…" (Fran)

Most participants (4/5) also worried about their child eventually finding out that they have CF and going on the internet where there are many negative stories. One worried about how to tell her child that they gave her CF:

"I think the hardest thing to tell her is that it's something that came from us…that we've kinda given her this." (Sophie)

Some (2/6) also thought about their own future and in particular they wondered about pregnancies and worried whether they would have another child with CF

"The other worry for us was having another child that had cystic fibrosis, so that was a concern…I just wonder whether other parents would be the same. They got one child with CF, maybe it's their first child and they worry about what to do…for future…future children." (Carole)

One very upsetting aspect of future pregnancies is the possible scenario of finding out the baby they are carrying has CF, and having to decide whether to keep the baby:

"We're faced with the decision of if we get pregnant and at the eleven weeks scan we find out if it's a CF baby or not, we got to decide whether to…terminate or keep." (Sophie)

5.3 Comforting myself

Some participants (3/6) who did say they thought about their child's future expressed their hopes that they will live a long time, although it was said with uncertainty:

"I think…if he takes responsibility and embraces it in the right way then I think he will live a normal length of time…" (Fran)

"Yes…I'm not sad about it really, I think…he's supposed…on his current treatment, if nothing changed, he's supposed to have a life expectancy of 50…" (Lorraine)

Some (2/6) hoped that their child may be able to live a near normal life:

> *"I think my children will live a near normal life. Apart from the fact that their lives won't be normal because to keep them living a normal life they will need to have a lot of medication and all the physio and the other things that keep them healthy…" (Louisa)*

Another expressed a hope that her child will embrace CF and be inspirational to others:

> *"I'd like her to be a 'strictly come dancing' dancer! (Laughter) that's what I keep telling her. But I think, I'd like her to be a bit of a beacon for CF, I would. I'd like her to embrace her disability…I would want her to proud of it more than anything I think." (Sophie)*

Although the same participant also described feeling saddened about knowing her child might not live a long time:

> *"I remember I walked into a shop and there were these two old ladies sat down eating their dinner and I got really really upset and had to leave…and (partner's name) mum said: 'what's wrong?' I said: 'She's never going to get that old, that life expectancy says 40, she's never going to get to be an old lady and do old lady things and go on Morecambe trips and stuff'." (Sophie)*

Core theme six: Quiet hopes and holding back

Core theme six describes what participants know about new developments and how they feel about them. Essentially there was a sense that participants had quiet hopes about future treatments but for particular reasons they were holding back from knowing more about them and investing hopes in them. Four subthemes emerged; A vague awareness of new developments; feeling pleased vs worry about funding; a healthy cynicism vs hopes for a new treatment; focusing on the now vs feeling hopeful for the future. There are also smaller themes described in some of the subthemes.

6.1 A vague awareness of new developments

This subtheme highlights that most participants (5/6) had a general awareness about gene mutations and a vague sense of how this might be connected to new developments in treatment. This subtheme is broken down into smaller themes as participants talked about different aspects of new developments and each one is described.

6.1.1. Knowing my child's gene mutation and class

Most participants (5/6) were able to identify one of their child's gene mutations:

"Ruby's is the most common. It's Delta F508." (Sophie)

One mother was aware that her child had two gene mutations and knew which mutation classes his genes belonged to:

"He has, he's got Class One, which is the unusual gene that my partner has, the 409 plus T, something like that, and the Class two, the deltaF508, so he's a mixture of Class One and Class Two." (Fran)

Some participants (3/6) recalled that they did not think it important to know their child's gene mutation or class before treatment developments emerged:

"It didn't make a difference to our lives…" (Lorraine)

"It hadn't been a high priority for us to find out…" (Louisa)

Since the development of the new treatment, there was awareness that knowing the child's gene mutation has started to potentially become much more important (4/6):

"But with the…appearance of drugs that do target a specific gene and I have heard of things like Ivacaf or I don't know if that's the same, anyway I've heard of various names, sometimes it's the brand names, sometimes it's something else…but I know there's lots of, lots of development going into new drugs that target specific mutations and so it is becoming more and more important that we do know what their gene mutation is… I think more recently it is becoming a priority so I have asked about it." (Louisa)

Another participant felt that knowing the gene mutation was important to prevent false hopes:

"If I didn't know which mutation he was…and then somebody came up to me and said: 'oh yes, if you've got this strand….the treatment in and they're going to be fine…and you're like I don't know what he's got and then you found out that it wasn't going to be…you'd be gutted wouldn't you?" (Lorraine)

Another said that she had always known that her child's gene mutation was very important, right from the initial diagnosis and before she had heard of the new treatments:

"I think it made a huge difference to me to know about Ruby's gene mutation, 'cos when we were told there was 150 genes, different mutations, I panicked it was going to be this gene that was in the minority and there were gonna be no research done on it, but it's not. I think we were quite relived to find out what gene mutation it was, because then we knew that it was the most common one to research, the most common one to look at for a cure, the most common one for other children so we could speak to other parents and they'd have the same gene. So it was very important to know what gene it were." (Sophie)

6.1.2: What I know about Ivacaftor and possible new developments

All participants (6/6) had heard about the new treatment (Ivacaftor otherwise known as Kalydeco), though they didn't always know the name:

"I've heard about the treatments that they're looking into…not sure of the name of it, I think it's…witcaledco and…another…I think…is it the iva..iva…something like that. I don't quite know…where they are in…how's it going." (April)

Some (3/6) felt that they had little bit of an understanding about how the new treatment might work and what form it might be in, although it was said with uncertainty:

"Is it the path…the protein that controls the cell membranes…is faulty so they get…the fluid exchange in the lungs and the other sort of…organs isn't as good. So….I think…the treatments for trying to open up these gateways so that they sort of improve…ions and things?" (Carole)

"It's some drug that will go into your genes, into cells and correct the fault, not a new gene. So correct the fault instead of replace the gene…and…yes…can't remember what its name is…corrector treatment is it called…Ivlacoror or…" (Lorraine)

Some participants (2/6) admitted that they did not know anything about how the treatment works:

"I haven't read into how it actually works or how they take the treatment…" (Louisa)

Though one participant was much more knowledgeable:

"…the Kalydeco which obviously helps the Classification Threes…" (Fran)

6.1.3 Not needing to know

Almost all participants (5/6) said that they had deliberately not enquired further into the new treatment after discovering that it was not relevant to their child's mutation:

> *"So I know about the new treatment, Ivacaf? But again I probably don't know that much about it because it…because it's not relevant, I don't feel it's that relevant to me. Although it's relevant to me in that it's a new treatment that's targeting a specific mutation, but it's not ours, it's not the one that affects Abi. I've sort of said I don't need…I don't need to have that information in my brain! I don't need to know too much about it because it's not going…it's not helping Abi specifically." (Louisa)*

One participant told how she had asked some questions about the treatment before stopping as she had decided it was not relevant just now:

> *"I've asked a certain amount, but if you like, we've shelved it until it becomes relevant. Because there's no point in asking all these questions about Kalydeco, because they're not relevant to us, we can't use it, it's not going to benefit Max at all, so no point in asking about it in so far as it helps the classification threes." (Fran)*

6.1.4: What I know about other possible developments

Some participants (2/6) reported some awareness of a possible new treatment that was currently in clinical trials:

> *"I know there's a study, is there a study going on at the moment because one of the dieticians was telling me about it. The study is going on for the similar drug that's already out there for the other mutation." (Sophie)*

One participant was very well informed about what treatments were being developed and their relevance for her son's mutation classes (one and two):

> *"There's nothing out there on the market yet to help those classifications, they're in the pipeline but they're not there yet. The urm, Altaluren which is being researched for Class Ones I believe is not successful as they hoped it would be for Class One mutations, they're still testing it but it's not effective as I think they hoped it would be. The other drugs, the ones they're working on to help Delta F508 and Class Twos, again we believe is not as helpful for Max because he hasn't got two Class Two mutations, he's a mixture of One and Two and the new drug they're working on just now apparently works very well for*

those who've got two Class Twos genes, they're not for those who've got a mixture." (Fran)

6.1.5 Gene therapy is still on the mind

For some participants (3/6), gene therapy – seen in the past as the next big breakthrough in treatment but currently much less discussed - was something that was on their minds:

"There's all of the gene therapy stuff that is going on that…that's a brilliant thing to be doing." (Louisa)

"Gene therapy was going to be where people with CF inhaled somehow or took into their body somehow, a new gene without the fault in and then…your body would just take instruction from the non-faulty gene. Brilliant. Sorted. But then that all went quiet and I don't think the doctors ever thought…I think they think it might still come eventually." (Lorraine)

One participant still saw gene therapy as potentially important, though far off, so it was important to make sure her child adheres to treatment so that they can benefit from it in the future:

"…to keep him…his lungs healthy so that if gene therapy is developed and it's effective and it's brought into clinical practice then he's going to be in a good enough condition, his lungs are going to be suitable to receive that treatment and it's going to be more effective than if maybe there was scarring and his lung function wasn't as good as it could be. So it's important for us to keep him fit and healthy." (Carole)

6.2 Feeling pleased vs worrying about funding

All participants (6/6) reported feeling really pleased about the new treatment and were happy that it was already benefiting some children with CF, even though it was of no benefit to their own child:

"Oh it's brilliant! I think it's really positive and I'm really pleased that there are people who can be treated with that." (Lorraine)

Some participants (3/6) also felt excited about what the development of this new treatment might mean for their own child in the future:

"I'm really pleased to hear when there's been a positive breakthrough in research and there's new developments, new treatments coming along. It's always really good news for me. It's a good outlook for him." (Carole)

"It's really positive and it's sparked further interest from the developers, pharmaceuticals and stuff…to develop further treatments so…it does make you feel that they are working towards a cure." (Louisa)

At the same time, several participants (3/6) also expressed concerns about whether the NHS would continue to fund future treatments:

"…I don't want to put my hopes onto it, just in case…it doesn't go well and then I think about the NHS in Wales when it got…when they refused to fund the treatment…they actually did a petition and now the NHS fund it. But one of my worries is because this new one that they're looking at is for the, is Oscar's mutation, which is the most common one, because there's so many people are the NHS going to want to put so much money into it when they refused just a tiny minority which was in Wales?"… "It's silly stuff like the budget cuts and recession and you've got to think, will they have the money, will they want to put it into CF when it's not well known and no one…speaks about it as well as some of the other diseases in the world." (April)

6.3 Healthy cynicism vs hopes for the new treatments

This subtheme highlights that participants (5/6) remained a little sceptical as to how much of a difference Ivacaftor and similar new treatments can make:

"I just can't see how a new drug can totally change, I don't know why." (Sophie)

"I guess… how much it's going to affect the people who can be treated with it, how great…how great is it really? There's obviously some results and that's why it's been used and has been approved but….they don't really know what it means yet do they?" (Louisa)

Some participants (3/6) felt it would be difficult to put all their trust in this one drug:

"I think it would be hard to put the… putting a trust in this one little thing which…you can't see the insides unless you're having regular x-rays on the inside." (April)

Most (4/6) thought that they would still carry on with pre-existing treatments even if they were still on the new treatment:

> *"I'd probably still do her physio, I probably would, 'cos I think I'd just want to do that extra bit, just in case anything happens." (Sophie*)

As a counterpoint to this scepticism, all participants (6/6) expressed hopes around what a new treatment that their child was eligible for would do for them:

> *"I would imagine he has a better quality of life, because even though he might live as long without the new drugs, if they came to light, I think with the new drugs his quality would be better because he wouldn't get as many chest infections, and those he's got, he'd be able to fight off bacteria and mucus a lot earlier." (Fran)*

One participant expressed a hope that a new drug would effectively take away their child's CF:

> *"What I think I would want is, I would want the new drug (short pause) I'd want the drug to make her like us." (Sophie)*

Another found it hard to identify how much a new treatment like Ivacaftor could help her child if he were taking it now, but appreciated his future could be different:

> *"It's hard to say because it's all could, should, would isn't it…his current life wouldn't change that much except for 20 minutes not having to do treatments, but his future might change yes, yes."(Lorraine)*

6.3.1 Willingness to try a treatment vs feeling uncertain

In the second part of subtheme 'hopes for new treatments' two smaller themes were also identified and each one is described. The first highlights that most participants (4/6) despite any scepticism were clear that they would be happy for their child to take a new treatment and believed that this would be safe:

> "*I'd say yes, I just want to go for it. Yes." (April).*

> *"I wouldn't say no to any magic drug that will make them much much better and be able to deal with their condition much better." (Louisa)*

Though others (2/6) remained uncertain and would need to know more:

> *"I think it's something that I couldn't jump straight into and yes…I'd want to know what it was and what it planned to do." (Chris)*

"I guess all the questions would be what are the long terms, whether there are the negatives in terms, not maybe in terms of CF, but maybe other things, how is it going to affect their other parts of his body perhaps. It may be great for his lungs and it might even help with his digestive system, but does it have any effect on his joints, are there any pay-offs shall we say?" (Fran)

One participant reported needing to have proof:

"if it was something where it was proven that we didn't have to physio and we didn't…he wouldn't get…his lungs wouldn't clog up and stuff like that then…I don't know…yes, I think I probably would…" (Chris)

Assessing the child's health at the time would be another important deciding factor

"…if he's coping very well with what's he got and there are possibly some negative pay-offs to go on the new drugs that I'm not aware off, then we might have to weigh up the pros and cons and then decide…" (Fran)

"I think it's purely about the state that Oscar was in. For me if he was healthy now…if they kind of said would you like to do it tomorrow if he was healthy now, then I don't know….I think it would be what state Oscar's in and how I felt when they told me about it." (Chris)

6.3.2 My child would still have CF

The second smaller theme; several participants (3/6) said that even if their child were on a new and effective treatment, CF would still be there, and some of the worries about the future that they shared earlier in the interview would still be present:

"I'd still always worry that she had cystic fibrosis and would still worry the same, even though the new drug would replace…I'd still worry about her getting poorly…because once it's in your head that's what she's got I don't, even if a drug helped her, I would still be thinking: 'well, if she's got a cold what if it went to her lungs and it damaged her?'…" (Sophie)

"I don't like to think about the future…getting to university, getting a job because I just kind of think well…we'll take it as it goes and if we'll get there then that's great and then you…I don't want to think about it and then get upset if one of these stages didn't happen so…" (April)

6.4 Focusing on the now vs feeling hopeful for the future

All participants (6/6) described the importance of focusing on the present. Their child cannot benefit from the new treatment so they still have a while to wait for the new drugs in clinical trials to become available:

> *"I think I'm just really at the moment trying to raise Harry as best as we can…dealing with the treatments, the current treatments that's available…but knowing in the background that there are new developments and hopefully they'll be ready for him for when he needs them." (Carole)*

One had decided there was little point in thinking too much about new treatments in clinical trial:

> *"For me it's…research is research, yes it's fantastic but it's not concrete and solid in the ground yet. I want facts and…research can obviously be developed and then fall…for me…I live for now and we…I…well me personally…I just kind of whatever's on offer now, that what we'll do then…if it's something in 10 years' time, then that what'll we do then." (Chris)*

Another preferred to focus on the here and now to avoid raising false hopes:

> *"I just focus on the here and now I suppose, I don't like to get my hopes up…I don't want to think there's something coming out that going to work and then either it don't work or the studies have not gone as they thought. I rather just think: 'This is how it's got to be', going forward and then something comes along that's a bonus." (Sophie)*

For several participants (4/6), part of focusing on the here and now involved continuing to adhere to current treatments, reasoning that if and when new treatments are available, the healthier their child is the more they will benefit:

> *"…you do your work now on his lungs and the physio and keep him healthy and then if something does come along in the future, his lungs will be in a pristine condition…to hopefully benefit the greatest if something did come along." (April)*

One participant, though, felt awareness of new treatments in development made no difference to current adherence:

> *"No. No because current treatments are still a million times better than if he was 30 years ago." (Lorraine).*

The second part of this subtheme reflects most participants (5/6) feelings of hope about future developments:

> *"It's only a matter of time before they find a combination of drugs that will help those with a mixture of genes…" (Fran)*

Several participants (4/6) reflected on their appreciation for how far treatment for CF has progressed and felt positive about the future:

> *"I'm very positive about how…how things have developed in the last 10 years and how they will continue to develop in the next years." (Louisa)*

> *"I was talking to a friend yesterday that there's a job for CF physio job coming in adults and in the past, the emphasis was on paediatric care in CF whereas there's obviously much more adults with CF now and they're living longer…so it just shows that things are looking better…"(Carole)*

> *"…someone's born with CF in 50 years, then they're just going to think…oh, you got CF, just have this drug and then you'll be fine. I bet you." (Lorraine)*

In contrast, one participant believed that CF will always be without a cure:

> *"I don't think there'll be a cure, I don't think that's what I'm expecting, I just think something to lessen the symptoms and make their day to day life obviously…a little easier…" (April)*

The mother of a child on Ivacaftor

The final participant's pen portrait and themes are now described.

