ASSESSING THE IMPACT OF ADMINISTERING THE 'CHALLENGES OF LIVING WITH CYSTIC FIBROSIS' QUESTIONNAIRE ON THE SELF-EFFICACY OF CAREGIVERS OF CHILDREN WITH CYSTIC FIBROSIS

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By

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ABSTRACT

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Tulsi P Patel

INTRODUCTION

Cystic Fibrosis is a multi-organ, genetic disorder with the most pronounced effects observed in the lungs, pancreas and the gastrointestinal system. From a young age, patients suffer from chronic pulmonary infections, poor digestion and malabsorption of nutrients from the gut. As a result, patients have to perform numerous therapeutic tasks regularly to remain healthy. These treatments come at the expense of a considerable treatment burden and burden of care to the patient and family involved.

METHODS

The 'Challenges of Living with Cystic Fibrosis Questionnaire' (CLCF-Q) has been developed to measure the burden felt by caregivers of children with CF. The aim of this study was to investigate the effect of administering the CLCF-Q to caregivers during their child's annual review on the carer's self-efficacy. Self-efficacy was measured by the 'Cystic Fibrosis Self-Efficacy Questionnaire' (CFSE-Q). Participants were randomised into the intervention or control groups. Those in the intervention group completed the CLCF-Q during the annual review process and received feedback at a later date. All participants completed the CFSE-Q at baseline and endpoint to measure their self-efficacy.

RESULTS

Thirty seven participants (17 Intervention, 20 Control) completed the whole study. The CLCF-Q identified some of the burdens faced by caregivers of children with CF. Total self-efficacy scores ranged from 36-53 in the intervention group and 37-51 in the control group at baseline. End-point scores ranged from 38-54 for the intervention group and 31-53 for the control group. Statistical analysis revealed that the self-efficacy of the intervention group remained stable over time and that of the control group decreased significantly by the end of the study. Self-efficacy scores decreased for six items and increased for six items in the intervention group. Only two items had higher scores in the control group and 10 items had lower scores at the end of the study.

DISCUSSION

The CLCF-Q may be used as a tool to measure burden in caregivers of children with CF. It helps health care providers recognise the burdens faced by these families. The feedback process aims to involve the caregiver and encourage them in areas of treatment which they find challenging. The combination of these two processes as an intervention may be used to maintain a high level of self-efficacy in caregivers of children with CF. The CFSE-Q was able to detect changes in self-efficacy and may be used in future self-efficacy trials in caregivers of patients with CF.

TABLE OF CONTENTS

LIST OF IMAGES

LIST OF TABLES

LIST OF ABBREVIATIONS

AAD- Adaptive aerosol delivery

ACBT- Active Cycle of Breathing Techniques

ACT- Airway clearance techniques

AD- Autogenic Drainage

BOS- Bronchiolitis Obliterans Syndrome

BPT- Behavioral Parent Training

CF- Cystic fibrosis

CFQ-R- Cystic Fibrosis Questionnaire- revised version

CFSE-Q- Cystic Fibrosis Self-Efficacy Questionnaire

CFTR- Cystic fibrosis transmembrane regulator

CHQ- Child Health Questionnaire

CLCF-Q- Challenges of Living with Cystic Fibrosis Questionnaire

CT- Computed tomography

FEF_{25-75}- Forced Expiratory Flow between 25% and 75% of the vital capacity

FEF max- Maximal Forced Expiratory Volume

FET- Forced expiration Technique

FEV_1- forced expiratory volume in one second

FVC- Forced Vital Capacity

GSE-S- General Self-Efficacy Scale

HRQOL- Health Related Quality of Life

IQR – Inter Quartile Range

IRT- Immunoreactive trypsinogen

NIH- National Institutes of Health

PAP Analysis- Pancreatic Associated Protein Analysis

PEP- Positive expiratory pressure

PERT- Pancreatic Enzyme Replacement Therapy

SD- Standard deviation

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CHAPTER ONE

INTRODUCTION

INTRODUCTION

1.0. INTRODUCTION

Cystic fibrosis (CF) is the most common life threatening genetic disease to affect the Caucasian population.[1,2] It is a multi-system disease primarily affecting the lungs, in addition to other organs such as the pancreas, gut and male reproductive organs.[3] Most patients with CF suffer from chronic, infective lung disease from a young age. Many also have nutritional deficiencies resulting from pancreatic insufficiency and malabsorption from the gut.[1,4] Therefore these patients are usually prescribed multiple daily medications and treatments such as antibiotics, nebulisers, nutritional supplements, pancreatic enzyme supplements and physiotherapy. When the disease was first discovered in the late 1930's, life expectancy was only a few months. As a result of these medical advances, patients with CF can now expect to live much longer.[1,4,5] Improvement in life expectancy is at the expense of a considerable treatment burden for patients and their families.[6-8]

A tool 'The Challenges of Living with Cystic Fibrosis Questionnaire' (CLCF-Q) has been developed to measure the burden faced by caregivers of children with CF.[9-11] This is a non-blinded, randomised control pilot study being carried out at Alder Hey Children's hospital, Liverpool. The CLCF-Q is being used as an intervention tool together with an interactive feedback session. The outcome measure is self-efficacy, assessed by the 'Cystic Fibrosis Self-Efficacy Questionnaire' (CFSE-Q). Self-efficacy is an important factor in health behavior, yet it has received little attention from medical professionals.[8,12,13] In this pilot study, we have explored the impact of completing the CLCF-Q during the CF annual assessment and the feedback session on the self-efficacy of caregivers of children with CF.

It is important to fully understand the complex nature of CF and the available treatment options. The remainder of this chapter will give a comprehensive insight into CF, including genetics, diagnosis, clinical features and management. This will highlight the nature and complexity of the disease and treatments. It will also describe the various burdens faced by patients and families with CF and how we can assess and measure them. The concept of self-efficacy and its' importance in CF will also be discussed.

CYSTIC FIBROSIS

2.0. CYSTIC FIBROSIS

2.1. EPIDEMIOLOGY

Cystic fibrosis is the most common fatal, hereditary disease affecting the Caucasian population. It is recessively inherited and has an incidence of 1 in 2,500 live births with a carrier rate of 1 in 25.[1] The risk of a carrier couple having a child with CF is 25%. The same parents have a 50% risk of having a child who is a carrier of CF.

In 2008, the UK CF Registry reported a total of 8513 patients registered as having CF in the UK. Out of these, 6082 patients had 'complete' data. 65% of patients were diagnosed with CF before their first birthday. Approximately 47% of patients with CF were female. According to the data, there are more males with CF surviving after 16 years than females in the UK (Figure 1, 2 & 3).[2]

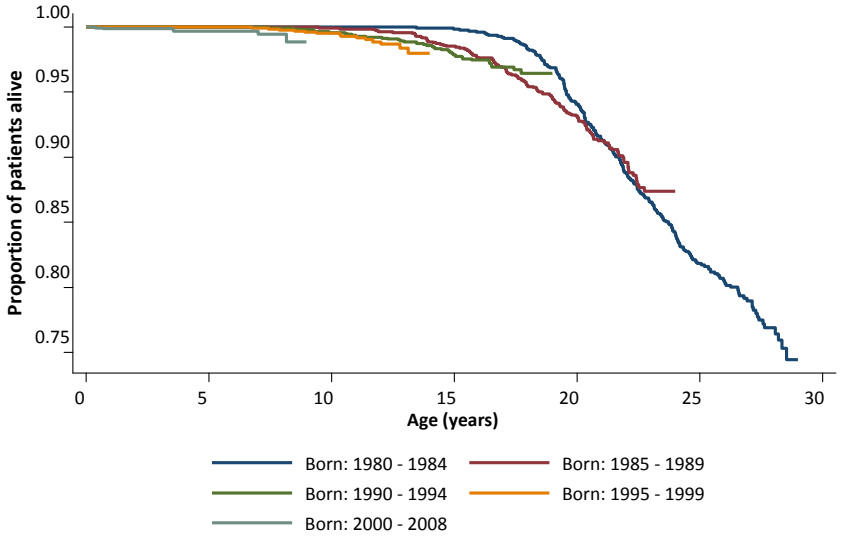


Figure 1. Actuarial survival by birth cohort

Graph representing birth cohort survival data for patients with CF in the UK[2]

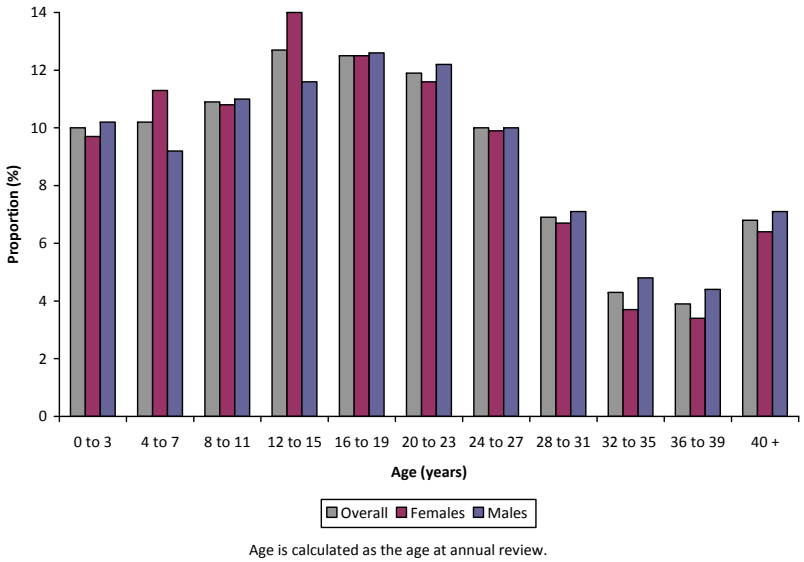


Figure 2. Age distribution

Graph representing the proportion of patients with CF in various age-groups[2]

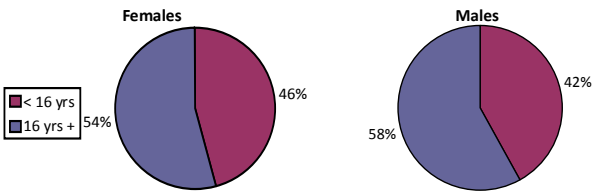


Figure 3. Age distribution by sex

Chart comparing age-distribution in male and female patients with CF [2]

2.2. GENETICS

The CF gene was identified in 1989. It is located on the long arm of chromosome seven.[14-16] The gene translates a protein, the Cystic Fibrosis Transmembrane conductance Regulator (CFTR), which is a chloride channel regulator, with a key role in the movement of water and salts across cell membranes of exocrine epithelial cells.[17] To date, over 1500 mutations of this gene have been found.[4] The most common mutation is ΔF508, which is present in approximately three quarters of cases of CF in Northern European and American societies.[17,18] The various genetic mutations are broadly classified into five different classes (I-V) based on their functional alterations.[4,17] The type of genetic mutation is partly responsible for the severity and prognosis of the disease (Table 1).[19,20]

Class	Molecular consequence	Example	Phenotypic consequence
I	nonsense or frameshift mutations that result in no significant protein product	G542X	typical CF phenotype
II	protein product does not negotiate intracellular trafficking pathways	phe508del R1066C A561E	typical CF phenotype
III	protein product transported to the cell membrane but no significant ion transport function	G551D	typical CF phenotype
IV	protein product transported to cell membrane and functions at a low level	R117H R334W	associated with pancreatic sufficiency
V	reduced mRNA expression, protein product normal	5T variant of intron 8 poly T region. 3272-26 A>G	associated with a mild phenotype, occasionally normal sweat electrolytes and single organ pathology (CBAVD)

Table 1. Classes of CFTR mutations with molecular and phenotypic consequences[20] (There are exceptions to this classification. i.e. Some Class I mutations may result in mild phenotypes and some Class V mutations may result in severe phenotypes)[19]

2.3. NEWBORN SCREENING AND DIAGNOSIS

2.3.a. Screening

Due to the common occurrence of CF, newborn screening programmes have been implemented in countries worldwide. It is a complex process and there are various methods being employed by different places.[21-24] Newborn screening for CF was first proposed in the 1970s, when it was found that infants with CF have raised serum immunoreactive trypsinogen (IRT) levels.[24,25] All programmes start off by measuring the IRT levels in the blood taken from a heel prick in the first week of life. An IRT measurement of more than 70ng/ml raises the suspicion of a diagnosis of CF.[24] This is

a very sensitive test and most infants with CF will be detected.[24,26] This is not a very specific test and therefore a second tier of testing is required.[27]

Screening protocols differ after the first IRT measurement.[21,22,24,26] Some choose to wait and repeat IRT measurements when the infant is 27-28 days old. Infants whose IRT levels still remain elevated are then referred for further testing and diagnosis.[21,22,24,26] Others proceed to genetic analysis to identify common genetic mutations for CF. After the discovery of the genetic mutations responsible for causing CF in 1989, this method has become more popular. This has the disadvantage of identifying carriers of CF, which may be useful for the patient and family in the future, but it may also cause undue stress.[21,22,24,26,28] Genetic testing in a multicultural society may also lead to ethnic discrimination, as some CFTR mutations are more common within some ethnicities than others. Testing for a few mutations would be disadvantageous to ethnic minorities within the area.[22] Another alternative is to use Pancreatic Associated Protein (PAP) analysis. This is a relatively new measure and is still being tested.[22]

Not all patients are identified through newborn screening. Some present with clinical features suggestive of CF and have to undergo investigations to confirm the diagnosis.

2.3.b. Diagnosis

The final step is sweat testing, which is the definitive diagnostic test for CF. The sweat of patients with CF has a high concentration of sodium and chloride due to defects in reabsorption of these electrolytes from the sweat ducts. Elevated concentrations of sodium in the sweat can be seen in other diseases besides CF. Therefore sweat testing relies on measuring the concentration of chloride in the sweat.[28-30] The procedure for carrying out a sweat test is very precise and should be carried out by trained personnel from a laboratory that has adequate experience of this procedure.[22,24,30] Sweating is stimulated by pilocarpine iontophoresis. A collection of at least 55mg-75mg of sweat (15µl if using the coil method) is required by most laboratories to produce reliable results. Sweat must be collected over a period of 20-30 minutes at a rate of more than 1g/m^2/min to obtain a sufficient sample.[22,28,30] An elevated sweat chloride concentration of more than 60mmol/L is indicative of a

diagnosis of CF. A sweat chloride concentration below 40mmol/L excludes CF as a diagnosis. An intermediate result is considered an equivocal diagnosis.[22,24,28,30]

2.3.c. Other physiological measures

In rare cases, when a diagnosis is in doubt even after sweat testing and genetic testing, various other methods have to be employed to reach a diagnosis. The European Cystic Fibrosis Society Neonatal Screening Working Group has produced a set of guidelines regarding the further management of infants with an equivocal diagnosis of CF.[23] The sweat test can be repeated to confirm whether the result was actually equivocal. Further genetic analysis using a wider range of mutations may be carried out to identify rare combinations. Following this, if a diagnosis is still in doubt, a clinical assessment by a trained clinician should take place to identify any evidence of the presence/absence of clinical features suggestive of CF. Tests such as airway cultures, chest radiography and CT, bronchoscopy and fecal elastase should also be performed.[23] Measurements of defects in ion transport can also be carried out as further investigations. Table 2 gives details on the various options available.[23]

Test	Technical details	What it involves for the infant	Availability[a]
Nasal Potential Difference (PD)	Ion transport across airway epithelium can be assessed by measuring the baseline PD. The impact on the PD of perfusing different solutions and drugs provides further information to differentiate CF from non-CF recordings.	The exploring electrode is placed in the nose. A reference electrode is placed either subcutaneously or over abraded skin on the forearm. Solutions are perfused through the exploring electrode into the nose and can be swallowed.	Very few centres are able to undertake this measurement in infants although it is more widely available in older children and adults.
Intestinal Current Measurements (ICM)	A biopsy is mounted in the laboratory in a device (Ussing chamber) that enables measurement of transepithelial ion transport. Various aspects of ion transport can be examined.	Biopsy of rectal mucosa. This procedure is painless and well tolerated by young infants. Does not require general anaesthesia or sedation.	This technique requires a dedicated laboratory service with highly skilled technicians. Available in limited number of centres in Europe.
Small bowel biopsy	Similar measures of transepithelial transport processes can be undertaken in the laboratory on upper gastro-intestinal (GI) mucosal biopsies.	Upper GI biopsy; requires general anaesthesia in most cases.	Limited (only currently available in Sheffield, UK; contact Prof Chris Taylor).

Table 2. Tests to measure defects in ion transport[23]

2.3.d. Disadvantages of newborn screening

Newborn screening does have its disadvantages. Families waiting for a definitive diagnosis after a positive screening test may suffer from stress and anxiety. Inconclusive results may cause stress and uncertainties within families who aren't sure whether or not their child has CF.[22,26] False negative screening results may lead to a delayed diagnosis and a false sense of security in families.[27] Those with a mild phenotype who are identified by screening may be unnecessarily subjected to

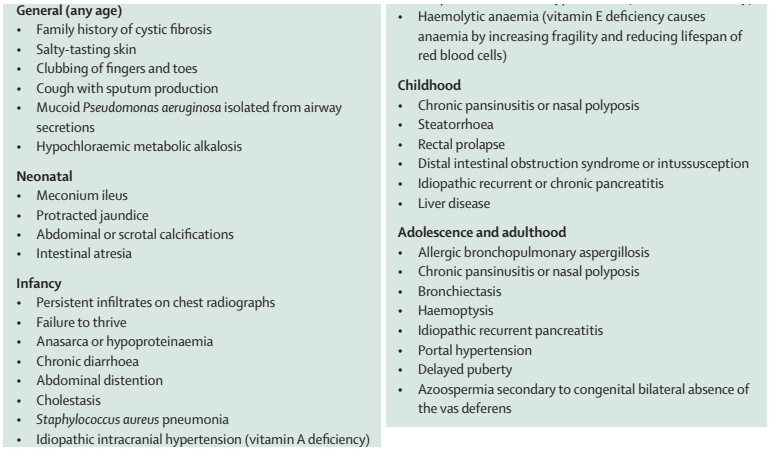
investigations and treatments.[27] One study in Wisconsin found that those who were identified by newborn screening were more likely to be infected with *Pseudomonas aeruginosa* at a younger age than those who had a clinical diagnosis. They also found that the screened group had a greater deterioration of the chest radiographs over time when compared to the clinical diagnosis group.[31]

2.3.e. Advantages of newborn screening

Newborn screening does, however, have its' advantages. Infants with CF identified by screening benefit from better nutrition and treatment at an early stage and improved growth as compared to children with a clinical diagnosis.[27] Those identified through screening have less severe lung disease than those identified as a result of clinical symptoms, unfortunately this was not found to be of long-term benefit.[22.27] A delayed clinical diagnosis is also quite stressful for parents due to the frequent unexplained episodes of illness before diagnosis. An earlier diagnosis by screening avoids this and also allows families to undergo genetic counselling for future pregnancies and cascade family testing. The long-term management of children identified through newborn screening is cheaper than that of those with a clinical diagnosis.[22,26,27]

2.4. PATHOPHYSIOLOGY AND CLINICAL FEATURES

CF manifests itself in a number of ways, with some features appearing as the disease progresses.[1,4] Abnormalities in the CFTR protein results in reduced transport of chloride ions and an associated reduction in the transport of sodium ions and water across epithelial cell membranes.[3,32,33] These effects are most significantly seen in the respiratory, hepatobiliary, gastrointestinal, pancreatic and reproductive tracts.[3] The table below (Table 3) lists some of the signs and symptoms commonly seen in CF.[4]



General (any age)
- Family history of cystic fibrosis
- Salty-tasting skin
- Clubbing of fingers and toes
- Cough with sputum production
- Mucoid *Pseudomonas aeruginosa* isolated from airway secretions
- Hypochloraemic metabolic alkalosis

Neonatal
- Meconium ileus
- Protracted jaundice
- Abdominal or scrotal calcifications
- Intestinal atresia

Infancy
- Persistent infiltrates on chest radiographs
- Failure to thrive
- Anasarca or hypoproteinaemia
- Chronic diarrhoea
- Abdominal distention
- Cholestasis
- *Staphylococcus aureus* pneumonia
- Idiopathic intracranial hypertension (vitamin A deficiency)
- Haemolytic anaemia (vitamin E deficiency causes anaemia by increasing fragility and reducing lifespan of red blood cells)

Childhood
- Chronic pansinusitis or nasal polyposis
- Steatorrhoea
- Rectal prolapse
- Distal intestinal obstruction syndrome or intussusception
- Idiopathic recurrent or chronic pancreatitis
- Liver disease

Adolescence and adulthood
- Allergic bronchopulmonary aspergillosis
- Chronic pansinusitis or nasal polyposis
- Bronchiectasis
- Haemoptysis
- Idiopathic recurrent pancreatitis
- Portal hypertension
- Delayed puberty
- Azoospermia secondary to congenital bilateral absence of the vas deferens

Table 3. Signs and symptoms of cystic fibrosis[4]

2.4.a. Pulmonary manifestations

The secretions in the lungs of a patient with CF are more viscous due to the DNA released by neutrophils and their degradation products. The DNA aggregates and causes the secretions to become more viscous.[3] As a result of the dysfunction in the CFTR, patients with CF have an abnormally low volume of isotonic liquid on their airway surfaces. This leads to reduced clearance of mucus from the airways. A build up of mucus in the airways predisposes the patient's lungs to recurrent bacterial infections which eventually become chronic infections.[32,34] Initially, infective organisms such as *Haemophilus influenzae* and *Staphylococcus aureus* are more prevalent. Organisms such as *Pseudomonas aeruginosa* and *Burkholderia cepacia* are more likely to be chronic later in life.[32,34] The host defense response in the lungs of patients with CF is primarily neutrophilic. Neutrophils lead to persistent airway inflammation eventually resulting in airway dilatation and bronchiectasis. Permanent damage to the structure of the lungs ensues.[32,34,35] Patients suffer from progressive tachypnoea, wheezing, dyspnoea and persistent cough. These symptoms may be intensified during pulmonary exacerbations. Eventually most patients develop respiratory failure, which is the main cause of mortality in patients with CF.[35]

2.4.b. Pancreatic disease

Pancreatic insufficiency develops in 85-90% of patients with CF with the exocrine function more commonly affected than the endocrine function.[4,33,36] There is decreased

secretion of chloride, sodium and bicarbonate ions. As a consequence, there is also a reduction in the secretion of water leading to dehydrated secretions with an increased protein concentration. The proteins are likely to precipitate within the pancreatic lumen and cause obstruction. This leads to acinar cell atrophy and a mild inflammatory reaction.[33] The pancreas of pancreatic insufficient patients has been found to secrete fewer digestive enzymes leading to malabsorption of fats, proteins and other nutrients, steatorrhoea, fat-soluble vitamin deficiency and malnutrition. Patients suffer from abdominal discomfort, bloating, flatulence and poor growth.[4,33,37] Vitamin deficiencies lead to anaemia, acrodermatitis, osteoporosis and bleeding disorders.[4] Eventually, the normal pancreatic tissue undergoes fibrotic changes with fatty replacement, cystic formations and calcification leading to destruction of the pancreas and cystic fibrosis related diabetes mellitus.[33,38] There has been some evidence showing that some patients who are pancreatic sufficient are likely to have deteriorating pancreatic function over time. Pancreatic acinar function is widely variable in this population of CF patients and eventually some patients do develop pancreatic insufficiency. These patients are also more susceptible to develop acute pancreatitis.[33]

2.4.c. Gastrointestinal manifestations

The gastrointestinal manifestations seen in CF are usually as a result of the viscous secretions present in the lumen.[39] Meconium ileus is present at birth in 10-15% of neonates with CF due to inspissated meconium in their bowels. This is due to defects in the transport of ions within the bowel, making the contents thick and sticky.[4,39] Later in life, especially during the second and third decade, 10-15% of patients will develop distal intestinal obstruction syndrome as a result of reduced gut motility, malabsorption and thickened faecal material obstructing the bowel lumen.[4,40] Other gastrointestinal manifestations seen in CF include fibrosing colonopathy, intestinal intussusception, appendicitis, rectal prolapse, gastro oesophageal reflux and an increased incidence of Crohn's disease.[39-41]

2.4.d. Hepatobiliary disease

After respiratory failure, liver disease is the second most common cause of death in patients with CF, with 20-50% of patients with CF developing some form of hepatic

involvement.[39-41] Viscous biliary secretions can lead to obstruction and inflammation of the intra-hepatic biliary ducts. This eventually develops into fibrosis and cholestatis, resulting in focal biliary cirrhosis, with a prevalence of 40%.[39,40,42] Gallbladder abnormalities can be seen in up to 50% of cases of CF and 12-24% of patients develop cholelithiasis. Obstruction of the gallbladder lumen with viscous secretions, leads to atresia, which is also known as microgallbladder and presents in 23-30% of CF patients.[39-41]

