WHAT CHILDREN WITH CYSTIC FIBROSIS KNOW ABOUT THEIR ILLNESS

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DECLARATION

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SECTION A: PREFACE

The theme that links the work I present in this dissertation is that of communicating about cystic fibrosis. Section C, the main body of research, involves the development of a booklet of questions aimed at accessing the knowledge that children with cystic fibrosis have of their illness. The purpose of this is to aid communication between adults and children, to ensure that information can be presented to children at an appropriate level, and that children can be involved in decision making about their treatment. Section B, the literature review focuses upon genetic screening for cystic fibrosis. Many of the papers examined discuss issues of communication within the field of genetics, exploring how to convey complex features such as genetic probability and risk to a number of different populations. Finally, Section D is a case study of work that I carried out with a family whose child with cystic fibrosis had stopped going to school.

The seeds of my interest in this field were sown when I joined the South West Thames regional paediatric cystic fibrosis team at Queen Mary's Hospital for Children, Carshalton, in April 1996. The service had not had a clinical child psychology post attached to it previously, and many of the families and patients were somewhat wary of a psychologist joining the team. Paediatric psychology is not yet a well-established profession. For many people, the fact that cystic fibrosis (CF) is a physical illness makes the presence of a mental health professional in the team anxiety provoking. Concerns that were voiced to me by patients and family members included: the fear that seeing a psychologist would imply that they were not coping successfully with CF, a fear of being intruded upon and judged, a fear of being

diagnosed with a mental illness or of having their children taken away, a fear of being seen as weak, and a fear that by talking about CF they might 'open up a can of worms'.

However, after having written an explanatory leaflet about the role of a clinical child psychologist within a CF team and attending CF clinic on a weekly basis, I began to be approached by people during their clinic appointment for advice and support. What interested me in particular was that the approach, which usually came from parents, often took the form of a question about communicating with their children, or other people, about CF.

The parents of younger children, in particular those at transition points such as going to a child minder, starting school etc, requested advice on how to prepare their children for this change and how to cope with potential questions and comments from 'outsiders'.

Mid school aged children were often asking their parents questions that the parents felt ill equipped to answer, such as why they have to spend time on treatment and their peers do not, why they have CF but their siblings do not, and what the future holds for them. One mother described how she nearly crashed her car as she was travelling in the fast lane of the M25 when her eight-year-old daughter asked, apparently quite cheerily, from the back seat "Mummy, is cystic fibrosis going to kill me?"

Many adolescents and their families were struggling with issues around adherence to treatment. Parents would describe having warned their child about the consequences of failing to follow the treatment regime but that this did not seem to have much effect. Other parents talked proudly about the way in which their teenager coped with the treatment requirements, but expressed concerns about their emotional wellbeing saying that they rarely talked about what it was like living with cystic fibrosis.

Consultations with colleagues also often centred around issues of communication. Questions would be asked such as 'when should we discuss fertility issues?' 'Does this behaviour indicate that the child does not understand what is happening?' 'How much should we say to this family about the child's physical deterioration?'

I often found myself discussing with adults, either parents or health professionals, where a child might be in terms of his or her understanding about their illness, and the impact that this might be having upon their behaviour and psychological well being. I would suggest that the starting point of any discussion with a child would be to find out how much they knew about CF and their current state of health. However, having suggested this I began to realise how hard it is to access children's knowledge about CF. Simply going in to a session with the child and asking direct questions about their understanding of CF felt inappropriate, turning a therapeutic session into a test and putting at risk the therapeutic relationship. Information could be obtained from talking with the child as part of a psychological intervention, but the process could be time consuming and the areas of knowledge covered limited. For example, in an intervention with a young child helping them to take digestive enzymes before eating it would be relevant to explore their understanding of the nature of CF and its

treatment requirements, but it would be most unusual to start discussing fertility issues.

No measures for accessing children's knowledge of CF were available, those that had been used previously were lengthy and detailed questionnaires that had been designed for use with adults and teenagers. From my discussions with children and adolescents with cystic fibrosis I felt that there was a wealth of knowledge and understanding about the condition that might not be captured by a questionnaire or set of closed questions. Instead, I began to wonder if there might be a way of accessing this information so as to allow children to express their understanding in their own words.

My main study 'What Children with Cystic Fibrosis Know about Their Illness', which is presented in Section C, does therefore explore whether it is possible to design a tool to access children's knowledge of CF. I want to see if the same tool can be used across the age range of 4 to 18, and if the information gathered can be used to examine how knowledge develops with age. Nowhere in the CF literature have I been able to find guidelines for parents or clinicians about what might be reasonable to expect a child of a particular age to understand about their illness. While much research has shown that age alone should not be used to determine a child's understanding of illness, parents and professionals still need guidance about what to expect, and given appropriate caveats, age can be a useful way of presenting developmental information. I also hope that the tool may have a future as a clinical aide, providing a new approach to exploring potentially distressing issues with children and young people with chronic illnesses.

The literature review that I present in Section B stems from a somewhat more personal experience. I took some leave from my job in the autumn of 1999 to accompany my husband while he spent three months working in San Francisco. I was pregnant with our second child and therefore arranged to receive antenatal care from within the American health care system. During my second visit to the obstetrician, she took a blood sample and told me that this was to test for genetic abnormalities including Downs syndrome and cystic fibrosis. She had no knowledge of my personal or professional background and provided no counselling and very little information prior to the blood test. I had had blood tests to screen for Downs syndrome during my first pregnancy in England and I readily signed the consent form for these tests. But having left the office I began to dwell upon the possibility that I might be identified as a carrier of a cystic fibrosis mutation. I was aware of becoming quite anxious over the next few days and of thinking about the children and families that I work with. I was able to use my knowledge of the genetics and familial patterns of CF to reassure myself that it was unlikely that both I and my husband would be carriers of a CF mutation. However, I did retain a low level background anxiety until I received the results of the test, which was negative.

This experience began to make me think about the process of screening for cystic fibrosis. It also made me recall a piece of work that I had carried out with a mother of a child with CF, who amongst other things, wanted to talk to me about whether to try for another baby. I felt, with hindsight, that I was not as knowledgeable as I should have been about the screening process. Within the South West Thames region, couples with a family history of CF are referred to St George's Hospital for specialist genetic counselling. It is not appropriate for psychologists working within a clinical

CF team to provide such input. However, we should be aware of the provision that is available and the psychological issues involved in making the decision to be screened or not as well as the emotional consequences of such a decision. I therefore decided that I would use the opportunity of carrying out a literature review to help me develop my knowledge and understanding of genetic screening for cystic fibrosis.

Finally, the case study that I discuss in Section D provides an example of how traditional clinical child psychology input can be adapted for use within the field of paediatric psychology. The case presented is of a girl with cystic fibrosis who develops a serious school refusal problem. As with many 'standard' school refusal cases, there were numerous issues such as attitudes towards schooling, peer relationships, family stresses, specific anxieties, and inter-professional tensions that contributed to the girl's difficulties in attending school. However, the fact that she had cystic fibrosis and that she and her family never openly discussed this added a further complicating factor into the equation.

In addition to using cognitive behavioural techniques for helping the girl to manage her anxiety and develop a graded return to school programme, much of the psychological intervention focused around issues of communication. The anxieties within the family in relation to CF were all unspoken or denied. While much attention was given to the girl's physical state, her emotional wellbeing was less of a focus for her parents or the problems were attributed solely to external factors such as being bullied by classmates or picked on by teachers. Addressing the impact of CF upon the family required sensitive management, and only had a limited effect. There were no clinical tools to help provide some structured input around these painful

issues. It is a case where had there been a tool such as the one I designed for my main study, the issues facing people with CF might have been addressed more openly.

In undertaking this doctorate my aim has been to reflect upon my practice, to increase my knowledge of CF, to develop my research skills, and in particular to explore systematically a series of clinical 'hunches' about children's knowledge of CF. The results of the study do suggest that children with cystic fibrosis have a more sophisticated understanding of their illness than might be expected given their age, and that given the right tool they are able to talk clearly and openly about their disease. The advantage of this finding for children with cystic fibrosis is that it should encourage clinicians to talk to them about their condition more openly, to inform them appropriately and to involve them in decision making about their treatment. This should in turn lead them to feel more empowered, confident and respected. However, it should also be noted that until research has examined the relationship between an individual's knowledge of cystic fibrosis and their psychological wellbeing, it can not be assumed that knowledge is entirely positive and the potential risks of increased knowledge such as increased anxiety should not be dismissed automatically.

Despite advances in the field of genetics, a cure for cystic fibrosis remains a distant prospect. Cystic fibrosis continues to be a chronic, life limiting illness, requiring complex and time consuming daily treatment coupled with periodic invasive and distressing medical interventions. The impact of this upon children and families must be immense. The fact that most of those affected by this illness cope with it so successfully is a huge credit to the individuals and their families. The presence of a clinical psychologist on a CF team should never be seen to take away from this

achievement. However, for people living under these circumstances it is natural that at times they may need the additional support, advice and input that a psychologist can provide. My hope is that this doctorate will, in some small way, add to the growing body of evidence of the importance of involving clinical child psychologists in teams providing care to children with cystic fibrosis and their families.

SECTION B: A LITERATURE REVIEW OF GENETIC SCREENING FOR CYSTIC FIBROSIS

CHAPTER ONE: INTRODUCTION

The CF gene was discovered in 1989 on chromosome 7. Since then more than 600 CF causing mutations have been discovered. From a simple blood test up to 85% of these can now be identified within an individual. As a result the opportunities for screening individuals for CF mutations have grown rapidly. Screening can be provided within the womb, at or shortly after birth, or at any other stage of life. The implications of screening for CF can be profound and genetic counselling is widely recommended to precede any decision to undergo the screening process. In this review I shall examine:

- The debate around the nature of genetic counselling
- The different techniques for CF screening
- Attitudes towards CF screening
- Knowledge and behaviour in relation to CF screening
- The psychological impact of CF screening.

Cystic Fibrosis

Cystic Fibrosis (CF) is the most common genetic disorder in Northern Europe. It is an autosomal recessive disorder that causes dysfunction at a cellular level, affecting the transport of chloride across the cell membrane. Extracellular secretions and mucus become thick and dehydrated, they can not be moved through the body efficiently and eventually block many of the body's organs. Thick mucus also

becomes attractive to bacteria and fungi thus massively increasing the individual's susceptibility to infection and disease. The main organs to be affected by CF are the lungs, pancreas and gastrointestinal tract. Treatment for CF has developed considerably over the last few decades, increasing the life expectancy for an individual with CF from 2 years in the 1940's to 30. 1 years in 1995. It is predicted that babies born with CF in the 1990's will have a life expectancy of over 40. However, although encouraging, these figures must be looked at in the light of increasing life expectancy figures within the general population, and it should be acknowledged that the treatment involved can be both time consuming and stressful.

The Genetics of Cystic Fibrosis

The defective gene responsible for Cystic Fibrosis, the CF Transmembrane Conductance Regulator (CFTR), was identified in 1989 on the long arm of chromosome 7. As a recessive genetic disorder, carrying one defective allele has no impact upon an individual's health, and CF only occurs when both of the alleles are mutations. Since the gene was identified more than 600 mutations that can cause CF have been found and the number continues to grow. However, one mutation, Delta F 508 is thought to account for up to 70% of the cases of CF, at least within the Caucasian population. Another 15-20 mutations account for a further 15% of cases of CF. Thus, within the Caucasian population over 80% of the CF causing genetic mutations can now be identified. The incidence of CF within other populations is significantly lower than the Caucasian population, as is the detection rate for the defective CFTR gene. For example, the incidence of CF within the Asian population is 1 in 32,000 births but only 30% of the mutations are identifiable. Over 25000

Americans have Cystic Fibrosis and it has been estimated that there are now over 6000 Britons with CF (Webb & David, 1994).

Summary

Cystic fibrosis is a genetic disease that occurs when both alleles of the CF Transmembrane Conductance Regulator gene are defective. This leads to disruption of the absorption of chloride at a cellular level and a thickening of the mucus throughout the body. Since the discovery of the CF gene in 1989 it is now possible to identify over 85% of the genetic mutations that cause CF within the Caucasian population. Thus the option of genetic screening for CF is now in place and can occur pre or post natally or within the general population. The following chapters explore the issues surrounding genetic screening and the provision of genetic counselling for CF in particular.

CHAPTER TWO: GENETIC COUNSELLING

With the growth in understanding of genetic conditions and the development of genetic testing, demand has grown for genetic counselling. However, little is known about the theoretical backgrounds or practical approaches used by genetic counsellors. Michie and Marteau (1995) define genetic counselling as a communication process aimed at helping people with problems associated with genetic disorders or the risk of these in their family. They add that its most uncontroversial goal is to improve the quality of life of the families that seek such help. Fraser (1974) provided a more specific definition of genetic counselling, which according to Michie and Marteau has had a wide-ranging influence upon the field of genetic counselling. The process of counselling should, in Fraser's view, help an individual or family to:

1. Comprehend the medical facts, including the diagnosis, the probable course of the disorder and the available management;
2. Appreciate the way heredity contributes to the disorder and the risk of recurrence in specified relatives;
3. Understand the options for dealing with the risk of recurrence;
4. Choose the course of action which seems appropriate to them in view of their risk and their family goals and act in accordance with that decision; and
5. Make the best possible adjustment to the disorder in an affected family member and/or to the risk of recurrence of that disorder.

The principle of non-directive counselling has been a defining factor within the field of genetic counselling. Michie and Marteau point out that all professional medical bodies dealing with genetic issues have adopted it as their key principle. The aim of a

non-directive approach is to ensure that while individuals or families are provided with information, the decisions that are made about whether to test, which tests to have, and how to act upon the results are made freely and independently by the counsellees. However, not only are there questions about how effectively individual counsellors follow a non-directive approach, there is also a debate within the field of medical genetics as to whether non-directive genetic counselling is even possible.

Clarke, in a Viewpoint article in The Lancet, 1991, contends "that an offer of prenatal diagnosis implies a recommendation to accept that offer, which in turn entails a tacit recommendation to terminate a pregnancy if it is found to show any abnormality". He goes on to argue that this occurs because of the social context and attitudes towards genetic abnormalities and is irrespective of the counsellor's individual attitudes or behaviour. He suggests that there is a fundamental inconsistency in bodies such as the Royal College of Physicians who argue that the role of genetic screening is to reduce the incidence of genetic disease, while at the same time offering impartial and non directive support to families who come for testing. In Clarke's opinion "the Holy Grail of non-directive counselling is unattainable ... the offer and acceptance of genetic counselling has already set up a likely chain of events in everyone's mind". However, despite the debate, genetic counselling, including that for cystic fibrosis, is flourishing.

Summary

Genetic counselling within the CF screening field is aimed at helping people make informed decisions about whether to screen for CF, how to respond to and cope with the result that they receive, and to consider their future reproductive plans. A

non-directive approach is the core principle within genetic counselling for CF, and yet little is known about the approaches used by the individuals who provide such a service, or the impact that they have upon their clients. The field of genetic counselling is one that is likely to be a target for further research in the future.

CHAPTER THREE: SCREENING FOR CYSTIC FIBROSIS

Screening for Cystic Fibrosis can occur in a number of ways including prenatally, neonatally and through large scale general population screening. Ryley, Goodchild & Dodge (1992) reviewed the nature of the tests available.

Prenatal testing involves obtaining a chorionic villus sample (CVS) from the mother's uterus at about 10 weeks gestation. The CF gene is then examined and specific mutations identified. However, Ryley et al. (1992) point out that "it is not uncommon for one parent in a CF family to carry an unidentifiable CF mutation thus making antenatal diagnosis impossible". They go on to suggest that genetic counselling should be provided prior to future pregnancies in such families and that "personnel who are thoroughly familiar with the disease and its ramifications" should carry it out. What they fail to mention is the risk involved in the initial CVS procedure that can often be as high as a 1 in 100 chance of miscarriage, regardless of the status of the foetus.

Neonatal screening should be, according to Ryley et al. (1992) "specific for CF, quick, simple and inexpensive". However, they then add that no such test is available. The definitive test for the disease is the sweat test in which the elevated levels of sodium and chloride ions within a child's sweat are identified. The procedure to collect sufficient amounts of sweat to be tested although not invasive can be uncomfortable and time consuming. Results can often be equivocal resulting in repeat tests over time, and highly trained personnel are required to carry out the procedure. Consequently the test is "not suitable for mass routine screening. The

approach therefore has been to filter out most normal children using less specific methods and to apply the sweat test to the few who are positive on initial screening".

The test used in mass screening of newborn infants is the blood immunoreactive trypsin test (IRT). It has been recognised that neonates with CF have two to three times the level of IRT in their blood compared to non-CF newborns. Tests to pick up IRT levels have been developed to be used on dry blood spots. Thus "routine IRT screening for CF could be integrated easily into existing neonatal screening programmes for the detection of phenylketonuria and hypothyroidism" (Ryley et al., 1992). The debate as to whether to introduce mass neonatal screening for CF is raging at present. Charities, patient pressure groups and some clinicians argue strongly for the introduction of mass blood spot screening. They suggest that early diagnosis improves the long term outcome for children with CF, reduces the psychological distress involved in the often long search for a diagnosis in those infants who are not tested at birth, and allows families to make informed choices about their future reproductive plans (Ravenscroft, 1996).

Others have argued that the demand for mass neonatal screening cannot be justified (NIHC, 1999). They are particularly influenced by research that has failed to "provide sufficient evidence that identifying CF patients earlier than the current average age of diagnosis improves outcomes". They also argue that the psychological effects upon those families whose babies are recalled for further tests after a first positive IRT test should be considered.

Population screening has also become a potential option for health authorities and clinicians to consider. Having identified the CF gene it is now possible for individual carrier status to be examined. An individual's carrier status can be examined from a simple mouth swab or a blood sample. However, as only approximately 85% of the CF causing mutations can be identified within the Caucasian population this means that individuals cannot be given a clear message that they are not carriers of a CF mutation. Instead they will be given information about "reduced risk", a concept that is notoriously difficult for people to understand fully.

By 1999 the National Institutes of Health Consensus Development Conference concluded "interest in CF genetic testing is limited in the general population and that agreement to participate in genetic education and testing procedures occurs primarily among pregnant women and persons with family histories of genetic disorders". The final statement from the conference did not advocate CF testing for the general population "given the low incidence and prevalence of CF and the demonstrable lack of interest in the general population". They did however recommend that screening be made available to certain target populations including those with a family history of CF, couples planning a pregnancy, couples seeking prenatal care and partners of people with CF.

Summary

Most genetic screening for CF occurs during pregnancy or shortly after birth, although the option of more widescale population screening is also available. Prenatal testing carries with it a risk of miscarriage and cannot always guarantee that the foetus is free of cystic fibrosis. Neonatal screening is not yet sufficiently refined

to be able to distinguish between babies with raised IRT levels who have CF and those who do not, resulting in the recall and further testing of babies without the disease. Similarly population screening cannot confirm unequivocally that an individual is not a carrier of a CF causing mutation, providing instead a more complex message of a lower likelihood of carrying a CF gene. However, despite the elements of risk and uncertainty associated with CF screening, it has been recommended for certain target groups within the USA (NIHC, 1999) and in 2001 the British government recommended that a national programme of neonatal screening for CF should be introduced.

CHAPTER FOUR: PSYCHOLOGY OF SCREENING

The main purpose of screening programmes according to Marteau (1994) is to detect disease or an increased risk of disease in order to reduce the morbidity and or mortality associated with the disease. However, a second goal is to reassure those for whom no problem is detected. A third aim is to help ensure that those people with positive results make the best possible adjustment following the new information. Exploring these issues from a public health viewpoint would suggest that participating in genetic screening programmes would confer advantages, such as reassurance of their low risk status, upon the vast majority of individuals and would therefore lead to a high level of participation within such programmes. However, an individual perspective acknowledges that there can be disadvantages as well as advantages stemming from participation in genetic testing. Studies have shown that the response of individuals to test results do not according to Marteau (1994) "easily fit the stimulus-response model" in which negative or low risk results predict low levels of distress while positive or high risk results produce high levels of distress and subsequent behaviour change. She describes studies that have shown that individuals with negative results have increased levels of distress post testing and "experience problems of a similar magnitude to those receiving high risk results" (Tibben, Vegter, Niermeijer, Kamp, Roos, Rooijmans, Frets & Verhage, 1990). Other studies have shown that test results whether positive or negative do not predict levels of distress six months after testing (Tibben, 1993). Reactions to positive results can also be more extreme than would be expected given the implication of the test results. For example, carriers of the recessive gene for Tay Sachs disease have been shown to

worry more about their own future health post testing despite having been informed that carrier status has no risk to their health whatsoever (Marteau, van Duijn & Ellis, 1992). On the other hand, some studies have shown that individuals given results indicating that they are at high risk of coronary heart disease due to their levels of cholesterol fail to contact a doctor because they forget the results or think that they are unimportant (Schucker, Bailey & Heimbach, 1987).

Psychological models can help with the planning and implementation of screening programmes. They can also aid the interpretation of individual responses to test results, and the assessment of the overall effectiveness of such programmes. Evaluating the psychological impact of screening is a complex process covering issues such as:

- The recall and interpretation of information,
- The attitudes and expectations of both counsellors and patients,
- The pre and post screening levels of adjustment of the patients,
- The decisions made or behavioural changes implemented by the patients following their test results.

Summary

The response shown by individuals to their genetic test results can be more complex than traditional psychological models would predict. Some individuals informed of their low risk status have been shown to display ongoing high levels of distress (Tibben et al., 1990) while others receiving high risk results have been found to have

difficulty in recalling their results or acting upon them (Schucker et al., 1987). Psychological models are being used to examine the responses of those who undergo genetic screening and to evaluate the overall impact and effectiveness of genetic screening programmes.

CHAPTER FIVE: ATTITUDES TOWARDS CF SCREENING

In 1999 the report from the US National Institutes of Health Consensus conference on CF screening (NIHC, 1999) stated that public knowledge of CF and interest in genetic testing for CF is low and mainly limited to women who are pregnant or people with a family history of CF. It also commented upon the complexity of the concepts involved in understanding genetic testing and its results such as "test sensitivity, carrier status, patterns of inheritance, risk/probability, and genotype phenotype correlations". However, the studies discussed in this chapter do suggest that knowledge about CF and the screening process can be successfully conveyed to a wide range of people.

High School Students

Durfy, Page, Eng, Chang & Waye assessed the knowledge and attitudes of high school pupils towards screening for cystic fibrosis in 1994. A questionnaire was given before and after a lesson on the clinical features of CF, its treatment, the mode of inheritance, and methods of screening for it. The authors described as low the 40% success rate of students answering questions about CF prior to the lesson. However, given that they are children, with no direct experience of the disease, and that relatives of people with CF (Lafayette, Abuelo, Passero & Tantravahi, 1999), and medical practitioners (Rowley, Loader, Levenkron & Phelps, 1993), show considerable gaps in their knowledge of CF, this figure could be considered surprisingly high. The information session led to 90% of the pupils correctly answering most of the questions. The finding would be more robust if the pupils had received the knowledge questionnaire again at a later date to determine how much information had been

retained, particularly as reproductive decision making is likely to be some time ahead for most high school students.

The questionnaire assessing the students' attitudes towards screening was presented again, four months after the lesson on CF. During the intervening period all the 101 students provided mouthwash swabs for analysis for carrier status. The results, which indicated that three pupils were carriers of the most common CF gene mutation, were presented to the class just before the questionnaire on attitudes was readministered. The pupils were found to be positive in their attitudes towards screening for CF. Nearly 90% of the pupils said that screening should be available to the general population even though rare mutations might be missed, and that they would have themselves tested if there was no charge. Over 60% of the pupils indicated that they would be tested even if there was a small cost involved. On the other hand, only 13% of the participants thought that during high school was the best time to screen someone for being a carrier of a genetic disease. In addition, a striking feature of this study was that only 5 of the 101 students tested actually requested their own results, a finding that seems to contradict their positive attitudes towards testing. The authors comment upon aspects of the design of the study that may have contributed to this, but the hypothesis that they focus upon is that the low rate of requesting information about carrier status reflects a high level of student disinterest. The authors briefly acknowledge that "being identified as a carrier could be a frightening or worrisome prospect to some" but fail to give this the attention it deserves, particularly given that they found an "increased uncertainty toward carrier screening and prenatal diagnosis for CF after learning class results". They also fail to address some of the ethical issues raised by their study such as the lack of counselling prior to testing, the

responsibility they hold about informing the three students with a positive carrier status or the residual distress that might be experienced by the group following the information that three of them are carriers of the CF gene. Instead they simply conclude, "more research is required to determine whether high school students are an appropriate target population for generalised screening for CF carrier status".

College Students

The knowledge and attitudes of college students towards the clinical and genetic features of CF were investigated by Neiger, Abuelo & Passero in 1992. Twenty-five college students were given a seventeen item questionnaire on CF to complete three weeks prior to attending a lecture on the disorder. At the end of the lecture the questionnaire was readministered. Eight questions covered students' knowledge of CF and there were significant increases in knowledge scores on seven of the eight items after the lecture. The questions covered issues such as the availability of carrier testing, population screening, newborn screening, and the course of the illness. The other nine questions explored the attitudes of the students towards genetic testing for CF and the decisions they would take about terminating a pregnancy if the foetus was found to have CF. The students showed very positive attitudes towards genetic testing for CF. 19 of the 25 indicated that they would take a test of their carrier status, 22 would test their foetus for CF, and all 25 would test their newborn baby for its carrier status. In addition, 15 of the 25 students said they would abort a foetus with CF.

The lecture did not produce any significant changes in attitudes towards genetic testing and aborting a foetus diagnosed with CF. However, fewer students were

interested in neonatal testing or terminating an affected pregnancy after the 80-minute lecture. While none of the students had any exposure to cystic fibrosis, ten of them had experience of other chronic illnesses within their families. Of the students with a family experience of chronic illness, only 60% would consider a pre-pregnancy genetic test to help them make decisions about family planning compared to 100% of the students with no experience of chronic illness. This would suggest that exposure to a chronic illness within a family member could create a greater ambivalence in attitude towards genetic testing than is felt by those with no experience of illness.

Family History of Cystic Fibrosis

People with a family history of cystic fibrosis have been identified as a high risk, target population for screening for CF. However, clinical observation has suggested that the uptake of screening amongst relatives of people with CF has been surprisingly low. Lafayette et al. (1999) chose to explore this phenomenon. They carried out a study of the knowledge and attitudes towards genetic testing for CF amongst close relatives of people with CF attending a specialist clinic at Rhode Island Hospital. 238 relatives (siblings, half siblings, uncles and aunts) were identified as being at high risk of being carriers of a CF mutation. A questionnaire covering knowledge of the clinical and genetic aspects of CF, attitudes towards genetic testing and pregnancy termination in general and specifically in relation to CF, as well as their perception of the burden of bringing up a child with CF, was sent to the 173 relatives for whom addresses were supplied. In addition, these relatives were invited to undertake free carrier testing via mouthwash.

The response rate to this questionnaire was 47% (75 forms were returned). Of those who responded 60% underestimated their risk of carrying the CF mutation but at the same time 63% overestimated their chance of having a child with CF. Almost all respondents (93%) indicated that they would be interested in carrier testing yet only 29% actually took up the offer of testing. Most (70%) indicated that they would use prenatal testing of a foetus if they and their partner were both carriers, but only 6 (8%) said that they would abort a foetus with CF. In their discussion Lafayette et al. (1999) write "most relatives do not have a working knowledge of autosomal recessive inheritance... and that outreach educational and genetic counselling efforts by medical professionals to at risk relatives are needed". They conclude that individuals with CF and/or their parents are unable to convey the complex information involved to their relatives and "it is unreasonable to expect them to do so".

The authors also commented on the results indicating that "the intention to use prenatal testing does not seem to be linked to the intention to terminate a pregnancy if the foetus were found to have CF". This finding concurs with that of the Neiger et al. (1992) study in which students with a family history of chronic illness were less likely to indicate that they would terminate an affected pregnancy than students with no family history of illness. Lafayette et al. (1999) admit that the lack of anonymity in their study may have made relatives feel uncomfortable in recording their true attitudes towards terminating an affected foetus. They also suggest that the relatives may have been under the impression that prenatal diagnosis could affect the management of the pregnancy and the outcome for the baby, and that they may not understand the risk of miscarriage involved in prenatal testing. They also acknowledge that for some people the desire to know whether the foetus is affected

may be important and they may not wish to wait until the end of the pregnancy to find out. This seems to be given secondary importance as an explanation, and yet the chance to avoid months of uncertainty, to receive reassurance (in most cases), and to prepare for a child with a chronic illness (in a few cases), seems instinctively powerful. The authors themselves comment "unfortunately we did not ask what their reasons for using prenatal diagnosis would be". This would be an interesting area to explore further. It is also of note that studies in which affected foetuses have been identified have shown high rates of pregnancy termination (Mennie et al., 1993; Brock, 1996). The implication of this is that while the concept of aborting an affected foetus seems unthinkable to many people in theory, when faced with the reality of bringing up a child with a serious life shortening illness, a more pragmatic approach is taken.

The results from the Lafayette et al. (1999) study create an interesting dilemma for those researchers and clinicians who argue that screening for cystic fibrosis should be introduced on grounds of cost effectiveness. Mennie et al. (1992) carried out a "crude cost-benefit calculation", in which they argued that "even if only half the parents of an affected foetus chose termination, this would still lead to the reduction in live born incidence of 1 case of CF for every 10,000 tests". Their figures suggested that the total cost for 10,000 tests is £80,000, while the lifetime cost of treating an individual with CF is roughly £125,000 and therefore prenatal screening "represents good value". If the figures from Lafayette et al. (1999) are used the "good value" argument is lost. It does seem that the purpose of screening is seen quite differently by service users and service providers, even when the providers are in favour of introducing screening programmes.

Health Professionals

However, a study of the knowledge and attitudes of obstetricians and family practitioners in Rochester, New York towards CF screening raised concerns about the attitudes of professionals to CF screening (Rowley, Loader, Levenkron & Phelps, 1993). All doctors in the region with obstetric responsibilities were informed by letter about the study and told that it would be the focus of their next weekly staff conference. Those staff who attended the meetings (which are supposedly obligatory) were given a CF knowledge questionnaire followed by a presentation about CF carrier screening in which they were offered a free full screening service for all their patients. Their attitudes towards screening were then assessed with a second questionnaire. Only 81 out of an expected 124 doctors attended the meetings. They had considerable gaps in their knowledge of CF. Only a quarter knew the average life expectancy for a person with CF, and fewer than half knew the incidence of CF. Rowley et al. (1993) concluded that their "misunderstanding of CF inheritance ill fits them to be genetic counsellors for CF".

In addition, while over two thirds of the doctors had positive attitudes towards carrier screening, agreeing with the statement "if screening is free, all women of reproductive age, pregnant or not, should be offered screening", 14% of those who attended the presentation said that they were not willing to participate in the study. Coupled with the 43 doctors who did not attend the presentation (35% of providers of antenatal care), almost 50% of women in the area would not be offered screening as part of their care. These figures would be likely to increase further once the free screening services provided for the study are no longer available. Once again, Rowley et al.

(1993) concluded that they were concerned by the attitudes and behaviour of service providers which make it hard to "translate advances in molecular medicine into prenatal screening services".

In contrast, Modell, a British general practitioner wrote in the British Medical Journal (1993) that general practice has a "pivotal role" in screening for cystic fibrosis. He argued that "general practitioners are family doctors and family studies form the basis of genetic practice". The vast majority of the population is registered with a GP, most practices have an age-sex register that can provide a database for screening, and GPs are usually the first health professionals approached to discuss family planning issues. Modell stated that the role of a general practitioner in CF screening was to:

1. Train a member of the practice team to be able to give basic genetic counselling
2. Provide information to those offered testing elsewhere
3. Offer testing when relevant to life situation
4. Counsel single carriers
5. Provide cascade screening for relatives of patients and carriers
6. Refer carrier couples for genetic counselling
7. Support couples after termination of pregnancy.

He concluded that "if carrier testing is focused on relevant groups in the practice population then in spite of the recent NHS reforms, which have added considerably to the workload of general practitioners, general practice will be able to accept a central role in cystic fibrosis screening". It is hard to know how far this paper represents the general view of family doctors within Britain or whether it is an entirely personal opinion presented by one general practitioner. But it does present a more positive

professional response to the issue of CF screening than has been identified within the United States.

Summary

Studies looking at high school students (Durfy et al., 1994), college students (Neiger et al., 1992), and people with a family history of cystic fibrosis (Lafayette et al., 1999) have all found these groups to have positive attitudes towards genetic testing for CF. A study of doctors in Rochester, New York (Rowley et al., 1993) also identified that over two thirds of the doctors taking part had positive attitudes towards CF screening. However, a positive attitude towards screening did not translate into a full use of the service (Lafayette et al., 1999) or a willingness to provide the service (Rowley et al., 1993). Those with a family history of CF or other chronic illnesses while positive in their general attitude towards CF screening were found to be less likely to consider terminating an affected pregnancy than those with no previous exposure to a chronic illness. However, when faced with the reality of an affected foetus the rates of termination have been high (Mennie et al., 1993; Brock, 1996). This would suggest that it might be inappropriate to draw conclusions about the actual use of screening services from studies that explore attitudes towards screening on a hypothetical basis.

Considerable gaps in CF related knowledge were identified within all groups studied, including groups of doctors and family members. However, those studies that included an educational component (Durfy et al., 1994; Neiger et al., 1992) found that knowledge scores about CF could be significantly increased by providing a teaching session. Whether such knowledge can be retained over time was not examined and this would be a useful area for future research.

CHAPTER SIX: THE IMPACT OF SCREENING FOR CYSTIC FIBROSIS

Prenatal Screening

Brock's 1996 review of five years of CF screening in Edinburgh compares two ways of delivering screening services in antenatal clinics. In the two-step model the woman is tested first and her partner is only tested if she is found to be a carrier. Couple screening on the other hand tests both partners at the same time and they are only informed of their high-risk status if both are found to be carriers. While women were positive in their attitudes towards two step screening, with 83% of those eligible taking up the offer of testing, those who were identified as carriers (4% of those tested) had "considerable short term anxiety with the consequent need for counselling".

The clinicians found that it was essential to employ a specialist full time genetic counsellor when offering the two step method of screening, and Brock (1996) concluded that this has major cost implications for nationwide screening of this type. Couple screening on the other hand "has the potential to be highly economic". Only those couples with a 1 in 4 risk of having an affected foetus are informed of their status, cutting the need for counselling from 4% to 0. 1%. Brock (1996) writes that in this case "counselling can easily be provided by obstetricians seeing couples for discussion of prenatal diagnosis". The implication of this is that no specialist counselling services would be required. Yet most obstetricians have not had specific training in counselling skills, may well be seen by patients as having an opinion however non directive they may try to be, and are unlikely to have sufficient time to

devote to the counselling process. Specialist counselling services should still be available for all those at risk of an affected pregnancy and should not be seen as an expensive or replaceable luxury.

While Brock (1996) addressed the practicalities of service delivery, Mennie et al. (1992) used the same population to assess the psychological impact of screening. Women were asked to complete the twelve item general health questionnaire (GHQ). This short form of the GHQ is a useful screening tool for identifying stress in individuals but does not provide any detail about the nature of their distress. Within the two step screening model 53% of women who were identified as carriers of the CF gene had positive scores on the GHQ at the time they received their results compared to 31% of women who were informed that they were not carriers. Once the women who were carriers received the news that their partners were not carriers their scores on the GHQ returned to the same level as the non-carrier women.

In their paper Mennie et al. (1993) examined the psychological impact of screening upon couples in which the woman tested positive for a CF mutation and her partner's test result was negative. Due to the incompleteness of carrier screening whereby 15% of mutations cannot be identified, these couples "face a 1 in 640 risk of having an affected child, substantially greater than their starting risk of 1 in 2500". These couples were asked to complete the short form of the GHQ (Goldberg & Williams, 1988) and the Symptom Rating Test (Kellner & Sheffield, 1973) which can provide separate scores for depression, anxiety, inadequacy and somatic symptoms. As reported before, there was no significant difference in GHQ scores between women who were carriers and those who were not at the time of booking into clinic. There

was a significant difference at the point where the women were informed of their carrier status, but this was then wiped out once their partners received a negative test result. There were no significant differences in GHQ scores six weeks after the test or six weeks after the birth of the baby. The male partners of the female carriers and their controls showed no difference in GHQ scores at any of the four assessment points.

Scores from the SRT indicated that female carriers had significantly higher generalised psychological disturbance, anxiety and depression in comparison to non-carriers when informed of their test results. However, as with the GHQ, the scores of the carriers returned to control levels once they received their partner's negative test result and remained at this level through future testing points. When the female carriers were informed of their status, their partners were found to have significantly higher scores of anxiety and inadequacy compared to the partners of controls. The anxiety scores of the partners returned to control level upon receiving their own negative test results, but the inadequacy scores did not change significantly. Mennie et al. (1993) acknowledged that "pre-natal carrier screening delivered in two stages does generate some psychological disturbance". But they went on to point out that the GHQ and SRT scores were "well below those reported for psychiatric patients", a finding that differs from previous studies on screening for neural tube defects (Robinson, Hibbard & Laurence, 1984). They do not comment in their conclusion about the residual high level of inadequacy scores amongst the partners of women who carry the CF mutation. This would obviously be an area to explore further before stating categorically that any distress associated with screening is short lived for both women and men.

Bekker, Denniss, Modell, Bobrow & Marteau (1994) carried out a similar study looking at the short term psychological impact of screening for cystic fibrosis amongst adults of reproductive age attending a GP practice in inner London. They found that people receiving a positive test result showed higher levels of anxiety than those with negative test results. However this anxiety had dissipated after three months even without a partner being tested for their carrier status. However, Bekker et al. (1994) were concerned by their findings with regard to understanding the test results. They found that 17% of those receiving a negative test result believed that this meant they had no chance of having a child with CF. While five out of fourteen carriers (36%) thought that they "probably, but not definitely, carried the gene for cystic fibrosis". They concluded "in the longer term the greatest problem of population screening would appear to be one of false reassurance rather than anxiety".

The UK Cystic Fibrosis Follow–up Study Group examined the long term effects of carrier screening in a three year study (Axworthy, Brock, Bobrow & Marteau, 1996). They sent questionnaires to all those individuals found to be carriers of the CF mutation from six British health centres and maternity hospitals that had carried out screening in the early 1990s. They also approached two matched non-carrier controls for each carrier. The questionnaire explored understanding of test results, perception of health, reproductive intentions and behaviour, feelings about the test result and anxiety. They found that "3 years after testing 20% of carriers and 50% of those with a negative test result cannot accurately recall the result". As with previous studies the inaccuracies of recall were to do with understanding that the result was definite for carriers but indefinite for those given negative results. Axworthy et al. (1996)

suggested that this might be due to "the way people process information about risks. When given a probability, many people simplify it- either it will happen or it won't happen- perhaps in an attempt to reduce the cognitive effort needed to process probabilities". In addition it could be "consistent with a tendency for people to recall information that is personally threatening in a way that reduces the threat".

There were no differences between carriers and screen-negative participants in terms of general anxiety, but there was a significant difference between them in terms of their perception of their health. CF carriers perceived their current health to be significantly poorer than controls. There were no differences in the reproductive plans or behaviour of carriers and non-carriers. The authors argued that these findings should not deter the continuation or initiation of CF screening programmes, but they do "highlight the need for more research on the psychological processing of risk information and its association with health behaviours".

Studies in America have identified similar difficulties in explaining the results of CF screening to the general public. Tatsugawa et al. (1994) interviewed and screened women in the first 18 weeks of pregnancy who attended hospital for their antenatal care. Prior to being given information leaflets or videos about CF and genetic screening, the women had some awareness of the chronic and serious nature of CF but little understanding of its genetics. The educational intervention improved their knowledge scores significantly but there remained serious misunderstandings. In particular only 72% of those interviewed recognised that the statement "carriers might develop CF" was false and only 52% understood that CF testing couldn't detect all CF carriers. 10% of those who received a negative test result "still had the misconception

that their risk was now zero". The authors described these results as "disturbing" and concluded that further information and counselling would have to be provided to these individuals. They also used these results to make "modifications to our educational materials and result letters to further stress the point that a negative test result does not eliminate the chance that a person could be a CF carrier". Whether simply providing more information to this group of people will have an impact upon either their understanding or their behaviour remains questionable.

How to convey information about CF screening was investigated by Leonard, Bartholomew, Swank & Parcel (1995). They wanted to see whether they could "enhance knowledge of cystic fibrosis and risk status more effectively using social cognitive theory generated role modeling vs. a more traditional informational approach". They designed two brochures, the first was a traditional one with information about CF, its treatment, risk of being a carrier, availability and nature of screening etc. The second provided a role model in which the same information was given but with the story of a family with a child with CF interspersed throughout. The brochures were alternated on a weekly basis within a hospital antenatal unit, and were given to pregnant women and their partners upon arrival. A questionnaire was developed to assess knowledge of CF, health beliefs and attitudes towards genetic testing. The results suggested that the modeling component enhanced people's perception of the risk of having a child with CF and the severity of the disease. It also reduced the perception of barriers to testing. However, there was no difference between groups when it came to deciding whether to be tested or not. Approximately 12% of subjects from each group decided to pursue testing. This figure is very low, especially in comparison to the British studies (Mennie et al., 1992; Bekker et al.,

1994) which achieved uptake figures above 70%. The authors acknowledge this discrepancy, but fail to account for it adequately. They suggest that it may be that women would be more positive in their attitudes towards testing prior to being pregnant, and yet the Mennie et al. (1993) study was carried out on pregnant women attending a hospital antenatal unit.