Jade

Jade is 28 years old and is a full time housewife who lives with her fiancée. She has two children Luke and Millie. Luke is nine years old and has CF. Millie who is three does not. Her story of how Luke came to be diagnosed was very unusual. Jade described a very difficult beginning. She had another son who was older than Luke, but who sadly passed away. It was through the autopsy that they found out he had CF. Prior to him passing away, he and Luke were always poorly with severe chest infections that required medical attention. She recalled never understanding why they were ill and reached a point of not taking them outside for fear of more infections. The knowledge that her elder son had CF and died from CF related symptoms had led to Luke being tested at the age of two where it was confirmed that he also had CF. Although it was a very sad time

for her, she described feeling hugely relieved to finally have an explanation for why her sons were always poorly, and knowing that it was CF's fault, not hers for what had happened to her sons.

Luke has a gene that makes him eligible for Ivacaftor and at the time of the interview he has been taking Ivacaftor for one year. She viewed her son as being so lucky to have an opportunity to try Ivacaftor as she appreciated it was not something that was widely given. I could see that she really valued Ivacaftor and has seen how much it has helped her son. The biggest change for her was seeing him transform from a frail and exhausted young boy who always needed to rest and could not keep up with his friends, to a boy who is so full of life, always playing outside with his friends, and having his rebellious moments such as slamming his bedroom door when he was at home

I found Jade to be very warm and engaging, and someone who wasn't afraid to show her emotions. There were a number of times when she expressed sadness that made me really appreciate how much CF had affected her. She said that CF can be a very lonely condition to have and even though Ivacaftor has helped her son massively, the other aspects of CF such as not being able to mix with others who have CF or see parents of children with CF face to face is still very much an issue for them.

Jade's themes

Some of the themes that emerged from Jade's account were related to earlier themes from previous participants and described above. These themes have numbers in front of them to correspond with the earlier themes.

<u>1.0 The arrival of CF…two years late</u>

Compared to most of the participants (apart from Fran) who received the diagnosis of CF soon after childbirth, Jade's son was diagnosed two years later and only after her first son had died. She lived for two years with two sick boys, not knowing why they were poorly, and blamed herself:

> *"I always thought that it's my fault cause you know everyone else can keep their kids well and they're all running around and they got their coats off and it's not really warm to have their coats off…and mine always got sick and they always got poorly and they always got chest infections…they just seemed to pick up every virus that was going round."*

> "*We've got pictures and he just looks like he has just been starved…cause he was he had black rings under his eyes, he was poorly and when you look at his*

baby pictures you look back thinking how could you, how could we have not noticed or how could nobody else?"

1.1 Feeling nothing else but just relief

The majority of participants experienced strong feelings of shock and distress when they first discovered that their child had CF, but for Jade she only felt a massive sense of relief:

"*When we found out that Luke had CF it was…this is going to sound really bad but it was just a massive relief because at the time he was so small he was really frail he was really poorly you could just tell he never really thrived."*

2.5 Information overload and not knowing what to believe

Similar to previous participants, Jade described experiencing an information overload in the beginning but felt this also involved working out which stories surrounding CF to believe:

"we got overloaded with information…you know you've got all the facts about everything that's good and everything that's bad and got such a massive…you got one person saying 'oh I'm 35 and I'm fine and I got two children' and then you got someone else saying 'we never had children' or 'I lost my sister' or whatever…how can one side be so good and the other side so bad? It's just such a contrast."

2.6 Seeking vs avoiding information

Jade shared a similar view to several participants in that she initially wanted to know everything before eventually retreating from this, thinking that it's not important to know everything for several reasons:

Feeling afraid

"I don't think it's important to know everything about CF…I would rather not frighten myself with what could be or what could happen…I suppose a lot of it is fear, it's fear of knowing how things might happen."

It might not even happen

"It might not happen. This is the thing about it. Not everything happens, not everything happens to each child…even like none of the doctors I've ever spoken to said have said to me that Luke will be absolutely fine in the next three years but this could happen because you don't know that it's going to happen."

After her child had started Ivacaftor, Jade decided that she did not want to know about anyone else who has taken the treatment:

> *"I don't know anyone else that has the treatment erm I suppose it's a little bit by choice because I kind of didn't want to know I didn't want to…if it was bad or worse for anyone else or if it hadn't work because I wouldn't like to know, I wouldn't like to know anything bad…"*

3.1 Becoming aware of CF and beginning to take responsibility

As Luke is now nine years old, Jade said that he has a growing awareness that he has CF, although the gravity of it remains unclear to him for now:

> *"he knows that he has CF and he knows he has physio and medication for the CF but he doesn't know what it consists of or how much it means or doesn't mean to him, he doesn't, he doesn't know how serious it is I suppose he knows he's poorly because of it."*

Jade also described being in a state of transition where she is encouraging her son to take responsibility for his treatments, something that has not yet happened for other participants but one that was a future worry for them:

> *"he's just been given a nice new piece of equipment that does his own physio for him which is a nice thing for Luke to do but it's also teaching him to be responsible and doing this own thing and making sure it's done correctly and the way it's meant to be done rather than I've got five minutes I could be outside playing with my friend so I'll do it really quick and say I have done it."*

5.3 Moving away from having a healthy cynicism

Jade shared her initial feelings of wondering how much Ivacaftor could help her child, a similar reaction that previous participants described ('a healthy cynicism'):

> *"I think at first we were not sure it was going to work and you know, it's just a trial and it's not guaranteed or anything…"*

However, there was a sudden and visible change observed in her son:

> *"In the first two weeks you didn't kind of see anything then it was just all of the sudden he just kind of woke up one day and he was like running around and chasing around…and you know he was just like, I suppose he was just like every other little boy."*

Having her son take Ivacaftor was 'like a dream':

> *"You're always waiting for the one phone call to say: 'you're not going to believe this but we have found a cure, it's a miracle' and do you know, Luke is kind of like that at the moment. It's kind of like living the dream."*

A changing self through using Ivacaftor

Jade frequently reflected on what her son used to be like and often compared that to what he is like now:

> *"Luke was the one that kind of just sat and he was always really…he never really wanted to run off or run around and I suppose it was because he kind of struggled I suppose running around…"… "He just seemed to have no energy before whereas now he seems to be…not like hyperactive but he seems to just be more active and more like his friend…"*

She described the marked improvements that Ivacaftor already brought within Luke in a space of a year:

> *"You can totally tell…he's just grown in his shoulders and he's just bigger, he just seems to be bigger and stronger…it just seem to have made such a massive difference"…"now he's happy to go on his bike, we've had new tyres on his bike cause he's worn the tred down on the bike, and before…this bike we've had you couldn't even tell its been used, he could ride the bike…but he would just stay at the side of you and stop and you know whereas now he's way in front, you're shouting at him cause he's out of sight you know you're shouting at him and he's carrying on peddling with his friends and he's really keeping up with them all."*

Jade also said that Luke no longer hides how he's really feeling because since taking Ivacaftor, those feelings don't exist:

> *"He used to, he kind of sat behind a window basically watching his friends play and holding back and coming in because he's so tired and he's had to say to them I'm thirsty I'm going in for a drink when really he's coming in for a sit down because he's so tired and he didn't want people to know and would say to me, just don't tell them that, just tell them I've gone to the toilet…but he doesn't need to do that kind of stuff anymore."*

Comparing current treatments to Ivacaftor

Jade reflected that before Ivacaftor, she could tell that there was something wrong with him as he looked pasty but since Ivacaftor, he looked much different:

"you kind of knew and looking back that was obviously a really sad time but like I think you kind of pretend it wasn't there and it was all fine…which it was, it was ok, the medication was obviously working but it wasn't like how the Ivacaftor worked…you can totally see the difference within him; he's like, if you look at that picture there…he's totally, his face is fuller and that one there…you can see that he's pastier…paler."

Treatments can still be time consuming but Ivacaftor has changed me

Jade reported that Ivacaftor is taken on top of current treatments and the treatments can still be really difficult:

"It can take sometimes on a bad day it can take up to an hour just to get him through his medication…that the kind of difficult part of CF, it's the treatments."

But Ivacaftor has helped her to manage it all:

"…the Ivacaftor has made everything a lot more easier because it's made a change in me and Luke and you can kind of think you know he got to live with everything else but he's so much better with it. He's living life."

5.3.2 CF does still live on

Previous participants who expressed that they would still worry about their child regardless of whether they were on new treatment echoes Jade's observation that she continues to worry about her son's health despite being on Ivacaftor and making great improvements:

"I'm really happy that he's doing really well on the treatment but at the back of your mind you're always worrying and waiting for that one tiny little slip that can change absolutely everything."

CF is blinding and a lonely condition

For most participants, part of adjusting to CF involved putting into perspective and appreciating that it was not a visible disability. Jade's view was opposite which was further reinforced since being on Ivacaftor:

"…everyone says: 'oh it's like he's not even poorly I know that he has everything else but if he didn't you wouldn't know' and I'm thinking that's great, that's brilliant but it's very very lonely I suppose cause he can never

speak to anyone with CF so he doesn't know another child that feels like him."…"You have still got the loneliness of CF…"

"It's frustrating because it's quite like I say for children to see it you know cause they don't see other children with CF because it's not something you see it's not something you visually see…and it's quite difficult because its blinding, it's kind of like people walk in a blind because they don't see it."

4.0 Future worries

Jade did not identify many worries about her child's future apart from one main worry which was that she was worried about how her son will cope with treatment adherence in the future, a worry that was expressed by most of the other participants:

"…and with CF you're always thinking about what they would do, how will they cope on their own and how will they manage to do the medication correctly you know how will they feel about it doing in their own home with other people and if they went to university how will they cope with doing it in front of strangers."

CHAPTER V

7.0 DISCUSSION

The first section of this chapter will revisit the research design and aims, before it will summarise the main findings, discuss the relationship to the literature, and provide an overall summary. The second section will focus on methodological considerations, before clinical implications and areas of future research. A conclusion will then follow.

7.1.1 Revisiting research aims

The current study aimed to explore parents' knowledge of the genetics of CF, views about current and future treatments, and thoughts about their child's future. Two methods were utilised; a short questionnaire which was completed by 30 mothers who attended the Leeds paediatric CF unit, and semi structured interviews with 7 parents that were then subjected to TA. The questionnaires were used to situate the sample and provide some background information on parental knowledge of CF. The qualitative analysis from the interviews were used to explore further parents' understanding of CF, their experiences of using current treatments, and how they viewed treatment developments in CF.

7.1.2 Summary of findings

The qualitative analysis has generated a number of themes; an unknown territory, struggling with paradoxes, information overload, taking control but feeling uncertain, dealing with an uncertain future, and quiet hopes and holding back. The first three themes emerged independent of the interview schedule and do not particularly answer the research questions, however they were included in the analysis as they provide a context for the rest of the themes. It felt appropriate to merge these themes into a narrative that describes how participants coped with the diagnosis and adjusted to CF, although they are described separately. This narrative is shared first before the remaining themes which particularly address the research questions. Finally, the view of the mother of a child on Ivacaftor is summarised and discussed.

Core theme one: An unknown territory

This research was not intended to focus on parents' experiences of receiving their child's diagnosis and how they adjusted to CF but it felt important to include this in the analysis as all participants used the day that their child was diagnosed as a starting point in the interview. This is not surprising given that research has shown that the diagnosis of CF can be a life-changing event for parents (e.g. Carpenter et al, 2004; Glasscoe et al,

2008). For the majority of the participants, the initial diagnostic period was characterised by shock and distress. One participant also expressed grief, which has previously been reported in the literature as parents acknowledge several losses, such as the loss of their child's health, freedom to live life as they wish (e.g. Lowes et al, 2004), and the loss of the future as it becomes dependent on treatments and the child's adherence to them (e.g. Jessup et al, 2010). Following the initial diagnosis, most participants described a period of feeling negative emotions including grief and depression that lasted some time, and further described that they isolated themselves away from others. Indeed, one participant described that in the beginning she felt reluctant to go outside as she did not want people to see how much pain she was in over her child's diagnosis. A couple of participants also shared that they felt unequipped and overwhelmed by the prospect of looking after a baby who was sick.

All participants felt that they initially tried to manage by recalling what they knew about CF, with some having no knowledge or understanding of CF before they started to mentally prepare for the worst. Several participants reported that they knew someone who had CF but said that they did not have a well-developed understanding. It seemed that participants then began to fear the worst and the fear of deterioration and death was significant at the time of diagnosis, followed by their worry about their child's capabilities, and how life was going to change. Parents' fear of their child dying following a diagnosis is not uncommon (e.g. Jessup et al. 2010; Moola et al 2011). Alongside this fear, one participant reported that she was worried about her personal and professional identity and thought that she would have to give up her interests and job. The role of parents entering into a full time caregiver and the losses that they experience that define their identity such as friendships, interests, and career has been frequently documented in the literature (e.g. Hodgkinson et al, 2002). However, not all participants dwelled on the possibility of death. One participant instead spoke about having an appreciation of the time he has with his son, and another participant described feeling determined to beat the odds and keep her child alive for as long as she could.