2.5. THERAPIES

When CF was first recognised as a disease in 1938, life expectancy from it was less than a year. At present day, as a result of medical advances in diagnosis and treatment, most patients with CF can expect to live into their forties.[1,5] As with other chronic diseases, treatment for CF is more inclined towards self-management by the patient and family at home.[43] Following a definitive diagnosis, treatment should be commenced as soon as possible to delay progression of the disease.[4,44] There are many options available, targeting various aspects of the disease. A multidisciplinary approach involving specialist physicians, nurses, physiotherapists, dietitians, clinical psychologists and the community team is vital in achieving these goals. Frequent assessment of the disease status and monitoring of treatment improves quality of life and survival.[4,45,46] The mainstay of treatment is to limit infective pulmonary exacerbations and optimise the nutritional status of the patient.[1,4]

2.5.a. Long-term respiratory management

2.5.a.i. Airway clearance and reducing viscoelasticity of airway secretions

The increased viscoelasticity of the mucus in the lungs promotes retention of the secretions resulting in obstruction of the airways.[3,32,34] Chest physiotherapy is the best method to clear these viscous chest secretions. There are various methods of physiotherapy recommended to clear the chest including simple breathing exercises, postural drainage, manual techniques such as percussion and vibration, and the use of instruments such as acapela and positive expiratory pressure (PEP) masks.[4,35,47] Conventional clearance techniques such as postural drainage, percussion and vibration often involves the assistance from another individual (e.g. a carer or physiotherapist). Some of the more recently developed airway clearance techniques

(ACT) including active cycle of breathing techniques (ACBT), forced expiration technique (FET), PEP masks, autogenic drainage (AD) and exercise can be self-administered.[3,48] A recent Cochrane review revealed that there was no difference between newer ACT's and conventional methods of physiotherapy except for individual preference. Most patients preferred using the newer ACTs compared to the older methods as these allowed for more independence, comfort and a wider choice.[48] Clearing airway secretions relieves obstruction and the symptoms of shortness of breath and reduced exercise tolerance, therefore it should be initiated as soon as possible after diagnosis.[4,47,48] Physiotherapy and ACT's should be tailored to the individual needs of the patient and family to ensure maximum compliance and effectiveness of the treatment.[45,47] Physical exercise is also conducive to airway clearance. It is widely encouraged by CF specialists as an adjunct to physiotherapy.[47]

Recombinant Human DNAse is capable of digesting the DNA released by the neutrophils in the airways. This reduces the viscosity of the airway secretions, thus decreasing airway obstruction, thereby improving lung function and limiting pulmonary infective exacerbations.[3,35] Hypertonic saline can also be used to rehydrate ciliary surfaces, aiding airway clearance and improving lung function.[49]

2.5.a.ii. Bronchodilator therapy

Bronchial hyperresponsiveness is more common in patients with CF than in healthy individuals with about 50% of patients suffering from it.[3,35] Bronchodilators such as beta-2-agonists (Salbutamol, salmeterol) and anticholinergics (Ipratropium bromide, tiotropium) are useful in relieving this obstruction. In the short-term they have been seen to improve the FEV_1 in 50-60% of patients. However, 10-20% of patients using bronchodilators have been found to have worsening lung function as a response to bronchodilators. Beta-2-agonists have also been found to be more effective than anticholinergics.[35,50] Despite the frequent use of bronchodilators in patients with CF, many more studies are still required to prove their effectiveness in the long-term. It has been recommended that patients should be prescribed a bronchodilator if there is an improvement of 10% or more in their FEV_1 after administration of the bronchodilator.[3,35,49,50] The patient should also be monitored during the course of the treatment to ensure that lung function is not deteriorating.[50]

2.5.a.iii. Antibiotic therapy

Many patients with CF are given regular antibiotics with a view of reducing infective pulmonary exacerbations and slowing the progress of pulmonary obstruction.[3] Infective organisms such as *Staphylococcus aureus* and *Pseudomonas aeruginosa* are quite common in patients with CF. They are associated with pulmonary inflammation and a decline in lung function. As a result, antibiotic prophylaxis has been advocated in certain patients with CF.[49,51,52] *Staphylococcus aureus* is more common in infancy. Antibiotics such a Flucloxacillin have been shown to reduce infection with this organism if started at a young age and continued for up to six years, although there is a lack of evidence showing whether this leads to improved lung function. Prophylaxis with anti-staphylococcal antibiotics may also have an association with increased infection with *Pseudomonas aeruginosa*. This finding may be incidental and has yet to be confirmed.[51,53] Infection with *Pseudomonas aeruginosa* is more common in CF as individuals grow older. Quinolones such as Ciprofloxacin are bactericidal antibiotics used against infection with *Pseudomonas aeruginosa*. Macrolides such as Azithromycin do not have a killing action against *Pseudomonas aeruginosa*. Nevertheless, they are effective against lung disease caused by this organism. A six months course of Azithromycin has been seen to have a small but significant effect on improvement of lung function in individuals with CF. Patients had fewer pulmonary infective exacerbations and a reduced need for other oral antibiotics. These effects were also seen beyond the six-month treatment period. Unfortunately, there is a high possibility of macrolide resistance among this organism.[52] The emergence of antibiotic resistant organisms creates constant challenges for the members of the CF team. To limit this, intermittent rather than daily administration of antibiotics has been suggested for such patients.[4,3,35]

2.5.b. Management of acute respiratory exacerbations

Infective exacerbations in CF are associated with increased symptomatology. Repeated episodes may lead to a deterioration in pulmonary function and a decline in life expectancy. Patients may require admission to hospital where they are likely to be started on two parenteral antibiotics for 14-21 days.[3,35] Some centres are now moving towards home intravenous therapy, which requires training and supervision but is also seen to be as effective as inpatient management.[35] The patient should be investigated

appropriately to identify the infective organisms and antibiotics altered/prescribed accordingly. Combination therapy with more than one drug is now being suggested for acute exacerbations to reduce antibiotic resistance. Antibiotic therapy is usually combined with intensified airway clearance techniques, physiotherapy, bronchodilator therapy and improved nutrition, either oral or parenteral.[3,35]

2.5.c. End of life treatment

2.5.c.i. Lung transplantation

The leading cause of death in patients with CF is respiratory failure due to irreversible damage to the structure of the lungs.[35] Since the 1980s, lung transplantation has become an option for patients with end-stage lung disease, including patients with CF.[54,55]

There are recommended guidelines for putting patients with CF on the transplant list.[55] There are three factors to consider when assessing CF patients for lung transplantation; a life expectancy of two years or less, poor quality of life and the absence of contraindications to transplantation.[56] The risk of death within two years is increased by a forced expiratory volume in one second (FEV_1) of less than 30% predicted for height. Female sex and younger patients are also at a higher risk of early death. Assessing quality of life is difficult in paediatric patients as there aren't many widely accepted methods of measuring it. Even so, if it is likely that the patient in question will benefit from an improved quality of life after having a lung transplant, they should be added to the transplant list. There are many contraindications to having a lung transplant. Some absolute contraindications include renal failure, as post-transplant immunosuppressive drugs may not be well-tolerated, active malignancy, coexistent HIV infection and ventilator dependant respiratory failure.[55-58]

Patients who have received lung transplants have a 70% survival rate in the first year and 45% at four years after the transplant.[57] Early organ rejection or severe infection due to immunosuppression are the main causes of death soon after transplantation. Bronchiolitis obliterans syndrome (BOS) is a condition that develops in 50% of long-term lung transplant survivors. It is the result of chronic rejection, and is irreversible. Other complications are usually due to the use of immunosuppressive agents such as

cyclosporine, prednisolone, azathioprine and tacrolimus. Patients are susceptible to infections, malignancies and drug relation organ damage especially to the kidneys.[55,59]

2.5.c.ii. Palliative care

Due to a shortage of donor organs, many patients with CF will die while on the waiting list for a lung transplant. Therefore, there is a need for palliative care of terminally ill CF patients.[60] Dyspnoea, fatigue, anorexia, pain, anxiety and unconsciousness are all symptoms commonly experienced by CF patients towards the end of their lives.[61] CF patients tend to fluctuate in terms of their symptoms at this stage of their lives.[62] In contrast to palliative care for other diseases, most patients with CF continue having their regular therapies including antibiotics, oxygen and physiotherapy till the end.[55,60,63] Opiates may be used to relieve dyspnoea and pain experienced by patients.[62,64] Benzodiazepines are successful in reducing anxiety, dyspnoea and seizures due to hypercapnia caused by respiratory failure.[61,62] Non-invasive ventilation is usually only used in patients waiting for a lung transplantation, but has also been used in palliative care for CF patients.[55,62] Consideration has to be given to adequate communication with the patient and the family about what symptoms and treatments to expect during the palliative care period. The patient and the family should be able to choose where they would like to be treated; either at home, in the hospital or in a hospice. Counselling and support should be provided for the patient and family during the process and bereavent counselling should be offered to the family after the death of the patient.[62,63]

2.5.d. Nutritional management

It is a well-recognised fact that well-nourished patients with CF have better pulmonary function than those who are malnourished.[4,36,65] Disease progression in CF is the main cause of nutritional problems for patients. Declining pulmonary function, pancreatic insufficiency, intestinal malabsorption, hepatobiliary and intestinal complications all contribute to malnutrition. Patients with CF also have increased energy demands due to increased energy losses in stool and urine, increased energy expenditure and a decreased intake as compared to the normal population.[46,65,66] As a result, there is a loss of lean body mass, nutritional deficiencies and depressed immune function.[36,46,65]

2.5.d.i Pancreatic Enzyme Replacement Therapy

Once pancreatic insufficiency has been identified, Pancreatic Enzyme Replacement Therapy (PERT) should be initiated. PERT should be given with all food and milk products. Preparations with acid resistent enteric coating prevents the enzymes from being inactivated by stomach acids. These are preferred to powder preperations as they are not associated with mouth or perianal excoriations in addition to being acid resistant.[36,66] Histamine-2 receptor blockers or proton pump inhibitors may be administered concomittantly to reduce the acidity of the small intestine and to activate the enzymes.[36] Enzymes can be taken before the meal or spread throughout the meal depending on its' size.[36,66] Breast feeding is encouraged in infants and those with pancreatic insufficiency should be started on 5,000-10,000 IU lipase per 60-120ml of standard formula feed or per breastfeed. The dose should be titrated against symptoms of malabsorption and growth rates.[44,66] PERT is associated with fibrosing colonopathy and therefore doses should not exceed 10,000 IU lipase/kg bodyweight/day.[36,66]

2.5.d.ii. Nutrition

Newborn screening has helped identify children with CF at an early stage. This has lead to earlier nutritional interventions and the prevention of malnutrition in infancy.[27] Due to fat malabsorption, patients with CF require a greater fat intake (35-40%) than the normal population (≤ 30%).[36] As patients with CF have high energy requirements, they are encouraged to eat energy dense foods with high protein and fat contents and to avoid, as much as possible, low fat and low calorie foods. Patients should have up to 150% of the recommended daily requirements to achieve normal growth and adequate energy.[36,44,46,65,66] Fat soluble vitamin supplements (Vitamins A, D and E) are generally required to prevent deficiencies. There are no recommendations on supplementation of vitamin K as yet.[36,44,66] Supplementation for minerals and electrolytes such as calcium, iron and zinc are recommended if deficiencies are identified. Patients should take sodium supplements to prevent hyponatraemia due to excessive losses in sweat associated with warm weather.[36,44,66]

2.5.d.iii. Interventions for patients with nutritional failre

2.5.d.iii.1. Oral supplementation

Growth should be regularly monitored for every CF patient. Those who are not growing well may need to take oral calorific supplementation in addition to their normal diet. These should not be used as a substitute for normal meals.[36,66]

2.5.d.iii.2. Enteric nutrition

After the failure of oral supplementation to provide adequate nutrition, enteric feeding should be considered. Enteric feeding is recommended in children with a weight for height ratio of less than 85%, and in adults with a BMI of less than 19. It is associated with improvements in weight, height, improved strength and the development of secondary sexual characteristics.[65,66] There are various options available for enteric feeding, of which nasogastric and gastostomy tubes are the most popular. Overnight feeding encourages normal feeding behaviours during the day.[65,66] Up to 120-150kcal/kg/day may be required to achieve significant growth. Growth should be monitored and feed volumes should be titrated against growth rates to ensure adequate supplementation.[36] Enteral feeding routes should be used on a long-term basis to achieve significant improvements in growth and health.[65,66] There is still a lack of evidence regarding the need for pancreatic enzymes during enteral feeds. The CF Trust recommends that half the normal dose of enzymes should be given prior to the feed, and the remainder should be given in the middle or at the end of the feed.[66]

2.5.d.iii.3. Behavioural interventions

Some children with CF develop adverse mealtime behaviours, which should be identified as early as possible. School age children (five to ten years) are particularly at risk of these behaviours and special attention should be given at these times.[36] Behavioral interventions may be effective in altering feeding behaviours and promoting healthy attitudes towards meals in children of three to twelve years. Education of the patients as well as the family is very important in this respect.[36]

2.6. MONITORING

2.6.a. Respiratory monitoring

2.6.a.i. Pulmonary function tests

It is essential to regularly assess the respiratory status of patients with CF to monitor disease progression. Pulmonary function is an important indicator of disease progression and severity in patients with CF. Routine pulmonary function tests with spirometry in age appropriate children (five years and above) allows early detection of deteriorating health.[35,45] Spirometry enables the measurement of the forced vital capacity (FVC), forced expiratory volume in one second (FEV_1), maximal forced expiratory flow (FEF max) and forced expiratory flow between 25% and 75% of the vital capacity (FEF_{25-75}).[45,56] Serial measurements enable the monitoring of pulmonary function over time. As patients with CF develop lung disease, their FEV_1 will start to decline.[35] An FEV_1 of less than 30% predicted should be an indication of end stage lung disease and for referral for a lung transplantation.[56] FEV_1 is the best clinical predictor of mortality in patients with CF. It is a standardised measure and the most commonly used indicator of lung function worldwide. It has been used in numerous studies as an outcome measure.[35,45,56]

2.6.a.ii. Other pulmonary investigations

Imaging techniques such as chest X-Rays and computed tomography scans (CT) provide a means of visual evaluation of lung conditions. Chest X-rays have been used frequently to identify progressive disease, although they are not very sensitive in identifying changes during acute pulmonary exacerbations and early stages of the disease. CT is a more sensitive and specific imaging technique than chest X-ray. It is easier to identify pathology and disease progression on a CT scan than on X-ray.[35] Sputum cultures can be used to assess which organisms have colonised the individual. Samples can be obtained from a cough swab or from expectorated sputum. Depending on the results of these tests, appropriate therapeutic interventions can be suggested or adjusted for the treatment of pulmonary infections.[35]

2.6.b. Nutritional monitoring

2.6.b.i. Anthropometric measurements of growth

Early recognition of poor growth leads to early interventions and better health in patients with CF. Particular importance should be given soon after diagnosis, in the first year of life and in the peripubertal growth period (Girls 9-16 years, boys 12-18 years).[36] Patients' growth should be monitored regularly by measuring their height/length and weight and in children under five, head circumference as well. Measurements should be recorded on relevant centile charts to track the rate of growth and compare it to the normal population.[36,44.45,66] Calculations such as, body mass index (BMI), percentage weight for age, height for age, weight for height and weight, height and BMI standard deviation scores (Z Scores) are used to quantify growth and sequential measurements can be used to measure progress of growth.[45,66] Mid-arm circumference and triceps skin fold thickness are measurements of lean body mass development and subcutaneous fat reserves.[36,66]

2.6.b.ii. Pancreatic assessment

Pancreatic insufficiency should be identified as soon as possible. In Northern Europe, 95% of infants with CF are pancreatic insufficient by the end of their first year. However a considerable number are born pancreatic sufficient. Therefore, after diagnosis of the newborn, pancreatic function must be assessed by measurement of stool fecal elastase. A fecal elastase concentration of less than 100μg/g is indicative of pancreatic insufficiency. This assessment should be repeated during the first year of life if found to be normal at diagnosis.[44,66] In patients who are found to be pancreatic sufficient, annual assessments of pancreatic functions should be carried out in order to identify deteriorating pancreatic function.[36,45]

2.6.b.iii. Other measurements of nutrition

Fat-soluble vitamin (vitamins A, D, E and K) levels should be regularly measured to identify vitamin deficiencies. Plasma levels of vitamins A, D and E should be assessed annually. Prothrombin time is used to measure vitamin K levels.[36,45,66] Calcium and iron levels should also be measured to identify deficiencies.[36]

A 24-hour dietary diary can be used to assess dietary intake. Parents/patients report all meals, snacks and supplements ingested within a normal 24-hour period. This allows for a quantitative analysis of daily nutrition.[36,45]

2.6.c. Outpatient assessments

Patients with CF should be seen at an outpatient CF clinic every six to eight weeks or at least once every three months.[67] Patients should be segregated as much as possible from each other to minimise cross infection.[45,67] The following standards for outpatient assessments have been recommended by the CF Trust, UK:[67]

- Height and weight measurements
- Spirometry
- Sputum cultures for *Burkholderia cepacia, Staphylococcus aureus* and *methicillin resistant Staphylococcus aureus (MRSA)*
- Chest X-Ray if clinically indicated
- Consultation and examination from a CF specialist consultant including medication and symptom reviews
- Physiotherapist review
- Dietitian review
- CF nurse specialist review
- A social worker should be made available if required
- Access to a clinical psychologist with experience in CF

Patients with CF should also have a detailed annual review every year to assess progression of disease and reassess treatments if required.[45,67] The annual review is a more detailed process than the routine outpatient CF clinic and covers all aspects of CF.[45,67]

THE BURDEN IN CYSTIC FIBROSIS

3.0. THE BURDEN IN CYSTIC FIBROSIS

3.1. THE CONCEPT OF BURDEN

The adverse consequences of the role of caring, on a carer is known as the burden of care.[68,69] Carers may feel that they are expected to do a lot 'more than their fair share' of tasks, as their caregiver role is an additional role to their usual role within the family. They may also feel restrictions on their time, finances and personal activities, leading to dissatisfaction with their role.[68-70]

Burden is considered to be a multidimensional concept, arising from patient behaviours, symptoms, role strains within the family and treatment demands.[6,68,70] A number of studies have investigated the caregiver burden associated with psychiatric diseases.

3.1.a. Burden of care in psychiatry

Carers of adult patients with psychiatric disorders have reported anxiety attributable to the care recipient. Psychiatric illnesses are often associated with negative illness-related behaviours which carers may find difficult to understand and accept. They fear social stigma and discrimination against the patient, and many resort to social isolation to avoid this embarrassment. The role of caring can also lead to financial problems. There is a loss of income due to the added responsibility of caring and/or the care recipient was originally the main income provider and is no longer in a position to earn a living. Caring for a relative can also lead to a strain in the relationship between the family members due to feelings of anxiety, guilt, rejection and resentment. Despite this, most carers have been found to be quite tolerant of their affected family members.[69,70]

Another study found that parents of children with psychiatric symptomatology were more likely to report a higher perceived burden than parents of children without psychiatric symptomatology. These parents felt the greatest effects on personal well-being, stigma and restrictions on personal activities.[71]

3.1.b. Burden of care in paediatric conditions

The emphasis on community-based care is gradually increasing for children with physical and mental disabilities and chronic diseases such as CF, diabetes and asthma. The responsibility of care usually lies with the child's main caregivers. Most of these children have to perform complicated, time-consuming treatment tasks on a routine basis. These intensive treatment demands can have an adverse effect on compliance and the general well being of the patient and their families.[7] Ziaian et al compared treatment time and hassle in children with CF, diabetes and asthma.[7] They found that children with CF and diabetes spent significantly more time per day performing treatment tasks than children with asthma. Although all children were found to have 'low' levels of hassle, those with CF and diabetes reported significantly higher hassle scores than those with asthma. These significant differences probably arise as a result of the more complex nature of the disease and treatment tasks in CF and diabetes than in asthma.[7]

A Danish study on severely mentally retarded children revealed that families of these children spent an average of 7 hours 11 minutes per day providing care, supervision, occupation and training for these children. 80% of parents reported disturbed sleep as a direct result of their child. Three quarters of the children in this population required constant supervision and could not be left alone. This had an adverse effect on time spent on other social and recreational activities.[72]

A more recent study compared healthy children with children with severe mental and physical disabilities such as cerebral palsy, autism, Sanfillipo syndrome, lissencephaly and osteogenesis imperfecta.[73] This study revealed that mothers of affected children spent significantly more time per day performing personal care tasks for their children than those of healthy children. This was seen to have an adverse effect on the occupational lives of these mothers, which in turn impacts on the family income.[73]

3.1.c. Burden of care in CF

Advances in medical care have resulted in a longer life expectancy for children with chronic life threatening diseases. The progressive nature of these conditions has meant that children require intensive, specialized, and often time consuming care. With recent trends towards community treatment, the duty of care usually lies with the parents and carers at home. Not surprisingly, this contributes to increasing family stress, marital disharmony, depression, role and routine disturbances.[74-76] A number of studies have been carried out looking at the psychosocial impact of CF on carers.

3.1.c.i. The impact of diagnosis

An early diagnosis of CF brings with it nutritional benefits and the chance to initiate treatment as early as possible. For parents and carers this can be a very stressful period, coming to terms with the diagnosis, making adjustments in their routines, learning and putting into practice new treatment regimes. Parents are likely to develop psychological symptoms during this period.[77,78] A study published in 2007 revealed that parents were at a higher risk of depression if their child was nine months or younger at the time of diagnosis. Compared to results from parents with healthy children, both parents of a child with a recent diagnosis of CF were more likely to suffer from dysphoria during the first year after diagnosis.[77] Similar results were shown in another study, where parents of a child with a recent diagnosis reported significantly more depressive symptoms and marital stress than those of healthy children[78]

3.1.c.ii. Stressors and role strains

Children with CF have specific needs and requirements. This gives rise to specific stresses and strains within their families, especially between parents.[75] Quittner et al investigated the relationships between CF specific tasks, normal parenting tasks, role strains within the family and parental depression in parents of children with CF.[78] Parents of children with CF reported that their children were more demanding and less adaptable than the normal population. The most stressful situation for parents was found to be 'outings in the community,' followed by CF specific stressors such as 'maintaining diet,' 'medications,' 'chest physical therapy,' and 'finances related to CF.' Least stressful situations were 'siblings,' 'parents' relationships with friends and

family' and 'child's relationships with peers.'[78] A high correlation was found between CF specific stressors and parental depression.[78] Maternal depression was associated with marital role strain. This study also found that unemployed mothers were more likely to report role strains and depressive symptomatology than working mothers.[78] Fathers reported more difficulties performing CF related medical tasks and higher levels of financial stress as compared to mothers. CF specific parenting was associated with depression in fathers in this study group.[78]

A 1998 study compared stressors and role strains between parents of children with CF and a control group of parents of healthy children.[75] It found that parents of children with CF reported significantly more conflicts in child-rearing, more child care tasks, unequal role division between partners, and fewer positive daily interactions between them as compared to parents of healthy children.[75] The CF group also spent less time on recreational activities and more time on child-care and medical tasks. The mothers in the CF group reported the highest levels of stress among all participants.[75] Associations were found between higher role strain and decreased time and satisfaction from recreational activities.[75] Surprisingly, there was no significant difference found in marital satisfaction and depression between the CF group and healthy controls.[75]

3.1.c.iii. Coping strategies

When faced with such strains and stresses, people develop various coping mechanisms, hopes and fears that affect adjustment processes. An Australian study identified the coping strategies used by parents of children with CF and their vicarious hopes and despairs. They also investigated the associations between these and child and parental adjustment.[79] Coping strategies reported by more than 70% of the study population as being used 'a lot' or 'a medium amount' were:

- Acceptance ('accepting the fact that the stressful event has occurred and is real')[79]
- Active coping ('exerting effort to remove or circumvent the stressor')[79]
- Planning ('thinking about how to confront the stressor)[79]
- The use of Emotional support ('seeking moral support, sympathy or understanding').[79]

Strategies reported as being used 'a little bit' or 'not at all' by over 70% of participants were:

- Substance abuse ('using alcohol or other drugs to disengage from the stressor')[79]
- Behavioural disengagement ('withdrawing effort from the attempt to attain the goal the stressor is interfering with')[79]
- Denial ('attempting to reject the reality of the stressful event')[79]
- Religion ('engaging in religious activities').[79]

The use of self-blame ('criticizing or blaming oneself for the stressor') as a coping mechanism by 42% of parents was associated with more parental depression and anxiety and poor child and parent adjustment. Self-distraction tactics also had a similar effect of increased parental depression, anxiety and emotional distress.[79] Reports of vicarious hope were more likely than reports of vicarious despair to be associated with better child physical function and mental health, lower parental depression, anxiety and emotional impact as a result of CF.[79] Poor child health was seen to be associated with parents who had high vicarious despair scores.[79]

3.2. TREATMENT BURDEN AND ADHERENCE IN CYSTIC FIBROSIS

3.2.a. Treatment burden

Treatment of CF is a complex process. Most patients have to take medications, dietary supplements, and nebulisers and perform physiotherapy on a regular basis. During episodes of acute illness, self-management at home can become even more challenging for the patient as well as the family.[6,7,8] Patients and their families have to devote a lot of time and effort to perform treatment tasks everyday. Thus it comes as no surprise, that patients with CF and their families may report a high treatment burden.[6,7,8] Treatment burden in CF is influenced by the number and frequency of daily treatments tasks, the complex nature of performing the treatments and the time spent on them. It can also be influenced by the effect of the treatments on the patient's and family's life.[6,7]

Sawicki et al investigated the relationship between perceived treatment burden and treatment activities in adult patients with CF. They carried out a survey on treatment activities performed the day before by the patients. They also assessed how often the

patients reported performing these treatments.[6] Treatment burden was measured using the treatment burden subscale of the 'Cystic Fibrosis Questionnaire- revised version' (CFQ-R).[6] This is a CF specific Health Related Quality Of Life (HRQOL) questionnaire.