A qualitative study exploring the experience that relatives of people with CF had when counselled and tested for CF carrier status was carried out by Callanan, Bloom, Sorenson, DeVellis & Cheuvront (1995). Telephone interviews and focus groups took place with ten couples who had been screened for CF at their request. Their experiences were positive. They described their counsellors as knowledgeable and able to give clear explanations about genetic issues. They appreciated the neutrality of the counsellor and recognised the role as one of educator and identifier of important issues that could not be addressed in other settings. Those participants who were informed of their carrier status described themselves as "disappointed but not surprised" and only one woman reported any long term negative effects i. e. concern about her own health and the impact of her carrier status upon her relationship. Couples had significant knowledge of CF owing to their exposure to it within their families, and their knowledge of reproductive issues and genetics post-counselling was accurate. Callanan et al. (1995) did recognise that their subjects might not be representative of most relatives of people with CF given that they had requested screening themselves and agreed to be interviewed about the experience. However, they argued that "genetic counseling prior to testing for individuals and couples with a positive family history of CF is a successful model for providing effective education and counseling".

The Health Belief Model

According to O'Connor and Cappelli (1999) the Health Belief Model (HBM) might help to explain individual responses to screening for CF. The HBM hypothesizes that motivation to take preventive health action depends upon an individual's perception of their susceptibility to the disease, the disease severity, the effectiveness of taking any action and the obstacles or barriers to taking such an action. O'Connor and Cappelli (1999) explored the health beliefs of a group of healthy adults and their impact upon screening for cystic fibrosis carrier status. As in previous studies (Durfy et al., 1994; Leonard et al., 1995), the results of this study indicated that the vast majority of people thought that the public should be informed about the availability of carrier screening but only a minority (40%) indicated that they would be interested in finding out their own carrier status. Health Belief Model variables did appear to have some influence over this pattern of response. Somewhat surprisingly perceiving CF to be a very severe illness was associated with a reduced likelihood of accepting carrier screening. The authors suggested that while a "moderate estimation of severity is necessary to prompt individuals to take action, the perception that being a carrier would be highly disruptive of future family planning may have an inhibiting effect on participation". They suggested that there may be an "optimal level of fear" necessary to prompt action, and that education programmes designed "to scare individuals into seeking carrier testing may actually be contraindicated". In addition, those participants who indicated that they would be interested in pursuing carrier testing for themselves saw fewer barriers and more benefits to testing than those who did not want to be tested. However, as the authors point out this study asked

participants hypothetical questions about their use of genetic screening. The impact of the Health Belief Model upon motivation to seek carrier testing would be more accurately assessed in a 'real life' situation.

Newborn Screening

Fewer studies on the impact of neonatal screening have been carried out because as Mischler et al. (1998) point out it is a controversial area "primarily because of uncertainty regarding whether early diagnosis improves the long-term prognosis". Thus those studies evaluating newborn screening have tended to focus upon physical issues. Green et al. (1993) reviewed the cystic fibrosis neonatal screening programme in East Anglia. They commented upon the fact that despite an increase in birth rate of 20% in the region during the time of the study, the incidence of cystic fibrosis halved. They found that newborn screening had an impact upon family size, with a strong birth order effect. They concluded that "early diagnosis and counselling have discouraged some couples from having more children". While this was obviously a medical paper covering specific genetic issues, it is interesting to note that the psychological impact of screening was not even mentioned.

Mischler et al. (1998) explored in more depth the impact of a state wide neonatal screening programme in Wisconsin upon reproductive decision making. Questionnaires were sent to the families of all infants who had been given a sweat test as well as all those who were diagnosed with CF. Following education and counselling the families with children with cystic fibrosis showed excellent understanding of the genetics of CF and the risks of future children having the disease. They retained the information successfully in the course of a year. They

were also aware of the availability of prenatal testing but only 26% of those who had a second pregnancy during the course of the study chose to test the foetus for CF, and the three affected foetuses were all carried to term. The authors write that the reasons for this pattern of reproductive behaviour are too complex to be fully addressed within their paper, but they do acknowledge that "the implications of prenatal diagnosis and possibly being faced with a decision about abortion have unique complexities for families with a child who has CF".

Wertz, Rosenfield, Janes & Erbe (1991) explored the attitudes towards abortion of parents of children with cystic fibrosis. They found that 20% of parents of children with CF would abort an affected foetus. Most parents felt that the option of prenatal testing and selective abortion should be available to all families affected by CF, but that it was not an acceptable option for themselves. They would however abort a foetus with severe mental retardation or one that would be bedridden for life. Thus the severity of a condition appears to be a significant factor in the decisions these families make about abortion. However, these families will be making reproductive decisions at a time when their child is likely to be least affected by CF, not yet facing the physical and psychological struggles that develop with age. The authors found that parents did have high levels of optimism about CF. They expected their child to reach the age of 40, to have a full time job and to marry. In addition, few parents believed that their CF doctors would support the abortion of a foetus with CF. Wertz et al. (1991) wondered whether this may "reflect the clinically necessary optimism conveyed by paediatricians, rather than the doctors' actual views" and they recommended that counsellors discuss these issues with families. It would have been interesting in this study to have had a measure of illness severity and family history of

CF to investigate whether experience of the more serious effects of CF has an impact upon reproductive decision making.

Within the Mischler et al. (1998) study those families whose child had a false positive screening result i.e. who were called for sweat tests which then proved negative, understood that their child did not have CF but carried a single CF mutation. However, some were confused and thought that their child could be susceptible to developing CF. In addition, those parents who chose to have themselves tested for carrier status did not understand that their risk of having a child with CF was greater than that of the general population. Indeed those who were tested and found not to be a carrier of a common mutation had great difficulty in comprehending that there remained a residual risk. In a similar analysis to Axworthy et al. (1996) Mischler et al. (1998) concluded that "relative risk perception is a difficult concept for people who normally do not think about probabilities". They go on to say that further research needs to address "whether there are alternative genetic counseling approaches that are more effective in conveying this information". But in the mean time they stated "it is essential that centres doing the definitive sweat testing have counsellors available to explain the genetic implications for the newborn's family, parents and siblings, extended family members, and ultimately the newborns themselves when they reach an age at which information is relevant to them".

Baroni and colleagues (1997) investigated the impact of the Wisconsin neonatal screening programme upon parenting stress. They wanted to look at the effect of both false positive IRT test results, and early diagnosis of asymptomatic infants. In an exploratory, pilot study, parents were asked to complete the Parenting Stress Index

(PSI), a well established and frequently used psychological measure. The PSI scores for families that had experienced a false positive test result suggested that parents may display hypervigilance and emotional repression, indicating that they rather than their child may be quite highly stressed despite a relatively low PSI score. No differences in levels of parenting stress were found between parents of children with CF who had been diagnosed early through the blood spot technique or those whose children were diagnosed through traditional methods. The stress levels for both groups of CF parents were similar to those of parents of healthy controls. However, there were several individual families with high-risk scores that were subsumed within the whole group statistics, so the authors suggested that within-group variation needs to be explored in order to obtain clinically useful information. While the results were only those of an initial pilot study, Baroni et al. (1997) did suggest that even at this stage there is a need for parenting stress to be assessed within a paediatric context. They also emphasised the need for services to be targeted not only towards those families with CF, but also the families of the healthy children who had false positive IRT tests.

Genetic Counselling for People with CF

The dramatic improvements in the treatment of cystic fibrosis that have increased the life expectancy for a baby born in the 1990's to 40 means that reproductive issues for individuals with cystic fibrosis must now be considered. Given that any child of a person with cystic fibrosis will inevitably carry one mutant allele, genetic counselling and screening of the partner becomes a high priority. Little has been written about this small population, but Lemke (1992) did write a paper exploring the implications for genetic counselling when an adult with CF considers having a child. 97-98% of men with cystic fibrosis are infertile while many women with CF also experience

reduced fertility and have difficult pregnancies. Thus, the decision to have a child is complicated not only by one parent having a life limiting chronic illness but also by probable difficulties in conception and carrying a pregnancy to term. Lemke (1992) identified a number of issues that a genetic counsellor would need to address with CF adults and their partners. Couples would need to be reminded about the nature of autosomal recessive inheritance and the risks of having a child with CF. If the partner of the person with CF is not a carrier of an identifiable CF mutation, there remains a 2% chance that the baby will have CF, while the chance is 50% if the partner is identified as a carrier. The risks and benefits of testing a foetus need to be discussed as do the requirements of caring for a baby whether it has CF or not. An additional issue that she highlights is that of paternity of a child when the male partner is the one with CF. She writes that "because most males with CF are infertile, the genetic counsellor may want to have a private conversation with the pregnant woman to discuss the paternity of the pregnancy. If the man with CF is not the father of the pregnancy, it is highly likely that this will be detected". The psychosocial issues that Lemke (1992) comments upon include concerns about body image, sexuality and sexual function, as well as "issues related to their own health and shortened lifespan". She concludes that "the genetic counsellor must meet the challenge to present the most accurate and current information available, while at the same time remain sensitive to the concerns of CF adults". This is an area of counselling that requires great skill and expertise, addressing so many highly emotional issues as it does. It is therefore unfortunate that there do not appear to have been any papers since 1992 exploring this field. Feedback from the couples as well as more detailed studies of the psychological issues around CF and reproduction would be extremely valuable.

Summary

British studies have found that following CF screening there is an increase in psychological distress, particularly anxiety and depression, when women are informed that they are carriers of the CF gene. However, this distress is short term, dissipating naturally within a three month period (Bekker et al., 1994) or upon being informed that their partner is not a carrier of the most commonly identified CF genes (Mennie et al., 1993). Being informed of one's carrier status is however associated with an increased likelihood of long term concern about one's own current health status (Axworthy et al., 1996).

The British studies also examined the way in which people understood their results at the time they received them and remembered their results over time. There were difficulties with the recall of results three years after testing (Axworthy et al., 1996) and concerns about the understanding of results at the time they were received. For example, some people given a negative test result did not grasp that their likelihood of carrying a CF gene was reduced but not eliminated completely and others informed of their positive carrier status did not understand that this was a certainty.

The results from a US study of the Wisconsin neonatal CF screening programme (Mischler et al., 1998) also found that some individuals had difficulty understanding the concept of relative risk for themselves or the impact of carrier status upon their child's health.

These results would suggest that while CF genetic counselling should continue to provide emotional support to those given 'bad' news, it also needs to develop its

educational component to address issues such as the recall of information and the accurate interpretation of test results.

CHAPTER SEVEN: CONCLUSION

The decision to screen for cystic fibrosis, whether it be neonatally, prenatally, or within the general population is highly complex. Debates continue to rage about what form of screening should be provided, when and to whom it should be offered and how best to maximise the positive effects and limit the negative consequences of such a course of action (NIHC, 1999; Modell, 1993; Ravenscroft, 1996; Rowley et al., 1993). Studies have shown that the public attitude towards CF screening is somewhat ambivalent. While a wide range of people including high school students, college students, pregnant women, relatives of those with cystic fibrosis, and members of the general public have indicated that they think carrier screening and prenatal testing should be made available to all, fewer participants within these studies have indicated that they would accept such offers of screening for themselves (Durfy et al., 1994; Neiger et al., 1992; Mennie et al., 1992; Lafayette et al., 1999; O'Connor & Cappelli, 1999). However, hypothetical choices about genetic screening and pregnancy termination that are made in the course of a research study which indicate an antipathy towards such a course of action (Wertz et al., 1991) appear to differ from the choices made by the individuals when actually faced with such a decision. The results from the Edinburgh screening programme were that all pregnancies which were found to have an affected foetus were terminated (Brock, 1996).

Similarly, the attitudes of professionals towards screening for cystic fibrosis differ considerably. The consensus statement from the panel of the US National Institutes of Health (1999) did not recommend the universal provision of neonatal screening for CF, while the professionals within the British charity the CF Trust have argued

vociferously for the introduction of such a screening programme (Ravenscroft, 1996). Modell (1996) wrote in the British Medical Journal of the central role that General Practitioners could and should take in the provision of CF screening services to their patients. Yet Rowley et al. (1993) found a considerable lack of knowledge about, and interest in, CF screening amongst doctors providing antenatal care within New York state. Modell (1996) does argue that the emphasis within the British health care system upon family doctors/general practitioners places those professionals in an ideal position with regards to CF screening through their knowledge of the wider family, access to family data, and long term doctor patient relationships. US gynaecologists and obstetricians do not have such a relationship with their patients and may not be able to integrate CF screening into their practice as naturally as Modell suggests could occur within the British system of general practice.

Some studies have explored directly the psychological consequences of screening for cystic fibrosis. In general the results have shown that being informed of one's carrier status produces a small but significant increase in anxiety that dissipates with time or upon being informed of ones partner's negative status (Bekker et al., 1994; Mennie et al., 1992). However, a small minority of individuals remain anxious and concerned about the impact of carrier status upon their current health (Axworthy et al., 1996), as do the parents of infants who are identified as carriers of a CF mutation following neonatal screening (Baroni et al., 1997). In addition, numerous studies (Bekker et al., 1994; Mischler et al., 1998; Tatsugawa et al., 1994) have uncovered poor retention of knowledge and mistakes in the interpretation of test results by those who are informed of their carrier status. The particular difficulties appear to be in relation to understanding that negative test results can never be definite.

Psychological models such as the Health Belief Model (O'Connor & Cappelli, 1999) and Social Learning Theory (Leonard et al., 1995) have been used to try to enhance our understanding of people's uptake and reactions to screening, as well as their memory and understanding of the results. In addition, they have been used to develop educational material and counselling input to counteract negative effects.

Genetic counselling remains a woolly term. There is little consensus about whom should provide it, what it's content should be, or how to evaluate it. Even the purpose of genetic counselling continues to be debated. Is its primary role to reduce the incidence of genetic disorders within the general population or to provide information and support to individuals and families affected by genetic disease? Further work needs to be carried out within this field to establish clear guidelines covering the provision of genetic counselling.

The rapid development in our understanding of genetics, the human genome, and genetic disease, is an immensely exciting aspect of modern medicine. However, for those directly affected by genetic disease, the developments may leave them facing questions and dilemmas that they would not have had to address previously. If we are to ensure that this new field of medicine is not damaging to the psychological wellbeing of many of its potential 'beneficiaries', it is essential that the medical progress is matched by developments in the counselling and psychological inputs to this ever growing group of people.

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ABSTRACT

The aim of this study is to design a tool to access the knowledge that children with cystic fibrosis have of their illness. This will help clinicians to develop an understanding of each child's existing knowledge so that new information can be targeted appropriately and children can be involved in making decisions about their treatment and their future.

A booklet was designed in which a series of questions from another (fictional) child with CF is presented to participants who are asked to help this other child learn more about the illness. Questions covered issues such as the nature of CF, its treatment requirements, the future impact of the illness, the psychological effect of CF upon family members, and the genetics of the disease. The booklet was structured to encourage children to use their own words to describe their experiences of living with CF in an attempt to gather qualitative data from the study. However, a scoring system was also developed to allow a quantitative analysis of the data to take place. The relationship between knowledge, compliance with treatment and family factors was examined, as was the way in which knowledge of CF develops with age.

The booklet was administered to children between the ages of four and eighteen. Young children gave fairly brief responses but displayed an impressive early knowledge base, understanding that CF is an illness they are born with, that will not go away, and that they have to undergo treatment to stay well. After the age of eight children begin to develop an awareness of the genetic nature of the illness, establish a clear rationale for treatment, and recognise the psychological impact of the disease upon themselves and their parents. Adolescence sees a jump in understanding of the impact of CF upon life expectancy, as well as further developments in grasping the nature and genetics of CF and recognising the wide ranging emotional impact that it has upon the whole family. However, gaps in knowledge remain, particularly in relation to issues of fertility.

Knowledge of CF was very strongly associated with age and with cognitive maturity, as would be expected given the cognitive advances made by children between the ages of four and eighteen. Knowledge of CF was not found to be associated with any of the other factors explored in the study.

The booklet was well received by participants and their parents, and suggestions are made for its use as a clinical tool.

SECTION C: WHAT CHILDREN WITH CYSTIC FIBROSIS KNOW ABOUT THEIR ILLNESS

CHAPTER ONE: INTRODUCTION

Children with chronic illnesses were once seen as passive recipients of medical care who needed to be protected from the reality of their situation by the adults around them. The traditional, Piagetian, view that children are limited in their understanding of illness by their level of cognitive development may unwittingly have contributed to them being under informed or excluded from making decisions about their treatment. However, recent progress in the understanding of developmental processes ensures that children are now being recognised as active participants in all aspects of their care. "Contemporary views seek not only to inform children, but also to involve them in decision making about treatment" (Eiser, 1994). Rushforth (1996) argues that there is scope for involving even very young children in decision making and management of their illness. She states that "The key would appear to be exploration and acknowledgement of the child's existing level of understanding and previous experience, which is then built on by careful explanation and constant re-checking of understanding". The main aim of this study is to investigate whether a tool can be developed to help this exploration by identifying a child's existing level of understanding of cystic fibrosis.

1. 1 Cystic Fibrosis

Cystic fibrosis (CF) is the most common directly inherited disease in Britain. Approximately 1 in 2500 babies born has CF, and one in twenty five of the Caucasian population is a carrier of the CF gene. The disease affects the exocrine glands of the body, which produce abnormally thick mucus that in turn impacts upon the functioning of many of the major organs of the body. The most seriously affected organs are the lungs and the pancreas. The lungs are unable to move the mucus through them as rapidly as required. It therefore accumulates within the lungs, attracts bacteria and fungi thus causing obstruction and infection. Over time the lungs become permanently damaged and respiratory failure is the main cause of death in 90% of CF patients. The pancreas also becomes blocked by mucus. The production of digestive enzymes is reduced, and those that are produced have difficulty reaching the colon. Thus most CF patients are unable to digest fat, protein or fat-soluble vitamins. Other organs that can also be affected include the liver and the reproductive system.

The treatment for Cystic Fibrosis is rigorous and time consuming. It is aimed at preventing exacerbation of infections and maintaining general functioning. It can include chest physiotherapy two to four times daily, prophylactic oral and nebulised antibiotics, inhaled anti-inflammatory medication, up to 30 digestive enzyme capsules to be taken prior to each meal or snack, supplementary fat soluble vitamins, and a high calorie diet (patients are thought to require 150-200% of a healthy person's recommended daily intake of calories). Any deterioration in functioning can result in an increase in frequency and intensity of treatment, which often takes place in

hospital. While great strides have been made in the understanding and treatment of CF, there is still no cure for it. The life expectancy for a person with CF has increased from 18 years in 1976 to 30 years in 1995, with a prediction that babies born with CF in the 1990s will live into their forties (NIHC, 1999).

1. 2 Coping with Cystic Fibrosis

When a child is diagnosed with Cystic Fibrosis, both the child and the family "must adjust to multiple demands and potential stresses"(Stark, Jelalian & Miller, 1995). While traditional commentators have talked about the psychological maladjustment likely to be associated with having a chronic childhood illness, more recent work has emphasised the coping skills of children and families living with chronic illness (Wallander & Varni, 1998). As Stark and her colleagues wrote "our current knowledge of children's adjustment in CF is not indicative of global difficulties in functioning or of increased overall pathology, but rather of specific areas of vulnerability and stress". Similarly, many families adjust very successfully to living with a child with cystic fibrosis, but others struggle to adapt at certain times (Wilson, Fosson, Kanga & D' Angelo, 1996), and all families have increased levels of childcare tasks in comparison to families of healthy children. Ievers and Drotar (1996) found that parents of children with CF experience more stress than parents of healthy children. They spend more time on childcare and medical activities, with less time available for recreation. Mothers of teenagers with CF have a significantly higher level of depression compared to mothers of healthy controls (Bywater, 1981). Adolescents with CF also show a tendency towards higher levels of depression than age matched healthy controls, and 35% have been found to be non-compliant with their treatment regime (Czajkowski & Koocher, 1987).

Koocher, McGrath & Gudens (1990) attributed non-adherence in CF to a number of factors including inadequate knowledge, psychosocial resistance and educated non-adherence. Henley and Hill (1990) presumed that "compliance will be enhanced by a broad, factual, and applied knowledge of the condition (CF) and its treatment". Nolan, Desmond, Herlich & Hardy (1986) commented in their paper upon uncontrolled studies that "suggest or imply that disease knowledge contributes to the development of patient independence, treatment compliance, disease control, efficient family functioning, and attainment of a realistic and appropriate life-style". Lask (1992) wrote "it has been suggested that CF patients resist information about their disease as a way of defending against the threat of a fatal illness. Equally it has been claimed that knowledge contributes to independence and compliance. However, very few studies have investigated how much CF patients and their relatives know about their disease".

1. 3 Educating patients about cystic fibrosis

In their study of adult patients' knowledge of cystic fibrosis, Conway, Pond, Watson & Hamnett (1996) discussed a number of reasons why children with the disease might be poorly informed about it. These included parental anxieties about a) expressing complex feelings, b) discussing problems related to everyday life with CF, or c) sharing knowledge of sensitive issues such as survival and reproduction. As a result, Conway et al. (1996) hypothesised that parents may wait to be approached by their child who in turn may experience very similar anxieties about discussing their disease. Similarly, professionals may resist discussing the more serious manifestations and implications of the disease with their patients. "The multi-

disciplinary nature of cystic fibrosis care may result in no team member taking responsibility for informing patients about specific issues". In addition "as the children grow and their understanding increases, parents and professional carers may neglect to upgrade appropriately information given to the younger patient". Nolan et al. (1986) on the other hand, suggested that "sensitive practitioners understandably resist repeatedly reminding patients with cystic fibrosis of their disease and its possible consequences; instead they rely on patients approaching them for information". They acknowledged that as a result there can often be confusion about whose role it is to inform patients about cystic fibrosis, and that patient education is haphazard. Indeed they went further in claiming that "a conspiracy of silence around these difficult problems may develop, which may in turn perpetuate ignorance among both patients and their parents, and lead to tensions and other difficulties within the family".

Altschuler in her book Working with Chronic Illness (1997) writes that "children are aware of the seriousness of illness from a young age, and given the opportunity, are keen to discuss what they know". She goes on to provide a number of recommendations about how we can "assist children in exploring their thoughts" and yet she does not suggest that the starting point should be to discover what the child already knows and understands. It is likely that when being informed about their illness children and young people may be given material that is either already very familiar to them, or too complex, or unwanted at that particular time. Perrin and Perrin (1983) have identified that most paediatric healthcare professionals "pitch much of their information-giving at mid-school age level, irrespective of the age, knowledge level or previous experience of the child". Rushforth (1996) interviewed

paediatric nurses about their practice when talking to children about their illness and she found that less than 50% of them would find out what the child already knows before giving them their own explanation. And yet as Rushforth said "establishing a child's existing level of knowledge as a baseline for information giving is fundamental to good practice". The purpose of this study is therefore to try to develop a tool for exploring the existing knowledge of children and young people with cystic fibrosis.

CHAPTER TWO: THEORETICAL BACKGROUND

2. 1 Stage based theories of children's understanding of illness

Studies of the way in which children develop an understanding of illness have been closely linked to general theories of developmental psychology, and in particular that of Piaget. Just as Piaget suggested that a child's cognitive skills develop through a series of stages that occur in a predetermined order and within a specific time frame, so many of the theories of how children develop their understanding of health and illness are similarly stage based.

Bibace and Walsh (1980) asked healthy children a series of questions about how people develop colds. They linked the results of their study directly to Piaget's theory of cognitive development. Thus they argued that children under the age of 6 who are at the pre-operational level of cognitive development understand the cause of illness in terms of phenomenism and contagion i. e. from a remote phenomenon such as the sun or from objects nearby that 'magically' trigger an illness. By the concrete operational stage between the ages of 7 and 10 children understand the causes of illnesses in terms of contamination and internalization i. e. through a harmful external action that then has an impact upon the internal body. Once children reach the formal operational stage beyond the age of 11, they become able to think in terms of physiologic and psychophysiologic explanations i. e. that illness is caused by both external and internal events and involve a breakdown of internal processes.

La Greca and Schuman, (1995) describe very clearly the stages that children are thought to move through in terms of their understanding of illness causation. They write, "Typically preschoolers regard illness causation as 'magical' or as a consequence of 'bad' behaviour. During the early school years, illness is often viewed as a result of specific external agents (such as germs) or of failure to adhere to health rules. During adolescence, a major cognitive shift occurs as the individual becomes more cognizant of the complexities of health and illness, and more sophisticated about the interaction of both internal and external factors in illness recovery. Not until late adolescence or early adulthood, however, does an adequate appreciation of the hypothetical future consequences of illness emerge".

Perrin and Gerrity (1981) wrote "That children develop concepts about health and illness slowly and according to a predictable developmental process seems straightforward". They then asked children between the ages of 5 and 13 questions about the causes, prevention and treatment of illness and followed this with an assessment of their general cognitive level using traditional Piagetian tasks of conservation, transformation, and abstract thinking. Their results indicated that there is a predictable sequence of development of illness related concepts that is "consistent with cognitive development in general, but less sophisticated...as compared to notions about phenomena of the physical world". The developmental sequence that they identified was very similar to that discussed by Bibace and Walsh (1980). Children between 4 and 7 believed that illness was caused by magical events and were often seen as a form of punishment. Early concepts of contagion also began to emerge but young children took it too far and considered all illnesses to be

contagious. 8 to 11 year olds developed a germ theory of illness, recognising that biological agents are involved in the causation of illness, but also becoming aware that not all illnesses are contagious. Early adolescence sees a growth in understanding that specific body parts can become dysfunctional, while later adolescence involves the recognition that psychological factors can influence disease causation and recovery.

Perrin and Gerrity (1981) drew the inference from their study that adults' explanations to children may "assume a more sophisticated understanding of the central concepts than the children actually understand", and that "cognitively advanced concepts can be taught and learned only to a limited extent". In addition, they commented upon other research indicating that hospitalised children have less advanced concepts of illness causality than their healthy peers and they concluded that "educational efforts either not be undertaken during this time (hospitalisation) or be even further scaled down in their level of cognitive sophistication".

Lansdown and Benjamin (1985) identified a similar stage based process of children's learning about the concept of death. They read a story about an old lady who died to 105 children aged 5 to 9. They found that almost all 8 and 9 year olds had a clear and full understanding of the concept of death. Younger children were able to understand concepts such as the separation involved and insensitivity of the body, but did not comprehend the irreversibility or universality of death until the age of 8 or 9. However, the study also identified a considerable proportion of younger children who did appear to have an almost complete understanding of the main components of

death. They concluded that "even a 4 or 5 year old will be able to discuss death in a way that superficial readings of the literature might lead one to imagine is unlikely".

Clunies-Ross and Lansdown (1988) followed this study with one exploring the concepts of death held by children with leukaemia. They did not identify any significant differences in the development of an understanding of death between these children and their healthy peers. However they did notice a trend for younger children with leukaemia to have a better understanding of the cessation of bodily function and irrevocability of death than did healthy children of the same age. They also commented upon the marked individual differences that they noticed within their study which they felt were masked somewhat by the statistical analysis and they advised that anyone working clinically with children with life threatening illnesses take this into account.

Crisp, Ungerer and Goodnow wrote in 1996 that "age is the variable most often used to explain changes in children's understanding of illness. It has the appeal of being easily used in clinical practice, of having been shown empirically to correlate with shifts in understanding, and of being linked to a general theory of cognitive development (Piagetian theory)". However, in 1986 Burbach and Peterson had reviewed the existing stage based studies of children's understanding of illness and wrote that "age is simply not an accurate predictor of cognitive maturity". As a result their review focused only upon the 11 studies "that examined children's illness concepts as a function of cognitive-developmental level". Yet the conclusions that they drew did not seem to make such a powerful distinction between age and

cognitive maturity, "overall, there is a clear relationship between chronological age/cognitive maturity and children's illness concepts".

While age has been found to correlate with developmental shifts in understanding it is important that studies do not simply equate chronological age with cognitive maturity. Instead they need to recognise the individual differences that can occur and include a specific measure of cognitive development. The study by Crisp et al. (1996) illustrates this well. Their study comparing the knowledge of children categorised as illness 'experts' and illness 'novices' found no differences between these two groups within the 10-14 year old age group. However, they also found that the illness 'expert' group had a "deficit in cognitive functioning". This allowed the authors to write "instead of concluding that experience does not enhance level of illness understanding, it could reasonably be argued that experience has a positive influence by maintaining age-appropriate development of illness understanding within the context of deficits in other cognitive domains".

Finding an appropriate measure of cognitive development is a difficult task, and the measures used in some of the stage-based studies have been criticised. Hergenrather and Rabinowitz (1991) identified "methodological difficulties" that made findings from the stage-based studies difficult to interpret. They found that when categorising responses within the Piagetian framework developed by Bibace and Walsh (1980) "similar behaviors are cited as evidence for different kinds of operational thinking". For example, viewing illness as a punishment for rule breaking was categorised as pre-operational thinking by some studies and as concrete operational thinking by others. In addition, Gelman and Baillargeon (1983) found no correlation between

standard Piagetian tasks such as conservation, transformation or perspective taking and concluded "the standard tasks do not provide a good or reliable measure of cognitive level". Crisp et al. (1996) also point out that the Piagetian cognitive model "represents only large changes" in children's cognitive development. They go on to argue that, in addition to chronological age, measures that are sensitive to differences in cognitive functioning need to be used in studies that explore the development of children's understanding of illness. Measures such as mental age or IQ are commonly used as an index of ability (Carr, 1999).

2. 2 Criticisms of stage based theories

Piaget's theory of cognitive development has been criticised for being incomplete and rigid. "Many tests of Piaget's theory have shown that under certain simplified experimental conditions, or following certain enriching developmental experiences or training experiences, children at a given stage can perform tasks that require the cognitive capacities which Piaget's theory attributes to a later stage" (Carr, 1999). The stage based cognitive developmental model of children's understanding of health and illness, has similarly been criticised on many levels. It is seen as a model that focuses upon the limitations of a child's understanding, examining what they do not know rather than what they can achieve (Kendrick, Culling, Oakhill & Mott, 1986). It has also been particularly challenged for failing to take into account the role of experience in contributing to a child's understanding of illness (Carey, 1985). In addition, the stage model is seen as implying that "children's concepts of illness are qualitatively different from those of adults, and such differences need to be taken into account in communications with children" (Eiser, 1994). Eiser goes on to argue that "Developmental changes in concepts of illness may be the result of experience and do

not need to be explained in terms of a 'stage' theory. The fact that children of a particular age discuss illness in a particular way does not imply that they are absolutely unable to understand different kinds of explanations". She suggested that the framework provided by stage theorists, and the descriptions of children's understanding within it, could be accepted without having to conclude that the changes within, represented fundamental cognitive shifts.

2. 3 The impact of experience upon understanding of illness

Goldman, Whitney-Saltiel, Granger & Rodin (1991) felt that previous studies looking at children's representations of illness had focused exclusively upon the concept of causality and failed to explore other aspects of illness that adults understand such as its timing, consequences and cure. They found that while children might not have as full or mature an understanding of these features of illnesses as adults, they still used the same framework of concepts as adults. This is seen as a challenge to the cognitive developmental model, which suggests that a cognitively immature child cannot hold a complex representation of illness. In addition, their finding that 4 to 6 year old children had a more detailed understanding of illness cures than causes was interpreted as a sign that experience may have an impact upon understanding.

Crisp et al. (1996) looked at the impact of experience on children's understanding of illness. They argued that previous studies had failed to specify the type of experience that children had had with illness and that the comparisons between healthy and ill children might not be appropriate. They chose to compare the levels of understanding between chronically ill children who may have experienced frequent hospitalisations (illness experts), and acutely ill children who have only experienced one recent

hospitalisation (illness novices). As they put it, "both groups of children are likely to have given some thought to it"(illness). Overall they found that experience with a chronic illness has a facilitating effect upon the understanding of illness amongst children between the ages of 7 and 10. The conclusions drawn were that assumptions about a child's level of understanding could not be based upon age or cognitive developmental level alone and that the impact of experience could be expressed in different ways depending on factors such as age, development and type of illness.

2. 4 Alternative developmental theories

The studies that have challenged the Piagetian theories of development of children's understanding of illness have instead introduced the idea that appropriate information giving and experience can enhance children's understanding of illness. Eiser (1989) linked these ideas with the developmental theories of Carey which suggest that children and adults have the same underlying schemata for understanding the world, the only difference being the amount of knowledge that they have to develop each schema. Eiser suggested that the most significant factor in determining a child's understanding of illness is his or her acquisition of knowledge rather than the level of cognitive development that the child has reached. Nelson (1986) has argued that "the key to understanding the child's mind, and thus cognitive development, is to be found by examining what children know". In other words, the day to day experience of children, and their structuring of this into scripts, determines how they understand the world. Nelson suggests that these scripts generate expectations about events and that they become more elaborate and detailed with age and experience without undergoing fundamental changes. For example, if a child experiences a serious illness or bereavement, they integrate this into their existing medical script, and display a

greater understanding than would be expected given their cognitive level. Rushforth (1999) has suggested that the developmental theories of Vygotsky may be the most appropriate ones to help make sense of the way in which children's understanding of illness is formed. As she states "Vygotsky recognised fully that at a given point in time there is a limit to what a child can understand. But unlike Piaget, he valued enormously the notion of able instruction". In other words, a child alone may be limited in what he or she can understand by age or cognitive development, but with "instruction", whether this be formal teaching or experience, the capacity for the child to understand a range of concepts may be greatly increased.

2.5 Summary

Traditional studies (Bibace & Walsh, 1980; Perrin & Gerrity, 1981; Lansdown & Benjamin, 1985) exploring the way in which children's understanding of illness and death develops have been based within a Piagetian framework. They have concluded that a child's understanding of illness develops through a number of pre-determined stages, often mirroring those of Piaget's theory of general cognitive development. For example, children at a pre-operational stage in their cognitive development have been found to understand the causes of illness in terms of phenomenism or contagion, as a punishment for bad behaviour or a remote, 'magical' event. Whereas children at the formal operational stage of cognitive development are able to recognise the complexities of illness and understand its causes in both physiological and psychophysiological terms.

However, more recent studies (Kendrick et al., 1986; Goldman et al., 1991; Crisp et al., 1996) have challenged the stage based theories, suggesting that they focus too

much upon what a child does not know as opposed to what they can achieve, and in particular that they fail to take into account the experience of illness that a child has had. Alternative developmental theories have been used to account for differences in the way in which ill and healthy children understand illness. These have included Carey's schema based theory and Vygotsky's theory emphasising the role of 'able instruction' and cognitive 'scaffolding'.

2.6 Conclusion

It is no longer possible to argue that a child of a certain age will be automatically at a particular level in their understanding of illness. Individual differences in terms of the child's level of cognitive development (usually described in terms of mental age or IQ) and experience of illness must also be taken into account before any conclusions can be drawn about a child's understanding of illness. As Crisp et al. (1996) wrote "the only way to be certain of a child's level of understanding appears to be on the basis of individual assessments" made with each child's chronological age, level of cognitive development and experience of illness in mind.

CHAPTER THREE: CHILDREN'S KNOWLEDGE OF SPECIFIC ILLNESSES

3. 1 Juvenile Rheumatoid Arthritis.

The Piagetian framework has been used to analyse the knowledge that children have of specific illnesses. Berry, Hayford, Ross, Pachman & Lavigne (1992) looked at what children with Juvenile Rheumatoid Arthritis (JRA) know about their illness and how their understanding develops with age. They asked 54 children between the ages of 6 and 17 five open ended questions about JRA. These covered issues such as the nature of the illness, its cause, and treatment. A scoring system was developed that was based upon the developmental categories identified by Bibace and Walsh in their 1980 study. Berry et al. (1992) reported that their results indicated that children's conceptual understanding of JRA could be reliably measured and that it could be seen to follow a developmental progression. However, they also noted that "a substantial number" of adolescents who would be expected to display evidence of formal operational thinking with regard to their illness were in fact performing within the concrete operational and sometimes even the pre-logical stage when answering questions about JRA. Similarly, approximately 25% of those children at the concrete operational stage of development were displaying pre-logical forms of understanding about their condition. In addition, within subject variation in understanding was evident depending upon what question was asked, and numerous inaccuracies and misconceptions were evident in the answers provided by the children. The authors

did not provide any explanation for the wide variation in answers that they elicited, but instead concluded "findings from this study highlight the importance of periodically evaluating each child's level of understanding as a means of improving educational efforts".

3. 2 AIDS

Walsh and Bibace(1991) "examined causal reasoning about AIDS in children representing three major phases of cognitive development" and found that the thinking of children about the causes of AIDS reflects their level of cognitive development and parallels the way that they think about illness in general. They used these results to suggest that educational packages should be developmentally based and gave specific examples to back this up.

3. 3 Asthma

Gibson, Henry, Vimpani & Halliday (1995) studied asthma knowledge, attitudes and quality of life in adolescents. Over 4000 children in year 8 at school in Australia filled in an asthma questionnaire during a science lesson. 23% of the children taking part reported that they suffered from asthma and 9% were receiving steroid treatment, which indicates a severe condition. The level of asthma knowledge was generally found to be low, with particular deficits in knowledge about asthma recognition and managing exercise induced asthma. While those teenagers with asthma had significantly higher knowledge scores than those without, they still fared poorly and knew considerably less than informed parents. Higher levels of asthma knowledge were associated with positive attitudes and internal locus of control in asthma. The attitudes of teachers and non-asthmatic teenagers to those with asthma were generally

positive. The authors used this to propose that training and information giving should be provided within the school setting.

A study of British school aged children with asthma, found that they knew little about the causes of asthma and few were prepared to avoid the physical or psychological triggers that were likely to precipitate an attack, such as avoiding homes with pets (Eiser, Town & Tripp, 1988). Another British study undertaken by Finn and Rickard (1996) in which adults' knowledge of childhood asthma was explored found similar gaps and misconceptions in their knowledge. It was suggested that the "knowledge and understanding of asthma could and should be better enhanced during the innumerable contacts that health services have with young asthma sufferers and their parents before they reach adolescence".

3. 4 Diabetes

Studies in diabetes have found that while children may obtain quite high scores on pen and paper factual knowledge tests, their ability to apply this knowledge appropriately is greatly reduced. Thus Johnson, Pollack, Silverstein, Rosenbloom, Spiller, McCallum & Harkavy (1982) found that, despite high knowledge scores, 80% of the children they tested made errors in their urine testing and 40% did not inject themselves correctly. Also, Lorenz, Christensen & Pichert (1985) found that children with diabetes could not select an appropriate meal for themselves from a restaurant canteen.

3. 5 Cancer

Other studies have focussed more upon the links between knowledge and experience of childhood illnesses. As part of a larger study looking at quality of life issues for children with cancer, Eiser, Havermans, Craft & Kernahan (1995) designed a questionnaire to assess the child's perception of their illness experience. Preliminary semi structured interviews were carried out with 15 children from which a set of statements were drawn and compiled into a questionnaire which was then given to 41 other young cancer sufferers. By using such statements the authors argue that they are reflecting the concerns of teenagers with cancer in their own language, and are not just exploring issues thought to be important by adults. Eight general areas of concern were explored including impact upon physical activities, peer relationships and preoccupation with illness. The only subscale in which age had a significant impact upon the answers given by the children was that of physical appearance, in which older children rated the illness as having had a greater effect upon their appearance than younger children.

Sargent and colleagues in their collaborative study in 1995 explored the perceptions of healthy siblings in families affected by childhood cancer. They found that while many of the children brought up issues of concern to them that had been identified in previous studies, they did not complain about a lack of information about cancer or convey a sense of being excluded from medical briefings. The authors concluded that "this may indicate that families and health professionals are becoming more skilled at informing siblings about cancer and its treatment".

Oakley, Bendelow, Barnes, Buchanan & Nasseem Husain (1995) looked at the cancer related knowledge and beliefs of children and young people. Their argument was that traditional forms of eliciting information from children about their understanding of illness were likely to be missing great swathes of children's knowledge. They wrote "there is a need to develop new methods of encouraging children to provide information on the ways in which they see health", as they put it "obtaining data from children requires a special approach". They asked 9 and 10-year-old children to "write or draw anything you know about cancer". Children used both writing and drawing to convey their considerable knowledge of cancer such that Oakley et al. (1995) concluded that drawings are a "valuable research tool".