Core theme two: struggling with paradoxes

As participants slowly adjusted to the presence of CF, they described their daily struggles with paradoxes. Most participants described that their day to day life was normal and that they generally forgot CF existed, although a couple shared that they still had days where they were sad about the diagnosis. It was only with a physical sign of CF such as a cough that CF suddenly came back into participants' reality. There appeared to be contradiction as although participants spoke of forgetting that their child's CF existed, they still described their general worries on a day to day basis such as

their child's appetite and exposure to germs. Furthermore, as part of bringing normality to their lives, participants all spoke of refusing to let CF take over and hold their child back from living life akin to their non CF peers. Even then, there was another contradiction as participants placed restrictions to protect their child's health. In essence most participants tended to move between forgetting CF exists to worrying about aspects of CF such as their child's diet and monitoring their health, and from being adamant about preventing CF from holding their child back from living a 'normal' life to placing restrictions on them; for example, one participant described not letting her child go to nursery and interact with other kids. There was a sense that participants really struggled with CF at times. On one hand they were desperate to construct some sense of normality in their lives, to allow their child to live life normally, and to protect their child's freedom from being taken over by CF. On the other hand, their worries about CF and their child's health declining frequently took over their desire for normality and they found themselves placing restrictions on their child to protect them.

Through time, participants reported that they eventually tried to make the best of the situation and their on-going struggles with paradoxes by using a number of coping strategies that they used to adjust to CF. This included having an appreciation that CF happened to their first child. Several participants felt that this helped them to adjust to CF because they could not compare life with a healthy baby to a life with an unhealthy baby, and this in turn enabled them to construct their own sense of normality. Parents' perception that life with their firstborn baby who has CF is 'normal' has been widely reported in the literature (e.g. Glasscoe et al, 2011). Another coping strategy was to compare CF to other conditions and put it into perspective. All participants expressed gratitude that CF was not a physical or an 'obvious' disability, echoing a theme from Moola's (2012) study on parents of children with CHD and CF, 'learning to put things into perspective', where parents compared their child's condition against others who were thought to be more ill. This in turn helped them to view CF as being manageable and bearable to live with.

Core theme 3: Information overload

A part adjusting to CF involved participants managing their information needs. All participants reported that they searched to understand CF in the beginning, with several stating that they attended an information clinic at the CF unit the day after they received their child's diagnosis. From here, participants quickly reported that there was an 'information overload' about their child's CF, and CF in general. They described feeling overwhelmed by the amount of information that there was and the realisation of 'what CF meant', and further felt distressed by the knowledge that their child could experience

endless health problems. Participants described engaging in seeking vs avoiding information behaviour as a coping strategy to manage their information needs. They seemed to managed their information needs by asking themselves two questions; 1) will it cause me emotional harm and 2) is it relevant to me now? A discussion of the subthemes information overload, seeking vs avoiding information, and relevance is provided next.

Information overload

Parents' information needs regarding their child's chronic condition has not been extensively explored in the literature although this is starting to change. Research has highlighted that providing information can help parents to adjust to the shock of a diagnosis, and make treatment decisions (e.g. Lambert, 2009). However, there is an increasing recognition that parents' information needs vary and that excessive information can have an undesired effect. Fisher (2001) investigated parents' needs for information through conducting a review of literature on chronic illness and generated three main themes; 'the need for normality and certainty', 'the need for information', and 'the need for partnership'. The first two themes were of particular interest. The 'need for normality and certainty' described parents feeling overwhelmed by the lack of control that they had over their child's illness during the initial diagnosis. One way for parents to regain normality was to take control over issues that were within their jurisdiction and this included management of information. Within the theme 'the need for information', Fisher (2001) summarised that overall parents were keen to develop an understanding about their child's condition, however they often reported their dissatisfaction with the information provided by health care professionals. Parents' reasons for dissatisfaction included receiving an insufficient amount of information (e.g. Cohen, 1993, Jerrett et al, 1996) and receiving too much information quickly (e.g. Diehl et al, 1991). Fisher (2001) has provided an understanding about parents' need for normality and certainty following the initial diagnosis that they attempt to achieve through managing their information needs. Some of the findings discussed echoes the experiences reported in the current study. A limitation that Fisher (2001) acknowledged in her study was that the studies reviewed were from the USA and Canada and therefore they have different healthcare systems compared to other countries such as the UK. Fisher (2001) appreciated and cautioned that parents' need for and experience of receiving information might differ across different countries.

A more recent study following Fisher's (2001) focus on parents' information needs was conducted by Hummelinck (2006) who explored the information needs of 20 sets of parents of children between the ages of 0-16 with a chronic condition. Semi-

structured interviews were employed which were then subjected to content analysis. The chronic conditions involved cancer, asthma, leukaemia, epilepsy, diabetes, severe eczema, and CF. The authors reported four main themes; 'the development of parents' need for information over time', 'the nature of parents' need for information', 'reasons for wanting information', and 'their opinion on the adequacy of information provided'. Of particular interest, the first theme described parents' needs for information changed over time. At diagnosis parents reported the difficulty to process information due to being unable to comprehend medical vocabulary used and to feeling devastated at having the diagnosis in the first place. They also reported that there was an overload of information which led them to feel confused and anxious. As parents gradually developed confidence over time, they began to identify their information needs and sought to meet them. From here, Hummelinck (2006) distinguished two types of information that parents were dissatisfied with which were similar to Fisher's findings (2001); inadequate information and information overload especially during the diagnostic period. Interestingly the former tended to be reported by parents of children with a diagnosis that could be managed in primary care such as asthma, while the latter tended to be reported by parents of children who had a diagnosis that required multidisciplinary input such as leukaemia and CF. Parents reported that information overload led to increasing feelings of anxiety and insecurity about their ability to cope. Of further interest, Hummelinck et al (2006) said that parents' wishes for information were not taken into account as those with children whose chronic condition demanded a high level of support from healthcare professionals (e.g. CF) were given more information despite not asking for it, compared to those with children whose diagnosis required less support. The participants in the current study have reported similar experiences of being overloaded with information, and described the process of receiving information as both difficult and overwhelming. The second theme in Hummelinck et al study (2006) highlighted that parents had multiple reasons for wanting to know different types of information; for example, some parents said that it was important to know everything in order to prepare themselves for the future and to be able to answer their child's questions about their condition. In the current study, most participants reported feeling the opposite and described their fear of knowing about negative events that might happen to their child as driving them to avoid knowing too much about CF. In contrast, the male participant in the current study shared the same view as some of the participants in Hummelinck et al's study (2006) in that he wanted to know everything to prepare himself for what's around the corner and to be able to answer all of his son's questions. Hummelinck et al's study (2006) discussed the importance of professionals recognising parents' needs for information and tailoring the

provision of information accordingly, however appreciated that parents themselves often do not know their information needs might be until much later on.

Seeking vs avoiding information:

The majority of participants adopted a seeking vs avoiding information stance that emerged from the initial diagnosis and continues. Their descriptions of information overload during the diagnostic period may have shaped their strategy of seeking vs avoiding information which could influence how they currently process information on CF and new developments, manage day to day life with CF and approach thinking about their child's future.

Participants in the current study reported that they initially searched for information to understand their child's diagnosis in several ways including asking questions to the paediatricians, reading educational books, and/or using the internet to learn about CF. This seeking of information is not uncommon; for example, parents of children with leukaemia were keen to know everything at the diagnosis (Patistea et al, 2003). Research has highlighted that information seeking is a coping strategy adopted by some individuals in order try to resume control and reduce feelings of uncertainty over their chronic condition (e.g. Eherman et al, 2009).

In the current study, participants' initial desire to understand CF changed to avoidance as they became aware of the gravity of CF and the likelihood that their child will encounter a range of health problems that could lead to an overall steady decline in their health. Consequently, this caused them great distress and from here the majority of participants adopted an avoidance strategy. Fisher (2001) in her study cited Cohen's belief (1993) that parents actively limited information about their child's illness that was viewed as having the potential to cause them emotional harm and incapacitate them. Another way participants in the current study managed their information needs was to actively seek positive stories about children with CF who were doing well. This could be to instil hope in the face of a harsh reality that CF might bring to them.

A recent study by Toruner et al (2013) explored information seeking behaviour and decision making processes in 15 parents of children with cancer who were hospitalised, using semi structured interviews that were then analysed using inductive content analysis. The authors were particularly interested in exploring factors that influenced parents' search for information on cancer and associated treatments. Two core categories 'information seeking' and 'decision making' along with six themes were identified. Within the former core category, parents described a need to learn about their cancer, their child's prognosis and treatments management. The authors highlighted several factors that influenced parents' information seeking behaviour with the most

significant factor as having inadequate information about their child's condition in the first place. In contrast, participants in the current study spoke more about dealing with excessive information and from there they engaged in avoiding information. Based on participants' experiences, two main reasons for avoiding information were identified (a) information overload and (b) to prevent self from feeling afraid and distress. Participants reported that they used the question of relevance to judge what information they need to know and what information they can disregard.

Relevance:

Under the subtheme knowing everything vs knowing what's relevant, all participants identified that they tended to judge information that they need to know based on its relevance to them personally and whether they had time to read it. This behaviour has continued since diagnosis; in relation to Ivacaftor, most participants expressed that that they did not need to know about it because it did not target their child's gene mutation and therefore could not provide any benefit for them. Indeed one participant described not wanting to hold that extra information in her head when she knew it was not going to benefit her child personally. With regards to new treatments in clinical trials, one participant discussed not wanting to know until there are concrete facts and evidence that the treatment works for their child. In contrast, one participant seemed to have a good understanding about different drugs in clinical trials and whether they will benefit her child. Hummelinck's (2006) and Toruner's (2013) view that parents have different individual information needs is supported by the findings in this study.

Several participants also identified that they generally liked to keep up to date with some of the latest research on CF, but described that they would skim through it as and when the opportunity presents itself and they wouldn't follow it as it would be too time consuming.

- *Models of coping*

Together the themes 'an unknown territory', 'struggling with paradoxes' and 'information overload' describe a process of adjustment that participants went through. Participants' experiences suggested that they went through a number of stages like the ones suggested in the literature; for example, Carpenter and Narsavage's (2004) model of 'falling apart, pulling together, and moving beyond' mentioned earlier.

Stephney et al (2011) proposed a more recent and universal conceptual phase model to explain how parents cope following their child's diagnosis of a chronic condition and used asthma as an example. A key strength is that this model is based on the authors' review of the literature on parents' experiences of paediatric chronic illness which led

them to identify three key phases that parents go through following the initial diagnosis. In the first phase; 'emotional crisis', parents may experience a range of emotions including denial, grief, self-doubt, and powerlessness. The second; 'facing reality' occurs when parents start to deal with their emotions from the initial diagnosis and try to resume normality in their lives by regaining control over their child's illness and bring order to the disruption caused (Maltby et al, 2003). Parents attempt to achieve this through adopting a number of coping strategies, and a key strategy is to manage their information needs and seek information about asthma and asthma management (e.g. Trollvik et al, 2004), similar to what participants in the current study reported. The final phase 'reclaiming life' occurs when parents begin to feel confidence in managing their child's condition and treatment tasks. Here they start to create new routines that merge living with a chronic condition and its demands with the normal day to day life in order to bring stability to their lives. The authors also proposed that phases are not linear; instead parents may move between different phases at different times because chronic conditions such as asthma are episodic in their nature; for example, the child may be well the majority of the time but unexpectedly their health may suddenly deteriorate and lead to hospital admissions. This may generate previous feelings of distress and powerlessness in parents that were felt when their child was first diagnosed and/or they may actively seek information again over what caused their child to become unwell and treatments needed to regain control. A particular strength of this model is that the authors reviewed the coping strategies that parents described across a range of chronic conditions as opposed to only focusing on one type of chronic condition.

- *The concept of chronic sorrow*

Participants in the current study described a process of making a gradual adjustment to CF and gave an indication that they have accepted CF as a part of their lives. However, for the majority of the participants CF still managed to evoke tears throughout the interviews. This might be unsurprising for several participants whose child's diagnosis of CF were fairly fresh, but there were other participants who have lived with the diagnosis for some time including one participant who had 7 years post diagnosis. This perhaps reflects the nature of participants' adjustment to and acceptance of CF. The concept of chronic sorrow developed by Olshanky (1962) could offer some insight into why participants expressed sadness in the interviews when they talked about their child's CF. Olshanky (1962) first introduced the term chronic sorrow to explain the recurrent sadness of parents of children with impaired cognitive abilities. The sadness was sporadic and emerged when there were reminders of their child being heavily dependent on parents to care for them and the loss of a fantasied child (Gordon, 2009).

Chronic sorrow has typically been explored in parents of children who are less able mentally and more rarely in parents of children with a chronic condition. Bowes et al (2008) interviewed 17 parents of children with type 1 diabetes and explored their long-term emotional adaptation to their child's diabetes 7 to 10 years post-diagnosis. The authors found that most of the parents had unresolved sadness from diagnosis and critical periods such as hospitalisations and health visits brought up the devastation that was felt at the time of the diagnosis to the surface. Although CF has not been explored in relation to chronic sorrow, it is likely that there may be features of chronic sorrow among the participants in the current study. As participants recalled their child's diagnosis and previous hospital admissions, and spoke of reminders of CF, they became visibly distressed.

The next three themes are summarised and discussed in relation to the literature and the research questions.

Research question 1:

- *What are parents' beliefs and views about current treatments and what do they perceive their children's' futures to be like*

The themes 'taking back control but feeling uncertain' and 'dealing with uncertain future' best answers research question 1. The former theme is described and discussed first before the latter.

Core theme 4: Taking control but feeling uncertain

In essence, all participants described the importance of their child's adherence to treatment, although they also questioned and worried about the efficacy of the treatments. The mediating factors to ensuring their child's adherence appeared to be their fear of what might happen to their child if they did not take treatments, and more importantly their trust in the experts. Participants also reported having mixed feeling about current treatments and further identified treatments that they liked and disliked. The process of administering treatments was described as being 'normal' by participants, although they shared their frustrations that treatments were often demanding and time consuming. The subthemes are discussed more in-depth below with links to the current literature.

Adherence

Participants in the current study described what their child's daily treatment regimen entailed and were also able to explain the purpose of each treatment for suggesting that overall they had a good awareness about current treatments. Adherence

was viewed as being paramount by most participants in the current study, although for some this was not always put into practice. One participant reported having approximately *"10 treatment days off a year"*. Several participants also recalled times when they had missed treatments due to time constraints and forgetfulness that led to feelings of guilt after. Participants also had mixed views about the time it took to administer treatments. One participant described treatments that took a matter of minutes, whereas another reported it took quite some time. Interestingly, one participant also said it depends on whether particular aspects of treatments are viewed as 'treatment' such as meal times.