Ziaian et al assessed the time and effort spent on treatment tasks and the effect of this on the HRQOL in a paediatric CF population. Investigators used telephone interviews to identify all treatment tasks carried out in the preceding 24 hours. They noted the time, nature and hassle involved with each treatment task. FEV_1 scores were used to assess disease severity and HRQOL was measured by using the child and parent versions of the 'Child Health Questionnaire' (CHQ).[7]

Children reported a mean of 5.8 separate treatment tasks per day, whereas adults reported a median of seven medications per day, most of which were oral medications.[6,7] Patients with CF were found to spend a considerable amount of time each day performing treatment related tasks including oral medications, nebulisers and physiotherapy. Adults spent a mean of 108 minutes per day and children reported a mean of 73.6 minutes per day spent on treatment tasks.[6,7] Both studies revealed a significant association between treatment activities and treatment burden. Patients who perform more treatment tasks and spend more time per day on them are more likely to report a higher treatment burden.[6,7] A 2005 study in Belgium used the CFQ-R to compare HRQOL reports between children with CF and their parents. This study revealed that parents reported a higher treatment burden than their children, even though both groups rated disease severity similarly. The children reported a higher overall HRQOL than their parents.[80] Similar results were reported by Britto et al in 2004 where adolescents with CF reported a higher overall HRQOL than their parents.[81]

3.2.b. Adherence

Time constraints and treatment demands play a significant role in the lives of patients and families with CF. It starts at a young age and carries on into adulthood. This has been seen to adversely affect compliance with treatment.[6,7] Treatment compliance in chronic diseases such as CF has been estimated around 50%.[82,83]

A number of studies have investigated treatment compliance in patients with CF. Performing physiotherapy was found to be the most problematic for patients.[82-85] Arias Llorente et al reported a 38% compliance rate for physiotherapy as compared to 100% compliance for digestive medication (PERT) in adults and children with CF.[84] Abbott et al reported poor compliance for physiotherapy by 47% of their study population of adults with CF.[85] Reasons given for poor compliance with physiotherapy include:

- Not effective[83-85]
- Too much time, commitment and effort involved[82-85]
- Not required[84]
- Exercise is used as a substitute for physiotherapy[83-85]
- Embarrassment and social intrusion[82-85]
- Forgetting[82-85]

The greatest compliance was seen with pancreatic enzyme supplements.[84,85] Modi et al also reported good compliance with pancreatic enzymes with 85% of their patients.[82] Arias Llorente et al found that the discrepancy between compliance rates of physiotherapy and digestive enzymes may be due to the fact that approximately 90% of the participants felt that digestive medications had a positive impact on their quality of life as compared to approximately 50% who felt that physiotherapy had very little/ no effect on their quality of life.[84]

Compliance for oral and inhaled medications ranged between 50% and 83%.[83,84] Reasons for non-compliance included:

- Forgetting[82-85]
- Bad taste/texture[82-84]
- Too much time and effort[83,84]
- Social intrusion[84]
- Not effective[84]

Overall, researchers have found that treatments with visible short-term benefits and the early appearance of adverse effects due to non-compliance are the ones that are adhered to most commonly. This may be the reason why pancreatic enzymes have a high rate of compliance, as the unpleasant effects of steatorrhoea are reduced after taking PERT.[84,85]

Nebuliser devices such as the I-neb and the Prodose are part of the newer 'adaptive aerosol delivery' (AAD) breath activated systems that are smaller, portable and faster ways of administering drugs via a conventional nebuliser.[86,87] They also have microchip technology, which allows health care professional to monitor adherence in patients using them.[86,87]

McNamara et al studied AAD nebuliser adherence in a population of children with CF who had been infected with *Pseudomonas aeruginosa*.[87] Over a period of one year, data was downloaded from the I-neb's of 28 children who had been prescribed long-term antibiotics. Patients and families were also regularly counseled regarding their adherence.[87] Monthly adherence was found to be between 60-70% over the duration of the year. The I-neb data revealed that mean (Standard Deviation (SD)) treatment time increased significantly over the year from 3.8 (2.1) minutes in the first month of treatment to 5.9 (4.2) minutes by the end of the year ($p < 0.05$). Adherence was also found to be better in the evenings than in the mornings when families were at their busiest.[87]

Latchford et al used the Prodose AAD device to monitor adherence in adult patients with CF who had been prescribed antibiotics.[86] Data was collected from the devices of 38 patients for a period of 12 weeks.[86] This study found adherence rates to be as low as 50% and participants were only fully adherent to their nebulised antibiotics for a mean (SD) of 31.6% (29.4) of days. Younger patients had poorer adherence than the older ones.[86]

Adherence can be influenced by a number of factors depending on the particular treatment in question, the patient and their family.[82-88] Poor adherence can lead to ineffective treatment, progression of disease, increased mortality and poor quality of life.[88] It is important for medical professionals to measure adherence and intervene and counsel patients appropriately to improve treatment compliance.[86,87]

3.2.c. Measuring treatment burden in CF

3.2.c.i The Cystic Fibrosis Questionnaire (CFQ)

Modi et al have validated a CF specific questionnaire, the Cystic Fibrosis Questionnaire (CFQ), to measure HRQOL of patients with CF and carers of children with CF.[89] It was first designed in France.[89,90] The CFQ-child version is designed for children/adolescents with CF aged 6-13. The CFQ-parent version is designed for parents of children aged 6-13, with CF. The CFQ-child contains eight domains:[89]

- Physical symptoms (6 items)
- Emotional functioning (8 items)
- Social functioning (5 items)
- Body image (3 items)
- Eating disturbances (3 items)
- Treatment Burden (3 items)
- Respiratory symptoms (4 items)
- Digestive symptoms (1 item).

The CFQ-parent contains eleven domains:[89]

- Physical symptoms (9 items)
- Emotional functioning (5 items)
- Vitality (5 items)
- School functioning (3 items)
- Eating disturbances (3 items)
- Body image (3 items)
- Treatment burden (2 items)
- Respiratory symptoms (6 items)
- Digestive symptoms (3 items)
- Weight (1 item)
- Overall health perception scale (3 items).

Both questionnaires are scored out of 100, with higher scores indicating better HRQOL.[89] The treatment burden subscales are quite small and limited measures, yet these questionnaires have been used to measure treatment burden in patients with CF.

3.2.c.ii. Development of the Challenges of Living with Cystic Fibrosis Questionnaire (CLCF-Q)

The Challenges of living with Cystic Fibrosis Questionnaire (CLCF-Q) is a parent reported outcome measure. It was developed to assess the extent of the challenges faced by carers of children under 13 years with CF. It measures the time and effort invested in caring for a child with CF.[10,11] It consists of 62 items divided between 10 sections:

- Family Lifestyle
- CF Background
- Child's Character
- Challenges to Family Life
- Hopes and Worries
- CF Routines
- Community Support
- CF Clinic and Pharmacy Visits
- Inpatient and Day Patient stays
- CF Treatments

The CLCF-Q makes use of four and five point Likert scales as well as some 'Yes, No, Don't know' nominal scales. A few items require more descriptive responses.

Development began with a consensus panel of CF healthcare professionals, who designed a score sheet for the time spent on CF care everyday. This was followed by a focus group of eight parent caregivers of children with CF aged 13 years and younger. They discussed the carers' views on what they felt were important challenges faced by them routinely in CF care. From this discussion, a thematic analysis was performed. This analysis was presented back to the focus group, now an action research group. It was further discussed and critiqued. A questionnaire with various themes was constructed. A second focus group of three parent caregivers further reviewed this first draft questionnaire. It was then tested by seven caregivers, from both focus groups, to assess its' validity and acceptability. [9]

The instrument then had to be refined. Nine carers, who were not familiar with the initial development stages, were asked to complete the questionnaire prior to a

cognitive interview with a researcher. They were asked how and why they answered the questions the way they did. This was to assess how clear the questions were to a new set of participants. Changes and adjustments were made in response to their interview answers and feedback.[9]

Finally, a pilot study was conducted to assess validity, floor ceiling effects and test-retest reliability. 30 participants completed the questionnaire twice, seven days apart. Convergent validity was tested by comparing the CLCF-Q scores with three burden specific scales on the CF-Q. The predictive validity of the CLCF-Q was also examined by looking for associations between the CLCF-Q scores and clinical severity as determined by FEV_1% predicted and the presence or absence of *Pseudomonas aeruginosa* infection.[9]

The CLCF-Q was administered to the principal caregiver during the annual review process. The annual review is generally viewed as a stressful situation by the family. This was taken into consideration when designing the questionnaires. This study has not yet been published and is in the process of being written up.[9]

SELF-EFFICACY

4.0. SELF-EFFICACY

Self-efficacy is a term used to describe the belief held by a person, about how capable he/she is to perform a particular task.[8,91-94] Albert Bandura quotes it as:

'... Beliefs in one's capabilities to organize and execute the courses of action required to manage prospective situations.'[95]

It can influence a persons' behaviour, goals, perseverance for particular tasks, choices, emotional thoughts and feelings. It plays a role in encouraging new behaviours and discouraging, resuming or maintaining old ones.[13,95]

'Outcome expectations' refers to the belief held by an individual about whether a particular behaviour will lead to a particular result. Self-efficacy expectations can be altered by the magnitude, strength and generality of the task and the individual.[12,13]

- Magnitude refers to how difficult one grades various tasks. Those with low magnitude expectations may attempt easier tasks than those with higher magnitude expectations.[13]
- Strength refers to one's own judgement of the probability to perform that particular task.[13]
- Generality refers to how an individual adapts self-efficacy expectations about one experience to other similar, but not identical experiences.[12,13]

A person will carry out a certain behaviour based on their efficacy expectations and outcome expectations. If a person believes that he/she is capable of performing a given task, which will lead to a desirable goal, then he/she is more likely to proceed and succeed. Individuals with higher self-efficacy for particular tasks are more likely to develop purposeful behaviours and persist with them, leading to better performance. Conversely, those with low self-efficacy may get anxious about certain tasks, which may lead to avoidance behaviour or failure.[8,13,91,92,94]

4.1. SOURCES OF EFFICACY BELIEFS

Self-efficacy can be influenced by a number of factors; mastery experiences, vicarious experiences, verbal persuasion and physiological and emotional states.[8,13,91,92,95]

4.1.a. Mastery experiences

This is one of the most effective methods to boost one's self-efficacy. Previous successful experiences enhance self-efficacy. Individuals develop the skills and coping mechanisms for the future, whereas failure may have the opposite effect.[8,13,91,95] A study on patients recovering from a recent MI, showed that those who performed successfully on a treadmill test had higher self-efficacy for physical activities at home.[12]

4.1.b. Vicarious experiences

This is learning from the observation of others' experiences. From vicarious experience, observers may acquire the necessary skills and strategies to improve their behaviour in similar situations resulting in higher self-efficacy. The observer should be able to relate to the model being observed. If they feel superior or inferior to the model, the experience may not have a positive effect on the self-efficacy of the observer. An upward comparison is a situation where the observer feels that the model is performing better than themselves. A downward comparison is a situation where the observer feels that the model is performing worse than they themselves are capable of. Therefore, the observation of the successes/failures of an individual seen as an equal, may lead to enhancement/discouragement of ones' own self-efficacy.[8,13,91,92,95,96]

4.1.c. Verbal persuasion

Verbal encouragement from others may boost self-efficacy. Individuals may feel more confident, which results in more energy being spent to achieve the end goal. Those who are verbally discouraged may lose confidence and not attempt the said task, resulting in failure.[13,95] In Ewart's study on post-MI patients, those who received detailed feedback and explanation of their treadmill test results from health care

professionals were found to have higher self-efficacy for a wide range of physical activities.[12]

4.1.d. Physiological and emotional state

These are the emotional and physical responses to certain experiences. Situations, which lead to high psychological arousal, such as feelings of stress and anxiety, may be interpreted as signs of inadequacy, leading to a decrease in self-efficacy and eventual failure. Whereas individuals who are generally happier, seem to report higher levels of self-efficacy. Physical challenges which trigger fatigue, weakness and pain may also be viewed negatively and reduce self-efficacy.[8,13,91,95,96] Patients who suffered from angina-like pain during their post-MI treadmill test were more likely to be discouraged and report a lower self-efficacy.[12]

4.2. INDIVIDUAL APPRAISAL OF EFFICACY INFORMATION

How individuals interpret information and experiences may also have an effect on self-efficacy. Different people interpret situations in different ways. What one person learns from an experience may not be what another person may learn from the same experience. Self-efficacy is influenced by the features of the experience that are retained most in the individuals' memory, the importance given to various aspects of the experience, the reliability of the source of information and how much the success of the experience is attributed to self effort or to chance and external factors.[13,95] Those with strong efficacy beliefs are more likely to persist in their efforts and aim to overcome obstacles after experiencing failure.[95,97]

4.3. SELF-EFFICACY IN PARENTING

Parenting can be stressful at times. The role of a parent is constantly changing as their child develops. Most parents have multiple roles to play, within the family as well as outside of it. The parental role, strains and tests parenting self-efficacy.[95]

'Parenting self-efficacy involves a parent's beliefs in their ability to influence their child and the environment, in ways that would foster the child's development and success'[98]

Parents with a high parenting self-efficacy have more confident and effective parenting skills such as improved parent-child interactions, increased maternal

sensitivity, warmth and responsiveness. These strategies promote positive childhood development.[97-100] In 1998, Tucker et al found that the group of mothers with a higher mean parental self-efficacy had more positive interactions with their children. Compared to the group of mothers with lower parental self-efficacy, these mothers exhibited fewer negative physical behaviours and more praise statements towards their children.[99] Teti and Gelfand found that higher maternal self-efficacy was associated with higher maternal behavioural competence including showing increased sensitivity and warmth towards their infants.[100] A study carried out on a group of Mexican immigrants in America also found a predictive association between self-efficacy and parental warmth and control towards their children.[101] Maternal parental self-efficacy has also been seen to be positively related to responsiveness towards their children.[102,103]

Parenting self-efficacy can be affected by environmental and family contexts such as cultural background, socio-economic status, surrounding neighbourhoods, social support and the children themselves.[97,101] Ardelt and Eccles showed that mothers from disadvantaged social groups with strong self-efficacy beliefs were more likely than mothers from advantaged social groups, to engage in promotive parenting activities to enhance their child's social, mental and emotional development. Children of these mothers also had higher self-efficacy and greater academic successes than their peers whose mother's had lower self-efficacy beliefs.[97] Children with challenging problems such as learning difficulties and behavioural problems could also affect parental self-efficacy.[97,104] Sanders and Woolley found that mothers of children attending a clinic for childhood disruptive behaviour had lower task specific self-efficacy than non-clinic mothers.[104] Parenting self-efficacy in turn impacts directly on child self-efficacy and indirectly on child anxiety through observation of the parent by the child.[97,105] Adolescents of parents with high parental self-efficacy have been found to have fewer behavioural problems and to be better adjusted. Parents with low parental self-efficacy have also reported more behavioural problems in their children.[101,103] Thus parental self-efficacy may be used as a predictor of parenting behaviours, childhood outcomes and as an indicator of risk to the family.[98,104]

Interventions such as parent training courses can be a useful tool to boost parental self-efficacy. In Tucker et al's 1998 study, mothers who received Behavioural Parent

Training (BPT) in the form of Webster-Stratton's intervention reported an improvement in their parental self-efficacy and reduction in parental stress from pre-intervention to 1 year after they received their training.[99] Miller-Heyl et al also found similar results of an increase in parental self-efficacy after an intervention with their 'DARE to be you' program which provided training to parents of 'high-risk' children in the form of decision making, problem solving, communication skills, and conflict management skills.[106]

4.4. SELF-EFFICACY IN CHRONIC DISEASE MANAGEMENT

With the advent of modern medicine, there has been a trend for chronic disease management to move from an inpatient hospital setting, to self-management at home.[43,107] Self-management in paediatric care involves both the patient and carer carrying out behaviours to ease the impact of the disease on the family's life and improve their quality of life as a whole.[8,43,107-109] Most families develop certain coping mechanisms to lead as normal a life as possible. Skills such as self-monitoring, decision-making and communication between the family as well as members of the health care team are important in the self-management of a disease.[8,43,108,109] Continued learning, development and maintenance of complex skills are important to improve care as the disease progresses.[8,43] Many of these skills are specialised and are not natural, instinctive skills, but rather they have to be taught to the parents and children by the health care professionals.[8,43,107,108]

Self-efficacy in chronic disease management can help empower the patient and family with the skills and confidence required to manage the disease. Those with a higher self-efficacy are more likely to actively participate in learning and developing new skills and maintaining old skills to improve care for the child involved.[8,12,13] Studies have also revealed how self-efficacy can be used as a tool to predict long and short-term health-care behaviors, and by boosting it, we can enable patients and their families to play a bigger role in the management of their disease.[13,96,109-113] In most chronic diseases, there is an interplay between various behaviors required for self-management. For each individual, each of these behaviors will vary in terms of difficulty.[8,92,109,114] Health care providers are in a good position to intervene and influence overall self-efficacy for self-management. They hold positions of respect and are generally viewed as experts in the field. Encouragement by them can have a

great influence on patient's self-efficacy.[12,115] For example, the self-efficacy of men recovering from a myocardial infarction was greatly influenced after receiving counseling from a physician and nurse about their performance on a treadmill test.[12] Health education and promotion programs give patients and families encouragement and a means of adapting and changing behaviors. It improves confidence leading to more effective treatment.[8,12,13,91,92]

Ewart et al. reported that soon after a myocardial infarction, men whose self-efficacy was high or boosted by successful exercise testing, were more likely to perform better at everyday home activities such as walking, climbing stairs and running than those whose tests were unsuccessful. Explanation of their results by health professionals, further enhanced their self-efficacy and they were able to perform better at other tasks such as sexual intercourse and lifting which are dissimilar to the treadmill test.[12] Similar results were obtained from a study on patients with COPD. Those who were given specific training (cognitive, behavioural or combined training) and advice from health professionals had a significantly higher perceived self-efficacy for physical activities such as walking, as compared to the group who only received advice and no training. The groups who received behavioural training had a greater improvement in their self-efficacy than those who received only cognitive training.[115]

4.5. SELF-EFFICACY IN CYSTIC FIBROSIS

Bartholomew et al investigated whether self-efficacy expectations and outcome expectations would be able to predict self-management behaviours in caregivers and adolescent patients with CF.[8] They used a previously validated tool to develop and determine the psychometric characteristics of a measure of self-efficacy expectations for the self-management of CF. The caretaker self-efficacy tool had five sub-scales: Medical judgement and communication, coping, family communication, compliance and acceptance. The adolescent self-efficacy tool had four sub-scales: communication with health care team, acceptance and coping, medical judgement and communication, and compliance. Outcome expectations were measured using the Outcome Expectations Instrument for Cystic Fibrosis, which measures the expectations of positive outcomes from self-management activities on a five-point scale ranging from *'Not at all sure'* to *'Very sure.'* The Marlowe Crowne Social Desirability Scale was used to measure the extent of the effect of social desirability on

the self-report of self-efficacy. This consisted of 33 items with a true/false ranking. Self-management activities were assessed using the Self-Management Questionnaire for Cystic Fibrosis caretaker and adolescent versions, which have 50 self-reported items relating to CF care with a five-point scale ranging from *'1-Never used'* to *'5-Always used.'* Caretakers were interviewed to obtain socio-demographic data and the clinical health of the patients was assessed by using the National Institutes of Health (NIH) scoring system.[8] Families of 199 patients were recruited into the study. Patients were aged 18 years or younger. All 199 caretakers completed the questionnaires. In addition, 55 adolescents (13-18 year olds) completed the adolescent versions of the questionnaires.[8] Carers who had a higher self-efficacy and younger children to care for, reported more self-management behaviours. The three sub-scales of the caretaker self-efficacy measure which most significantly predicted self-management were 'medical judgement and communication,' 'family communication' and 'compliance.' For adolescents, those with high outcome expectations as well as high self-efficacy performed more self-management behaviours. Outcome expectation was a better predictor of self-management behaviours than self-efficacy alone. The 'medical judgement and communication,' 'communication with the health care team' and 'coping and acceptance of CF' sub-scales of the self-efficacy measure were also significant predictors of self-management, as was social desirability.[8] This study shows that behaviours such as communication with the family and the health care team and monitoring of treatment and disease severity have a significant effect on self-management behaviours. Putting emphasis on these aspects during patient and caregiver education may lead to better self-management of the disease. Having high self-efficacy and outcome expectations to carry out certain behaviours are also important factors in self-management. Once again, health education should take these factors into consideration, by persuading families that they are capable of carrying out certain health-care practices, and that these practices will eventually lead to positive results.[8]

Bartholomew et al's 1993 study and Parcel et al's 1994 study were both part of a larger health education interventional study on the same participants.[8,109] Parcel's aim was to identify the extent to which self-management behaviours were influenced by knowledge about CF, self-efficacy and outcome expectations through a cross sectional study.[109] Demographic information was collected during interviews with the

participants. The National Institutes of Health (NIH) scoring system was used to assess the clinical health status of the patient. The height and weight of the patients was obtained from their hospital records. The Self-Management Questionnaire for Cystic Fibrosis was used to measure self-management behaviours. The Coping/Problem-Solving Scale was designed for this study to assess how caregivers dealt with moderately stressful problems and situations relating to their child's CF as well as other non-CF related situations. Caregiver's self-efficacy and outcome expectations were measured using the same scales that were used in Bartholomew's study above. The caregivers completed a 53-item knowledge survey to assess their knowledge about CF. This was the only questionnaire that was completed in hospital to ensure that the knowledge was the participants' own and not from another source. The remaining questionnaires were completed at home by the participants and returned after completion.[109]

The overall mean scores indicated that caregivers reported a high rate of self-management behaviours, although monitoring skills were used less frequently than other self-management skills. Most participants had high outcome expectations and a high self-efficacy for dealing with medical regimens, family communication and acceptance of CF as a disease. Unfortunately the self-efficacy for coping abilities with the disease process was found to be quite low in the study group. A mean of 58% of items on the knowledge survey were answered correctly indicating that there is a big gap in the CF knowledge of carers in this population.[109]

Reports of gastrointestinal and respiratory monitoring behaviours were associated with reports gastrointestinal and respiratory treatment behaviours respectively. This study also found that poor pulmonary function could promote respiratory monitoring behaviours, which in turn influenced gastrointestinal monitoring behaviours. Caregivers with high self-efficacy also reported more frequent respiratory monitoring behaviours. Self-efficacy was found to be indirectly related to gastrointestinal monitoring through respiratory monitoring. Knowledge about CF was positively related to coping mechanisms and better growth of the child with CF. It was not associated with monitoring or treatment behaviours by caregivers. Outcome expectations were seen to be positively related to self-efficacy reports by the caregivers.[109]

This study found that self-efficacy was the most important factor in predicting 'self-management behaviour for monitoring and treating respiratory problems' in carers of children with CF. They also concluded that educational programmes for CF addressing self-efficacy are more likely to result in improved self-management of treatment, than those programmes, which solely aim to increase knowledge about CF. As seen above in Bartholomew et al's study, encouraging beliefs in caregivers that certain treatments will eventually have a positive effect on their child's health may lead to an improvement in their self-efficacy and self-management behaviours.[109]

A Norwegian study investigated the relationship between pulmonary function and perceived health status and global quality of life in adults suffering from CF. They also examined the effect of self-efficacy on these relationships.[116] This was a cross sectional, questionnaire based study. The questionnaire was administered to most participants during their outpatient clinic appointments. Some participants received it via post and returned it to the clinic shortly after. The questionnaire consisted of various sections and scales measuring the aspects that the investigators were interested in. The first part consisted of socio-demographic data, clinical information, CF related problems and patient satisfation with health and social services. The second part consisted of various scales. The Quality of Life Scale measured the individual's satisfaction with various aspects of his/her life. It is rated on a seven point scale ranging from '*Delighted'* to *'Terrible.'* Higher scores indicate better quality of life. Perceived health status was measured by the St George's Respiratory questionnaire which identifies patient symptoms, activities affected by shortness of breath and the impact of the disease on social and psychological functioning. Higher scores indicate poorer health status. The Generalised Self-Efficacy Scale was used to measure the perceived self-efficacy of the patients. A higher score indicates better self efficacy. The Forced Expiratory Volume in one second (FEV_1%) was used to give an objective measure of the participant's health status.[116] The questionnaire was completed by 86 patients. A significant relationship was found between pulmonary function and perceived health status. Pulmonary function was only related to global quality of life indirectly through percieved health status. Self-efficacy was found to be significantly related to both perceived health status and global quality of life, although there was no direct interaction between self-efficacy and pulmonary function. Those with higher self-efficacy reported better perceived health and and a better global

quality of life. This study found that self-efficacy is as important as pulmonary function in the perceived health status and global quality of life in a patient with CF. This should be taken into consideration by health care professionals when treating such patients.[116]

4.6. MEASURING SELF-EFFICACY

4.6.a. The General Self-Efficacy Scale

The General Self-Efficacy Scale (GSE-S) was developed by Jerusalem and Schwarzer in 1979 in German. It consisted of 20 items. In 1981 it was reduced to 10 items and adapted into 28 languages. The 10 items are scored on a four point Likert scale. A minimum score of 10 and maximum score of 40 can be awarded. The GSE-S has been previously validated and tested for reliability with a Cronbach's alpha value of 0.86.[117,118]

The General Self-Efficacy Scale:

- I can always manage to solve difficult problems if I try hard enough
- If someone opposes me, I can find the means and ways to get what I want
- I am certain that I can accomplish my goals
- I am confident that I could deal efficiently with unexpected events
- Thanks to my resourcefulness, I can handle unforeseen situations
- I can solve most problems if I invest the necessary effort
- I can remain calm when facing difficulties because I can rely on my coping abilities
- When I am confronted with a problem, I can find several solutions
- If I am in trouble, I can think of a good solution
- I can handle whatever comes my way

Response Format:

1= Not at all true 2= Hardly true 3= Moderately true 4= Exactly true[117,118]

4.6.b. Development of the cystic fibrosis self-efficacy questionnaire (CFSE-Q)

Measuring self-efficacy in chronic diseases can help us predict the health related behaviours of patients and caregivers. By using interventions, we can try and modify/improve their self-efficacy, thus giving them a sense of empowerment over their situation and improving care and overall quality of life for the patient and the family.[13,96,109-113]

For this study, the general self-efficacy scale (GSE-S) had been adapted to be used by caregivers of children with CF, creating the CFSE-Q. Four additional questions have been added to the original scale, giving a total of 14 questions. Participants can score a minimum of 14 and a maximum of 56. A higher score indicates a better self-efficacy.