3. 6 Summary

There are a relatively small number of studies of children's knowledge of specific diseases. Many of those that do exist suggest that children's understanding of their illness is limited, that they have misconceptions about their disease and that their understanding is below what would be expected of their cognitive developmental level (Berry et al., 1992; Walsh and Bibace, 1991; Gibson et al., 1995). It has taken sensitive and original techniques for accessing children's knowledge of specific illnesses (Oakley et al., 1995; Eiser et al.,1995) to start the process of challenging long held views of children's inability to understand their own illness. It is hoped that the new technique for eliciting information from children with cystic fibrosis that is presented in this study may contribute to the growing body of evidence that children are more perceptive about their illness than they have previously been given credit for.

CHAPTER FOUR: KNOWLEDGE OF CYSTIC FIBROSIS

4. 1 Studies of Young Children's Knowledge

There have been few studies of what children with Cystic Fibrosis know about their illness. Those that have been carried out have tended to look at the knowledge of teenagers and young adults rather than that of younger children. Lask (1992) cites studies by Kulczycki and Falkman that explored the knowledge of CF patients between the ages of 4 and 15. Both studies found wide variation in knowledge of the disease. However, the studies were published in 1969 and 1977 respectively and no further studies have been carried out with young children.

4. 2 Studies of Teenagers/Young Adults' Knowledge

The studies exploring the knowledge of teenagers and young adults about cystic fibrosis have taken the form of questionnaires or multiple choice tests.

4. 2. 1 Nolan, Desmond, Herlich & Hardy (1986)

Nolan et al. (1986) designed a questionnaire looking at subjects' knowledge of the disease, its treatment, and genetics/reproductive issues. The authors sought content validity "the extent to which the questionnaire incorporates the domain of CF knowledge" for their measure by appointing "a representative panel of experts consisting of seven clinic staff members ... together with two patients and parents from another clinic". They were individually asked to specify what knowledge about cystic fibrosis was important for patients and "their responses were used as a guide to

formulating items for the questionnaire". The measure included scored components on cystic fibrosis knowledge as well as unscored questions related to sources of information, CF concerns, and whether discussions about death had taken place. The scored components included open ended questions, multiple choice and true-false alternatives. The authors wrote that "arbitrary weights were assigned by the investigators to scored questions based on overall importance and relevance". However, they did not specify the grounds on which these judgements were made nor did they record whether they made any adjustments to the scores from the multiple choice questions to take into account the possibility that participants were guessing.

The questionnaire was administered to 25 patients between the ages of 10 and 21 and their parents. The results indicated that both patients and parents had a relatively good knowledge of the disease and its treatment, but performed less well on questions about genetics and reproduction. Age and educational level were predictive of performance. There was a significant difference in the scores of those subjects under the age of 13 compared to those over 13, particularly in the area of genetics and reproduction. Nolan et al. (1986) wrote that this result was not surprising because "apart from the fact that knowledge accrues with age, there is evidence that prior to the onset of the formal operational period of cognitive development at 12 or 13 years of age, children are unable to grasp realistic concepts of illness, its complexities, and its multifactorial causes and ramifications".

Patients and their parents were also asked in this study who had taught them most about cystic fibrosis, whether they had discussed dying, and whether they wanted more information about the illness. Over half the patients said that they had learnt

most about CF from their parents, a quarter of whom in turn reported that they had learnt most about CF from non-clinic sources such as medical texts. A third of both patients and parents said that they would like more information from clinic staff, especially doctors. Eight out of eighteen patients (those above the age of 13) said that they had talked about dying, six with their parents and two with their doctor. 64% of parents said that they had talked about dying with their child, but in five patient-parent pairs there were discordant results where the parents reported having discussed death while the patient denied this. The authors of the study suggested that this could be due to guilt ridden parents feeling that they should have talked to their child and so reporting erroneously that they had done so, or patients who used denial as a coping strategy, failing to acknowledge that such a conversation had taken place. This coupled with the fact that parents were most often the main informants about CF for patients led the authors to recommend that "apart from ensuring that parents are fully informed about cystic fibrosis, it is just as important to provide them with advice from the earliest stages about how to transfer this information to their children in a way that is threatening neither to the parent nor the child".

4. 2. 2 Hames, Beesley & Nelson (1991)

Hames et al. (1991) used the questionnaire devised by Nolan et al. (1986) and said that they "modified" it for use with a British population. But, they did not report the changes that they made, whether linguistic or more substantial. Their measure had the same format of open-ended, multiple choice and true-false questions as that of the Nolan et al. (1986) questionnaire. However, Hames et al. (1991) did not indicate whether they also weighted the scores that they received, how they had scored the open-ended questions, or discuss any of the psychometric properties of the measure.

Hames et al. (1991) used the questionnaire with twenty-two of their patients aged between 11 and 20. Patients and parents "showed similar degrees of knowledge and also of ignorance". As in the Nolan et al. (1986) study, patients and parents were found to have a reasonable knowledge of the disease and its treatment, but a limited understanding of the genetics of CF. A comparison of responses from patients under 13 and over 13 demonstrated that the percentage of correct answers about the disease was similar in both groups, but the younger group knew less about treatment and genetics. Patients under 18 said they got equal teaching about CF from their parents and from clinic staff, while those over 18 said that most information came from clinical staff. A third of patients wanted more information, in particular about employment and "the future". Patients over the age of 13 indicated that they would like to receive information from staff rather than their parents. Hames and colleagues used this study to inform their practice within clinic. They recognised that "discussion had been inadequate on what to expect in the future, death and the everyday problems of cystic fibrosis", and they reported that since the study staff had a much clearer understanding of their role and responsibility in informing patients and parents about CF.

4. 2. 3 Henley and Hill (1990)

Henley and Hill (1990) developed a CF knowledge test consisting of 63 multiple choice questions covering an extensive series of CF related issues. They reported on the psychometric properties of their measure in some detail. They carried out a reliability analysis on the test scores of patients, parents and siblings and concluded that the coefficients of "0.91, 0.94, and 0.95, respectively, reflected high levels of

internal consistency among the performances of all family groupings". They had established content validity while designing the measure. They carried out an extensive search of professional and lay literature on CF, had the "content and technical adequacy of items evaluated by experts in the fields of pediatrics and education" and had three senior paediatric consultants assess whether items would be correctly answered by parents. Construct validity "was assessed by studying differences in knowledge among groups believed to differ in their levels of content mastery". The questionnaire was found to discriminate between the answers given by paediatricians, physiotherapists, nurses, medical students and domestic/clerical staff.

Henley and Hill (1990) also wrote that in order to "overcome threats to validity such as 'cheating' and guessing, family members were asked to respond spontaneously, to use the 'I don't know' option when in doubt, and not to exchange answers with each other". They do not comment upon how successful they believe their participants were at following this advice, and the study might have been stronger had they been able to ensure that the questionnaires had been completed individually. However, given the impressive size of the sample (n = 60 families), the fact that patients, parents and siblings were all asked to complete the questionnaire and that the study took place across a wide geographical area, this would have been an onerous task.

The results of Henley and Hill's study indicated that "75% of parents and patients could be interpreted as reflecting satisfactory levels of medical knowledge". The knowledge of siblings was significantly lower which would suggest that there was no extensive collaboration between parents and their children when completing the questionnaire. However, the study also highlighted numerous errors and

misconceptions held by family members. Of particular concern was the fact that the errors occurred when people were asked to apply their factual knowledge to practical situations. For example, most subjects knew that pancreatic enzymes were used to aid digestion and the absorption of fats, but many could not make the link between fat intake and enzyme dosage and several were unaware that enzymes needed to be taken at every meal. Other sources of concern to the authors included the fact that 72% of patients and 33% of parents thought that nebulized antibiotics should be given before physiotherapy, while 40% of parents gave themselves a 75% chance of having another baby with CF in any future pregnancy despite correctly answering that there is a 1 in 4 chance of having another affected baby. All of these findings led Henley and Hill (1990) to suggest that inadvertent noncompliance with treatment may be widespread and that simply testing someone's factual knowledge of a disease does not provide an accurate measure of what is truly understood. They recommended that similar studies be carried out in other clinics so that "specific errors, gaps, and misconceptions in knowledge of cystic fibrosis can be accurately identified for correction by the health care team. In this way inadvertent noncompliance with therapy in the cystic fibrosis patient can be minimized".

4. 2. 4 Meghdadpour & Steele (1992)

Similar conclusions were drawn by Meghdadpour and Steele (1992) after they administered a CF knowledge questionnaire to teenagers and their parents. The results indicated a wide variation in the quality of answers with particular difficulties in the field of genetics and reproduction. 40% of their participants did not know what a gene is and 30% were confused about male and/or female reproductive capacity. The authors wrote that the results "indicate a need to make periodic assessments of

knowledge base and to address the deficits in an individual fashion". They used their questionnaire to draw up "individual education plans" for each child and his or her family.

4. 2. 5 Conway, Pond, Watson & Hamnett (1996)

The most recent study of CF related knowledge was published by Conway et al. (1996). Their questionnaire had been "independently approved by a consultant specialist in cystic fibrosis, a professor of medicine, a cystic fibrosis research fellow, and a social worker as being representative of what adults with cystic fibrosis might be reasonably expected to know". The questions were presented in a true-false format but included a 'don't know' response "to reduce guessing".

Conway and colleagues restricted their study to adults with cystic fibrosis with ages ranging from 16 to 40. They thought that given the gaps in the knowledge of adolescents with cystic fibrosis these may well be carried through into adulthood and that the errors and misconceptions limit the "sense of control and independence that must accompany the patients' graduation to adult life". Eighty patients were asked to complete a 25 item multiple choice questionnaire covering issues such as lung and digestive tract involvement in CF, reproduction and sexuality, as well as more unusual complications and genetics. Overall "patient knowledge about the effects of cystic fibrosis on the lungs and the gastrointestinal tract were good but nonetheless showed some important gaps in understanding". They too found that 25% of patients thought nebulized antibiotics should be given prior to physiotherapy, 51% did not know whether severe constipation can be the result of taking too few enzymes, and 15% of male participants did not believe that they were sterile. Like Henley and Hill (1990)

they suggested that "poor knowledge about treatment in CF could be contributing to unintentional poor compliance with treatment regimens". An additional feature of this study was that clinic staff members were asked to predict the scores that each of the participants would obtain in order to determine whether staff could identify those individuals who might need additional information about their condition. The results showed that "the core members of the cystic fibrosis team were poor judges of individual patient's overall knowledge". The specialist nurses were however the most accurate predictors of knowledge scores which the authors felt "undoubtedly reflects the time they spend in direct contact with the patients, and the insufficient informal contact between doctor and patient. This is a reminder to optimise available resources".

4. 2. 6 Bluebond-Langner (1996)

Using a purely qualitative approach, Bluebond-Langner investigated the perspective, including knowledge, of well siblings of children with cystic fibrosis. She carried out in-depth interviews and observations of 40 families of children with CF over a two year period. She found that the views and understanding of siblings varied according to the extent of the illness in the child with CF. She identified a series of stages in the understanding of siblings from a point of seeing CF as "a condition one does things for" during the early years without an exacerbation, to a recognition of CF as a "chronic, progressive, incurable disease that shortens the life span, including one's own sibling" during the terminal phase of the disease. She did not find age to be a factor in determining a well sibling's view of the disease, and she did not even discuss issues of cognitive development. But she did write that age does "affect the sophistication with which the well siblings express their views". Instead she

concluded that the views of well siblings were the response to a complex interplay of four factors, (1) the patient's condition and experiences with CF, (2) the sibling's assessment of the patient's condition and experiences, (3) the parents' responses to the physical and psychological demands of CF, and (4) the siblings' assessments of those responses.

4. 3 Gaps in the Studies

While these studies have been essential in highlighting the need for patients with cystic fibrosis and their parents to be well informed about their illness, and have identified a number of commonly held errors and misconceptions, there remains a gap in knowing what children with CF know about the disease. No study since 1977 has asked children under the age of 9 about their knowledge of CF. Those studies that have been carried out have mostly involved older adolescents and young adults and they have generally made no distinction between the results of adults and children. All the studies have asked participants to fill in forced choice multiple choice questionnaires. While this does provide clear information about the participants' factual knowledge, it does not allow any more qualitative data to be obtained which could provide a greater flavour of what children think and feel about their illness. The way in which children's knowledge of CF develops has also not been explored in any of the existing studies, nor have any of them tried to gather data on children's understanding of the emotional impact of CF upon themselves and their families. In addition, all of the previously cited papers have looked at CF knowledge in isolation, thus no study has explored the link between CF knowledge and adherence with treatment, or CF knowledge and family environment.

4.4 Conclusion

Studies carried out in the USA (Nolan et al., 1986), Great Britain (Hames et al., 1991; Conway et al., 1996), and South Africa (Henley & Hill, 1990), have all explored the knowledge that people with cystic fibrosis and their families have of their illness. The results have been remarkably similar, indicating that patients and their parents have a relatively good understanding of the disease and it's treatment, but are less well informed about reproductive issues, the genetics of CF, and the future course of the illness. Specific errors and gaps in knowledge have been identified; particularly those concerned with the practicalities of treatment such as when to carry out nebulized treatment and how to calculate enzyme usage at mealtimes. These studies have made recommendations about how to improve clinic procedures and professional-patient communication.

The studies reported in this chapter have focussed upon the knowledge of adolescents and young adults, with no studies exploring the knowledge that children with CF have of their illness having been published for over twenty years. This study will examine the knowledge of children and adolescents with cystic fibrosis between the ages of 4 and 18. It will use a booklet of open-ended questions in the hope that the participants will be able to give more detailed responses than is possible with a multiple-choice structure. The study will also examine the relationship between participants' knowledge of CF, age, cognitive development, severity of illness, compliance with treatment and family environment.

CHAPTER FIVE: AIMS AND HYPOTHESES

5.1 Introduction

Children's understanding of illness, its cause, course, cure etc has been a topic of particular interest to researchers since the 1980's. Originally children's knowledge was thought to progress through a series of orderly, pre-ordained stages, mirroring the stages of the child's more general cognitive development. However, more recent studies have emphasised the role that experience and teaching has upon a child's understanding of illness, and there is now increasing recognition of the extent to which even some very young children can make sense of and understand illness.

Strangely, while general studies of children's understanding of illness have flourished since the 1980s, no study exploring what children with cystic fibrosis know about their illness has been published since 1977. There have been some studies (Nolan et al., 1986; Hames et al., 1991; Henley & Hill, 1990; Conway et al., 1996) looking at how older adolescents and young adults understand their cystic fibrosis, which have identified areas of strengths and weaknesses in terms of CF knowledge. However, these studies have tended to use very structured forced choice and multiple choice questionnaires. They have also looked at knowledge in isolation rather than explore associations between knowledge and other factors within the child's environment. The purpose of this study is therefore to focus upon the knowledge that children and adolescents have of their cystic fibrosis and to examine whether such knowledge is associated with family factors and compliance with treatment.

5. 2 Aims of the Study

The aims of this study are:

1. To develop a tool to access children's knowledge of cystic fibrosis
2. To investigate how children with cystic fibrosis understand their condition
3. To explore the way in which children's knowledge of CF develops
4. To examine the relationship between children's knowledge of cystic fibrosis and family environment
5. To look at the relationship between children's knowledge of cystic fibrosis and compliance with treatment.

5. 3 Hypotheses

1. While some studies exploring children's understanding of illness and the way in which it develops have restricted the age range that they have studied (Clunies-Ross & Lansdown, 1988; Gibson et al., 1995) others have applied their measures across a wide age range of children (Berry et al., 1992; Sargent et al., 1995). The open-ended structure of the measures used in these studies have contributed to their applicability across age ranges as broad as 4-18. Therefore the first hypothesis of this study is that: ***It should be possible to develop a single tool to access the knowledge that children with cystic fibrosis have across the age range of four to eighteen.***

2. A child's knowledge of illness has been shown to be positively correlated with age and cognitive development in numerous studies (Bibace & Walsh, 1980; Perrin & Gerrity, 1981; Hansdottir & Malcarne, 1998). However, illness

knowledge has also been found to be enhanced by exposure to/experience of an illness (Gibson et al., 1995; Crisp et al., 1996). Whether this increase in knowledge is the result of increased contact with health professionals, the child's increased attention to and interest in the illness or some other factor is unclear (Rushforth, 1996). However, it would suggest that within the CF population those children with more severe disease as characterised by lower lung function, weight and height and a greater number of hospitalisations, would be likely to have an increased knowledge of cystic fibrosis. Thus the second hypothesis of this study is that: ***A greater knowledge of cystic fibrosis will not only be associated with age and cognitive development but also with experience of cystic fibrosis as measured by lower lung function, height and weight, and a greater number of hospitalisations in the previous year.***

3. The studies looking at the knowledge that adolescents and young adults have of their illness have highlighted the extent to which they receive most of their information about CF from their parents (Nolan et al., 1986; Hames et al., 1991; Bluebond-Langner, 1996). Family functioning particularly in areas of cohesion, expressiveness and organisation have been found to be associated with compliance with treatment "perhaps open expression of concern helps families appraise their situation realistically and manage it better" (Moos & Moos, 1994). In addition, cohesion and expressiveness are the features of family functioning "most closely associated with children's cognitive and social development, cognitive competence and verbal communication" (Moos & Moos, 1994). Thus the third hypothesis of this study is that: ***A greater knowledge of cystic fibrosis***

will be associated with higher levels of family cohesion, expressiveness and organisation.

4. The studies looking at patients' knowledge of cystic fibrosis have commented upon the association between knowledge and compliance with treatment. Henley & Hill (1990) wrote, "it is presumed that compliance will be enhanced by a broad, factual and applied knowledge of the condition and its treatment". Koocher et al. (1990) explored compliance with treatment issues within a CF population and identified three different types of non-adherence. The three aspects were inadequate knowledge, psychosocial resistance and educated non-adherence. The fourth and final hypothesis of this study assumes the counter point to that identified by Koocher, McGrath & Gudens. and supposes that: ***A greater knowledge of cystic fibrosis will be associated with higher levels of compliance with treatment.***

CHAPTER SIX: METHODOLOGY

6.1 Ethical Approval and Safeguards

A research proposal was drawn up outlining the purpose and methodology of the proposed study. This was submitted to the ethics committee of City University and the Local Research Ethics Committee of Merton and Sutton which monitors all research work carried out at Queen Mary's Hospital for Children, Carshalton. Both committees granted ethical approval. The local hospital committee emphasised that care should be taken to ensure that any potential hazards inherent within the research were minimised and that safeguards be in place should any difficulties develop. The possible hazards were thought to be:

- Potential participants might feel forced into taking part in the study as it was being carried out in their treatment centre
- Distressing children by asking them questions they can not answer or do not want to think about at present
- Distressing parents by exploring painful issues about cystic fibrosis with their children
- Contributing to difficulties in family communication if either parents or children wish to discuss matters further before the other is ready to do so.

The safeguards that were put in place to address these issues included:

- Sending potential participants and/or parents a detailed information form in which the independent nature of the study was emphasised so that they would not worry that their treatment might be compromised if they refused to take part

- Assuring all participants that they do not have to answer any question they do not want to and can ask to stop at any time
- Allowing parents to be present throughout the interview and allowing them to look at the booklet before agreeing to their child taking part in the study
- Assuring parents that the researcher would stop the interview as soon as a child showed signs of distress
- Providing a space for debriefing with the researcher after the interview should either the participant or parent request it
- Providing verbal feedback immediately after the interview and written feedback at the end of the study to any individuals who would like it
- Providing access to another member of the Child and Adolescent Psychology Department should either an individual or family suffer ongoing distress that could not be appropriately addressed by the researcher in her clinical capacity
- Ensuring that all material from the study is kept separately from the participant's medical/clinical records, and is held in locked filing cabinets in different premises from their medical notes.

6. 2 Measures

6. 2. 1 The "I can help others learn about cystic fibrosis" booklet

A booklet was designed to access children's knowledge of cystic fibrosis. It was structured to ensure that participants could use their own words to describe what they know and could answer a question in as much detail as they liked. Twenty-two open-ended questions around five main themes were included. The themes covered were:

- The nature of cystic fibrosis
- Treatment requirements

- Genetic aspects of the disease
- The future impact of the illness
- Feelings associated with CF.

Each question was on a separate sheet of paper and was accompanied by a comic style illustration drawn by a specialist children's illustrator. Separate booklets were developed for boys and girls, with male and female characters accordingly. The illustrations were identical other than the male character (Sam) having short hair, and the female (Sophie) having long hair. The pictures were designed to make the booklet more child friendly, but were not designed to lead the participants in any way. For example, the illustration to the question "How did Sam/Sophie get cystic fibrosis?" was of him/her in a supermarket with a shopping trolley taking a can labelled CF off a shelf. It was assumed that no participant would be likely to think that CF could be bought from a shop, and if they were led by the picture it would be very clear from the content of their answer.

The questions were presented to the participants as those of another child with cystic fibrosis who knows nothing about the disease but wants to find out about it. The booklet was entitled "I Can Help Other People Learn About Cystic Fibrosis" and the front sheet said "Sam/Sophie is a boy/girl just like you. He/she has cystic fibrosis but does not know anything about it. He/she has lots of questions about it, do you think you could try to answer them for him?" The purpose of this was to make the questions less threatening or upsetting while at the same time still allowing for detailed information to be collected. Thus instead of directly asking a child "how old will you grow up to be?" the child's knowledge of life expectancy in CF can be

gathered more sensitively by asking "how old do you think Sam/Sophie will grow up to be?" The questions were brief and open ended so that they could be presented to participants across the age range of four to eighteen and would ensure that considerable qualitative data could be obtained. (Appendix 2).

The questions in the booklet were initially drawn up by the author, based upon her clinical experience, and were then discussed with a range of other professionals. A paediatric cystic fibrosis nurse specialist, paediatric dietician and paediatric physiotherapist, all of whom work on the same team as the author, were asked to draw up their own list of questions, and additions were made to the author's list where gaps were identified. The list of questions was then shown to the paediatric chest physician on the team for his comments and suggestions. In addition, another clinical child psychologist working with children with cystic fibrosis in a different setting was consulted and her suggestions were incorporated into the booklet. Finally, two children with cystic fibrosis, one aged 11 and one aged 16, and their mothers were shown the booklet and asked to comment on any omissions they thought should be addressed. Despite considerable encouragement to make suggestions they reported that they felt all areas were covered and they did not have any questions to add other than the 11 year old who thought a question about the dangers of smoking should be included. The author however decided that this was not a specific aspect of CF related knowledge and did not warrant a question itself. But it was hoped that it might be addressed spontaneously by participants in their answers to other questions such as "what should Sam/Sophie do each day to keep well?"

6.2.1.1 Scoring the Booklet

All participants completed the "I can help others learn about cystic fibrosis" booklet. As this was an exploratory study no scoring system was developed prior to the data being collected. The purpose of the research was to investigate what children know and how this knowledge develops, so no assumptions were made about what might be expected from their answers. Instead, the answers to each question were examined by the researcher and clustered together according to their depth and accuracy. There were no restrictions upon the number of clusters that could be obtained for each question; the aim was not to try to fit answers into a pre-determined structure but rather to explore what details and themes might be identified from the data. Thus the first question which asked participants "What is cystic fibrosis?" was found to produce five clusters of answers. In ascending order of depth and accuracy, they were as follows:

1. Five participants said that they did not know what CF is
2. Five participants mentioned some aspect of CF treatment e. g. "it is where you have to take tablets and if you don't you get a tummy ache"
3. Seven participants described it as something that affects your lungs and digestive system
4. Three participants indicated that it is an illness you are born with that affects the lungs
5. Three participants talked of it as a genetic illness affecting the lungs and digestive system.

In contrast, question number 3 which asked participants "What are the things that Sam/Sophie has to do each day to keep well?" was analysed in terms of the number of treatments mentioned by each participant. In this case seven clusters of answers were established ranging from two participants who mentioned one aspect of treatment, through six participants who listed four aspects of treatment, to the one participant who described seven aspects of treatment.

Once all the data had been clustered, the researcher then looked at the clusters in each question and made a decision about how to rate them in terms of quality. They were then ranked in numerical order from 1 which was the least detailed upwards according to the number of clusters obtained for each question (range 3-7). Participants' scores were thus available for each question, and a total CF knowledge score was obtained by adding together their scores on each individual question. The lowest total score that a participant could obtain i. e. if they had scored 1 for every single question, was 22. The highest possible score that a participant could obtain i. e. if their answer had fallen within the highest category each time, was 111.

6.2.1.2 Booklet Reliability

A test of inter-rater reliability was carried out to determine whether the researcher analysed the data appropriately. Six participants' responses to the booklet were randomly selected (participant numbers were pulled out of a hat) to be the focus of the reliability study. Two colleagues, a clinical child psychologist with some experience of working with people with CF and a cystic fibrosis nurse specialist, were asked to rate the data. They were presented with the answers of the six participants as well as the title of clusters developed for each question and they were asked to assign each

participant's answer to the cluster that matched it most closely. They repeated this for all twenty two questions. In addition they were then asked to rate the clusters of answers to each question from least to most sophisticated. These results were then compared to the judgements made by the researcher and inter-rater reliability figures were calculated.

6.2.2 The British Picture Vocabulary Scale (BPVS)

Studies exploring the development of children's understanding of illness have shown that chronological age is not the only variable that should be explored. Measures of a child's cognitive development should also be included (Bibace & Walsh, 1980; Perrin & Gerrity, 1981), as should their experience of illness (Goldman et al., 1991; Crisp et al., 1996). Walsh and Bibace (1991) do argue that there is no "pure measure" of cognitive development and that chronological age is as good as any more formal Piagetian measure of conservation, transformation or logical thinking. Crisp et al. (1996) also argue that the traditional Piagetian measures are not sufficiently refined to be able to detect small changes in cognitive development. However, rather than reverting to using chronological age as an approximation of cognitive development, they measured cognitive function with the Peabody Picture Vocabulary Test–Revised (Dunn & Dunn, 1981). The present study used the British Picture Vocabulary Scale (BPVS) which is based upon the PPVT-R but has been modified for use with, and standardised upon, a British cohort.

In the manual accompanying the BPVS Dunn and Dunn (1982) describe it as a measure of a subject's "receptive (hearing) vocabulary for standard English". They go on to say "vocabulary has been found to be the best single index of school success

and to be one of the most important contributors to measures of intelligence". However, Dunn and Dunn are at pains to point out that even though the BPVS provides a measure of mental age "it is not a comprehensive test of general intelligence" and should not be "equated with innate or fixed ability".

Dunn and Dunn write in the manual that "there are no direct measures of test-retest reliability" and so "internal consistency reliabilities have been used to establish standard errors of measurement". The split half reliability of the BPVS has been found to be in the range of 0.75 to 0.86 with a median of 0.80. While there have been few studies exploring the reliability and validity of the BPVS per se, Dunn and Dunn report that there are "over 100 published studies on the reliability of the PPVT, demonstrating that its reliability remains stable for many special groups. Validity studies have shown that it also correlates well with other vocabulary tests and individual intelligence tests".

The BPVS is a brief and unthreatening test for children in which participants are shown a series of cards with four pictures on and they are asked to point to one picture on each card that best represents a stimulus word that they hear. It is quick to administer and score. Ideally for this study which covers a wide age range, it can be used with children aged between 2 and 18. Raw scores are converted into three different types of normative scores, the "two deviation-type norms... standardised score equivalent and percentile ranks" and "the developmental–type norm, age equivalents". For the purposes of this study only the mental age equivalence scores were used as the measure of each child's cognitive functioning.

6.2.3 The Medical Compliance Incomplete Stories Test (MCIST)

The MCIST is a tool for assessing the attitudes of children and adolescents towards medical compliance situations. It consists of five unfinished stories in which the main character is faced with a medical dilemma which involves him or her making a decision about whether to follow medical advice or not. The situations range from one where the protagonist is told that it is time for him to have a top up vaccination to a situation where a boy is told that he may die if he does not have his foot amputated. Participants are asked to complete each story verbally and to predict what happens to the main character. (Appendix 3)

Each story is scored separately using a series of objective criteria. Responses are measured in terms of compliance, optimism and self-efficacy. An overall competency/compliance score is obtained by summing the three sub-scores across the five scenarios.

Koocher designed the MCIST in 1982 as a way of screening for non-compliance in a large American paediatric population. Czajkowski and Koocher (1987) used a population of hospitalised adolescents with cystic fibrosis to explore the relationship between MCIST scores and objective measures of medical compliance such as participation in physiotherapy, adherence to dietary requirements and taking medication. All three MCIST sub-scores and the overall score were found to have a strong and significant positive correlation with observed compliance measures. 97% of subjects were correctly classified from their MCIST scores into compliant and non-compliant groups. Commentators have suggested that it may not be very accurate at identifying non-compliance in an adolescent population. They cite the Czajkowski

and Koocher (1987) study in which 35% of adolescents were identified as non-compliant, and say that this is an unrealistically low score. However, given that their population were in hospital being very closely monitored and directly supervised for much of the time, the percentage of adolescents who would be found to be non-compliant with their independent treatment at home is likely to be considerably higher.

Given the time and resources available for this study, objective measures of compliance such as observation or monitoring of medication were never going to be feasible. The MCIST on the other hand could be relatively quickly and easily administered and has already been used with patients with cystic fibrosis. The stories needed slight modification in terms of language for a British population. Thus in story one the term "booster shot" was changed to "injection" and in story two the word "sick" was changed to "ill". In addition, the age range of participants in the Czajkowski and Koocher (1987) study was 13-23, and no other published studies have used the MCIST with a younger population. Thus it was unclear whether the measure would be suitable for younger children, but a decision was taken to use it and investigate the lower age range to which it could apply.

6.2.4 The Family Environment Scale (FES)

The FES is a ninety item self-completed questionnaire for individuals that measures their perceptions of the family environment. It yields ten subscales assessing three dimensions of family functioning. "The relationship and system maintenance dimensions primarily reflect internal family functioning, whereas the personal growth dimensions primarily reflect the linkages between the family and the larger social

context" (Moos & Moos, 1994). Parents and/or adolescent children within a family can complete the scale. In this study parents (in all cases the mother) were asked to complete the scale for participants under the age of 16. Those participants aged 16 and above completed the FES themselves. (Appendix 4).

The FES was chosen as the measure of family functioning in this study for several reasons. Firstly, it is one of the most widely used tools for assessing family functioning, with considerable content, face and construct validity (Varni, Katz, Colegrove & Dolgin, 1996). Secondly, it has been used extensively in research with families affected by a range of chronic illnesses, including cystic fibrosis (Patterson, 1990; Wilson, Fosson, Kanga & D' Angelo, 1996). Wilson et al. (1996) found the FES sub-scales of cohesion, expressiveness and organisation to be the three most important clinical dimensions in families with a child with CF. Similarly, Fosson (1991) identified four FES sub-scales that measured variables important in family adaptation to CF: cohesion, expressiveness, organisation and control. Thirdly, the areas of family functioning that the FES explores are likely to be of particular relevance to this study. Thus, when measuring children's knowledge of their illness, the family variables of expressiveness, cohesion, conflict, organisation and control may well be related to the extent of a child's knowledge and understanding of their illness.

6.3 Pilot Phase

The questions being explored in the pilot phase were:

1. Could the booklet be used for interviewing both young children and older adolescents?

2. Did the booklet obtain meaningful information?
3. Were any alterations required before the booklet could be used in the main study?
4. How long did it take to carry out the interview, including completing all tests and questionnaires?
5. Was the time involved acceptable to participants and their parents?

A seven year old boy and a fourteen year old girl were interviewed. The format used was identical to that being planned for the main study, with the additional request that they answer some questions about the study once they completed it. The two subjects were chosen because they spanned the age range of the population being investigated and were due to be seen at the clinic on the day chosen to begin the study.

Both participants were interviewed in clinic, the boy prior to his clinic appointment and the girl once she had finished her clinic visit. Both were accompanied by their mothers who were also included in the feedback session. Following the main interview they were asked for their opinion of the booklet and whether any changes should be made to it, whether they thought the time involved was acceptable and whether they had any other suggestions to make.

The feedback was generally extremely positive. Both participants said they enjoyed the interview. The girl aged 14 said "I thought the CF sheet was quite helpful, it made me realise the things I don't know about CF" (she mentioned not knowing how creon works, if she has CF why her sister does not, and how medication works). She was asked if she thought the format of the booklet was too young for her to which her reply was "the pictures look a bit babyish, but when you look at them they are quite

funny. The questions felt quite relevant to me". She did not make any suggestions for alterations. The boy aged 7 said "they were alright [the questions] they were good and they make me healthy when I say them". He was asked if there were any things he did not like, any questions that were boring or upsetting to which he replied "I didn't like the pin". This was a reference to the story in the MCIST where a child is told that he needs a vaccination. He did not indicate that any changes should be made to the CF booklet.

The interview with the boy took 50 minutes and with the girl took an hour and five minutes. Both participants said that the interview was long but OK. The girl said she did not get bored and it was worth taking the time to think carefully. She did say that she was tired at the end of clinic.

Both mothers fed back that it was quite a strain to add such a time consuming interview to clinic. They suggested that it would be better carried out on a different day and preferably at home. They also said that they found the FES hard to fill in as they did not like making absolute statements about their family life.

The researcher noticed that with the booklet there was a difficulty in knowing when a participant had finished their answer. At times they stopped because they were thinking of additional elements to include in their answer, while at other times they had said all that they wished to.

The content of the answers were examined by the author and a colleague within the Child and Adolescent Psychology Department. The answers were judged to be meaningful and it was felt that the questions were appropriate for both age groups.

It was decided that no changes, other than a few minor presentational ones, needed to be made to the study before implementing it with the whole clinic population. These included:

a) Asking participants to tell the researcher when they have completed their answer for each question in the booklet
b) Acknowledging that it might be tiring carrying out the interview during a clinic visit and suggesting that it be carried out on a different occasion
c) Acknowledging to parents that the FES might feel a little uncomfortable to fill in.

6. 4 Recruitment

Having obtained ethical approval for the study from both City University and the local Merton and Sutton hospital research ethics committee the author wrote to all parents of children and adolescents between the ages of 4 and 16 (n = 37), and all adolescents aged 17 and 18 (n = 3) who attend the South West Thames regional paediatric cystic fibrosis clinic at Queen Mary's Hospital for Children, Carshalton. Patients attend this specialist clinic on a quarterly basis for a routine check up and review of treatment. However, the clinic takes place once a week and parents and patients are aware that they can attend clinic whenever they have a CF related concern.

The letter that the families received before attending clinic described the purpose and structure of the study, the time involved and the safeguards in place such as the ability to withdraw at any time and full confidentiality. The letter emphasised in particular that the study was an independent piece of work that was not a standard part of their clinic visit and that there was no expectation or pressure for them to consent to participate. The letter made it clear that no aspect of their or their child's treatment would be affected by their decision. (Appendix 1).

There were 40 individual children and adolescents eligible to take part in the research, of whom 23 were interviewed. This represents a 58. 5% uptake. Only 5 individuals (12. 5% of those approached) openly refused to take part. A further 3 potential participants (7. 5%) replied that they would be interested in taking part but did not have the time to do so as one was working and two were going on holiday. An additional 9 families (22. 5%) were written to but not seen in clinic. Of the 40 children and adolescents that were eligible to take part 25 were girls and 15 were boys. Of the 23 participants who agreed to take part, 14 were girls (56% of those eligible to take part) and 9 were boys (60% of those eligible to take part).

Demographic data such as social class, family structure and number of children with cystic fibrosis within each family was collected on all participants. Health data was also obtained in the form of an average measure of lung function for the previous year, numbers of hospital admissions, as well as centile scores for height and weight that were then calculated as measures of each participant's body mass index (BMI).

Written consent was obtained from the parents of all participants under the age of 16 and from all adolescent participants aged 16 and above.

Sample Bias

The nature of the recruitment of participants into this study has some inherent biases within it that do weaken the study. Firstly, the clinical role of the researcher had an impact upon the way in which she recruited participants. While she wrote to the families of all those patients eligible to take part, she actively recruited only those who attended CF clinic during the recruitment period. Thus some potential participants were lost because the researcher felt uncomfortable about approaching families within her clinical caseload at home. In addition, the responses of potential participants when approached about being recruited into the study could have been influenced by their existing and ongoing clinical relationship with the researcher. For example, some families might have experienced some sense of obligation towards the clinical team that led them to enrol in the study when they were not sure that they wanted to. In contrast, others might have taken part in the study had it been carried out by someone with no links to their clinical service. The researcher did try to address the issues above, particularly those related to families feeling under pressure to take part in the study, in the initial information that she sent out. But the effectiveness of this information was not explored in the study, and a concern must remain about the nature of the recruitment.

In addition, the fact that participants were recruited from only one CF centre reduces the likelihood that the sample is going to be representative of the general CF population. Within this small sample, participants were relatively healthy with only

six of the twenty-three having required a hospitalisation in the previous year, and none at the severe stage of the disease as defined by a lung function (FEV1) below 40%. Figures from the national CF database indicate that within Scotland 6% of children with CF have the disease in the severe form. The participants in this study have all received their care from one very stable (in terms of staff turnover) clinical team. There is no opportunity within this study to compare the knowledge of patients from different clinical teams that may be providing different care and information to their clients. Similarly, the participants in this study all live within a fairly defined geographical area (South West Thames). The area served by the South West Thames CF clinic was classified by the National Statistics Office in 1999 as Urban Fringe and was one of fifteen areas described as the most prosperous in Great Britain. The key characteristics of the most prosperous areas were "higher than average values for all indicators of affluence including the proportion of the population where the household head is in social class 1 or 2, households with two or more cars, and households with two earners" (Griffiths & Fitzpatrick, 1999). There is no way of knowing within this study whether the scores obtained by this sample would be similar to those from other geographical regions such as a deprived inner city area.

6.5 Procedure

All patients aged between 4 and 18, and their parents, who attended the cystic fibrosis clinic at Queen Mary's Hospital for Children, Carshalton, between June and August 1999 were informed about this study. They were sent an information sheet two weeks prior to their routine clinic appointment with details of the purpose and procedures involved in the study. Upon arrival at clinic they were approached by the researcher and asked if they would be interested in taking part. Those who did not want to take

part then continued with their clinic appointment, while those who indicated that they were interested arranged a time for the interview to be carried out.

All interviews began with a few minutes of informal conversation aimed at putting the participants at ease. Parents were given the Family Environment Scale (FES) to fill in, and a joke was made about how they could not get away with just observing the interview as they would be working as hard as their child. The researcher then explained to participants that the interview was not a test, that there were no right or wrong answers and that what she wanted to get was each individual's point of view. The measure were then administered in the following order:

1) The British Picture Vocabulary Scale (BPVS)

2) The 'I can help others learn about cystic fibrosis' booklet N. B. Informal feedback was requested at this stage on the participants' views of the booklet

3) The Medical Compliance Incomplete Stories Test (MCIST)

The BPVS was presented first as it was thought to be an appropriate introduction to the interview as it provided a good way of establishing rapport and gave plenty of opportunity for positive feedback. The 'I can help others learn about cystic fibrosis' booklet was the key component to the study and was therefore presented next. Despite having piloted the measures on two participants aged 7 and 14 who coped well with the demands of the interview, the researcher was aware that the time involved was quite considerable and that younger participants might struggle to complete all the measures. The researcher felt that if there were interviews that were not completed it would be more useful to have the booklet information rather than the MCIST results. Consequently, the order in which the measures were presented to

participants was not randomised. There is evidence from the results obtained of an order effect with the MCIST completed in less depth than the other measures. The MCIST results are therefore not as robust as they would have been had the presentation of measures been randomised and any future study should take this into account.

The answers provided by participants were written down verbatim during the interview. However, all interviews were also audio tape-recorded other than one when the cassette recorder broke down. It was explained to participants that the recordings were taking place in case they gave so much information that the researcher could not write it down during the interview and needed to go back to it later. They were also told that the tape would not be listened to by anyone else on the CF team and would be destroyed at the end of the study.

The researcher provided positive feedback throughout the interview, praising participants for their effort, thoughtfulness and achievements. At the end of the interview they and their parents were thanked and asked if they had any questions themselves or wanted to talk about any aspect of the interview.

CHAPTER SEVEN: THE SAMPLE

7.1 Participants

As noted above, 40 children and adolescents were eligible to take part in the study and were sent information sheets prior to attending clinic. Thirty-one eligible children attended clinic during the data collection period. Twenty-three individuals from twenty families were interviewed (74% of those who attended clinic). Three others, girls aged 4, 9 and 17 (10% of those who attended clinic) said that they were interested in taking part but were unable to find a convenient time to be interviewed. While they (or their parents) may have been genuinely keen to take part, it is also possible that they felt uncomfortable openly refusing to take part in the study, especially as the researcher was a member of their clinical team, and so found other more subtle ways of not taking part. Five individuals refused to take part (16% of those who attended clinic), three boys aged 8, 13 and 17, and two girls aged 8 and 12. While they were not asked to explain their decision, the parents of three of the children spontaneously gave their reasons for refusing to let their child participate. The 8-year-old girl has learning difficulties in addition to CF and her mother thought she might become distressed in a 'testing' environment. The 8-year-old boy had said he was not interested in the study and the 13-year-old boy had recently been an inpatient so his mother said "he just wants to forget about CF".