Adherence to treatments (including in CF) has been extensively examined in the literature, however the focus has tended to be on adults with chronic conditions, although understanding the rates of adherence in children has started to develop and parents' perspectives are becoming incorporated (e.g. O'Toole, 2012) especially in paediatric populations where the child's adherence to treatments is dependent on the parents. Research has suggested that parental involvement is one explanation for why childrens' rate of adherence is higher than adolescence (e.g. Foster et al, 2001; Modi et al, 2008). When children reach adolescence their adherence to CF treatment often (though not always) decreases (e.g. Zindani et al, 2006; Riekert et al, 2007; Masterson et al, 2011). Lomas (2014) conducted a recent review of available research on non-adherence in CF and reported that generally there is a higher rate of adherence to simpler treatments such as enzymes therapy and exercise (e.g. Abbotts et al, 1996) compared to treatments that take longer such as ACTs and nebulisers (e.g. Abbotts et al, 2009). A possible explanation for participants' mixed views on treatment time in the current study could be based on treatment tasks prescribed. Participants of younger children were required to administer nebulisers which were reported to take longer, compared to participants of older children who did not need to take them.

Worry over treatments and trusting the experts

The majority of participants stressed the importance of adherence and yet all questioned current treatments' effectiveness and expressed worry over whether they worked. Participants reported that there were particular treatments (mainly physio and antibiotics) that were difficult to understand in terms of how they helped their child, compared to treatments such as Creon where immediate benefits could be seen as their child quickly put on weight. A possible hypothesis offered is that most participants in the current study did not view their children as being 'poorly' because they did not exhibit many symptoms of CF and therefore this may have led them to question whether treatments were helping. Indeed one participant stated that her child did not even seem

to be poorly in the first place. Children with CF generally do show fewer symptoms than adults with CF because the condition has not usually deteriorated. Despite the worry over treatments' effectiveness, participants ensured that their child took them. This finding is different to previous research such as Conn et al (2005) who examined parents' attitudes to asthmatic treatments and found that their concerns about prescribed treatments resulted in them stopping their child from taking them.

There may be two mediating factors that could explain participants' insistence of administering treatments despite feeling uncertain over them. The first was their fear that stopping treatments would be harmful to their child and the second was their relationship with the CF team. With the latter, all participants frequently praised the Leeds paediatric CF team for knowledge, care, and dedication to their child that helped them to trust them. Research has highlighted that trust and communication are two significant elements within the context of parents' relationships with health care providers (e.g. Thorne et al, 1993; Kirschbaum et al, 1996). Pyke-Grimm et al (2006) explored 36 parents of children with cancer treatment decision making (TDM). The authors employed semi structured interviews that were then subjected to content analysis. A number of themes were identified as having a positive influence on the parents' TDM. Of a particular interest, the theme 'parent-physician' relationship was described. Within this theme, some parents identified that physicians' knowledge about cancer made it easier for parents to allow them to have the final say over what treatments their child should have. Parents also reported they had a good relationship with the physician who kept the parents informed and asked their opinions about their child's treatments. Furthermore, parents identified that physicians who were approachable and very supportive influenced the decisions that parents made about their child's treatments. Participants in the current study have all reported similar views with the Leeds team and it may be that the quality of this relationship helped parents to trust the team and follow treatment recommendations even when they had doubts.

Process of administering treatments

Several participants reported that they had initial concerns about administering treatments to their child, including wondering how their child will sit still and take the nebulisers, and where they would do physio when they were out in public. A feeling of worry about remembering to administer treatments and ensuring that these were done correctly was reported by several participants. Indeed one participant spoke of using a chart to record the treatments that she needed to administer for 7 months before she felt confident enough. This was associated with participants' sense that they were solely responsible to keep their child healthy against a serious condition. In the literature

parental concerns about treatments have tended to be about whether treatments are necessary and safe for their children (e.g. Conn et al, 2005; Conn et al, 2007). In contrast, little research has explored parents' views about their ability to administer the treatments themselves. The current study has started to shed some light on this.

Over time, most participants reported that they became accustomed to the treatments and felt treatments were normal for them. Indeed one participant said that treatments were second nature. For some, they reiterated that having their firstborn baby with CF helped in some respect to adjust to the act of giving treatments and perceive this as normal. They identified that they did not have any means of comparing their current life and so did not know any different. Some participants viewed several CF treatments as 'normal behaviour', and no different to what the rest of the non CF population should be doing; for example, exercise (even though the cost of not doing exercise and the demand of doing them is greater as without exercise mucus becomes clogged and can cause life-threatening respiratory problems). Normalisation was also achieved when participants reported their childrens' view of treatments as normal and when their children were co-operative with taking treatments. Normalising medical treatments and incorporating them into their daily routine has been highlighted frequently among parents in the literature (e.g. Hodginskon et al, 2002, Glasscoe et al, 2008). The concept of normalisation has been rooted in the literature on chronic illness for decades (e.g. Anderson, 1981; Deatrick et al, 1990; Rechner, 1990). Robinson's research (1993) although relatively old, provides some understanding about how parents of chronically ill children use normalisation to manage day to day care of their condition through adopting a 'normalcy lens' where they pay more attention to normal aspects of their lives, and de-emphasise difficult aspects associated with the chronic condition. Robinson (1993) found that parents used the skill of minimising issues associated with a chronic condition and reframing to help them normalise their experiences. In her research, she discussed an example of a father who minimised difficult issues associated with his son's asthma by viewing his son (who spent a long time in a hospital with life threatening allergies, eating problems and asthma) as normal: "he's normal, it's just his throat". In the current study, although it was not explicitly reported, the skill of minimising may have been adopted by some participants, as one participant added that there was not much to do compared to a parent of a child without CF, after she described in detail her child's daily treatment regimen.

On the other hand, whilst participants worked hard to construct their own sense of normality and largely viewed treatments as being fairly easy to administer, they reported that treatments were time consuming and demanding. Research has reported that CF respiratory therapies alone can up to several hours per day for patients (e.g.

Hunter et al, 2003), and the view of treatments being time consuming has been reiterated throughout the literature on other chronic health conditions (e.g. Peterson-Sweeney et al, 2003; Tong et al, 2010). It is therefore unsurprising that participants in the current study reported similar views. One participant said that she could not switch off until the evening when all the required treatments were completed, a feeling that was also reported in Jessup et al's (2010) study with CF patients and their parents, as they described CF as a battle that was *"not a Monday to Friday one with weekends off".* Nonetheless, one participant, though they also shared similar views to other participants, appreciated that the demands of administering treatments multiple times daily was a small price to pay in exchange for her son being as healthy as he can be.

Feeling ambivalent

Parents' views of current CF treatments have not been widely investigated in the literature and the current research has attempted to address this. Overall a theme of feeling ambivalent about current treatments was found. Participants had mixed feelings about different types of CF treatments used and there was a sense that some treatments were more liked than others. Treatments that allowed participants or significant others to spend time with the child and have the potential to be fun and enjoyable such as exercise were particularly liked. Equally, Creon was liked by participants because they could see the immediate benefit of their child taking them. Treatments that participants felt were relatively easy to do such as ACTs were tolerated and most reported that they used the usual chest tapping physiotherapy. Participants' reports of chest physiotherapy being tolerated and as generally okay are different to a previous study by Williams et al (2007) who explored children, adolescents and parents' views of chest physiotherapy using semi structured interviews. The authors found that chest physiotherapy was viewed as restrictive and problematic by the participants.

Treatments that were difficult to administer and caused their child discomfort such as nebulisers, blood tests, and x-rays were disliked by participants in the current study. There was a sense these treatments were viewed as invasive and as difficult to explain to their child why they were needed. This view has been documented among parents of other chronic conditions for example, Tong et al (2010) interviewed 20 parents' perspectives of caring for their children with a chronic renal disease. Using thematic analysis, the authors reported 4 main themes, and of particular interest the theme 'absorbing the clinical environment' highlighted parents' dislike of invasive procedure as they described that they felt helpless and distressed seeing their child have needles and tubes inserted in them.

Core theme 5: Dealing with an uncertain future

Core theme five describes participants' three main approaches to thinking about the future (a) open avoidance, (b) comforting themselves and (c) quietly worrying about key stages they may encounter as their child ages.

Parents' views of their chronically ill child's future are becoming explored and this current study contributes to this. The majority of participants reported that they did not tend to think about the future and a number of reasons were shared as to why. The main reason provided was fear of looking ahead and becoming aware about negative events that their child might experience. Indeed, the process of thinking about the future was both a scary and an upsetting one for most participants. Several participants reported that there was no point in thinking about the future and preparing themselves because everything bad related to CF might not actually happen to their child. There was a sense that participants would only think about the unpleasant events that might happen in their child's future if and when they were confronted. A couple of participants describing refusing to think ahead because they were unsure about whether their child might reach different stages in life for example, they did not want to think about their child attending university because they were not sure whether their child will live that long. Instead, most participants stated that they preferred to live in the present. Although it was not explicitly said by the participants in the current study, previous research has highlighted parents' views that their child's future was lost at diagnosis (e.g. Glasscoe et al, 2008, 2011, Jessup et al, 2010) and it might be that the participants felt the same as they could not bring themselves to think about the future. In contrast, one participant, the father, described genuinely wanting to make the most of now which involved not dwelling on thinking about the future. He reported that he did think about the future, but preferred to concentrate on the time that he has with his son and he described the importance of making the most of life properly, which also involved being spontaneous. His reason for not thinking too much about the future was different to previous research on fathers of children with CF who reported that they avoided thinking about the future because they did not want to acknowledge their child's deteriorating health (e.g. Peck et al 2005). Instead, the male participant in the current study echoes previous research such as Moola (2011) who highlighted that within the context of a reduced life expectancy, fathers particularly emphasised the importance of spending time with their children who had CF.

Despite participants' reports of not thinking far ahead, most described their hopes and beliefs about their child's future e.g. *"I think…if he takes responsibility and embraces it in the right way then I think he will live a normal length of time…"*, suggesting that they perhaps do not strictly avoid thinking about the future as they had

described. As most expressed their thoughts about their child's future, there was a sense that it was said without conviction suggesting that they did not believe it would happen. Instead, from listening to the participants and observing their body language, it felt as though they were comforting themselves and trying to be optimistic about their child's future. Most participants tended to describe a hope that their child will live long and did not really express hopes about what they would like their child to do as they become older. In contrast, one described a clear hope that her child will become a beacon for CF and inspirational to others. She wanted her to be proud of her disability and not view CF as something that holds her back.

The final subtheme highlights that participants identified a range of worries that they had about their child's future suggesting that despite their best efforts to avoid thinking about the future in order to protect themselves from becoming distressed or to comfort themselves by thinking positively, they had privately thought about the things that they were worried about. Participants' main worry centred on whether their child will accept their condition, take responsibility to adhere to treatments and maintain their health such as following a healthy diet. There was a worry that their child might rebel against their treatments and as part of managing that, almost all participants had reported that they were keen to educate their child about their treatments and the importance of adherence. Linked to the need for educating their child about CF in the near future, some participants described their worry about how to tell their child that they have CF and where it came from. Indeed, one participant stated that she did not know how she was going to tell her child that her CF was inherited from her. Another main source of worry was the fear of their child's health deteriorating as they age. This included both the worry that their child will develop other conditions for example diabetes, and worry about needing more complex and invasive treatments such as lung transplant. Other worries included thinking about their own future pregnancies and whether their next baby will have CF. There seemed to be a particular stage during a pregnancy that the participants worried about getting to which was having a scan and finding out whether their child has CF or not. Participants worried about the decision that they will be faced with; the decision to keep or abort the baby.

Together, the three subthemes describe an overall narrative where at one end of a spectrum participants avoided thinking about the future mainly due to fear of what negative events their child might encounter and to protect themselves from feeling distressed. At the other end, participants described their hopes that their child should live a long life, although it was said with a sense of uncertainty and could be seen as their attempts to comfort themselves against the harsh reality that CF is a life limiting condition. In between both ends of the spectrum were a range of specific worries that

participants had about their child's future which felt 'hidden' as participants needed to be prompted and this was in contrast to their earlier descriptions of avoiding the future.

Research question 2 and 3:

- *What do parents know and understand about CF and in particular the genetic characteristics influencing the presentation in their children?*
- *Whether knowledge of new developments in treatment for CF changes parents' beliefs about their child's CF and the importance of current and future treatment*

The theme 'quiet hopes and holding back' best answers one of the key research questions on knowledge of the genetics of CF and views about new treatments. Included in this theme are parents' hopes of how a new treatment can help, and the importance of maintaining adherence.

Core theme six: quiet hopes and holding back

This is the first study to attempt to understand participants' views in light of the new generation of treatments such as Ivacaftor, and consequently there is no comparable research available. Research on Ivacaftor has thus far focussed on clinical outcomes (e.g. Davies at al, 2013).

Most participants were able to identify one of their child's gene mutations, and it is unclear whether they were reporting the common gene mutation or if they only knew one of the two gene mutations. In contrast, one participant reported both gene mutations of her child and the mutation classes that the genes fell into. Knowing their child's gene mutation has started to become important to some participants, as they had recognised that treatments are changing to become more focused on gene mutation and/or class mutation specific.

All participants reported that that were aware of Ivacaftor but this seemed to be undeveloped as some were unable to correctly identify the name, and several expressed having a poor understanding of it. Most participants stated that they did not want information on Ivacaftor because it did not target their child's gene mutation and therefore it was not relevant to them. In addition, they had minimal awareness that there were other drugs in clinical trials but again chose not to know too much about this. This need not to know is predominantly linked to an earlier subtheme described; 'knowing everything vs knowing what's relevant', however some participants also described not wanting to know about other treatments in order to protect themselves against false hopes. Indeed one participant reported that she did not want to find out about drugs in

clinical trials in case she unintentionally invested hopes in it only to discover that the treatment did not work. Interestingly for several, gene therapy was discussed first by at least three participants when they were asked what they knew about new developments, before they started to talk about Ivacaftor. Although they recognised that gene therapy was far off from development, they expressed a hope that it will materialise. Indeed, one participant described the importance of child's adherence to treatment in order to benefit from gene therapy in the future.