Four additional items in the CFSE-Q:

1. I face problems on a daily basis
2. I do have the support I need to solve problems
3. I can only solve a problem if I expected it to happen
4. I never feel my views are fully appreciated

Three items on the CFSE-Q (items 2, 9 and 13) were scored in a reverse manner. Eg. If a participant gives themselves a four, the resulting score would be one due to the nature of the question:

- I face problems on a daily basis
- I can only solve a problem if I expected it to happen
- I never feel my views are fully appreciated

4.7. ASSESSING THE IMPACT OF ADMINISTERING THE 'CHALLENGES OF LIVING WITH CYSTIC FIBROSIS' QUESTIONNAIRE ON THE SELF-EFFICACY OF CARE-GIVERS OF CHILDREN WITH CYSTIC FIBROSIS

These studies show that self-efficacy plays an important role in the management of chronic diseases such as CF. This particular study explores the impact of an intervention tool on the self-efficacy of carers of children with CF. The following chapters will discuss the methodology and results from this study.

4.7.a. Aims

The aim of this randomised controlled pilot study is to investigate the effect of administering the 'Challenges of Living with Cystic Fibrosis' Questionnaire (CLCF-Q) to caregivers of children with CF during the annual review and the subsequent feedback process on their self-efficacy as measured by the 'Cystic Fibrosis Self-Efficacy' Questionnaire (CFSE-Q).

4.7.b. Hypothesis

H_1 There will be a difference in the change in CFSE-Q scores at the beginning and end of the study period between the intervention and control groups.

H_0 There will be no difference in the change in CFSE-Q scores at the beginning and end of the study period between the intervention and control groups.

4.7.c.Duties of the investigator

A literature review was done for the purpose of this study. Databases such as Pubmed were used to extract the relevant articles.

Recruitment for this study began in March 2010 by Miss L Patel who recruited 18 participants. In August 2010, I became the new study investigator. The study process carried on, new participants were recruited into the study and they joined the old participants in completing the study cycle. Data was collected, recorded and analysed using appropriate software.

CHAPTER TWO

METHODS

5.0. METHODS

5.1. THE ANNUAL REVIEW PROCESS AT ALDER HEY CHILDREN'S HOSPITAL

Alder Hey Children's Hospital, Liverpool, is a regional CF centre. The CF team consists of consultant specialist respiratory physicians, CF nurses, paediatric physiotherapists, clinical psychologists and dietitians. New patients and their families are personally introduced to the team who play an active role in their child's life. Once a year, patients have an annual review, which consists of blood tests, pulmonary function tests, sputum cultures, height and weight measurements, chest X-rays, abdominal ultrasounds and hearing tests. A few weeks later, their combined results from the annual review are discussed with them during a clinic appointment, where they will have a consultation with each of the professionals from the CF team. Patients are reviewed in the CF clinic every six to eight weeks to monitor progress, treatment and growth. To minimise cross infection with other CF patients, each patient and family is allocated to one room. All the professionals will see them in that room in turn. The clinics and annual reviews are also separated into *Pseudomonas* and non-*Pseudomonas* days, to minimise spread of infection with this organism. After each clinic, the team conducts a multi disciplinary team meeting where they discuss all the patients who attended that clinic. This allows everyone to familiarise themselves with the state of all the patients and develop efficient management strategies. When patients are acutely ill, they are admitted into the wards for more intensive treatment.

5.2. METHODOLOGY

This is a single centre randomised controlled trial undertaken by MPhil students and staff from the Institute of Translational medicine, University of Liverpool and staff from the Institute of Child Health, Alder Hey Children's NHS Trust.

5.2.a. Ethical approval

This study was approved by the North West Research Ethics Committee. Ethical approval was initially gained in March 2010 by Miss L Patel, the previous MPhil student (2009-2010). In August 2010, I was approved as the new chief investigator for the study.

5.2.b. Inclusion criteria

- Main parent or carer of a child with CF
- Child must have been diagnosed with CF over a year ago
- Child must be between 1-13 years of age
- Child must be a registered patient at Alder Hey Children's hospital, Liverpool
- Parents/carers must fully understand the information being given to them and be willing to participate and provide voluntary consent

5.2.c. Exclusion criteria

- Parents, carers or children with learning difficulties or co-existing physical or mental illnesses which would prevent them from successfully participating in the study
- Families with complex social problems involving child care

5.2.d. Randomisation

Participants were parents/carers of children with CF. They were randomly allocated to either the intervention or control group by an independent observer using sealed envelopes. Ten participants were randomised at a time using a set of 10 sealed envelopes out of which five were for the intervention group and five for the control group. An independent observer randomly selected envelopes and their sequence was recorded. During the annual review, participants were allocated to the next available group.

5.2.e. Participant recruitment and study process

The study was described to the carers during a clinic appointment. Carers were told what the study entailed, what was being measured and how it was going to be measured. They were informed about who was running the study and were given contact details in case they required more information. Printed information sheets and invitation letters were also given to them to take home (Appendix 9.1.a. & 9.1.b.) . This gave them time to reflect on the study. During their child's annual review, a study investigator approached them. The study was once again explained to them. Any questions were answered. If they agreed to participate, written consent was obtained (Appendix 9.2.a) .

The questionnaires were completed by the participants during the annual review and returned to the study investigator at the end of the day. This was considered 'Baseline.' Carers randomised to the intervention group filled out the CFSE-Q (Appendix 9.2.b.) and the CLCF-Q (Appendix 9.2.c.). The CLCF-Q is ten pages long and generally takes about 30 minutes to complete, but as it was filled out during the annual review, the process took longer as it was continuously being interrupted by clinical procedures and examinations. The CFSE-Q is a shorter questionnaire and takes two minutes to complete. Those randomised to the control group only filled out the CFSE-Q.

At the next clinic appointment, participants from the intervention group were asked to complete a 'Self feedback form for carers' based on how they felt they have scored in the CLCF-Q (Appendix 9.2.e.). The investigator, based on the participant's actual responses in the CLCF-Q, completed a similar form (Appendix 9.2.d.). The feedback forms have 13 statements relating to various questions in the CLCF-Q. Each statement in the investigator version had a column for a score, calculated from the participants' answers from the CLCF-Q. The scores were then ranked as being "false", "sometimes true" and "true", represented by colour-coded boxes, according to the traffic light system, and faces depicting different expressions (Table 4). Notes were also made from the answers in the CLCF-Q. The participant version did not have a column for scores. Participants used the same colour coded system to rank the various statements as "false", "sometimes true" and "true". The investigator and participant forms were compared and verbal feedback was given to those in the

intervention group by a study investigator. The scores were discussed and any notes made by the study investigator from the CLCF-Q were also mentioned and discussed. The participants were encouraged to discuss any problems with the relevant health care professionals in the CF team. A written copy of the feedback was sent out to the participants' home addresses together with a letter thanking them for their participation (Appendix 9.1.c.). Those in the control group did not receive structured feedback through the forms. The results of the annual review were discussed with all participants during the clinic session with the CF team. This signifies the completion of the first part of the trial.

DOMAIN	SUB-DOMAIN	ITEM ON CLCF	HIGHEST POSSIBLE SCORE	SCALE		
				TRUE ☺	😐	FALSE ☹
Family Lifestyle 'We work well as a family'	We share challenges	4. How does your family divide childcare relating to CF	5	4-5	3	1-2
	We work together	5d. How would you describe your general family lifestyle? (Work together-Work as individuals)	15	11-15	6-10	1-5
		6. How well do you think you are juggling the demands of CF with the needs of your family?				
		7. How well do you think your family as a whole handles the challenges of CF?				
CF Background 'My child's CF is well managed'		12. Over the last two weeks, has your child been? (Unwell-Well)	5	4-5	3	1-2
Child's Character 'My child is well behaved'		15. My child makes more demands on me than I expected	35	23-35	13-22	1-12
		17. My child sleeps throughout the night				
		19. My child makes friends easily				
		20. My child is easily upset by things generally				
		21. My child is very moody				
		23. My child is popular with his/her peers				
		24. My child reacts very strongly when something happens that s/he doesn't like				
Challenges of Family Life 'We face CF together as a family'	I have a lot of support	25. How supported do you feel by the following groups of people?	30	21-30	11-20	1-10
	We don't worry about infections	26. To reduce the risk of cross infection, the CF team advises that people with CF avoid contact with other people with CF. How much does this affect contact with other CF families?	5	4-5	3	1-2
	CF doesn't impact on our life too much	27. Caring for a child with CF can involve extra expense. How difficult is it for you to manage this?	15	11-15	6-10	1-5
		28. To what extent do you think CF has changed your work pattern?				
		29. How often have you had a disturbed night's sleep in the past 2 weeks?				
Hopes and Worries 'My hopes for everyday life are bigger than my worries about everyday life'	I am hopeful about our day to day life	32. It is difficult to predict what the future holds in relation to CF. To what extent does this uncertainty affect your family's approach to life?	30	21-30	11-20	1-10
		33. How much does the responsibility of looking after a child with CF affect you?				
		34. How much is your child's growth a worry for you? A. Height				
		B. Weight				
		35. To what extent are you worried that your child might become infected with pseudomonas when s/he is outside the home, e.g. at friends' houses, at school?				
		36. How worried are you about a change in your child's lung function?				

	I don't worry about everyday life	32. It is difficult to predict what the future holds in relation to CF. To what extent does this uncertainty affect your family's approach to life?	30	21-30	11-20	1-10
		33. How much does the responsibility of looking after a child with CF affect you?				
		34. How much is your child's growth a worry for you? A. Height				
		34. How much is your child's growth a worry for you? B. Weight				
		35. To what extent are you worried that your child might become infected with pseudomonas when s/he is outside the home, e.g. at friends' houses, at school?				
		36. How worried are you about a change in your child's lung function?				
Hopes and Worries 'My hopes for the future are bigger than my worries for the future'	I have a lot of hope for the future	31. Some say that living with CF is like a balance of hope and worry: What hopes do you have for your child?	30	21-30	11-20	1-10
	I don't worry about the future	31. Some say that living with CF is like a balance of hope and worry: What hopes do you have for your child?	30	21-30	11-20	1-10
CF Routines 'Managing my child's CF is easy	We have a good routine	39. How easy was it to establish the CF care routine after your child was diagnosed?	35	23-35	13-22	1-12
		40. How much of a problem is it to manage the daily routines for CF now? a. Mealtimes- getting him/her to eat enough				
		40. How much of a problem is it to manage the daily routines for CF now? b. Digestion- tummy problems (wind, pain, diarrhoea)				
		40. How much of a problem is it to manage the daily routines for CF now? c. Taking enzymes/creon				
		40. How much of a problem is it to manage the daily routines for CF now? d. Taking vitamins/oral antibiotics				
		40. How much of a problem is it to manage the daily routines for CF now? e. Doing physiotherapy				
		40. How much of a problem is it to manage the daily routines for CF now? f. Doing nebulised medications				
	I never feel overwhelmed	41. With all the things that need to be done, it may be overwhelming at times. How true has this been for you over the last 2 weeks?	5	4-5	3	1-2
Community Support 'I have a good relationship wit h the CF team in the community'		43. What quality of relationship do you have with your local GP/surgery?	20	14-20	7-13	1-6
		44. How helpful is your local pharmacist?				
		45. What sort of relationship do you have with you child's minder/nursery/school?				
		46.How comfortable are you with how your child's minder/nursery, or school gives medications to your child?				
CF Clinic and Pharmacy Visits 'I find it easy getting my child's medication'		53. How consistent are the messages you get from different members of the CF team?	10	8-10	4-7	1-3
		54. How much information would you like to have from the CF team about your child's condition or treatment?				

Table 4. Feedback sheet scoring system

At the third clinic appointment, all participants were given the second CFSE-Q to complete. This was considered to be the 'End-point.'

Figure 4 summarises this information in the form of a flow chart.

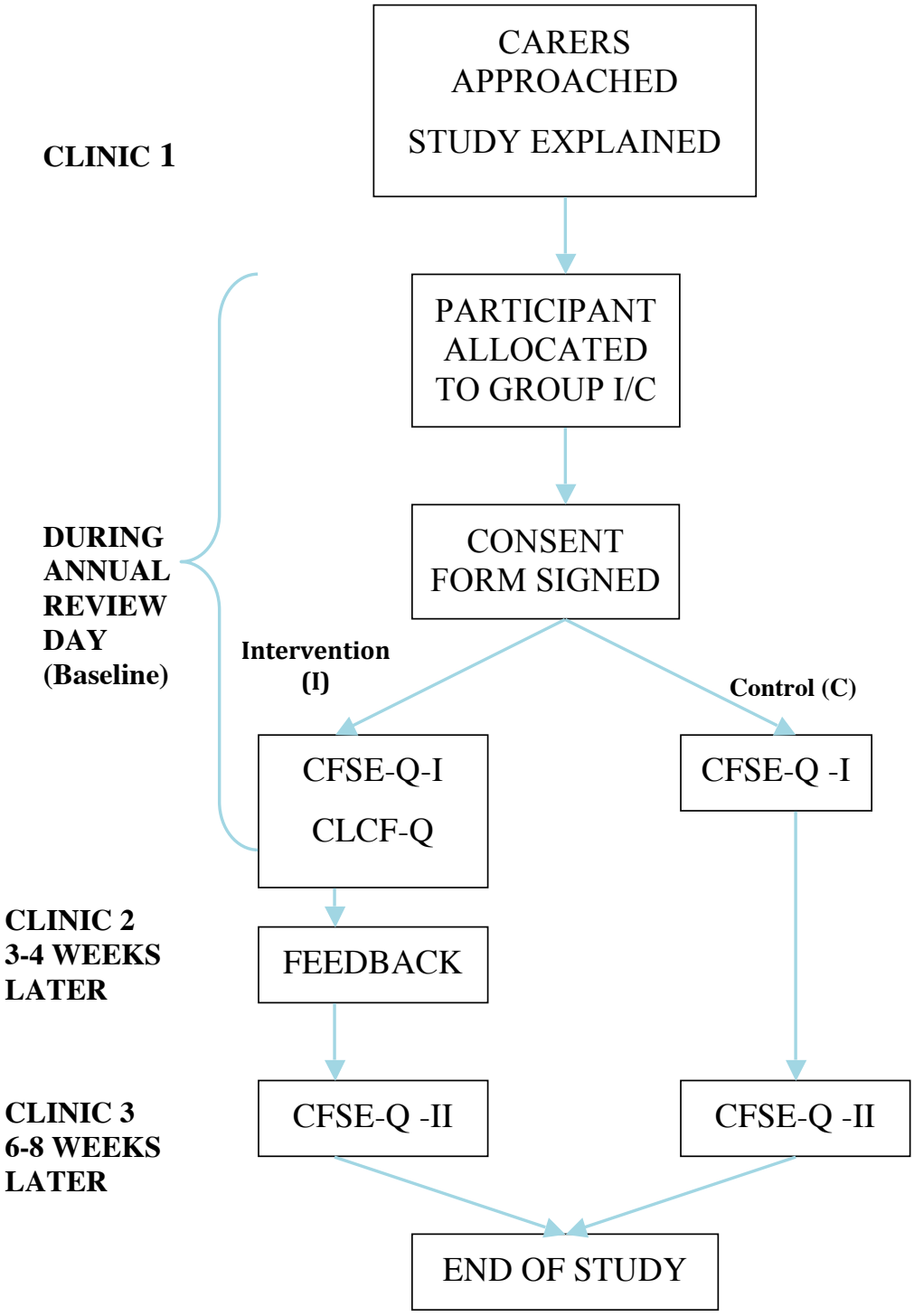


FIG. 4. SUMMARY OF RECRUITMENT AND PARTICIPATION EVENTS

5.2.f. Confidentiality

All questionnaires were collected and kept secure to protect the identity of the patients and carers. Only the study investigators had access to the data.

5.2.g. Statistical Analysis

The results were analysed using SPSS PASW Statistics 18. The means and standard deviations (SD) were used to analyse data that were normally distributed and the median and interquartile ranges (IQR) for skewed data. The paired student's t-test was used to analyse data between two time points. Correlation studies and r values were used to identify relationships between two variables.

CHAPTER THREE

RESULTS

6.0. RESULTS

6.1. DEMOGRAPHICS

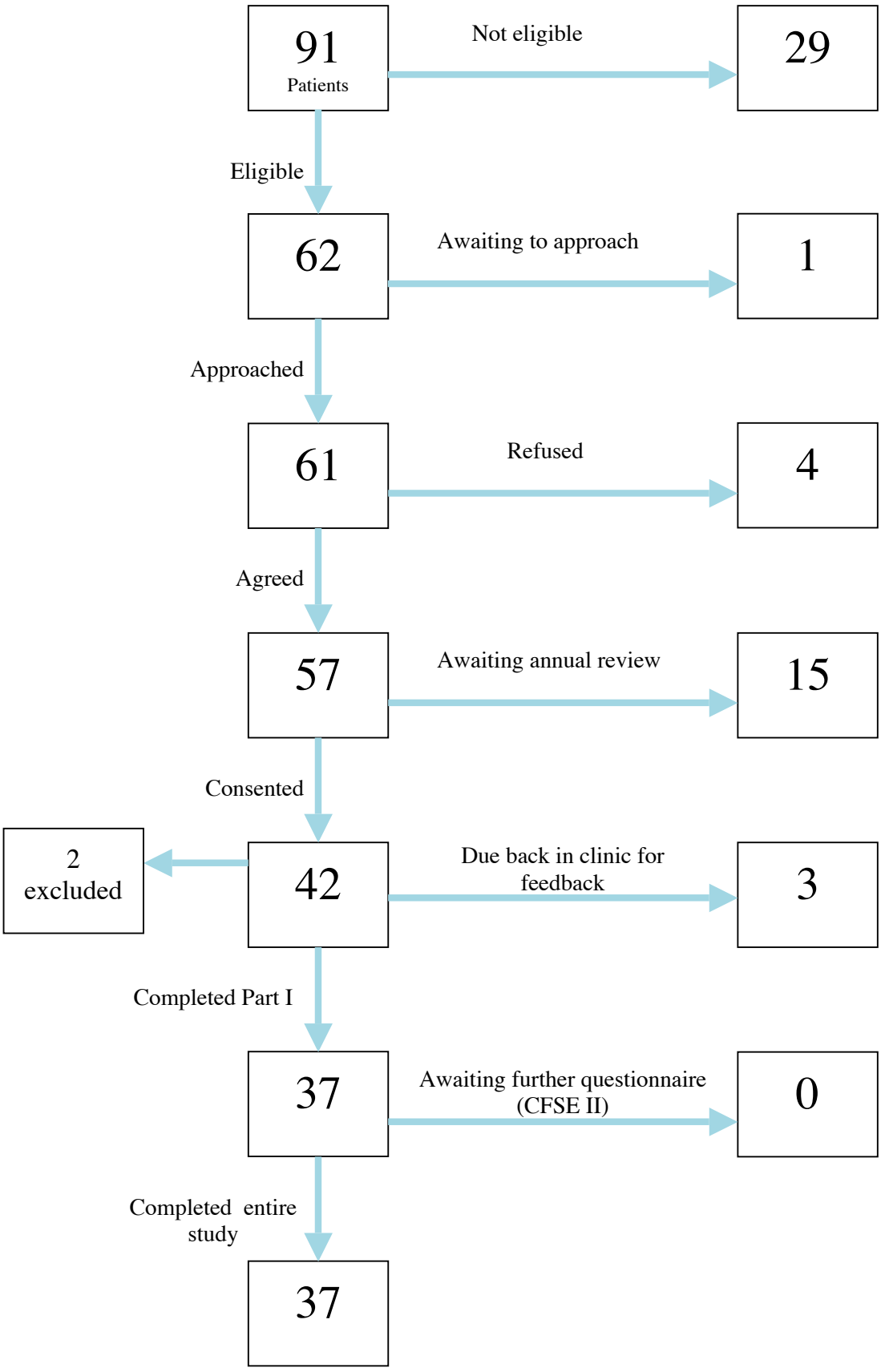
Out of the 91 CF patient and carer groups registered to have their annual review at Alder Hey Children's Hospital, Liverpool, 62 were eligible to participate in this study. Twenty-nine carers were not eligible as they had children who were either too young (n=3), too old (n=20), had a sibling in the trial (n=3) or had children in care (n=3). During clinic appointments, 61 eligible carers were approached; four refused to participate leaving 57 carers who agreed. Out of these, signed consent was gained from 42 carers during their child's annual review, where they also completed the relevant questionnaires. From these, two participants were excluded from final analysis; the first, as the CFSE-Q was completed by different carers at both time points, and the second, as the carer did not complete the first CFSE-Q during the annual review and refused to participate any further. As of 19th July 2011, 37 participants had completed the study (Figure 5).

There were 21 participants randomized to the control group and 19 to the intervention group (Table 5).

		INTERVENTION	CONTROL	TOTAL
Relationship of carer to child	Mother	17	16	33
	Father	1	5	6
	Other	1 (Grandmother)	0	1
Sex of child	Girl	9	10	19
	Boy	10	11	21
Age groups of children	1-4	5	3	8
	5-8	7	5	12
	9-13	7	13	20

Table 5. Demographics of the study population

Figure 5. Participant numbers (as of 19th July 2011)



6.2. CFSE-Q RESULTS

6.2.a. The CFSE-Q

The following data is only from the 37 individuals who have completed the entire cycle of the study (Intervention group n=17, Control group n=20). The CFSE-Q was completed well by the participants. Most questionnaires were fully completed. Only three responses were missing from the whole set of questionnaires. Participants understood the instructions and the questions well and did not have any difficulties completing the questionnaire. Scores were normally distributed (Figure 6). No floor or ceiling effects were observed as none of the participants achieved the lowest or highest possible scores. Repeatability was not assessed in the present study.