Of the original 40 eligible individuals who were written to nine did not attend clinic during the data collection period and so were not included in the study. This represents 22.5% of the total eligible sample and had they been included in the study

it would have been strengthened. While the recruitment of these potential participants would have been slightly different from the recruitment of those who attended clinic it would have been useful to have a larger sample. The researcher could have been more proactive in following up these potential participants through further letters, telephone contact, or arranging home visits independently of their clinic attendance. However, the researcher felt constrained by her clinical role and did not feel comfortable approaching these families in any way that might be perceived by them to be pressurising. The researcher should perhaps have enlisted the help of someone who was not involved with the CF clinic to contact these nine potential participants and explore whether they were happy to take part rather than simply leaving them out of the study.

7.2 Power Analysis

A power analysis to estimate the appropriate sample size for this study was not carried out as studies reported previously that looked at CF related knowledge (Nolan et al., 1986; Hames et al., 1991) had sample sizes of 28 and 22 respectively, which were similar to the sample size of 23 in this study. However, the methodology of the study would have been strengthened had a power analysis been included from the start.

7.3 Demographic and Health Data

A range of demographic data was collected about each participant. In addition to their gender, chronological age and mental age, measures of the participants' physical health status were recorded. In particular each participant's average lung function (FEV1) over the last year and number of hospital admissions or episodes of intravenous drug treatment that had occurred in the previous year were recorded. In addition their current weight and height was recorded and measured as a percentile of

what would be expected for a healthy child of the same age. Each participant's body mass index(BMI) which is a measure of the relationship between weight and height was also calculated. Family factors were also examined, including the social class of the family, marital status of the parents and number of siblings with cystic fibrosis. Table 1 below presents the main demographic data for each of the twenty-three participants.

Table 1. Demographic and Health Details of Participants

Participant	Gender	Chronological age	Mental age	% Lung function	Number of hospital admissions	Social class	Marital status of parents	Number of siblings with CF
A	F	4	4	89	0	2	Divorced	0
B	F	4	5	66	0	3	Married	0
C	F	5	5	108	0	2	Divorced	0
D	F	6	6	70	0	2	Married	0
E	M	6	6	90	0	2	Married	0
F	M	6	6	86	0	2	Married	2
G	F	7	6	87	0	3	Married	0
H	M	7	7	69	0	2	Married	0
I	M	7	7	93	0	1	Married	0
J	M	8	7	66	3	5	Married	1
K	F	8	9	78	0	2	Married	2
L	M	9	8	85	2	5	Married	1
M	M	11	9	111	0	5	Divorced	0
N	F	11	10	78	0	3	Married	0
O	F	13	11	111	0	3	Married	0
P	F	13	12	74	2	3	Married	1
Q	F	14	13	91	0	2	Divorced	0
R	F	14	13	67	3	1	Married	0
S	M	14	13	89	0	3	Married	0
T	F	15	13	73	3	3	Married	1
U	F	16	16	72	0	3	Married	0
V	F	16	17	72	1	2	Married	0
W	M	18	18	88	0	3	Married	0

7.3.1 Health Data

The average lung function of each participant over the previous year was obtained as a measure of the child's well being. The measure used is that of Full Expiratory Volume (FEV1) which provides a reading of the amount of air that a child can expel from their lungs in the first second after having taken a deep breath in. It is recorded in terms of the percentage of what would be expected to be blown out by a healthy child of the same gender, age and height. The lung function of each participant is presented above, in Table 1. The range of lung function scores for the male participants is 66-111% (mean 90%). The range of lung function scores for the female participants is also 66-111% (mean 82%).

The national CF database based in Dundee is in the process of collecting health data on every child with CF from every CF clinic across the country. They have not yet received the necessary data from all the English CF centres, but they have received this data from all Scottish centres. The Scottish data is classified into mild, moderate and severe disease according to lung function figures. The same criteria were used to classify the participants in this study. Table 2 below presents the data for this sample and compares it to that of the Scottish paediatric CF population.

Table 2: The disease severity of this sample compared to the Scottish paediatric CF population.

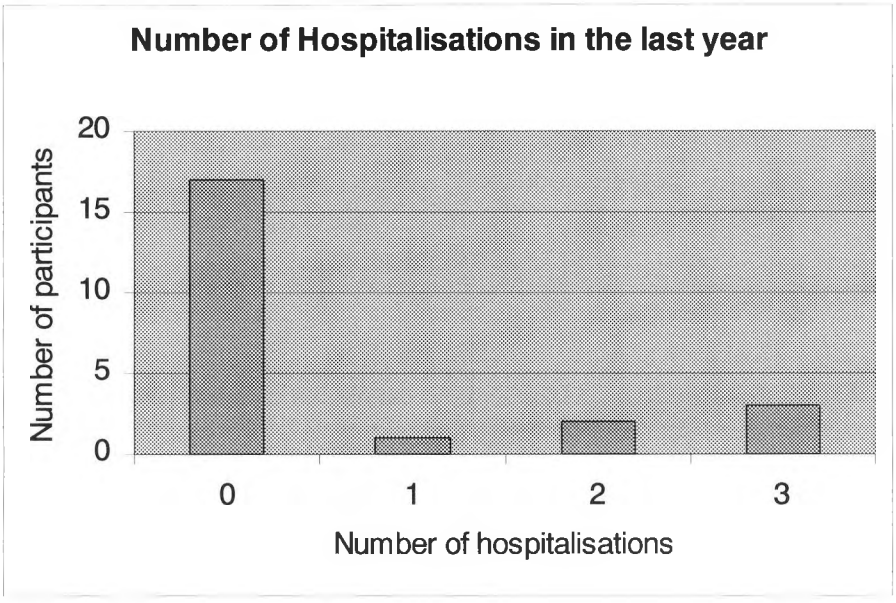
Disease severity	This sample	Scottish CF population
Mild (FEVI 85% and over)	52%	47%
Moderate (FEV1 41%-84%)	48%	47%
Severe FEV1 (40% and under)	0%	6%

As table 2 displays 52% of the participants in this study can be described as having mild CF compared to 47% of the Scottish CF population. 47% of the Scottish children are reported to have moderate CF (lung function 41-84%) while this sample had 48% of participants with moderate CF. There were 6% of Scottish children with CF within the severe range (lung function 40% and below) but no children in this sample fell into the severe category.

The number of hospitalisations or episodes of intravenous drug treatment that each participant had experienced in the previous year was recorded as it was thought to be a measure that could be used as a reflection of the participants' physical health. As Figure 1 on the following page shows, the majority of participants (74%) had had no hospital admissions or intravenous drug treatment within the last year. Six of the twenty-three participants had experienced at least one hospital admission within the

last year. The Scottish database figures show that 55% of the CF population aged 18 and under in Scotland had no admissions to hospital or IV treatments in the year 2001.

Figure 1: Number of Hospitalisations Experienced by Participants in the Previous Year



Height and weight were included within the health data as an indicator of physical wellbeing within cystic fibrosis.

Figure 2: Height Centiles of Participants

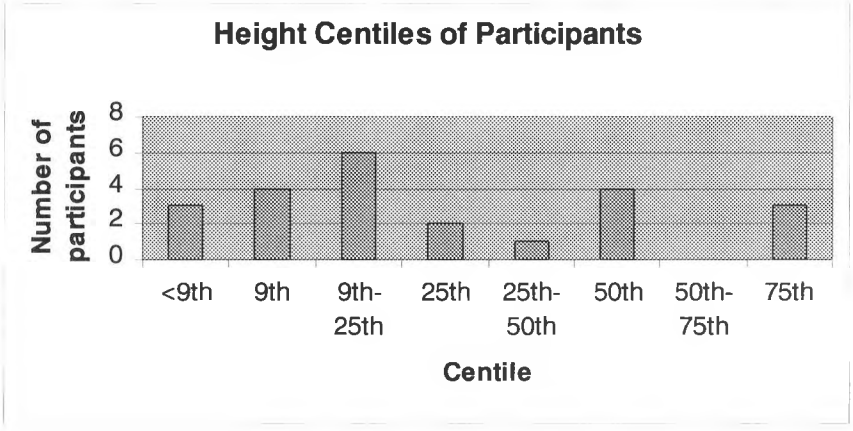
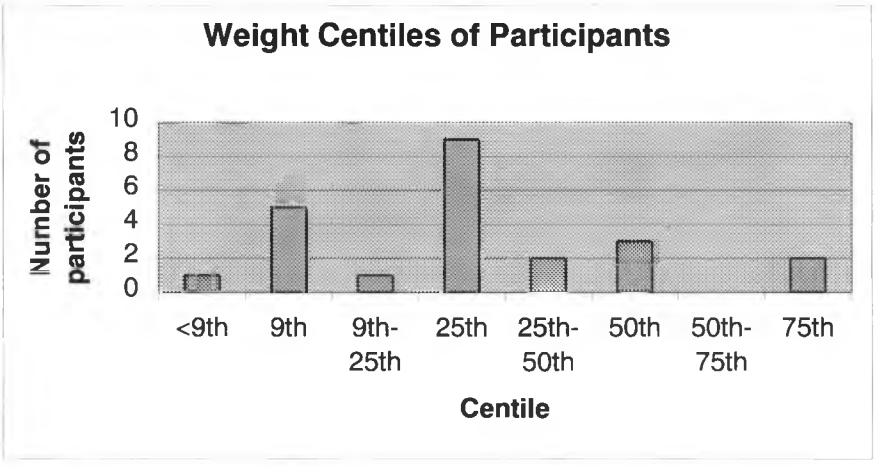


Figure 3: Weight Centiles of Participants



7.3.2 Family Demographic Data

Parental occupation was recorded and the social class of the family of participants was determined from this in accordance with the British Registrar General's grading scheme. The classification is derived from two types of information, occupation and status in employment. The classification used in the 1990s by the office of National Statistics (Griffiths & Fitzpatrick, 1999) is shown below:

Social class 1: Professional e.g. accountants, lawyers, doctors etc

Social class 2: Managerial and technical/intermediate e.g. proprietors, sales managers, researchers etc.

Social class 3: Skilled e.g. plumbers, clerks, production fitters etc.

Social class 4: Partly skilled e.g. warehousemen, cashiers (retail) etc.

Social class 5: Unskilled e.g. building labourers, cleaners etc.

The social class of participants in this study (determined by parental occupation) is presented below in Figure 4. The social class of participants is then compared in Table 3 to the national figures for social class presented in the General Household Survey 2000 published by the Office of National Statistics.

Figure 4: Social Class of Participants

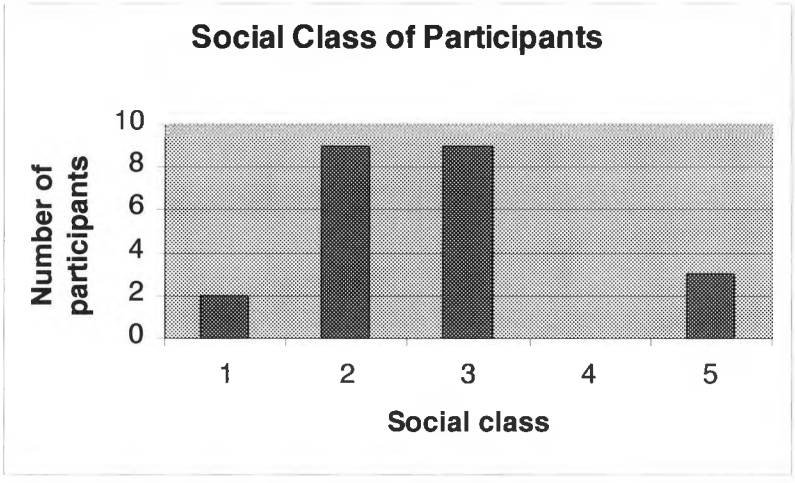


Table 3: The social class of this sample compared to the British population

Social Class	This sample	Great Britain
1	9%	5%
2	39%	16%
3	39%	55%
4	0%	18%
5	13%	6%

This pattern is likely to be an accurate reflection of the population attending the South West Thames cystic fibrosis clinic. The clinic is located in an "Urban Fringe" area between London and the Home Counties, in an area recognised by the Office for National Statistics as one of the fifteen most prosperous areas in Great Britain.

Data covering family status was also collected. It was noted whether participants came from a family where the parents were living together or had separated. Four of the participants came from families headed by single mothers. Three had undergone a divorce and one had never been married. The parents were married and living together in the other 16 families. Data from the General Household Survey 2000 indicates that across Great Britain 74% of families are headed by parents who are married or cohabiting while 26% are headed by lone parents. The figures from this sample are that 80% of the participants come from families where the parents are together and a single parent heads 20% of the families.

7.4 Representativeness of the Sample

The sample used in this study does not provide a fully representative cross section of the general paediatric cystic fibrosis population. The participants were all recruited from one hospital in an affluent area of Great Britain. Of the families in the study the percentage coming from social classes 1 and 2 (48%) is more than double the percentage of families in social classes 1 and 2 within the general British population (21%). Only 13% of the families in this study came from social classes 4 and 5 whereas the general British percentage is 24%. There was a less marked difference between the sample and the general population in terms of family status, although

there was still a lower percentage of single parent households within the sample population (20%) compared to the general British population (26%).

There were also differences in terms of health data between the sample used in this study and the Scottish paediatric CF population. Since the year 2000 health data on all patients from the UK with cystic fibrosis has been collected by the CF Database based in Dundee. No data has yet been published but figures have been collated for the Scottish paediatric population and were provided to the researcher by Drs Anil and Gita Mehta of the CF database. Dr Anil Mehta reports that the Scottish population represents 10% of the UK population and provides an accurate reflection of the UK demographic figures. The participants in this study were in better health than the Scottish CF population. There were no participants in this study classified as being severely affected by CF i.e. having a lung function below 40% whereas this group makes up 6% of the Scottish CF population. 11% of the Scottish population has a Body Mass Index (BMI) below the 5th centile compared to only 4% of the sample in this study. In addition, 74% of participants in this study had not required hospitalisation in the year prior to the study whereas only 55% of the Scottish population had not had any episode of hospitalisation in the year 2001.

This sample also failed to obtain a gender split representative of the general CF population. The Scottish data is that 51.5% of the paediatric CF population is female and 48.5% is male. Within this study 61% of the participants were female and 39% were male. While these figures are an accurate reflection of the gender divide within the 40 children and adolescents that were eligible to take part in this study, it does not reflect the more even gender split within the general CF population.

7.5 Conclusion

The sample used in this study was more affluent, healthy and had a higher proportion of female participants than the CF paediatric population as a whole. The participants were also all recruited from a single treatment centre by a researcher who provides a clinical service to this centre. The study would have been strengthened had it obtained a more representative sample from a larger number of treatment centres.

CHAPTER EIGHT: QUANTITATIVE RESULTS

8.1 British Picture Vocabulary Test (BPVS)

The BPVS provides a measure of each participant's mental age. Table 1 in the previous chapter presents the chronological and mental ages of each participant in this study. While the mean chronological and mental ages matched for both males (CA=9, MA=9) and females (CA=10, MA=10), only nine of the twenty-three participants (39%) actually had matching chronological and mental ages. Three participants had mental ages that were one year above their chronological age. Three participants had mental ages that were two years below their chronological age and eight had mental ages that were one year below their chronological age. While it is not the remit of this study to explore the impact of cystic fibrosis upon cognitive development, it is interesting that the number of participants with a mental age below their chronological age is almost four times as high as the number of participants with a mental age that is higher than their chronological age.

8.2 Medical Compliance Incomplete Stories Test (MCIST)

All participants were asked to complete this test despite the fact that it has only been used with teenagers previously (Csajkowski & Koocher, 1987). Children aged seven and under were unable to complete the test in a meaningful way. Five of those interviewed (55%) failed to complete the whole test, either saying that they did not know the answers, or making comments such as "that one is too hard". The other four (44%) managed to complete all the stories, but tended to provide very brief responses from which it was hard to obtain meaningful data. Their response to prompts for further details tended to be that they did not know any more. For example, subject 16,

a five-year-old girl gave the following response to story 4 *"Jill decided to be brave because she didn't want to cry"*. The prompt 'what happened next to Jill?' elicited a further response of *"She still felt poorly, she got poorlier"*. The limited content of her answer and its fairly tangential relationship to the original story thus made it hard to rate her answer. A decision was therefore taken to remove the data provided by children aged seven and under from the statistical analysis.

Eight to eleven-year-olds on the other hand were all able to complete all five stories that were presented to them. One nine-year-old boy gave pretty fanciful answers such as *"He took the tablets and they made him much better. Then one day the dog took the tablets too and it was much better. (laughs) I am just imagining, the dog felt much better and the parents didn't know why it was OK"*. This may have been a result of tiredness, he was observed to be losing concentration and becoming fidgety, although he did not accept the researcher's offer to stop. However, the other four participants within this age group all provided relevant if fairly brief responses. For example, an eight-year old girl completed story 1 about a child being told to have a booster vaccination as follows *"He started screaming because he was scared and he was trying to get away. But when it was done he said 'that didn't hurt' and he wasn't scared of injections anymore"*. It was decided that the answers provided by this age group did indicate that they had understood the task that they were given, so their responses were rated and the data included within the quantitative analysis.

All participants aged twelve and over also completed all five stories presented to them. It was possible to rate their answers, but the subjective impression of the researcher was that many of the answers lacked the depth or thought that participants

had displayed when answering the 'I can help others learn about cystic fibrosis' booklet. For example, subject 20, a 15 year old girl who talked in considerable detail about her CF related fears when answering questions from the booklet, was less focused in her response to story 3 of the MCIST. This story is about a boy who is advised to have his foot amputated in order to save his life. She completed the story in the following way *"I don't know, it is horrible. His foot gets cut off and he is sad but he gets used to it. But he still feels sad from time to time".*

The MCIST was the last measure to be presented to participants, often after they had been taking part in the interview for up to an hour. Participants may well have been physically tired as well as emotionally drained. The 'I can help others learn about cystic fibrosis' booklet which they had just completed did require much from participants, and it may be that the demands of the MCIST, with its quite distressing and extreme content, in addition to the previous measures were too great. While younger children simply said that they did not want to or did not feel able to complete the MCIST stories, the older children may have felt inhibited from doing so. Instead they may have tried to rush through the MCIST without giving it the thought that they would do had they been presented with it first.

In addition, the researcher was also tired by the time the MCIST was reached. The demands of writing down verbatim the answers to the 'I can help others learn about cystic fibrosis' booklet while at the same time monitoring participants' reactions to it felt quite considerable. Thus at the point of administering the MCIST her concentration may have been somewhat reduced, and her ability to prompt appropriately in order to obtain the most meaningful responses from participants may

have been diminished. This does represent a clear order effect which would have been eliminated had the order in which the measures were presented been randomised, and this should be taken into account in future studies.

8.3 Family Environment Scale (FES)

The FES results from this study are presented below beside the results for normal and distressed families that Moos and Moos (1994) present in the FES manual.

Table 4: FES Means for CF, Normal, and Distressed Families

Subscale	CF	Normal	Distressed
Cohesion	7. 25	6. 73	5. 25
Expressiveness	5. 8	5. 54	4. 71
Conflict	3. 55	3. 18	4. 02
Independence	6. 4	6. 66	6. 03
Achievement Orientation	4. 85	5. 47	5. 33
Intellectual-Cultural Orientation	6. 05	5. 56	4. 62
Active-Recreational Orientation	5. 95	5. 33	4. 15
Moral-Religious Emphasis	4. 25	4. 75	4. 51
Organisation	4. 75	5. 47	5. 07
Control	4. 55	4. 26	4. 61

There are some interesting differences between CF and normal families, such as the higher levels of cohesion, intellectual-cultural orientation, and active-recreational orientation, and lower levels of achievement orientation, moral-religious emphasis and organisation amongst CF families. However, it should be noted that none of the scores for the CF families fall outside one standard deviation of those obtained by the normal or distressed families. The trends identified within this study could be explored further with a larger sample size and a British control/normal family

population in order to determine whether there are indeed any significant differences in family environment between families with a child with cystic fibrosis and those without. Comparisons could then be made between a British and American population as Wilson, Fosson, Kanga & D' Angelo (1996) have already presented data upon the family environment of 31 CF families from Kentucky. It is worth noting that the trends within this study of higher levels of cohesion, expressiveness and control amongst CF families is similar to that of the Wilson et al. (1996) study, although, perhaps as a result of their slightly larger sample size, they found significant differences between CF and control families on these measures.

8.4 The I Can Help Others Learn About Cystic Fibrosis Booklet

8.4.1 Feedback from Participants and Parents

Participants were asked informally for their opinion of the booklet upon completion. As the researcher asked for verbal feedback and participants knew that she had designed the booklet herself, the feedback is unlikely to be impartial. A more appropriate way to have gathered such feedback would have been to ask participants and their parents to complete an anonymous questionnaire and return it in a stamped addressed envelope.

The researcher did assure participants that she wanted to hear their real views of the booklet and that if they had any criticisms or suggestions for changes it would be very helpful to hear. She also probed in particular about whether participants had found the questions boring or upsetting.

Feedback was recorded from 18 participants. Seven participants aged eight and under gave feedback which was almost all positive. The most frequent comment was that the questions were *"good"*. These were then elaborated by several children with comments such as *"they made me feel good"*, *"they were good because of the thinking"* and *"it was quite good for me understanding my CF and getting in good shape"*. One four year old girl said *"The questions were good, they are alright to ask a four year old. They made me feel sad but I am alright now"*.

A nine year old boy fedback that the questions were *"Boring, very boring indeed although some of it was quite funny. They weren't annoying or upsetting"*. And a sixteen year old girl said that the booklet felt as if *"it was probably directed to younger people than me, but it was OK for me. You can't make it too complicated or people won't understand them"*. While a fourteen year old girl made the following suggestion *"the questions covered most things but it should cover how friends feel, they might think she is taken over by it"*.

Apart from the three comments above, the feedback from the eleven participants aged nine and over was all positive. There were numerous remarks such as *"It was pretty important"* and *"it was really interesting"*. Comments such as *"It highlighted the main point about CF and makes you think about it. It is quite good to give your opinion"*, *"Some of the questions were quite hard but they make you think about it and make you realise how you have to take care of yourself and how you can only give advice if you are doing it yourself"* and *"it made me proud, I didn't know I knew all that, no one has ever asked me those questions before"* reflect the thoughtful approach taken by participants.

None of these participants claimed that the questions were boring or upsetting. One girl said that they felt relevant to her, another that *"At first they were a bit general but then they became more personal even though we were talking about Sophie. They weren't really upsetting, talking about someone else helps"*. Another teenager said that the questions *"were not really babyish, simple words are better, they make the questions easier to answer and the pictures make it amusing"*.

Feedback from the parents was not directly sought or recorded verbatim. However some parents gave their views to the researcher despite this. As with the children their feedback was almost all positive. One mother was concerned that her son had not answered certain questions correctly and said that she felt he had not conveyed the full extent of his knowledge. Others however had a different reaction, with several saying that they were surprised by how much their child knew. Others also commented on the way in which their child opened up and discussed CF in a way that they did not usually. All parents, including those who felt that it was not appropriate for them to be present during the interview, were interested in the content of their child's answers and many wanted to know if their child's understanding was age appropriate or if there were major gaps that they should be addressing.

8.4.2 Reliability

The reliability of the scoring system for the 'I can help others learn about cystic fibrosis' developed by the researcher was examined. As described in the methodology two independent raters, a clinical child psychologist and a cystic fibrosis nurse specialist were asked to complete two tasks. Firstly, they were asked to

examine the clusters that the researcher had established and to rank them numerically in terms of levels of sophistication (Ranking). Secondly they were presented with the answers to each question provided by six participants and were asked to place them into one of the clusters/categories of answers already established by the researcher (Categorisation). The results from the two raters and the researcher were then analysed using the Pearson correlation coefficient. The results from each task are presented below.

Ranking: The correlation between the researcher and rater 1 was 0.87, while the correlation between the researcher and rater 2 was 0.89. The correlation between raters 1 and 2 was 0.88. These figures are quite high, and reflect a high level of inter-rater agreement. However, it should be noted that a corrleation is a measure of association and that it does not record exact agreement. In the case of this study, the level of exact agreement between the researcher and rater 1 was 63%, but where there were disagreements about which number to assign to a cluster, the discrepancy between the researcher and rater 1 was small. Thus while rater 1 and the researcher disagreed in 41 out of 110 cases, in only 10 of these cases did their number assigned differ by more than one. Similarly, the level of exact agreement between the researcher and rater 2 was 74%, but the ranking that they assigned to a cluster only differed by more than 1 on seven occasions.

Categorisation: The correlation between the researcher and rater 1 was 0.96 and between the researcher and rater 2 was 0.94. The correlation between rater 1 and rater 2 was 0.93. As described in the ranking section, a correlation does not provide a figure of exact agreement but rather one of association. The researcher and rater 1

agreed which category a participant's answer fell into in 121 out of 132 cases (92%), of the eleven cases where they disagreed, in only three of those cases did the category that they assigned differ by more than one. The researcher and rater 2 agreed exactly in 112 out of the 132 cases (85%), but had a discrepancy of more than one in only three of the twenty cases in which they disagreed.

These reliability figures are extremely high and indicate that the coding system established by the researcher is able to be applied successfully and consistently by different raters. Despite this very high level of inter-rater reliability both raters provided informal feedback that they had found the process of ranking the categories of answers in terms of sophistication quite difficult. They said that the process might have been simpler if they had been given copies of all the participants' answers. For example, both raters interpreted the category 'I don't know' in question 13 "Will Sam/Sophie have children of his/her own?" to be a sign of the participants' uncertainty about whether they would be able to have children, whereas in fact it was an inability to answer the question. This therefore had an impact both upon the categories into which they placed the answers of participants and the rank that they gave the categories. The concerns of the raters, although unnecessary, did reflect the fact that there was by no means a perfect match between raters and the researcher on either task. However, as already described, where differences did occur they almost all differed by only one point on the scale for example placing an answer into category 3 when the rater had put it in category 4. As the reliability figures make clear, there were no examples of any extreme differences of opinion between either of the raters or the researcher on either task.

8.5 Statistical Analysis

By clustering the answers given by participants to the questions in the 'I can help others learn about cystic fibrosis' booklet and ranking the clusters numerically, (as described in the methodology), it was possible to undertake a quantitative analysis of the data from the booklet. The scores obtained by participants to each question were added together to give them a full CF knowledge score. As noted before, the lowest score that a participant could obtain was 22, and the highest was 111. The full knowledge scores of this group ranged from 37 to 108. It should be noted that a score of 111 does not reflect a perfect knowledge of cystic fibrosis but is simply the sum of the most detailed and accurate answers provided by this particular group of participants.

8.5.1 Multiple Comparisons

Participants' full CF knowledge scores were correlated with 24 other variables, using the Pearson correlation coefficient. The other variables that knowledge was correlated with included chronological and mental age, health and social variables, the three subsets and the full score of the MCIST, and all ten categories of the FES. Daly, Bourke & McGilvray (1991) write in their book Interpretation and Uses of Medical Statistics that "If a set of data is dredged for any significant relationships without reference to the purpose for which the study was set up, some relationships will appear statistically significant purely due to chance". Similarly, Armitage & Berry (1987) write in their book Statistical Methods in Medical Research "if the data are subjected to what is sometimes called a dredging procedure-a search for significant contrasts which would not have been thought of initially-there is a real danger that a number of comparisons will be reported as significant, but that they will almost all

have arisen by chance". They go on to explain that "a number of procedures have been devised to reduce the chance of this happening. They are referred to as methods of making multiple comparisons or simultaneous inference". They conclude by saying that "multiple-comparison methods are deliberately conservative in order to reduce the probability of too many significant differences arising by chance in any one study". A common approach to managing the issue of multiple-comparisons is to increase the level of significance viewed as acceptable, and specifically to divide p 0.05 by the number of comparisons/correlations that were carried out (the Bonferroni connection). In this study it would mean that only those correlations in which p=0.002 would be seen as significant. However, statisticians and academic psychologists advised the researcher that this is a very conservative and quite draconian approach and that it is common practice to simply increase the level of significance deemed to be acceptable from p 0.05 to p 0.01. This more pragmatic approach seems to be particularly appropriate in this study given that a "dredging" procedure did not take place. The vast majority of the correlations that were carried out were directly related to the purpose of the study, with only five of the FES subtests and three of the MCIST subtests not being referred to in the hypotheses. However, it must also be recognised that 24 comparisons is a considerable number, and there should be some adjustment made to the level of significance deemed acceptable.

8.5.2 Statistical Analysis

Given a level of significance where (p=0.01), positive correlations were found between knowledge and chronological age (r = 0.94) and knowledge and mental age (r = 0.90).

There was no significant correlation between CF knowledge and compliance with treatment. No family environment factors were found to be significantly correlated with participants' knowledge of cystic fibrosis. Similarly, demographic variables such as social class or family structure, and health variables such as lung function, height, weight and number of hospitalisations, showed no association with CF knowledge scores.

The effect of gender upon knowledge scores and compliance with treatment was explored using *t*-tests, but no effect was identified.

CHAPTER NINE: QUALITATIVE DATA

9.1 Introduction

While a quantitative analysis of the data provides important information about the variables that interact with knowledge of cystic fibrosis, the qualitative details contained within the answers from the 'I can help others learn about cystic fibrosis' booklet were also an important feature of this study. The role of qualitative analysis within paediatric psychology is becoming increasingly prominent. Eiser (1996) suggested that "the dichotomy between quantitative and qualitative approaches is unhelpful ... often it will be most beneficial to combine the two approaches". As she points out "children have their own views, and these need to be sought and considered", but the small sample sizes involved in paediatric psychology "does not lend itself easily to the traditional scientific approach ... we may need to rely on qualitative approaches to elicit information from children".

9.2 Qualitative Research in Paediatrics

Fiese and Bickham (1998) have argued that qualitative research provides an opportunity within paediatric psychology to gain a better understanding of relatively rare medical conditions and to focus upon the meaning of events for patients and their families. In addition, "qualitative research also holds promise in exploring the perspectives of the often overlooked consumers of pediatric care, the children themselves". According to Todd (1998) "the same data set, for example interviews, could be analysed according to both qualitative and quantitative means .... Qualitative methods can be used to add richness and detail to a quantitative study".

Fiese and Bickham (1998) describe qualitative research at its most basic level as "a process by which the data of words and pictures are analyzed in a systematic way". In this study the tape-recorded interviews with children with cystic fibrosis were transcribed and the verbatim material was examined. The clustering of answers to each question allowed a quantitative analysis to take place. But it also illustrated the variety of ways in which the questions could be answered as well as providing an idea of how the answers developed in terms of depth. Examining individual answers ensured that the variety of language and detail used by participants could be recognised at the same time as identifying individual misconceptions or errors in the understanding of cystic fibrosis.

9.3 The role of the researcher

The first step within a qualitative analysis is for the researcher to "identify any potential biases and to determine the degree of their involvement in the data collection phase [given that] all interviews are conducted in a context that includes not only the questions being asked, but also the characteristics of the relationship between interviewer and interviewee" (Fiese and Bickham, 1998). It is therefore important to note that the researcher in this study was also the clinical psychologist within the paediatric cystic fibrosis team. As a result she had met all the interviewees on numerous occasions and in many cases had provided them with considerable individual input. This may have led to the sample being skewed towards those with an interest in, and appreciation of, the role of psychological factors in coping with cystic fibrosis. Of the twenty-three participants twenty had been seen by the psychologist in her clinical role. Whereas of the five individuals who refused to take part in the study three had been offered but refused input from the psychologist.

Participants or their parents might have felt some obligation to take part in the study given the clinical role of the researcher. They might also, despite the researcher's attempts to appear neutral, have had some sense that she was looking for a certain kind of answer, possibly similar in content to her clinical input. It is also important to note that the cognitive-behavioural orientation of the researcher who designed the booklet of questions about CF is likely to have influenced the nature of the questions asked.

In addition, the fact that the researcher knew all the participants may have had an impact upon the data analysis that she carried out. The researcher knew the chronological age of each participant and may also have had preconceived ideas about their intellectual capabilities, both of which could influence the way in which she interpreted their answers. The researcher was also well aware of the hypotheses of the study, and inevitably would want the results to confirm them, thus perhaps casting doubt about the impartiality of the researcher in her data analysis role. The reliability study should go some way to challenging these concerns in that two people analysed the data 'blind' and produced results that were very similar to that of the researcher. But had the study been more methodologically tight the difficulties posed by the researcher knowing the participants, carrying out the interviews and undertaking the main data analysis would have been addressed in part by ensuring that the researcher would not have carried out all the tasks.

However, despite there being several provisos about the way in which the information was collected, the material provided by the children is rich in content and worthy of investigation.

In designing the booklet to access children's knowledge of cystic fibrosis five main areas of knowledge were explored. These were:

a) The nature of cystic fibrosis

b) Treatment requirements

c) Genetic aspects of the disease

d) The future impact of the illness

e) Feelings associated with CF.

The answers provided by the participants to each of these areas will be examined in turn. The way in which the answers develop in terms of detail and complexity will be outlined, as will patterns and themes that run through the answers.

9.4 The nature of cystic fibrosis

The first question in the booklet asked participants **"What is cystic fibrosis?"**

The answers provided by participants fell into five categories, each of which is outlined below.

Category 0: Participants said that they did not know the answer

Category 1: Participants described CF in terms of some aspect of its treatment e.g. *"you have to take your creons, you have to take your puffer, you have to have your physio, you have to concentrate with your nebuliser"* (participant G, girl aged 7).

Category 2: Participants described CF as an illness affecting the lungs and/or digestive system e.g. *"It is an illness that can attack your lungs and stomach"*

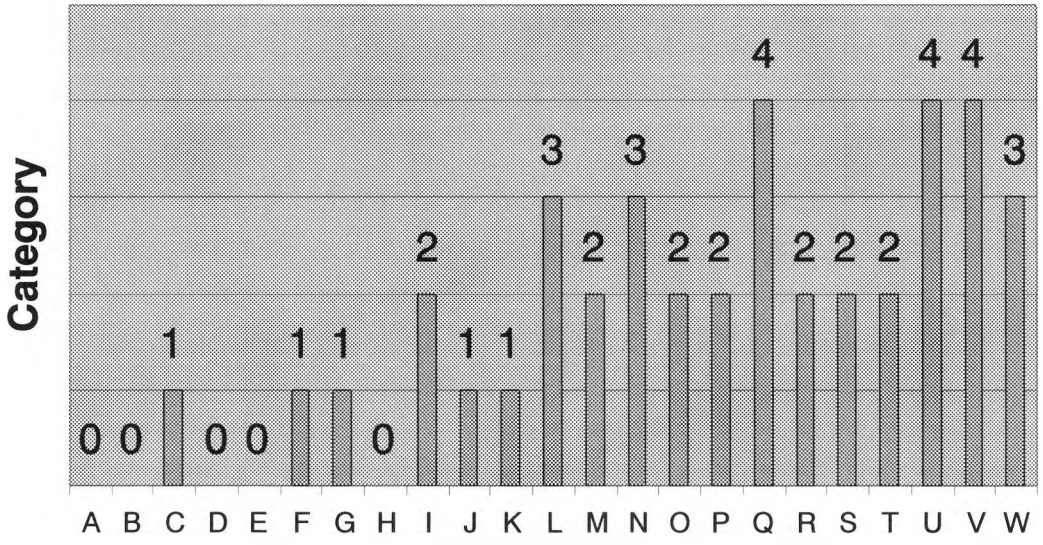
(participant O, girl aged13); *"your lungs, and parts of your intestine, the pancreas or whatever you call it"* (participant M, boy aged 11).

Category 3: Participants described CF as an illness that one is born with that affects the lungs and digestive system e.g. *"A lung dysfunction you are born with so you have trouble with your chest and breathing. You also have trouble with your digestion"* (participant W, boy aged 18).

Category 4: Participants described CF as a genetic illness affecting the lungs, digestive system and other organs e.g. *"it is a genetically transmitted disease that affects many parts of your body. The DNA for the lungs makes the mucus thicker than normal so it is harder to get rid of and to breathe. It also blocks up the pancreas so enzymes can't be released"* (participant U, girl aged 16).

Figure 5 on the next page illustrates the category that each participant's response fell into.

Figure 5: Development in understanding the nature of CF



CA	4	4	5	6	6	6	7	7	7	8	8	9	11	11	13	13	14	14	14	15	16	16	18
MA	4	5	5	6	6	6	6	7	7	7	9	8	9	10	11	12	13	13	13	13	16	17	18

Figure 5 shows that, with the exception of participant I, participants with a chronological age between 4 and 8 (mental age range 4-9) were either unable to explain what cystic fibrosis is or did so by describing it in terms of the treatment it involves. Describing CF as an illness affecting the lungs and digestive system was the most common response from participants aged 9-15 (mental age range 8-13). However, three of the nine participants within this age range gave more accurate descriptions that included the fact that they were born with CF and in the case of participant Q that CF is a genetically transmitted disease. The two sixteen year old participants with mental ages of 16 and 17 gave answers that fell into the most accurate category (Category 4) in which CF is seen as a multi-organ disease that is genetically transmitted. The eighteen-year-old, participant W, failed to mention the genetic nature of CF. The accurate descriptions of CF provided by the two 16-year-

olds and one 14-year-old appear to be very impressive. But it is possible that they could be the result of having rote learnt such definitions within a classroom setting or from a text book, and it is worth noting that of the two sixteen year olds, one wants to be a doctor and one a pharmacologist. This study does not have any direct way of distinguishing between rote-learning and genuine understanding. However, within this small group of well informed youngsters, pockets of confusion and misconceptions were apparent *"it is a genetic disease where the pancreas produces thicker mucus so it gets blocked. I'm not sure how the lungs are included but it is harder to breathe and you have to do physio to dislodge the mucus. Apparently it often goes with diabetes"* (participant V, girl aged 16).

It is interesting to note that no respondents commented upon the fact that the mucus of a person with CF is thick throughout the body. They focused instead upon the parts of the body that are most directly affected by the thick mucus and which treatment is concentrated upon. Also, even the most well informed teenagers did not describe the illness in terms of its impact at a cellular level. Thus while they have a good understanding of the manifestations of the illness, they lack an understanding of the underlying cause. Whether they need to have such an understanding could be debated. They grasp the genetic element of the condition and its impact upon their daily lives, so what an understanding of the cellular defects would add is unclear. However, it does suggest that if the three teenagers in this study with an accurate and detailed understanding of the nature of CF are producing rote-learnt descriptions, they have either not been taught or not read about the cellular defect, or they are only retaining the information that seems personally relevant to them.

An area that might begin to impact upon the lives of children with cystic fibrosis as they grow up is that of reproduction. CF affects the reproductive organs of both males and females with major repercussions upon fertility, particularly amongst males. Yet this was not mentioned by any of the respondents. The wording of the question 'which parts of the body need special care?' may have led to a focus upon treatment rather than organ involvement, although other organs, such as the liver, that are not usually directly treated were mentioned. Denial may also have been a factor, with respondents not wishing to focus upon or acknowledge the involvement of additional organ systems that could have distressing implications for their future. Or there may be a genuine lack of knowledge or limited interest in the involvement of an organ system that as yet has little relevance in their day to day lives.

What is interesting is that seven of the participants (30%) mentioned the heart as a part of the body that needs special care *"your heart has to work two times as hard"* (participant N, girl aged 11). And yet the heart is one organ that is relatively unaffected by CF. The heart remains physiologically and functionally normal in CF. Only at the very end stage of the illness are some signs of right sided growth found. However, even these changes in response to the increased demands from the lungs do not have a serious impact upon the functioning of the heart. Hence, people with CF who undergo heart lung transplantation only do so because the surgery is too complex to transplant the lungs alone, and the CF patient's heart is usually then donated to someone requiring a heart transplant. It is true that the heart rate will increase when a person with CF experiences an acute infection, just as it will in someone without CF. This increase is a normal, healthy, physiological reaction to a bacterial or viral infection and the rate returns to normal once the infection has been managed. It is

interesting that those participants who mentioned the heart as being affected by CF tended to fall within the younger age group, with no one over the age of 13 including it in their list of affected organs. As the heart is probably the first organ to be recognised by young children as critical to survival, its inclusion may therefore represent a younger child's attempt to understand the serious nature of cystic fibrosis.

9.5 Treatment Requirements

Within this section of the booklet, participants were asked a general question about what Sam or Sophie would have to do each day to keep well, and whether this would make the cystic fibrosis go away. These were then followed by more specific questions about why they have to follow certain features of treatment such as physiotherapy and taking medicines as well as why they need to attend clinic or stay in hospital.

9.5.1 The role of treatment

The responses to the question **"Will they** (the treatments) **make Sam/Sophie's cystic fibrosis go away?"** fell into four categories.

Category 0: Don't know

Category 1: Aware that treatment will not make the CF go away. For example, *"no, because you are born with it"* (participant C, girl aged 5).

Category 2: Aware that treatment is aimed at controlling symptoms rather than curing CF. For example, *"because you can get rid of the bugs but you can't get rid of CF"* (participant I, boy aged 7).