Prior to interviewing the participants, it was anticipated that perhaps some might express sadness or resentment that Ivacaftor only has the potential to benefit a small percentage of CF individuals, which their child is excluded from as they do not have the gene targeted. Participants' reactions to Ivacaftor were in fact the opposite - they described feeling pleased for those who can benefit from it. One participant described the 'brilliance' that *"Ivacaftor is out there and making a difference to people's lives"*. Most participants also expressed excitement at the prospect that Ivacaftor could spark a new line of developments that may target their child's gene mutation, although they maintained a position of not wanting to know until it materialised. In contrast, one participant expressed a worry that new treatments targeting her own child's gene mutation would not happen because they were in a minority and questioned whether scientists would be motivated to develop a treatment that might only benefit a handful of people. In reality, her child had a common mutation and this raised questions as to whether the participant had a developed understanding about her child's gene mutation and its prevalence, and whether she was aware that Ivacaftor was designed to target a less common gene mutation. On the other hand, contrasting with feeling pleased for others and hopeful that more new treatments will develop, most participants reported feeling worried about the costs of new developments and were further concerned whether this would be funded by the NHS. Participants shared that they had heard about previous stories of NHS Wales refusing to fund Ivacaftor before a petition led them to endorse the treatment and fund it. Several participants felt that in the context of a changing NHS, the recession, and the fact that is CF is not a well-known condition compared to other health conditions, Ivacaftor and future treatments may not be viewed as being important enough to be funded. Consequently, several participants spoke about focusing on the available current treatments rather than learning about and pinning hopes on new treatments.

When participants were asked about Ivacaftor and possible future treatments, the majority expressed some scepticism as to whether the treatments will work and make a great deal of difference. Indeed one participant questioned how one tablet could change everything for the better. Several described feeling unable to place all their trust in the

new developments and indicated that they would continue with current treatments in addition to any new treatment because they wanted their child to be safe, and also because they would still be unable to see how the new treatment works. The latter echoes their earlier concerns described over whether current treatments work as they cannot see what the treatments are doing. Despite having a healthy cynicism, most participants expressed hopes for the new treatments that their child might one day benefit from in protecting them from infections, improving their quality of life, and reducing current treatment tasks. Another expressed that it was difficult to consider how much of a difference a new treatment could make to her child's life at present, before stating that perhaps her child will get 20 minutes currently spent on a single treatment task time back. However she went on to appreciate that being on a new treatment that targets the gene defect could change his future outlook. There may be a possibility that perhaps the potential of Ivacaftor and other possible new treatments are not fully realised and appreciated by parents of CF children because they rarely exhibit symptoms of CF in their childhood, compared to older children and adults with CF. Overall, there was a sense that participants had quiet hopes but these were not dwelled on in case new treatments either do not materialise or do not work.

The majority of participants expressed their willingness to let their child try a new treatment provided that it has been tested and approved for safety. In contrast, two participants described their need to have all the facts, proof that treatment works, and would enquire whether treatment comes with any risks. One participant also spoke about wanting to know what part of their child's CF a new treatment would positively affect and what this would mean for the rest i.e. would a new treatment only target the lungs or digestive systems or both. Seeing the current state of their child's health would also be taken into consideration, and if their child was viewed as being healthy enough they may feel that a new treatment such as Ivacaftor is unnecessary.

Despite possible advances in treatment and the changes that they could bring - for example, less treatment tasks and improved quality of life - most participants described that CF will still live on and so their worries about their child's health and the future will continue to exist. One participant said the condition itself will still exist and live in their child so that element of worrying will always be there. Another felt she would probably continue to take each day as it comes even if her child was on a treatment in case the future does not pan out well echoing previous research findings (e.g. Glasscoe et al, 2011). Most participants concluded that they know they will not benefit from Ivacaftor and that drugs in clinical trials are not yet accessible. With this in mind, participants identified that focusing on the present is particularly important, although they remain hopeful about future treatments materialising one day that will

help their child. There was an overall sense of participants' appreciation of how far CF has improved and will continue to improve.

Comparing the view of mother of a child on Ivacaftor to other participants' views:

For most participants there was a certain degree of homogeneity in that they all had children under the age of 5 and all had the same gene mutation DeltaF508. Unsurprisingly, these participants shared many views and worries however given their unique experiences some differences were also observed. To further understand parents' knowledge about the genetics of CF and views of current and new treatments particularly Ivacaftor, a parent (Jade) whose child was taking the new treatment was included. A focus was on her perspective about whether and how Ivacaftor has made a difference, although like other participants Jade started with how her child was diagnosed.

The arrival of CF was different for Jade compared to rest of the participants in that CF lived undetected for at least two years and she also blamed herself for her childrens' poor health. The impact on delayed diagnosis for CF has been poorly described. A study by Kharrazi et al (2005) sought out over 20 diagnostic stories from families of children with CF and found that families who sought out medical help but were misdiagnosed felt resentment and frustration towards health professionals. In contrast, relief was the primary emotion that Jade felt when she received the diagnosis as she realised it was the CF gene responsible for why her children were so poorly and she finally absolved herself of blame. These emotions were different to how the other participants experienced their child's diagnosis. Relief has not been described widely in the literature as a diagnosis of a chronic condition often evokes negative emotions yet there are conditions that might be undetected for a period of time causing distress to parents (e.g. Jerrett et al 1996). A study by Moola et al (2011) described parents of children with a chronic heart disease who were undiagnosed for some time and who shared their concerns and beliefs about their child's poor health but were often met with scepticism by health professionals. In their persistence to seek out an explanation, they finally received a diagnosis and described feeling relief.

Jade also shared a similar experience of information overload following the diagnostic period however she also highlighted that she experienced an additional difficulty of knowing what information to believe as there were many contradictions. In order to manage her ability to assimilate information well and to protect herself from feeling fearful and distressed Jade continues to engage in 'seeking vs avoiding information' as described by other participants. Interestingly, with regards to Ivacaftor Jade described limiting herself from knowing whether it had worked well with other

people in case it hadn't. Acceptance of CF and treatment adherence had been voiced as a significant worry for most participants when the time comes for their child to take ownership, however, Jade reported that her son was starting to become less dependent on her to administer his treatments and she described that this transition was generally going well. Although Jade was slowly beginning to give up some of her responsibility over administering treatments, she still reported that treatments can be time-consuming.

As Jade began to describe her views and experiences of Ivacaftor, her narrative no longer became comparable to that of the other participants' whose children were not taking the treatment. Initially Jade shared some of the healthy cynicisms that other participants reported about Ivacaftor, especially when she was told by the team that it was just a trial with no guarantees, but this quickly ebbed once she saw the visible transformation in her son's appearance and energy in one year. She reiterated how her son's weight increased and that he looked healthy, a transformation that was also noticed by extended family members and friends. The biggest change Jade reported was his personality in that he became energetic and was now able to keep up social activities with his friends. Furthermore, he was no longer hiding his breathlessness and constant exhaustion that prevented him playing with his friends. Prior to Ivacaftor, Jade described him as a helpless *"baby who just slept most of the time and struggled to eat"* and she marvelled at how he was now like a 'normal' young boy who had the energy to slam his bedroom door. Prior to interviewing Jade, I had assumed that Ivacaftor replaced all current treatments and so it was a surprise to hear that it was prescribed in addition to existing treatments. This was not viewed negatively - such as being more time consuming. Instead she reported that Ivacaftor made CF significantly more bearable for him and although his treatment tasks and consequently routine has increased, it had made him better and he was now living life. She appreciated on reflection that she had perhaps lived in a fantasy, forgetting that CF exists and maintaining that everything was ok on the current treatments. Ivacaftor has brought radical changes to her son's health and personality in ways that she never saw from the current treatments.

Despite the positive changes Jade saw in her son, she described that CF lives on and her future worries of his health deteriorating and treatment adherence are still there. She described the nature of CF meant that she had spent a long time watching out for signs that preceded an infection and acknowledged that this was something she would continue to do. Jade also highlighted that Ivacaftor did not change everything as the loneliness was still there for both her and her son in that they could not interact with others who live with CF. Interestingly, the appreciation of CF not being a physical disability highlighted by participants in this study and within the literature (e.g. Glasscoe, 2008; 2011) was not shared by Jade. Finally, Jade's understanding about the

genetics of CF were not explored due to the topic being focused on Ivacaftor in the interview, but she did complete the questionnaire that was used in this study and on that she ticked the box indicating that she was unsure of what her child's gene mutation was.

7.1.3 Summary of key themes and the relationship to the wider literature

In the current study, there were a number of key themes - 'an unknown territory', 'struggling with paradoxes', and 'taking control but feeling uncertain' - that echo themes highlighted in the wider literature on parents of children with other chronic conditions. The adjustment process that participants in the current study described mirrors that highlighted in the wider literature especially on framework of adjustment processes (e.g. Stephney et al, 2011).

Two particular themes in the current study could be viewed as being potentially specific to CF; 'information overload' and 'dealing with uncertain future' appears to be potentially specific to CF. Parents in the current study described that their information needs were not adequately met. Instead they described feeling overwhelmed by the amount of information they received about their child's CF the day after diagnosis was given. This particular finding has been highlighted in previous studies on parents of children with CF, for example in Hummelinck et al's study (2006) as described earlier. Studies on parents of children with other chronic conditions such as cancer have reported feeling the opposite (e.g. Ringner et al, 2011). Freeman et al (2004) were keen to understand significant problems that parents typically faced and resources that they particularly valued during key phrases of their child's illness; diagnosis, hospitalisation/surgery, hospital discharge, adjuvant treatment, recurrence (of illness), end of life, and remission. 137 parents of children with a brain tumour were asked to complete a survey by rating items on their experiences of interacting with healthcare providers, medical information/educational needs, utilising healthcare, and psychosocial concerns at the key phrases of their child's illness identified as either problematic or helpful. Parents were also asked to rate the level of stress of each item from 0-10. Of particular interest, the authors found that for at least half the sample, the most common and important problem encountered was having limited access to information specific to each key phrase especially at diagnosis, recurrence of illness, end of life, and remission. Parents reported that their information needs at those stressful times were unmet. It is important to acknowledge that whilst the participants in the current study have identified the CF team as being valuable, the team are actively encouraging parents to learn about CF. In doing so, parents may feel overwhelmed. Further research is warranted to investigate this further.

With regards to the theme dealing with an uncertain future, participants in the current study described feeling uncertain and fearful about the future. They moved between openly avoiding thinking about the future knowing that CF is a life limiting condition and trying to comfort themselves against the harsh reality of it. There was an element of self-protection. This particular theme has not been highlighted in studies on parents of children with other chronic conditions such as diabetes and this may be because those conditions, although long-term, are not normally life-threatening if individuals follow prescribed treatments well. A chronic condition where parents may feel those similar emotions is paediatric cancer. Although advances in treatments for cancer have increased survival rates (National Cancer Institute, 2009), the future remains a source of fear and uncertainty for parents. Fletcher et al's qualitative study (2010) with 9 mothers and 3 health care professionals, explored factors affecting mothers' abilities to cope with paediatric cancer. Of interest, a subtheme 'being fearful and protective' was developed which described mothers' fear of their child's relapse and death, and their fear of the unknown and what the future might bring despite their child being in remission. The mothers in Fletcher's study described feeling afraid of the future and in particular whether cancer was going to make a re-appearance. In the current study, participants described feeling afraid to look into the future and think about whether their child will live. In this way, cancer and CF are not so dissimilar in that they both instil in parents the fear of and feeling uncertain about their child's future.

7.1.4 Potential issues rising in CF

Before an overall summary of the research findings is provided, there may be several potential issues to consider. The way CF is seen is arguably entering a transition as new treatments targeting mutation class become more widely available and prognosis is associated with class of CF rather than having CF itself. The introduction of Ivacaftor may also increase public awareness about the different classes of gene mutations in CF. Indeed, in the current study participants were beginning to be aware of the importance of knowing their child's gene mutations and of future developments in treatments which may target them. The participants expressed happiness that Ivacaftor can help some individuals, but also expressed concerns over whether the new treatment would actually be effective. There was also worry over whether new treatments for their child's gene mutations would be developed and whether they would become routinely available on the NHS. CF healthcare professionals may find themselves needing to spend more time with parents and manage their worries and hopes effectively. It may therefore be helpful to explore other health conditions that have experienced a similar revolution in treatment options. Treatment for paediatric cancer has certainly become increasingly effective, and

one area in particular has undergone significant change which are targeting specific genetic mutations that underlie particular cancers, and developing treatments for these. As yet, the psychological impacts of these new developments have not been greatly explored. Instead, paediatric cancer studies with parents have largely tended to examine their care-giving experiences (e.g. Bally et al, 2014) and their decision making processes (e.g. Lipstein et al, 2012), with Lipstein et al's study finding that parents largely preferred to share the task of making decisions around their child's treatments with healthcare professionals. Lipstein et al (2012) also found that parents' decisions were influenced by changes in health status. Research is needed to explore parents' views of new developments in treatments for various health conditions to better understand potential issues that might surface for parents, strategies that they might use to cope, and to inform health teams in their work with them.

One other area of research on the psychological impact of new treatments for paediatric cancer with relevance of the current study is the importance of parental hope. Granek et al (2013) explored the trajectory of parental hope when a child has a difficult to treat cancer. The authors interviewed 35 parents of paediatric cancer with a poor prognosis at three times points; within 3 months of diagnosis, at 6 months and at 9 months. Using grounded theory the authors presented two main themes; future-orientated hope and present-orientated hope. Under the former theme, parents expressed a hope for a cure and treatment success. The authors found that hope for a cure was a strong motivator for parents to keep going even their child was particularly poorly. Interestingly, hope for their child's future appeared to change over time. At diagnosis, parents hoped that their child would live a long life. As their child began to respond to treatment, hope changed to focus on future goals such as hoping that they will reach milestones such as going to university and getting married. The theme present-orientated hope reflected parents' hope about the day-to-day quality of life for their child. They expressed hopes that their child would not be in any pain or suffering. They also expressed hopes that there would be no complications or side effects from any treatments that their child was current receiving. Granek et al's study (2013) has highlighted the importance of hope in the face of an adverse health condition. Although not explicitly explored in the current study, participants also expressed quiet hopes of a cure for their child, and much of the parental discourse around the new treatments could be seen in terms of generating hope for a better future.

7.2 Overall summary: Answering the research aims

All participants' narratives began at diagnosis. For most, the diagnostic period was characterised by shock and distress, apart from one participant who experienced a strong sense of relief following a delayed diagnosis. Participants moved through stages of coping highlighted in the literature leading to an eventual adjustment and making the best of the situation. Minimising the significance of CF was achieved through comparing CF to worse conditions and refusing to let it hold their child back from life.

A key element adjusting to the presence of CF involved participants reclaiming control over their child's diagnosis by managing their information needs. In the beginning, participants' keenness to learn about their child's condition quickly changed to feeling overwhelmed as there was an overload of information by healthcare providers followed by a realisation of what CF was and the range of life-threatening health events yet to come for their child. From here, participants began to engage in seeking vs avoiding information, which they seemed to have carried forward even after the diagnostic period. Participants avoided information through considering the likelihood of emotional harm caused by knowing something, and questioning the personal relevance. This shaped their knowledge about Ivacaftor with most reporting that they did not know too much about it as it did not benefit their child in the first place. Their knowledge about other treatments in clinical trials was again actively limited in order to protect themselves from false hopes. For most participants, they reported their preference to make do with current treatments that are available to their child and focus on living in the present. In contrast, for some, gene therapy was still on their minds who expressed a hope that it will materialise one day.