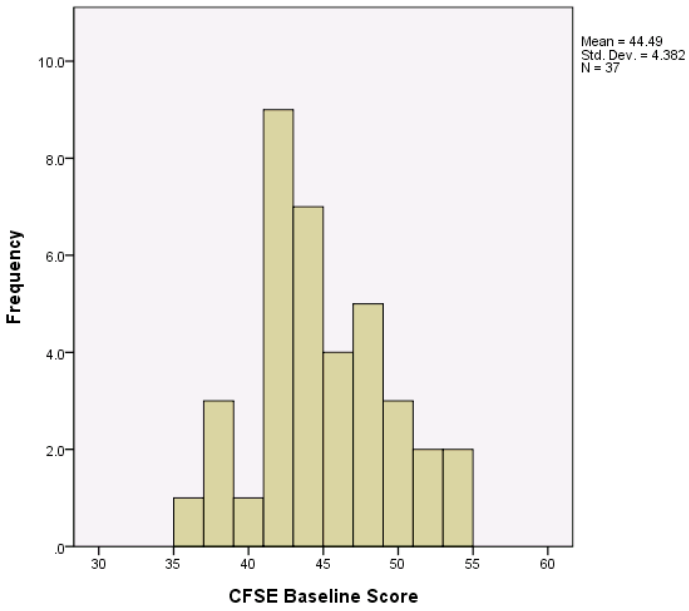


Figure 6. CFSE-Q baseline scores

6.2.b. Participant analysis

Total scores for participants ranged from 36-53 in the intervention group and 37-51 in the control group at baseline. End-point scores ranged from 38-54 for the intervention group and 31-53 for the control group. Self-efficacy scores at baseline were similar for both groups (Figure 7 & 8).

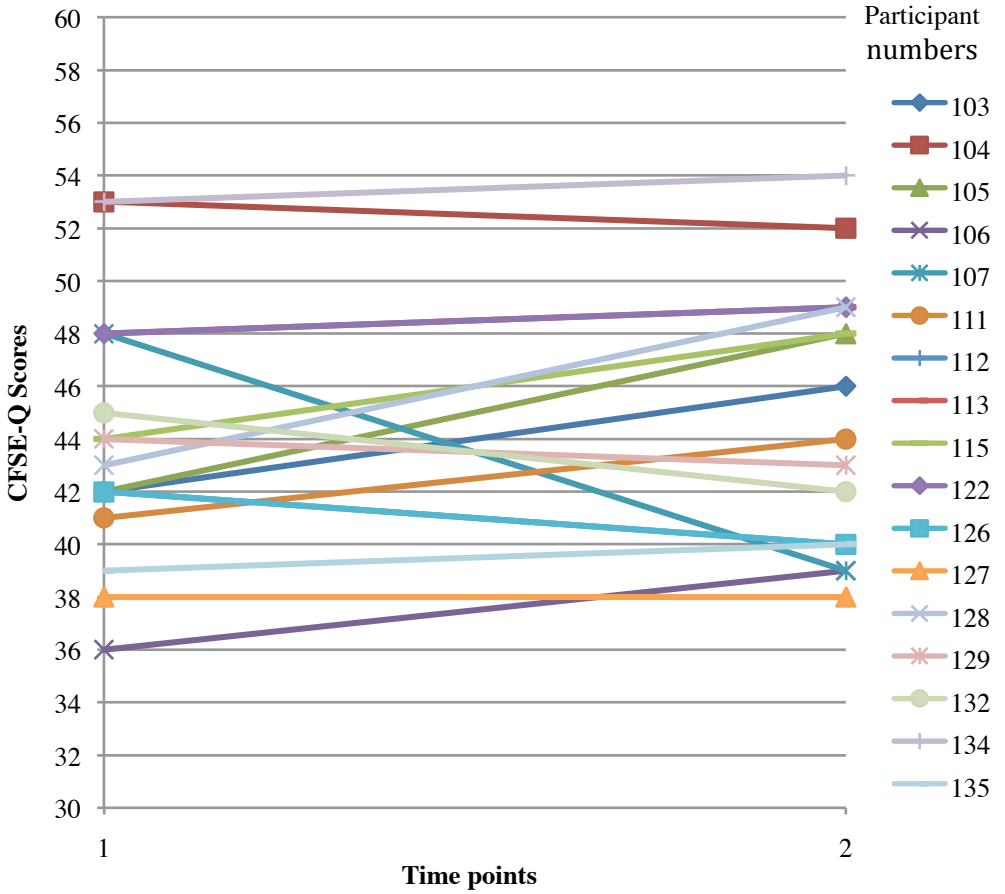


Figure 7. CFSE-Q Scores- Intervention group

Chart representing individual participant self-efficacy scores in the intervention group at baseline (time point 1) and at end-point (time point 2)

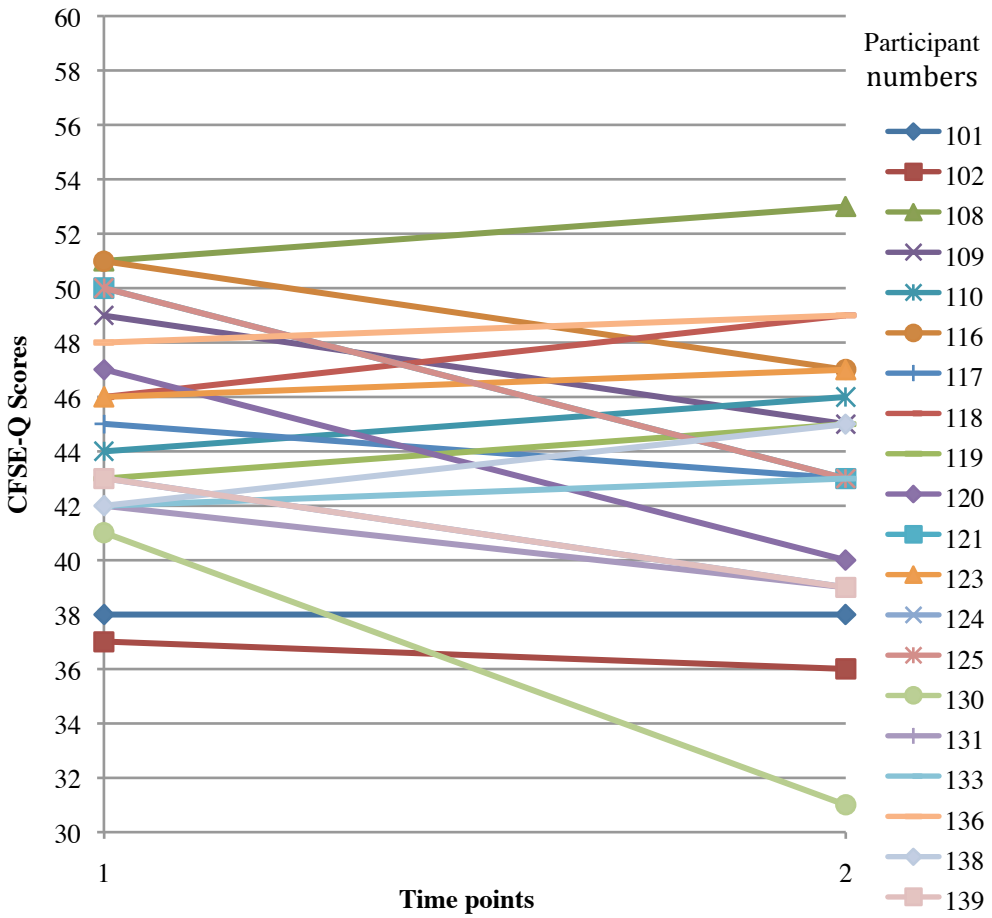


Figure 8. CFSE-Q Scores- Control group

Chart representing individual participant self-efficacy scores in the control group at baseline (time point 1) and at end-point (time point 2)

The self-efficacy scores of the intervention group at the end of the study had increased more than those of the control group as compared to what they were at baseline. The total change in self-efficacy score for all the participants of the intervention group was 12 and -30 for the control group. Median (IQR) change in self-efficacy score 1.0 (-1.5 to 3.5) versus -0.5 (-4.0 to 2.0); U= 120.0, p=0.1). This was not statistically significant, thus we can accept the null hypothesis that there is no difference between the change in self-efficacy scores at baseline and end-point between the two groups.

6.2.c. Group Analysis

6.2.c.i Intervention group

The mean (SD) CFSE scores for the intervention group were 44.0 (4.8) at baseline and 44.7 (5.0) at the end point. A paired student's t-test revealed that the scores remained stable between time points ($p= 0.4$). Self-efficacy was maintained throughout the trial (Figure 7).

6.2.c.ii Control group

The mean (SD) CFSE scores for the control group were 44.9 (4.1) at the start and 43.0 (5.1) at the end. The scores differed significantly between the two time points ($p< 0.05$), with a decline in self-efficacy scores at the end of the trial (Figure 8).

6.2.d. Item analysis

The majority of the CFSE-Q scores for the items were three or four, indicating a relatively high level of self-efficacy amongst the study population. Total scores for items at baseline ranged from 88-127. Item 2 (I face problems on a daily basis) had the lowest overall score. Item 7 (I do have the support I need to solve problems) had the highest overall score (Table 6A). Total CFSE-Q scores at end point ranged from 85 to 130, with items 2 and 7 having the lowest and highest scores respectively. (Table 6B)

COMBINED CFSE-Q BASELINE SCORES STATEMENT		SCORE Percentage (N=37)				Total score
		1 Not at all true	2 Hardly true	3 Moderately true	4 Exactly true	
1	I can always manage to solve difficult problems if I try hard enough	0% (0)	3% (1)	49% (18)	46% (17)	124
2	I face problems on a daily basis	16% (6)	41% (15)	32% (12)	11% (4)	88
3	If someone opposes me, I can find the means and ways to get what I want	5% (2)	16% (6)	57% (21)	22% (8)	109
4	It is easy for me to stick to my aims and accomplish my goals	5% (2)	30% (11)	41% (15)	24% (9)	121
5	I am confident that I could deal efficiently with unexpected events	0% (0)	3% (1)	59% (22)	38% (14)	124
6	Thanks to my resourcefulness, I know how to handle unforeseen situations	3% (1)	3% (1)	59% (22)	35% (13)	121
7	I do have the support I need to solve problems	0% (0)	11% (4)	35% (13)	54% (20)	127
8	I can solve most problems if I invest the necessary effort	0% (0)	3% (1)	57% (21)	41% (15)	125
9	I can only solve a problem if I expected it to happen	11% (4)	14% (5)	46% (17)	30% (11)	109
10	I can remain calm when facing difficulties because I can rely on my coping abilities	3% (1)	11% (4)	54% (20)	32% (12)	117
11	When I am confronted with a problem, I can usually find several solutions	3% (1)	5% (2)	59% (22)	32% (12)	119
12	If I am in trouble, I can usually think of a solution	0% (0)	0% (0)	62% (23)	38% (14)	125
13	I never feel my views are fully appreciated	5% (2)	16% (6)	46% (17)	32% (12)	113
14	I can usually handle whatever comes my way	0% (0)	0% (0)	65% (24)	35% (13)	124

Table 6A. Combined CFSE-Q Scores- Baseline

COMBINED CFSE-Q ENDPOINT SCORES STATEMENT		SCORE Percentage (N=37)				Total score
		1 Not at all true	2 Hardly true	3 Moderately true	4 Exactly true	
1	I can always manage to solve difficult problems if I try hard enough	3% (1)	0% (0)	51% (19)	46% (17)	125
2	I face problems on a daily basis	27% (10)	27% (10)	41% (15)	5% (2)	85
3	If someone opposes me, I can find the means and ways to get what I want	8% (3)	11% (4)	65% (24)	14% (5)	106
4	It is easy for me to stick to my aims and accomplish my goals	0% (0)	14% (5)	59% (22)	27% (10)	116
5	I am confident that I could deal efficiently with unexpected events	3% (1)	0% (0)	54% (20)	41% (15)	121
6	Thanks to my resourcefulness, I know how to handle unforeseen situations	0% (0)	3% (1)	68% (25)	30% (11)	121
7	I do have the support I need to solve problems	3% (1)	5% (2)	30% (11)	54% (23)	130
8	I can solve most problems if I invest the necessary effort	0% (0)	3% (1)	49% (18)	49% (18)	128
9	I can only solve a problem if I expected it to happen	11% (4)	27% (10)	32% (12)	24% (9)	97
10	I can remain calm when facing difficulties because I can rely on my coping abilities	5% (2)	11% (4)	35% (13)	49% (18)	121
11	When I am confronted with a problem, I can usually find several solutions	3% (1)	14% (5)	57% (21)	27% (10)	113
12	If I am in trouble, I can usually think of a solution	0% (0)	3(1)	54% (20)	43% (16)	125
13	I never feel my views are fully appreciated	5% (2)	16% (6)	49% (18)	32% (12)	112
14	I can usually handle whatever comes my way	5% (2)	3% (1)	54% (20)	38% (14)	120

Table 6B. Combined CFSE-Q Scores- End point

The total scores for each item and differences between time points for each group were calculated (Table 7). As the intervention group had only 17 participants who had completed the study, compared to 20 in the control group, their total scores were less than those in the control group for all the questions. Six items (Statements 1, 3, 5, 6, 9 and 11) in the intervention group had lower scores at the end of the study than at baseline. Two items (Statements 4,12) had the same score at both time points and the remaining had higher scores at the end of the study. Only two items (Statements 1 and 6) in the control group had a higher score at the end of the study as compared to the beginning. Ten items (Statements 2, 3, 4, 7, 8, 9, 10, 11, 13 and 14) had lower scores at the end of the study and items 5 and 12 had the same score at both time points. The total difference for all items in the intervention group was 12 and -38 for the control group. The mean difference for the intervention group was 0.86 and -2.71 for the control group. There was no significant difference between the two groups ($p= 0.5$).

	STATEMENT	GROUP (Intervention=I ; Control=C)	TOTAL SCORES		DIFFERENCE
			BASELINE	END POINT	
1	I can always manage to solve difficult problems if I try hard enough	I	62	59	-3
		C	62	66	4
2	I face problems on a daily basis	I	39	43	4
		C	49	42	-7
3	If someone opposes me, I can find the means and ways to get what I want	I	47	45	-2
		C	62	61	-1
4	It is easy for me to stick to my aims and accomplish my goals	I	53	53	0
		C	68	63	-5
5	I am confident that I could deal efficiently with unexpected events	I	57	54	-3
		C	67	67	0
6	Thanks to my resourcefulness, I know how to handle unforeseen situations	I	57	55	-2
		C	64	66	2
7	I do have the support I need to solve problems	I	56	63	7
		C	71	67	-4
8	I can solve most problems if I invest the necessary effort	I	56	60	4
		C	69	68	-1
9	I can only solve a problem if I expected it to happen	I	48	47	-1
		C	61	50	-11
10	I can remain calm when facing difficulties because I can rely on my coping abilities	I	52	57	5
		C	65	64	-1
11	When I am confronted with a problem, I can usually find several solutions	I	54	53	-1
		C	65	60	-5
12	If I am in trouble, I can usually think of a solution	I	58	58	0
		C	67	67	0
13	I never feel my views are fully appreciated	I	53	54	1
		C	60	58	-2
14	I can usually handle whatever comes my way	I	56	59	3
		C	68	61	-7

Table 7. Total CFSE-Q scores and differences

6.3. THE CLCF-Q

6.3.a. Completing the CLCF-Q during the annual review

Out of 19 participants in the intervention group, 17 had completed the whole study. Not all participants completed all items in the CLCF-Q. Therefore, some items cannot be accounted for 100% of the participants. Participants were given the questionnaire during their child's annual review. They carried it around with them and completed it along the way, together with the first CFSE-Q. As they were consulting various specialists throughout the annual review, completing the questionnaire usually took the whole morning. When asked for feedback, many complained that the CLCF-Q is too long and complicated. Some felt that they were too stressed during the annual review to pay enough attention to their answers and would have preferred if they could take the questionnaire home and complete it there.

6.3.b. Results from the CLCF-Q (N=17)

The CLCF-Q has managed to identify quite a few of the burdens faced by caregivers and families of children with CF. Most caregivers were living together with their spouse/partner. For approximately 50% of families, there was equal role division within the family regarding to CF childcare. Most caregivers had no difficulty to marginal difficulty with balancing CF and family demands and majority of families handled the challenges of CF with varying degrees of ease. Over 75% of children from this group were relatively well in the two weeks preceding the annual review, despite the fact that 82% had a history of previous hospital admissions usually for IV antibiotics. Only four children had hospital admissions in the three months preceding the annual review. While in hospital the biggest source of stress for carers was disruptions to family life and the least stressful was communication with healthcare professional.

The majority of the caregivers did not find their child to be very challenging. Most children were moderately demanding, very determined, friendly and popular with peers and most settled down with new routines easily, which is an important aspect in CF care as treatment regimes can change frequently.

The main sources of support for the families were the CF team in the hospital and friends and family members. Caregivers were satisfied with the information and care

they were getting from the CF team. The GP and community pharmacy were moderately supportive.

One of the most difficult challenges to family life was the effect CF had on the caregivers' occupational lives. 71% of caregivers reported that CF had changed their work pattern quite a bit; most had to give up work or felt unable to carry on working full time. CF also caused quite a few caregivers to feel stressed and overwhelmed. Despite this most caregivers were quite hopeful and had few worries about their child's futures. Regarding daily life, most carers had moderate hopes and worries.

Oral medications and pancreatic enzymes required the least amount of effort from carers and children. Physiotherapy was found to be the most time consuming and difficult treatment for both parties. Despite this, most families reported very few problems managing daily routines for CF.

The following sections give more details and frequencies of each individual subsection of the CLCF-Q.

6.3.b.i. Family Lifestyle

The family lifestyle section consisted of seven items relating to family demographics, roles within the family and general family lifestyle (Table 8). Only 18% (n=3) of the intervention group were lone caregivers, the remaining 82% (n=14) were living with a spouse or a partner. The majority of the families were caring for two children (71%, n=12) and only had one child with CF (82%, n=14). Three families were looking after both of their children with CF. Over 50% (n=9) of participants reported that childcare relating to CF was equally divided between the family, 47% (n=8) felt that they were doing most or all of the work by themselves, 41% (n=7) felt that their general family lifestyle was more relaxed than stressed, 71% (n=12) lead quite a busy lifestyle, 59% (n=10) reported an organised family lifestyle and 71% (n=12) had quite fixed routines within the family and felt that the members of the family generally work together. The majority of participants (65%, n=11) felt that their families were quite chatty and 71% (n=12) reported a relatively sporty/active family lifestyle.

Only one participant (6%) felt that they had great difficulty juggling the demands of CF with the needs of their family. The remaining 16 participants (94%) reported no

difficulty to marginal difficulty with this. Most caregivers (71%, n=12) felt that their families seem to handle the challenges of CF with varying degrees of ease (Table 8).

ITEM	SCALE	PERCENTAGE (N=17)
1. Are you?	A lone caregiver	18% (3)
	Living with spouse or partner	82% (14)
	A lone caregiver living with family	0% (0)
2. How many children do you care for?	1	12% (2)
	2	71% (12)
	3	12% (2)
	4	0% (0)
	5	6% (1)
3. How many children with CF do you have living with you?	1	82% (14)
	2	18% (3)
4. How does your family divide childcare relating to CF?	1 (I do it all)	24% (4)
	2	24% (4)
	3 (Equal shares)	53% (9)
	4	0% (0)
	5 (My partner/others do it all)	0% (0)
5. How would you describe your general family lifestyle?	1 (Relaxed)	18% (3)
	2	24% (4)
	3	41% (7)
	4	6% (1)
	5 (Stressed out)	6% (1)
	1 (Disorganised)	0% (0)
	2	0% (0)
	3	35% (6)
	4	53% (9)
	5 (Organised)	6% (1)
	1 (Busy)	35% (6)
	2	35% (6)
	3	29% (5)
	4	0% (0)
	5 (Laid Back)	0% (0)
	1 (No fixed routines)	0% (0)
	2	6% (1)
	3	18% (3)
	4	53% (9)
	5 (Fixed routines)	18% (3)
	1 (Chatty)	29% (5)
	2	35% (6)
	3	24% (4)
	4	0% (0)

	5 (Quiet)	6% (1)
	1 (Sporty/active)	24% (4)
	2	47% (8)
	3	18% (3)
	4	6% (1)
	5 (Not sporty/active)	6% (1)
	1 (Work together)	41% (7)
	2	29% (5)
	3	18%(3)
	4	6% (1)
	5 (Work as individuals)	6% (1)
6. How well do you think you are juggling the demands of CF with the needs of your family?	1 (No difficulty)	24% (4)
	2 (A little)	29% (5)
	3 (Marginal)	41% (7)
	4 (Definite)	0% (0)
	5 (Great difficulty)	6% (1)
7. How well do you think your family as a whole handles the challenges of CF?	1 (Very easily)	6% (1)
	2 (Easily)	29% (5)
	3 (Marginally easily)	35% (6)
	4 (Some difficulty)	29% (5)
	5 (Great difficulty)	0% (0)

Table 8. Responses to questions on family lifestyle

6.3.b.ii CF Background

This section consisted of six questions relating to the current and previous health of the patient. All 17 children in the intervention group required enzymes in their food due to pancreatic insufficiency. At the time of the annual review, none of the children had been diagnosed with CF-related diabetes.

Only three participants reported their child's FEV_1% predicted/ blow scores (89%, 67%, 75%). Some children (29%, n= 5) were still under five years old at the time of their annual review and therefore were not yet capable of producing reliable results. Ten patients (59%) had grown various organisms on their cough swabs/ sputum samples in the three months preceding the annual review. There was one (10%) report of *Aspergillus*, six (60%) of *Pseudomonas* and three (30%) of MRSA. 76% (n=13) of children were well or mostly well for the two weeks before their annual review. The remaining 24% (n=4) were a mixture of well and unwell. Fourteen children (82%) had a history of hospital admissions in the past. Most (93%, n=13) were admitted to receive IV antibiotics. Other reasons for admission were: portacath fitted (36%, n=5), gastrostomy tube fitted (7%, n=1), Oxygen (14%, n=2), portacath removed (7%, n=1) and removal of adenoids (7%, n=1).

6.3.b.iii Child's Character

The child's character was assessed through a set of 11 questions identifying various aspects of the patient's personality. The children in this population were not found to be very challenging for their caregivers (Table 9). Most carers found that their children were moderately demanding (29% n=5), very determined (59% n=10), quite friendly (59% n=10), and popular with their peers (53% n=9). Only 35% (n=6) felt that their child was so active that it exhausted them. The remainder felt neutral or disagreed with this statement (statement 22). Children were not found to be very moody or very easily upset and the majority of children settle down with new routines relatively easily (53% n=9). Children falling asleep (65% n=11) and staying asleep at night (59% n=10) was not problematic for most caregivers.

ITEM	PERCENTAGE (N=17)				
	1 Strongly agree	2 Agree	3 Neutral	4 Disagree	5 Strongly disagree
14. My child is very determined; when s/he wants to do something s/he usually keeps trying until s/he succeeds	59% (10)	29% (5)	12% (2)	0% (0)	0% (0)
15. My child makes more demands on me than I expected	12% (2)	24% (4)	29% (5)	29% (5)	6% (1)
16. My child goes to bed easily	24% (4)	41% (7)	24% (4)	6% (1)	6% (1)
17. My child sleeps throughout the night	41% (7)	18% (3)	24% (4)	18% (3)	0% (0)
18. It takes a long time for my child to settle with new routines	6% (1)	12% (2)	29% (5)	47% (8)	6% (1)
19. My child makes friends easily	29% (5)	59% (10)	6% (1)	6% (1)	0% (0)
20. My child is easily upset by things generally	0% (0)	24% (4)	24% (4)	41% (7)	12% (2)
21. My child is very moody	0% (0)	6% (1)	35% (6)	35% (6)	24% (4)
22. My child is so active it exhausts me	6% (1)	29% (5)	18% (3)	41% (7)	6% (1)
23. My child is popular with his/her peers	35% (6)	53% (9)	12% (2)	0% (0)	0% (0)
24. My child reacts strongly when something happens that s/he doesn't like	18% (3)	41% (7)	24% (4)	18% (3)	0% (0)

Table 9. Parent/caregiver perception of child's character

6.3.b.iv. Challenges to Family Life

Six questions investigated the sources of support to families and some of the problems faced by families as a result of CF.

The majority (94%, n=16) of participants felt very supported by the CF team at the hospital. The pharmacy (88%, n=15) was also quite supportive for most caregivers. The GP, on the other hand was very supportive for only 53% (n= 9) of participants (Table 10A). On further inquiry during the feedback process, some reasons for this were:

- The GP was not involved directly in the management of CF and only provided the prescriptions for medications, if needed
- The GP did not seem well informed about CF
- There was hardly any contact with the GP

Family and friends seem to be an important source of support for families with children with CF. Most (65%, n=11) felt very supported by their family and 59% (n=10) had good support from their friends. The group that was least supportive to these families' were parents of other children with CF. Some of the reasons for this were:

- They didn't personally know other families with CF
- For the sake of infection control some families tried to keep away from others with CF (18%, n=3) (Table 10B).