Category 3: Aware that treatment is aimed at controlling symptoms and preventing deterioration. Also aware that research is being undertaken to try to cure CF. For example, *"No, you can't make CF go away, it is all in the genes. But you can make yourself feel better and stop getting worse. Who knows, maybe one day they will find a cure"* (participant P, girl aged 13).

Figure 6 below illustrates the category that each participant's response fell into.

Figure 6. Development of awareness that treatment does not cure CF.

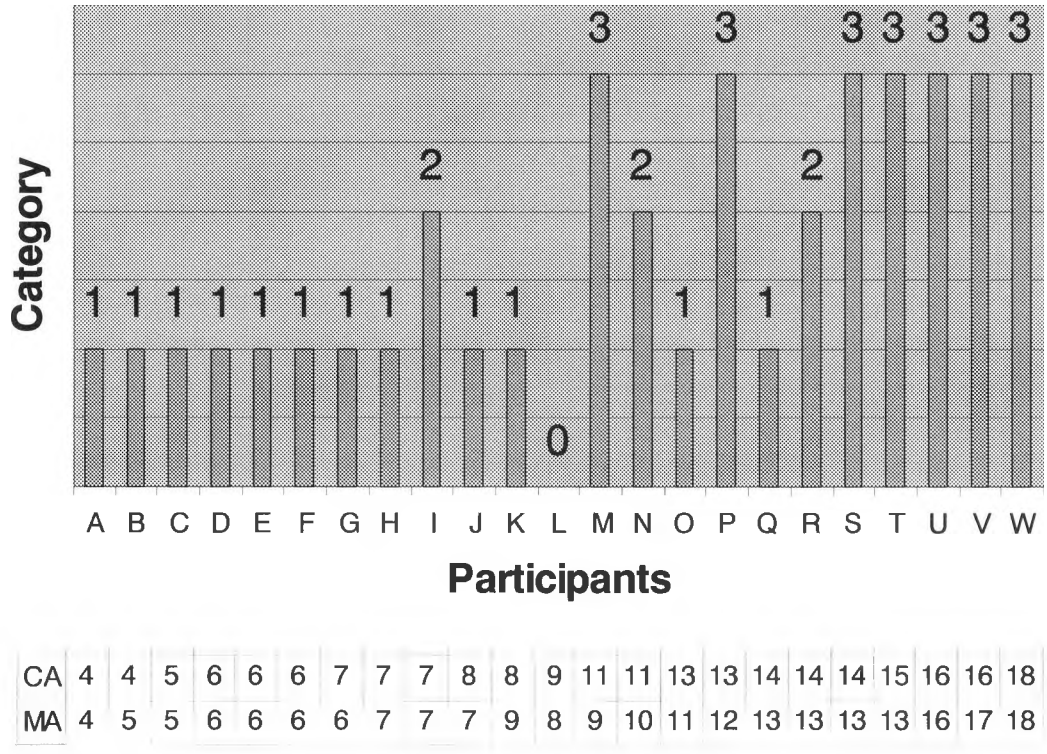


Figure 6 shows that with the exception of one 9year-old boy with a mental age of 8 all the participants in this study recognise that CF treatment does not make CF go away. This finding poses a challenge to the traditional Piagetian view that only children at the formal operational stage of cognitive development (usually aged 11 and over) grasp the concept of a chronic illness that requires ongoing treatment but is not curable. This sample had all participants except one (chronological age range 4-18, mental age range 4-18) reporting that the treatment they undertake can not cure their illness. A more detailed understanding of the role of treatment as controlling symptoms and preventing deterioration is only established within this group around the chronological age of 11 (mental age 9-10). But the findings do suggest that within this sample even the younger children still have a basic understanding of the role of treatment and do not have unrealistic or over-optimistic expectations about what their treatment can achieve.

9.5.2 The elements of treatment

All participants were able to mention at least one form of treatment *"bash bash"* i. e. physiotherapy (participant A, girl aged 4), with the majority listing four or five treatments and one 16 year old girl reporting seven daily treatments. Treatment of the digestive aspects of CF appears to be the easiest for children to grasp. Taking digestive enzymes was the most commonly reported form of treatment, being mentioned by 20 out of the 23 participants. Only two participants said that they did not know why it was important for Sam or Sophie to eat lots of food and to take tablets before eating. Even the two 4 year-old children (participants A and B) were able to explain that a failure to take enzymes before food would cause tummy aches. Physiotherapy was mentioned by 18 out of 23 participants. However, six of the nine

participants under the age of 8 (participants A, C, D, E, H, I), were unable to answer why Sam or Sophie should have medicine each day. A further seven participants were only able to give a general statement about the purpose of medication such as *"so he keeps up his strength and don't go in hospital and so he don't get ill"* (participant M, boy aged 11).

Experience may play a part in these findings. Eating food and consuming digestive enzymes beforehand is likely to be the treatment most often undertaken by a child with CF. Failure to do so also has the most immediate impact upon a child, causing stomach aches to develop rapidly. Whereas taking medication, although a daily event is not as constant a procedure as taking digestive enzymes and a failure to comply with taking medication only impacts upon the individual at a later stage.

Younger children, under the age of 12, seem to explain the rationale behind aspects of their treatment in terms of keeping well, staying healthy etc. They do not distinguish much between the different forms of treatment, and instead focus upon avoiding unpleasant outcomes. *"Otherwise he will get a tummy ache"* (participant H, boy aged 7), *"if he doesn't he will get things stuck up in his mouth and his neck. He doesn't want the line [long line for the administration of intravenous drugs] to have to go back in"* (participant J, boy aged 8).

Teenagers have a more detailed understanding of the purpose behind their treatment, although the emphasis upon avoiding unpleasant complications remains. *"Mucus can't be got rid of like normal people just breathing. Physio helps the lungs get rid of the mucus so her lungs and airways don't get blocked and stop her breathing. Mucus*

encourages the spread of infection. The more mucus she has festering on her lungs the more vulnerable she is to infections" (participant U, girl aged 16). *"She has to eat lots of food because CF people don't digest food as well as normal people so you have to eat about twice as much to put weight on. It fights infections so you don't have to go into hospital"* (participant R, girl aged 14).

All bar one six-year-old girl knew at least one reason why they had to attend cystic fibrosis clinic, with the majority being able to list at least two reasons. The most common explanation was that Sam/Sophie needed to go for a check up, but medication changes and lung function measures were also commonly cited as reasons for going to clinic. One fifteen year old girl (participant T) presented six reasons for going to clinic *"so the doctor can see how well she is and if she needs more help like new tablets or treatments so she doesn't get iller. So she can have x-rays and phlegm samples to see what bugs are growing in her lungs. To have a lung function to see how well her lungs are. To see the dietician to see if she needs to change her diet and to see her weight and her height. And to see her therapist if she has got any problems"*.

The cohort was a relatively healthy sample. Only six of the twenty-three participants (26%) had been hospitalised in the previous year, with many of the others not having been admitted since birth. However, only four of the youngest participants (aged 7 and under) said that they did not know why Sam/Sophie might need to stay in hospital. Failing to follow treatment plans was mentioned by seven and eight year olds *"because if you don't take your creons or puffer or physio or nebuliser you will get a chest infection and end up in hospital"* (participant G, girl aged 7). It was also

cited by eleven and fourteen year olds, but they also acknowledged that a deterioration in health could occur spontaneously, regardless of compliance with treatment *"because he hasn't been eating or drinking or taking his tablets. Sometimes you just have to go if you get pseudomonas or a bad cold"* (participant M, boy aged 11). However, it is important to note that some young children (five 4 to 9 year olds) did not attribute a stay in hospital to any failure on their part to follow the treatment regime. Instead they provided explanations such as *"when she has her check up there might be something wrong with her like a pain and that makes her stay in hospital"* (participant K, girl aged 8). By later adolescence, participants recognised that hospitalisation was linked to the need for more intensive treatment following a deterioration in health. *"When you catch an infection it is better to come into hospital. You need an IV drip. It's not nice, the food is appalling, but in the long run it's for the best"* (participant W, boy aged 18). They however did not comment upon any association with treatment adherence despite being the group most likely to experience a hospitalisation following difficulties with the treatment regime.

While there is no strong evidence from these results to suggest that younger children engage in magical thinking to explain a hospitalisation, the results suggest that some do blame themselves for such an event by attributing it to a failure to comply with treatment. Eleven and fourteen year olds seem to have a balanced viewpoint in which they acknowledge that non-adherence with treatment can influence health, but also that a deterioration in health leading to hospitalisation can be quite outside their control. While older teenagers on the other hand appear to avoid acknowledging the role of treatment adherence in maintaining health.

9.6 The Future Impact of the Illness

At a later stage in the booklet, participants were asked a general question about what would happen to Sam/Sophie as he/she grows up, followed by a specific question about how old Sam/Sophie would grow up to be. The answers to the question **"what will happen to Sam/Sophie as he/she grows up?"** fell into the three categories outlined below.

Category 0: Participants described getting bigger, growing up normally, often saying that they would be like their parents. They did not make any reference to cystic fibrosis. For example, *"she will grow up just like Mummy"* (participant A, girl aged 4); *"he will be a grown up, he will be a Dad, he will get married, he will look after people"* (participant F, boy aged 6).

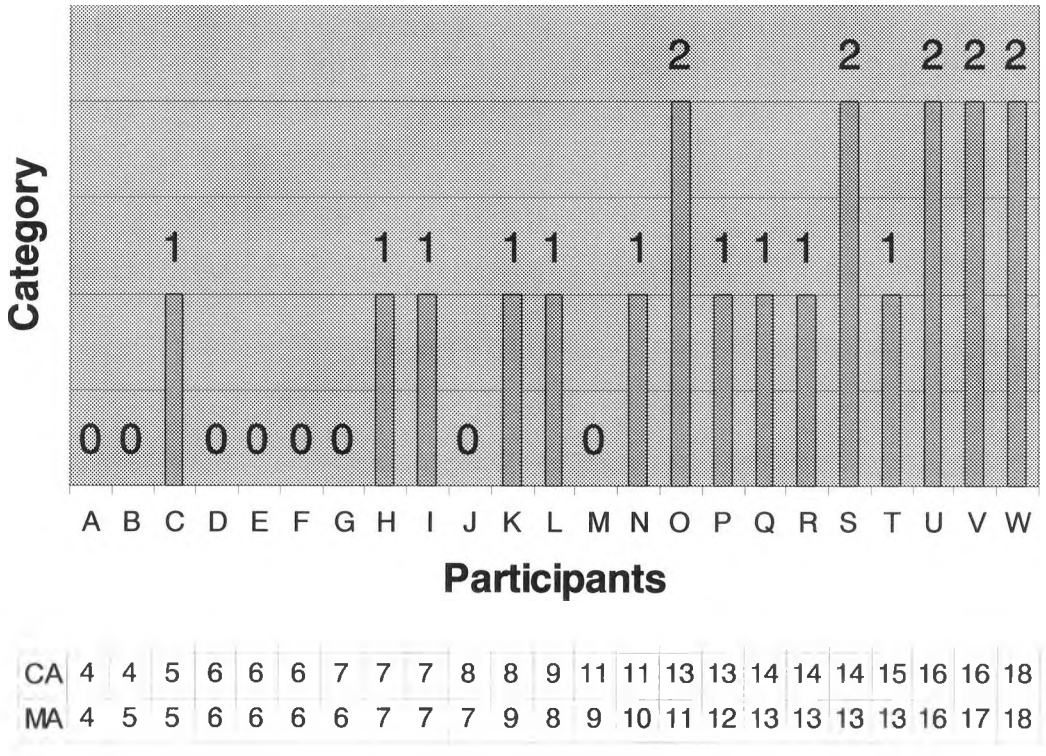
Category 1: Participants acknowledged that CF would remain with them and require increased treatment for which they would have to take on more responsibility. For example, *"She will still have to take her medicine, she will still have it but her lungs might grow so she might have to take a higher dose of turbo inhaler and Creon"* (participant Q, girl aged 14); *"She will understand more about her lungs and how to treat them. It will get harder as she wants to go out but she will learn her own responsibility. She'll do her own physio, be more independent, organise her own appointments, get her own medicine, fill in lots of forms, get a magazine called Sixty-Five Roses. She will have to explain her illness to her friends and maybe her boyfriend. Everything will be her own responsibility; she won't get*

no help. She might get upset and say why me, why aren't I like everybody else? She might ask more questions" (participant T, girl aged 15).

Category 2: Participants acknowledged that their CF would deteriorate, although they did add that this was dependent upon how well the treatment regime is followed. For example, *"unless there is a breakthrough she will always have CF. Depending on how well she keeps herself she should remain healthy but she will always have to go to hospital and keep doing her treatment. Even then she will deteriorate but hopefully her treatment will stop her getting seriously ill"* (participant U, girl aged 16).

Figure 7 below illustrates the category that each participant's answer fell into.

Figure 7 Development in understanding their future



These results show that, with one exception, the seven participants with mental ages of six and under (chronological age range 4-7) viewed their future as one that would mirror the experiences of their parents. When thinking about the future, even in this setting with its focus upon CF, they did not comment upon CF. In contrast, the three participants with a chronological and mental age over 16 all acknowledged that CF would remain with them and would deteriorate, although they did see the deterioration as being closely linked to treatment compliance.

The answers to the question **"How old do you think Sam/Sophie will grow up to be?"** also fell into three main categories.

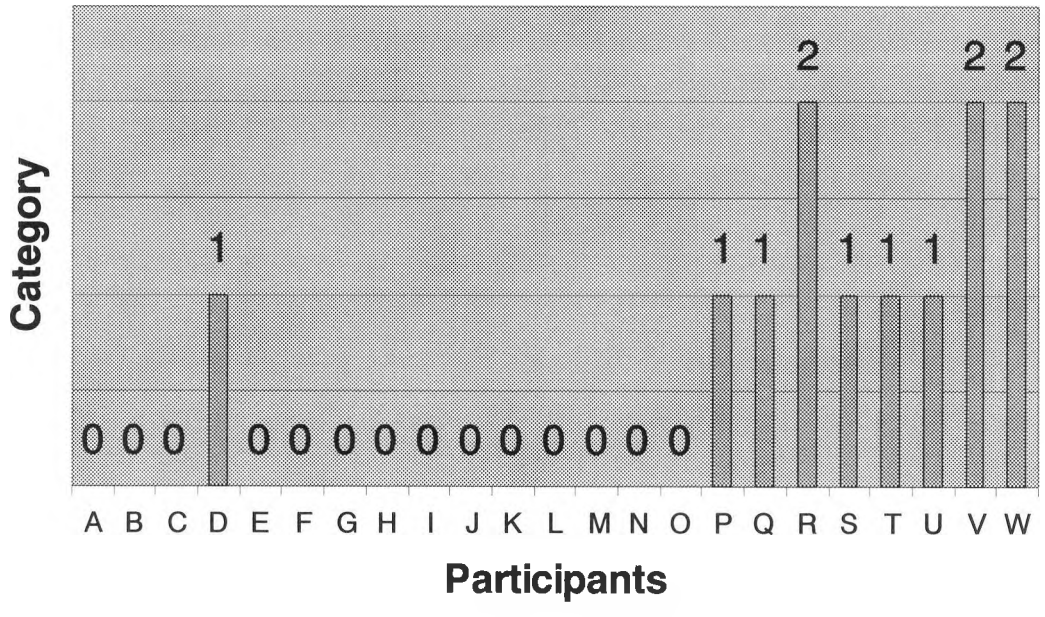
Category 0: Participants gave a wide variety of age ranges and did not indicate that they understood that CF affects life expectancy. For example *"You can only get up to 100 or 101. Mummy's granny is still alive and she is going to be 101 soon"* (participant I, boy aged 7); *"16 I expect"* (participant B, girl aged 4).

Category 1: Participants acknowledged that CF has an impact upon life expectancy, but it is hard to predict, or only occurs in certain circumstances. For example, *"With any adult it depends how well she keeps herself. If she is not doing her treatment she could get very ill and die of CF. But if she can keep herself well and stop infection then she can grow as old as any person"* (participant U, girl aged 16); *"There's no real way of saying. Ages ago there was no medication and children died at 3 months. Now the medicine is getting better and better so you can't really pick an age there is no way of knowing"* (participant P, girl aged 13).

Category 2: Participants reported the accurate life expectancy figures for someone with CF. For example, *"Around 45, but it depends how bad your CF is and if she has a car crash"* (participant R, girl aged 14); *"That depends on how bad her CF is, how well and how often she has her treatment. The average age is now around 40, but treatment is improving all the time. I'm going to live to 96. I might not live as long as other people but I'm not going to die at 20 which is what I was told in a biology class. I was told it was about 20 but that's not true"* (participant V, girl aged 16).

The category that each participant's response fell into is illustrated below in Figure 8.

Figure 8: Development in understanding the impact of CF on life expectancy



CA	4	4	5	6	6	6	7	7	7	8	8	9	11	11	13	13	14	14	14	15	16	16	18
MA	4	5	5	6	6	6	6	7	7	7	9	8	9	10	11	12	13	13	13	13	16	17	18

Figure 8 shows that within this sample the knowledge that CF has an impact upon life expectancy develops around the chronological age of 13 (mental age 12). One thirteen year old (participant O with a mental age of 11) and thirteen of the fourteen participants aged 4-11 (mental age range 4-10) did not report that CF has an impact upon life expectancy. The only participant within this age range who did indicate that she is aware that CF reduces life expectancy (participant D) had recently experienced the death of her grandmother who had lived with her, and this does seem to have led to some focus upon the consequences of her own CF *"30. Nanny died when she was 60. Not as old as nanny because she's got cystic fibrosis and nanny didn't".*

Unfortunately the question "How old will Sam/Sophie grow up to be?" is not the same as asking participants directly about the life expectancy for people with CF and there is therefore some room for interpretation and ambiguity within the responses obtained. The question that participants answered does not specifically mention CF or the life expectancy associated with it. There is therefore a chance that participants might not consider CF in the answers that they provide. Alternatively, they might see themselves or Sam/Sophie as exceptions to the rule, and thus provide an answer that does not appear to indicate a knowledge of the life expectancy associated with CF despite actually having such knowledge. The responses from participants over the age of 13 do suggest that they generally interpreted the question as the researcher had intended in that they convey their understanding that some people with CF do not live as long as those without it. However their answers then tend to focus upon the impossible task of predicting the point at which an individual will die. They do not therefore provide a numerical figure despite the fact that they may know the average life expectancy for the CF population. The results displayed in Figure 8 should

therefore be viewed with some caution in that they may be underestimates of participants' knowledge.

9.6.1 Coping with the knowledge of reduced life expectancy

While there may be some doubt as to the factual interpretation of the comments provided by the participants aged 13 and over, they do give a real insight into how adolescents with cystic fibrosis cope with the knowledge of their own reduced life expectancy. Outright denial does not seem to be an issue with this group of young people. Participant W simply reported a relatively low life expectancy *"between 40 to 50 seems to be the range really"*. But all the other participants P-V gave more revealing answers. Participant R talked about Sophie living to *"about 45"* but then added that *"it depends how bad your CF is and if you have a car crash"*. Participants P and V focused upon the progress in treatment that has occurred in the past and so *"now the medication is getting better and better you can't really pick an age, there is no way of knowing"*. Participants Q, S, T, and U focused instead upon *"the kind of CF"* that Sam/Sophie has and the quality of adherence to treatment they display. They indicated that if they had *"good"* CF and stuck to their treatment regime religiously then *"he should live to the average age of a normal person, hopefully"*. However, there were signs from their comments that they recognised this was unlikely. For example, they all emphasised that Sam/Sophie would have to be *"really fit"*/*"very healthy"* and they also all finished their statements about growing as old as any person with the word *"hopefully"*. The responses from the teenagers would suggest that this group is aware of the impact of CF on life expectancy, but that on a day to day basis they have a number of strategies that help them not to dwell upon this.

This finding may explain why this question was unexpectedly comfortable for the researcher to ask. Initially she approached the question with a sense of discomfort and unease for fear of upsetting the participants. Indeed it had been this anxiety that had led the researcher to word the question in the way that she did rather than asking directly about life expectancy. However, it became clear that the participants were not distressed and were quite willing to discuss this question in exactly the same style as they answered all the other questions. Any future studies may therefore wish to address the issue of life expectancy in a more open, direct and less ambiguous way.

It seems that the strategies that these participants use to contain the distress, which must almost inevitably accompany the knowledge that they will not live as long as others, are very successful. These strategies include:

- Thinking that we all face potential life threatening events
- Focussing upon developments in treatment
- Seeing themselves as exceptions to the rule
- Concentrating upon their own treatment regime
- Hoping for the best.

Perhaps the most poignant comments are the ones from subject T, a fifteen-year-old girl who had been hospitalised three times in the previous year. She said, *"Depends on how she looks after herself and how well she will be. Don't know how old she will grow up to be. Lin [CF nurse specialist] says I could be 95 with all the treatment that is coming out. I don't know, it is what I want to think"*. In those few words she conveys both a longing to believe that she has as long a life expectancy as anyone else

and a realisation that she does not. She was the only participant who went on to ask questions about this issue at the end of the interview. She asked what the current life expectancy is and what happens to people with CF as they get more ill. In addition, her response to a later question asking if there is anything else that Sophie should know about CF indicates her need for further information and support in coping with her realistic understanding of her life expectancy. *"Things about death and that. One thing that really worries you is dying. Knowing about the things that can happen to you when you get really ill, that you will not just suddenly die, those things are important for Sophie to know".*

9.7 Genetic Aspects of the Disease

In this section participants were asked **"How did Sam/Sophie get cystic fibrosis?"** Their answers fell into four categories.

Category 0: Don't know

Category 1: Participants reported that they were born with it. For example, *"She was just born with it"* (participant C, girl aged 5)

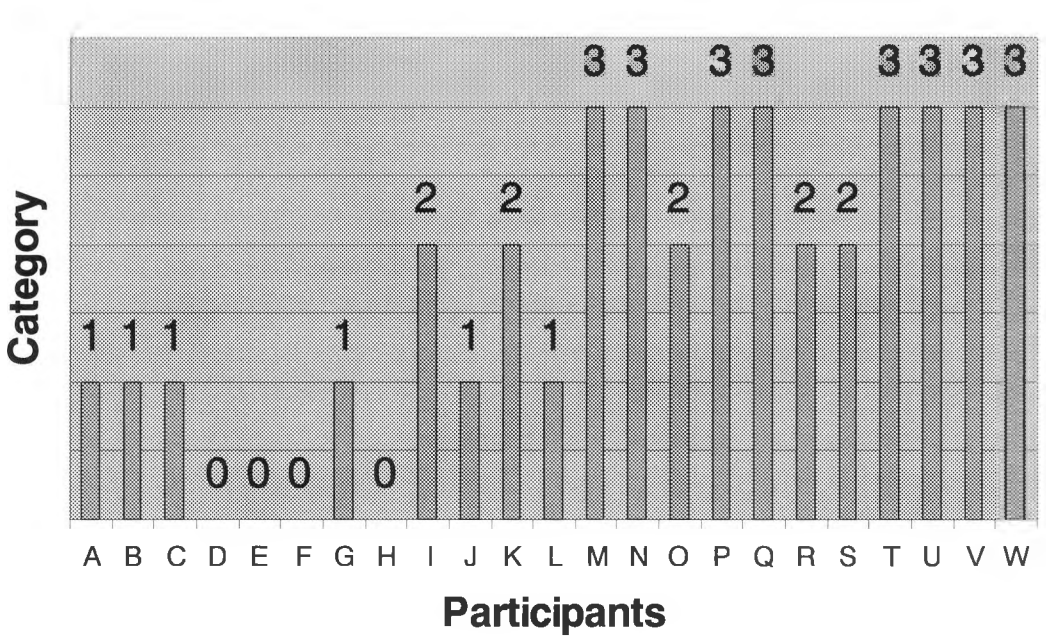
Category 2: Participants mentioned genes but were unable to display a clear understanding of genetic inheritance. For example *"It is in her genes. You get one gene from your Mum and one gene from your Dad. You can't catch CF"* (participant O, girl aged 13); *"She inherited it off her mother and father because they both have a cystic gene and so she was created as a cystic child"* (participant Q, girl aged 14).

Category 3: Participants used genetic terminology accurately and displayed a clear understanding of how CF is acquired. For example, *"She has had it from*

when she was born. If both parents are carriers of the gene, I don't know which gene it is, but if the two parents are carriers then there is a 1 in 4 chance that each child will have CF" (participant V, girl aged 16); *"she didn't catch CF, during fertilisation the CF carrier genes from her mother and father combined so that her DNA was faulty and caused her to have CF"* (participant U, girl aged 16).

The category that each participant's response fell into is displayed below in Figure 9.

Figure 9: Development in understanding how CF is acquired.



CA	4	4	5	6	6	6	7	7	7	8	8	9	11	11	13	13	14	14	14	15	16	16	18
MA	4	5	5	6	6	6	6	7	7	7	9	8	9	10	11	12	13	13	13	13	16	17	18

The results are a little erratic within the younger section of the sample. For while the two 4 year-olds and one 5 year-old (participants A, B & C) reported that Sam/Sophie was born with CF, the three six year olds and one seven year old (participants D, E, F & H) said that they did not know how Sam/Sophie had got CF. Participants G, J and

L reported that Sam/Sophie was born with CF while participants I and K tried to explain the genetic aspects of CF acquisition. However, the older section of the sample was more uniform in that all eleven participants aged 11 and over (mental age 9 and over) used genetic terminology and eight of them did so accurately. The seven-year-old (participant I) who displayed a certain level of genetic understanding did not in fact use genetic terminology but instead used age appropriate language to convey his sense of CF being an inherited illness *"He was just born with it. Because you've got bad stuff in you which creates a baddie. His Mummy gave him a part of her bad bit and his Daddy gave him another part of his bad bit and they joined together, mixed up, and made him have cystic fibrosis".*

Participants were also asked, **"If Sam/Sophie's parents had another baby would the baby have cystic fibrosis?"** The understanding displayed by the participants was much more patchy in this case. Only the two 16 year old girls indicated that they could assess the probability accurately *"Possibly but not definitely, there is a 1 in 4 chance with each baby"* (participant V), and *"Not necessarily. It depends how the genes from the mother and father mix to make the baby. It is more likely not to have CF as it is a recessive gene so you need both. It is more likely to have a dominant gene and be a carrier. If both parents had CF the baby would too"* (participant U). Twelve other participants acknowledged that there was uncertainty about whether another baby would have CF, but only three attempted to explain it in terms of genetic probability *"There is a chance, you can have 2 in 4 or 3 in 4 children. Sometimes you can have 3 children with it and your fourth doesn't so it is sort of 50-50"* (participant P, girl aged 13). The other nine simply talked in terms of chance or conveyed their own uncertainty *"I suppose so, yeah. If they are two carriers. I'm not sure really"*

(participant W, boy aged 18) and *"Not necessarily. If the next baby had CF it is just chance and could be just complete coincidence"* (participant S, boy aged 14). Most of the children under the age of eight gave definite answers, two indicating that the baby would have CF and six that the baby would not.

It is interesting to note that the 7 year old boy who conveyed an understanding of how he had inherited CF also shows a quite remarkable, though not completely accurate, understanding in response to this question. He said, *"You can't tell. My sister hasn't got CF and I have so you can't tell. Jane might have had two parts of good bits or Mummy might have given her part of her bad bit so she might not be a carrier anymore because she gave her bit to Jane and lost it all. And Daddy gave Jane his good part"*.

Following the question about parents having a child with CF, participants were then asked **"Will Sam/Sophie have children of his/her own?"** This question was trying to access not only further evidence of the children's understanding of genetic issues within CF, but also to explore their understanding of fertility. Two participants (J and H) said that they did not know the answer to this question, while fourteen (A-G, K, M, N, and P-S) said yes, Sam/Sophie would have children. Seven others (participants I, L, O, and T-W) gave answers indicating a possibility that they would have children. Not one participant said that it would not be possible for Sam/Sophie to have children.

Of those participants who said Sam/Sophie would have children, three (G, K, M) said the child would have CF *"Yes, but they will have CF. It is up to him if they have children and they want them to have CF"* (participant M, boy aged 11). Five

participants (B, D-F, and N) said the child would not have CF *"Yeah, when he is older. His children won't have CF but he will still have it"* (participant E, boy aged 6), while six participants (A, C, P-S) indicated that the outcome for the child was uncertain *"Of course he will have children. Sometimes they will have CF and sometimes not, it just depends whose genes are stronger, the mother or the father"* (participant S, boy aged 14).

The seven participants who were less certain about whether Sam/Sophie would have children generally had some idea that the health of the person with CF was a factor in determining whether they would reproduce. *"CF doesn't affect fertility directly but in order to carry a child Sophie needs to be healthy enough to support herself and her baby"* (participant U, girl aged 16). *"Possibly. From what I have been told it is possible but comes down to how well you have done your treatment and how well you are"* (participant W, boy aged 18). However, only two girls aged 15 and 16 made any mention of compromised fertility within CF *"Yeah, but I don't think males have a chance. But females may not be able to have children either. I don't really know. My Mum said I may not be able to have children, but I was little, I can't remember"* (participant T, girl aged 15). *"If she wants to. I have heard somewhere that people with CF are sometimes unable to have children, but only from a magazine. If her husband is a carrier then it is a 1 in 2 or 1 in 3 chance the baby will have CF. If both have CF the baby will too. It might be better to adopt"* (participant V, girl aged 16).

9.8 Feelings Associated with Cystic Fibrosis

The purpose of the final section of the booklet was to explore the emotional impact of cystic fibrosis upon the children with the disease and their families, from the child's

perspective. Standardised measures were not used because it would have altered the style of the interview and would not have provided the opportunities to gather the information in the participants' own words.

Initially participants were asked, **"How does cystic fibrosis make Sam/Sophie feel?"** The responses from participants fell into four categories.

Category 0: Don't know

Category 1: Participants gave a brief description of a single distressing emotion. For example, *"She doesn't want it, she feels sad because she doesn't want it"* (participant C, girl aged 5).

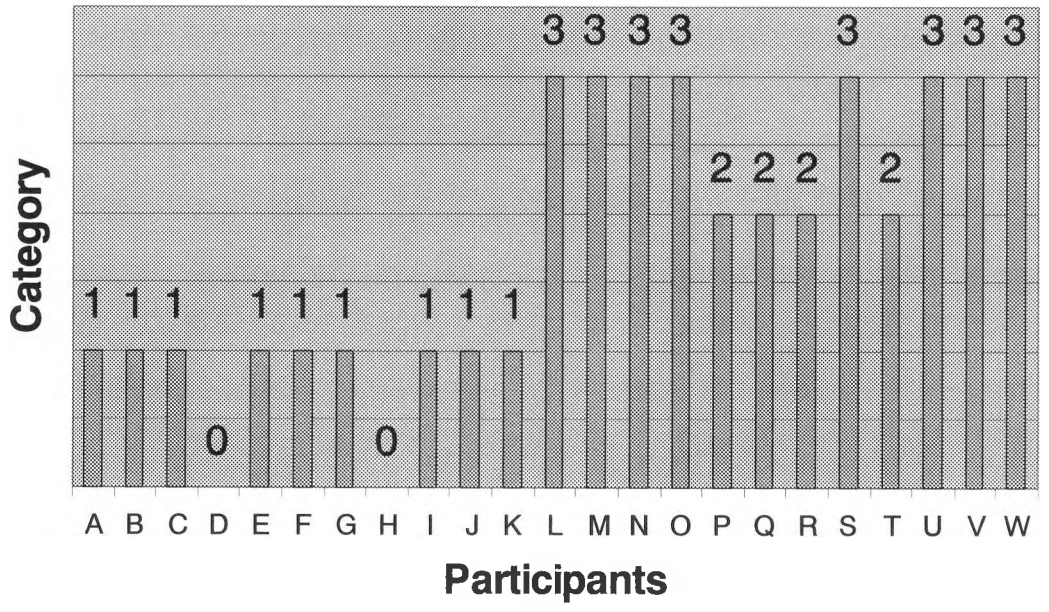
Category 2: Participants reported feeling distressed by CF for much of the time but were also able to recognise that they had episodes of feeling more optimistic. For example, *"Sometimes it can make her depressed. It might make her not very confident. She might be scared that people might take the mickey or bully her. But when she has found out quite a bit of stuff she might feel a bit more confident about what she has got and that they are trying to find a cure for it"* (participant Q, girl aged 14). *"Probably a bit upset, depressed, worried, nervous, anxious and disappointed. But sometimes she might feel quite happy when you get time off school"* (participant R, girl aged 14).

Category 3: Participants reported feeling normal for much of the time but acknowledged episodes of increased CF related distress. For example, *"It should not make him feel sad or worried. It doesn't stop you from doing anything. You just*

have to take lots of tablets, but you get used to it after a while" (participant M, boy aged 11). *"It doesn't make her feel any different from other people. She can do what they do like running and athletics. Sometimes it makes her worry and she can feel upset if friends take the mickey and say ha ha we are normal and you are not. She might worry about what happens if she doesn't do her physio"* (participant N, girl aged 11).

The category that each participant's response fell into is illustrated in Figure 10 below.

Figure 10: Development in ability to describe the emotional impact of CF upon oneself.



CA	4	4	5	6	6	6	7	7	7	8	8	9	11	11	13	13	14	14	14	15	16	16	18
MA	4	5	5	6	6	6	6	7	7	7	9	8	9	10	11	12	13	13	13	13	16	17	18

Figure 10 shows that all those participants aged eight and under either did not answer the question or gave fairly brief responses that indicated that they are distressed by CF but did not explore this in any depth. Those aged nine and over were instead much more detailed in their responses to this question and spontaneously gave examples to illustrate their feelings. Six of the eleven participants aged over 11 (participants N, P, Q, R, V and W) spontaneously commented upon fears of being bullied and having 'the mickey taken'. The researcher did not ask whether this was something that they had personally experienced, or if it was an internal concern about possible events in the future. Thus it is hard to determine whether bullying is a feature for many of these youngsters with CF or just something that they fear could happen. Either way it is an issue that may need to be addressed by clinicians more than is occurring at present.
The older group (11 and over) also either admitted feeling different from their peers, or were vociferous in their comments that Sam/Sophie should not feel different from others. Adolescence is a period when the peer group becomes highly influential upon a young person and a sense of difference or isolation can be very damaging to an individual's self esteem. It is of interest to note that the more visible manifestations of CF such as taking enzymes before eating and following a high fat, high calorie diet, were the aspects of CF that participants highlighted when talking about being bullied or feeling different from others. If treatment adherence is to be maintained during adolescence clinicians may have to devise strategies to help 'hide' its more obvious features.

Similar patterns of detail were found in the answers to the question **"How does cystic fibrosis make Sam/Sophie's parents feel?"** Children under the age of 11 gave fairly

brief, though often revealing comments about parental reactions to CF *"Sometimes they feel happy and sometimes they feel sad. They feel sad because they have to look after her. I don't know why they feel happy but they love their daughter"* (participant K, girl aged 8). *"Sad because they have to do all the work"* (participant G, girl aged 7). All the children under 11 except the two who did not answer the question (participants D and H) said that CF made their parents feel sad, but half of them also described their parents as happy as well. It is interesting that within this cohort even the two four year olds were able to recognise that CF has a distressing emotional impact upon their parents, but also that this distress is not constant and their parents are able to experience happiness as well.

The participants aged 11 and over (mental age 9 and over) were able to talk about their parents' emotional reactions to CF in some depth. They talked about the shock and distress that parents might feel at the point of diagnosis, although interestingly none of them mentioned parental guilt at this time *"I should imagine very sad at the start, almost like they felt they had got it themselves"* (participant W, boy aged 18). They described the fears that their parents had about the future and the possible premature death of their child, *"Sometimes my Mum thinks, before I had my inhaler to stop my coughing, she used to wonder if I would be there in the morning"* (participant N, girl aged 11). *"Probably at times of serious illness they would feel very stressed about what will happen as they don't have any control"* (participant U, girl aged 16). There were numerous comments about parents feeling protective of their child with CF *"very protective over Sophie"* (participant Q, girl aged 14), *"I think they would probably be more protective of her and worried about her health"* (participant V, girl aged 16). These were also often coupled with descriptions of the pressure that parents

are under *"They have to help her so it can't be easy for them. When she is in hospital they have to visit her all the time and they might miss work and there can be a lack of pay"* (participant P, girl aged 13). *"It is harder for them to care for her so they get more tired"* (participant V, girl aged 16). *"Maybe a bit pressured so then they pressure Sophie to do things. They may be wary of doing new things"* (participant T, girl aged 15).

These participants do appear to be very perceptive about their parents' feelings. However, some of their comments suggest that parents may be trying to use strategies to shield their children from their own feelings. Certainly some of the adolescents within this sample appear to be well aware of their parents distress even though their parents may try to hide it, *"They put a brave face on in front of the person with CF"* (participant R, girl aged 14); *"I don't know what my Mum and Dad feel, they don't open up. She feels let down, like her hard work is going to waste"* (participant T, girl aged 15); *"I know my parents worry about me and my CF but they don't like me to know that they worry"* (participant V, girl aged 16).

An interesting gender difference is that the older boys, while able to recognise their parents' distress at the time of diagnosis, go on to say that over time that distress diminishes. *"It takes a while to come to terms with it, but they get used to it"* (participant W, boy aged 18). *"Once they see him taking his tablets and living a healthy life they will feel pretty confident, they will feel OK"* (participant S, boy aged 14). The older girls on the other hand were able to acknowledge that their parents' distress was ongoing. Perhaps the parental strategies of hiding their feelings from their children are more effective with boys than girls.

Participants were then asked, **"How does cystic fibrosis make Sam/Sophie's brothers and sisters feel?"** Children under the chronological age of 11 tended to use the same brief descriptions as they had used for their parents, i. e. sad or sad and happy. However, their answers indicated that several of them were able to distinguish the reactions of their siblings from those of their parents. *"A little bit sad because they don't like him having it because they know what it is like, they have seen what he has to do and they think oh no that is tiring and it is"* (participant I, boy aged 7). *"Sad because they have to put up with her coughing all night and happy because they get all our food, the high fat stuff"* (participant K, girl aged 8). Four participants (M, N, P, and S) had some difficulty in acknowledging that siblings might be affected by CF *"It shouldn't really affect them unless they have CF too"* (participant P, girl aged 13). *"Basically they should just treat him as normal, like a normal person in a normal loving family"* (participant S, boy aged 14).

Eight participants (K, L, M, Q, R, U, V, W) ranging in age from 8 to 18 mentioned that siblings might feel jealous of the person with CF. *"Probably quite jealous that their parents spend time looking after her and if she goes into hospital they might stay with her. When Sophie grows up they might resent her in some way, she has had her mother and father look after her and treat her in a different way"* (participant O, girl aged 13). However, half of them also acknowledged that their siblings also experienced other, perhaps more sympathetic, feelings towards them, *"If he goes on a CF holiday and they can't come, they feel a bit jealous, but they are also relieved that they don't have to take tablets. They might also feel worried about him, and they still love him even though he has got CF"* (participant M, boy aged 11). In addition, three

girls aged between 11 and 14 described their siblings as feeling sad, anxious and protective towards them, *"They will feel protective of her and a bit scared that she might die or whatnot"* (participant Q, girl aged 14).

The final section of the booklet asked if there were aspects of the disease that Sam/Sophie might like or dislike, if there were important aspects of cystic fibrosis that had not been mentioned so far, and who they could talk to if they wanted to know more about CF. In response to the question **"Are there any things Sam/Sophie may like about cystic fibrosis?"** nine children, (participants A-D, G, J, L, M and W) all aged under 11 apart from participant M (CA, 11; MA, 10) and participant W (CA 18; MA 18) said no. *"Not really, there is nothing nice about it. Ask anyone, they wish they didn't have it. There is nothing nice but you just go along with it, let nature take its course"* (participant W, boy aged 18). Two girls aged 5 and 6 (participants C and D) said that they liked their treatment because it made them feel better *"She feels happy about taking her nebuliser because she wants to get better"* (participant C, girl aged 5). Twelve participants were able to acknowledge certain advantages associated with having CF such as going on CF holidays, to CF parties, missing school, having a high calorie diet, and not getting fat. However, there was a sense in which the listing of advantages did not appear very convincing to the participants, and their reaction was summed up by one 13 year old girl who said *"You get holiday funds, you get to eat lots of crisps and chocolates. When you are in hospital you get lots of attention and presents. It makes your friends jealous, they say I wish I had it and I think no you don't"* (participant P).

The question **"Are there any things Sam/Sophie may not like about having cystic fibrosis?"** at first sight seems somewhat repetitive as participants had been indicating their dislike of CF throughout the interview. However, in fact it allowed them to summarise their feelings towards CF, to explain in their own words the aspects of the illness they disliked the most, and in some cases it also provided additional new information about coping with the disease. The younger children, those under 9, tended to focus their dislike upon the daily treatment routine *"Yes, bash bash [physiotherapy], medicine, and getting poorly with it"* (participant A, girl aged 4). A mixed age range of children focused upon invasive treatments and physical deterioration as the element that they most disliked *"Having to put the line in, it hurts. He hates having to have his tablets, it is annoying to sit at a table and have to take tablets before eating"* (participant J, boy aged 8). *"If he gets really ill and goes into hospital and has all these injections. But with time he will get used to it. Having to take lots of tablets but he gets used to that too"* (participant M, boy aged 11).