It seemed that participants had mixed views about current treatments, with particular treatments being favoured over others that were invasive and difficult to administer. Treatments were normalised however participants reported that they were demanding and time consuming at the best of times. Participants questioned and worried about the efficacy of current treatments and stated that they could never see how they were helping their child. Participants viewed the CF team as being highly knowledgeable, supportive and dedicated to their child's health and these were factors that influenced them to trust them and follow treatment recommendations. All participants stated that they openly avoided thinking about the future however they identified a number of hidden worries about their child's future particularly around their child's acceptance of CF and adherence to treatments. As part of managing their concerns and possibly anxiety about the future, participants tried to comfort themselves about their child's future.

Participants were sceptical of the notion that Ivacaftor or a new treatment could make a significant difference although expressed hopes of a new treatment that their child can benefit from to improve their quality of life and reduce treatment tasks. The participant whose child was taking Ivacaftor reported and hugely appreciated the radical changes seen in his health and behaviour. The participant also acknowledged that Ivacaftor did not change everything; for example, the worry about CF, future fears and associated aspects such as segregation from other CF individuals still exists. Nonetheless, the advances in treatments for CF have led to participants feeling pleased for those who can benefit from them and further excited at the possibility further treatments will develop that their child may directly benefit from.

7.3 Methodological considerations

This section will outline the methodological considerations of the current study. First, the limitations are described before moving on to the strengths.

7.3.1 Limitations

Recruitment process and sample

The recruitment process for the questionnaire involved CF clinicians informing parents during routine outpatient consultations about this research, and administering the questionnaires to those who consented to take part. The researcher did not play an active role in this part of the process and therefore was reliant on the CF clinicians. Consequently, there is a possibility that although the researcher ensured that the CF clinicians were well informed about the purpose of the research, the inclusion criteria, and kept in regular contact, they may have approached parents who they felt that they had a particularly good relationship with, were approachable, and were more willing to complete a questionnaire. Additionally, routine outpatient consultations varied for parents, with some taking place fortnightly and others being seen once every 6 weeks. This in turn might have influenced the type of sample generated for the interviews as CF clinicians were also responsible for informing participants about the interviews.

The sample generated for the interviews were predominantly mothers. This is not surprising given that research has focused on mothers' views and experiences of chronic conditions including CF compared to fathers who have been less documented in the literature. The current study had hoped to amend this, however as anticipated, mothers made up the sample, and only one father took part. There was also a certain degree of homogeneity in that all participants who took part in the interview were parents of children with the most common gene mutation. It would have been interesting to see whether there were any differences between parents of children with other gene mutations in their knowledge about the genetics of CF, and their views about current treatments and future treatments. However as the gene DeltaF508 is the most common gene, it is not surprising that the sample generated participants of children with that gene. The inclusion criteria was also amended to involve a participant of a child with a different gene mutation and who was eligible for Ivacaftor as I felt that this participant may have had a different perspective.

There were also some difficulties with contacting potential participants who expressed an interest to the CF clinicians about taking part in the interviews, in that most participants did not respond to my email as they were not able to access their email and/or their email was sent to the 'Junk' folder in their email account. Although this was

later addressed, this had meant that the process of meeting participants to be interviewed took much longer than anticipated and consequently, some valuable time to analyse participants' transcripts were lost.

Leeds CF unit

This research has focused on parents who attended the Leeds CF centre and whilst it has provided an understanding about how well the CF team inform and educate parents about the genetics of CF, the findings reported in this study are strictly limited to this particular CF centre. There may be clear differences in parents' knowledge and understanding of CF across different CF centres in the UK, particularly as the Leeds CF centre held educational sessions to help parents understand CF well. This type of service might not be held in all of the CF centres and therefore the findings should be taken with the view that it applies to the Leeds CF centre only.

Researchers' bias

My bias have been described in section 2.27 however there were several biases that might have influenced my analysis and interpretation of participants' transcripts. A particular bias was that I was a psychologist in clinical training (PICT) first and foremost who worked in a health setting. I am skilled in active listening and helping patients to make sense and work though their presenting difficulties. Although this has helped me to be sensitive to participants in the interviews and show empathy, it is possible that when participants became aware of my role as a PICT, they may have become influenced in how they responded to my questions and what aspects of their narratives that they chose to share with me. Indeed, one participant explicitly stated that she was nervous about the fact that I was a PICT. Furthermore, due to being a PICT, I was perhaps more cued to seeing particular emotions such as distress, anxiety, and denial in participants and may have been more quicker assume what they were feeling and why than perhaps someone who had less experience and/or had more objectivity.

Furthermore, at the time of doing the thematic analysis and writing up the results, I had worked with two adult inpatients with CF on my placement for a brief period of time. As I listened to their difficulties and distress that was strongly related to their CF, I had thought about the participants that I interviewed and their children. Indirectly and unintentionally, it is likely that this may have influenced how I then interpreted participants' stories and views shared. Nonetheless, I attempted to address this by using reflection and my personal experiences of having a disability to constantly question what participants expressed in the interviews both verbally and non-verbally to ensure I interpreted well.

Quality checks

Section 5 describes the quality checks that were carried out to ensure that my analysis and interpretations were accurate enough and valid. A particular quality check that is useful when using qualitative designs is seeking respondent validation (e.g. Smith, 2008). Although three participants responded to my summary of themes, it would have been ideal if more had responded. However, due to recruitment problems identified earlier in section 2.7.2, there was a delay in sending out the summary.

7.3.2 Strengths

Contribution to the literature

Ivacaftor has attracted a great deal of attention among professionals who work with CF patients and their families. There is a growing body of research on Ivacaftor, however the focus has been on reporting the outcomes of using Ivacaftor in clinical trials. There is no research that has explored the perspectives of CF patients who can directly benefit from Ivacaftor or their families. This is the first research to explore what parents currently know about the genetics of CF since 1986 (Nolan et al, 1986) and more importantly what they know about Ivacaftor, new treatments, and what it means to them.

Exploring more than one aspect of CF

A key strength of the current research is that more than one aspect of CF was explored. Indeed the diagnostic period, adjusting to CF, views of current and future treatments, future outlooks, and finally knowledge of CF were explored. Research has typically investigated particular aspects on their own for example, living with CF (e.g. Jessup et al (2010), and treatment adherence (e.g. O'Toole, 2012). In contrast the current research explored a range of key aspects of CF, which provides a context for parent's views of new treatments. The study generated rich understanding of some of the processes that parents experience and engage in from the start of the diagnosis that may shape how they then approach things later on. For example, parents' experiences of information overload during the diagnostic period have led them to seek and avoid particular types of information based on personal relevance and as a coping strategy. This in turn has influenced how they view new developments and could explain why they have a limited understanding about Ivacaftor and other possible drugs in clinical trials.

The current research adds to the previous literature in providing different perspectives on issues previously covered, for example it has highlighted parents' uncertainty and worry about the effectiveness of current treatments (rather than focus on how they find treatments time consuming, which has been heavily described already) and also how they approach thinking about their child's future which has been less explored in the CF literature. Altogether, our understanding of CF is becoming more evolved.

Using a mixed design

Research on CF has typically used either a quantitative or qualitative design (e.g. Hummelinck et al., 2006; Grob., 2008; Jessup et al, 2010; Glasscoe et al, 2008, 2011) and consequently there is little research on CF that has used both. Quantitative research has tended to provide a general and arguably surface level understanding of CF, whereas qualitative research has tended to focus on a small sample of participants providing a rich understanding but that is difficult to generalise. The current study has used a mixed design. The survey has generated an understanding of what participants know about their child's CF and of new developments. The use of a qualitative design in addition has provided an in-depth understanding of parents' views and experiences of current and future treatments. Together, the mixed design have enabled me to identify both at a wider and deeper level what parents know about the genetics of CF, their views, and experiences of different aspects of CF.

Sample

Although there were several limitations with the sample which have been outlined in the limitations section, a particular strength is that a mother of a child taking Ivacaftor was recruited. This has enabled me to explore any similarities and differences between her knowledge and views of CF and those of the other participants, and has also provided an understanding of how much impact Ivacaftor has for example, on Jade's future worries and acknowledgement that CF continues to exist despite the effects of the new drug. This has helped to understand how much of a difference Ivacaftor can make from the parents' perspective.

Researcher's reflexivity and quality checks

Braun and Clark (2006) argued for the importance of researchers reflecting on their position as a researcher and on any potential bias when using qualitative designs. I have engaged in the process of reflexivity throughout and particularly more during the interview and qualitative analysis stage. Supervision and peer supervision were utilised

to ensure that my analysis and interpretations were valid. This was particularly helpful as one potential bias proved to be more significant and challenging to contain at times than I had initially thought, the presence of my own disability. Prior to the interviews, I was aware of the influence that my disability might have on how I approached and interpreted participants' transcripts, however I did not anticipate the possibility of having a strong reaction on occasions. One particular occasion was when participants reported that they actively limited their understanding of new developments, and several expressed a hesitation about letting their child try them. I experienced a mixture of frustration and confusion as I thought about how I personally would feel elated and keen to know more if there was a treatment targeting deafness on the market or in development. Using reflection, I was aware that I was in danger of making inaccurate judgements about participants' views that could influence my analysis and interpretations. Preventative measures were taken by sharing my interpretations in supervision, peer supervision, and to another clinical psychologist who did not have any involvement in this research. Consequently, I came to appreciate that I was perhaps thinking from a patient's perspective and not from a parent's perspective, which shifted how I then thought about participants' views. The process of reflexivity has helped me to ensure that I accurately interpreted participants' narratives without judgement.

8.0 Clinical implications

The study has highlighted a number of areas where based on the experiences of the current participants, changes to current practice might be considered.

Increase healthcare providers' understanding on CF and awareness:

The current research was not intended to elicit participants' experiences of receiving their child's diagnosis, however at least two participants talked about having a delayed diagnosis by over a year and stated that healthcare providers in their local areas were not able to recognise signs of CF. They suggested that they did not have a good enough knowledge of CF. These participants also implied feeling dismissed when they queried about their child's health. Based on this finding, it is important for CF clinicians to increase local healthcare providers' awareness of CF and how to identify the signs. They might also need to broaden this work to include areas of UK where there are no CF centres making it difficult for parents in those areas to receive a diagnosis. Equally it is important for healthcare providers to take ownership for their own learning about CF.

Increase parents' understanding of the genetics of CF

To date, treatments for CF have been based on symptom reduction and as such parents' understanding of their child's gene mutation and classes until recently not been viewed as being particularly important. It is now becoming increasingly important to challenge this view, as new treatments in development are becoming mutation specific. CF clinicians are encouraged to ensure parents are aware of their child's gene mutations and classes and about possible new developments in treatment as this might emphasise the importance of adherence and give them hope (although it is appreciated that parents may then choose to not know more). In turn, their child might be more encouraged to pursue interests and education. CF clinicians across the UK are encouraged to spend more time reviewing parents' knowledge of their child's CF and where needed, to hold informative sessions to help parents develop their understanding of CF.

Tailoring information provision

The current study has highlighted that although most parents who attended the CF information clinic the day after the diagnosis viewed this as being important, it was largely exhausting and overwhelming. Whilst it is appreciated that newly diagnosed patients need to get started on treatments straight away and parents might not always know what they want to know, a review of when and what information about CF is given might be useful. Some parents may prefer to attend a clinic a few days after diagnosis.

Participants' reports of information overload should be taken on board and health care providers who are not doing so already are encouraged to review with parents what they would like to know and offer them the option to provide smaller amounts of information. This may help parents to feel less threatened by the amount of information and in turn less distressed and less avoidance of knowing information.

Helping parents to understand current treatments

Participants reported that although they understood what currents treatments were for, they described worry over whether they work and attributed this to being unable to see how it helps them. Whilst it is difficult to show parents exactly how current treatments work (aside from the yearly annual visits where their child's progress on treatment is reviewed), it might be helpful for healthcare providers to hold this in mind and consider whether this is an influencing factor for those who have low adherence rates to treatments.

Increase opportunities for parents to meet

Several participants reported that they felt isolated and wanted to meet other parents of children with CF. Participants stated that they did use a CF forum on the internet to talk with parents, however they preferred to meet face to face. Although educational classes for parents are held every once in a while at the Leeds paediatric CF centre, it might be beneficial for CF clinicians to develop a social support group for parents of CF patients that had an open membership giving flexibility to parents to attend when they can and on a monthly or bi-monthly basis. This may help parents to form meaningful relationships, feel less lonely and more supported dealing with their child's CF and the associated impacts.

Signposting parents for emotional support

The current study also highlighted that most participants felt distressed during the diagnostic period and struggled to adjust in the first few months. Whilst it is understandable and natural for parents to feel a range of negative emotions, it may be worth CF clinicians bearing in mind services that parents can access to help them to manage their emotions, such as short term counselling and/or psychological services. Although CF centres such as Leeds are well informed about the importance of direct or indirect clinical psychological care (e.g UK CF Trust, 2011), they are encouraged to continue to monitor and offer support - especially as for some several years post diagnosis are still distressed.

9.0 Summary

Overall, the current study highlight that there are areas of parents' knowledge of CF and their views of current treatments that needs to be addressed. The most important finding is parents' experience of information overload that essentially leads to them limiting their understanding about CF. This strategy could also explain why parents do not have a basic understanding of Ivacaftor.

10.0 Future research

The current research has explored the view of parents' of children aged 5 and under concerning knowledge of CF and their perspectives on current and future treatments. It would be interesting to compare their views to parents of adolescents and/or adults with CF where responsibility for making decisions around current and future treatments is likely to be shared. Furthermore, it would be interesting to see how older CF patients and their parents manage their information needs and understanding of Ivacaftor and/or new developments in clinical trials are different to the parents in this current study.

Another possible area of future research would be to compare parents' knowledge of CF and their views about different aspects of CF across CF centres. Indeed, Leeds actively promotes parents' understanding of CF through running computer based education classes. It would be interesting to see the level of difference between parents who attend the classes and parents who may not be able to at other CF centres.

The sample in the current study only included one mother of a child on Ivacaftor. Future research could expand on this by exploring more parents of children who are taking Ivacaftor in order to gain a greater understanding of how they view Ivacaftor and how much of an impact it has had on them.

Finally, although it was not a focus in this research I had noticed in the interviews that participants briefly touched upon the differences between them and their partners in how they both experienced their child's diagnosis, their views of current and future treatments, future outlooks, and how they manage their information needs. Differences in parents' coping styles and how they understand CF could be a worthwhile area to explore that might provide a valuable understanding and in turn inform healthcare providers on how to meet their needs.