25. How supported do you feel by the following groups of people?	**PERCENTAGE (N=17)**			
	1 Very supported	**2**	**3**	**4 Not at all supported**
Family members	65% (11)	24% (4)	12% (2)	0% (0)
Friend	59% (10)	85% (5)	12% (2)	0% (0)
Another parent whose child has CF	6% (1)	24% (4)	12% (2)	29% (5)
CF team	94% (16)	6% (1)	0% (0)	0% (0)
GP	53% (9)	29% (5)	12% (2)	6% (1)
Pharmacy	88% (15)	12% (2)	0% (0)	0% (0)

Table 10A. Sources of support for families with a child with CF

The extra expenses involved with managing CF did not seem to cause any difficulty with 35% (n=6) of carers, although 12% (n=2) did feel that these expenses were very hard to deal with. CF also managed to change the work pattern of many of the carers (Table 10B). CF had a great impact on the work patterns of 41% (n=7) of participants and 29% (n=5) felt that it had some effect on their work:

- 'All my attention/energy goes on my child'
- 'Changed to part time from full time'
- 'Part time, annual leave used for appointments and illness. Employers not helpful or colleagues when off with son'
- 'Felt couldn't work in his early years'
- 'I am a pensioner'
- 'Unable to take on further work commitments. Need to come home a lot during working hours'
- 'Gave up work to look after both boys'
- 'Off work with stress for 6 months'
- 'Reduced work load'

ITEM	SCALE	PERCENTAGE (N=17)
26. To reduce the risk of cross infection, the CF team advises that people with CF avoid contact with other people with CF. How much does this affect contact with other CF families?	1 (A great deal)	18% (3)
	2 (Some)	6% (1)
	3 (Moderate)	12% (2)
	4 (A little)	12% (2)
	5 (Not at all)	53% (9)
27. Caring for a child with CF can involve extra expense. How difficult is it for you to manage this?	1 (Very difficult)	12% (2)
	2	6% (1)
	3 (Moderately)	29% (5)
	4	18% (3)
	5 (Not at all difficult)	35% (6)
28. To what extent do you think CF has changed your work pattern?	1 (A great deal)	41% (7)
	2 (Some)	29% (5)
	3 (Moderate)	0% (0)
	4 (A little)	12% (2)
	5 (Not at all)	12% (2)
29. How often have you had a disturbed night's sleep in the past 2 weeks?	1 (Every night)	18% (3)
	2 (Frequent)	6% (1)
	3 (Some)	12% (2)
	4 (Few)	29% (5)
	5 (No nights)	35% (6)

Table 10B. Challenges faced by families

The carers were asked how they know when they need a break:

- 'Feel stressed and tired'
- 'Moody, loose temper easily, very tired'
- 'I may feel stressed and tearful for nothing at all'
- 'When we loose sight of ourselves and are overwhelmed'
- 'I get stressed out'
- 'Don't get a break'
- 'I don't. Just keep going'
- 'Take routine breaks'
- 'Tired'

6.3.b.v. Hopes and Worries

Daily and future hopes and worries were assessed through eight questions. The majority of the caregivers seemed hopeful regarding their child's future. From the feedback sheets, 13 (76%) caregivers scored highly on their hopes for their child's future. The same proportion didn't have many worries about their child's future either. These two sections of the feedback sheet were based on the answers to Question 31 (Table 11A).

31. Some say that living with CF is like a balance of hope and worry: What hopes do you have for your child?	**PERCENTAGE (N=17)**			
	1 Very hopeful	**2**	**3**	**4 Not hopeful**
S/he will adjust well to secondary school	65% (11)	24% (4)	12% (2)	0% (0)
S/he will go on to higher education	71% (12)	18% (3)	12% (2)	0% (0)
S/he will have a job	65% (11)	24% (4)	6% (1)	6% (1)
S/he will have a family of his/her own	35% (6)	29% (5)	24% (4)	12% (2)
S/he will continue to be as well as s/he is now	47% (8)	18% (3)	24% (4)	12% (2)
There will be an advance in science that will help my child	41% (7)	35% (6)	18% (3)	6% (1)

Table 11A. Hopes and worries for the future

When asked how the uncertainty of the future with CF affected the family's approach to life, some of the carers responded as below:

- 'We worry how his health will be'
- 'Worry about life expectancy'
- 'His CF/Autism combined mean we have to plan for the future more than for other children'
- 'Find it hard to live a normal life'
- 'Anxiety for me affects the rest of the family'
- 'Nothing apparent to F, but I question whether she will ever be able to lead a normal life i.e. job and family'

Regarding day-to-day life, most carers had 'moderate' hopes and worries (Table 11B). Carer's main worries were:

- ‘His chest’
- ‘That it will get worse and affect her breathing, preventing her from doing her activities’
- ‘The unknown’
- ‘Staying well’
- ‘M’s lung function has decreased slightly’
- ‘Weight’
- ‘Lung damages that’s irreversible’
- ‘Him needing a new port’
- ‘Damaging lungs’
- ‘Not being well later in life’
- ‘The future’
- ‘Lack of scientific development’
- ‘That he grows pseudomonas and needs a portacath fitted’
- ‘Becoming infected with something’
- ‘F picking up bacteria’s that require hospital stays for’

Carers felt most positive about the following:

- ‘A cure’
- ‘Everything’
- ‘Research’
- ‘Treatment getting better’
- ‘M’s ability to cope with CF’
- ‘C has improved on his eating’
- ‘That M is happy and well looked after by the CF team’
- ‘How well he has been’
- ‘J has not been in hospital for 10 years’
- ‘How well she has cleared infection in the past’
- ‘Not much at the moment’
- ‘Care from the CF team’
- ‘How well he keeps at present’
- ‘Advances in medicine’
- ‘Her happiness/ attitude to life’

ITEM		PERCENTAGE (N=17)				
		1 A great deal	2	3 Moderately	4	5 Not at all
32. It is difficult to predict what the future holds in relation to CF. To what extent does this uncertainty affect your family's approach to life?		6% (1)	29% (5)	35% (6)	6% (1)	24% (4)
33. How much does the responsibility of looking after a child with CF affect you?		18% (3)	47% (8)	18% (3)	18% (3)	0% (0)
34. How much is your child's growth a worry for you?	**Height?**	18% (3)	6% (1)	24% (4)	24% (4)	29% (5)
	Weight?	18% (3)	12% (2)	35% (6)	29% (5)	6% (1)
35. To what extent are you worried that your child might become infected with Pseudomonas when s/he is outside the home, e.g. at friends' houses, at school?		12% (2)	12% (2)	53% (9)	6% (1)	6% (1)
36. How worried are you about a change in your child's lung function?		18% (3)	35% (6)	35% (6)	12% (2)	0% (0)

Table 11B. Daily hopes and worries

6.3.b.vi. CF Routines

Four questions evaluated the thoughts and difficulties caused by CF routines in families with children with CF. The management of CF demands various treatments such as medications, physiotherapy, nebulisers and nutritional supplements. The family has to work these into their daily routine. The majority of the participants reported no or a few problems in the daily routine (Table 12A). Digestion difficulties seemed to be a constant problem for two (12%) families. Physiotherapy and mealtimes caused constant problems for one (6%) family each. Taking oral medications such as vitamins and antibiotics caused the least amount of problems for these families (59%, n=10).

40. How much of a problem is it to manage the daily routines for CF now?	**PERCENTAGE (N=17)**				
	1 No problem	**2**	**3**	**4**	**5 A constant problem**
a. Mealtimes- getting him/her to eat enough	35% (6)	18% (3)	24% (4)	18% (3)	6% (1)
b. Digestion- tummy problems (wind, pain, diarrhoea)	29% (5)	29% (5)	29% (5)	0% (0)	12% (2)
c. Taking enzymes/creon	47% (8)	29% (5)	18% (3)	6% (1)	0% (0)
d. Taking vitamins/ oral antibiotics	59% (10)	29% (5)	12% (2)	0% (0)	0% (0)
e. Doing physiotherapy	29% (5)	18% (3)	24% (4)	18% (3)	6% (1)
f. Doing nebulised medications	29% (5)	12% (2)	29% (5)	12% (2)	0% (0)

Table 12A. Problems with CF routines

From above, we can see that the CF routine is quite complex. Six participants (35%) found it moderately easy to establish a CF care routine when their child was first diagnosed. One (6%) carer found it very easy and another one found it quite difficult to establish a routine. Eleven (65%) felt that the treatments for their child were all justified but five (29%) felt quite overwhelmed with the demands of CF that have been placed on them (Table 12B).

ITEM	SCALE	PERCENTAGE (N=17)
39. How easy was it to establish the CF care routine after your child was diagnosed?	1 (Very easy)	6% (1)
	2	24% (4)
	3 (Moderately easy)	35% (6)
	4	29% (5)
	5 (Not at all easy)	6% (1)
41. With all the things that need to be done, it may be overwhelming at times. How true has this been for you over the last 2 weeks?	1 (Very true)	29% (5)
	2	24% (4)
	3 (Neutral)	24% (4)
	4	12% (2)
	5 (Not at all true)	12% (2)
42. Do you think doing all these treatments for your child are justified?	1 (Completely justified)	65% (11)
	2	12% (2)
	3 (Not sure)	24% (4)
	4	0% (0)
	5 (Not at all justified)	0% (0)

Table 12B. Thoughts on CF routines

6.3.b.vii. Community Support

Five items obtained information on support from the community including child minders, GP and pharmacies. Most carers (88%, n= 15) had a moderately good to a very good relationship with their local GP and only one (6%) carer reported a very bad relationship with their GP. Most pharmacists (82%, n=14) were also seen to be very helpful (Table 13). Most carers (53%, n=9) had a very good relationship with their child's minder/nursery/school and seemed to be quite comfortable with them administering medications to their children. Despite this, only six (35%) carers got support by these groups for other treatments for their children, such as physiotherapy and nebulisers. One parent claimed that the child's grandmother administered medication and physiotherapy. Another child had the community physiotherapist providing services in school once a week.

ITEM	SCALE	PERCENTAGE (N=17)
43. What quality of relationship do you have with your local GP/surgery?	1 (Very good)	41% (7)
	2	12% (2)
	3 (Moderately good)	35% (6)
	4	6% (1)
	5 (Not at all good)	6% (1)
44. How helpful is your local pharmacist?	1 (Very helpful)	82% (14)
	2	18% (3)
	3 (Moderately helpful)	0% (0)
	4	0% (0)
	5 (Not at all helpful)	0% (0)
45. What sort of relationship do you have with your child's minder/nursery/school?	1 (Very good)	53% (9)
	2	29% (5)
	3 (Moderately good)	6% (1)
	4	6% (1)
	5 (Very good)	0% (0)
46. How comfortable are you with how your child's minder/ nursery or school gives medications to your child?	1 (Very comfortable)	53% (9)
	2	12% (2)
	3 (Moderately comfortable)	18% (3)
	4	6% (1)
	5 (Not at all comfortable)	0% (0)
47. Do you get support from your child's minder/nursery or school above and beyond creon or inhalers e.g., physio or nebs	Yes	35% (6)
	No	47% (8)

Table 13. Community support

6.3.b.viii. CF Clinic & Pharmacy Visits

At Alder Hey, most CF patients attend the routine CF clinic every eight to nine weeks and spend two to three hours on each visit being seen by various medical professionals from the CF team. These visits and hospital pharmacy visits were assessed by seven questions. Most participants felt that the messages they got from the team members were very consistent (53%, n=9) and adequate for their needs (76%, n=13). The hospital pharmacy was used rarely or on some occasions by the CF patients. The waiting times were moderate to very unacceptable for the families (Table 14).

ITEM	SCALE	PERCENTAGE (N=17)
51. How often do you use the hospital pharmacy?	Most visits	0% (0)
	Sometimes	41% (7)
	Rarely	59% (10)
52. Please think about your last visit to pharmacy. How acceptable was the wait for medicines?	1 (Very acceptable)	0% (0)
	2	24% (4)
	3	53% (9)
	4	18% (3)
	5 (Very unacceptable)	6% (1)
53. How consistent are the messages you get from different members of the CF team?	1 (Very consistent)	53% (9)
	2	29% (5)
	3 (Moderately consistent)	12% (2)
	4	0% (0)
	5 (Not at all consistent)	6% (1)
54. How much information would you like to have from the CF team about your child's condition or treatment?	1 (More information)	6% (1)
	2	18% (3)
	3 (The same as now)	76% (13)
	4	0% (0)
	5 (Less information)	0% (0)

Table 14. CF Clinic and pharmacy visits at Alder Hey

6.3.b.ix. Inpatient and Day Patient Stays

Participants whose children had been admitted to hospital completed five questions regarding details of recent hospital admissions. Four (24%) children had hospital admissions in the three months preceding the annual review. Out of these, one child had four separate admissions, the remainder had only one admission each. Most of the admissions were for IV antibiotics. One child was admitted for the removal of her adenoids. This was the first admission for one child, two had previous admissions and one carer did not answer this question. Two children had been admitted for routine/prevention measures and two for treatment purposes. Two carers found this admission to be stressful, one found it to be very stressful and one was not at all stressed. In order of increasing stress, the following situations were rated:

- Communication with healthcare professionals
- Staying in overnight
- Child's loneliness
- Getting good care in hospital
- Getting the intravenous line in
- Disruptions to family life

Other types of stress were:

- The worry and risk of cross infection
- Buying meals for themselves during their child's admission turned out to be quite expensive for one carer

6.3.b.x. CF Treatments

This section of the CLCF-Q details most of the main treatment options for the management of CF. It consists of three questions with numerous subsections each. Inconsistencies were found in the answers from this section. Certain treatments were not reported as being carried out in question 60, but in the subsequent questions, participants rated difficulty levels for them (Table 15A, 15B and 15C) For example, only four (24%) participants reported administering inhalers to their children (Table 15A). Seven (41%) reported difficulty levels for themselves (Table 15B) and six (35%) reported difficulty levels for their children (Table 15C) when taking inhalers. Similar inconsistencies can be found for other treatments as well.

Patients spent the most time each day doing physiotherapy (mean 33.8 mins/day). It was also the task that was found to be the most difficult for carers as well as the children and the one that required the most effort. Only one participant reported having overnight feeds and oxygen therapy each. The time per day reported for these treatments was nine minutes and two minutes respectively. Presumably, the participants meant nine and two hours, but did not realise that the units for the column were in minutes.

60. Over the last two weeks how much has your child needed the following treatments to keep him/her well?		SECTION 1A Prescribed? Percentage %(N)			SECTION 1B Treatment taken: Time required		SECTION 1C: Treatment taken: Effort required Percentage %(N=17)		
		Yes	Yes but not done	No	Minutes per day doing task (Mean)	Number of days of treatment over past week (Mode)	Minimal effort	Moderate effort	High effort
Calculating doses		29 (5)	6 (1)	47 (8)	9.8	7	18 (3)	18 (3)	
Extra day time feeding/ calorie supplements		24 (4)		41 (7)	12.3	7	12 (2)	6 (1)	
Inhalers		29 (5)		47 (8)	4.2	7	12 (2)	18 (3)	
Insulin injections for diabetes				76 (13)					
IV antibiotics at home		6 (1)	6 (1)	65 (11)	15	7			6 (1)
IV antibiotics in hospital			6 (1)	71 (12)					
Nebulised medications	Antibiotics	47 (8)		47 (8)	9.1	7	18 (3)	24 (4)	
	DNAse	18 (3)		59 (10)	4.3	7	18 (3)		
	Hypertonic saline	6 (1)		59 (10)	3	4			
	Salbutamol			59 (10)					
Non-prescribed (alternative remedies e.g., herbal remedies)				12 (2)					
Oral antibiotics (back-up and specific)		71 (12)		18 (3)	3.5	7	59 (10)	12 (2)	
Other medicines (lactulose, antacids, vitamins etc)		82 (14)		18 (3)	4.2	7	59 (10)	18 (3)	
Overnight feed through a gastrostomy or nasogastric tube		6 (1)		71 (12)	9	5		6 (1)	
Oxygen therapy delivered by mask or nasal specs		6 (1)		76 (13)	2	7	6 (1)		
Pancreatic enzyme supplements (creon)		88 (15)		6 (1)	16.6	7	59 (10)	24 (4)	
Physiotherapy		71 (12)	12 (2)	6 (1)	33.8	7	35 (6)	29 (5)	24 (4)
Ursodeoxycholic acid (URSO) for liver involvement		24 (4)		59 (10)	6.8	7	12 (2)	12 (2)	
Collecting and preparing medicines and cleaning equipment		82 (14)		12 (2)	15.5	7	41 (7)	29 (5)	

Table 15A. CF Treatments

61. We want to know how hard it has been for YOU to manage these treatments.		How hard has it been for YOU to manage these treatments? Percentage (N=17)			
		Very difficult	Somewhat difficult	Not at all difficult	Does not apply
Extra day time feeding/ calorie supplements		0% (0)	12% (2)	35% (6)	35% (6)
Inhalers		0% (0)	12% (2)	29% (5)	41% (7)
Insulin injections for diabetes		0% (0)	0% (0)	0% (0)	76% (13)
IV antibiotics at home		6% (1)	6% (1)	18% (3)	53% (9)
IV antibiotics in hospital		0% (0)	0% (0)	12% (2)	59% (10)
Nebulised medications	Antibiotics	0% (0)	24% (4)	29% (5)	35% (6)
	DNAse	0% (0)	6% (1)	12% (2)	59% (10)
	Hypertonic saline	0% (0)	6% (1)	0% (0)	65% (11)
	Salbutamol	0% (0)	0% (0)	0% (0)	71% (12)
Non-prescribed (alternative remedies e.g., herbal remedies)		6% (1)	0% (0)	29% (5)	47% (8)
Oral antibiotics (back-up and specific)		6% (1)	12% (2)	65% (11)	0% (0)
Other medicines (lactulose, antacids, vitamins etc)		6% (1)	12% (2)	71% (12)	0% (0)
Overnight feed through a gastrostomy or nasogastric tube		0% (0)	6% (1)	0% (0)	71% (12)
Oxygen therapy delivered by mask or nasal specs		0% (0)	12% (2)	0% (0)	71% (12)
Pancreatic enzyme supplements (creon)		0% (0)	24% (4)	65% (11)	0% (0)
Physiotherapy		6% (1)	35% (6)	41% (7)	0% (0)
Ursodeoxycholic acid (URSO) for liver involvement		0% (0)	12% (2)	12% (2)	59% (10)

Table 15B. CF Treatments- Difficulties for caregivers

62. How do you think YOUR CHILD has managed these aspects of the CF routine over the last two weeks?		How hard has it been for YOUR CHILD to manage these treatments? Percentage (N=17)			
		Very difficult	Somewhat difficult	Not at all difficult	Does not apply
Extra day time feeding/ calorie supplements		6% (1)	6% (1)	29% (5)	53% (9)
Inhalers		0% (0)	12% (2)	24% (4)	53% (9)
Insulin injections for diabetes		0% (0)	0% (0)	0% (0)	82% (14)
IV antibiotics at home		6% (1)	0% (0)	12% (2)	71% (12)
IV antibiotics in hospital		0% (0)	0% (0)	6% (1)	71% (12)
Nebulised medications	Antibiotics	0% (0)	24% (4)	24% (4)	47% (8)
	DNAse	0% (0)	0% (0)	18% (3)	59% (10)
	Hypertonic saline	0% (0)	6% (1)	0% (0)	65% (11)
	Salbutamol	0% (0)	12% (2)	6% (1)	59% (10)
Non-prescribed (alternative remedies e.g., herbal remedies)		0% (0)	0% (0)	12% (2)	71% (12)
Oral antibiotics (back-up and specific)		6% (1)	29% (5)	53% (9)	12% (2)
Other medicines (lactulose, antacids, vitamins etc)		6% (1)	18% (3)	59% (10)	6% (1)
Overnight feed through a gastrostomy or nasogastric tube		0% (0)	12% (2)	0% (0)	76% (13)
Oxygen therapy delivered by mask or nasal specs		0% (0)	6% (1)	0% (0)	82% (14)
Pancreatic enzyme supplements (creon)		6% (1)	12% (2)	82% (14)	0% (0)
Physiotherapy		6% (1)	41% (7)	47% (8)	0% (0)
Ursodeoxycholic acid (URSO) for liver involvement		0% (0)	0% (0)	24% (4)	76% (13)

Table 15C. CF Treatments- Difficulties for patients

6.4. FEEDBACK RESULTS

The investigator feedback sheet was scored based on the participant's CLCF answers (Table 3). The investigators scored most of the domains in the feedback sheet as being 'true,' except in the 'Hopes and Worries for everyday life' domain. Here, most participants received an intermediate score (65%, n=11), 53% (n=9) were also found to be quite overwhelmed with their duties as a carer (Table 16A).

Overall, the self-feedback forms had lower scores than the investigator version (Table 16B). Like the investigator version, the majority of the items were rated as being 'true' by the caregivers. Unlike the investigator version, a higher proportion of caregivers (although not the majority) had scored domains as being 'false.' Caregivers expressed more worry about infections than investigators thought they did. Caregivers also felt more hopeful about daily life and reported fewer feelings of being overwhelmed than the investigators thought they felt. The differences between both the feedback scores were calculated (Table 16C).

DOMAIN	SUB-DOMAIN	HIGHEST POSSIBLE SCORE	SCALE (Investigator) Percentage (N=17)		
			TRUE ☺	😐	FALSE ☹
Family Lifestyle 'We work well as a family'	We share challenges	5	53% (9)	24% (4)	24% (4)
	We work together	15	59% (10)	41% (7)	0% (0)
CF Background 'My child's CF is well managed'		5	76% (13)	24% (4)	0% (0)
Child's Character 'My child is well behaved'		35	65% (11)	35% (6)	0% (0)
Challenges of Family Life 'We face CF together as a family'	I have a lot of support	30	94% (16)	6% (1)	0% (0)
	We don't worry about infections	5	65% (11)	12% (2)	24% (4)
	CF doesn't impact on our life too much	15	41% (7)	35% (6)	24% (4)
Hopes and Worries 'My hopes for everyday life are bigger than my worries about everyday life'	I am hopeful about our day to day life	30	24% (4)	65% (11)	12% (2)
	I don't worry about everyday life	30	24% (4)	65% (11)	12% (2)
Hopes and Worries 'My hopes for the future are bigger than my worries about the future'	I have a lot of hope for the future	30	76% (13)	24% (4)	0% (0)
	I don't worry about the future	30	76% (13)	24% (4)	0% (0)
CF Routines 'Managing my child's CF is easy'	We have a good routine	35	59% (10)	41% (7)	0% (0)
	I never feel overwhelmed	5	24% (4)	24% (4)	53% (9)
Community Support 'I have a good relationship with the CF team in the community'		20	82% (14)	18% (3)	0% (0)
CF Clinic and Pharmacy Visits 'I find it easy getting my child's medication'		10	82% (14)	18% (3)	0% (0)

Table 16A. Investigator feedback scores

DOMAIN	SUB-DOMAIN	SCALE (Caregiver) Percentage (N=17)		
		TRUE ☺	😐	FALSE ☹
Family Lifestyle 'We work well as a family'	We share challenges	76% (13)	18% (3)	6% (1)
	We work together	76% (13)	24% (4)	0% (0)
CF Background 'My child's CF is well managed'		82% (14)	18% (3)	0% (0)
Child's Character 'My child is well behaved'		71% (12)	29% (5)	0% (0)
Challenges of Family Life 'We face CF together as a family'	I have a lot of support	65% (11)	29% (5)	6% (1)
	We don't worry about infections	0% (0)	41% (7)	59% (10)
	CF doesn't impact on our life too much	18% (3)	35% (6)	47% (8)
Hopes and Worries 'My hopes for everyday life are bigger than my worries about everyday life'	I am hopeful about our day to day life	65% (11)	29% (5)	6% (1)
	I don't worry about everyday life	29% (5)	47% (8)	24% (4)
Hopes and Worries 'My hopes for the future are bigger than my worries about the future'	I have a lot of hope for the future	59% (10)	35% (6)	6% (1)
	I don't worry about the future	24% (4)	41% (7)	35% (6)
CF Routines 'Managing my child's CF is easy'	We have a good routine	71% (12)	29% (5)	0% (0)
	I never feel overwhelmed	24% (4)	41% (7)	35% (6)
Community Support 'I have a good relationship with the CF team in the community'		82% (14)	6% (1)	6% (1)
CF Clinic and Pharmacy Visits 'I find it easy getting my child's medication'		76% (13)	24% (4)	0% (0)

Table 16B. Caregiver self-feedback scores

DOMAIN	SUB-DOMAIN	DIFFERENCE BETWEEN FEEDBACK SCORES (Caregiver) - (Investigator)		
		TRUE ☺	😐	FALSE ☹
Family Lifestyle 'We work well as a family'	We share challenges	4	-1	-3
	We work together	3	-3	0
CF Background 'My child's CF is well managed'		1	-1	0
Child's Character 'My child is well behaved'		1	-1	0
Challenges of Family Life 'We face CF together as a family'	I have a lot of support	-5	4	1
	We don't worry about infections	-11	5	6
	CF doesn't impact on our life too much	-4	0	4
Hopes and Worries 'My hopes for everyday life are bigger than my worries about everyday life'	I am hopeful about our day to day life	7	-6	-1
	I don't worry about everyday life	1	-3	2
Hopes and Worries 'My hopes for the future are bigger than my worries about the future'	I have a lot of hope for the future	-3	2	1
	I don't worry about the future	-9	3	6
CF Routines 'Managing my child's CF is easy'	We have a good routine	2	-2	0
	I never feel overwhelmed	0	3	-3
Community Support 'I have a good relationship with the CF team in the community'		0	-2	1
CF Clinic and Pharmacy Visits 'I find it easy getting my child's medication'		-1	1	0

Table 16C. Difference between feedback scores

6.5. FEEDBACK AND SELF-EFFICACY

The investigator feedback scores were used to represent the overall performance of the intervention when analysing the relationship between the intervention process and the self-efficacy scores. The scatterplot graph below (Figure 9) shows that there is a moderately positive relationship between the two variables ($r=0.547$, $p< 0.05$). Feedback scores account for approximately 30% of the variability of self-efficacy scores at baseline.