Nine other participants (K, M, O-S, U, V) all teenagers apart from participant K (CA 8; MA 9) and participant M (CA 11; MA 9) commented upon both practical and emotional aspects of the disease that they disliked. *"All the physio and tablets, when you are not feeling well and have to go into hospital. All the puffers as well. Also the disadvantages, you can't run as fast, you can't grow too fast. The worries of your life expectancy with CF and when you are not well, the general worry"* (participant R, girl aged 14). Other worries that were reported by this group included:

- Being treated like a baby/sympathised with/fussed over,
- Being teased or bullied,
- Feeling embarrassed in public,

- Having to explain about CF to others,
- Feeling different from their peers,
- Being away from home,
- A lack of spontaneity i. e. having to do their treatment before going out with friends,
- Causing family stress,
- Physical pain,
- Worrying about CF and it's future effects.

The penultimate question **"Are there any other important things about CF that Sam/Sophie really need to know?"** was interpreted by some participants in terms of what advice they should give to Sam or Sophie about coping with CF, while others used it as an opportunity to identify areas that they would like further information on. The kind of advice given included:

a) Reminders about following the treatment regime *"Just look after herself, do her physio, take her tablets and eat enough food and drink and she will grow up to be healthy"* (participant N, girl aged 11).

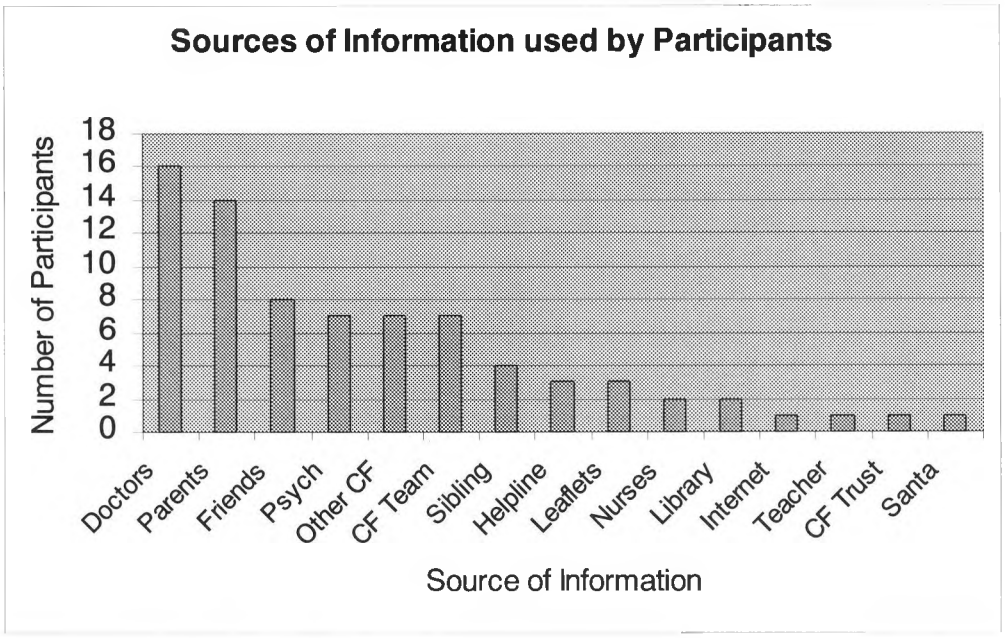
b) Advice about behaviour when older *"You don't smoke when you grow up or you will die"* (participant F, boy aged 6).

c) Recommendations not to dwell on having CF and to try to lead as normal a life as possible *"Don't let it affect you. Don't let it take control of you one way or the other. If you ignore it it still ends up controlling you, but don't stay at home when it is cold. Live your life but realise that it is there. Don't let anyone tell you that you can't do something because you have got it"* (participant V, girl aged 16).

Those who interpreted the question as a way of highlighting areas that they wanted to know more about focused upon what the future holds for them. *"Things about death and that. One thing that really worries you is dying. Knowing the things that can happen when you get really ill, that you won't just suddenly die"* (participant T, girl aged 15). Other issues that they wanted to know more about were how to talk to friends and boyfriends about CF, what to expect and how to cope when older and more ill, getting a job and having children. One participant, the 18-year-old boy, pointed out that the timing of information giving is important *"Things you will want to know all through your life but just at certain times. For example, talking about having a child at 18 to 20 is better than younger. When you are a child you don't ask questions"* (participant W).

The last question of the booklet asked participants **"If Sam/Sophie wanted to talk more about cystic fibrosis who could he/she talk to?"** All bar one four year old (participant A) were able to mention at least one source of information/support whom Sam or Sophie could approach. Four of the participants (B, D, E, and H) only mentioned one source, six others listed two or three sources (C, F, I, J, M, P), while the majority (12 participants) described at least four such sources. There were fifteen sources of support that were mentioned and they are presented on the next page in Figure 11.

Figure 11: Sources of further information mentioned by participants



As Figure 11 illustrates, doctors were the most frequently cited source of support closely followed by parents and then friends and other members of the CF team. Interestingly, the CF Trust which is the main charity supporting people with CF, and which produces large amounts of information and material about CF was only mentioned as a source of support by one participant (and another participant mentioned Santa Claus). It is possible that the Trust is not seen by these participants as providing an interactive service that they could use. Alternatively, the Trust may not be directing its material specifically towards young people, preferring instead to support the families and professionals that care for the children with the disease. It would be interesting to see whether the response would have been more positive if the parents of the participants had been asked whom they could talk to.

These results serve as a reminder to health professionals, especially doctors, within a cystic fibrosis team that children see them as the main source of information about

CF. While the fact that doctors are seen by these participants as the main source of information and support may not appear to be a very surprising result, it is worth remembering that previous studies have assumed that for children with CF their parents are their main source of information, acting as a channel between health professionals and children.

CHAPTER TEN: DISCUSSION

10.1 Children's Knowledge of CF

The children and adolescents in this study do appear to have a basic but impressive understanding about their cystic fibrosis from an early age. As expected, knowledge accrues with age and cognitive maturity. The older children in the study were able to discuss the impact and implications of their illness in greater depth and detail than the younger children. In some areas, such as the impact of CF upon life expectancy, the understanding within this cohort only developed in early adolescence. With just one exception the fourteen participants aged 11 and under (mental age 10 and under) did not display any recognition that life expectancy is influenced by CF. Of the nine participants aged 13 and over all except one (who had a mental age of 11) recognised that CF reduces life expectancy. However in many other areas, such as the nature of the illness or the rationale for treatment, a simple understanding was present amongst several of the very young participants.

For example, the results of this study have shown that twenty-two of the twenty-three participants know that their treatment regime is aimed at keeping them well but does not make their CF go away. In addition, nineteen of the twenty-three participants, including the three youngest participants with chronological and mental ages of 4 and 5, reported that they were born with CF. The four participants (chronological and mental ages of 6 and 7) who did not report that they were born with CF responded to the question 'How did Sam/Sophie get CF?' by saying that they did not know. There was no evidence that any of the twenty-three participants in this study engaged in magical thinking about the causes of cystic fibrosis.

There were eleven participants in this study with chronological ages of 11-18 (mental age range 9-18) and they were all using genetic terminology such as 'gene' and 'carrier' in their explanations of how Sam/Sophie acquired CF. Only two of the twelve participants under the age of 11 used such terminology. However, only one sixteen-year-old girl (mental age 17) was able to provide an accurate figure of the probability that if Sam/Sophie's parents had another child that child would have CF. In other words, despite beginning to use genetic terms in their conversation, the participants in this study were not able to translate this language into an accurate understanding of genetic processes and probabilities.

The understanding of fertility issues amongst the participants of this study was also poor. Not one participant said that Sam/Sophie would not be able to have children despite the evidence that 99% of males with CF are infertile, and that fertility is significantly compromised amongst women with CF (Stark et al., 1995). Two participants aged 15 and 16 (mental ages 13 and 17) referred to difficulties with conception and pregnancy, but still reported that it would be possible for Sophie to have a child of her own. This may be an accurate reflection of the information that the participants have received from health professionals. It must be difficult to say to a teenager that they are likely to be infertile and instead may be easier to emphasise the fact that pregnancy is possible and that progress is occurring in the field of infertility. It could also be a reflection of the publicity that infertility treatment has received in our society. The developments within IVF have received a great deal of attention and although the individual success rates remain low there is a commonly held view that people can now use technology to ensure that they have a baby. Alternatively, these participants may have been given accurate information about their

own fertility but not retained it either because they are not yet interested in fertility or because they are so distressed by the issues that they develop a form of denial. Whatever the underlying reasons, it is a matter of some concern that within this small group of children and adolescents the important issue of fertility seems to be so poorly understood in comparison to other ways in which CF affects their lives.

The participants in this study, with the exception of one six year old and one seven year old, were all able to describe at least briefly the emotional impact that CF has had on both themselves and their parents. The twelve participants aged 9-18 talked in greater depth and acknowledged a wider range of emotions than the eleven participants aged 4-8 who simply described feeling sad or cross about their CF. The ability to talk in greater depth and detail about the feelings generated by CF amongst parents and siblings was noted in participants aged 11 and over.

The cross sectional design of this study, coupled with the small sample size means that it is not possible to make statements about how CF knowledge develops over time within children and adolescents. However, within this cohort of twenty-three children and adolescents aged between 4 and 18, it is interesting to note that there are certain points at which an increase in knowledge appears to be associated with a particular age. For example, the chronological age of 11 (mental age 10) was a dividing point within this sample between participants who could and could not describe how CF is acquired. Similarly the chronological age of 14 (mental age 13) was a point at which all those aged 14 and above knew that CF limits life expectancy but only two of those under the age of 14 did so.

While misconceptions do exist amongst all age groups, for example confusion about where mucus is produced in the body, or how digestive enzymes work, most children rarely gave highly inaccurate or inappropriate answers, responding instead that they did not know an answer. Edwards and Davis (1997) wrote in their book Counselling Children with Chronic Medical Conditions that " If children feel anxious or 'put on the spot', they are more likely to say they do not know rather than risk getting an answer wrong". The younger children in particular in this study did respond that they did not know an answer quite often, and it is possible that they were made to feel uncomfortable by the interview process. On the other hand the feedback that they gave did not indicate that this was the case. Edwards and Davis go on to say "Other reasons for children saying they do not know an answer when in fact they do, include feeling shy, upset, confused by a complex or badly structured question, or being asked something they consider odd or unusual". While none of these reasons should be discounted completely, an alternative explanation could be that the children in this study felt sufficiently comfortable with the researcher and the style of questioning that they were able to admit to not knowing an answer. The fact that they would answer some questions but not others does suggest that they were engaged in the interview, and it is hoped that they were not inhibited from displaying the full range of their knowledge. What is beyond doubt is that there was little evidence of magical thinking or self blame amongst the younger children, and not one child gave any indication that they saw CF as a punishment for their misdeeds.

The results of this study mirror those of previous studies exploring the knowledge that people with cystic fibrosis have of their illness (Nolan et al., 1986; Hames et al., 1991; Conway et al., 1996). Like this study, the previous studies found that older

teenagers and young adults with CF have a relatively good knowledge of the disease and its treatment, but are less informed about genetics and reproductive issues. They also wanted more information about what to expect in the future in terms of their physical health and psychosocial adjustment. This study has indicated that within this sample much of this knowledge is acquired by the time children reach the age of 12 and that it is firmly consolidated by the age of 15. It also suggests that children from an early age are acquiring age appropriate information about CF, which they go on to refine and expand as they develop.

There are some striking similarities in the qualitative data of this study and that of the Bluebond-Langner (1996) study of the well sibling's perspective of CF. She identified a pattern of development of siblings' understanding of cystic fibrosis which she linked to the stage in disease trajectory of the child with CF. Initially the well siblings view CF as a condition one does things for. This view is similar to that of the younger children in this study who understand CF in terms of the treatment involved. The cohort in Bluebond-Langner's study then go on to view CF as a disease with episodes of acute illness and recovery. Similarly the cohort in this study develop an understanding of CF as an inherited illness with multi-organ involvement. Next the siblings in Bluebond-Langner's study gradually become aware that CF is "a chronic, progressive, incurable disease that shortens the life span, but not of one's own sibling, at least in the near future". It is only at the final stages of their sibling's illness that they acknowledge that CF shortens the life span of their own sibling. The adolescents with CF in this study also develop an understanding that CF reduces life expectancy, but as the siblings did they tend to think that it will not affect them or that they will be an exception to the rule.

It is interesting to note that this study has found that age has an impact upon knowledge whereas Bluebond-Langner (1996) did not identify such an effect. She found that the extent of the illness in the child with CF was the key factor in determining the extent of their well siblings' knowledge of CF. She began her study in 1985 when the average life expectancy for a person with CF was 19, and she interviewed siblings of children at all stages of the illness, including some whose sibling had already died. Thus her population had a much greater variation in terms of physical status, and contained many more very seriously ill children, than the population in this study. Had it been possible in this study to interview children at a more serious stage of their disease a similar pattern of results might have developed.

10.2 CF Knowledge and Health Factors

The hypothesis that CF knowledge would be positively correlated with illness experience as measured by various health related factors such as lung function and number of hospitalisations or episodes of IV drug treatments was not supported by the results of this study. The thinking behind the hypothesis was that those participants in poor health would receive more treatment and be in contact with the CF team professionals more often than those in better health. It was thought that they would be both more focussed upon their health and would have greater access to the knowledge held by the CF health professionals. That no association between hospital admissions and CF knowledge was found could lead to a number of potential explanations. It could suggest that:

1. Those children and adolescents in worse health than their peers do not focus more upon their illness and do not seek out further information.

2. CF health professionals do not take the opportunity of an in-patient admission to update the CF related knowledge of the patient.
3. CF health professionals do try to educate their in-patients but the information is not retained by the patient. Certainly a busy ward is not an ideal learning environment and the patient's poor health coupled with the anxiety associated with an admission may contribute to difficulties in processing and retaining information.
4. Or it may be that parents and staff are educating the children and adolescents about CF on an out-patient basis so successfully that an in-patient admission has little impact upon CF knowledge.

These results appear to support the traditional Piagetian views of illness related knowledge in that age and cognitive maturity alone have been found to be positively correlated with CF knowledge. However, as already discussed, the sample used in this study imposes several constraints. The sample is both small and skewed in terms of CF health related variables and demographic variables. There were no participants in this study that fell into the severely affected category despite 6% of the Scottish CF population having lung functions below 40%. The percentage of participants within this study that had not been hospitalised in the last year (74%) was considerably higher than the national figures for the Scottish CF population (55%), and the percentage of the study participants with a BMI below the 5th centile (4%) was lower than the national Scottish figure of 11%. In other words, this sample was considerably more healthy than would be expected from national figures.

Crisp et al. (1996) studied the effects of experience upon children's understanding of illness. They compared groups that had both had either a current or a recent hospitalisation, but where their 'novice group' were in hospital "for an acute illness after relatively uneventful health histories" while their 'expert group' had "experienced prolonged illness and repeated hospitalisation". They used these groups because "illness then is an issue which both groups of children are likely to have given some thought to, a situation that may not be the case when the comparison is between sick and healthy controls". Given that in this study only three of the participants had been hospitalised 'repeatedly' i.e. three times in the previous year, that seventeen of the participants had not been hospitalised once in the previous year and fifteen of them had not been hospitalised since an admission at or shortly after birth, it is hard to argue that this sample could constitute an illness expert group. For the effects of illness experience upon CF knowledge to be fully investigated a comparison would have to be made between a sample of children within the severely affected range and those who are mildly affected by the disease.

10.3 CF Knowledge and Family Factors

The results from the FES when looked at in terms of mean scores for each subscale indicate that the families of the children with cystic fibrosis who were interviewed for this study adapt very well to the pressures that they are faced with. There is no significant difference between their scores and those of the normal family sample presented in the FES manual.

Wilson et al. (1996) proposed a homeostatic model of family adaptation to CF in which they argued that the stresses associated with coping with CF resulted "on

average in stronger families and children with higher self-esteem and lower depression". However, this "contrasted sharply with their response to external stress". They found that when additional stresses such as financial concerns or relationship difficulties were added to a family already coping with a child with CF there was a marked deterioration in family adaptation as well as the child's physical and psychological wellbeing.

While this study did not include any measure of external stresses facing the families of participants, an informal review of the families who were identified as conflict oriented or disorganised did include those known to be experiencing marital difficulties and severe housing problems. Thus it may be that those families that are currently identified as conflict oriented and disorganised may experience more adaptive patterns of family environment at times of reduced stress. The homeostatic model should help clinicians to avoid labelling families as 'non-copers' or 'difficult', instead encouraging them to acknowledge and explore with families the impact of external stresses as well as strategies for coping with them.

There is little scope within this particular study to explore whether the family environment has an impact upon the coping skills of a child with cystic fibrosis, as no measures of coping skills were applied. However, previous studies have found a link between family cohesion and organisation and a child's compliance with CF treatment (McCubbin et al., 1983). The FES manual reports in its section on physical illness that "High support is consistently linked to better youth adaptation, including more self esteem, social competence and sociability; less depression, anxiety, pain,

and physical symptoms; and fewer behavioural problems. More organisation is also associated with these outcomes".

The hypothesis of this study that greater knowledge of CF would be associated with higher levels of family cohesion, expressiveness and organisation and with lower levels of family conflict was not supported by these results. No significant associations were found between knowledge and any family factor measured by the FES. One explanation for this is that children and adolescents with CF may obtain their information about CF from sources outside their family and therefore family factors have little influence upon their knowledge of CF. Alternatively, educating children and adolescents about CF may be such an integral part of CF treatment that it is carried out by parents irrespective of the family environment.

10.4 CF Knowledge and Compliance with Treatment

The hypothesis that greater CF knowledge would be associated with greater levels of compliance with treatment was not supported by the findings from this study. The hypothesis was developed because knowledge has been identified as an important factor in compliance with treatment. Koocher et al. (1990) linked non-adherence to treatment regimes to inadequate knowledge, while Henley and Hill (1990) and Conway et al. (1996) concluded that poor knowledge contributed to unintentional poor compliance with treatment. It is thus a natural step to hypothesise the opposite, namely that higher levels of knowledge would be associated with greater compliance.

However, Koocher et al. (1990) also identified factors such as psychosocial resistance and educated non-compliance that contribute to problems in adherence with treatment regimes. In addition, knowledge alone has long been known not to be the only factor

influencing health related behaviours. The Health Belief Model (Rosenstock, 1966) hypothesises that taking preventative health action depends not on knowledge alone, but upon an individual's perception of their susceptibility to disease, the severity of the disease, the benefits of taking any action, and the obstacles in the way of acting. Thus the finding in this study that there is no association between knowledge and compliance with treatment may indicate that factors such as those described in the Health Belief Model are more powerful than knowledge alone in influencing the adherence of these children to treatment.

It should also be noted that the measure of compliance with treatment, the MCIST, was judged to be unsuitable for children under the age of seven, thus reducing the number of participants eligible for the analysis of the relationship between knowledge and compliance. In addition, the MCIST was administered at the end of the interview at a point when the participants may well have been tired and unable to give the measure their full attention. It may be that the MCIST scores are not as robust as they could be, and that future studies would establish different results.

10.5 Limitations of the Study

10.5.1 The Sample

Twenty three participants is a small sample size that restricts the forms of quantitative analysis that can be used and limits the conclusions that can be drawn from the qualitative data. However, many areas of paediatric psychology research are hampered by small sample sizes as, luckily, the number of children with chronic illnesses such as cystic fibrosis remains quite low. Thus it is not unusual to see studies that have been conducted with less than twenty participants (Noble-Jamieson

et al., 1996). Within the field of CF, sample sizes of 40 (Czajkowski & Koocher, 1987), 29 (Blair et al., 1994), and 23 (Fischer-Fay et al., 1988) occur quite frequently. Those studies cited previously that explore patients' knowledge of CF have used sample sizes of 28, 22, and 18 respectively (Nolan et al., 1986; Henley & Hill, 1990; Hames et al., 1991). Thus while this study's sample size is by no means unusual for studies of CF related knowledge a power analysis was not carried out to determine the appropriate size of the sample for this research project.

An additional matter of concern in relation to the sample is that it may be unrepresentative of the paediatric CF population as a whole. The 23 participants were all recruited from the same CF centre covering the South West Thames region of London and the Home Counties. This is a prosperous region of the country and a much higher proportion of the children come from families in social classes 1 and 2 (48%) than the national average (21%). Additionally, this sample had only 13% of the families representing classes 4 and 5 compared to the national figure of 24%. Also, the gender divide within this sample is not an exact reflection of the national gender divide. The national figures for Scotland show that 51.5% of children with CF are female and 48.5% are male. Whereas the figures for this study are that 61% of participants were female and 39% were male. In addition, the sample was relatively healthy. No participants were classified as having severe CF (lung function below 40%) and none of them required the frequent hospitalisation, oxygen therapy or heart lung transplantation associated with end stage cystic fibrosis. Thus there has been no opportunity to assess whether the booklet is sensitive to the issues facing those children whose illness is at a more severe stage.

An additional issue to be aware of was the nature of recruitment of participants into the study. The researcher was the clinical child psychologist attached to the CF centre, so all the participants knew her and many had worked with her on an individual basis. This may have had an impact upon both the number of patients agreeing to take part in the research and the nature of the sample recruited. The nature of the impact of the researcher's clinical role with the population from which participants were recruited is hard to define. Instinctively it seems possible that families may have felt pressurised to consent to take part in the study despite the assurances of the researcher that their decision would have no impact upon the clinical treatment provided. However it is also possible that families could have felt inhibited in taking part in a study carried out by a member of their clinical team and they might have consented to take part had the study been carried out by someone with whom they had had no previous contact and who would have no ongoing role in their treatment. Whatever the impact of the researcher's clinical role, it does seem that the recruitment process would have been easier to interpret and less open to bias if the researcher had not had a clinical role with this population.

10.5.2 The Booklet

The 'I can help others learn about cystic fibrosis' booklet requires children to answer a series of questions about the impact of CF upon a fictitious child whom they are told is just like them. An assumption has been made that in answering these questions each participant is using their own experience and knowledge of how CF affects their lives and that their answers therefore mirror what they would say if they were asked the question about themselves. Anecdotally this appears to be the case. Many children gave responses in the first person "I don't like the pin", or made it very clear

that they were giving specific examples about their own experiences "I don't know what my parents think, they never open up". However, this was not explored directly and it could be argued that the responses of the participants do not necessarily reflect their knowledge of how CF affects their own lives. This could perhaps have been addressed more directly by including an additional question at the end of the booklet such as "In what way are your experiences the same or different to those of Sam/Sophie?"

Also the premise that there could be a child with cystic fibrosis who is just like the participant but does not know anything about cystic fibrosis is somewhat far fetched, especially when applied to adolescents. Indeed the booklet itself appears to be targeted towards a younger age group. In designing a tool that uses language and illustrations that are likely to appeal to young children, the risk is that the content may be seen by teenagers as rather childish and immature. While the feedback from the teenagers who took part did not indicate that this was their view, the process of obtaining the feedback was not one that was likely to encourage frank opinions. Each participant was simply asked to give his or her feedback directly to the researcher at the end of the interview.

The researcher did consider this during the design phase of the study, and tried to identify other introductions that might be more appropriate for teenagers. For example, they could have been told that the questions were from a recently diagnosed patient. But this might have led to teenagers adjusting their answers for age or assuming that the subject of the booklet had very mild CF if they had only been diagnosed in adolescence. Alternatively they could have been told that the questions

were those of a sibling of someone with CF who had only now become interested in their sibling's condition. However, this began to feel as if it was changing the nature of the booklet, however minimally, and a key aim of the study was to try to design a tool that could be used across the paediatric age range.

The booklet has succeeded in differentiating the answers of teenagers from those of younger children, which would suggest that the adolescent participants were not put off from giving full and detailed answers. However, it would have been useful to ask them to complete another measure, perhaps a questionnaire from a previous study of CF knowledge to see whether the booklet does give them the opportunity to display the full extent of their knowledge.

In addition, while the booklet was designed to encourage participants to talk freely and without constraint about their illness, it could be argued that it does not provide information about their understanding of the practical application of their knowledge. Henley and Hill (1990) and Conway et al. (1996) commented upon discrepancies between the knowledge scores obtained by patients with CF and their application of this knowledge. They gave examples such as patients correctly answering detailed questions about the function of digestive enzymes yet not knowing that they should take the enzymes before snacks. This issue is not addressed in this study. The purpose of this study was to engage in a more exploratory approach to looking at what children with CF know about their illness. However, more practical applications could perhaps become a focus for further work in future.

10.5.3 Scoring the booklet

A formal system for interpreting the data from the booklet in a quantitative manner has not been established. While the booklet could be used immediately as a clinical tool to obtain information from individual children about their knowledge of CF, the scoring system of the booklet would have to be refined before it could be used as a research tool more widely. For example, if a researcher wanted to compare the knowledge scores of children from different hospitals or countries there would need to be an established set of categories into which to place their answers. It would not be sufficient to examine, cluster and rate the answers provided by each group, as occurred in this study, because the groups could have differing numbers of clusters to each answer which would then lead to different knowledge scores. In addition, the higher scores in this study reflect the higher levels of knowledge amongst this particular population, but another centre using the same system to analyse their results could obtain similar knowledge scores and yet their subjects could in fact have a quite different depth of knowledge. In other words, the current system of clustering answers from a particular group allows comparisons of knowledge to be made within that group but does not allow for inter-group comparison as there is no scoring system with any objective measures of accuracy.

For measures of accuracy to be established, further exploratory work would need to be carried out with more children with CF. Researchers would need to examine the clustering of answers to each question and determine which ones covered the greatest number of responses from the widest sample of participants. They would also need to rate them in terms of sophistication. From this they would then have to try to develop a scoring system similar to those of the MCIST or comprehension section of the

WISC (Wechsler Intelligence Scales for Children). These tests establish a spectrum of answers based upon detail and accuracy, and the responses of participants are then given numerical scores according to where they fall within that spectrum.

However, a positive feature of this study is that the qualitative data could be used to contribute to the process of defining both the continuum and the scoring criteria within it. For example, the data looking at how children's understanding about the impact of CF upon life expectancy develops has shown that there is a continuum from no awareness, through an awareness that it can reduce life expectancy but only in certain cases, to a point where a child is aware that it reduces the life expectancy of all those who have it. This information could then be used to help define the scoring continuum to be used in response to the question "How old will Sam/Sophie grow up to be?"

An additional difficulty in scoring the booklet is that it does not take into account each participant's experience of CF. For example, a young child who is very well may respond to the question 'What does Sam have to do each day to keep well' with a list of four elements of treatment, while an older child who is more ill might list six elements. However, the four elements listed by the well child might constitute all his treatment requirements while the child who is more ill could have omitted several important aspects of her treatment, and yet she would be given a higher knowledge rating. Clearly this would not be a matter of concern if the booklet were to be used as a clinical tool in which the clinician would already have information about the child's state of health. However, if it is to be developed into a quantitative measure of CF knowledge this issue will have to be addressed.

CHAPTER ELEVEN: CONCLUSION

The "I can help others learn about cystic fibrosis" booklet has been used successfully to access a small sample of children's knowledge of cystic fibrosis. Participants ranging in age from 4 to 18 used the questions in the booklet to display their knowledge and understanding of their illness, and reported informally that it was an enjoyable process.

This study has shown that, as predicted, within this cohort of children, knowledge of cystic fibrosis develops in terms of detail and sophistication with age and cognitive maturity. The participants over the chronological age of 11 and mental age 9 within this study were able to describe CF in some depth. They were aware of the range of treatments and their purpose, used genetic terminology when talking about CF, recognised that CF reduces life expectancy and acknowledged the emotional impact that CF has upon their whole family. The participants in this study with chronological ages under 11 did not indicate an awareness of the impact of CF upon life expectancy, or use genetic terminology, and they were less able than the older participants to talk about the emotional impact that CF has upon their wider family, particularly their siblings.

Having said that, even the three youngest participants in this study (chronological and mental ages of 4 and 5) could discuss their cystic fibrosis with a greater maturity than traditional 'stage' theorists would have predicted. All the participants in the study

were able to describe at least one element of their treatment, and with the exception of one nine year old boy all the participants knew that their treatment would not make their CF go away. Two thirds of the participants under the chronological age of 11 and mental age of 9, including the two 4 year olds and one 5 year old were able to recognise that they had been born with CF. The third of participants under 11 who did not know that they were born with CF simply answered that they did not know how they had got CF. Not for this group of participants is CF a magical illness sent to them as a punishment for misdeeds.

The older teenagers (participants aged 15 and over) in this study appear to have a knowledge base that is quite similar to that of young adults as identified in previous studies (Nolan et al., 1986; Henley & Hill, 1990; Conway et al., 1996). They have a good understanding of the nature of CF, the way in which they inherited it, the role of treatment, and its impact upon their own life expectancy. This study has shown that they also recognise that CF has a psychological impact upon both themselves and their families. However, as in the previous studies this study has also identified that teenagers have gaps in their knowledge about certain aspects of CF. These include patterns of inheritance, reduced fertility and progression of the illness.

The hypotheses of this study that greater knowledge of CF would be associated with worse health, family factors such as cohesion and expressiveness and increased levels of compliance with treatment were not supported by the results. Knowledge of CF was not found to be associated with any factors other than chronological and mental age. This result does at first sight appear to support the traditional Piagetian approach to making sense of children's understanding of illness in which it is argued that

knowledge accrues with age and cognitive maturity and that children have to go through a series of predetermined stages before they can develop a sophisticated understanding of illness. However, the qualitative details within this study do not fit some of the classic Piagetian predictions. For instance, the Piagetian model would suggest that children at a preoperational and concrete operational stage of cognitive development would view the causes of illness as a matter of magical association or contamination, and yet two thirds of participants in this study with a mental age below nine were able to report that they were born with CF. Similarly, the Piagetian model would predict that children at a preoperational stage of cognitive development would view treatment as being in place to cure an illness and yet all except one participant in this study were able to report that CF treatment does not make CF go away. These qualitative results do perhaps indicate that the experience of having cystic fibrosis does influence a child's understanding of their CF and allows them to develop a more mature understanding of their particular illness than would be expected given their chronological and mental ages.

The fact that no quantitative results indicated that greater experience of illness is associated with greater knowledge of CF could be due to the small and skewed sample of participants that was used in this study. The hypothesis was that those participants with lower lung function, weight and height and higher numbers of hospitalisations would be 'illness/CF experts', receiving more treatment, focussing more upon their CF and having greater exposure to the CF team and their knowledge of the illness, than those in better health. However, the sample used in this study was relatively healthy. No participants fell into the category of severe CF (lung function below 40%) and 74% had had no hospitalisation in the previous year compared to

55% of the Scottish paediatric population. It is possible therefore that there were insufficient numbers of 'illness experts' in this sample or that the measures used to identify the 'illness experts' were not able to differentiate between the 'experts' and 'novices'. Further research with a larger and more representative sample of CF patients should allow this issue to be addressed more conclusively.

The "I can help others learn about cystic fibrosis" booklet could be of use as a clinical tool immediately. It is easy to administer, was appealing to the children and adolescents taking part in the study, and produces information both about participants' areas of expertise and knowledge as well as potential gaps or misconceptions in their understanding of CF. It can be used as a starting point for conversations about a wide variety of issues facing children with CF, such as the rationale behind changes in treatment, reasons for hospitalisation, the psychological responses of family members, and the future impact of the disease. It provides adults with a tool for accessing children's knowledge, but also gives children the opportunity to tell adults what they need more information about. It could be used to track a child's knowledge over time, or to explore patterns of knowledge within different family members. It could potentially play a part in a child's CF annual assessment or in the pre-transplant assessment of children with CF. Its role in enhancing adult-child communication could be considerable, and in so doing it would be likely to increase the involvement of children in the decision making and management of their own illness.

In terms of its clinical use it would not have to be restricted to use by a clinical psychologist. Any member of a CF team could administer it with a child provided they followed a series of specific instructions. These would include:

- Having a specific, clinically based reason for wanting to explore a child's knowledge of CF and discussing this with the child and their family
- Obtaining written consent from the child and his or her parents
- Allowing at least an hour for the interview
- Carrying out the interview in a private space and ensuring that no interruptions took place
- Making it clear to the child that there are no right or wrong answers
- Listening to the child, not interrupting them and not trying to teach them by providing alternative or additional information in the course of the interview
- Providing support to any child who became distressed in the course of the interview and seeking additional input from a mental health professional if necessary
- Giving feedback to children and parents upon request
- Terminating the interview upon request from the child, and ensuring that all children know this before the start.

In practice, the members of the CF team most likely to administer the booklet would be clinical psychologists, social workers and clinical nurse specialists. These team members would be well placed to support children and families should they become distressed as a result of completing the booklet. However, given that the booklet did not appear to cause distress to any of the participants in this study, the booklet should be available to all team members provided they follow the criteria outlined above.

There is no reason to think that this tool could not be applied to all children with cystic fibrosis. But it should be noted that within this study the booklet was not used

with children at a very serious stage of their illness. Ideally further studies should be undertaken with such a population and should include 'consumer satisfaction' measures to determine whether useful information can be obtained without causing unnecessary distress to seriously ill children and their families.

As a quantitative measure further refinements would be required before this booklet could be put into general use. A scoring system would have to be developed that could take into account the age of the child and stage of their illness. Pilot studies involving larger and more representative samples would have to be undertaken, as well as investigations into the reliability and validity of the measure.

Questions remain as to the purpose of developing a quantitative measure of children's knowledge of cystic fibrosis. In clinical terms it is important to establish the knowledge that individuals have about their illness and its treatment, but this does not have to be within a quantitative context. The pressures for children and families coping with CF are already very considerable without the added burden of having some measure of CF knowledge to measure up to. Making comparisons between children's knowledge would seem to add little to their clinical management. Clinicians could benefit from having an outline description of what most children know about CF at what age, and how that knowledge develops with time as this could help them to direct their information giving more appropriately. However, as in this study, such information could probably be most successfully gleaned from qualitative rather than quantitative data.

There may be more persuasive arguments for developing a quantitative measure of CF knowledge for research purposes. Knowledge is seen as an important factor in influencing health related behaviours (Rosenstock, 1966), and informing children about their illness and preparing them for medical procedures has had a central role in paediatrics for many years. Yet it is also known that knowledge alone does not determine health related behaviours such as compliance with treatment (Koocher et al., 1990). The results from this study indicate that knowledge is associated with age and cognitive maturity but not with other factors such as family environment or health and demographic variables. However, the sample used in this study was small and not fully representative of the CF population as a whole. If other studies using more participants from a more representative pool of CF patients were carried out they might be able to identify additional factors that are associated with children's knowledge of CF. This could help clinicians to consider how best to inform children and teenagers about their illness and how to support families in this process. Further research exploring the relationship between CF knowledge, coping skills and individual psychological functioning could help to clarify the impact of knowledge upon the individuals coping with CF and the way in which they put their knowledge to use. This in turn could help us as clinicians to empower the children and teenagers that we work with.

This study, along with previous studies, has highlighted the fact that some of the young people who took part in the study are aware of gaps in their knowledge of cystic fibrosis. It also indicates that they want further information on issues such as the future course of their illness and their ability to start families of their own. The conclusion could be drawn that clinicians are failing in their duty to inform patients

about their illness. It could be argued that they make judgements about what information to give to children and young people and what to withhold, thus failing to inform them appropriately and potentially leaving them feeling anxious and uncertain about their futures. Research exploring the relationship between knowledge, expectations and individual psychological wellbeing could contribute additional important information to the debate clinicians face on a daily basis about how best to balance the need to inform patients appropriately against the desire to avoid causing unnecessary distress.

The number of clinical psychologists working with people with cystic fibrosis has expanded rapidly over the last few years and looks set to continue to do so. The CF Trust has argued that regional units should not be accorded specialist status without ring fenced input from a clinical psychologist. There is an ever increasing awareness of the psychological impact that cystic fibrosis has upon patients and their families. Clinical psychologists and other mental health professionals working within this field are already skilled in talking to patients and their families about the many difficult issues that they have to cope with. And yet addressing issues such as life expectancy, fertility and physical decline remains challenging even for experienced mental health professionals. It is hoped that the 'I can help others learn about cystic fibrosis' booklet may become seen by professionals as a helpful tool with which to structure and initiate such conversations, and that it may be developed into a meaningful research measure that can be used to extend our understanding of cystic fibrosis and the way in which patients understand and cope with it.

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SECTION D: CASE STUDY

A SCHOOL REFUSER WITH CYSTIC FIBROSIS

Confidentiality: Certain details and all names within this case study have been changed in order to preserve anonymity. The family concerned gave written consent to allow this case to be written up and presented in this document.

CHAPTER ONE: REFERRAL AND PRESENTING PROBLEM

No formal written referral was received due to the integrated nature of the multi-disciplinary paediatric cystic fibrosis team. Instead the author, in her role as clinical psychologist within the CF team, was alerted to this case and asked to become involved by the consultant paediatrician upon her return from maternity leave in May 1998. The consultant reported that Kate Baker, a 12 year-old girl with cystic fibrosis had developed extreme difficulties in attending school at the point of transfer from primary to secondary school. She had been absent from school for over two terms and showed no signs of returning. The consultant paediatrician asked the author to see the family and try to help them to establish a programme to reintegrate Kate into school.

Presenting problem

Kate was a 12 year-old girl with cystic fibrosis who had missed almost the whole of her first year at secondary school (Sept 97-May 98). Her CF was extremely well controlled, she had a lung function of over 100% for the entire time that she had been absent from school and she had required no treatment in addition to her daily prophylactic care. Her difficulty in attending school appeared to be anxiety related.

She reported features of panic such as sweating, nausea, and wobbly legs, accompanied by a desire to scream and run away. These symptoms had escalated to a point where each morning Kate would be unable to leave the bathroom due to feelings of sickness and diarrhoea. The symptoms could be triggered by the mention of school, often beginning the evening before she was due to attend school, and only being resolved once she was assured by her mother that she would not have to go to school. Kate had difficulty identifying why she was so anxious about attending school. She said that there had been a teacher who had shouted at the whole class on the first day of term, but he had not singled her out. She was worried that she did not know her way around the new school and she said that some of the older pupils had been very big. But she had moved up with several children from primary school whom she liked and she had not been alone at any point during her first day. Kate did acknowledge that she was worried that if she went to school her house might be bombed and her parents killed. She recognised that this thought was irrational but said that she could not ignore it and it became worse every time she watched the news and saw items about war and bomb attacks throughout the world.

Kate had attended the first full day of term at her secondary school and two further half days later that week. She was in school on only two further occasions for the rest of the term. She did not attend school once during the second term at which point her parents made the decision to move her to a different school. They were unhappy with the way that they had been treated by the school, they felt that they had been seen as incompetent parents unable to control their child, and they also knew of a new school in which Kate's year was the first intake. This meant that there were no children in the years above her and a high teacher-pupil ratio. They hoped that these unique

features would address some of the concerns about school that Kate had identified. However, in the first half of term Kate attended school on only three occasions. The new school convened a multi-professional meeting at the end of the first half of term in order to develop a graded return to school programme for Kate to be implemented in the second half of term. Members of the CF team were asked to attend, and the author, together with the CF nurse specialist did so. This was the author's first point of contact with the Baker family after returning from leave. A number of decisions were made at this meeting.

1. Kate would attend school for one hour a week
2. The special needs coordinator (SENCO) at the school would provide Kate with one to one tuition in this hour and would give her work to do at home
3. The hospital to home tuition service would provide additional support for Kate once her hours at school were increased
4. The author would meet with the family for at least eight sessions to assess and treat Kate's difficulties in attending school
5. A meeting would be arranged to review progress after six to eight weeks, and Kate's attendance would be gradually increased.

CHAPTER TWO: ASSESSMENT

The focus of the assessment was influenced by the literature review that the author carried out. Fowler, Johnson and Atkinson (1985) found that children with chronic illnesses were absent from school more often than their healthy counterparts. Eiser (1993) considered various explanations for this. They included bouts of genuine illness, outpatient hospital appointments, parental fears of infections within school, and inaccurate beliefs about the illness and its consequences. School absence among children with chronic illnesses is not simply related to the seriousness of the condition. It has also been found to be linked to social factors, mothers' level of education, and parents' attitude towards school. Weitzman (1986) commented that many absences are not justified and can be attributed to inaccurate parental perceptions of the vulnerability of their child.

Herbert (1996) commented upon the different terminology used to describe children who do not attend school, but concluded that school refusal is now the term used most frequently to describe those children who do not attend school due to psychological factors such as anxiety, depression, or family dysfunction. These children differ from those who truant from school in that truants are unlikely to be anxious about attending school. Herbert characterised school refusal as:

- Severe difficulty attending school, often resulting in prolonged absence,
- Severe emotional upset including excessive fearfulness, temper outbursts, or complaints of feeling ill when faced with the prospect of going to school,
- Staying at home with the parents knowledge,
- Absence of antisocial behaviours such as stealing, lying and destructiveness.