11.0 CONCLUSION

1. The survey showed that all participants knew that CF was an inherited condition and that they were carriers of the CF gene. Most were able to identify their child's gene mutation with a small number stating that they were unsure. Furthermore, only several named both of their child's gene mutations. It is unclear whether most knew both of their child's gene mutations or whether they decided to report the common one of the two genes. DeltaF508 was the gene mutation that was commonly reported by participants both in the survey and in the interview. This gene is classed as being a severe gene mutation, however most believed that their child's gene mutation were either mild or moderate, with only a handful believing that it was severe. It is unclear what participants used to judge the severity of their child's CF.
2. The survey showed that most participants had a reasonably good understanding of some aspects of CF. For example, they were aware that it impacted on their child's lungs and pancreas. Most participants also understood that mutation class was important to know as it determined what treatments were available to their child. Despite this, most remained unsure of which mutation class their child's genes fell into and how severe they were. The survey has highlighted that there are areas in participants' knowledge that are perhaps not well developed.
3. The survey showed that few participants were aware of new developments in CF treatments and a possible explanation was suggested in the qualitative study in the participants' management of their information needs, for some at least a deliberate strategy as a result of their experience of information overload. This involved seeking and avoiding particular types of information, based on personal relevance and the likelihood of whether it will cause them distress.
4. The current research suggests that participants' management of their information needs influenced their understanding of Ivacaftor, with most having heard of the treatment but after recognising that it will not be of personal benefit to them they chose to refrain from knowing more. Consequently, they can be viewed as having an undeveloped understanding of Ivacaftor. Similarly most were unclear about other new developments in clinical trials and listed the same reasons mentioned above as an explanation. Interestingly, gene therapy was still on some participants' minds and there were indications that this was viewed as being a more effective or a realistic treatment than drugs similar to Ivacaftor.
5. The information overload that led participants to understand what CF meant also influenced how participants approached thinking about their child's future, with

most choosing to avoid thinking about the future and focusing on the now, although they shared a range of specific worries about the future suggesting that avoidance was not always successful.

6. All participants in the interviews were pleased about the development of Ivacaftor and happy for those who can benefit from it. Participants were hopeful at the prospect of new treatments being developed for their child, but were more worried about whether this will be routinely available on the NHS and whether their child will be able to access this. For this reason, they chose to focus on current treatments available. The worry about new treatments being funded was not reported by the mother of a child on Ivacaftor.
7. Despite feeling pleased and hopeful, most participants questioned the effectiveness of new treatments, something that they also did with their child's current treatments and described their worry over whether current and future treatments would work. Their worry stemmed from not being able to see how current treatments were helping their child. In the end, participants reported that they adhered to treatments and would continue to do so because they trusted the CF team. In contrast, the mother of the child on Ivacaftor could clearly see the benefits of Ivacaftor, more so than she could ever with current treatments.

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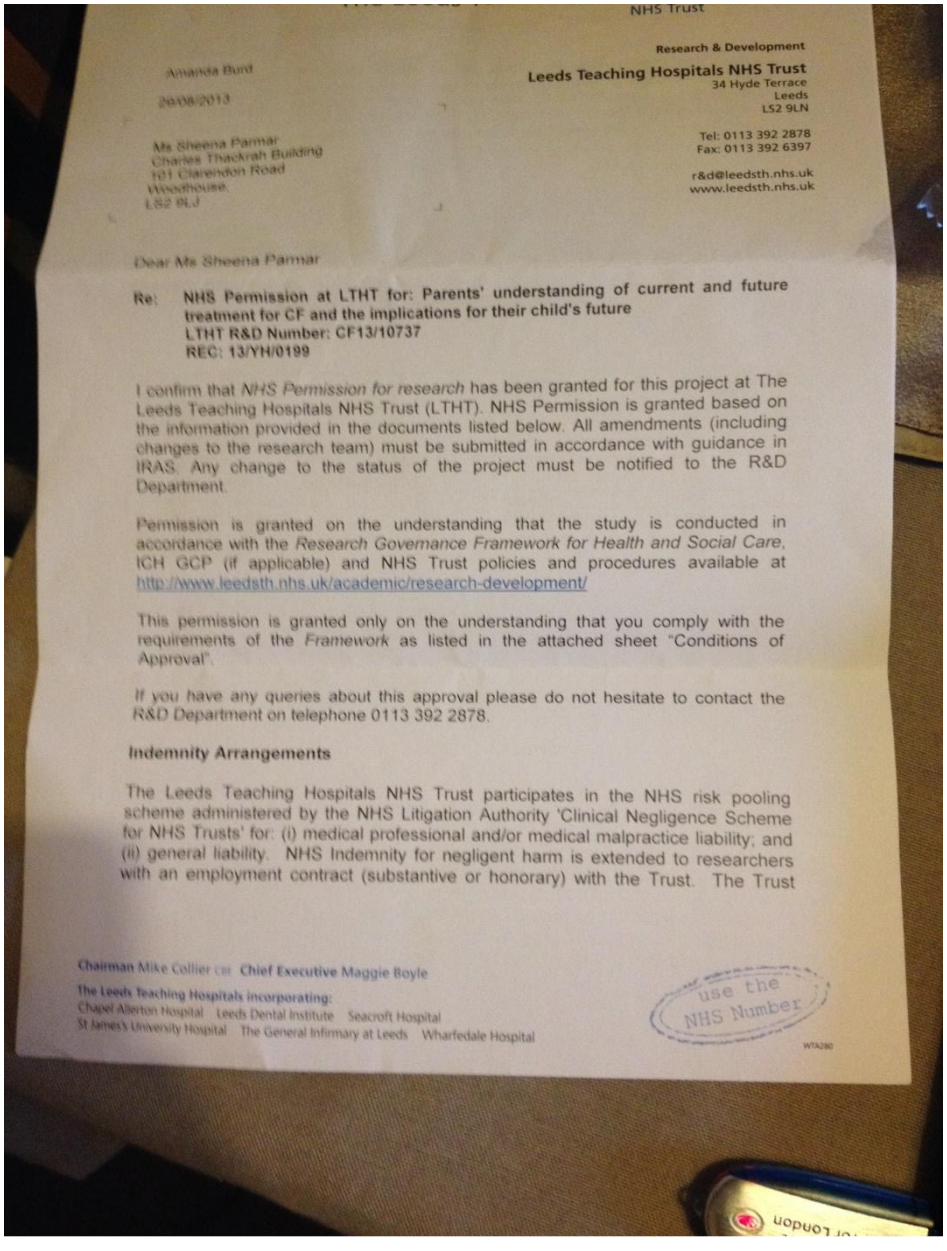
APPENDIX A: ETHICAL APPROVAL LETTER



NHS Trust

Research & Development

Amanda Burd

29/08/2013

Leeds Teaching Hospitals NHS Trust
34 Hyde Terrace
Leeds
LS2 9LN

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r&d@leedsth.nhs.uk
www.leedsth.nhs.uk

Ms Sheena Parmar
Charles Thackrah Building
101 Clarendon Road
Woodhouse,
LS2 9LJ

Dear Ms Sheena Parmar

Re: NHS Permission at LTHT for: Parents' understanding of current and future treatment for CF and the implications for their child's future
LTHT R&D Number: CF13/10737
REC: 13/YH/0199

I confirm that *NHS Permission for research* has been granted for this project at The Leeds Teaching Hospitals NHS Trust (LTHT). NHS Permission is granted based on the information provided in the documents listed below. All amendments (including changes to the research team) must be submitted in accordance with guidance in IRAS. Any change to the status of the project must be notified to the R&D Department.

Permission is granted on the understanding that the study is conducted in accordance with the *Research Governance Framework for Health and Social Care*, ICH GCP (if applicable) and NHS Trust policies and procedures available at http://www.leedsth.nhs.uk/academic/research-development/

This permission is granted only on the understanding that you comply with the requirements of the *Framework* as listed in the attached sheet "Conditions of Approval".

If you have any queries about this approval please do not hesitate to contact the R&D Department on telephone 0113 392 2878.

Indemnity Arrangements

The Leeds Teaching Hospitals NHS Trust participates in the NHS risk pooling scheme administered by the NHS Litigation Authority 'Clinical Negligence Scheme for NHS Trusts' for: (i) medical professional and/or medical malpractice liability; and (ii) general liability. NHS Indemnity for negligent harm is extended to researchers with an employment contract (substantive or honorary) with the Trust. The Trust

Chairman Mike Collier CBE Chief Executive Maggie Boyle

The Leeds Teaching Hospitals incorporating:
Chapel Allerton Hospital Leeds Dental Institute Seacroft Hospital
St James's University Hospital The General Infirmary at Leeds Wharfedale Hospital

use the NHS Number

WTA280

APPENDIX B: QUESTIONNAIRE

Understanding Cystic Fibrosis

ID:

This short questionnaire aims to explore how familiar parents are with the genetics of CF. It shouldn't take longer than 5 or 10 minutes to complete, and we would be very grateful if you could answer each question.

1. **There are many different types of genetic mutation that cause CF. Can you write the type of CF mutation that your child has? (e.g. DF508, W1282X, etc.)**

   Mutation type: _____ Not sure ☐

2. **CF mutations are usually grouped in to five different classes by doctors. Can you tick the CF mutation class you think your child has?**

Class 1	Class 2	Class 3	Class4	Class 5	Not sure
☐	☐	☐	☐	☐	☐

3. **What difference do you think the mutation class in CF has on the following?**

	No difference	A little difference	Some difference	A great difference	Not sure
General health	☐	☐	☐	☐	☐
The severity of symptoms	☐	☐	☐	☐	☐
The type of symptoms	☐	☐	☐	☐	☐
Treatments available now	☐	☐	☐	☐	☐
Treatments available in the future	☐	☐	☐	☐	☐
How effective the treatments are	☐	☐	☐	☐	☐

4. **Compared to CF generally, do you have a sense that your child's mutation class is:**

Mild	Moderate	Severe	Not sure
☐	☐	☐	☐

5. **Which of the following do you think your child's mutation affects?**

Lungs	Kidneys	Bladder	Pancreas	Intestines	All of them	Not sure
☐	☐	☐	☐	☐	☐	☐

6. Can you describe how cystic fibrosis is inherited?

Both parents are carriers of the gene that causes cystic fibrosis	From the mother only	From the father only	Not Sure
☐	☐	☐	☐

7. Are you aware of any new approach to treatment for CF?

Yes	No
☐	☐

Thank you for completing this questionnaire.

If you have any questions about anything in this questionnaire. please do not hesitate to ask me and myself or one of the doctors will contact you.

APPENDIX C: INTERVIEW SCHEDULE

Interview Schedule

Going to ask some questions about your child with CF and generally interested in your experience of CF, current treatments and the future.

Knowledge:

1. Can you tell me what you know about CF; the actual fault of it?
   What can you tell me about your child's cystic fibrosis?
   Are you aware about mutation classes?
   Knowing the type of mutation gene – is that important to you?
   Does it make a difference to you?
   (if no comment) Perhaps start with what life is like on a day to day basis?

2. How well would you say you understand your child's cystic fibrosis?
3. How important to you to know everything about cystic fibrosis
   (e.g. type and cause, if not why not and if so why?)

Impact and Symptoms:

1. Can you tell me a little bit about what life is like for you and your child having CF?
2. How do you view your child symptoms*?*

Current Treatment

1. Can you tell me a little abit about the current treatments that your child receives?
   Are there any negative and/or positive impacts of current treatments?
   What the process of going to hospital/CF unit like?
   Wait round at hospital?
   How does it make you feel seeing your child receive treatments?
   Child's reaction to treatments?

2. How well would you describe your knowledge about the current treatments that your child receives?
   (Can you tell me more about this?) e.g. do they know about each treatment and what it is for?

3. How would you describe your views about them?
   (E.g. are they effective?)
   Would you say that they make a difference to your child's health?
   Am I right in thinking then your child adheres to the treatments/not?
   Can you tell me more?

4. Are there any impacts that these treatments have on your child and/or you and family? Can be positive and/or negative?

Future based on current treatments

1. Based on the current treatments, can you tell about the future and what you see it being like?
   Is your child's future is something that you tend to think about?
   (E.g. Can you tell me some more about this?
   What is it that you think/don't think about?
   Can I ask about whether you have any hopes & dreams for your child?

Have they expressed any to you?

2. Is the future something you ever discuss with your child?
   Can you tell me more about this?

New Treatment for CF

1. Are you aware of any new treatments for CF?
   (Can you tell me more about this?)
   (What is it that you are aware of?)

2. Upon hearing about the new treatment, how would you describe your reaction/how it makes you feel?

3. How would it make you feel to allow your child to try a new treatment? E.g. Ivacaftor

4. If your child was on it, would there be any positive impacts on yours and their daily living with CF?
   Would it add any burdens?

Future based on new treatment

1. Based on the hearing about the new and upcoming treatment that may be available, are there any changes in how you view your child's life and future?

2. Are there any changes in how you view the current treatments your child receives?

3. Are you aware of whether your child is able to receive this treatment?
   (Are you aware of any eligibility criteria?)

4. What would it mean to your child, you and your family if your child was able to receive this treatment?

Is there anything else that you would like to tell me that we haven't covered already?

(Off record)

Thank you for participating in the study. It would be really helpful to know how you have found this interview.

APPENDIX D: INFORMATION SHEET FOR QUESTIONNAIRE

Leeds Institute of Health Sciences
FACULTY OF MEDICINE AND HEALTH

UNIVERSITY OF LEEDS

Information sheet for questionnaire: **Version 2**
Date: 10.01.14

A survey of parents' understanding of current and future treatment for CF, and the implications for their child's future

As part of my Doctorate in Clinical Psychology, I am conducting a research project that focuses on Cystic Fibrosis. I would like to invite you to participate, however before you decide whether you wish to take part, please read the information below that explains what this research is about, why it is being conducted and what your participation would involve.

If you have any questions please do not hesitate to contact me. The contact details are at the bottom of page 3.

Thank you for taking the time to read this.

What is the purpose of this research?
At present, current treatments for CF are mainly focussed on reducing the symptoms. However, some of the new treatments in development work by addressing some of the underlying causes. One of these, Ivacaftor has recently been approved for use in the UK by the NHS in January 2013.

What we know about this new treatment is that it works by addressing the problems caused by one of type of mutations that cause CF. There are many different types of mutations in CF; because this particular drug only works for one of them, not everyone will benefit at the present time. With this in mind, it has become important for people with CF and their parents to gain a better understanding of the genetics and mutations that underlie CF.

To date, there is very little literature that tells us how much parents know about mutations and the genetics of CF, and the possible implications of the new treatments being developed. This research aims to understand how much parents of children with CF know about these topics, and to ask for their thoughts about the implications for them and their child.

Who can participate?
I would like to invite mothers of children with CF, who are aged 10 or younger and who have had a diagnosis of CF for at least six months to take part in this research.

If you would like to know more about this, please contact myself using the contact details at the bottom of page 3.

What will I need to do if I choose to participate?
If you would like to take part, you will be asked to complete a questionnaire asking for your thoughts about the genetics of CF. This should not take longer than 10 minutes to complete and you can complete this here on the CF unit.