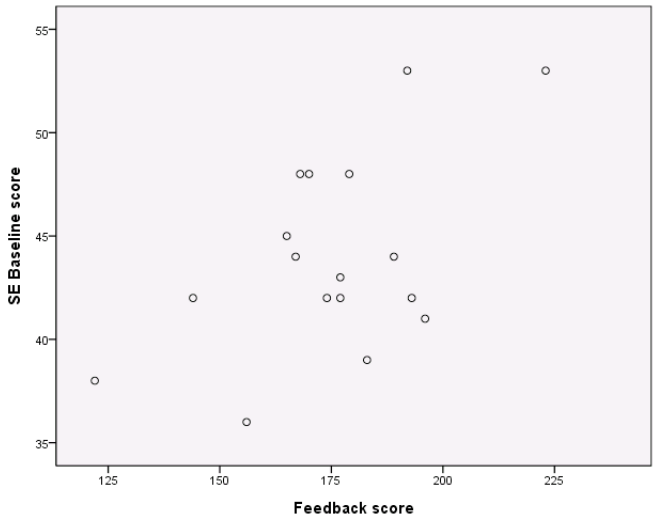


Figure 9. Relationship between baseline self-efficacy score and feedback score

CHAPTER FOUR

DISCUSSION

7.0. DISCUSSION

The aim of this exploratory pilot study was to investigate the effect of administration and feedback of the CLCF-Q to caregivers of children with CF on their self-efficacy. The CLCF-Q was developed to be used during the annual review process to measure the burden of care faced by these caregivers. The self-efficacy was measured by the CFSE-Q.

Whilst most parents are the primary caregivers of their children, parents of children with chronic diseases find that a lot more is demanded of them in terms of the care giving process. For some families, the illness may develop into a dominating presence in the household, leading to sacrifices of other daily routines. This extra burden can have negative effects on their physical and psychological health.[119,120] CF is a complex disease with a complicated treatment approach. The management of CF requires the cooperation and communication between everyone involved in the care of the patient with CF. Caregivers have to invest a lot of time and effort to adequately care for their child with CF. They have to administer medication, perform physiotherapy and other treatments and monitor their child's health on a regular basis. The CF team has a major role to play in all aspects of the life of a patient with CF. They prescribe treatment plans, educate the patient as well as the family about the disease, the various management options and the outlook for CF. They also monitor their patient's health by using clinical examinations and investigations. Families with CF require the support from the healthcare team as well as social support from the community to help them deal with the challenges they face as a result of CF.

7.1. STUDY FINDINGS

7.1.a. Demographics

Studies have found that mothers are more likely than fathers to take on the main care giving role for children with chronic diseases.[78,120] Demographics from those recruited into this study show that 83% (n=33) of participants were mothers. From the CLCF-Q data, 53% (n=9) reported an equal share in CF duties between the family/partners. Regardless, when asked who the main carer was prior to handing out the questionnaires, most mothers volunteered over their partners.

7.1.b. The CLCF-Q

Data from the CLCF-Q revealed some of the burdens faced by caregivers of children with CF. The majority of the caregivers felt that their families were quite active and led a generally busy lifestyle. Most also felt that members of the family worked together and that the family, as a unit, seemed to handle the challenges of CF quite well. All except one caregiver reported very little difficulty juggling the demands of their family with the demands of CF on a personal level. These families seem to work together and support each other with everyday tasks as well as CF-related tasks, which has the effect of easing the burden on the main caregiver. Family and social support, marital satisfaction and good relations within the family have all been found to be associated with better adjustment in mothers of children with chronic diseases.[121]

Over half of the children in the intervention group had been infected with *Aspergillus, Pseudomonas* or MRSA in the three months leading up to their annual review. 82% had been admitted to hospital in the past, with the most common reason for admission being IV antibiotics. Despite this, in the two weeks preceding the annual review, 76% of the study population in the intervention group reported that their child had been well or mostly well. Many studies have found no significant relationships between disease severity and parental adjustment. Stress as a result of the child's illness has been found to have a strong association with maternal adjustment problems. General and daily life stressors also have an effect on maternal adjustment.[121]

Caregivers of children with chronic diseases and disabilities suffer from a considerable amount of stress. A strong social support team has been seen to improve outcomes for caregivers.[119] The main sources of support for the families in the intervention group were the CF team in the hospital and their pharmacists. The GP was supportive to over half the study population in the intervention group and family and friends were also an important source of support for over half the families. During the feedback process, families were asked whether they felt that they required more support from any particular group in table 10A. Most were happy with the amount of support they received. The lack of support from the GP and other families with children with CF was noted and reasons have been given in the results section.

The burden of care had quite a prominent effect on the work pattern of the carers. Many felt that they had to sacrifice their working life to look after their child/children with CF. There was even one report of the lack of support and understanding from employers and colleagues. With fewer work commitments, the income of the family may also decrease. Some carers did feel that they were struggling to deal with the extra expenses as a result of their child's CF. All these challenges to family life can lead to increased amounts stress for the carer. Some carers even reported disturbed sleep at night usually due to stress. Despite these overwhelming feeling of stress, most carers had quite moderate hopes and worries about daily life and were very optimistic about their child's future. Parental perceptions of hopes, worries, threats, challenges and competence all influence the coping strategies they adapt to deal with various experiences. This in turn will affect outcomes and ultimately personal adjustment and well-being as well.[121]

Mealtimes, digestion problems, IV antibiotics at home, some oral medications and physiotherapy seemed to be a constant problem and caused difficulty for some families. Other than this, most families felt that they had a good treatment routine, which they coped well with. Physiotherapy was the most time-consuming treatment procedure for most patients with a mean of 33.8 minutes spent per day performing it. This is likely to contribute to a high treatment burden as long, complicated treatments have been found to be associated with reports of higher treatment burdens.[6,7]

7.1.c. The feedback process

The feedback process was designed to be easily understood and acceptable by caregivers. It involved the caregivers and recognised their efforts in caring for their child's disease. Where difficulties were identified, consultations with the appropriate professionals helped to encourage the caregivers of their abilities to provide adequate treatment. By using the theory of verbal persuasion to influence self-efficacy, the purpose of the feedback process was to guide the caregivers and involve them in planning and carrying out management behaviours for their affected child.[13,95]

Verbal persuasion encourages individuals and boosts their confidence in their capabilities to perform a given task successfully. A person who is encouraged positively by others is more likely to invest the effort required to achieve their goals.

Similarly, those who are discouraged will dwell on their deficiencies and are more likely to give up easily when faced with a challenge. For verbal persuasion to be effective it should not give an individual unrealistic expectations of their skills that could lead to possible failure, disappointment and a decline in perceived self-efficacy. Verbal persuasion should give an individual the confidence and strength to learn and develop appropriate skills to be successful in the tasks they perform.[95]

After completing the CLCF-Q most families welcomed the opportunity to get structured feedback. Many felt that this was very helpful and were eager to talk to the study investigator about the problems they faced. They were encouraged to discuss these problems with the appropriate healthcare professionals and to review the written feedback they received at home. Some of these problems may not necessarily be viewed by caregivers as legitimate problems to discuss with the CF team. The CLCF-Q helps healthcare professionals to identify these problems. Some participants did feel that this was a show of support and recognition of their difficulties by the CF team.

Despite the fact the most participants scored themselves lower than the investigator on the feedback form, the majority of them had quite a positive outlook on the challenges they faced. Many were surprised that the investigator had given them a higher rating and after discussion with the investigator, the reasons for the discrepancy became clear. The investigator scoring system took into consideration aspects of the CLCF-Q, which may not necessarily have been taken into consideration by the participants when rating themselves on the feedback sheet. For example, to assess the sub-domain 'CF doesn't impact on our lives too much,' investigators considered expenses associated with CF, changes in work pattern due to CF and disturbed night's sleep in the past two weeks (Table 4). Participants may have considered other aspects of life that CF has an impact on such as social, marital and family life, leisure activities and hobbies. When these issues were identified, they were highlighted and discussed with the caregivers.

7.1.d. Self-efficacy

There is a link between self-efficacy and depression and anxiety.[98] Individuals with a low sense of self-efficacy have few aspirations and set low standards for themselves. They believe that they are not capable of achieving their goals and focus on their

personal deficiencies and failures. They feel that they are unable to exercise control over threats and challenges in life and find it difficult to discard these disturbing thoughts and feeling. They do not derive pleasure and satisfaction from life and thus are more vulnerable to depression and anxiety. These individuals also have difficulties establishing and maintaining social relationships due to a low sense of self-worth and self-efficacy. Without adequate social support they are unable to gain encouragement and learn from observation of others, leading to worsening depression, anxiety and self-efficacy.[95] Caregivers of children with chronic diseases are likely to suffer from anxiety and depression.[119] Improving caregivers' self-efficacy may lead to a lower prevalence of depression and anxiety in them.

The CFSE-Q is an altered version of the previously validated GSE-S. This is the first time that it has been used in a population of caregivers of children with CF. There were no floor and ceiling effects observed in the CFSE-Q, as none of the participants from either group achieved the highest or lowest possible scores. The CFSE-Q can be used to identify any improvement or deterioration in self-efficacy, as this study population did not reach the limits of the questionnaire. The scores from both groups at baseline and end point were well spread and showed a normal distribution (Fig 6,7 & 8). The CFSE-Q was also able to measure changes in self-efficacy scores between two separate time points. It has shown promise as a tool to measure the self-efficacy in caregivers of children with CF.

The principal finding was that the self-efficacy score of the intervention group had remained steady over both time points ($p=0.4$) (Figure 7). That of the control group had shown a significant decrease with time ($p< 0.05$) (Figure 8). These data suggest that the CLCF and its' feedback by the investigator had the effect of stabilising the self-efficacy in those carers who had completed it. A few of the participants in the control group had quite a considerable decline in their self-efficacy scores. This may be due to significant life events and stressors, which were not formally assessed in this group. The mean self-efficacy score of the control group had dropped by 1.9 points at the end of the study. Considering the small sample size ($n=20$), this may represent a clinically significant change in self-efficacy score. Although there was no significant difference between the changes in scores of both groups, the intervention

group did have a more positive change in their self-efficacy score than the control group at the end of the study (12 and -30 respectively).

Item analysis of the CFSE-Q revealed that six items out of fourteen in the intervention group had better scores at the end of the trial compared to only two items in the control group. The CLCF-Q had a positive effect on nearly half of the items on the self-efficacy scale and managed to stabilise the score of two items.

The annual review gives health care professionals as well as caregivers the opportunity to assess and review the effects of their management of the child with CF. It can be a stressful process for the family involved. The self-efficacy of the intervention group had remained constant between two time points. This was the group that completed the CLCF-Q and received feedback on it. This may be the reason for the observed stability in self-efficacy. The CLCF-Q and feedback may be a mechanism that enables caregivers to maintain their confidence in themselves as well as their abilities to perform caregiving duties during a stressful period in their year. Without this intervention, the self-efficacy of the control group decreased significantly by the end of the study. Their burdens were not formally recognised by the investigators. The annual review may have had a negative effect on the participants' confidence in the control group thus leading to a decrease in their self-efficacy scores.

There was a moderately positive relationship between baseline self-efficacy scores and feedback scores, which represented overall performance on the intervention process. Participants who reported fewer burdens in the CLCF-Q were more likely to have higher self-efficacy scores at baseline. By using the CFSE-Q to measure self-efficacy, we may be able to identify caregivers who feel overburdened by their caregiving duties.

Self-management for chronic diseases requires the patient and family to learn new skills, practice, refine and maintain them. Those with a high sense of self-efficacy and who believe that they can exercise some control over their own health are more likely to set goals for recommended treatments, make lifestyle changes, monitor behaviours, motivate themselves, persevere with their efforts and refine and maintain their skills. Self-efficacy gives patients and families the confidence they need to manage their

disease at home. In this way, the measurement of self-efficacy can be used to predict health care behaviours and by influencing self-efficacy we can attempt to modify patient's health care behaviours.[95]

7.2. LIMITATIONS

Approximately 40% of the references used in this thesis have been drawn from the decade 1992-2002. As a result of advances in research and improvements in CF care, data from this decade may not represent a true picture of CF care today. Conversely nearly 50% of references used are more recent. This may have contributed to more reliable and up to date data in this thesis.

The CFSE-Q is a modified version of the GSE-S. The four additional questions could have affected the reliability and validity of the self-efficacy measure. This effect was not investigated in this study.

Self-efficacy data was not measured in the 'normal' (caregivers of healthy children). It would have been interesting to compare self-efficacy data between the caregivers in our study and those from the healthy population.

From figure 8 we can see that a few participants in the control group had drastic drops in their self-efficacy scores. This may have been due to other life stressors/events, which were not measured as a part of this study.

The CLCF-Q has already undergone the development and validation phases, but this data has not yet been published.[9] As this is a pilot study, there is still potential to make adjustments and changes to the structure of the questionnaire.

The CLCF-Q is a parent reported measure. Many items request detailed information. This may have introduced some recall bias into the results.

The CLCF-Q is a long, 10-page questionnaire. The longest and most complicated set of items is at the end of the questionnaire, spanning three pages and requesting the respondent to recall fine details in some cases (e.g. Time and effort spent per day on each treatment). Questionnaire length has been seen to have an effect on response rates. Longer questionnaires have lower response rates than shorter ones. Responses in longer questionnaires are more likely to be random, incomplete and of poor quality, especially towards the end of the questionnaire. A complicated group of questions

towards the end of a long questionnaire is likely to generate a set of similar, unreliable answers.[122-124]

The timing of the questionnaire, during the annual review may also have an adverse effect on the quality of the answers. The annual review is a stressful process for most caregivers. As they complete the questionnaire during their three hour morning appointment with various members of the health care team, they are likely to be distracted and anxious and may not give their full attention to the questionnaire, thus compromising its' reliability. Many did say that they did not appreciate the added hassle of completing the CLCF-Q during the annual review and would have preferred to take it home to complete. This was also stated as a reason for some caregivers withholding consent for the study. Other eligible caregivers did not appear for their child's annual review or clinic appointments and were therefore not included in the study.

'The Hawthorne effect' is a term used to describe a change in behaviour in research subjects as a result of the awareness that researchers are observing their behaviour. It is seen in behavioural interventional studies where subjects know that their behaviour is being observed.[125] In this study, participants were aware that their self-efficacy was being measured. This may have led to increased consciousness in personal mannerisms and coping mechanisms resulting in altered behaviours during the study period, thus introducing some bias into the results.

The timeline of this study may also introduce some bias into the results. As a result of missed, rescheduled and delayed appointments, not all participants were followed up at equal time intervals. Most followed the time line suggested in figure 4, but there were others whose time lines were considerably different. The effect of the CLCF-Q and feedback process must be investigated in such cases. We do not know how long self-efficacy interventions take to have an effect or even how long this effect lasts for.

The results from this study may be limited by the small sample size. The strength of the present findings needs to be confirmed in a larger study with power to reduce a type I error.

Other limitations involve those of the investigator. Four and a half months after this pilot study was started, the interviewer changed. Both individuals underwent similar

training and overlapped for a period of four weeks, where the second interviewer was trained. Regardless, this change in interviewers may have led to a difference in the quality of certain aspects of the study such as the feedback process, which was written and verbal.

Transferring the data from the questionnaire to the database and spreadsheets on the computer may also be a source of error in this study. All precautions were taken to minimise this source of error. While the data was being input into the computer, it was checked twice. At a later time point, the data was once again checked by comparing the computer and questionnaire scores of randomly selected participants.

7.3. FURTHER WORK

The length and complexity of CLCF-Q may need to be reviewed so that more caregivers welcome it and are more willing to complete it. Time may need to be set-aside during the annual review specifically for the purpose of completing it in an appropriate environment. These adjustments may improve the reliability of the caregivers' answers.

If the variable time-lines of participants have an effect on the results of the study, the study design may have to be reviewed and altered to prevent this from occurring. Perhaps adherence to a strict time line should be implemented, but this may have adverse effects on the numbers of participants.

For future studies, a larger sample size would improve the reliability of the results and may also influence study design. Assessing the long-term effects of interventions such as the CLCF-Q and feedback process would also be beneficial to this group of patients and caregivers.

Self-efficacy should be measured regularly as it is constantly being influenced by external sources. Identifying low self-efficacy and intervening at the earliest possible opportunity may result in an improved standard of care for patients with CF.

7.4. CONCLUSION

The CLCF-Q has shown promise as a tool to identify the burden of care felt by caregivers of children with CF. It can be used to recognise problematic areas and introduce interventions to reduce the burden of care in this population. Measuring the

burden of care in such a population can also be used as outcome criteria for clinical trials in patients with CF.

The CLCF-Q and the feedback process have had the effect of stabilising the self-efficacy of those who completed the whole intervention process. By influencing self-efficacy, these interventions could possibly be used as a tool to try and modify health behaviours of caregivers of children with CF to achieve improvements in their child's health.

The CFSE-Q has also proved its' ability as a reliable measure of self-efficacy in caregivers of children with CF. Even though the additional four items have not been formally validated, the CFSE-Q was able to identify changes in self-efficacy in this population. The use of the CFSE-Q may extend beyond this study. It could be used to measure self-efficacy in other situations and studies in a similar population. It could also be used to identify caregivers with low self-efficacy who are potentially at risk of providing inadequate care to the patient.

Successful incorporation of the CLCF-Q into the annual review process may be a long-term strategy to maintain a high level of self-efficacy for caregivers of children with CF. As these children grow older, they will start to be more responsible for their own treatment. Strategies that improve their self-efficacy would benefit the quality of care and reduce adverse mental health issues arising from managing a chronic disease.

CHAPTER FIVE

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8.0. REFERENCES

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CHAPTER SIX

APPENDIX

9.0. APPENDIX

9.1. CORRESPONDENCE WITH PARTICIPANTS

9.1.a. Information letter to caregivers

Alder Hey

Eaton Road
Liverpool
L12 2AP

Telephone: 0151 228 4811
www.alderhey.com

2011

Dear Parent or Guardian

RE: The PROMISE Study (Parent Reported Outcome Measures Improve Self Efficacy)

Last year, we validated a questionnaire which explores the time and effort that goes into caring for a child with Cystic Fibrosis. This questionnaire is called the Challenges of Living with Cystic Fibrosis (CLCF). It has been suggested that Patient/Parent Reported Outcome measures such as the CLCF may have positive effects on the Self Efficacy of participants. We would like to explore this theory in further detail and in order to do so we need your help.

You may have already been approached regarding your participation in the PROMISE Study. Consider this letter as a formal invitation to participate in the PROMISE Study.

Please find enclosed in this pack a, 'PARTICIPANT INFORMATION SHEET.' If you do wish to participate in this study please read the information sheet carefully. Please do not hesitate to contact me if you have any questions regarding this study and your participation.

You will be approached at your next Annual Review appointment to discuss your possible involvement and discuss any queries you may have. Only participants who understand the information provided and voluntarily consent will be included in the study.

I look forward to seeing you at your next appointment and I thank you in advance for your cooperation and time.

Yours sincerely,

Miss Tulsi Patel

Medical Student/MPhil Student
E-mail: t.p.patel@student.liverpool.ac.uk
Telephone: 0151 228 4811 Ext 4532

9.1.b. Participation information sheet

Participant Information Sheet

1. **Title**
   Does administering a parent reported outcome measure during the annual review improve the self efficacy of the carer of a child who has cystic fibrosis?

2. **Investigators**
   Dr Kevin Southern PhD MBChB
   Dr Claire Glasscoe PhD
   Miss Latifa Patel (Medical/MPhil Student)
   Dr Clare Dixon PhD
   Miss Tulsi Patel (Medical/MPhil Student)

3. **Version number and date**
   Version 3
   Last amended 8th March 2010

4. **Invitation**
   You are invited to participate in a research study. Before you make a decision, it is important for you to understand why the research is being done and what it will involve. Please read the following information carefully. You may wish to discuss this with your family and friends. I would like to stress that you do not have to accept this invitation and should only agree to take part if you want to. Thank you in advance for taking your time to read this information. If there is anything you do not understand or if you have any questions please do not hesitate to contact Elinor on the details below.

5. **What is the purpose of the study?**
   Last year, we validated a questionnaire which explores the time and effort that goes into caring for a child with Cystic Fibrosis (CF). This questionnaire is called the Challenges of Living with Cystic Fibrosis Questionnaire (CLCF-Q). We believe that Patient/Parent Reported Outcomes such as the CLCF-Q may have positive effects on your Self Efficacy. Self efficacy is your belief in your own ability to succeed. We would like to explore this theory.

6. **Why have I been chosen to take part?**
   All patients currently registered on the Cystic Fibrosis list have been considered for the study. As a parent/carer of a child with Cystic Fibrosis you are being invited to join the study.

7. **Do I have to take part?**
   Participation is completely voluntary and you are free to withdraw at anytime. You do not need to give us a reason. If you are unsure of what is involved please contact Elinor on the details at the end of the document.

8. **What will it involve for my family and me if I take part?**
   The principal investigator is Dr Kevin Southern.

If you choose to participate in the study Tulsi Patel will take informed consent from you at your child's annual review appointment. The study will be explained to you in detail and there will be an opportunity for you to raise any questions you may have.

You will be randomly allocated into 2 groups. If you are in Group 1 you will be asked to complete 3 questionnaires and 1 feedback form. If you are in Group 2 you will be asked to complete 2 questionnaires and one feedback form.

The Self Efficacy Questionnaire should take no longer than 10 minutes to complete and will be issued to all participants. The CLCF Questionnaire may take up to 30 minutes to complete and will only be issued to you if you are allocated into Group 1. All questionnaires will be issued during clinic time and will be collected back in at the end of clinic time.

Throughout the study you will be given opportunities to voice your opinions and any concerns you may about the study. With your permission we may use these to support the results of this study. Your name will not appear in the study and all information your give will remain anonymous. You can withdraw any comments at any point during the study.

9. How time consuming is this going to be?

You will not be required to attend any extra sessions/appointments so you do not need to make any extra journeys to Alder Hey. However, you may be asked to participate in an interview at the end of the study. This again will be scheduled during a clinic session.

10. Expenses and/or payments

You will incur no expenses. The time used to complete the study will be taken from your currently scheduled clinic times.

11. Are there any benefits in taking part?

You may benefit from the feedback given in the clinic appointment after your Annual Review regarding your questionnaire answers. You may also find the results of the study of use.

12. Are there any risks in taking part?

You are under no risk or disadvantage.

13. What if I am unhappy or there is a problem?

If you are unhappy, or if there is a problem, please do let Elinor know. Her contact details are below and she will try to help you. If you remain unhappy or have a complaint which you feel you cannot come to Elinor with then you should contact the Research and Development Manager Dot Lambert on 0151 252 5673 or dot.lambert@rlc.nhs.uk. When contacting the Research Governance Officer, please provide details of the name or description of the study (so that it can be identified), the researcher(s) involved, and the details of the complaint you wish to make.

14. Will my participation be covered by an insurance scheme?

You are taking part in a NHS Research Ethics Committee approved study and are fully covered.

15. Will my participation be kept confidential?

All data will be collected on the paper questionnaires and then transferred anonymously onto a secure computer program. It is completely anonymous and confidential and individual data will never be discussed. It will only be used in connection with the above named study. Only the principal researcher will have access to the data.

Once the study has been evaluated the data will be used to come to a conclusion about the participant population as a whole and never individually.

16. How will my personal data be used?

Relevant sections of your child's medical notes and data collected during the study may be looked at by individuals from regulatory authorities or from the NHS Trust, where it is relevant to you taking part in this research.

During the study it may also be necessary for the researchers to look at your child's medical records and access personal data. This will be to aid your participation in the study.

Personal data will not be removed from the hospital premises and will not be mentioned anywhere in the study. Your child's personal data will be kept confidential at all times.

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

The results will be written up and you will be able to access the grouped data but not the individual anonymous data.

This study may be published at a later data and you will be informed and acknowledged for all your help and support. Copies of the study will be forwarded to you at your request.

18. What will happen if I want to stop taking part?

You can withdraw at anytime, without explanation. Results up to the period of withdrawal may be used, if you are happy for this to be done. Otherwise you may request that they are destroyed and no further use is made of them. Your routine treatment will not be affected in anyway.