He added that the prevalence of school refusal is unclear but thought to be low, with peaks at ages 5-6 and 11-12. Children who refuse to go to school show a normal distribution of intelligence and there is no particular association with special educational needs.

King, Ollendick and Tonge (1995) wrote that "the formulation of school refusal is complex, variable and highly individualized". They suggested that clinicians should focus upon the functions served by the absence from school rather than the symptoms themselves. Kearney and Silverman(1990) outlined four possible functions of school refusal:

- Avoidance of specific fears or general over anxiousness in relation to school
- Escape from aversive social situations
- Attention seeking behaviour or separation anxiety
- Rewarding experiences provided out of school.

Thus, in addition to establishing a background history, the areas that the author wished to explore during the assessment included the family attitude towards CF and illness in general, the family attitude towards education, and the nature and function of Kate's school refusal. Four assessment sessions were carried out and despite the whole family being asked to attend each session Mr Baker did not attend any assessment session.

While the author interviewed the family for much of the assessment, she also asked Mrs Baker and Kate to draw up a genogram of their extended family. Barker(1992) recommended genograms as a way of gathering information and an understanding of how a family has come to where it is. He went on to describe genograms as "a useful

adjunct in both assessment and treatment". Mrs Baker and Kate agreed that taking a wider perspective, beyond specific school related issues, might help to develop a deeper understanding of the difficulties faced by Kate and the family. Work therefore began with the construction of a family tree. This was started during a session and completed by the family at home. They reported enjoying the process of constructing the tree and they showed considerable interest in the practical details. But they did not reflect upon the broader picture created by their family tree. Thus it was only following prompts from the psychologist that they began to think about the shape of the genogram and patterns within it. They commented on the number of family members who had died young, the arguments that led to certain members of the family not talking to each other, and the fact that Kate was reportedly the most loved member of the family. No distress was shown at this time and no comments were made about Kate's own life limiting illness.

1) Background history

Family structure: Kate Baker is an only child. She was born in September 1985. She is the daughter of Derek (born 1955) and Louise (born 1958). She was born at full term following a problem free pregnancy. However, she was diagnosed with cystic fibrosis within 24 hours.

No other family members have been diagnosed with CF, however Mrs Baker had an older sister who died of an unknown cause at the age of two, and Mr Baker had two cousins who died during childhood. There is also a history of Downs syndrome within the extended family, as well as premature deaths from meningitis and cancer.

Mr Baker runs his own small catering company. Mrs Baker does not work outside the home, and has not done so since Kate's birth. She does play a role in community activities that Kate is involved in, such as running Girl Guide groups and making costumes for a local children's theatre group.

Medical History: Kate was diagnosed with Cystic Fibrosis shortly after birth. She was kept in hospital for the first five weeks of her life following abdominal surgery. She has not been hospitalised since then.

Like all children with Cystic Fibrosis, Kate has to follow a rigorous treatment regime. She is expected to spend an hour a day on treatment and has to take up to forty tablets a day. However, Kate has been extremely well. She has had no hospitalisations since birth. Her weight and height are above average for girls of her age (between the 50^{th} and 75^{th} centiles) and her lung function has consistently been shown to be over 100% of that expected for girls of her age. This is reflected in her success in physical activities. She has represented her county in cross-country running and has always been top of her class in PE.

Kate has suffered from abdominal discomfort on a number of occasions. This has usually been managed by increasing her use of digestive enzymes and prescribing mild laxatives. The pain and diarrhoea which she has reported periodically and which does not respond to treatment has been rigorously investigated and no organic cause can be found.

School Attendance History: Kate attended the local nursery at the age of four and there were considerable difficulties in separation at this time. However, her transition to Primary school was trouble free. There were no difficulties with attendance for four years. However, problems arose at the start of Year 5 (9 years old) when new staff came into post. However, the family did reintegrate Kate into school without any professional input, by "bribing her with presents each day". Difficulties arose again in Year 6 when two of Kate's classmates circulated a book in which they wrote of their dislike of her. Kate began to resist going to school and also developed some anxiety about leaving home at all, saying she was scared something bad might happen to her mother.

The author became involved at this point. An assessment revealed that Kate had already resumed her friendships with her classmates and was able to provide a list of things she liked about school. However, she said she disliked assemblies and was worried that teachers would tell her off, despite never having been in trouble at school. Mrs Baker was disparaging about school staff in front of Kate and also talked of her concerns about sending Kate to school when she reported feeling unwell. The author discussed with Mrs Baker the impact of her ambivalence upon Kate's school attendance. Kate was taught a simple breathing relaxation technique, encouraged to develop positive coping self statements, and used a reward chart to record her progress in returning to school. Full time school attendance was rapidly achieved and continued until the point of transfer to secondary school.

2) The family attitude towards illness

Kate's early days were traumatic for Mrs Baker. She recalled the diagnosis of Kate's cystic fibrosis with palpable anger. She reported fighting for Kate to be treated and thinks that it was only her fighting spirit that saved Kate's life. She described spending the first five weeks of Kate's life with the baby in isolation. She could not recall the role of her husband during this time and she acknowledges that they coped with their distress separately.

Mrs Baker and Kate denied feeling distressed by Kate's current or future state of health. Mrs Baker talked about progress in the development of a cure for Cystic Fibrosis, and said that as a family they did not worry about health. Kate's response was that she never thought about CF and it never bothered her. While Kate did appear to present as unconcerned about her illness, Mrs Baker's self reports seemed at odds with her behaviour during the assessment, which conveyed high levels of anxiety in relation to Cystic Fibrosis. Attempts were made to explore this discrepancy but they were unsuccessful.

Mrs Baker viewed Kate's school related sickness and diarrhoea as having an organic origin. She claimed that she had been reassured by the hospital investigations that had found no physical obstruction/malfunction within Kate's digestive system. But she then felt that there must be a problem with the digestive enzymes that Kate takes before eating or that Kate had developed a food intolerance or allergy. She did acknowledge that there was a pattern of interaction between Kate's fears of attending school and the onset of symptoms, but she interpreted this as evidence of stress exacerbating a pre-existing physical problem. Kate said that she did not know why

she experienced these symptoms, but she denied sharing her mother's concern that there may be an underlying unidentified physical problem.

3) Family Attitude Towards Education

Kate was acknowledged to be bright, with considerable academic potential. She reported that her ambition was to go to college to do a course in catering and then join her father's business. Her mother talked about the importance of attending school in order to be able to meet this ambition, fulfil her potential, and mix socially. However at the same time she displayed signs of ambivalence towards the formal education system. She talked of other pupils holding Kate back academically, and she frequently requested advice on whether to arrange private tuition for Kate. She also said that she ensured that Kate remained socially integrated with her peers by providing her with opportunities to engage in activities outside school such as horse riding, dance classes and girl guides.

Mrs Baker had unhappy memories of school, having been bullied, and she struggled academically. She remained particularly resentful towards staff whom she felt had failed to support her and she recognised that this had resulted in an antagonistic reaction to many of Kate's teachers. Mr Baker had not been unhappy at school but had not done well and had built his business up without having academic qualifications.

4) Family Relationships

In the course of the assessment it became clear that there was an antagonistic relationship between Mr and Mrs Baker, coupled with an enmeshed/overinvolved relationship between Mrs Baker and Kate. Mrs Baker reported that a special bond

was created between herself and Kate in the very early days of her life. She denied that it was an exclusive bond, saying that Kate did lots of things with her father and friends. She did however describe Kate as her "little shadow" and talked of herself as Kate's best friend. She admitted feeling some anxiety when separated from Kate, explaining that she worried about how others would respond to her if she became ill. But she added that she thought that Kate was more anxious about being separated from her, a claim that Kate immediately denied. Kate often became hostile towards her mother during sessions and described herself as getting on better with her father.

Mrs Baker rarely mentioned her husband. She struggled to describe his individual characteristics, tending instead to comment on their interactions. She said that they "fought like cat and dog" but that they had been together for over twenty years and would never split up. They seemed to have little time together, either because of his workload or because Kate was with them. Mrs Baker agreed that they did not usually discuss parenting issues and that they had different ways of managing Kate's behaviour. Kate confirmed this saying that her mother "goes on and on about things, but always gives in to me in the end", while her father "ignores a lot but means what he says when he gets serious".

5) The nature and function of Kate's school refusal

The nature of Kate's difficulties in attending school closely matched the characterisation of school refusal presented by Herbert (1996). She had experienced a very prolonged absence from school, she displayed severe emotional upset, mostly in the form of somatic symptoms, when faced with the prospect of going to school, she

stayed at home with her parents' knowledge and she did not exhibit anti-social behaviours such as lying, stealing or destructiveness.

Examination of the function of Kate's school refusal indicated that she displayed elements from all four categories of the Kearney and Silverman (1990) functional model.

- Kate displayed specific anxiety in relation to school. She had a history of difficulties in relating to certain members of staff, and was concerned about her ability to perform successfully in certain subjects. She exhibited cognitive distortions in relation to school, in particular thinking that she was the focus for teachers' anger when this was not the case. She also showed symptoms of hyperventilation and panic attacks prior to attending school.
- Kate's school refusal allowed her to escape from or avoid aversive social situations. She had been the victim of some short term bullying during primary school and felt threatened by older children upon her entry into secondary school. While she and her mother did not state directly that she had difficulties in interacting with her peers, it was hypothesised that she was significantly less anxious when interacting with her peers in the presence of her mother.
- Separation anxiety and attention seeking behaviours were a major feature of Kate's school refusal. She reported numerous somatic complaints in addition to those linked to hyperventilation, and despite a somewhat hostile relationship with her mother she also acknowledged that she feared leaving home in case danger befell her parents. Her difficulties in going to school were likely to be exacerbated by the separation anxiety that her mother displayed, and the extra

attention that she received from her father who would stay at home in the mornings in an attempt to entreat her to get ready for school.

- Kate also had numerous rewarding experiences by staying away from school. There appeared to be few restrictions upon her at home during school hours. She was allowed to return to bed for a lie in, watch television, play computer games and accompany her mother throughout the day. In addition, her after school time activities were not restricted by her parents so she was able to continue with her much loved horse riding, cross country running and girl guides.

CHAPTER THREE: INITIAL FORMULATIONS

The author hypothesised that family anxiety about Cystic Fibrosis was holding Kate back from attending school. The diagnosis of CF within 24 hours of Kate's birth appears to have been devastating for Mrs Baker. Her only baby was undergoing major abdominal surgery very shortly after birth, and she recalls this time as one when she felt alone with her baby whom she perceived to be very close to death and whom she had to fight to get treated. The familial roles that were established during these early days seem to have been maintained ever since. Thus Kate is seen as a very fragile, very vulnerable child whose CF presents mainly with digestive rather than chest complications. Mrs Baker developed the role of Kate's protector while Mr Baker was marginalised, having apparently distant relationships with both his wife and daughter. Medical and educational staff are perceived by this family as intrusive authority figures who do not necessarily have Kate's best interests at heart.

That Kate's school attendance difficulties should have become so extreme and entrenched at the point of transfer to secondary school was hypothesised by the author to be linked to the developmental transition point that this event presents all families with. The transfer to secondary school can be seen as an obvious and public beginning of adolescence. Barnhill and Longo (1978) identify nine family transition points that all families with children have to negotiate. Point 4-5 involves "accepting adolescence with the changed roles associated with this, and the parents' need to come to terms with the rapid social and sexual changes occurring in their son or daughter". Kate's approaching adolescence was likely to have presented the family with an apparently threatening developmental stage. For healthy families adolescence is a process that allows the teenager greater independence from her family, an

increased role for her peer group and a preparation for adulthood (Herbert, 1991). Yet for the Baker family it was hypothesised that adolescence signifies "the beginning of the end". Greater independence for Kate could involve taking more responsibility for her own treatment, a process that almost inevitably involves at least a temporary deterioration in health. An increased role for her peer group could expose Kate to threats such as smoking and avoidance of treatment as well as interfering in the "friendship" between mother and daughter. Whilst the preparation for adulthood is full of promise for so many, for those with Cystic Fibrosis is often a preparation for physical deterioration and untimely death. For the Baker family there is perhaps a sense that if only Kate's adolescence could be prevented or at least delayed then it might just be possible to hold back the inevitable decline of CF. Secondary school therefore becomes a frightening symbol for Mr and Mrs Baker, and Kate, being able to sense their concerns, also begins to view school as a dangerous, threatening place to be that is best avoided.

An alternative formulation focuses less upon the role of CF and more upon the function of Kate being at home. Not only was there no strong parental alliance within this family, there were reports of considerable antagonism within the marital relationship. There is a question of whether Kate's role within the family went some way towards controlling this parental antagonism by allowing the parents to focus upon Kate's illness rather than their own relationship. Engfer (1988) identified that "mothers in marital relationships involving a considerable amount of conflict were prone to interpret the relationship with their infants as a substitute source of love and affection as well as being over-anxiously worried about the health and well being of their infants". If Kate was at home during the day then there would be less opportunity for Mr and Mrs Baker to be alone and more opportunity of keeping their

interactions focused around Kate. This role may have been understood by Kate at some level, thus making it hard for her to separate from her parents to attend school for fear of what might happen to their relationship in her absence.

CHAPTER FOUR: INTERVENTION

Treatment Plan

It was decided that the first area to work on would be family factors as it was thought that they could have a major role in undermining any other practical return to school strategies. The plan was to present the formulation to the family, and in particular to highlight the difficult developmental tasks facing them. It was thought that this might allow the family to voice their unspoken fears and in so doing help them to develop a greater understanding of their situation and exert more effective control over it.

Secondly, the graded return to school programme, discussed in the multi-professional meeting was to be developed by school staff with the support of local authority education officials. To support this Kate would be offered individual input to help her to understand and control her symptoms of panic. The cognitive behavioural model of panic outlined by Clark (1986) would form the basis for this work.

At the same time that Kate was being seen Mrs Baker would also be offered individual sessions to support her during the difficult process of encouraging her child to return to school. Mr Baker would be encouraged to attend these sessions in order to strengthen the parental alliance within the family.

Finally a full medical check up was arranged to ensure that the family had accurate and up to date information about Kate's current state of health.

Presenting the formulation

The author presented the CF centred formulation to Mrs Baker and Kate. Mr Baker had not turned up again, despite having been specifically invited by letter. Mrs Baker did not indicate that she found the formulation either illuminating or helpful. On the contrary, she responded angrily, describing feelings of having been intruded upon or abandoned by professionals. She also indicated that she was angry with her husband for not having attended any sessions. In addition, she said that Kate's abdominal pain had been escalating and she was not prepared to consider sending her to school until further medical investigations had been carried out.

The author commented upon Mrs Baker's anger and suggested that the formulation might have triggered considerable distress for her. She denied that she was worried about CF but did admit to feeling distressed that the focus of professionals seemed to be on forcing Kate back to school rather than investigating her medical problems. She also described her sense of fighting a lonely and losing battle.

Mrs Baker's sense of isolation was explored and the author acknowledged that she had not made any attempts to involve Mr Baker in sessions beyond inviting him and asking his wife and daughter to give him verbal feedback. Mrs Baker and Kate were asked to reflect upon why they thought it was so hard for Mr Baker to attend. Mrs Baker focussed upon his pressure of work and difficulty in getting time off work. Kate said that he had told her the sessions sounded silly and he did not think talking would help. Mrs Baker said that she had not heard him say this, but she did think that he might be wary of talking to a psychologist.

It was agreed that if Mr Baker was made more aware of the content of the sessions some of his anxieties might be addressed. The author wrote to him providing detailed feedback about the sessions carried out so far and outlining the future intervention plan. Mr Baker's views were requested and identified as being very significant. The author hoped that this letter might provide both a structure for Mr Baker to attend future sessions as well as some recognition of and reinforcement for the effort that Mrs Baker and Kate had put into the work.

Individual sessions with Kate

Attendance at sessions became much more erratic after the session when the formulation was presented. Thus while Kate was offered seven individual appointments she attended only three. Mr and Mrs Baker were also offered two sessions, both of which they cancelled. Kate's individual sessions began with a discussion about the cognitive-behavioural model of panic (Clark, 1986). Kate recognised that her breathing rate increased when she started to think about school and she worried that she would be sick or get diarrhoea. The second and third sessions involved teaching Kate techniques for managing her hyperventilation, introducing her to muscle relaxation techniques, and beginning to explore the links between her thoughts, feelings and behaviours (Kendall, 1994). Kate struggled to identify her cognitions but did say that she thought that the teachers might shout at her and everyone would think she was stupid. Unfortunately cognitive restructuring techniques were not introduced to Kate owing to her poor attendance at sessions.

CHAPTER FIVE: CONTACT WITH SCHOOL

Contact between professionals broke down during this period. No agency had been nominated to arrange meetings and there appeared to be no sense of collective responsibility. Hostility between the professional groups began to emerge and issues of funding became paramount. The hospital to home tuition service indicated that their offer to support Kate had been dependent upon the service being funded by the school. The school was not able to fund this and began to question whether the input that they were providing was cost effective. As a result of boundary changes no educational welfare officer was assigned to Kate's case. No meetings took place during the rest of the summer term so no extension of the programme took place.

However, while no professional coordination was taking place, Kate attended school for one hour a week as she had been asked to do in the action plan of May 1998. Kate appeared to manage these sessions without any visible anxiety. However, Mrs Baker stayed within the school for this hour and would try to talk to staff. She was also reported by the SENCO to be calling her several times a day. When the support offered by the hospital to home tuition service did not materialise Mrs Baker directed her anger towards the SENCO who in turn responded by becoming hostile towards Mrs Baker.

School ultimatum

The headteacher of Kate's school took a unilateral decision shortly before the start of the autumn term to write to Mr and Mrs Baker. She informed them that school staff

would be unable to provide Kate with any support that term and that they would expect Kate to attend school on a full time basis. The family were distressed by this letter. They felt that Kate had achieved what had been asked of her, but had received little recognition for this and was now being let down by staff who were failing to deliver the promises of a gradual return to school.

Crisis point

The relationship between the Bakers and the school broke down completely on the first day of the 1998 autumn term. Kate arrived on time and entered her classroom. However, on the way to assembly she passed her mother who was still in the building. Kate was reported to have become "hysterical" when asked to say goodbye to her. Mrs Baker also became distressed and was reported by staff to have slapped Kate on the face. Mrs Baker said that she did hit Kate but that she was trying to stop Kate from holding her breath and fainting. This incident took place in front of many pupils. The headteacher asked Mrs Baker and Kate to leave the premises and followed this with a letter suggesting that the family look for alternative places to educate Kate.

Psychology session

The breakdown in relationship between Mrs Baker and the school seemed to act as a trigger for Mr Baker to become involved. Mrs Baker requested a session with the author which was attended by Mr and Mrs Baker but not Kate. Having vented their anger they then began to describe concerns about Kate beyond her education. They described her as becoming "a bit of a recluse" saying that she had not made friends during a summer girl guides camp. They also reported that she was beginning to display anxiety about going outside and would no longer accompany her mother on

shopping trips or social visits. In addition Mr Baker mentioned that strain was being put on their marriage. The psychologist said that the issues they were coping with had become more extreme and that their daughter's mental health was now a matter of great concern. She made a request to consult with a Consultant Child and Adolescent Psychiatrist, to which they gave their permission.

CHAPTER SIX: PSYCHIATRIC CONSULTATION

A meeting was arranged between the psychologist, the consultant paediatric chest physician and the consultant child and adolescent psychiatrist from the local children's inpatient unit. A brief outline of the family history, Kate's medical history and contact with child psychology services was presented to the psychiatrist. Themes to have emerged from the assessment sessions were listed and the current sense of crisis and impasse were described. The psychiatrist was asked for his opinion on the long-term outcome for this girl and her family, advice on what intervention to try next and whether a period of inpatient assessment and treatment would be appropriate.

The psychiatrist commented in particular upon the sense of "stuckness" that could be felt within the family, the school and the health systems involved in this case. He suggested that acknowledging this and the way in which Kate's development was being arrested as a result was the new starting point for resuming work with the family. He reflected that the fear of change within the family was so great that it would lead family members to unconsciously resist all attempts to help. He added that "the more we encourage Kate to get better the stronger becomes the impulse to resist". Instead he recommended acknowledging the pessimism felt by the family and others, and discussing their realistic concerns that without rapid change Kate would be unlikely to resume full time education and would almost certainly experience psychological disability as an adult.

The psychiatrist also recognised that the graded return to school programme had collapsed as much as a result of professional issues as from factors within the family. He therefore felt that if the family could be encouraged to develop the strength and

motivation to overcome their resistance to change, an intensive and carefully monitored graded return to school programme could still be effective. However, he also said that an admission to an adolescent inpatient unit could be considered if no progress occurred.

Theoretical background

A further discussion was held between the psychologist and psychiatrist to discuss the theoretical underpinnings of the psychiatric advice. Follow up studies of school refusal have indicated that at least one third of clinical cases continue to have severe emotional or social difficulties into adulthood (Elliott, 1999). Kearney and Tillotson (1998) have found that where school refusal persists for two academic years, susceptibility to treatment is limited.

The use of paradox and the declaration of therapeutic impotence are devices commonly used within strategic family therapy, often at a time when more direct suggestions have failed to produce change within a family. These strategies hand responsibility for the symptoms over to the family and are particularly effective in cases where the family have become confrontational towards the therapist. The use of paradox can involve a positive reframing of symptoms, advice to continue or extend the patterns of behaviour, and a prediction of relapse. Announcing therapeutic impotence is another form of paradox that can be used at a time of escalating tension between the clients and the therapists. It is "intended to put an end to the battle, and also avoids the therapists appearing as the initiators of change"(Barker, 1992). The paradoxical effect is that the family is challenged to prove that the apparent hopelessness of their case is wrong. Similarly within motivational enhancement

therapy (MET) Miller and Sanchez (1994), have identified "active ingredients" required to produce internally motivated change within individuals. These have included feedback of personal risk or impairment, emphasis upon personal responsibility for change and acknowledgement of where the client currently is within the cycle of change.

CHAPTER SEVEN: SERVICE RESPONSES TO THE CRISIS

Paediatrics

Following the meeting with the psychiatrist, the Baker family attended the next paediatric clinic. During this meeting the paediatrician and psychologist used the therapeutic strategies described above to present the following thoughts to the family:

- Kate's symptoms were extending beyond school refusal to more widespread social and psychiatric difficulties
- The likelihood that these difficulties would continue into Kate's adult life was high and would be greatly increased if Kate remained out of school for two years (she had already missed more than one whole academic year)
- All attempts by professionals to reintegrate Kate into school had failed
- The psychologist's work to develop a greater understanding of the situation faced by the family had not been helpful for them
- Medical investigations had failed to find an adequate explanation for Kate's abdominal pain
- The professionals involved felt extremely sad that they had been unable to help get Kate back to school and they could think of no further strategies to improve the situation.

The family sat silently throughout the presentation and were clearly shocked by its content. Their response was to acknowledge that they were aware that the situation was serious and that it seemed that no one could help them. The paediatrician made a follow up appointment for two weeks later, and finished the consultation.

Education

The Headteacher's threat to exclude Kate from school alerted the Educational Welfare Service to the serious nature of the breakdown in relationship between the school and the family. The senior educational welfare officer (EWO) for the school district assigned himself and a colleague to the case. A multi professional meeting was arranged for the week following the paediatric consultation.

At this meeting the Headteacher said that while they were not initiating formal exclusion procedures, they were not in a position to spend time individually with Kate and therefore wondered whether she was appropriately placed. She acknowledged that there had been a breakdown in the relationship between school staff and Mrs Baker. She said she understood the distress involved for the family, but it had been inappropriate for Mrs Baker to stay in school, and her telephone calls to staff had been time consuming and stressful.

The psychologist outlined the comments made to the family within the paediatric consultation of the previous week and said that the role for psychology at present was one of support for the family rather than providing active interventions.

The Educational Welfare Officers said that they were concerned by the idea of Kate being educated elsewhere and that they felt that she had not had the chance to follow a tight, carefully monitored graded return to school programme.

Mr and Mrs Baker agreed with this. They then said that they had been discussing the next steps for them to take as a family and that Kate herself had volunteered that she

would like to start attending school one morning a week and build up her attendance from there.

The professionals agreed this was a positive step, Kate joined the meeting, her timetable was reviewed and she chose the morning that she would like to start with. The EWO said that he would be happy to accompany Kate from home to school for the first two occasions and to meet her in school after that. The psychologist offered to be the professional that Mrs Baker could telephone to discuss any concerns, and to take on a liaison role between school and home while the programme was being established. It was also agreed that meetings should continue on a fortnightly basis.

CHAPTER EIGHT: OUTCOME AND FOLLOW UP

Six subsequent multi-professional meetings took place over the course of two terms. On each occasion Kate had met the goals set at the previous meeting and attended with clear plans about how she wished to extend the programme. She took the opportunity to report concerns that she had such as the seating arrangements in one of her lessons and her difficulties in catching up with languages that she had missed. The school staff responded rapidly to her concerns. The EWO summarised each meeting in a letter sent to all participants and he formally congratulated Kate on her progress. The psychologist talked with Mrs Baker on several occasions. The discussion was usually about how best to reinforce Kate's achievements although the psychologist also reflected upon the great changes that Mrs Baker had made.

Progress was inevitably interrupted by school holidays and also by two episodes when Kate was ill. However, these issues had been discussed prior to their occurrence, a mini relapse plan was implemented in which Kate reverted to the previous level for one week before building up again, and there was no disruption to the programme. After 15 school weeks Kate returned to full time school. Kate was awarded two prizes at the end of year prize giving, one for the greatest progress over the year, and one for achieving the highest test scores in mathematics of any girl in her year.

Kate has remained in full time attendance at school for three years. She has made many friends, is taking part in both the academic and sporting life of the school and says that she feels well and happy. Her parents report that she is much more settled at home and she is now confident in her ability to socialise without her mother being

present. Mrs Baker has resumed some activities that she had let lapse while Kate was at home, including visiting residents of a local nursing home and seeing her own friends. Mr and Mrs Baker report that they are getting on well together and they have spent a weekend away.

CHAPTER NINE: REFORMULATION

The nature of the events after the assessment add weight to the secondary rather than the primary formulation originally proposed by the author. The primary formulation argued that Kate's move to secondary school was a developmental transition point that the Baker family were unable to cope with. Underlying the family paralysis was the fear of Kate's CF deteriorating. If the family could hold Kate back from secondary school and the adolescence associated with it then the parents could maintain control over Kate's treatment, prevent the peer group from exerting their influence over Kate and protect her from harm. However, this formulation did not appear to ring true for the family. While Mrs Baker's angry response to the formulation would suggest that it was meaningful to her at some level, her comments highlighted her sense of isolation and the rift between her and her husband. Attempts to involve Mr Baker in the work were increased at this point. In addition, the family's contact with the CF team did not alter after the formulation had been presented to them. They continued to contact the team about Kate's abdominal symptoms but they did not display more specifically CF related concerns or initiate discussions about the long term implications of Kate's CF. Finally, the paradoxical strategy of admitting therapeutic failure, although presented by the CF consultant, did not appear to have any impact upon the family's relationship with the cystic fibrosis team. It was as if they simply made no links whatsoever between Kate's CF and her school refusal.

Instead, the deterioration in the relationship between the school and Mrs Baker, perhaps coupled with the paradoxical strategies, do seem to have triggered Mr Baker's involvement in the case. Mr Baker only attended sessions once he and his

wife had been asked to find an alternative place to educate Kate. The reasons for this were never explored, but the author hypothesised that this external threat of exclusion, threatened the stability of the pre-existing family relationships and finally forced Mr and Mrs Baker to come together as Kate's parents to get the family through this threatening time. The use of the paradoxical techniques did have the desired effect of placing the responsibility for change within the family, but the key element was that this became a joint parental activity. The author hypothesises that Kate instinctively and rapidly sensed that the relationship between her parents had undergone a fundamental change. In her parents moving from a generally distant and antagonistic relationship towards one that was more collaborative and supportive, Kate no longer needed to be at home in order to protect their marriage. Secure in her sense that things would be OK at home Kate was then able to return to school and needed only a few face-saving strategies to support her reintegration.

CHAPTER TEN: DISCUSSION

This case appeared to be developing into one of the one third of cases of school refusal that fail to respond to treatment and lead to severe social and psychiatric disabilities in the child that extend into adulthood. Kate had missed over a year of school, there was a breakdown in the relationship between the family and the school, inter agency hostility was rife, and the family were withdrawing from the help that was being offered to them. Numerous attempts at reintegration to school had been made and none had been effective in the long term. Admission to a psychiatric in patient unit was being seriously considered.

However, the use of the strategic manoeuvre of declaring therapeutic failure and impotence, together with the threat of Kate being excluded from school, reversed the pattern of behaviour within this case almost overnight. Palazzoli, Boscolo, Cecchin & Prata (1978) caution that the timing of such an intervention is crucial and should not be done too soon. They write that the time is right when "the angry obstinacy of the therapists together with the family's reinforcing of its disqualifications of the therapists, indicate escalation of the symmetrical battle". There is little doubt that this point had been reached in this case. Battles were escalating within and between all the systems involved, i. e. family, health and education. All the usual support structures for families coping with school refusal looked at one point as if they were going to collapse. The consultation with a psychiatrist experienced in systemic family therapy helped to identify this and provided ideas about how to break these destructive and repetitive patterns of behaviour. The use of paradox and the

declaration of therapeutic impotence were new and interesting ways of approaching the issue of motivation and responsibility for change with this family.

But what was not discussed in much detail in the course of the psychiatric consultation was the potential risk associated with this technique i.e. that the therapeutic relationship could have broken down completely. Within a mental health setting this could have serious consequences in terms of accessing services in future. But the team working with the Baker family was a paediatric one and the consequences of a breakdown in relationship could have been devastating. The CF team provides a tertiary, specialist service covering the entire South West Thames region. Had the Baker family associated the therapeutic impotence with the CF related care that the team provide as opposed to just the input around Kate's school refusal they would have lost all confidence in the medical input that Kate receives. This could have had a major impact upon areas such as compliance with treatment or clinic attendance, or could have resulted in the family having to travel out of region to receive their CF care.

The risks would have been reduced had the psychiatrist suggested using other approaches such as motivational enhancement therapy before embarking upon the risky strategy of paradoxical input. In addition, an alternative response that might have broken the sense of "stuckness" without threatening the relationship between the CF team and the Baker family would have been to transfer the management of Kate's school refusal to a local, non-paediatric child and adolescent mental health team. Whether the clinical psychologist from the cystic fibrosis team was the most appropriate member of the child psychology department to carry out the work with

this family in the first place is debatable. While a basic knowledge of the symptoms of cystic fibrosis and its impact upon families was essential, having a role within the paediatric team may have contributed to some of the difficulties the family had in addressing psychological rather than physical health issues. The ongoing contact that is a feature of the relationship between the psychologist and families within the cystic fibrosis service may have inhibited both parties in openly acknowledging underlying tensions that developed within the therapeutic relationship. In addition, the CF psychologist may have focused too much upon CF as a critical feature of the formulation, whereas with hindsight it appears to have had a much less central role than other family factors that other professionals less involved with CF might have identified at an earlier stage.

School refusal is a very public form of psychological distress. The legal requirements upon parents to educate their children ensure that when this does not happen numerous agencies become involved and the family can feel as if it is under both scrutiny and threat. In the case of the Baker family the normal parental task of keeping their child well, by virtue of Kate's cystic fibrosis, was already a more public and a more difficult task than for many other families. As a result they were likely to have a strong sense of ambivalence towards professional intervention in any area of their family life. At one level they must have recognised the inevitability of needing help from outside agencies, especially health professionals whilst at another level resenting the intrusion and implication of failure that professional input could imply. Had these issues been addressed with the family at the start of the intervention, and also discussed within the multi-professional meetings it is possible that some of the tensions and antagonism within the case might have been avoided. It might also have

resulted in a more open discussion about who held the responsibility for change thus possibly averting the need to use the risky strategy of declaring therapeutic impotence.

The role of inter-agency co-operation and co-ordination was another key factor in this case. While the declaration of therapeutic impotence was clearly a turning point for the family in terms of accepting responsibility for change and paradoxically increasing their motivation to change, it occurred at the same time that inter agency co-ordination was resumed. It would have been difficult for Kate to be reintegrated into school if the various services involved had not addressed their failure to co-operate in the past and determined to resume contact with each other. Through frequent, regular and planned meetings, the professionals involved were able to provide the family with input that was seen as supportive rather than intrusive and which was also consistent and co-ordinated. Co-operation between professionals also reduced the opportunity for any dysfunctional patterns of communication within the family to be reflected in professional interactions, and allowed effective communication strategies to be modelled within meetings.

Ultimately change occurred because the parents came together, strengthened their parental alliance and accepted responsibility for change. In so doing they developed the skills to challenge and reverse long standing patterns of behaviour, and allowed Kate the opportunity to start to look outwards. As Berg noted in a 1992 review of absence from school and mental health "it is only when the child realises that parents are determined to effect a return to school that real progress tends to be made".

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APPENDICES

APPENDIX ONE

EXPLORING CHILDREN AND YOUNG PEOPLE'S UNDERSTANDING OF CYSTIC FIBROSIS: A STUDY.

INFORMATION SHEET FOR PARENTS.

The main aim of this study is to find out what children and young people with cystic fibrosis know about their condition, and how this knowledge develops. The study will also try to see if there is a link between what children and young people know about cystic fibrosis and their compliance with treatment. Finally, the study will try to identify if there are any family factors that influence children and young people's knowledge of cystic fibrosis.

It is hoped that the study will increase our knowledge of how children and young people understand and cope with cystic fibrosis. If health professionals and parents have a way of getting more information about what children and young people know about their condition they can use this to help them to answer children's questions and communicate more effectively. It would allow adults to avoid giving children and young people information that they already have, while at the same time it would identify gaps in children's knowledge which could then be talked about. Not only might this improve communication between adults and young people with cystic fibrosis, it might also improve the child or young person's compliance with treatment and feelings about themselves.

I would see your child during a routine clinic visit. The total time involved would be approximately 45 minutes. I would begin by giving them a brief picture vocabulary test to get an idea of their verbal skills and to help them to relax at the start of the session. Next I would give them the "I Can Help Others Learn About Cystic Fibrosis" booklet. In this booklet your child is asked to see if he or she can help answer the questions of a fictional child with cystic fibrosis. There are twenty-two questions covering areas such as what cystic fibrosis is, how it can be treated, growing up with it, the feelings involved, and the genetics of the condition. The booklet is illustrated and designed to be fun for the child to complete. Finally I would give your child a test of their compliance with treatment in which they are told five very short but unfinished stories about medical situations where they are asked to make up the endings. I would write down your child's answers to all the questions asked during the session, but would also tape record the session to ensure that I do not miss any important information. While I was seeing your child, I would ask one parent to fill in a questionnaire about your family that would take approximately 20 minutes.

If you would like to look at the booklets etc before your child answers them, you are welcome to do so. You are also welcome to accompany your child during the study, or ask someone else to go in with them, if you think your child would feel more comfortable with you or another adult in the room with them. Your child will be told at the start of the session that they can ask to stop at any time.

The tape recording of the session will be destroyed once the information has been collected. All information obtained is confidential. However, if you would like to discuss your child's results, or if your child wants to talk about the session, a follow

up appointment could be arranged. Once the study is completed all families that took part in it will receive written feedback covering the general results and conclusions.

There is no obligation to take part in this study. Although the session would probably take place during a clinic visit in order to minimise the disruption to you and your family, it is not a routine part of clinic. If you would prefer me to visit you at home, this can easily be arranged. If you do not want your child to take part in this study, it will not affect the care that he or she receives from any member of the cystic fibrosis team. You are also able to withdraw your consent to your child taking part in this study at any time, without explanation, and without any impact upon the care that they receive. Your child will also be told that they can stop at any time and I would stop the session if I felt that any child was becoming tired or upset.

Please feel free to discuss this study with me at any time. I would be happy to try to answer any questions that you may have. You can call me on either 0181 296 4455 (The Child and Adolescent Psychology Department) or 0181 296 3067(Barbara, Dr Rosenthal's secretary). If I am not available when you call, please leave a message and I will try to get back to you as soon as possible. Thank you for taking the time to read this information sheet.

Frances Goodhart, BA (Hons), MSc, C.Psychol
Chartered Clinical Psychologist.

City University
Department of Psychology

EXPLORING CHILDREN AND YOUNG PEOPLE'S UNDERSTANDING OF CYSTIC FIBROSIS

Informed Consent Signature Sheet (parent)

I confirm that I have read and understood the description of the investigation given in the attached parent's information sheet and I give my consent to my child taking part in this study.

I understand that all information collected about my child and family will be kept strictly confidential and will not be disclosed to any third party with any identifying information without my further written consent. I also understand that I may withdraw my child from the study at any time without giving a reason and without incurring any penalty.

CHILD'S NAME:

CHILD'S DATE OF BIRTH:

YOUR NAME:

YOUR SIGNATURE:

DATE:

EXPLORING CHILDREN AND YOUNG PEOPLE'S UNDERSTANDING OF CYSTIC FIBROSIS: A STUDY.

PARTICIPANT'S INFORMATION SHEET.

The main aim of this study is to find out what children and young people with cystic fibrosis know about their condition and how this knowledge develops. The study will also try to see if there is a link between what children and young people know about cystic fibrosis and their compliance with treatment. Finally the study will try to see whether there are any family factors that might influence children and young people's knowledge of cystic fibrosis. It is hoped that this study will help to improve communication between adults and children and young people with cystic fibrosis.

You would have one session with me during one of your routine clinic visits, or I can see you at home if you would prefer this. You can ask for someone to come in with you if you would like an adult to be with you during our meeting. The session should last approximately 45 minutes. During this time I will begin by asking you to match words to pictures. Then I will ask you some questions about a child with cystic fibrosis. Finally I will tell you some unfinished stories about medical situations and ask you to complete them. I will write down your answers to all the questions, but I will also tape record the session so that I can check that I do not miss anything important that you say. I will ask one of your parents to fill in a questionnaire about your family while they are waiting for you.

The information that you and your parents give me will be confidential. The tape recording of our session will be destroyed as soon as I have checked it to see that I wrote down your answers completely. Your parents will have a chance to look at the questions before I give them to you so they might want to talk about the session with you once it has ended. If you or your parents would like a follow up appointment to discuss the results of our meeting, this can be arranged.

You do not have to take part in this study. Although it is taking place during clinic time it is not a part of clinic. If you decide that you do not want to be in the study it will not affect the treatment that you receive from any member of the cystic fibrosis team. If you do agree to take part, you can refuse to answer questions, and you can stop the session and withdraw from the study at any time. Your parents are also able to withdraw you from the study if they have any concerns (and if you are under 16).

I hope that you will think about taking part in this study. Please feel free to ask me any questions you may have about it. If you want to contact me, you can call me at the Department of Child and Adolescent Psychology on 0181 296 4455 or via Barbara,Dr Rosenthal's secretary on 0181 296 3067. If I am not available you can leave a message for me and I will try to get back to you as soon as possible. Thank you for taking the time to read this information sheet.

Frances Goodhart
Chartered Clinical Psychologist

City University
Department of Psychology

EXPLORING CHILDREN AND YOUNG PEOPLE'S UNDERSTANDING OF CYSTIC FIBROSIS.

Informed Consent Signature Sheet (participant)

I confirm that I have read and understood the description of the investigation given in the attached participant's information sheet. I give my consent to take part in the study.

I understand that all individual information collected about me will be kept confidential and will not be passed to third parties with any identifying information without my further consent in writing. I also understand that I may withdraw from the study at any time without giving a reason and without incurring any penalty.

YOUR NAME:

YOUR DATE OF BIRTH:

YOUR SIGNATURE:

DATE:

68. In our family each person has different ideas about what is right and wrong.
69. Each person's duties are clearly defined in our family.
70. We can do whatever we want to in our family.
71. We really get along well with each other.
72. We are usually careful about what we say to each other.
73. Family members often try to one-up or out-do each other.
74. It's hard to be by yourself without hurting someone's feelings in our household.
75. "Work before play" is the rule in our family.
76. Watching T.V. is more important than reading in our family.
77. Family members go out a lot.
78. The Bible is a very important book in our home.
79. Money is not handled very carefully in our family.
80. Rules are pretty inflexible in our household.
81. There is plenty of time and attention for everyone in our family.
82. There are a lot of spontaneous discussions in our family.
83. In our family, we believe you don't ever get anywhere by raising your voice.
84. We are not really encouraged to speak up for ourselves in our family.
85. Family members are often compared with others as to how well they are doing at work or school.
86. Family members really like music, art and literature.
87. Our main form of entertainment is watching T.V. or listening to the radio.
88. Family members believe that if you sin you will be punished.
89. Dishes are usually done immediately after eating.
90. You can't get away with much in our family.

FAMILY ENVIRONMENT SCALE

FORM R

Rudolf H. Moos

Instructions

There are 90 statements in this booklet. They are statements about families. You are to decide which of these statements are true of your family and which are false. Make all your marks on the separate answer sheet. If you think the statement is *True* or mostly *True* of your family, make an X in the box labeled T (true). If you think the statement is *False* or mostly *False* of your family, make an X in the box labeled F (false).