Do I have to take part?
Participation is not compulsory and therefore it is your decision whether you wish to take part or not. If you do wish take part, you can withdraw at any stage of the research. All you would need to do is let the researcher (myself) know. If you decide to participate and later wish to withdraw, any data collected from you up to that time will be retained.

Will my participation and information be confidential?
Any information that you give about yourself will be confidential and kept in a secure place. No identifiable information about you will be included in write up.

What will happen to the results of the study?
Your interview data will be analysed by myself and written up as part of my doctoral thesis to be kept in the library at the University of Leeds. The results of this research may also be disseminated to CF professionals; however no information that reveals you or your child's identity will be given.

What if there is a problem and I need to make a complaint?
If at any stage of the interview you become concerned, you can speak to the researcher who will do their best to address your concerns.

However, if you remain unsatisfied and wish to make a formal complaint, you can go through the NHS Complaints Procedure. Details can be obtained from the Leeds Cystic Fibrosis Unit. Alternatively you can contact Claire Skinner, Faculty Head of Research Support, Faculty of Medicine and Health Research Office, Room 10, Level 10, Worsley Building, University of Leeds. Clarendon Road, Leeds, LS2 9NL.

If you experience any distress during or after the questionnaire, and you wish to talk to someone, you can talk to a clinical psychologist who is a part of the Leeds CF team.

Contact Details:
Dr. Gary Latchford, Consultant Clinical Psychologist: (0113) 206 5897
Dr. Alistair Duff, Consultant Clinical Psychologist: (0113) 206 5897
Dr. Tim Lee, Consultant Paediatrician: (0113 3927125

What do I need to do next?
If you would like to ask any further questions about the research or if you would like to take part then please feel free to contact myself using the following contact details;

Sheena Parmar
Leeds Institute of Health Sciences
Charles Thackrah Building
University of Leeds
101 Clarendon Road,
Leeds
LS2 9LJ

Tel: 07717871521 (**please text or email** as I am hard of hearing and I will respond back to you)
Email; umspa@leeds.ac.uk

Thank you for taking the time to read this information.
If you would like to make a complaint about this research, please contact the Department of Clinical Psychology Administration Team at Leeds University who will make arrangements for you to do this. Please find contact details below;

Leeds Institute of Health Sciences
Charles Thackrah Building
University of Leeds
101 Clarendon Road,
Leeds
LS2 9LJ
0113 343 0829
Email Lydia.stead@leeds.ac.uk or Debby.williams@leeds.ac.uk

APPENDIX E: INFORMATION SHEET FOR INTERVIEWS

Leeds Institute of Health Sciences
FACULTY OF MEDICINE AND HEALTH

UNIVERSITY OF LEEDS

Participant Information Sheet version 2
Date:
November 2014

Parents' understanding of current and future treatment for CF and the implications for their child's future: an interview study

As part of my Doctorate in Clinical Psychology, I am conducting a research project that focuses on Cystic Fibrosis. I would like to invite you to participate, however before you decide whether you wish to take part, please read the information below that explains what this research is about, why it is being conducted and what your participation would involve.

If you have any questions please do not hesitate to contact me. The contact details are at the bottom of page 3.

Thank you for taking the time to read this.

What is the purpose of this research?
At present, current treatments for CF are mainly focussed on reducing the symptoms. However, some of the new treatments in development work by addressing some of the underlying causes. One of these, Ivacaftor has recently been approved for use in the UK by the NHS in January 2013.

What we know about this new treatment is that it works by addressing the problems caused by one of the type of mutations that cause CF. There are many different types of mutations in CF; because this particular drug only works for one of them, not everyone will benefit at the present time. With this in mind, it has become important for people with CF and their parents to gain a better understanding of the genetics and mutations that underlie CF.

To date, there is very little literature that tells us how much parents know about mutations and the genetics of CF, and the possible implications of the new treatments being developed. This research aims to understand how much parents of children with CF know about these topics, and to ask for their thoughts about the implications for them and their child. The research will explore parent's knowledge, views and beliefs about current and future treatments, and what this might mean for their child.

Who can participate?
I would like to invite mothers of children with CF, who are aged 5 or younger and who have been diagnosed for at least six months to take part in this research. As this research is primarily concerned with CF, I would like to ask mothers who wish to take part that their child has no other conditions. This is to help prevent mothers' views and beliefs about CF to become blurred or influenced by other treatments that their child may also be currently receiving for other conditions. Only a limited number of mothers can be considered for an interview due to the size of the study.

Furthermore, it is important to acknowledge that this research is exploring an area which understandably may cause some distress. While the researcher will do their best to ensure that mothers feel safe and comfortable during the interviews, it is possible that mothers may still experience distress. With this in mind, I ask mothers who have not had at least 6 months adjustment period since the diagnosis of their child's CF, to refrain from participating. If you would like to know more about this, please contact myself using the contact details at the bottom of the page.

What will I need to do if I choose to participate?
If you would like to take part, you will need to write down your contact details (please provide your email address and/or mobile number) and give this to your consultant, who will then pass it on to me. I will contact you and ask to meet you at a time that is convenient for you. You will be asked to read and sign a consent form.

You will be asked to take part in an interview with me, to talk about this topic. This discussion is a chance for you to talk about your thoughts and feelings about CF in general, and also specifically about your child's CF. The discussion will be audio taped and transcribed. It is planned to last one hour and will take place at a time that is suitable for you. You can choose whether you would like to have the interview at the CF unit, or at your home.

Do I have to take part?
Participation is not compulsory and therefore it is your decision whether you wish to take part or not. It will not affect the standard of care that you receive. If you decide that you would like to take part, you can also ask for parts of the interview data to be omitted from the analysis and in the final write up.

If you do wish take part, you can withdraw at any stage of the research. All you would need to do is let the researcher (myself) know. Any data collected from you up to that time will be retained.

Will my participation and information be confidential?
If you choose to take part, the interview will be audio taped and transcribed; however they will be kept in a secure place until this research is completed. Any identifiable information about you and your child will be removed and not included in the write up.

What will happen to the results of the study?
Your interview data will be analysed by me and written up as part of my doctoral thesis to be kept in the library at the University of Leeds. The results of this research will also be disseminated at CF conferences; however no information that reveals you or your child's identity will be given.

What if there is a problem and I need to make a complaint?
If at any stage of the interview you become concerned, you can speak to the researcher who will do their best to address your concerns.

However, if you remain unsatisfied and wish to make a formal complaint, you can go through the NHS Complaints Procedure. Details can be obtained from the Leeds Cystic Fibrosis Unit. Alternatively you can contact Claire Skinner, Faculty Head of Research Support, Faculty of Medicine and Health Research Office, Room 10, Level 10, Worsley Building, University of Leeds. Clarendon Road, Leeds, LS2 9NL.

If you experience any distress during or after the interview, and you wish to talk to someone, you can talk to a clinical psychologist who is a part of the Leeds CF team.

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Dr. Alistair Duff, Consultant Clinical Psychologist: (0113) 206 5897
Dr. Tim Lee, Consultant Paediatrician: (0113 3927125

What do I need to do next?
If you would like to ask any further questions about the research or if you would like to take part then please feel free to contact myself using the following contact details;

Sheena Parmar
Leeds Institute of Health Sciences
Charles Thackrah Building
University of Leeds
101 Clarendon Road,
Leeds
LS2 9LJ

Tel: 07717 871 521 (**please text or email** as I am hard of hearing and I will respond back to you)
Email; umspa@leeds.ac.uk

Thank you for taking the time to read this information.

If you would like to make a complaint about this research, please contact the Department of Clinical Psychology Administration Team at Leeds University who will make arrangements for you to do this. Please find contact details below;

Leeds Institute of Health Sciences
Charles Thackrah Building
University of Leeds
101 Clarendon Road,
Leeds
LS2 9LJ
0113 343 0829
Email Lydia.stead@leeds.ac.uk or Debby.williams@leeds.ac.uk

APPENDIX F: CONSENT FORM FOR THE INTERVIEW

The Leeds Teaching Hospitals NHS
NHS Trust

Consent Form

Participant Identification Number for this study: _____ **Date:**

Research Study: Parents' understanding of current and future treatment for CF and the implications
for their child's future

Sub questions:

1. Do they understand the genetics of CF?
2. Do they know about the new generation of drugs
3. Does this influence their process of adherence

Name of Researcher: Sheena Parmar, Dr Latchford
Dr Tim Lee and Dr Alistair Duff

Please initial box

1. I confirm that I have read and understand the information sheet dated _____ for the above study. I have had the opportunity to consider the information, ask questions and have had these answered satisfactorily. ☐
2. I understand that my participation is voluntary and that I am free to withdraw at any time without giving any reason, without my medical care or legal rights being affected. ☐
3. I understand that relevant sections of my data collected during the study, may be looked at by individuals from the research team at the University of Leeds or from regulatory authorities, where it is relevant to my taking part in this research. I give permission for these individuals to have access to my records. ☐
4. I agree to quotes being used in the writing of a report on the understanding that my anonymity is maintained. ☐
5. I agree to the interview being audio-taped. ☐
6. I agree to take part in the above study.

Name of Participant Date Signature

APPENDIX G: REVISED INTERVIEW SCHEDULE

Interview Schedule

I'm going to ask some questions about your child with CF and generally interested in your experience of CF, current treatments and the future.

Knowledge:

1. Can you tell me what you know about CF; the actual fault of it?
2. What can you tell me about your child's cystic fibrosis?
   - Are you aware about mutation classes?
   - Knowing the type of mutation gene – is that important to you?
   - Does it make a difference to you?
   - (if no comment) Perhaps start with what life is like on a day to day basis?
3. How well would you say you understand your child's cystic fibrosis?
4. How important to you to know everything about cystic fibrosis
   (e.g. type and cause, if not why not and if so why?)

Impact and Symptoms:

1. Can you tell me a little bit about what life is like for you and your child having CF?
2. How do you view your child symptoms*?*

Current Treatment

3. Can you tell me a little abit about the current treatments that your child receives?
   - Are there any negative and/or positive impacts of current treatments?
   - What the process of going to hospital/CF unit like?
   - Wait round at hospital?
   - How does it make you feel seeing your child receive treatments?
   - Child's reaction to treatments?
4. How well would you describe your knowledge about the current treatments that your child receives?
   - (Can you tell me more about this?) e.g. do they know about each treatment and what it is for?
5. How would you describe your views about them?
   - (E.g. are they effective?)
   - Would you say that they make a difference to your child's health?
   - Am I right in thinking then your child adheres to the treatments/not? Can you tell me more?
6. Are there any impacts that these treatments have on your child and/or you and family? Can be positive and/or negative?

Future based on current treatments

1. Based on the current treatments, can you tell about the future and what you see it being like?
   - Is your child's future is something that you tend to think about?
   - (E.g. Can you tell me some more about this?
   - What is it that you think/don't think about?

Can I ask about whether you have any hopes & dreams for your child?
Have they expressed any to you?

2. Is the future something you ever discuss with your child?
   Can you tell me more about this?

New Treatment for CF

1. How did you hear about Ivacaftor?
   How did your child get started on it?
   What was the process like for you to let your child try Ivacaftor?
   (Can you tell me more about this?)

2. How would you describe your reaction to Ivacaftor?
   How does it makes you feel knowing your child is on this?

3. Can you tell me whether there have been any changes since your child has been taking Ivacaftor?

4. Are there any positive impacts on yours and theirs on daily living with CF?
   Does it add/lessens any burdens?

Future based on new treatment

1. Based on Ivacaftor, are there any changes in how you view your child's life and future?

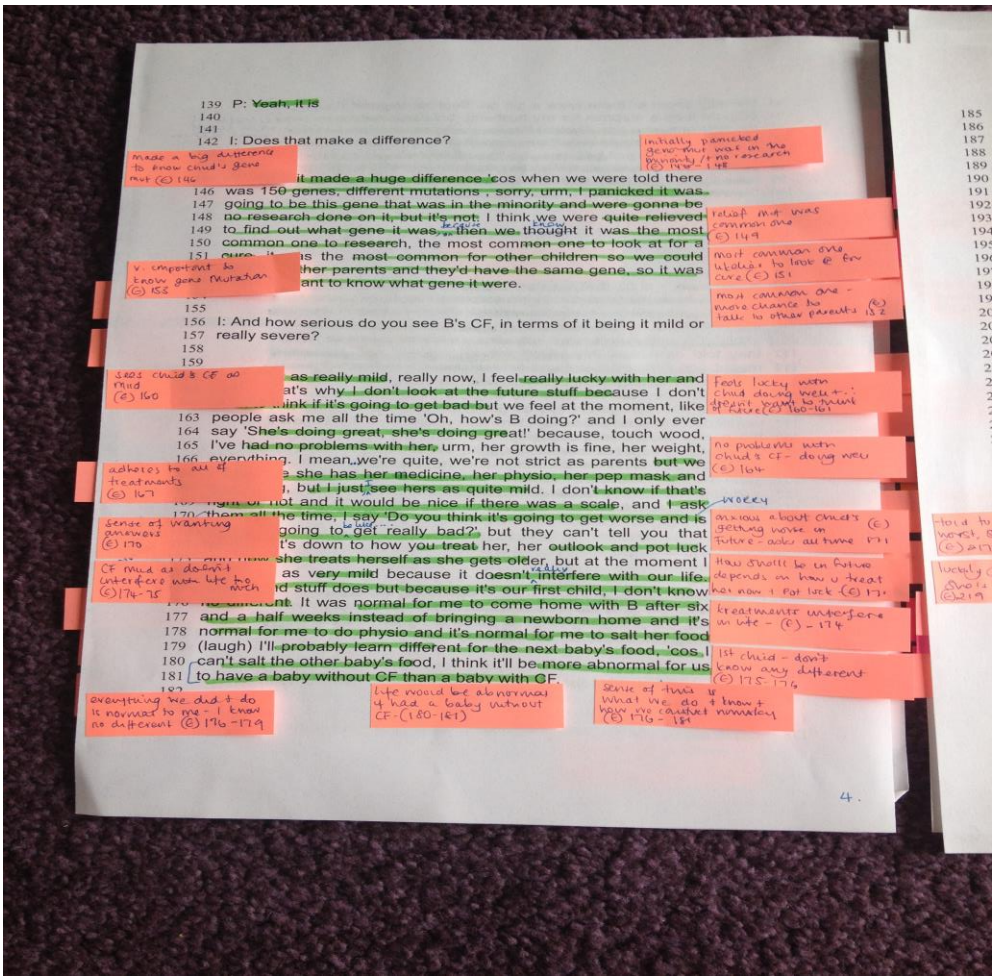
2. Are there any changes in how you view the current treatments your child receives?

Is there anything else that you would like to tell me that we haven't covered already?

(Off record)

Thank you for participating in the study. It would be really helpful to know how you have found this interview.

APPENDIX H: EXAMPLE OF INITIAL CODING (LOUISA'S TRANSCRIPT)



APPENDIX I: EXAMPLE OF FORMING CATEGORIES FROM GROUP ANALYSIS