19. Who can I contact if I have further questions?

ELINOR F BURROWS
RESPIRATORY DEPARTMENT
ALDER HEY CHILDREN'S NHS FOUNDATION TRUST
EATON ROAD
LIVERPOOL
MERSEYSIDE
L12 2AP

E-mail: elinor.burrows@alderhey.nhs.uk
Tel: 0151 252 5297

THANK YOU

9.1.c. Letter to caregiver accompanying feedback results

Alder Hey

Eaton Road
Liverpool
L12 2AP

Telephone: 0151 228 4811
www.alderhey.com

July 2010

Dear

RE: The PROMISE Study (Parent Reported Outcome Measures Improve Self Efficacy)

Following your participation in the PROMISE Study please find attached your follow up results from your child's Annual Review. These are simply for your information and we do not require you to do anything further.

On behalf of the research team I would like to thank you for taking part in the study.

Please do not hesitate to contact me if you have any queries.

Yours sincerely,

Miss Tulsi Patel

Medical Student/MPhil Student

E-mail: t.p.patel@student.liverpool.ac.uk
Telephone: 0151 228 4811 Ext 4532

9.2. QUESTIONNAIRES AND FORMS

9.2.a. Consent form

Consent Form

To be completed by the parent or legal guardian

Title of Study:

Does administering a parent reported outcome measure during the annual review improve the self efficacy of the carer of a child who has cystic fibrosis?

Name of Investigators:

Dr Kevin Southern
Dr Claire Glasscoe
Miss Latifa Patel
Dr Claire Dixon
Miss Tulsi Patel

I agree to take part in the above study and for the relevant information about my child to be used.

Child's Name.....(please print in CAPITAL letters)

	Please tick
I confirm that the above study has been fully explained to me	☐
I confirm that I was given opportunity to ask questions	☐
I confirm that I have received a copy of the, 'Participant Information Sheet'	☐
I confirm that I have received information on how to gain access to the findings of this study when available	☐

'I understand that relevant sections of my child's medical notes and data collected during the study, may be looked at by individuals from regulatory authorities or from the NHS Trust, where it is relevant to my taking part in this research. I give permission for these individuals to have access to my child's records'

Participation in this study is entirely voluntary and I have a right to withdraw from the study without giving a reason and in the knowledge that this will not affect my child's treatment in any way.

Name of parent/legal guardian.....(please print in CAPITAL letters)

Signature of parent/legal guardian.....

Signed in the presence of:

Name of witness.....(please print in CAPITAL letters)

Signature of witness.....

Date.....

Address.....

.....Postcode.....

08/03/2010 11:41:07 Version 5

9.2.b. Cystic Fibrosis Self-Efficacy Questionnaire (CFSE-Q)

Self Efficacy Questionnaire for Carers Instructions

The following questions are designed to assess your, 'self efficacy;' your belief in your own ability to succeed.

Please answer the questions below by ticking the box that most closely represents how you feel about the statement on the left. Only tick one box per statement.

1 = Not at all true

2 = Hardly true

3 = Moderately true

4 = Exactly true

The answers should be your own and should reflect how you feel, no one else. Do not spend too long thinking about the answers.

Self Efficacy Questionnaire for Carers

	Statement	Your Response			
		1 Not at all True	**2** Hardly true	**3** Moderately true	**4** Exactly true
1	I can always manage to solve difficult problems if I try hard enough				
2	I face problems on a daily basis				
3	If someone opposes me, I can find the means and ways to get what I want				
4	It is easy for me to stick to my aims and accomplish my goals				
5	I am confident that I could deal efficiently with unexpected events				
6	Thanks to my resourcefulness, I know how to handle unforeseen situations				
7	I do have the support I need to solve problems				
8	I can solve most problems if I invest the necessary effort				
9	I can only solve a problem if I expected it to happen				
10	I can remain calm when facing difficulties because I can rely on my coping abilities				
11	When I am confronted with a problem, I can usually find several solutions				
12	If I am in trouble, I can usually think of a solution				
13	I never feel my views are fully appreciated				
14	I can usually handle whatever comes my way				

Self Efficacy Questionnaire for Carers
Feedback

Thank you for taking the time to complete this questionnaire.

Do you have any queries or comments regarding this questionnaire or this study?

If you would like to discuss your queries or concerns please contact Miss Tulsi Patel on;

Institute of Child Health
Alder Hey
Eaton Road
Liverpool
L12 2AP

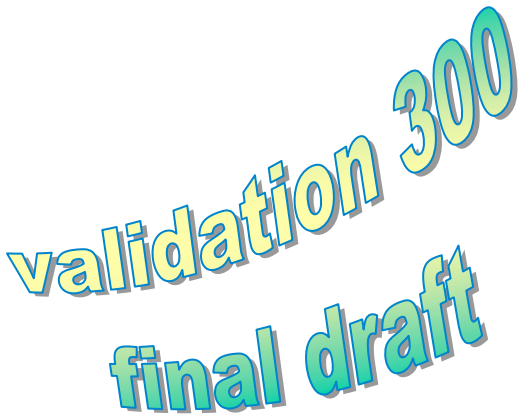
E-mail: t.p.patel@student.liverpool.ac.uk
Tel: 0151 228 4811 Ext 3536
Mobile: 078 288 199 69

9.2.c. The Challenges of Living with Cystic Fibrosis Questionnaire (CLCF-Q)

ID Number:

Challenges of Living with Cystic Fibrosis (CLCF)

A questionnaire for caregivers of children one year after diagnosis up to 13 years of age



Name of person completing form	Relationship to child with CF
Name of child with CF Boy/girl *Please circle*	Date of Birth of Child with CF _____/_____/_____ Day / Month / Year
When was your child diagnosed with CF? _____/_____ Month / Year	Does your child have a minder or baby sitter for part of the day? What nursery/school year/grade is your child in?
Today's Date _____/_____/_____ Day / Month / Year	

ID Number: ☐

In answering the questions on this and the next page, please consider your responses over the past two weeks

Family Lifestyle

1. Are you: A lone caregiver ☐ Living with spouse or partner ☐ A lone caregiver living with family ☐

2. How many children do you care for in your family? ☐

3. How many children with CF do you have living with you? ☐

4. How does your family divide childcare relating to CF?

1 2 3 4 5

I do it all — Equal shares — My partner/others do it all

5. How would you describe your general family lifestyle? *(please circle one number on each of the scales below)*

1 2 3 4 5

Relaxed — Stressed out

1 2 3 4 5

Disorganised — Organised

1 2 3 4 5

Busy — Laid back

1 2 3 4 5

No fixed routines — Fixed routines

1 2 3 4 5

Chatty — Quiet

1 2 3 4 5

Sporty/active — Not sporty/active

1 2 3 4 5

Work together — Work as individuals

6. How well do you think you are juggling the demands of CF with the needs of your family?

1 2 3 4 5

No difficulty — A little — Marginal — Definite — Great difficulty

7) How well do you think your family as a whole handles the challenges of CF ?

1 2 3 4 5

Very easily — Easily — Marginally easily — Some difficulty — Great difficulty

CF Background

8) Does your child need enzymes with food? Yes ☐ No ☐ Don't know ☐

9) Has your child been diagnosed with CF-related diabetes? Yes ☐ No ☐ Don't know ☐

10) Based on your most recent clinic visit, what is your child's FEV_1 % predicted/'blow score'? ☐

11) Has your child grown anything on a cough swab/sputum over the last three months?

Yes ☐ No ☐ Don't know ☐

If yes, please indicate which of the following:

Aspergillus ◯ Pseudomonas ◯ Burkholderia cepacia ◯ Other (please specify)_____

12) Over the last two weeks, has your child been: *(please tick one)*

Unwell ☐ Mostly unwell ☐ Mixture of well and unwell ☐ Mostly well ☐ Well ☐

ID Number:

13) Has your child ever had a hospital admission for any of the following? Yes ☐ No ☐
If yes then please tick all that apply)

IV Antibiotics ○ Portacath fitted ○ Gastrostomy tube fitted ○ Nasogastric tube ○ Oxygen ○

Other please specify_____

Child's Character

	Strongly agree	Agree	Neutral	Disagree	Strongly disagree
14) My child is very determined; when s/he wants to do something s/he usually keeps trying until s/he succeeds	1	2	3	4	5
15) My child makes more demands on me than I expected	1	2	3	4	5
16) My child goes to bed easily	1	2	3	4	5
17) My child sleeps throughout the night	1	2	3	4	5
18) It takes a long time for my child to settle with new routines	1	2	3	4	5
19) My child makes friends easily	1	2	3	4	5
20) My child is easily upset by things generally	1	2	3	4	5
21) My child is very moody	1	2	3	4	5
22) My child is so active it exhausts me	1	2	3	4	5
23) My child is popular with his/her peers	1	2	3	4	5
24) My child reacts very strongly when something happens that s/he doesn't like	1	2	3	4	5
	Strongly agree	Agree	Neutral	Disagree	Strongly disagree

Challenges to Family Life

25) How supported do you feel by the following groups of people? (*please tick boxes that best reflect your view).*

	Very supported			Not at all supported
Family members	1	2	3	4
Friends	1	2	3	4
Another parent whose child has CF	1	2	3	4

	Very supported			Not at all supported
CF team	1	2	3	4
GP	1	2	3	4
Pharmacy	1	2	3	4

Are there any other key people who are supportive to you?

_____

(For each of the following statements, please circle one number)

26) To reduce the risk of cross infection, the CF team advises that people with CF avoid contact with other people with CF. How much does this affect contact with other CF families?

1 2 3 4 5
A great deal Some Moderate A little Not at all

27) Caring for a child with CF can involve extra expense. How difficult is it for you to manage this?

1 2 3 4 5
Very difficult moderately Not at all difficult

28) To what extent do you think CF has changed your work pattern?

1 2 3 4 5
A great deal Some Moderate A little Not at all

If YES, in what way? _____

ID Number:

29) **How often have you had a disturbed night's** sleep in the past 2 weeks?

1 Every night 2 Frequent 3 Some 4 Few 5 No nights

30) How do you know when you need a break?

_____

Hopes and Worries

In answering the questions on this and the next section please consider your responses over the past two weeks

31) Some say that living with CF is like a balance of hope and worry:

What hopes do you have for your child?

	Very Hopeful			Not hopeful
S/he will adjust well to secondary school	1	2	3	4
S/he will go on to higher education	1	2	3	4
S/he will have a job	1	2	3	4

	Very hopeful			Not hopeful
S/he will have a family of his/her own	1	2	3	4
S/he will continue to be as well as s/he is now	1	2	3	4
There will be an advance in science that will help my child	1	2	3	4

32) It is difficult to predict what the future holds in relation to CF. To what extent does this uncertainty affect your family'**s approach to** life?

1 A great deal 2 3 Moderately 4 5 Not

If it does, in what way? _____

33) How much does the responsibility of looking after a child with CF affect you?

1 A great deal 2 3 Moderately 4 5 Not at all

34) How much is **your child's** growth a worry for you?

A) Height?

1 A great deal 2 3 Moderately 4 5 Not at all

B) Weight?

1 A great deal 2 3 Moderately 4 5 Not at all

35) To what extent are you worried that your child might become infected with pseudomonas when s/he is outside the home, e.g. at friends' houses, at school?

1 A great deal 2 3 Moderately 4 5 Not at all

36) How worried are you about a change in your **child's lung function?**

1 A great deal 2 3 Moderately 4 5 Not at all

37) What is your main worry? *(Please specify)*

_____

38) What do you feel most positive about? (*Please specify*)

_____

ID Number:

CF Routines

39) How easy was it to establish the CF care routine after your child was diagnosed?

1 2 3 4 5

Very easy Moderately easy Not at all easy

40) How much of a problem is it to manage the daily routines for CF now?

No problem A constant problem

a) Mealtimes—getting him/her to eat enough — 1 2 3 4 5

b) Digestion – tummy problems (wind, pain, diarrhoea) — 1 2 3 4 5

c) Taking enzymes/creon — 1 2 3 4 5

d) Taking vitamins/oral antibiotics — 1 2 3 4 5

e) Doing physiotherapy — 1 2 3 4 5

d) Doing nebulised medications — 1 2 3 4 5

No problem A constant problem

41) With all the things that need to be done, it may be overwhelming at times. How true has this been for you over the last 2 weeks?

1 2 3 4 5

Very true Neutral Not at all true

42) Do you think doing all these treatments for your child are justified?

1 2 3 4 5

Completely justified Not sure Not at all justified

Please specify any treatment you have a question about?

_____

Community support

43) What quality of relationship do you have with your local GP/surgery?

1 2 3 4 5

Very good Moderately good Not at all good

44) How helpful is your local pharmacist?

1 2 3 4 5

Very helpful Moderately helpful Not at all helpful

45) What sort of relationship do you have **with your child's minder/nursery/school?**

1 2 3 4 5

Very good Moderately good Not at all good

ID Number: ☐

46) How comfortable are you with how your **child's minder / nursery, or school gives** medications to your child?

1 2 3 4 5

Very comfortable Moderately comfortable Not at all comfortable

47) Do you get support **from your child's** minder/nursery or school above and beyond creon or inhalers e.g., physio or nebs? Yes ☐ No ☐

If yes, what kind of support? *Please specify*

CF Clinic & Pharmacy Visits

48) How many clinic visits has your child had since last research point? ☐ visits

49) How long on average do you spend at the clinic? (Number of hours) ☐ hours

50) How frequently does your child usually attend the CF clinic? every ☐ weeks

51) How often do you use the hospital pharmacy? Most visits ☐ Sometimes ☐ Rarely ☐

52) Please think about your last visit to pharmacy. How acceptable was the wait for medicines?

1 2 3 4 5

Very acceptable Very unacceptable

53) How consistent are the messages you get from different members of the CF team?

1 2 3 4 5

Very consistent Moderately consistent Not at all consistent

54) How much information would you like to **have from the CF team about your child's condition** or treatments?

1 2 3 4 5

More information The same as now Less information

ID Number: ☐

Inpatient and Day Patient Stays

55) How many times was your child with CF admitted to hospital over the last three months for a day or more? ☐

What was it for?

56) Was this the first time your child was admitted to hospital because of his/her CF? Yes ☐ No ☐ **Don't know** ☐

57) Was this admission routine/preventative or for treatment/intervention for symptoms?

Routine/prevention ☐ Treatment ☐ N/A ☐

58) How stressful was this admission for you and your family?

1 2 3 4 5

Very stressful Stressful Not at all stressful

59) Here is a list of types of stress you may have **experienced during your child's admission.** Please rank them in order of their stressfulness.

Not applicable because my child has not had an admission in the last three months. ☐

Type of stress during last admission:	Rate 1-6 for stressfulness with 1 high and 6 low
• staying in over night	☐
• disruptions to family life	☐
• getting good care in hospital	☐
• getting the intravenous line in	☐
• **child's loneliness**	☐
• communication with health care professionals	☐

other type of stress _____

ID Number:

CF Treatments

In answering the questions on this and the next two pages please consider your responses over the past two weeks

60) Over the last two weeks how much has your child needed the following treatments to keep him/her well?	SECTION 1A Prescribed? *Please tick a circle. If yes, complete Section B & C.*			SECTION 1B Treatment Taken : Time Required *Please estimate the time spent doing this treatment*		SECTION 1C Treatment Taken : Effort Required *Please tick the circle that represents the amount of effort required to do each treatment*		
Please tick one circle next to each question in section A and estimate time taken (section B) and effort (section C) involved	Yes	Yes but not done	No	Minutes per day doing the task	Number of days of treatment over past week	Minimal Effort	Moderate Effort	High Effort
Calculating doses	O	O	O			O	O	O
Extra day time feeding /calorie supplements	O	O	O			O	O	O
Inhalers	O	O	O			O	O	O
Insulin injections for diabetes	O	O	O			O	O	O
IV antibiotics at home	O	O	O			O	O	O
IV antibiotics in hospital	O	O	O			O	O	O
Nebulised medications:								
Antibiotics	O	O	O			O	O	O
DNAse	O	O	O			O	O	O
Hypertonic Saline	O	O	O			O	O	O
Salbutamol	O	O	O			O	O	O
Non-prescribed (alternative remedies e.g., herbal remedies)						O	O	O
Oral antibiotics (back-up & specific)	O	O	O			O	O	O
Other medicines (lactulose, antacids, vitamins etc)	O	O	O			O	O	O
Overnight feeds through a gastrostomy or nasogastric tube	O	O	O			O	O	O
Oxygen therapy delivered by mask or nasal specs	O	O	O			O	O	O
Pancreatic enzyme Supplements (creon)	O	O	O			O	O	O
Physiotherapy	O	O	O			O	O	O
Ursodeoxycholic acid (URSO) for liver involvement	O	O	O			O	O	O
Collecting & preparing medicines and cleaning equipment	O	O	O			O	O	O

ID Number: ☐

61) We want to know how hard it has been for YOU to manage these treatments. *(Please tick one circle from Section 2A for each treatment type that applies and then consider Section 2B)*	Section 2A: How hard has it been for YOU to manage these treatments?				Section 2B: Please tick the circle next to any treatment that YOU would like to talk about the next time you come to clinic. This may be how it is done or whether it is relevant.
	Very Difficult	Somewhat Difficult	Not at all Difficult	Does not apply	Yes, I would like to talk about this treatment at the next clinic or annual review
Extra feeding, calorie supplements	O	O	O	O	☐
Inhalers	O	O	O	O	☐
Insulin injections for diabetes	O	O	O	O	☐
IV antibiotics at home	O	O	O	O	☐
IV antibiotics in hospital	O	O	O	O	☐
Nebulised medications:					
Antibiotics	O	O	O	O	☐
DNAse	O	O	O	O	☐
Hypertonic Saline	O	O	O	O	☐
Salbutamol	O	O	O	O	☐
Non-prescribed remedies (e.g. alternative remedies)	O	O	O	O	☐
Oral antibiotics (back-up & specific)	O	O	O	O	☐
Other medicines (lactulose, antacids, vitamins etc)	O	O	O	O	☐
Overnight feeds through a gastrostomy or nasogastric tube	O	O	O	O	☐
Oxygen therapy by mask or nasal specs	O	O	O	O	☐
Pancreatic enzyme supplements (creon)	O	O	O	O	☐
Physiotherapy	O	O	O	O	☐
URSO for the liver	O	O	O	O	☐

ID Number:

62) How do you think YOUR CHILD has managed these aspects of the CF routine over the last two weeks? *(Please tick one circle from Section 3A for each treatment type and then consider Section 3B)*	Section 3A: How hard has it been for YOUR CHILD to manage these treatments?				Section 3B: Please tick the circle next to any treatment that YOUR CHILD would like to talk about the next time you come to clinic. This may be how it is done or whether it is relevant.
	Very Difficult	Somewhat Difficult	Not at all Difficult	Does not apply	Yes, my child would like to talk about this treatment at the next clinic or annual review
Extra feeding, calorie supplements	O	O	O	O	☐
Inhalers	O	O	O	O	☐
Insulin injections for diabetes	O	O	O	O	☐
IV antibiotics at home	O	O	O	O	☐
IV antibiotics in hospital	O	O	O	O	☐
Nebulised medications:					
Antibiotics	O	O	O	O	☐
DNAse	O	O	O	O	☐
Hypertonic Saline	O	O	O	O	☐
Salbutamol	O	O	O	O	☐
Non-prescribed remedies (e.g. alternative remedies)	O	O	O	O	☐
Oral antibiotics (back-up & specific)	O	O	O	O	☐
Other medicines (lactulose, antacids, vitamins etc)	O	O	O	O	☐
Overnight feeds through a gastrostomy or nasogastric tube	O	O	O	O	☐
Oxygen therapy by mask or nasal specs	O	O	O	O	☐
Pancreatic enzyme supplements (creon)	O	O	O	O	☐
Physiotherapy	O	O	O	O	☐
URSO for the liver	O	O	O	O	☐

THANK YOU

9.2.d. Investigator feedback sheet

UNIVERSITY OF LIVERPOOL

Alder Hey Children's NHS
NHS Foundation Trust

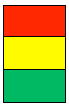
Feedback Form Information

What does this form mean?

The following form has been completed using the information you gave in the Challenges of Living with Cystic Fibrosis-Questionnaire.

A selected group of responses have been statistically analysed and have been allocated a score. This score can be found under the heading, "Score."

The scores have then been graded using the following key.



This statement is false
This statement is sometimes true
This statement is true

This form should give you an idea of how well you are coping with the challenges of living with cystic fibrosis.

What happens next?

You will be given a copy of this form and a copy of, "Self Feedback Form." to take home and review.

The results on this form should be compared to your results on the, "Self Feedback Form."

If you are concerned about your score and would like to know how you could face the challenges better you could talk to someone on the Cystic Fibrosis Care Team.

On the far right column under the heading, "Who can I talk to?" are a list of members of your child's Cystic Fibrosis Care Team who you can talk to, to discuss your results and any concerns you may have.

Domain		Score	Scale	Who can I talk to?
Family Lifestyle "We work well as a family"	"We share challenges"		True / False	Doctor CF Nurse Psychologist
	"We work together"		True / False	
CF Background "My child's CF is well managed"			True / False	Doctor CF Nurse Physiotherapist Dietician
Child's Character "My child is well behaved"			True / False	Doctor Psychologist
Challenges of Family Life "We face CF together as a family"	"I have a lot of support"		True / False	Doctor CF Nurse Psychologist
	"We don't worry about infections"		True / False	
	"CF doesn't impact on our life too much"		True / False	
Hopes and Worries "My hopes for everyday life are bigger than my worries about everyday life"	"I am hopeful about our day to day life"		True / False	Doctor CF Nurse Psychologist Physiotherapist Dietician
	"I don't worry about everyday life"		True / False	
Hopes and Worries "My hopes for the future are bigger than my worries about the future"	"I have a lot of hope for the future"		True / False	
	"I don't worry about the future"		True / False	
CF Routines "Managing my child's CF is easy"	"We have a good routine"		True / False	Doctor CF Nurse Psychologist Physiotherapist Dietician
	"I never feel overwhelmed"		True / False	
Community Support "I have a good relationship with the CF team in the community"			True / False	Doctor GP Pharmacist School/Minder
CF Clinic and Pharmacy Visits "I find it easy getting my child's medication"			True / False	Doctor Pharmacist CF Nurse
Inpatient and Day Patient Stays "I am happy with the amount of admissions my child has had"			True / False	Doctor CF Nurse Psychologist
CF Treatments "I am happy with my child's CF treatment"			True / False	Doctor CF Nurse Dietician

Notes:

Feedback Form for Carers
Feedback

Thank you for taking the time to review this feedback form.

Do you have any queries or comments regarding this form or this study?

If you would like to discuss your queries or concerns please contact Miss Tulsi Patel on;

Institute of Child Health
Alder Hey
Eaton Road
Liverpool
L12 2AP

E-mail: t.p.patel@student.liverpool.ac.uk
Tel: 0151 228 4811 Ext 3536
Mobile: 078 288 199 69

9.2.e. Caregiver self-feedback form

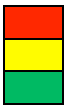
UNIVERSITY OF LIVERPOOL

Alder Hey Children's NHS
NHS Foundation Trust

Self Feedback Form for Carers
Instructions

The following form is designed to assess how well you think you are coping with the Challenges of Living with Cystic Fibrosis.

Please respond to the statements by ticking one box in the right column. You should choose your response using the following key.



This statement is false
This statement is sometimes true
This statement is true

The answers should be your own and should reflect how you feel, no one else. Do not spend too long thinking about the answers.

At your next appointment you will be given a copy of this form to take home.

Domain		Scale
Family Lifestyle **"We work well as a family"**	**"We share challenges"**	True / False 
	"We work together"	
CF Background **"My child's CF is well managed"**		True / False 
Child's Character **"My child is well behaved"**		True / False 
Challenges of Family Life **"We face CF together as a family"**	**"I have a lot of support"**	True / False 
	"We don't worry about infections"	
	"CF doesn't impact on our life too much"	
Hopes and Worries **"My hopes for everyday life are bigger than my worries about everyday life"**	**"I am hopeful about our day to day life"**	True / False 
	"I don't worry about everyday life"	
Hopes and Worries **"My hopes for the future are bigger than my worries about the future"**	**"I have a lot of hope for the future"**	
	"I don't worry about the future"	
CF Routines **"Managing my child's CF is easy"**	**"We have a good routine"**	True / False 
	"I never feel overwhelmed"	
Community Support **"I have a good relationship with the CF team in the community"**		True / False 
CF Clinic and Pharmacy Visits **"I find it easy getting my child's medication"**		True / False 
Inpatient and Day Patient Stays **"I am happy with the amount of admissions my child has had"**		True / False 
CF Treatments **"I am happy with my child's CF treatment"**		True / False 

Self Feedback Form for Carers Feedback

Thank you for taking the time to complete this feedback form.

Do you have any queries or comments regarding this form or this study?

If you would like to discuss your queries or concerns please contact Miss Tulsi Patel on;

Institute of Child Health
Alder Hey
Eaton Road
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L12 2AP

E-mail: t.p.patel@student.liverpool.ac.uk
Tel: 0151 228 4811 Ext 3536
Mobile: 078 288 199 69