You may feel that some of the statements are true for some family members and false for others. Mark T if the statement is *true* for most members. Mark F if the statement is *false* for most members. If the members are evenly divided, decide what is the stronger overall impression and answer accordingly.

Remember, we would like to know what your family seems like to *you*. So *do not* try to figure out how other members see your family, but *do* give us your general impression of your family for each statement.

Consulting Psychologists Press, Inc.
3803 E. Bayshore Road, Palo Alto, CA 94303

1. Family members really help and support one another.
2. Family members often keep their feelings to themselves.
3. We fight a lot in our family.
4. We don't do things on our own very often in our family.
5. We feel it is important to be the best at whatever you do.
6. We often talk about political and social problems.
7. We spend most weekends and evenings at home.
8. Family members attend church, synagogue, or Sunday School fairly often.
9. Activities in our family are pretty carefully planned.
10. Family members are rarely ordered around.
11. We often seem to be killing time at home.
12. We say anything we want to around home.
13. Family members rarely become openly angry.
14. In our family, we are strongly encouraged to be independent.
15. Getting ahead in life is very important in our family.
16. We rarely go to lectures, plays or concerts.
17. Friends often come over for dinner or to visit.
18. We don't say prayers in our family.
19. We are generally very neat and orderly.
20. There are very few rules to follow in our family.
21. We put a lot of energy into what we do at home.
22. It's hard to "blow off steam" at home without upsetting somebody.
23. Family members sometimes get so angry they throw things.
24. We think things out for ourselves in our family.
25. How much money a person makes is not very important to us.
26. Learning about new and different things is very important in our family.
27. Noboby in our family is active in sports, Little League, bowling, etc.
28. We often talk about the religious meaning of Christmas, Passover, or other holidays.
29. It's often hard to find things when you need them in our household.
30. There is one family member who makes most of the decisions.
31. There is a feeling of togetherness in our family.
32. We tell each other about our personal problems.
33. Family members hardly ever lose their tempers.
34. We come and go as we want to in our family.
35. We believe in competition and "may the best man win."
36. We are not that interested in cultural activities.
37. We often go to movies, sports events, camping, etc.
38. We don't believe in heaven or hell.
39. Being on time is very important in our family.
40. There are set ways of doing things at home.
41. We rarely volunteer when something has to be done at home.
42. If we feel like doing something on the spur of the moment we often just pick up and go.
43. Family members often criticize each other.
44. There is very little privacy in our family.
45. We always strive to do things just a little better the next time.
46. We rarely have intellectual discussions.
47. Everyone in our family has a hobby or two.
48. Family members have strict ideas about what is right and wrong.
49. People change their minds often in our family.
50. There is a strong emphasis on following rules in our family.
51. Family members really back each other up.
52. Someone usually gets upset if you complain in our family.
53. Family members sometimes hit each other.
54. Family members almost always rely on themselves when a problem comes up.
55. Family members rarely worry about job promotions, school grades, etc.
56. Someone in our family plays a musical instrument.
57. Family members are not very involved in recreational activities outside work or school.
58. We believe there are some things you just have to take on faith.
59. Family members make sure their rooms are neat.
60. Everyone has an equal say in family decisions.
61. There is very little group spirit in our family.
62. Money and paying bills is openly talked about in our family.
63. If there's a disagreement in our family, we try hard to smooth things over and keep the peace.
64. Family members strongly encourage each other to stand up for their rights.
65. In our family, we don't try that hard to succeed.
66. Family members often go to the library.
67. Family members sometimes attend courses or take lessons for some hobby or interest (outside of school).

APPENDIX THREE

MCIST INTRODUCTION

I am going to tell you a number of stories about people like you who are faced with making a decision about their health. Please listen carefully to each story and make up an ending for each one. Your story endings should tell what each person decides to do and what eventually happens to that person. There are no right or wrong answers. Simply make up an ending that seems sensible to you. Be sure to tell me what the person decides to do and what happens.

MCIST STORY 1

Bill went to the doctor for a checkup and was surprised when the doctor said "you haven't had a booster injection in quite some time. I think I should give you one today". What do you think happened next?

MCIST STORY 2

Helen had been ill for two days. She had a headache a tummy ache and felt as though she might have to be sick. Her mother took her to the doctor. What do you think happened next?

MCIST STORY 3

The doctor came into Mike's hospital room and told him that he needed to have a serious operation. His right foot was diseased and it would have to be taken off or else he might die. Mike knew that his foot was sore, but had not realized just how serious it was. What do you think happened next?

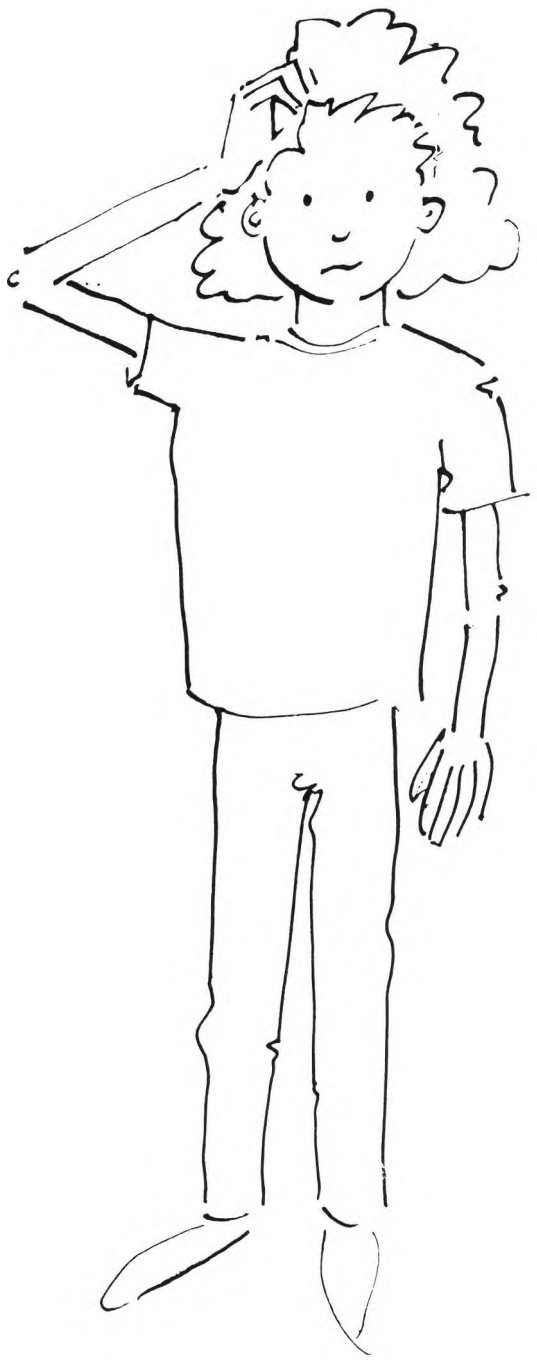
MCIST STORY 4

Jill had been ill for a long time, almost two years. Sometimes she felt better for a couple of months, and then she felt ill again for a time. Her doctor called her up one morning to tell her about a new treatment. It was so new that they did not know whether it would work for her or not. Sometimes it seemed to help some people with the same problem as Jill, but at other times it did not help or even seemed to make things worse. What do you think happened next?

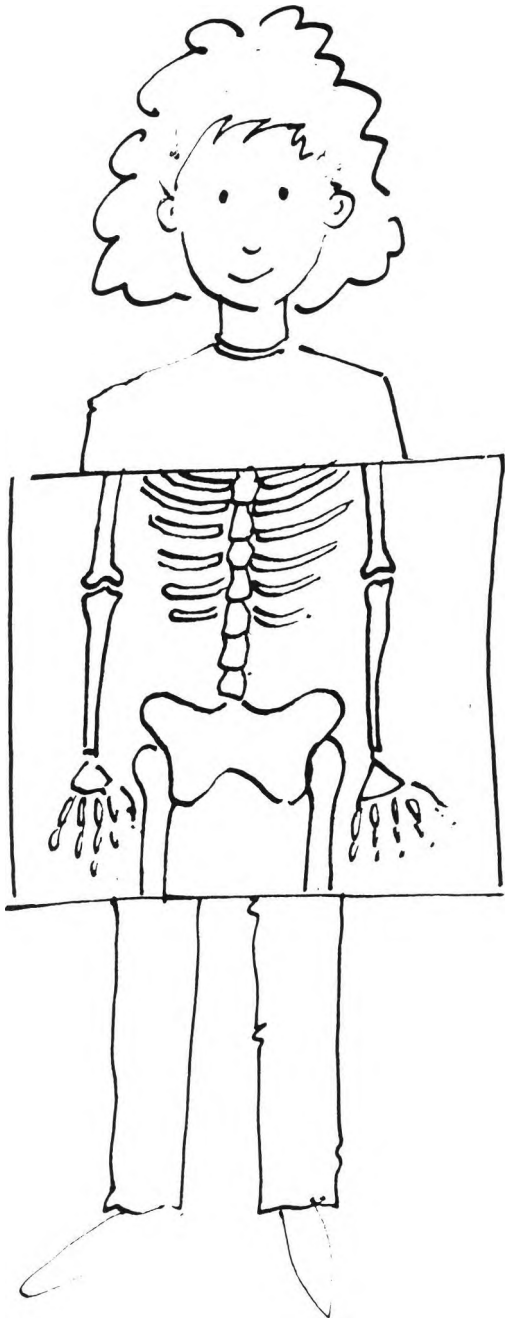
MCIST STORY 5

Henry was born with a disease that he will have for his whole life. A lot of the time he feels perfectly well and not ill at all. Sometimes he even forgets he has a medical problem. The doctors told him that he would have to do special exercises and take ten pills every day to try and stay well. What do you think happened next?

I CAN HELP OTHER PEOPLE LEARN ABOUT CYSTIC FIBROSIS

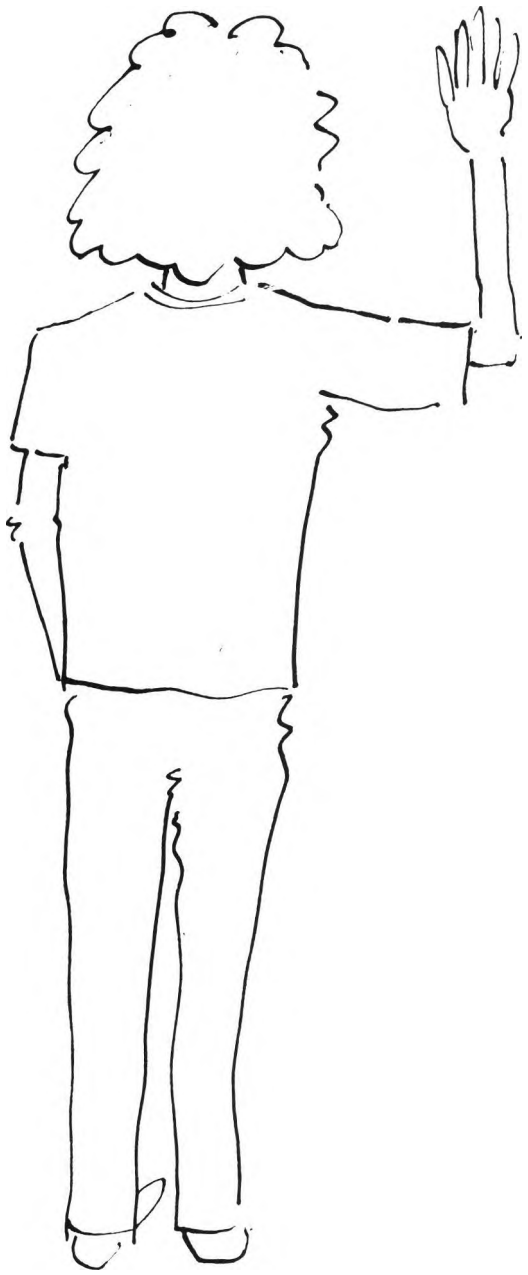


what is cystic fibrosis ?



what parts of the body need special care?

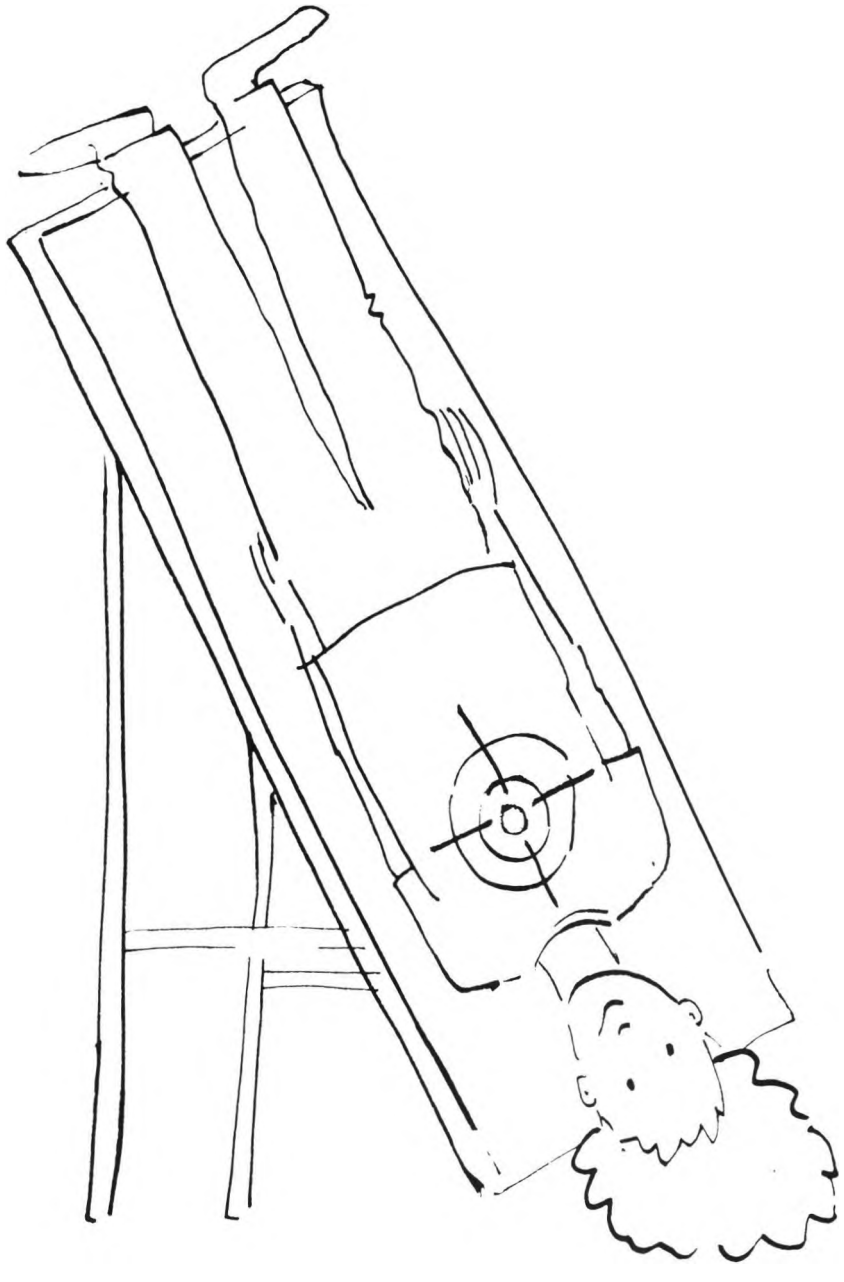
what are the things that Sophie has to do each day to keep well ?



will they make Sophie's
cystic fibrosis go away ?

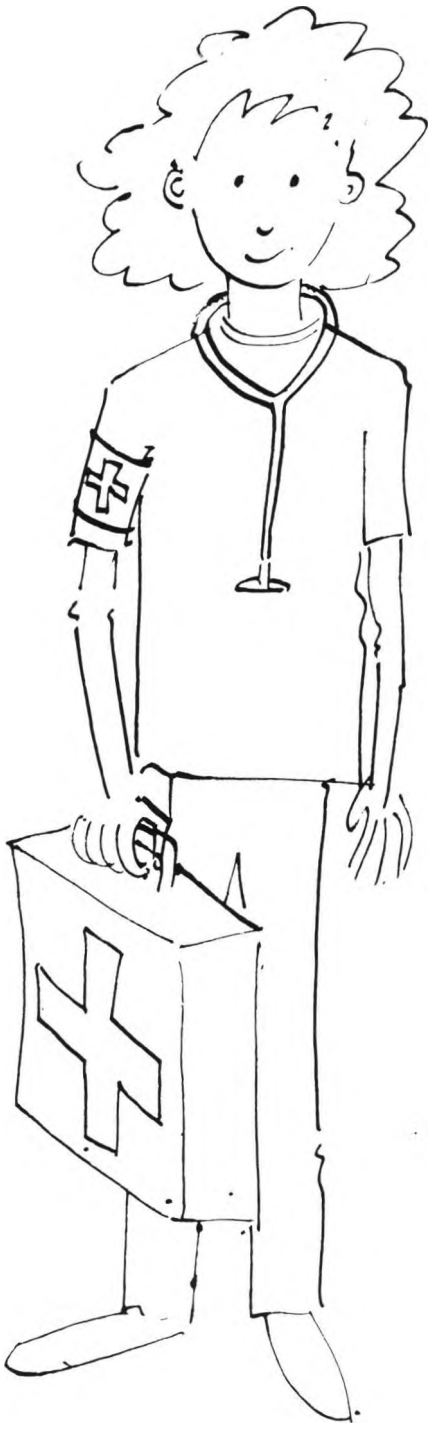
why is it important for Sophie to eat lots of food?

why is it important for Sophie to take tablets before eating her food ?

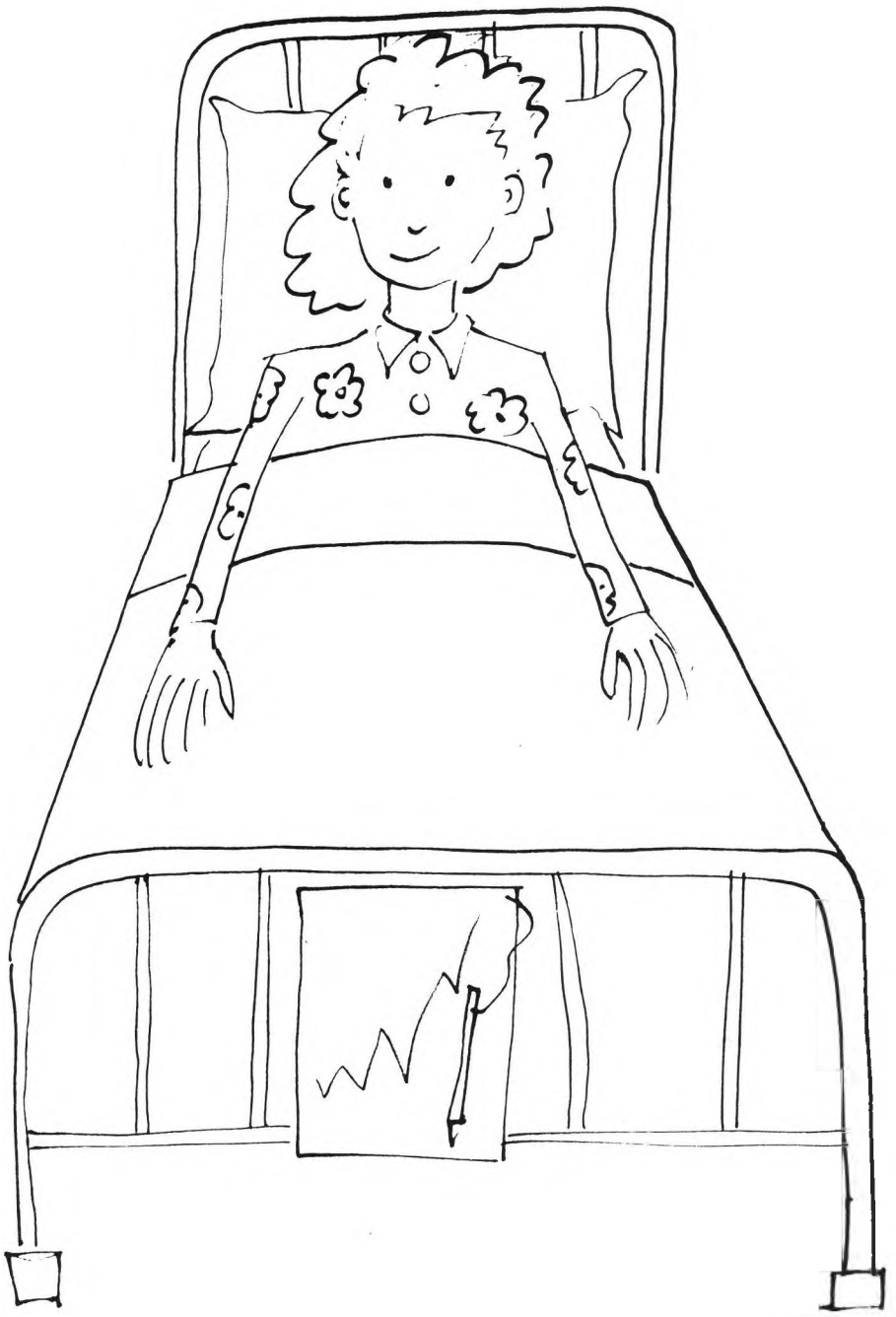


why is it important for Sophie to do her physiotherapy ?

why is it important for Sophie to have her medicine ?

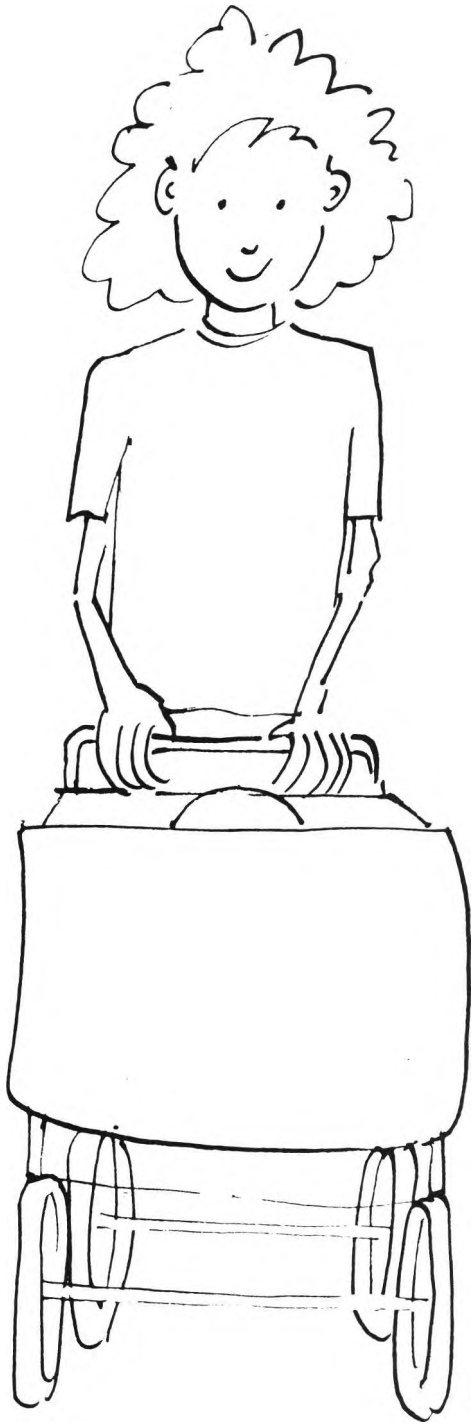


why is it important for Sophie to go to the clinic?

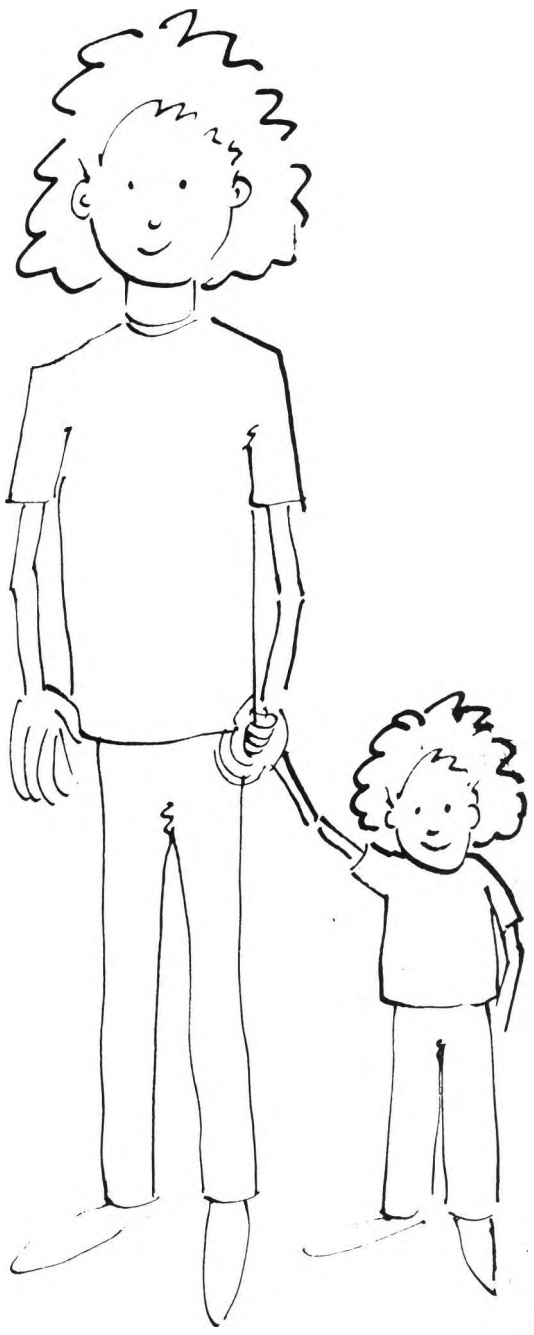


why might Sophie have to stay in hospital sometimes ?

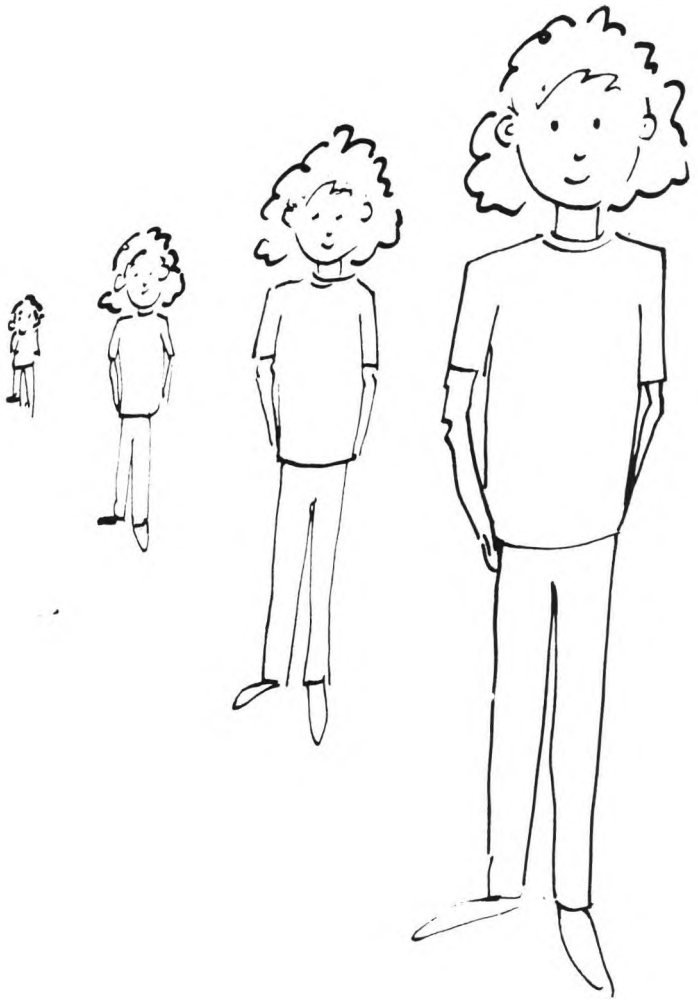
how did Sophie get cystic fibrosis ?



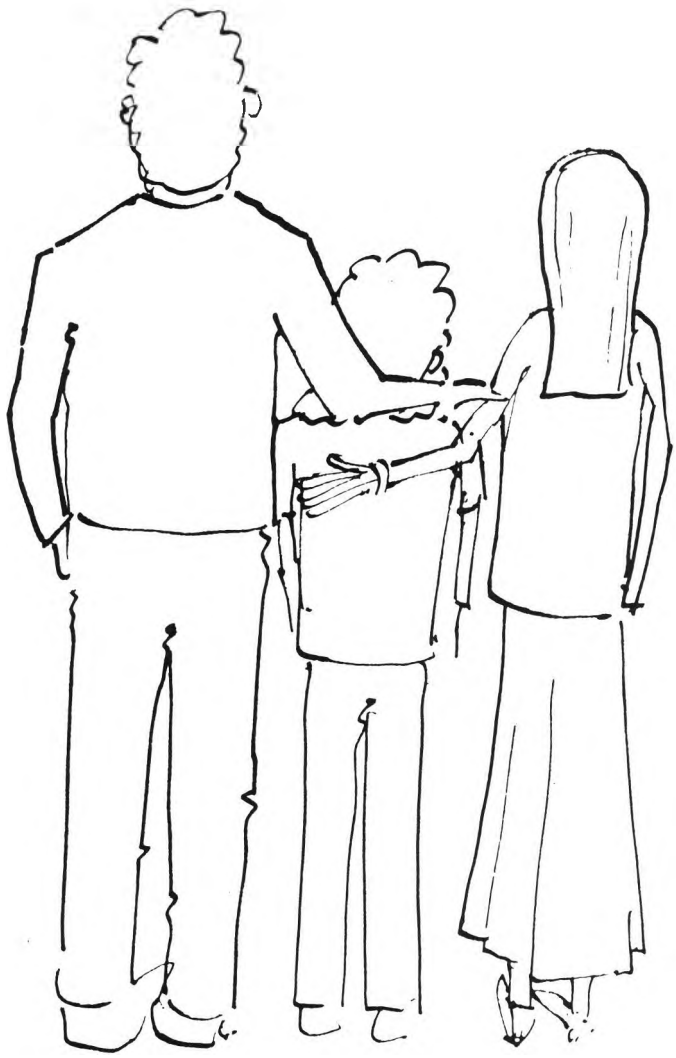
if Sophie's parents had another baby, would the baby have cystic fibrosis?



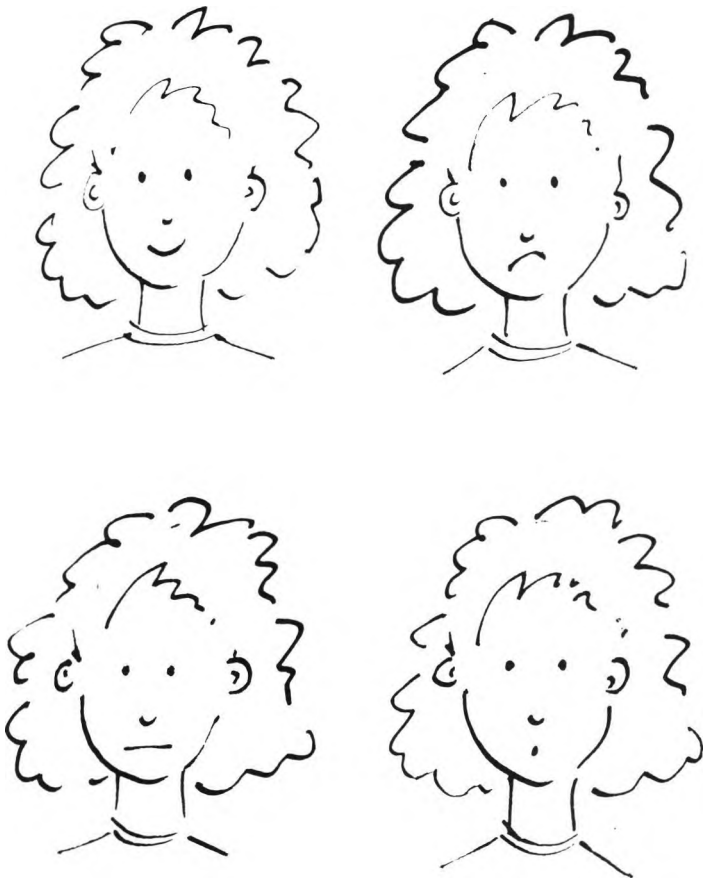
will Sophie have children
of her own ?



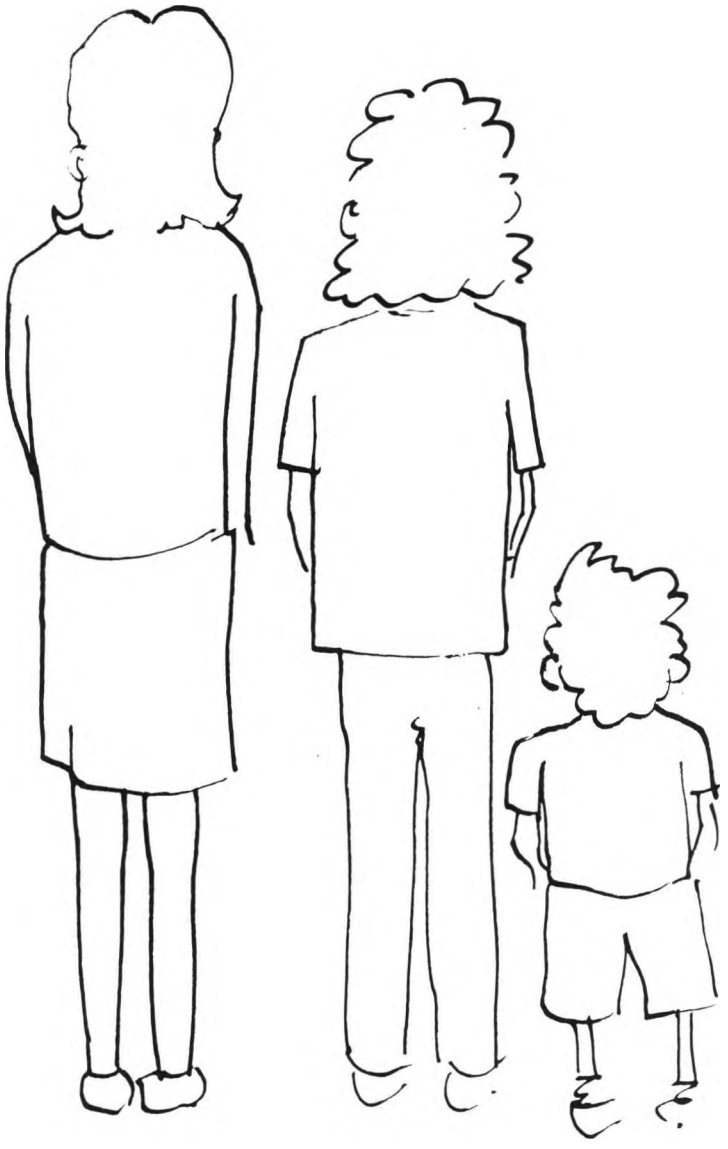
what will happen to
Sophie as she grows up?



how does cystic fibrosis make Sophie's parents feel?

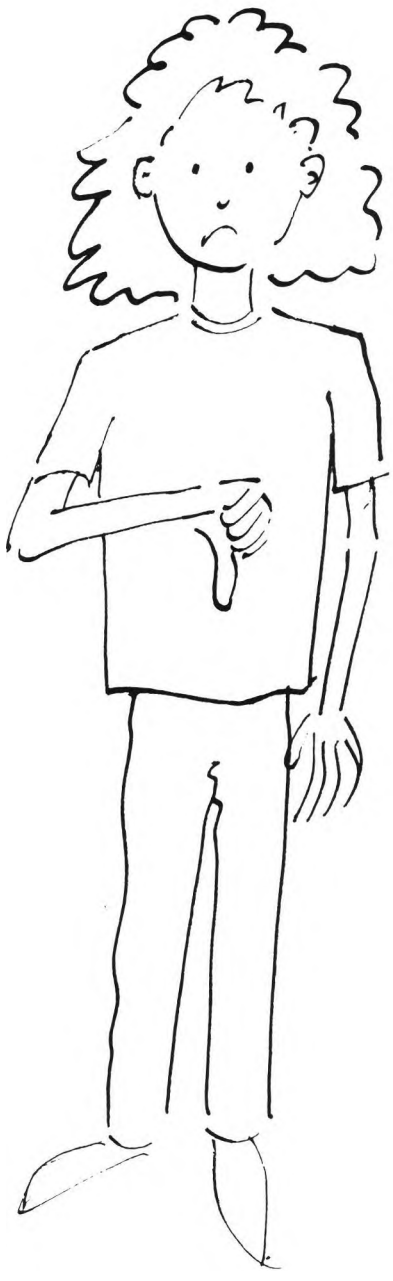


how does cystic fibrosis make Sophie feel ?

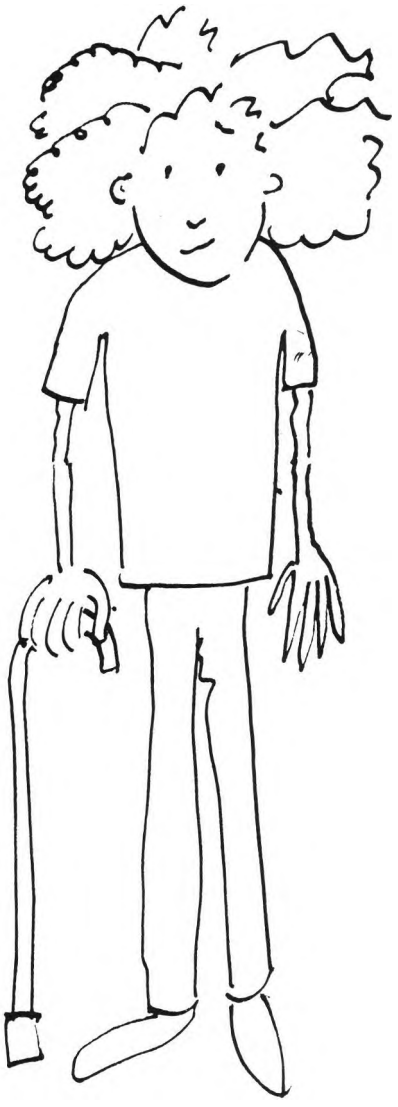


how does cystic fibrosis make Sophie's brothers and sisters feel ?

are there any things
Sophie may like about
cystic fibrosis ?

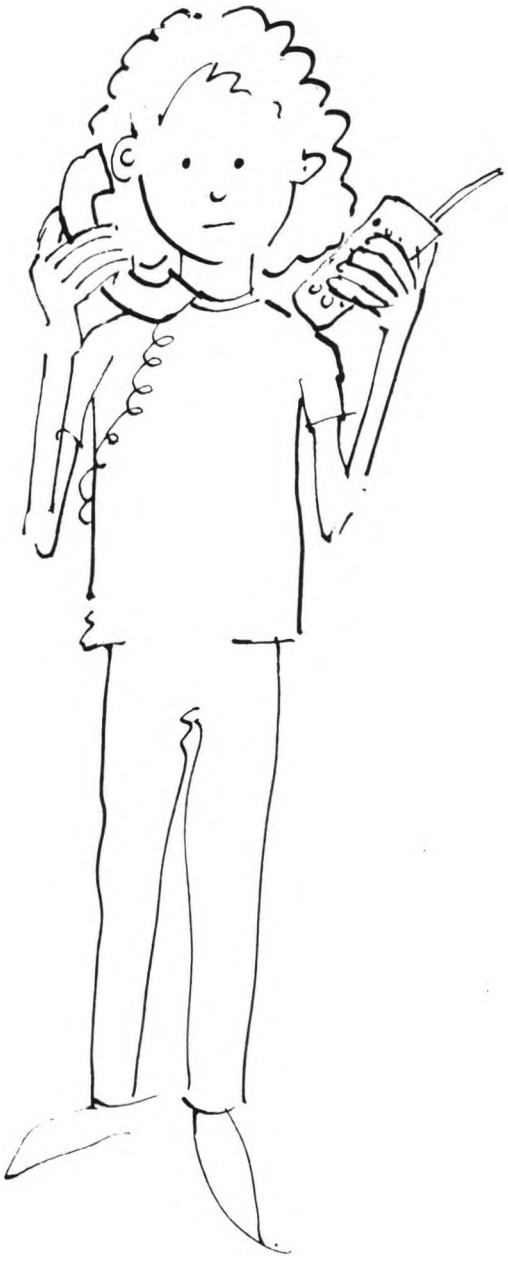


are there any things
Sophie may not like
about cystic fibrosis ?



how old do you think Sophie will grow up to be?

are there any other important things about cystic fibrosis that Sophie really needs to know ?



if Sophie wanted to talk more about her cystic fibrosis, who could she talk to ?